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SUBJECT TO COMPLETION, DATED MAY 14, 2018

PRELIMINARY PROSPECTUS

5,360,000 Shares



SCHOLAR ROCK
Scholar Rock Holding Corporation
Common Stock

We are offering 5,360,000 shares of our common stock. This is our initial public offering and no public market currently exists for our common stock. We anticipate that the initial public offering price will be between \$13.00 and \$15.00 per share. We have applied to list our common stock on The Nasdaq Global Market under the symbol "SRRK."

Investing in our common stock involves risks. See "Risk Factors" beginning on page 13 of this prospectus.

We are an "emerging growth company" as defined under U.S. federal securities laws and will be subject to reduced public company reporting requirements.

Neither the Securities and Exchange Commission nor any state securities commission has approved or disapproved of these securities, or passed upon the accuracy or adequacy of this prospectus. Any representation to the contrary is a criminal offense.

	<u>Per Share</u>	<u>Total</u>
Initial public offering price	\$	\$
Underwriting discounts and commissions ⁽¹⁾	\$	\$
Proceeds, before expenses, to us	\$	\$

⁽¹⁾ See "Underwriting" beginning on page 173 of this prospectus for additional information regarding total underwriter compensation.

Delivery of the shares of common stock is expected to be made on or about _____, 2018. We have granted the underwriters an option for a period of 30 days from the date of this prospectus to purchase up to 804,000 additional shares of our common stock. If the underwriters exercise the option in full, the total underwriting discounts and commissions payable by us will be \$ _____ million, and the total proceeds to us, before expenses, will be \$ _____ million.

Joint Book-Running Managers

Jefferies

Cowen

BMO Capital Markets

Co-Manager

Wedbush PacGrow

Prospectus date _____, 2018

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We and the underwriters have not authorized anyone to provide any information or to make any representations other than those contained in this prospectus or in any free writing prospectuses we have prepared. We and the underwriters take no responsibility for, and can provide no assurance as to the reliability of, any other information that others may provide you. We are offering to sell, and seeking offers to buy, shares of common stock only in jurisdictions where offers and sales are permitted. The information contained in this prospectus is accurate only as of the date of this prospectus, regardless of the time of delivery of this prospectus or of any sale of the common stock.

PROSPECTUS SUMMARY

This summary highlights information contained elsewhere in this prospectus. Before investing in our common stock, you should carefully read this entire prospectus, including our consolidated financial statements and the related notes included elsewhere in this prospectus and the section titled "Risk Factors." As used in this prospectus, unless the context otherwise requires, references to the "company," "we," "us" and "our" refer to Scholar Rock Holding Corporation together with its consolidated subsidiaries.

Overview

We are a biopharmaceutical company focused on the discovery and development of innovative medicines for the treatment of serious diseases in which signaling by protein growth factors plays a fundamental role. Our newly elucidated understanding of the molecular mechanisms of growth factor activation enabled us to develop a proprietary platform for the discovery and development of monoclonal antibodies that locally and selectively target these signaling proteins at the cellular level. We believe this approach, acting in the disease microenvironment, avoids the historical challenges associated with inhibiting growth factors for therapeutic effect. We believe our focus on biologically validated growth factors may facilitate a more efficient development path. We are advancing our lead product candidate, SRK-015, a selective first-in-class inhibitor of the activation of the growth factor myostatin in skeletal muscle, into clinical development for the treatment of spinal muscular atrophy, or SMA. We plan to initiate our Phase 1 clinical trial in May 2018. Utilizing our proprietary platform, we are also creating a pipeline of novel product candidates with the potential to transform the lives of patients suffering from a wide range of serious diseases, including other neuromuscular disorders, cancer, fibrosis and anemia.

Our Approach and Proprietary Platform

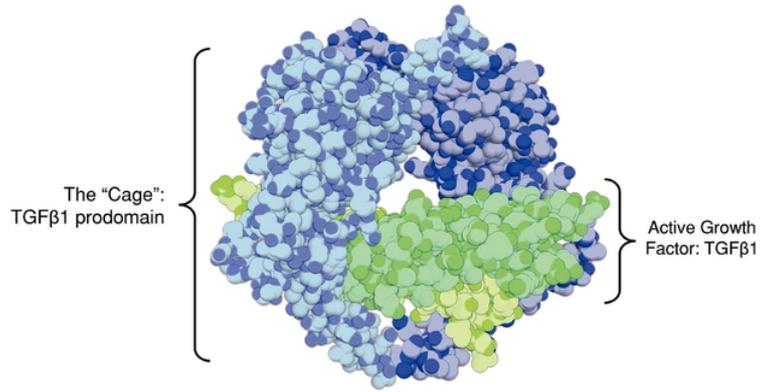
Our proprietary platform is designed to discover and develop monoclonal antibodies that have a high degree of specificity to achieve selective modulation of growth factor signaling. Growth factors are naturally occurring proteins that typically act as signaling molecules between cells and play a fundamental role in regulating a variety of normal cellular processes, including cell growth and differentiation. Current therapeutic approaches to treating diseases in which growth factors play a fundamental role involve directly targeting an active growth factor or its receptor systemically throughout the body and have suffered from a variety of shortcomings:

- § multiple growth factors often signal through the same or overlapping sets of related receptors, making it difficult to specifically modulate one pathway over another;
- § members of the same growth factor superfamily share considerable structural similarities, making it difficult to achieve specific inhibition of the targeted growth factor; this can result in broad systemic inhibition that can cause undesirable, and in many cases toxic, side effects; and
- § systemic and nonselective inhibition of a growth factor can block the growth factor's role in the disease process, but can also simultaneously interfere with its normal physiological roles.

Our innovative approach is rooted in our structural biology insights into the mechanism by which certain growth factors are activated in close proximity to the cell surface, which we refer to as "supracellular activation." We integrate these insights with sophisticated protein expression, assay development and monoclonal antibody discovery capabilities. We believe our proprietary platform can address the challenges of current therapeutic approaches to treating diseases in which growth factors play a fundamental role by:

- § targeting the natural activation mechanism to prevent activation of the growth factor rather than attempting to inhibit the growth factor after activation;
- § achieving heightened specificity for the targeted growth factor while minimizing interactions with structurally similar and related growth factors, thereby reducing the risk of unintended systemic adverse events; and
- § targeting the disease microenvironment, where we believe we can interfere with the disease process while minimizing the effects on the normal physiological processes mediated by the same growth factors.

Unlike many other proteins that are produced and secreted by cells in a mature, or active, form, many growth factors are expressed by cells in a precursor, or latent, form. For example, TGF β 1 is produced by cells as a single protein which is then enzymatically processed by the cells into two distinct and physically separated domains — the mature growth factor and the remaining portion of the original protein, referred to as the prodomain — which remain associated as part of a complex. This secreted complex is latent, or inactive, and must first be activated to carry out its normal function in a highly localized tissue or disease microenvironment. In a seminal peer-reviewed publication in 2011, Timothy A. Springer, Ph.D., one of our co-founders, elucidated a new understanding of the mechanism of supracellular activation as it applies to members of the TGF β superfamily, by solving a high resolution x-ray crystal structure of this latent form of TGF β 1, as illustrated in the graphic below.



Structural representation of the latent form of TGF β 1 wherein the prodomain wraps around the active growth factor.

This research explained at a molecular level why the secreted form of TGF β 1 is inactive. The prodomain, though physically separated from the mature growth factor domain, forms a "cage" around the active form of TGF β 1, blocking the growth factor from signaling through its receptor. Only when the cage is "unlocked" by a supracellular activation event can the growth factor be released and mediate its effects in the local microenvironment. Dr. Springer further hypothesized that this phenomenon likely holds true for most members of the TGF β superfamily, though the exact nature of the activation event, such as integrin binding or enzymatic cleavage, may differ among members of the superfamily. Importantly, while many growth factors are structurally very similar, their cages are structurally diverse, and this provides the basis for our approach to improved selectivity.

Our Pipeline Programs

Using our innovative approach and proprietary platform, we are creating a pipeline of novel product candidates that selectively inhibit the supracellular activation of growth factors believed to be important drivers in a variety of diseases, including neuromuscular disorders, cancer, fibrosis and anemia. Our proprietary platform includes (i) our know-how expressing and purifying latent protein growth factor complexes in quantity and quality sufficient to enable antibody discovery; (ii) strategies to identify rare antibodies that selectively bind targeted latent protein growth factor complexes; and (iii) assays developed by us in which to test the highly selective antibodies' ability to modulate the activation of specific latent

growth factors. In addition to such know-how, our proprietary platform is covered by two patent families, with issued patents projected to expire in 2034. We have worldwide rights to our proprietary platform and all of our product candidates, with the exception of certain early-stage antibodies that specifically inhibit the activation of TGFb1 in the context of regulatory T cells, which we licensed to Janssen Biotech, Inc., or Janssen, a subsidiary of Johnson & Johnson.

Our Lead Product Candidate and Additional Programs

SRK-015

We are advancing our lead antibody product candidate, SRK-015, a first-in-class inhibitor of the activation of myostatin, into clinical development for the treatment of SMA. Myostatin is a negative regulator of muscle mass expressed primarily in skeletal muscle tissue and a member of the transforming growth factor beta, or TGFb, superfamily, a group of more than 30 related growth factors that mediate diverse biological processes. Vertebrate animals that lack the myostatin gene display increased muscle mass and strength relative to their normal counterparts, but are otherwise healthy. We believe inhibition of the activation of myostatin may promote a clinically meaningful increase in muscle mass and strength. As a result, we have focused our initial development efforts for SRK-015 on the treatment of SMA. SMA is a rare, and often fatal, genetic disorder that typically manifests in young children, characterized by atrophy of the voluntary muscles of the limbs and trunk and dramatically reduced normal neuromuscular function. An estimated 30,000 to 35,000 patients suffer from SMA in the United States and Europe. In preclinical studies, we observed that SRK-015 promoted increased muscle mass and strength, while selectively avoiding interaction with other closely related growth factors that play distinctly different physiological roles. We believe that SRK-015 has the potential to be the first muscle-directed therapy to reverse or prevent muscle atrophy in SMA patients and could be used both as a monotherapy or in conjunction with the current standard of care. In March 2018, we filed an Investigational New Drug application, or IND, with the U.S. Food and Drug Administration, or FDA, for SRK-015, and in April 2018, the FDA notified us that our Phase 1 first-in-human clinical trial of SRK-015 may proceed. We plan to initiate our Phase 1 clinical trial in May 2018. The FDA has granted orphan drug designation for SRK-015 for the treatment of SMA.

TGFb1

Our second antibody program is focused on the discovery and development of highly specific inhibitors of the activation of TGFb1. TGFb1 is also a member of the TGFb superfamily, and increased signaling by TGFb1 is a key driver of a number of disease-relevant processes, including tissue and organ fibrosis, immune system evasion by cancer cells, and bone marrow fibrosis associated with hematological disorders. Historically, selectively targeting TGFb1 has been challenging due to off-target inhibition of other, closely related growth factors, TGFb2 and TGFb3. Pan-TGFb inhibition has been associated with a range of toxicities, most notably cardiac toxicity. In preclinical studies of our antibodies, we have observed inhibition of TGFb1 activation *in vitro* and immunomodulatory and antifibrotic activity in multiple disease models *in vivo*. In addition, we have completed a 28-day pilot toxicology study of our leading antibody and, to date, we have not observed any drug-related toxicity. In the same study, we tested pan-TGFb inhibitors and observed the toxicities, including cardiac toxicity that have been observed by others. We are actively evaluating a limited number of our selective inhibitors of the activation of TGFb1 in multiple disease models, and we intend to nominate a clinical candidate to initially pursue in one or more of our currently targeted indications of oncology, immuno-oncology and fibrosis, by the first half of 2019.

BMP6

Our third antibody program targets the signaling of bone morphogenetic protein 6, or BMP6, another member of the TGFb superfamily, which is involved in a diverse set of biological processes in various parts of the body. For example, in the liver, BMP6 signaling is a key controller of the body's ability to regulate iron levels. Given BMP6's important role in iron metabolism, we believe that targeting BMP6 signaling in a liver-selective fashion presents the potential to address both iron-restricted anemias and iron overload conditions. In preclinical studies of our antibodies targeting BMP6 signaling in the liver, we have observed

increased iron levels in the bloodstream of healthy animals and we are now evaluating a limited number of these antibodies in disease models of iron restriction.

Our Strategy

Using our proprietary platform to unlock the therapeutic potential of targeting growth factor signaling in the disease microenvironment, our goal is to deliver novel therapies to underserved patients suffering from a wide range of serious diseases, including neuromuscular disorders, cancer, fibrosis and anemia. To achieve this goal we plan to:

- § rapidly advance our lead product candidate, SRK-015, through clinical proof-of-concept;
- § advance our TGFb1 program into clinical development;
- § explore additional indications for our existing and emerging product candidates;
- § continue to leverage our proprietary platform to expand our pipeline beyond current lead programs;
- § selectively seek strategic collaborations to maximize the value of our proprietary platform and pipeline; and
- § attract and retain people that share our commitment to scientific excellence and a focus on patients.

We have worldwide rights to our proprietary platform and all of our product candidates and antibodies, with the exception of certain early-stage antibodies that specifically inhibit the activation of TGFb1 in the context of regulatory T cells, which we licensed to Janssen.

We have assembled an experienced management team, board of directors, scientific founders and advisory board who bring extensive industry experience to our company. The members of our team have deep experience in discovering, developing and commercializing therapeutics with a particular focus on rare diseases, having worked at companies such as Alnylam Pharmaceuticals, Inc., Avila Therapeutics, Inc., Biogen, Inc. and Dyax Corp. We were founded by internationally respected scientists, Drs. Timothy A. Springer and Leonard I. Zon of Harvard Medical School and Boston Children's Hospital.

Since our inception in 2012, we have raised over \$100 million through convertible preferred stock financings. Our investors include ARCH Venture Partners, Cormorant Asset Management, EcoR1 Capital, Fidelity Management and Research Company, Invus, The Kraft Group, Polaris Partners, Redmile Group and Timothy A. Springer, Ph.D.

Risks Affecting Our Business

Our business is subject to a number of risks, including risks that may prevent us from achieving our business objectives or may adversely affect our business, financial condition, results of operations, cash flows and prospects that you should consider before making a decision to invest in our common stock. These risks are discussed more fully in the section titled "Risk Factors" beginning on page 13 of this prospectus, and include the following:

- § We have limited operating history, have incurred net losses in every year since our inception and anticipate that we will continue to incur net losses in the future.
- § We will require additional capital to fund our operations and if we fail to obtain necessary capital, we will not be able to complete the development and commercialization of SRK-015 and any future product candidates.
- § Our business is highly dependent on the success of our lead product candidate, SRK-015, as well as any future clinical candidates that are generated from our other preclinical programs. All of our product candidates will require significant additional preclinical and clinical development before we may be able to seek regulatory approval for and launch a product commercially.
- § Our approach to the discovery and development of innovative medicines for the treatment of serious diseases in which signaling by protein growth factors plays a fundamental role is based on our proprietary platform, which is unproven and may not result in marketable products.

- § Clinical development involves a lengthy and expensive process, with an uncertain outcome. We may incur additional costs or experience delays in completing, or ultimately be unable to complete, the development and commercialization of any product candidates.
- § We face significant competition from other biotechnology and pharmaceutical companies, and our operating results will suffer if we fail to compete effectively.
- § If we lose key management personnel, or if we fail to recruit additional highly skilled personnel, our ability to identify and develop new or next generation product candidates will be impaired, could result in loss of markets or market share and could make us less competitive.
- § Our success depends in part on our ability to protect our intellectual property. It is difficult and costly to protect our proprietary rights and technology, and we may not be able to ensure their protection.
- § We rely on third parties to conduct certain aspects of our preclinical studies and will rely on third parties to conduct our clinical trials. If these third parties do not successfully carry out their contractual duties or meet expected deadlines or comply with regulatory requirements, we may not be able to obtain regulatory approval of or commercialize any potential product candidates.
- § Our future collaborations will be important to our business. If we are unable to enter into new collaborations, or if these collaborations are not successful, our business could be adversely affected.

Corporate History

We were incorporated in 2017 under the laws of the state of Delaware as the holding company for Scholar Rock, Inc., a wholly owned subsidiary of Scholar Rock, LLC. Pursuant to the terms of a corporate reorganization that was completed on December 22, 2017, all of the equity interests in Scholar Rock, LLC were exchanged for the same number and class of newly issued securities of Scholar Rock Holding Corporation and, as a result, Scholar Rock, LLC became a wholly owned subsidiary of Scholar Rock Holding Corporation. See the section titled "Restructuring" for additional information. Our principal executive offices are located at 620 Memorial Drive, 2nd Floor, Cambridge, MA 02139, and our phone number is (857) 259-3860. Our website address is <http://www.scholarrock.com>. The information contained in or accessible from our website is not incorporated into this prospectus, and you should not consider it part of this prospectus.

We own various U.S. federal trademark applications and unregistered trademarks, including our company name and our logo. All other trademarks or trade names referred to in this prospectus are the property of their respective owners. Solely for convenience, the trademarks and trade names in this prospectus are referred to without the symbols ® and ™, but such references should not be construed as any indicator that their respective owners will not assert, to the fullest extent under applicable law, their rights thereto.

Implications of Being an Emerging Growth Company

We are an "emerging growth company," as defined in the Jumpstart Our Business Startups Act, or the JOBS Act, enacted in April 2012. For so long as we remain an emerging growth company, we are permitted and intend to rely on certain exemptions from various public company reporting requirements, including not being required to have our internal control over financial reporting audited by our independent registered public accounting firm pursuant to Section 404(b) of the Sarbanes-Oxley Act of 2002, reduced disclosure obligations regarding executive compensation in our periodic reports and proxy statements, exemptions from the requirements of holding a nonbinding advisory vote on executive compensation and any golden parachute payments not previously approved. In particular, in this prospectus, we have provided only two years of audited financial statements and have not included all of the executive compensation related information that would be required if we were not an emerging growth company. Accordingly, the information contained herein may be different than the information you receive from other public companies in which you hold stock.

We will remain an emerging growth company until the earlier to occur of (1) the last day of 2023, (2) the last day of the fiscal year in which we have total annual gross revenues of at least \$1.07 billion, (3) the last day of the fiscal year in which we are deemed to be a "large accelerated filer," under the rules of the U.S. Securities and Exchange Commission, or SEC, which means the market value of our equity securities that is held by non-affiliates exceeds \$700 million as of the prior June 30th, and (4) the date on which we have issued more than \$1.0 billion in non-convertible debt during the prior three-year period.

In addition, the JOBS Act provides that an emerging growth company can take advantage of an extended transition period for complying with new or revised accounting standards. This provision allows an emerging growth company to delay the adoption of some accounting standards until those standards would otherwise apply to private companies. We have irrevocably elected not to avail ourselves of delayed adoption of new or revised accounting standards and, therefore, we will be subject to the same requirements to adopt new or revised accounting standards as other public companies that are not emerging growth companies.

THE OFFERING

Common stock offered by us	5,360,000 shares
Common stock to be outstanding immediately after this offering	24,440,536 shares
Option to purchase additional shares offered by us	804,000 shares
Use of proceeds	We estimate that we will receive net proceeds from the sale of shares of our common stock in this offering of approximately \$67.5 million, or \$78.0 million if the underwriters exercise their option to purchase additional shares in full, assuming an initial public offering price of \$14.00 per share, the midpoint of the price range set forth on the cover page of this prospectus, and after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us. We intend to use the net proceeds from this offering, together with our existing cash, cash equivalents and marketable securities, to fund research and development activities for SRK-015 through our planned Phase 2 clinical proof of concept trial, and to fund TGFb1, BMP6 and other preclinical research and development activities. We intend to use the remaining proceeds from this offering for working capital and other general corporate purposes. For a more complete description of our intended use of the proceeds from this offering, see "Use of Proceeds."
Risk factors	You should carefully read the "Risk Factors" section of this prospectus for a discussion of factors that you should consider before deciding to invest in our common stock.
Proposed Nasdaq Global Market symbol	"SRRK"

The number of shares of our common stock to be outstanding after this offering is based on 19,080,536 shares of our common stock (which includes 1,114,089 shares of restricted common stock) outstanding as of March 31, 2018, giving effect to the conversion of all outstanding shares of our convertible preferred stock into an aggregate of 15,109,950 shares of our common stock upon the completion of this offering, and excludes:

- § 7,614 shares of common stock issuable upon the exercise of a warrant outstanding as of March 31, 2018, at an exercise price of \$3.94 per share;
- § 660,319 shares of common stock issuable upon the exercise of stock options outstanding as of March 31, 2018 under our 2017 Stock Option and Incentive Plan at an exercise price of \$5.77 per share;
- § 355,539 shares of common stock issuable upon the exercise of stock options granted subsequent to March 31, 2018 under our 2017 Stock Option and Incentive Plan at an exercise price of \$7.17 per share;
- § 3,139,274 shares of our common stock that will become available for future issuance under our 2018 Stock Option and Incentive Plan upon the effectiveness of the registration statement of which

this prospectus forms a part, from which we have granted options to purchase 309,925 shares of common stock, subject to and effective upon the closing of this offering, at an exercise price equal to the public offering price per share in this offering; and

- § 235,743 shares of our common stock that will become available for future issuance under our 2018 Employee Stock Purchase Plan upon the effectiveness of the registration statement of which this prospectus forms a part.

Unless otherwise indicated, all information in this prospectus reflects or assumes the following:

- § the filing of our amended and restated certificate of incorporation and the effectiveness of our amended and restated by-laws upon the completion of this offering;
- § the conversion of all outstanding shares of convertible preferred stock into an aggregate of 15,109,950 shares of common stock upon the completion of this offering;
- § the conversion of the outstanding warrant to purchase 21,739 shares of convertible preferred stock into a warrant to purchase 7,614 shares of common stock;
- § no exercise of outstanding options or warrants after March 31, 2018;
- § a 2.8548-to-one reverse split of our common stock effected on May 11, 2018; and
- § no exercise by the underwriters of their option to purchase up to 804,000 additional shares of common stock in this offering.

SUMMARY CONSOLIDATED FINANCIAL DATA

The summary consolidated financial data set forth below should be read together with the consolidated financial statements and the related notes to those statements, as well as the section entitled "Management's Discussion and Analysis of Financial Condition and Results of Operations." We have derived the summary consolidated statement of operations data for the years ended December 31, 2016 and 2017 from our audited consolidated financial statements included elsewhere in this prospectus. We have derived the summary consolidated statement of operations data for the three months ended March 31, 2017 and 2018 and the summary consolidated balance sheet data as of March 31, 2018 from our unaudited consolidated financial statements included elsewhere in this prospectus. In the opinion of management, the unaudited consolidated financial statements have been prepared on a basis consistent with our audited consolidated financial statements and contain all adjustments, consisting of only normal recurring adjustments, that management considers necessary for the fair presentation of the financial information set forth in those statements. Our historical results are not necessarily indicative of results that may be expected in the future, and the results for the three months ended March 31, 2018 are not necessarily indicative of results to be expected for the full year ending December 31, 2018 or any other period.

	Year Ended December 31,		Three Months Ended March 31,	
	2016	2017	2017	2018
	(in thousands, except unit, share, per unit, and per share data)			
Consolidated Statement of Operations Data:				
Collaboration revenue	\$ 379	\$ —	\$ —	\$ —
Operating expenses:				
Research and development	12,477	19,944	3,568	6,701
General and administrative	4,112	5,085	1,173	2,315
Total operating expenses	<u>16,589</u>	<u>25,029</u>	<u>4,741</u>	<u>9,016</u>
Loss from operations	(16,210)	(25,029)	(4,741)	(9,016)
Other income (expense):				
Interest income (expense), net	(19)	44	13	144
Other income (expense), net	22	(10)	(2)	(20)
Total other income	<u>3</u>	<u>34</u>	<u>11</u>	<u>124</u>
Net loss	<u>\$ (16,207)</u>	<u>\$ (24,995)</u>	<u>\$ (4,730)</u>	<u>\$ (8,892)</u>
Net loss per unit, basic and diluted ⁽¹⁾	<u>\$ (10.11)</u>		<u>\$ (2.95)</u>	
Net loss per share, basic and diluted ⁽¹⁾		<u>\$ (15.30)</u>		<u>\$ (3.18)</u>
Weighted average common units outstanding, basic and diluted ⁽¹⁾	<u>1,603,088</u>		<u>1,603,088</u>	
Weighted average common shares outstanding, basic and diluted ⁽¹⁾		<u>1,634,100</u>		<u>2,795,497</u>
Pro forma net loss per share, basic and diluted (unaudited) ⁽¹⁾		<u>\$ (2.06)</u>		<u>\$ (0.50)</u>
Pro forma weighted average common shares outstanding, basic and diluted (unaudited) ⁽¹⁾		<u>12,108,377</u>		<u>17,905,447</u>

⁽¹⁾ See Note 17 to our consolidated financial statements included elsewhere in this prospectus for further details on the calculation of basic and diluted net loss per unit and share and unaudited pro forma basic and diluted net loss per share.

	As of March 31, 2018		
	Actual	Pro Forma ⁽¹⁾ (in thousands)	Pro Forma As Adjusted ⁽²⁾
Consolidated Balance Sheet Data:			
Cash, cash equivalents and marketable securities	\$ 47,763	\$ 47,763	\$ 115,250
Working capital ⁽³⁾	45,043	45,043	112,530
Total assets	54,159	54,159	121,646
Convertible preferred stock	109,232	—	—
Accumulated deficit	(66,417)	(66,459)	(66,459)
Total stockholders' equity (deficit)	(61,931)	47,358	114,845

⁽¹⁾ Pro forma amounts give effect to (1) the automatic conversion of all of our outstanding shares of convertible preferred stock into an aggregate of 15,109,950 shares of common stock upon completion of this offering, (2) the automatic conversion of the outstanding warrant to purchase 21,739 shares of convertible preferred stock into a warrant to purchase 7,614 shares of common stock, and the resulting reclassification of the warrant liability to additional paid-in capital, and (3) the vesting of a performance-based stock option award to purchase up to 8,757 shares of our common stock with vesting conditions contingent upon completion of this offering, and the resulting recognition of equity-based compensation expense.

⁽²⁾ Pro forma as adjusted amounts reflect pro forma adjustments described in footnote (1) as well as the sale of shares of our common stock in this offering at the assumed initial public offering price of \$14.00 per share, the midpoint of the price range set forth on the cover page of this prospectus, after deducting the estimated underwriting discounts and commissions and estimated offering expenses payable by us. A \$1.00 increase (decrease) in the assumed initial public offering price of \$14.00 per share, the midpoint of the price range set forth on the cover of this prospectus, would increase (decrease) the pro forma as adjusted amount of each of cash, cash equivalents and marketable securities, working capital, total assets and total stockholders' equity by \$5.0 million, assuming that the number of shares offered by us, as set forth on the cover of this prospectus, remains the same and after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us. Similarly, each increase (decrease) of 1,000,000 in the number of shares offered by us in this offering would increase (decrease) the pro forma as adjusted amount of each of cash, cash equivalents and marketable securities, working capital, total assets and total stockholders' equity by \$13.0 million, assuming the assumed initial public offering price remains the same and after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us.

⁽³⁾ We define working capital as current assets less current liabilities. See our consolidated financial statements and related notes appearing at the end of this prospectus for further details regarding our current assets and current liabilities.

REORGANIZATION

Reorganization and Convertible Preferred Stock

On December 22, 2017, we completed a series of transactions pursuant to which Scholar Rock Merger Sub, LLC, a wholly owned subsidiary of Scholar Rock Holding Corporation, was merged with and into Scholar Rock, LLC, or the Reorganization. In connection with the Reorganization:

- § Holders of Scholar Rock, LLC Series B convertible preferred units received one share of Scholar Rock Holding Corporation Series B convertible preferred stock for each outstanding Series B convertible preferred unit held immediately prior to the Reorganization, with an aggregate of 13,526,994 shares of Scholar Rock Holding Corporation Series B convertible preferred stock issued in the Reorganization;
- § Holders of Scholar Rock, LLC Series A-4 convertible preferred units received one share of Scholar Rock Holding Corporation Series A-4 convertible preferred stock for each outstanding Series A-4 convertible preferred unit held immediately prior to the Reorganization, with an aggregate of 3,906,738 shares of Scholar Rock Holding Corporation Series A-4 convertible preferred stock issued in the Reorganization;
- § Holders of Scholar Rock, LLC Series A-3 convertible preferred units received one share of Scholar Rock Holding Corporation Series A-3 convertible preferred stock for each outstanding Series A-3 convertible preferred unit held immediately prior to the Reorganization, with an aggregate of 5,579,709 shares of Scholar Rock Holding Corporation Series A-3 convertible preferred stock issued in the Reorganization;
- § Holders of Scholar Rock, LLC Series A-2 convertible preferred units received one share of Scholar Rock Holding Corporation Series A-2 convertible preferred stock for each outstanding Series A-2 convertible preferred unit held immediately prior to the Reorganization, with an aggregate of 5,066,915 shares of Scholar Rock Holding Corporation Series A-2 convertible preferred stock issued in the Reorganization;
- § Holders of Scholar Rock, LLC Series A-1 convertible preferred units received one share of Scholar Rock Holding Corporation Series A-1 convertible preferred stock for each outstanding Series A-1 convertible preferred unit held immediately prior to the Reorganization, with an aggregate of 2,000,000 shares of Scholar Rock Holding Corporation Series A-1 convertible preferred stock issued in the Reorganization;
- § Holders of Scholar Rock, LLC common units received one share of Scholar Rock Holding Corporation common stock for each outstanding common unit held immediately prior to the Reorganization, which were subject to a subsequent 2.8548-to-one reverse split with an aggregate of 1,603,088 shares of common stock issued;
- § Holders of Scholar Rock, LLC vested and unvested incentive units, irrespective of any strike price or voting rights on any such outstanding incentive units, exchanged one incentive unit for one share of common stock or restricted common stock, respectively, which were subject to a subsequent 2.8548-to-one reverse split. The restricted common stock was issued with the same vesting terms as the unvested incentive units held immediately prior to the Reorganization. An aggregate of 2,367,498 shares of common stock and restricted common stock were issued to the prior holders of incentive units; and
- § The outstanding warrant to purchase 21,739 Series A-3 convertible preferred units at an exercise price of \$1.38 per unit was converted to a warrant to purchase 21,739 shares of Series A-3 convertible preferred stock at the same exercise price per share.

We issued 13,055,555 shares of Series C convertible preferred stock on December 22, 2017.

Our Series C convertible preferred stock, Series B convertible preferred stock, Series A-4 convertible preferred stock, Series A-3 convertible preferred stock, Series A-2 convertible preferred stock and Series A-1 convertible preferred stock are designated as convertible preferred stock under our amended and

restated certificate of incorporation. All outstanding shares of our convertible preferred stock are convertible into shares of common stock at the then-effective conversion ratios. In connection with the Reorganization, by operation of law, we acquired all assets of Scholar Rock, LLC and assumed all of its liabilities and obligations. The purpose of the Reorganization was to reorganize our corporate structure so that Scholar Rock Holding Corporation would continue as a corporation and so that our existing investors would own our capital stock rather than equity interests in a limited liability company. For the convenience of the reader, except as context otherwise requires, all information included in this prospectus is presented giving effect to the Reorganization.

RISK FACTORS

Investing in our common stock involves a high degree of risk. You should carefully consider the risks described below, as well as the other information in this prospectus, including our consolidated financial statements and the related notes and "Management's Discussion and Analysis of Financial Condition and Results of Operations," before deciding whether to invest in our common stock. The occurrence of any of the events or developments described below could harm our business, financial condition, results of operations and growth prospects. In such an event, the market price of our common stock could decline and you may lose all or part of your investment. Additional risks and uncertainties not presently known to us or that we currently deem immaterial also may impair our business operations.

Risks Related to Our Business and Operations

We have limited operating history, incurred net losses in every year since our inception and anticipate that we will continue to incur net losses in the future.

We are a biopharmaceutical company with a limited operating history. We were formed in 2012 and our operations to date have been focused on organizing and staffing our company, business planning, raising capital, establishing our intellectual property portfolio and performing research and development of monoclonal antibodies that selectively inhibit activation of growth factors for therapeutic effect. Consequently, we have no meaningful operations upon which to evaluate our business and predictions about our future success or viability may not be as accurate as they could be if we had a longer operating history or a history of successfully developing and commercializing drug products. Investment in biopharmaceutical product development is highly speculative because it entails substantial upfront capital expenditures and significant risk that any potential product candidate will fail to demonstrate adequate effect or an acceptable safety profile, gain regulatory approval and become commercially viable. We have not yet demonstrated the ability to progress any product candidate through clinical trials, we have no products approved for commercial sale and we have not generated any revenue from product sales to date. We continue to incur significant research and development and other expenses related to our ongoing operations. As a result, we are not profitable and have incurred losses in each period since our inception. For the years ended December 31, 2016 and 2017, we reported a net loss of \$16.2 million and \$25.0 million, respectively. For the three months ended March 31, 2017 and 2018, we reported a net loss of \$4.7 million and \$8.9 million, respectively. As of December 31, 2017 and March 31, 2018, we had an accumulated deficit of \$57.5 million and \$66.4 million, respectively. We expect to continue to incur significant losses for the foreseeable future, and we expect these losses to increase as we continue our research and development of, and seek regulatory approvals for, our lead product candidate, SRK-015, and any future product candidates.

We anticipate that our expenses will increase substantially if, and as, we:

- § advance the development of our lead product candidate, SRK-015, into Phase 1 clinical development, and, if successful, later-stage clinical trials;
- § advance our other preclinical development programs into clinical development;
- § seek regulatory approvals for any product candidates that successfully complete clinical trials;
- § increase the amount of research and development activities to identify and develop product candidates using our proprietary platform technology;
- § hire additional clinical, quality control and scientific personnel;
- § expand our operational, financial and management systems and increase personnel, including personnel to support our clinical development, manufacturing and commercialization efforts and our operations as a public company;
- § maintain, expand and protect our intellectual property portfolio;
- § establish a sales, marketing, medical affairs and distribution infrastructure to commercialize any products for which we may obtain marketing approval and intend to commercialize on our own or jointly with third parties; and
- § invest in or in-license other technologies.

To become and remain profitable, we or any potential future collaborators must develop and eventually commercialize products with significant market potential. This will require us to be successful in a range of challenging activities, including completing preclinical studies and clinical trials, obtaining marketing approval for product candidates, manufacturing, marketing and selling products for which we may obtain marketing approval and satisfying any post-marketing requirements. We may never succeed in any or all of these activities and, even if we do, we may never generate revenue that is significant or large enough to achieve profitability. If we do achieve profitability, we may not be able to sustain or increase profitability on a quarterly or annual basis. Our failure to become and remain profitable would decrease the value of our company and could impair our ability to raise capital, maintain our research and development efforts, expand our business or continue our operations. A decline in the value of our company also could cause you to lose all or part of your investment.

Even if we succeed in commercializing one or more of our product candidates, we will continue to incur substantial research and development and other expenditures to develop and market additional product candidates. We may encounter unforeseen expenses, difficulties, complications, delays and other unknown factors that may adversely affect our business. The size of our future net losses will depend, in part, on the rate of future growth of our expenses and our ability to generate revenue. Our prior losses and expected future losses have had and will continue to have an adverse effect on our stockholders' equity and working capital.

We will require additional capital to fund our operations and if we fail to obtain necessary capital, we will not be able to complete the development and commercialization of SRK-015 and any future product candidates.

Our operations have consumed substantial amounts of cash since inception. We expect to continue to spend substantial amounts of cash to conduct further research and development and preclinical studies and clinical trials of SRK-015 and any future product candidates, to seek regulatory approvals for our product candidates and to launch and commercialize any products for which we receive regulatory approval. As of March 31, 2018, we had approximately \$47.8 million in cash, cash equivalents and marketable securities. Based on our current operating plan, we believe that the net proceeds from this offering, together with existing cash, cash equivalents and marketable securities, will be sufficient to fund our operating expenses and capital expenditure requirements into the second half of 2020. However, our future capital requirements and the period for which our existing resources will support our operations may vary significantly from what we expect, and we will in any event require additional capital in order to complete clinical development of any of our current programs. Our monthly spending levels will vary based on new and ongoing development and corporate activities. Because the length of time and activities associated with development of our product candidates is highly uncertain, we are unable to estimate the actual funds we will require for development and any approved marketing and commercialization activities. Our future funding requirements, both near and long-term, will depend on many factors, including, but not limited to:

- § the initiation, progress, timing, costs and results of preclinical studies and clinical trials for our product candidates;
- § the clinical development plans we establish for these product candidates;
- § the number and characteristics of product candidates that we develop;
- § the terms of any collaboration agreements we may choose to enter into;
- § the outcome, timing and cost of meeting regulatory requirements established by the U.S. Food and Drug Administration, or FDA, the European Medicines Agency, or EMA, and other comparable foreign regulatory authorities;
- § the cost of filing, prosecuting, defending and enforcing our patent claims and other intellectual property rights;
- § the cost of defending intellectual property disputes, including patent infringement actions brought by third parties against us or our product candidates;

- § the effect of competing technological and market developments;
- § the cost and timing of completion of commercial-scale outsourced manufacturing activities; and
- § the cost of establishing sales, marketing and distribution capabilities for any product candidates for which we may receive regulatory approval in regions where we choose to commercialize our products on our own.

We do not have any committed external source of funds or other support for our development efforts and we cannot be certain that additional funding will be available on acceptable terms, or at all. Until we can generate sufficient product or royalty revenue to finance our cash requirements, which we may never do, we expect to finance our future cash needs through a combination of public or private equity offerings, debt financings, collaborations, strategic alliances, licensing arrangements and other marketing or distribution arrangements. If we raise additional funds through public or private equity offerings, the terms of these securities may include liquidation or other preferences that adversely affect our stockholders' rights. Further, to the extent that we raise additional capital through the sale of common stock or securities convertible or exchangeable into common stock, your ownership interest will be diluted. In addition, any debt financing may subject us to fixed payment obligations and covenants limiting or restricting our ability to take specific actions, such as incurring additional debt, making capital expenditures or declaring dividends, including, for example, the covenants included in our existing loan and security agreement with Silicon Valley Bank. If we raise additional capital through marketing and distribution arrangements or other collaborations, strategic alliances or licensing arrangements with third parties, we may have to relinquish certain valuable rights to our product candidates, technologies, future revenue streams or research programs or grant licenses on terms that may not be favorable to us. We also could be required to seek collaborators for SRK-015 or any future product candidate at an earlier stage than otherwise would be desirable or relinquish our rights to product candidates or technologies that we otherwise would seek to develop or commercialize ourselves. If we are unable to raise additional capital in sufficient amounts or on terms acceptable to us, we may have to significantly delay, scale back or discontinue the development or commercialization of one or more of our product candidates or one or more of our other research and development initiatives. Any of the above events could significantly harm our business, prospects, financial condition and results of operations and cause the price of our common stock to decline.

Our business is highly dependent on the success of our lead product candidate, SRK-015, as well as any future clinical product candidates that are generated from our other preclinical programs. All of our product candidates will require significant additional preclinical and clinical development before we may be able to seek regulatory approval for and launch a product commercially.

We are very early in our development efforts. Because SRK-015 is our lead product candidate, if SRK-015 encounters safety or efficacy problems, development delays or regulatory issues or other problems, our development plans and business would be significantly harmed. SRK-015 is currently being advanced into a Phase 1 clinical development program for the treatment of spinal muscular atrophy, or SMA, which we plan to initiate in May 2018. All of our other programs are in preclinical development, and we have yet to nominate a clinical candidate from these programs.

SRK-015 and any future clinical product candidates will require additional preclinical and clinical development, regulatory review and approval in one or more jurisdictions, substantial investment, and access to sufficient commercial manufacturing capacity and significant marketing efforts before we can generate any revenue from product sales. We may not have the financial resources to continue development of, or to modify existing or enter into new collaborations for, a product candidate if we experience any issues that delay or prevent regulatory approval of, or our ability to commercialize, our product candidates, including:

- § negative or inconclusive results from our preclinical studies or clinical trials or the clinical trials of others for product candidates similar to ours, leading to a decision or requirement to conduct additional preclinical testing or clinical trials or abandon a program;

- § product-related side effects experienced by subjects in our clinical trials or by individuals using drugs or therapeutic biologics similar to our product candidates;
- § delays in submitting Investigational New Drug applications, or INDs, or comparable foreign applications or delays or failure in obtaining the necessary approvals from regulators to commence a clinical trial, or a suspension or termination of a clinical trial once commenced;
- § conditions imposed by the FDA, EMA or comparable foreign authorities regarding the scope or design of our clinical trials;
- § delays in enrolling subjects in clinical trials;
- § high drop-out rates of subjects from clinical trials;
- § inadequate supply or quality of product candidates or other materials necessary for the conduct of our clinical trials;
- § greater than anticipated clinical trial costs;
- § poor effectiveness of our product candidates during clinical trials;
- § unfavorable FDA, EMA or other regulatory agency inspection and review of a clinical trial site;
- § failure of our third-party contractors or investigators to comply with regulatory requirements or otherwise meet their contractual obligations in a timely manner, or at all;
- § delays and changes in regulatory requirements, policy and guidelines, including the imposition of additional regulatory oversight around clinical testing generally or with respect to our technology in particular; or
- § varying interpretations of data by the FDA, EMA and similar foreign regulatory agencies.

Our approach to the discovery and development of innovative medicines for the treatment of serious diseases in which signaling by protein growth factors plays a fundamental role is based on our proprietary platform, which is unproven and may not result in marketable products.

Our proprietary platform is designed to discover and develop monoclonal antibodies that have a high degree of specificity to achieve selective modulation of growth factor signaling. Our approach is rooted in our structural biology insights into the mechanism by which certain growth factors are activated in close proximity to the cell surface, which we refer to as "supracellular activation." We integrate these insights with sophisticated protein expression, assay development and monoclonal antibody discovery capabilities. However, the scientific research that forms the basis of our efforts to develop product candidates utilizing our proprietary platform is ongoing. We have not yet tested any monoclonal antibodies discovered through use of our proprietary platform in humans. Therefore, we may ultimately discover that our proprietary platform and any product candidates resulting therefrom do not possess properties required for therapeutic effectiveness. As a result, we may never succeed in developing a marketable product. If our product candidates discovered utilizing our proprietary platform prove to be ineffective, unsafe or commercially unviable, our entire proprietary platform and pipeline would have little, if any, value, which would have a material and adverse effect on our business, financial condition, results of operations and prospects.

We will need to grow the size of our organization, and we may experience difficulties in managing this growth.

As of May 7, 2018, we had 50 full-time employees. As our clinical development and commercialization plans and strategies develop, and as we transition into operating as a public company, we expect we will need additional managerial, clinical, regulatory, sales, marketing, financial, legal and other personnel. Future growth would impose significant added responsibilities on members of management, including:

- § identifying, recruiting, integrating, maintaining and motivating additional employees;

§ managing our development efforts effectively, including the clinical and FDA review process for SRK-015 and any future product candidates, while complying with our contractual obligations to contractors and other third parties; and

§ improving our operational, financial and management controls, reporting systems and procedures.

Our future financial performance and our ability to commercialize our product candidates, if approved, will depend, in part, on our ability to effectively manage any future growth, and our management may also have to divert a disproportionate amount of its attention away from day-to-day activities in order to devote a substantial amount of time to managing these growth activities.

We currently rely, and for the foreseeable future will continue to rely, in substantial part on certain independent organizations, advisors and consultants to provide certain services, including contract manufacturers and companies focused on antibody development and discovery activities. There can be no assurance that the services of independent organizations, advisors and consultants will continue to be available to us on a timely basis when needed, or that we can find qualified replacements. In addition, if we are unable to effectively manage our outsourced activities or if the quality, accuracy or quantity of the services provided is compromised for any reason, our clinical trials may be extended, delayed or terminated, and we may not be able to obtain, or may be substantially delayed in obtaining, regulatory approval of our product candidates or otherwise advance our business. There can be no assurance that we will be able to manage our existing consultants or find other competent outside contractors and consultants on economically reasonable terms, or at all.

If we are not able to effectively expand our organization by hiring new employees and expanding our groups of consultants and contractors, we may not be able to successfully implement the tasks necessary to further develop and commercialize SRK-015 or any future product candidates and, accordingly, may not achieve our research, development and commercialization goals.

If we lose key management personnel, or if we fail to recruit additional highly skilled personnel, our ability to identify and develop new or next generation product candidates will be impaired, could result in loss of markets or market share and could make us less competitive.

Our ability to compete in the highly competitive biotechnology and pharmaceutical industries depends upon our ability to attract and retain highly qualified managerial, scientific and medical personnel. We are highly dependent on our management, scientific and medical personnel, including Nagesh K. Mahanthappa, Ph.D., our Chief Executive Officer and President, Rhonda M. Chicko, C.P.A., our Chief Financial Officer, and Yung H. Chyung, M.D., our Chief Medical Officer. The loss of the services of any of our executive officers, other key employees, and other scientific and medical advisors, and our inability to find suitable replacements could result in delays in product development and harm our business.

We conduct our operations at our facility in Cambridge, Massachusetts. This region is headquarters to many other biopharmaceutical companies and many academic and research institutions. Competition for skilled personnel in our market is intense and may limit our ability to hire and retain highly qualified personnel on acceptable terms or at all.

To induce valuable employees to remain at our company, in addition to salary and cash incentives, we have provided restricted stock awards and stock options that vest over time. The value to employees of restricted stock awards and stock options that vest over time may be significantly affected by movements in our stock price that are beyond our control, and may at any time be insufficient to counteract more lucrative offers from other companies. Despite our efforts to retain valuable employees, members of our management, scientific and development teams may terminate their employment with us on short notice. Although we have employment agreements with our key employees, these employment agreements provide for at-will employment, which means that any of our employees could leave our employment at any time, with or without notice. We do not maintain "key man" insurance policies on the lives of these individuals or the

lives of any of our other employees. Our success also depends on our ability to continue to attract, retain and motivate highly skilled junior, mid-level and senior scientific and medical personnel.

Our internal computer systems, or those used by our contract research organizations, or other contractors or consultants, may fail or suffer security breaches.

Despite the implementation of security measures, our internal computer systems and those of our future contract research organizations, or CROs, and other contractors and consultants are vulnerable to damage from computer viruses and unauthorized access. While we have not experienced any such material system failure or security breach to date, if such an event were to occur and cause interruptions in our operations, it could result in a material disruption of our development programs and our business operations. For example, the loss of preclinical or clinical data could result in delays in our regulatory approval efforts and significantly increase our costs to recover or reproduce the data. Likewise, we may rely on third parties for the manufacture of our product candidates and to conduct clinical trials, and similar events relating to their computer systems could also have a material adverse effect on our business. To the extent that any disruption or security breach were to result in a loss of, or damage to, our data or applications, or inappropriate disclosure of confidential or proprietary information, we could incur liability and the further development and commercialization of our product candidates could be delayed.

Our employees, independent contractors, consultants, commercial partners and vendors may engage in misconduct or other improper activities, including noncompliance with regulatory standards and requirements.

We are exposed to the risk of employee fraud or other illegal activity by our employees, independent contractors, consultants, commercial partners and vendors. Misconduct by these parties could include intentional, reckless and/or negligent conduct that fails to comply with the laws of the FDA, EMA and other similar foreign regulatory bodies, provide true, complete and accurate information to the FDA, EMA and other similar foreign regulatory bodies, comply with manufacturing standards we have established, comply with healthcare fraud and abuse laws in the United States and similar foreign fraudulent misconduct laws, or report financial information or data accurately or to disclose unauthorized activities to us. If we obtain FDA approval of any of our product candidates and begin commercializing those products in the United States, our potential exposure under such laws will increase significantly, and our costs associated with compliance with such laws are also likely to increase. These laws may impact, among other things, our current activities with principal investigators and research patients, as well as proposed and future sales, marketing and education programs. In connection with this offering, we will adopt a code of business conduct and ethics, but it is not always possible to identify and deter misconduct by our employees, independent contractors, consultants, commercial partners and vendors, and the precautions we take to detect and prevent this activity may not be effective in controlling unknown or unmanaged risks or losses or in protecting us from governmental investigations or other actions or lawsuits stemming from a failure to comply with these laws or regulations. If any actions are instituted against us and we are not successful in defending ourselves or asserting our rights, those actions could result in the imposition of civil, criminal and administrative penalties, damages, monetary fines, individual imprisonment, disgorgement, possible exclusion from participation in government healthcare programs, additional reporting obligations and oversight if we become subject to a corporate integrity agreement or other agreement to resolve allegations of non-compliance with these laws, contractual damages, reputational harm, diminished profits and future earnings and the curtailment of our operations.

We have disclosed that there is substantial doubt about our ability to continue as a going concern.

In Note 1 to our consolidated financial statements, we disclose that there is substantial doubt about our ability to continue as a going concern. If we are unable to obtain sufficient funding, we could be forced to delay, reduce or eliminate all of our research and development programs, product portfolio expansion or commercialization efforts, and our financial condition and results of operations will be materially and

adversely affected and we may be unable to continue as a going concern. After the completion of this offering, future financial statements may disclose substantial doubt about our ability to continue as a going concern. If we seek additional financing to fund our business activities in the future and there remains substantial doubt about our ability to continue as a going concern, investors or other financing sources may be unwilling to provide additional funding to us on commercially reasonable terms or at all.

Risks Related to Research and Development and the Biopharmaceutical Industry

Preclinical development is uncertain. Our preclinical programs may experience delays or may never advance to clinical trials, which would adversely affect our ability to obtain regulatory approvals or commercialize these programs on a timely basis or at all, which would have an adverse effect on our business.

We have yet to nominate a clinical candidate for any of our programs, other than SRK-015. Before we can commence clinical trials for any product candidate in these programs, we must complete extensive preclinical studies that support our planned INDs in the United States, or similar applications in other jurisdictions. We cannot be certain of the timely completion or outcome of our preclinical studies and cannot predict if the FDA, EMA or other regulatory authorities will accept our proposed clinical programs or if the outcome of our preclinical studies will ultimately support the further development of our programs. As a result, we cannot be sure that we will be able to submit INDs or similar applications for our preclinical programs on the timelines we expect, if at all, and we cannot be sure that submission of INDs or similar applications will result in the FDA, the EMA or other regulatory authorities allowing clinical trials to begin.

Due to our limited resources and access to capital, we must prioritize development of certain programs and product candidates; these decisions may prove to be wrong and may adversely affect our business.

We may fail to identify viable new product candidates for clinical development from our current or future research programs for a number of reasons. If we fail to identify additional potential product candidates, our business could be materially harmed.

Research programs to pursue the development of our existing and planned product candidates for additional indications and to identify new product candidates and disease targets require substantial technical, financial and human resources whether or not they are ultimately successful. Our research programs may initially show promise in identifying potential indications and/or product candidates, yet fail to yield results for clinical development for a number of reasons, including:

- § the research methodology used may not be successful in identifying potential indications and/or product candidates;
- § potential product candidates may, after further study, be shown to have harmful adverse effects or other characteristics that indicate they are unlikely to be effective products; or
- § it may take greater human and financial resources than we will possess to identify additional therapeutic opportunities for our product candidates or to develop suitable potential product candidates through internal research programs, thereby limiting our ability to develop, diversify and expand our product portfolio.

Because we have limited financial and human resources, we intend to initially focus on research programs and product candidates for a limited set of indications. As a result, we may forego or delay pursuit of opportunities with other product candidates or for other indications that later prove to have greater commercial potential or a greater likelihood of success. Our resource allocation decisions may cause us to fail to capitalize on viable commercial products or profitable market opportunities.

Accordingly, there can be no assurance that we will ever be able to identify additional therapeutic opportunities for our product candidates or to develop suitable potential product candidates through internal research programs, which could materially adversely affect our future growth and prospects. We may focus our efforts and resources on potential product candidates or other potential programs that ultimately prove to be unsuccessful.

The successful development of biopharmaceuticals is highly uncertain.

Successful development of biopharmaceuticals is highly uncertain and is dependent on numerous factors, many of which are beyond our control. Product candidates that appear promising in the early phases of development may fail to reach the market for several reasons including:

- § preclinical study results may show the product candidate to be less effective than desired or to have harmful or problematic side effects;
- § clinical trial results may show the product candidates to be less effective than expected (e.g., a clinical trial could fail to meet its primary endpoint(s)) or to have unacceptable side effects or toxicities;
- § failure to receive the necessary regulatory approvals or a delay in receiving such approvals. Among other things, such delays may be caused by slow enrollment in clinical trials, patients dropping out of trials, length of time to achieve trial endpoints, additional time requirements for data analysis, or biologics license application, or BLA, preparation, discussions with the FDA, an FDA request for additional preclinical or clinical data, or unexpected safety or manufacturing issues;
- § manufacturing costs, formulation issues, pricing or reimbursement issues, or other factors that make a product candidate uneconomical; and
- § the proprietary rights of others and their competing products and technologies that may prevent one of our product candidates from being commercialized.

The length of time necessary to complete clinical trials and to submit an application for marketing approval for a final decision by a regulatory authority varies significantly from one product candidate to the next, and may be difficult to predict.

Even if we are successful in getting market approval, commercial success of any approved products will also depend in large part on the availability of coverage and adequate reimbursement from third-party payors, including government payors such as the Medicare and Medicaid programs and managed care organizations, which may be affected by existing and future health care reform measures designed to reduce the cost of health care. Third-party payors could require us to conduct additional studies, including post-marketing studies related to the cost effectiveness of a product, to qualify for reimbursement, which could be costly and divert our resources. If government and other health care payors were not to provide coverage and adequate reimbursement levels for one any of our products once approved, market acceptance and commercial success would be reduced.

In addition, if any of our product candidates are approved for marketing, we will be subject to significant regulatory obligations regarding the submission of safety and other post-marketing information and reports and registration, and will need to continue to comply (or ensure that our third-party providers) comply with current good manufacturing practices, or cGMPs, and good clinical practices, or GCPs, for any clinical trials that we conduct post-approval. In addition, there is always the risk that we or a regulatory authority might identify previously unknown problems with a product post-approval, such as adverse events of unanticipated severity or frequency. Compliance with these requirements is costly, and any failure to comply or other issues with our product candidates post-approval could adversely affect our business, financial condition and results of operations.

Clinical development involves a lengthy and expensive process, with an uncertain outcome. We may incur additional costs or experience delays in completing, or ultimately be unable to complete, the development and commercialization of SRK-015 or any future product candidates.

To obtain the requisite regulatory approvals to commercialize any product candidates, we must demonstrate through extensive preclinical studies and clinical trials that our product candidates are safe, pure and potent or effective in humans. Clinical testing is expensive and can take many years to complete, and its outcome is inherently uncertain. We may be unable to establish clinical endpoints that applicable regulatory authorities would consider clinically meaningful, and a clinical trial can fail at any stage of testing.

Differences in trial design between early-stage clinical trials and later-stage clinical trials make it difficult to extrapolate the results of earlier clinical trials to later clinical trials. Moreover, clinical data are often susceptible to varying interpretations and analyses, and many companies that have believed their product candidates performed satisfactorily in clinical trials have nonetheless failed to obtain marketing approval of their products.

Successful completion of clinical trials is a prerequisite to submitting a BLA to the FDA, a Marketing Authorization Application, or MAA, to the EMA, and similar marketing applications to comparable foreign regulatory authorities, for each product candidate and, consequently, the ultimate approval and commercial marketing of any product candidates. We do not know whether any of our clinical trials will begin or be completed on schedule, if at all.

We may experience delays in initiating or completing clinical trials. We also may experience numerous unforeseen events during, or as a result of, any future clinical trials that we could conduct that could delay or prevent our ability to receive marketing approval or commercialize SRK-015 or any future product candidates, including:

- § regulators or institutional review boards, or IRBs, or ethics committees may not authorize us or our investigators to commence a clinical trial or conduct a clinical trial at a prospective trial site;
- § we may experience delays in reaching, or fail to reach, agreement on acceptable terms with prospective trial sites and prospective CROs the terms of which can be subject to extensive negotiation and may vary significantly among different CROs and trial sites;
- § clinical trials of any product candidates may fail to show safety, purity or potency, or produce negative or inconclusive results and we may decide, or regulators may require us, to conduct additional preclinical studies or clinical trials or we may decide to abandon product development programs;
- § the number of subjects required for clinical trials of any product candidates may be larger than we anticipate, enrollment in these clinical trials may be slower than we anticipate or subjects may drop out of these clinical trials or fail to return for post-treatment follow-up at a higher rate than we anticipate;
- § our third-party contractors may fail to comply with regulatory requirements or meet their contractual obligations to us in a timely manner, or at all, or may deviate from the clinical trial protocol or drop out of the trial, which may require that we add new clinical trial sites or investigators;
- § we may elect to, or regulators, IRBs or ethics committees may require that we or our investigators, suspend or terminate clinical research or trials for various reasons, including noncompliance with regulatory requirements or a finding that the participants are being exposed to unacceptable health risks;
- § the cost of clinical trials of any product candidates may be greater than we anticipate;
- § the supply or quality of our product candidates or other materials necessary to conduct clinical trials of our product candidates may be insufficient or inadequate to initiate or complete a given clinical trial;
- § our product candidates may have undesirable side effects or other unexpected characteristics, causing us or our investigators, regulators, IRBs or ethics committees to suspend or terminate the trials, or reports from clinical testing of other therapies may raise safety or efficacy concerns about our product candidates; and
- § the FDA, EMA or other regulatory authorities may require us to submit additional data such as long-term toxicology studies, or impose other requirements before permitting us to initiate a clinical trial.

We could also encounter delays if a clinical trial is suspended or terminated by us, the IRBs of the institutions in which such trials are being conducted, or the FDA, EMA or other regulatory authorities, or

recommended for suspension or termination by the Data Safety Monitoring Board, or DSMB, for such trial. A suspension or termination may be imposed due to a number of factors, including failure to conduct the clinical trial in accordance with regulatory requirements or our clinical protocols, inspection of the clinical trial operations or trial site by the FDA, EMA or other regulatory authorities resulting in the imposition of a clinical hold, unforeseen safety issues or adverse side effects, failure to demonstrate a benefit from using a product or treatment, failure to establish or achieve clinically meaningful trial endpoints, changes in governmental regulations or administrative actions or lack of adequate funding to continue the clinical trial. Many of the factors that cause, or lead to, a delay in the commencement or completion of clinical trials may also ultimately lead to the denial of regulatory approval of our product candidates. Further, the FDA, EMA or other regulatory authorities may disagree with our clinical trial design and our interpretation of data from clinical trials, or may change the requirements for approval even after they have reviewed and commented on the design for our clinical trials.

Our product development costs will increase if we experience delays in clinical testing or marketing approvals. We do not know whether any of our clinical trials will begin as planned, will need to be restructured or will be completed on schedule, or at all. Significant clinical trial delays also could shorten any periods during which we may have the exclusive right to commercialize our product candidates and may allow our competitors to bring products to market before we do, potentially impairing our ability to successfully commercialize our product candidates and harming our business and results of operations. Any delays in our clinical development programs may harm our business, financial condition and results of operations significantly.

Our future clinical trials or those of our future collaborators may reveal significant adverse events not seen in our preclinical studies and may result in a safety profile that could inhibit regulatory approval or market acceptance of any of our product candidates.

Before obtaining regulatory approvals for the commercial sale of any products, we must demonstrate through lengthy, complex and expensive preclinical studies and clinical trials that SRK-015 and any future product candidate is both safe and effective for use in its target indication. Clinical testing is expensive and can take many years to complete, and its outcome is inherently uncertain. Failure can occur at any time during the clinical trial process. The results of preclinical studies and early clinical trials of our product candidates may not be predictive of the results of later-stage clinical trials. In addition, initial success in clinical trials may not be indicative of results obtained when such trials are completed. There is typically an extremely high rate of attrition from the failure of product candidates proceeding through clinical trials. Product candidates in later stages of clinical trials may fail to show the desired safety and efficacy profile despite having progressed through preclinical studies and initial clinical trials. A number of companies in the biopharmaceutical industry have suffered significant setbacks in advanced clinical trials due to lack of efficacy or unacceptable safety issues, notwithstanding promising results in earlier trials. Most product candidates that commence clinical trials are never approved as products and there can be no assurance that any of our future clinical trials will ultimately be successful or support further clinical development of SRK-015 or any of our future product candidates.

If significant adverse events or other side effects are observed in any of our clinical trials, we may have difficulty recruiting patients to our clinical trials, patients may drop out of our trials, or we may be required to abandon the trials or our development efforts of one or more product candidates altogether. We, the FDA, EMA or other applicable regulatory authorities, or an IRB may suspend clinical trials of a product candidate at any time for various reasons, including a belief that subjects in such trials are being exposed to unacceptable health risks or adverse side effects. Some potential therapeutics developed in the biotechnology industry that initially showed therapeutic promise in early-stage trials have later been found to cause side effects that prevented their further development. Even if the side effects do not preclude the product candidate from obtaining or maintaining marketing approval, undesirable side effects may inhibit

market acceptance of the approved product due to its tolerability versus other therapies. Any of these developments could materially harm our business, financial condition and prospects.

If we encounter difficulties enrolling patients in our clinical trials, our clinical development activities could be delayed or otherwise adversely affected.

We may experience difficulties in patient enrollment in our clinical trials for a variety of reasons. The timely completion of clinical trials in accordance with their protocols depends, among other things, on our ability to enroll a sufficient number of patients who remain in the trial until its conclusion. The enrollment of patients depends on many factors, including:

- § the patient eligibility and exclusion criteria defined in the protocol;
- § the size of the patient population required for analysis of the trial's primary endpoints;
- § the proximity of patients to trial sites;
- § the design of the trial;
- § our ability to recruit clinical trial investigators with the appropriate competencies and experience;
- § our ability to obtain and maintain patient consents; and
- § the risk that patients enrolled in clinical trials will drop out of the trials before completion.

For example, we are initially developing SRK-015 for the treatment of SMA, which is a rare disease, affecting only an estimated 30,000 to 35,000 patients in the United States and Europe. As a result, we may encounter difficulties enrolling subjects in our clinical trials for SRK-015 due, in part, to the small size of this patient population. In addition, our clinical trials will compete with other clinical trials for product candidates that are in the same therapeutic areas as our product candidates, and this competition will reduce the number and types of patients available to us, because some patients who might have opted to enroll in our trials may instead opt to enroll in a trial being conducted by one of our competitors. Since the number of qualified clinical investigators is limited, we expect to conduct some of our clinical trials at the same clinical trial sites that some of our competitors use, which will reduce the number of patients who are available for our clinical trials in such clinical trial site.

Delays in patient enrollment may result in increased costs or may affect the timing or outcome of our future clinical trials, which could prevent completion of these trials and adversely affect our ability to advance the development of our product candidates.

If product liability lawsuits are brought against us, we may incur substantial liabilities and may be required to limit commercialization of our product candidates.

We face an inherent risk of product liability as a result of testing SRK-015 and any of our future product candidates in clinical trials and will face an even greater risk if we commercialize any products, if approved. For example, we may be sued if our product candidates cause or are perceived to cause injury or are found to be otherwise unsuitable during clinical trials, manufacturing, marketing or sale. Any such product liability claims may include allegations of defects in manufacturing, defects in design, a failure to warn of dangers inherent in the product, negligence, strict liability or a breach of warranties. Claims could also be asserted under state consumer protection acts. If we cannot successfully defend ourselves against product liability claims, we may incur substantial liabilities or be required to limit commercialization of our product candidates. Even successful defense would require significant financial and management resources. Regardless of the merits or eventual outcome, liability claims may result in:

- § inability to bring a product candidate to the market;
- § decreased demand for our products;
- § injury to our reputation;
- § withdrawal of clinical trial participants and inability to continue clinical trials;
- § initiation of investigations by regulators;

- § costs to defend the related litigation;
- § diversion of management's time and our resources;
- § substantial monetary awards to trial participants;
- § product recalls, withdrawals or labeling, marketing or promotional restrictions;
- § loss of revenue;
- § exhaustion of any available insurance and our capital resources;
- § the inability to commercialize any product candidate, if approved; and
- § decline in our share price.

Our inability to obtain sufficient product liability insurance at an acceptable cost to protect against potential product liability claims could prevent or inhibit the commercialization of products we develop, alone or with collaborators. We need to obtain additional insurance for clinical trials as our lead product candidate SRK-015 enters the clinical development phase. However, we may be unable to obtain, or may obtain on unfavorable terms, clinical trial insurance in amounts adequate to cover any liabilities from any of our clinical trials. Our insurance policies may also have various exclusions, and we may be subject to a product liability claim for which we have no coverage. We may have to pay any amounts awarded by a court or negotiated in a settlement that exceed our coverage limitations or that are not covered by our insurance, and we may not have, or be able to obtain, sufficient capital to pay such amounts. Even if our agreements with any future corporate collaborators entitle us to indemnification against losses, such indemnification may not be available or adequate should any claim arise.

We face significant competition from other biotechnology and pharmaceutical companies, and our operating results will suffer if we fail to compete effectively.

The biopharmaceutical industry is characterized by intense competition and rapid innovation. Our competitors may be able to develop other compounds or drugs that are able to achieve similar or better results. Our potential competitors include major multinational pharmaceutical companies, established biotechnology companies, specialty pharmaceutical companies and universities and other research institutions. Many of our competitors have substantially greater financial, technical and other resources, such as larger research and development staff and experienced marketing and manufacturing organizations and well-established sales forces. Smaller or early-stage companies may also prove to be significant competitors, particularly as they develop novel approaches to treating disease indications that our product candidates are also focused on treating. Established pharmaceutical companies may also invest heavily to accelerate discovery and development of novel therapeutics or to in-license novel therapeutics that could make the product candidates that we develop obsolete. Mergers and acquisitions in the biotechnology and pharmaceutical industries may result in even more resources being concentrated in our competitors. Competition may increase further as a result of advances in the commercial applicability of technologies and greater availability of capital for investment in these industries. Our competitors, either alone or with collaborative partners, may succeed in developing, acquiring or licensing on an exclusive basis drug or biologic products that are more effective, safer, more easily commercialized or less costly than our product candidates or may develop proprietary technologies or secure patent protection that we may need for the development of our technologies and products. We believe the key competitive factors that will affect the development and commercial success of our product candidates are efficacy, safety, tolerability, reliability, convenience of use, price and reimbursement.

We anticipate competing with other companies that are focused on treating disease indications that our product candidates are also focused on treating. A competitor may develop technologies focused on the same disease pathway as our technology or may focus on treating the targeted disease in a completely different manner. To the extent a new drug is developed that is more efficacious than any product candidate developed by us, this could reduce or negate the need for our product candidate. In addition, while we believe our product candidates may be used in conjunction with existing or emerging standard of care in

certain disease indications, including SMA, as companies continue to improve upon existing standard of care, more efficacious drug therapies could become available, reducing or completely negating the benefit of our product candidates. Our competitors may also include companies that are or will be developing therapies for the same therapeutic areas that we are targeting within our early pipeline, including neuromuscular disorders, cancer, fibrosis and anemia.

Even if we obtain regulatory approval of our product candidates, the availability and price of our competitors' products could limit the demand and the price we are able to charge for our product candidates. We may not be able to implement our business plan if the acceptance of our product candidates is inhibited by price competition or the reluctance of physicians to switch from existing methods of treatment to our product candidates, or if physicians switch to other new drug or biologic products or choose to reserve our product candidates for use in limited circumstances. For additional information regarding our competition, see "Business — Competition."

Even if a product candidate we develop receives marketing approval, it may fail to achieve the degree of market acceptance by physicians, patients, third-party payors and others in the medical community necessary for commercial success.

If SRK-015 or any future product candidate we develop receives marketing approval, whether as a single agent or in combination with other therapies, it may nonetheless fail to gain sufficient market acceptance by physicians, patients, third-party payors, and others in the medical community. For example, doctors may deem it sufficient to treat patients with SMA with an SMN upregulator such as nusinersen, and therefore will not be willing to utilize SRK-015 in conjunction with such SMN upregulator. If the product candidates we develop do not achieve an adequate level of acceptance, we may not generate significant product revenues and we may not become profitable. The degree of market acceptance of any product candidate, if approved for commercial sale, will depend on a number of factors, including:

- § efficacy and potential advantages compared to alternative treatments;
- § the ability to offer our products, if approved, for sale at competitive prices;
- § convenience and ease of administration compared to alternative treatments;
- § the willingness of the target patient population to try new therapies and of physicians to prescribe these therapies;
- § the strength of marketing and distribution support;
- § the ability to obtain sufficient third-party coverage and adequate reimbursement; and
- § the prevalence and severity of any side effects.

If we fail to comply with environmental, health and safety laws and regulations, we could become subject to fines or penalties or incur costs that could have a material adverse effect on the success of our business.

We are subject to numerous environmental, health and safety laws and regulations, including those governing laboratory procedures and the handling, use, storage, treatment and disposal of hazardous materials and wastes. Our research and development activities involve the use of biological and hazardous materials and produce hazardous waste products. We generally contract with third parties for the disposal of these materials and wastes. We cannot eliminate the risk of contamination or injury from these materials, which could cause an interruption of our commercialization efforts, research and development efforts and business operations, environmental damage resulting in costly clean-up and liabilities under applicable laws and regulations governing the use, storage, handling and disposal of these materials and specified waste products. Although we believe that the safety procedures utilized by our third-party manufacturers for handling and disposing of these materials generally comply with the standards prescribed by these laws and regulations, we cannot guarantee that this is the case or eliminate the risk of accidental contamination or injury from these materials. In such an event, we may be held liable for any resulting damages and such liability could exceed our resources and state or federal or other applicable authorities may curtail our use of certain materials and/or interrupt our business operations. Furthermore, environmental laws and

regulations are complex, change frequently and have tended to become more stringent. We cannot predict the impact of such changes and cannot be certain of our future compliance. In addition, we may incur substantial costs in order to comply with current or future environmental, health and safety laws and regulations. These current or future laws and regulations may impair our research, development or production efforts. Failure to comply with these laws and regulations also may result in substantial fines, penalties or other sanctions.

Although we maintain workers' compensation insurance to cover us for costs and expenses we may incur due to injuries to our employees resulting from the use of hazardous materials or other work-related injuries, this insurance may not provide adequate coverage against potential liabilities. We do not carry specific biological waste or hazardous waste insurance coverage, workers compensation or property and casualty and general liability insurance policies that include coverage for damages and fines arising from biological or hazardous waste exposure or contamination.

Comprehensive Tax Reform Legislation Could Adversely Affect Our Business And Financial Condition.

On December 22, 2017, President Trump signed into law the "Tax Cuts and Jobs Act," or the TCJA, that significantly reforms the Internal Revenue Code of 1986, as amended, or the Code. The TCJA, among other things, contains significant changes to corporate taxation, including reduction of the corporate tax rate, limitation of the tax deduction for interest expense, limitation of the deduction for net operating losses and elimination of net operating loss carrybacks and modifying or repealing many business deductions and credits (including reducing the business tax credit for certain clinical testing expenses incurred in the testing of certain drugs for rare diseases or conditions generally referred to as "orphan drugs"). Our net deferred tax assets and liabilities were revalued at the newly enacted U.S. corporate rate. We continue to examine the impact this tax reform legislation may have on our business. The overall impact of the TCJA is uncertain and our business and financial condition could be adversely affected.

Our ability to use our net operating loss carryforwards and certain tax credit carryforwards may be subject to limitation.

As of December 31, 2017, we had net operating loss carryforwards for federal and state income tax purposes of \$50.4 million and \$49.8 million, respectively, which begin to expire in 2034. As of December 31, 2017, we also had available tax credit carryforwards for federal and state income tax purposes of \$1.1 million and \$0.7 million, respectively, which begin to expire in 2034 and 2020, respectively. Under Section 382 of the Code, changes in our ownership may limit the amount of our net operating loss carryforwards and tax credit carryforwards that could be utilized annually to offset our future taxable income, if any. This limitation would generally apply in the event of a cumulative change in ownership of our company of more than 50% within a three-year period. Any such limitation may significantly reduce our ability to utilize our net operating loss carryforwards and tax credit carryforwards before they expire. Private placements and other transactions that have occurred since our inception, as well as this offering, may trigger such an ownership change pursuant to Section 382. Any such limitation, whether as the result of this offering, prior private placements, sales of our common stock by our existing stockholders or additional sales of our common stock by us, could have a material adverse effect on our results of operations in future years. The reduction of the corporate tax rate under TCJA may cause a reduction in the economic benefit of our net operating loss carryforwards and other deferred tax assets available to us. Under the TCJA, net operating losses generated after December 31, 2017 will not be subject to expiration.

Our current operations are concentrated in one location, and we or the third parties upon whom we depend may be adversely affected by earthquakes or other natural disasters and our business continuity and disaster recovery plans may not adequately protect us from a serious disaster.

Our current operations are located in our facilities in Cambridge, Massachusetts. Any unplanned event, such as flood, fire, explosion, earthquake, extreme weather condition, medical epidemics, power shortage, telecommunication failure or other natural or manmade accidents or incidents that result in us being unable to fully utilize our facilities, or the manufacturing facilities of our third-party contract manufacturers, may have a material and adverse effect on our ability to operate our business, particularly on a daily basis, and have significant negative consequences on our financial and operating conditions. Loss of access to these facilities may result in increased costs, delays in the development of our product candidates or interruption of our business operations. Earthquakes or other natural disasters could further disrupt our operations, and have a material and adverse effect on our business, financial condition, results of operations and prospects. If a natural disaster, power outage or other event occurred that prevented us from using all or a significant portion of our headquarters, that damaged critical infrastructure, such as our research facilities or the manufacturing facilities of our third-party contract manufacturers, or that otherwise disrupted operations, it may be difficult or, in certain cases, impossible, for us to continue our business for a substantial period of time. The disaster recovery and business continuity plans we have in place may prove inadequate in the event of a serious disaster or similar event. We may incur substantial expenses as a result of the limited nature of our disaster recovery and business continuity plans, which, could have a material adverse effect on our business. As part of our risk management policy, we maintain insurance coverage at levels that we believe are appropriate for our business. However, in the event of an accident or incident at these facilities, we cannot assure you that the amounts of insurance will be sufficient to satisfy any damages and losses. If our facilities, or the manufacturing facilities of our third-party contract manufacturers, are unable to operate because of an accident or incident or for any other reason, even for a short period of time, any or all of our research and development programs may be harmed. Any business interruption may have a material and adverse effect on our business, financial condition, results of operations and prospects.

Risks Related to Government Regulation

The regulatory approval process for our product candidates in the United States, EU and other jurisdictions is currently uncertain and will be lengthy, time-consuming and inherently unpredictable and we may experience significant delays in the clinical development and regulatory approval, if any, of our product candidates.

The research, testing, manufacturing, labeling, approval, selling, import, export, marketing and distribution of drug products, including biologics, are subject to extensive regulation by the FDA in the United States and other regulatory authorities. We are not permitted to market any biological product in the United States until we receive a biologics license from the FDA. We have not previously submitted a BLA to the FDA, or similar marketing application to comparable foreign authorities. A BLA must include extensive preclinical and clinical data and supporting information to establish that the product candidate is safe, pure and potent for each desired indication. A BLA must also include significant information regarding the chemistry, manufacturing and controls for the product, and the manufacturing facilities must complete a successful pre-license inspection.

The FDA may also require a panel of experts, referred to as an Advisory Committee, to deliberate on the adequacy of the safety and efficacy data to support approval. The opinion of the Advisory Committee, although not binding, may have a significant impact on our ability to obtain approval of any product candidates that we develop based on the completed clinical trials.

In addition, clinical trials can be delayed or terminated for a variety of reasons, including delays or failures related to:

- § obtaining regulatory authorization to begin a clinical trial, if applicable;
- § the availability of financial resources to begin and complete the planned trials;
- § reaching agreement on acceptable terms with prospective CROs and clinical trial sites, the terms of which can be subject to extensive negotiation and may vary significantly among different CROs and trial sites;
- § obtaining approval at each clinical trial site by an independent IRB or ethics committee;
- § recruiting suitable patients to participate in a trial in a timely manner;
- § having patients complete a trial or return for post-treatment follow-up;
- § clinical trial sites deviating from trial protocol, not complying with GCP requirements or dropping out of a trial;
- § addressing any patient safety concerns that arise during the course of a clinical trial;
- § addressing any conflicts with new or existing laws or regulations;
- § adding new clinical trial sites; or
- § manufacturing qualified materials under cGMP regulations for use in clinical trials.

Patient enrollment is a significant factor in the timing of clinical trials and is affected by many factors. Further, a clinical trial may be suspended or terminated by us, the IRBs for the institutions in which such trials are being conducted, or the FDA, EMA or other regulatory authorities, or recommended for suspension or termination by the DSMB for such trial, due to a number of factors, including failure to conduct the clinical trial in accordance with regulatory requirements or our clinical protocols, inspection of the clinical trial operations or trial site by the FDA, EMA or other regulatory authorities resulting in the imposition of a clinical hold, unforeseen safety issues or adverse side effects, failure to demonstrate a benefit from using a product candidate, changes in governmental regulations or administrative actions or lack of adequate funding to continue the clinical trial. If we experience termination of, or delays in the completion of, any clinical trial of our product candidates, the commercial prospects for our product candidates will be harmed, and our ability to generate product revenue will be delayed. In addition, any delays in completing any clinical trials will increase our costs, slow down our product development and approval process and jeopardize our ability to commence product sales and generate revenue.

The FDA may disagree with our regulatory plan and we may fail to obtain regulatory approval of our product candidates.

The general approach for FDA approval of a new biologic or drug is dispositive data from two well-controlled, Phase 3 clinical trials of the relevant biologic or drug in the relevant patient population. Phase 3 clinical trials typically involve hundreds of patients, have significant costs and take years to complete.

Our clinical trials results may also not support approval. In addition, our product candidates could fail to receive regulatory approval for many reasons, including the following:

- § the FDA, EMA or comparable foreign regulatory authorities may disagree with the design or implementation of our clinical trials;
- § we may be unable to demonstrate to the satisfaction of the FDA, EMA or comparable foreign regulatory authorities that our product candidates are safe and effective for any of their proposed indications;
- § the results of clinical trials may not meet the level of statistical significance required by the FDA, EMA or comparable foreign regulatory authorities for approval;
- § we may be unable to demonstrate that our product candidates' clinical and other benefits outweigh their safety risks;

- § the FDA, EMA or comparable foreign regulatory authorities may disagree with our interpretation of data from preclinical studies or clinical trials;
- § the data collected from clinical trials of our product candidates may not be sufficient to the satisfaction of the FDA, EMA or comparable foreign regulatory authorities to support the submission of a BLA or other comparable submission in foreign jurisdictions or to obtain regulatory approval in the United States or elsewhere;
- § the FDA, EMA or comparable foreign regulatory authorities may fail to approve the manufacturing processes or facilities of third-party manufacturers with which we contract for clinical and commercial supplies; and
- § the approval policies or regulations of the FDA, EMA or comparable foreign regulatory authorities may significantly change in a manner rendering our clinical data insufficient for approval.

We have received Orphan Drug Designation from the FDA for SRK-015 for the treatment of SMA and we may seek Orphan Drug Designation for our future product candidates, and we may be unable to maintain the benefits associated with Orphan Drug Designation, including the potential for market exclusivity.

Regulatory authorities in some jurisdictions, including the United States and Europe, may designate drugs and therapeutic biologics for relatively small patient populations as orphan drugs. Under the Orphan Drug Act, the FDA may designate a drug or therapeutic biologic as an orphan drug if it is a drug or therapeutic biologic intended to treat a rare disease or condition, which is generally defined as a patient population of fewer than 200,000 individuals annually in the United States, or a patient population greater than 200,000 in the United States where there is no reasonable expectation that the cost of developing the drug or therapeutic biologic will be recovered from sales in the United States. In the United States, Orphan Drug Designation entitles a party to financial incentives such as opportunities for grant funding toward clinical trial costs, tax advantages and user-fee waivers. In addition, if a product that has Orphan Drug Designation subsequently receives the first FDA approval for the disease for which it has such designation, the product is entitled to orphan drug exclusivity, which means that the FDA may not approve any other applications, including a full NDA or BLA, to market the same product for the same indication for seven years, except in limited circumstances, such as a showing of clinical superiority to the product with orphan drug exclusivity or where the manufacturer is unable to assure sufficient product quantity.

Even though we obtained Orphan Drug Designation from the FDA for SRK-015 for the treatment of SMA, or if we obtain Orphan Drug Designation for any of our future product candidates in specific indications, we may not be the first to obtain marketing approval of these product candidates for the orphan-designated indication due to the uncertainties associated with developing pharmaceutical products. In addition, exclusive marketing rights in the United States may be limited if we seek approval for an indication broader than the orphan-designated indication or may be lost if the FDA later determines that the request for designation was materially defective or if the manufacturer is unable to assure sufficient quantities of the product to meet the needs of patients with the rare disease or condition. Further, even if we obtain orphan drug exclusivity for a product, that exclusivity may not effectively protect the product from competition because different drugs or therapeutic biologics with different active moieties can be approved for the same condition. Even after an orphan product is approved, the FDA can subsequently approve the same drug or therapeutic biologic with the same active moiety for the same condition if the FDA concludes that the later drug or therapeutic biologic is safer, more effective or makes a major contribution to patient care. Orphan Drug Designation neither shortens the development time or regulatory review time of a drug or therapeutic biologic nor gives the drug or therapeutic biologic any advantage in the regulatory review or approval process. In addition, while we may seek Orphan Drug Designation for our future product candidates, we may never receive such designations.

We may seek Breakthrough Therapy Designation or Fast Track Designation from the FDA, for certain of our product candidates, but receipt of either such designation may not actually lead to a faster development or regulatory review or approval process.

We may seek Breakthrough Therapy Designation or Fast Track Designation for certain of our product candidates.

A breakthrough therapy is defined as a product that is intended, alone or in combination with one or more other products, to treat a serious or life-threatening disease or condition, and preliminary clinical evidence indicates that the product may demonstrate substantial improvement over existing therapies on one or more clinically significant endpoints, such as substantial treatment effects observed early in clinical development. For products that have been designated as breakthrough therapies, interaction and communication between the FDA and the sponsor of the trial can help to identify the most efficient path for clinical development while minimizing the number of patients placed in ineffective control regimens. Products designated as breakthrough therapies by the FDA can also be eligible for accelerated approval.

Designation as a breakthrough therapy is within the discretion of the FDA. Accordingly, even if we believe one of our product candidates meets the criteria for designation as a breakthrough therapy, the FDA may disagree and instead determine not to make such designation. In any event, the receipt of a Breakthrough Therapy Designation for a product candidate may not result in a faster development process, review or approval compared to products considered for approval under conventional FDA procedures and does not assure ultimate approval by the FDA. In addition, even if one or more of our product candidates qualify as breakthrough therapies, the FDA may later decide that the products no longer meet the conditions for qualification and rescind the breakthrough designation.

If a product is intended for the treatment of a serious or life-threatening condition and the product demonstrates the potential to address unmet medical needs for this condition, the product sponsor may apply for Fast Track Designation. The FDA has broad discretion whether or not to grant this designation, so even if we believe a particular product candidate is eligible for this designation, we cannot assure you that the FDA would decide to grant it. The FDA has broad discretion whether or not to grant this designation, so even if we believe a particular product candidate is eligible for this designation, we cannot assure you that the FDA would decide to grant it. Even if we do receive Fast Track Designation, we may not experience a faster development process, review or approval compared to conventional FDA procedures. The FDA may withdraw Fast Track Designation if it believes that the designation is no longer supported by data from our clinical development program.

Our relationships with healthcare providers and physicians and third-party payors will be subject to applicable anti-kickback, fraud and abuse and other healthcare laws and regulations, which could expose us to criminal sanctions, civil penalties, contractual damages, reputational harm and diminished profits and future earnings.

Healthcare providers, physicians and third-party payors in the United States and elsewhere play a primary role in the recommendation and prescription of pharmaceutical products. Arrangements with third-party payors and customers can expose pharmaceutical manufacturers to broadly applicable fraud and abuse and other healthcare laws and regulations, including, without limitation, the federal Anti-Kickback Statute and the federal False Claims Act, which may constrain the business or financial arrangements and relationships through which such companies sell, market and distribute pharmaceutical products. In particular, the research of our product candidates, as well as the promotion, sales and marketing of healthcare items and services, as well as certain business arrangements in the healthcare industry, are subject to extensive laws designed to prevent fraud, kickbacks, self-dealing and other abusive practices. These laws and regulations may restrict or prohibit a wide range of pricing, discounting, marketing and promotion, structuring and commission(s), certain customer incentive programs and other business arrangements generally. Activities subject to these laws also involve the improper use of information obtained in the course of patient

recruitment for clinical trials. The applicable federal, state and foreign healthcare laws and regulations laws that may affect our ability to operate include, but are not limited to:

- § the federal Anti-Kickback Statute, which prohibits, among other things, knowingly and willfully soliciting, receiving, offering or paying any remuneration (including any kickback, bribe, or rebate), directly or indirectly, overtly or covertly, in cash or in kind, to induce, or in return for, either the referral of an individual, or the purchase, lease, order or recommendation of any good, facility, item or service for which payment may be made, in whole or in part, under a federal healthcare program, such as the Medicare and Medicaid programs. A person or entity can be found guilty of violating the statute without actual knowledge of the statute or specific intent to violate it. In addition, a claim including items or services resulting from a violation of the federal Anti-Kickback Statute constitutes a false or fraudulent claim for purposes of the federal False Claims Act, or FCA. The Anti-Kickback Statute has been interpreted to apply to arrangements between pharmaceutical manufacturers on the one hand and prescribers, purchasers, and formulary managers on the other. There are a number of statutory exceptions and regulatory safe harbors protecting some common activities from prosecution;
- § the federal civil and criminal false claims laws and civil monetary penalty laws, including the FCA, which prohibit, among other things, individuals or entities from knowingly presenting, or causing to be presented, false or fraudulent claims for payment to, or approval by Medicare, Medicaid, or other federal healthcare programs, knowingly making, using or causing to be made or used a false record or statement material to a false or fraudulent claim or an obligation to pay or transmit money to the federal government, or knowingly concealing or knowingly and improperly avoiding or decreasing or concealing an obligation to pay money to the federal government. Manufacturers can be held liable under the FCA even when they do not submit claims directly to government payors if they are deemed to "cause" the submission of false or fraudulent claims. The FCA also permits a private individual acting as a "whistleblower" to bring actions on behalf of the federal government alleging violations of the FCA and to share in any monetary recovery;
- § the federal Health Insurance Portability and Accountability Act of 1996, or HIPAA, which created additional federal criminal statutes that prohibit knowingly and willfully executing, or attempting to execute, a scheme to defraud any healthcare benefit program or obtain, by means of false or fraudulent pretenses, representations, or promises, any of the money or property owned by, or under the custody or control of, any healthcare benefit program, regardless of the payor (e.g., public or private) and knowingly and willfully falsifying, concealing or covering up by any trick or device a material fact or making any materially false statements in connection with the delivery of, or payment for, healthcare benefits, items or services relating to healthcare matters. Similar to the federal Anti-Kickback Statute, a person or entity can be found guilty of violating HIPAA without actual knowledge of the statute or specific intent to violate it;
- § HIPAA, as amended by the Health Information Technology for Economic and Clinical Health Act of 2009, or HITECH, and their respective implementing regulations, which impose, among other things, requirements on certain covered healthcare providers, health plans, and healthcare clearinghouses, known as covered entities, as well as their respective business associates, independent contractors that perform services for covered entities that involve the use, or disclosure of, individually identifiable health information, relating to the privacy, security and transmission of individually identifiable health information without appropriate authorization. HITECH also created new tiers of civil monetary penalties, amended HIPAA to make civil and criminal penalties directly applicable to business associates, and gave state attorneys general new authority to file civil actions for damages or injunctions in federal courts to enforce the federal HIPAA laws and seek attorneys' fees and costs associated with pursuing federal civil actions;

- § the federal Physician Payments Sunshine Act, created under the Patient Protection and Affordable Care Act, and its implementing regulations, which require some manufacturers of drugs, devices, biologicals and medical supplies for which payment is available under Medicare, Medicaid or the Children's Health Insurance Program (with certain exceptions) to report annually to the United States Department of Health and Human Services, or HHS, information related to payments or other transfers of value made to physicians (defined to include doctors, dentists, optometrists, podiatrists and chiropractors) and teaching hospitals, as well as ownership and investment interests held by physicians and their immediate family members;
- § federal consumer protection and unfair competition laws, which broadly regulate marketplace activities and activities that potentially harm consumers; and
- § analogous state and foreign laws and regulations, such as state anti-kickback and false claims laws, which may apply to sales or marketing arrangements and claims involving healthcare items or services reimbursed by non-governmental third-party payors, including private insurers, and may be broader in scope than their federal equivalents; state and foreign laws that require pharmaceutical companies to comply with the pharmaceutical industry's voluntary compliance guidelines and the relevant compliance guidance promulgated by the federal government or otherwise restrict payments that may be made to healthcare providers; state and foreign laws that require drug manufacturers to report information related to payments and other transfers of value to physicians and other healthcare providers or marketing expenditures; state and local laws that require the registration of pharmaceutical sales representatives; and state and foreign laws governing the privacy and security of health information in certain circumstances, many of which differ from each other in significant ways and often are not preempted by HIPAA, thus complicating compliance efforts.

The distribution of pharmaceutical products is subject to additional requirements and regulations, including extensive record-keeping, licensing, storage and security requirements intended to prevent the unauthorized sale of pharmaceutical products.

The scope and enforcement of each of these laws is uncertain and subject to rapid change in the current environment of healthcare reform, especially in light of the lack of applicable precedent and regulations. Federal and state enforcement bodies have recently increased their scrutiny of interactions between healthcare companies and healthcare providers, which has led to a number of investigations, prosecutions, convictions and settlements in the healthcare industry. Ensuring business arrangements comply with applicable healthcare laws, as well as responding to possible investigations by government authorities, can be time- and resource-consuming and can divert a company's attention from the business.

It is possible that governmental and enforcement authorities will conclude that our business practices may not comply with current or future statutes, regulations or case law interpreting applicable fraud and abuse or other healthcare laws and regulations. If any such actions are instituted against us, and we are not successful in defending ourselves or asserting our rights, those actions could have a significant impact on our business, including the imposition of civil, criminal and administrative penalties, damages, fines, disgorgement, individual imprisonment, possible exclusion from participation in federal and state funded healthcare programs, contractual damages and the curtailment or restricting of our operations, as well as additional reporting obligations and oversight if we become subject to a corporate integrity agreement or other agreement to resolve allegations of non-compliance with these laws. Any action for violation of these laws, even if successfully defended, could cause a pharmaceutical manufacturer to incur significant legal expenses and divert management's attention from the operation of the business. Prohibitions or restrictions on sales or withdrawal of future marketed products could materially affect business in an adverse way.

Obtaining and maintaining regulatory approval of our product candidates in one jurisdiction does not mean that we will be successful in obtaining regulatory approval of our product candidates in other jurisdictions.

Obtaining and maintaining regulatory approval of our product candidates in one jurisdiction does not guarantee that we will be able to obtain or maintain regulatory approval in any other jurisdiction, while a failure or delay in obtaining regulatory approval in one jurisdiction may have a negative effect on the regulatory approval process in others. For example, even if the FDA grants marketing approval of a product candidate, the EMA or comparable regulatory authorities in foreign jurisdictions must also approve the manufacturing, marketing and promotion of the product candidate in those countries. Approval procedures vary among jurisdictions and can involve requirements and administrative review periods different from, and greater than, those in the United States, including additional preclinical studies or clinical trials as clinical trials conducted in one jurisdiction may not be accepted by regulatory authorities in other jurisdictions. In many jurisdictions outside the United States, a product candidate must be approved for reimbursement before it can be approved for sale in that jurisdiction. In some cases, the price that we intend to charge for our products is also subject to approval.

We may also submit marketing applications in other countries. Regulatory authorities in jurisdictions outside of the United States have requirements for approval of product candidates with which we must comply prior to marketing in those jurisdictions. Obtaining foreign regulatory approvals and compliance with foreign regulatory requirements could result in significant delays, difficulties and costs for us and could delay or prevent the introduction of our products in certain countries. If we fail to comply with the regulatory requirements in international markets and/or receive applicable marketing approvals, our target market will be reduced and our ability to realize the full market potential of our product candidates will be harmed.

Even if we receive regulatory approval of any product candidates, we will be subject to ongoing regulatory obligations and continued regulatory review, which may result in significant additional expense and we may be subject to penalties if we fail to comply with regulatory requirements or experience unanticipated problems with our product candidates.

If any of our product candidates are approved, they will be subject to ongoing regulatory requirements for manufacturing, labeling, packaging, storage, advertising, promotion, sampling, record-keeping, conduct of post-marketing studies and submission of safety, efficacy and other post-market information, including both federal and state requirements in the United States and requirements of comparable foreign regulatory authorities. In addition, we will be subject to continued compliance with cGMP and GCP requirements for any clinical trials that we conduct post-approval.

Manufacturers and manufacturers' facilities are required to comply with extensive FDA, EMA and comparable foreign regulatory authority requirements, including ensuring that quality control and manufacturing procedures conform to cGMP regulations. As such, we and our contract manufacturers will be subject to continual review and inspections to assess compliance with cGMP and adherence to commitments made in any BLA, other marketing application, and previous responses to inspection observations. Accordingly, we and others with whom we work must continue to expend time, money, and effort in all areas of regulatory compliance, including manufacturing, production and quality control.

Any regulatory approvals that we receive for our product candidates may be subject to limitations on the approved indicated uses for which the product may be marketed or to the conditions of approval, or contain requirements for potentially costly post-marketing testing, including Phase 4 clinical trials and surveillance to monitor the safety and efficacy of the product candidate. The FDA may also require a risk evaluation and mitigation strategies, or REMS, program as a condition of approval of our product candidates, which could entail requirements for long-term patient follow-up, a medication guide, physician communication plans or additional elements to ensure safe use, such as restricted distribution methods, patient registries and other risk minimization tools. In addition, if the FDA, EMA or a comparable foreign regulatory authority approves

our product candidates, we will have to comply with requirements including submissions of safety and other post-marketing information and reports and registration.

The FDA may impose consent decrees or withdraw approval if compliance with regulatory requirements and standards is not maintained or if problems occur after the product reaches the market. Later discovery of previously unknown problems with our product candidates, including adverse events of unanticipated severity or frequency, or with our third-party manufacturers or manufacturing processes, or failure to comply with regulatory requirements, may result in revisions to the approved labeling to add new safety information; imposition of post-market studies or clinical trials to assess new safety risks; or imposition of distribution restrictions or other restrictions under a REMS program. Other potential consequences include, among other things:

- § restrictions on the marketing or manufacturing of our products, withdrawal of the product from the market or voluntary or mandatory product recalls;
- § fines, warning letters or holds on clinical trials;
- § refusal by the FDA to approve pending applications or supplements to approved applications filed by us or suspension or revocation of license approvals;
- § product seizure or detention or refusal to permit the import or export of our product candidates; and
- § injunctions or the imposition of civil or criminal penalties.

The FDA strictly regulates marketing, labeling, advertising, and promotion of products that are placed on the market. Products may be promoted only for the approved indications and in accordance with the provisions of the approved label. The FDA and other agencies actively enforce the laws and regulations prohibiting the promotion of off-label uses and a company that is found to have improperly promoted off-label uses may be subject to significant liability. The policies of the FDA, EMA and of other regulatory authorities may change and additional government regulations may be enacted that could prevent, limit or delay regulatory approval of our product candidates. We cannot predict the likelihood, nature or extent of government regulation that may arise from future legislation or administrative action, either in the United States or abroad. If we are slow or unable to adapt to changes in existing requirements or the adoption of new requirements or policies, or if we are not able to maintain regulatory compliance, we may lose any marketing approval that we may have obtained and we may not achieve or sustain profitability.

Coverage and reimbursement may be limited or unavailable in certain market segments for our product candidates, if approved, which could make it difficult for us to sell any product candidates profitably.

The success of our product candidates, if approved, depends on the availability of coverage and adequate reimbursement from third-party payors. We cannot be sure that coverage and reimbursement will be available for, or accurately estimate the potential revenue from, our product candidates or assure that coverage and reimbursement will be available for any product that we may develop.

Patients who are provided medical treatment for their conditions generally rely on third-party payors to reimburse all or part of the costs associated with their treatment. Coverage and adequate reimbursement from governmental healthcare programs, such as Medicare and Medicaid, and commercial payors is critical to new product acceptance.

Government authorities and third-party payors, such as private health insurers and health maintenance organizations, decide which drugs and treatments they will cover and the amount of reimbursement. Coverage and reimbursement by a third-party payor may depend upon a number of factors, including the third-party payor's determination that use of a product is:

- § a covered benefit under its health plan;
- § safe, effective and medically necessary;
- § appropriate for the specific patient;
- § cost-effective; and

§ neither experimental nor investigational.

In the United States, no uniform policy of coverage and reimbursement for products exists among third-party payors. As a result, obtaining coverage and reimbursement approval of a product from a government or other third-party payor is a time-consuming and costly process that could require us to provide to each payor supporting scientific, clinical and cost-effectiveness data for the use of our products on a payor-by-payor basis, with no assurance that coverage and adequate reimbursement will be obtained. Even if we obtain coverage for a given product, the resulting reimbursement payment rates might not be adequate for us to achieve or sustain profitability or may require co-payments that patients find unacceptably high. Additionally, third-party payors may not cover, or provide adequate reimbursement for, long-term follow-up evaluations required following the use of product candidates. Patients are unlikely to use our product candidates unless coverage is provided and reimbursement is adequate to cover a significant portion of the cost of our product candidates. There is significant uncertainty related to insurance coverage and reimbursement of newly approved products. It is difficult to predict at this time what third-party payors will decide with respect to the coverage and reimbursement for our product candidates.

Payment methodologies may be subject to changes in healthcare legislation and regulatory initiatives. For example, the Middle Class Tax Relief and Job Creation Act of 2012 required that the Centers for Medicare & Medicaid Services, the agency responsible for administering the Medicare program, or CMS, reduce the Medicare clinical laboratory fee schedule by 2% in 2013, which served as a base for 2014 and subsequent years. In addition, effective January 1, 2014, CMS also began bundling the Medicare payments for certain laboratory tests ordered while a patient received services in a hospital outpatient setting. Additional state and federal healthcare reform measures are expected to be adopted in the future, any of which could limit the amounts that federal and state governments will pay for healthcare products and services, which could result in reduced demand for certain pharmaceutical products or additional pricing pressures.

Moreover, increasing efforts by governmental and third-party payors in the United States and abroad to cap or reduce healthcare costs may cause such organizations to limit both coverage and the level of reimbursement for newly approved products and, as a result, they may not cover or provide adequate payment for our product candidates. There has been increasing legislative and enforcement interest in the United States with respect to specialty drug pricing practices. Specifically, there have been several recent U.S. Congressional inquiries and proposed and enacted federal and state legislation designed to, among other things, bring more transparency to drug pricing, reduce the cost of prescription drugs under Medicare, review the relationship between pricing and manufacturer patient programs, and reform government program reimbursement methodologies for drugs. We expect to experience pricing pressures in connection with the sale of any of our product candidates due to the trend toward managed healthcare, the increasing influence of health maintenance organizations, cost containment initiatives and additional legislative changes. At the federal level, the Trump administration's budget proposal for fiscal year 2019 contains further drug price control measures that could be enacted during the 2019 budget process or in other future legislation, including, for example, measures to permit Medicare Part D plans to negotiate the price of certain drugs under Medicare Part B, to allow some states to negotiate drug prices under Medicaid, and to eliminate cost sharing for generic drugs for low-income patients. While any proposed measures will require authorization through additional legislation to become effective, Congress and the Trump administration have each indicated that it will continue to seek new legislative and/or administrative measures to control drug costs. At the state level, legislatures are increasingly passing legislation and implementing regulations designed to control pharmaceutical and biological product pricing, including price or patient reimbursement constraints, discounts, restrictions on certain product access and marketing cost disclosure and transparency measures, and, in some cases, designed to encourage importation from other countries and bulk purchasing.

Ongoing healthcare legislative and regulatory reform measures may have a material adverse effect on our business and results of operations.

Changes in regulations, statutes or the interpretation of existing regulations could impact our business in the future by requiring, for example: (i) changes to our manufacturing arrangements; (ii) additions or modifications to product labeling; (iii) the recall or discontinuation of our products; or (iv) additional record-keeping requirements. If any such changes were to be imposed, they could adversely affect the operation of our business.

In the United States, there have been and continue to be a number of legislative initiatives to contain healthcare costs. For example, in March 2010, the Patient Protection and Affordable Care Act, as amended by the Health Care and Education Reconciliation Act of 2010, or collectively the ACA, was passed, which substantially changes the way health care is financed by both governmental and private insurers, and significantly impacts the U.S. pharmaceutical industry. The ACA, among other things, subjects biological products to potential competition by lower-cost biosimilars, addresses a new methodology by which rebates owed by manufacturers under the Medicaid Drug Rebate Program are calculated for drugs that are inhaled, infused, instilled, implanted or injected, increases the minimum Medicaid rebates owed by manufacturers under the Medicaid Drug Rebate Program and extends the rebate program to individuals enrolled in Medicaid managed care organizations, establishes annual fees and taxes on manufacturers of certain branded prescription drugs, and creates a new Medicare Part D coverage gap discount program, in which manufacturers must agree to offer 50% (and 70% commencing January 1, 2019) point-of-sale discounts off negotiated prices of applicable brand drugs to eligible beneficiaries during their coverage gap period, as a condition for the manufacturer's outpatient drugs to be covered under Medicare Part D.

Some of the provisions of the ACA have yet to be fully implemented, while certain provisions have been subject to judicial and Congressional challenges, as well as efforts by the Trump administration to repeal or replace certain aspects of the ACA. Since January 2017, President Trump has signed two Executive Orders and other directives designed to delay the implementation of certain provisions of the ACA or otherwise circumvent some of the requirements for health insurance mandated by the ACA. For example, the Trump administration has concluded that cost-sharing reduction, or CSR, payments to insurance companies required under the ACA have not received necessary appropriations from Congress and announced that it will discontinue these payments immediately until such appropriations are made. The loss of the CSR payments is expected to increase premiums on certain policies issued by qualified health plans under the ACA. A bipartisan bill to appropriate funds for CSR payments was introduced in the Senate, but the future of that bill is uncertain. Several state Attorneys General have filed lawsuits to stop the administration from terminating the subsidies, but their request for a restraining order was denied by a federal judge in California on October 25, 2017. Concurrently, Congress has considered legislation that would repeal or repeal and replace all or part of the ACA. While Congress has not passed comprehensive repeal legislation, two bills affecting the implementation of certain taxes under the ACA have been signed into law. The TCJA includes a provision repealing, effective January 1, 2019, the tax-based shared responsibility payment imposed by the ACA on certain individuals who fail to maintain qualifying health coverage for all or part of a year that is commonly referred to as the "individual mandate." Additionally, on January 22, 2018, President Trump signed a continuing resolution on appropriations for fiscal year 2018 that delayed the implementation of certain ACA-mandated fees, including the so-called "Cadillac" tax on certain high cost employer-sponsored insurance plans, the annual fee imposed on certain health insurance providers based on market share, and the medical device excise tax on non-exempt medical devices. Further, the Bipartisan Budget Act of 2018, or the BBA, among other things, amends the ACA, effective January 1, 2019, to close the coverage gap in most Medicare drug plans, commonly referred to as the "donut hole." Moreover, CMS has recently proposed regulations that would give states greater flexibility in setting benchmarks for insurers in the individual and small group marketplaces, which may have the effect of relaxing the essential health benefits required under the ACA for plans sold through such marketplaces. Litigation and legislation over the ACA are likely to continue, with unpredictable and uncertain results.

Other legislative changes have been proposed and adopted in the United States since the ACA was enacted. On August 2, 2011, the Budget Control Act of 2011, among other things, created measures for spending reductions by Congress. A Joint Select Committee on Deficit Reduction, tasked with recommending a targeted deficit reduction of at least \$1.2 trillion for the years 2013 through 2027, was unable to reach required goals, thereby triggering the legislation's automatic reduction to several government programs. This includes aggregate reductions of Medicare payments to providers of 2% per fiscal year. These reductions went into effect on April 1, 2013 and, due to subsequent legislative amendments to the statute, including the BBA, will remain in effect through 2027 unless additional congressional action is taken. On January 2, 2013, the American Taxpayer Relief Act of 2012 was signed into law, which, among other things, further reduced Medicare payments to several types of providers.

These laws, and future state and federal healthcare reform measures may be adopted in the future, any of which may result in additional reductions in Medicare and other healthcare funding and otherwise affect the prices we may obtain for any of our product candidates for which we may obtain regulatory approval or the frequency with which any such product candidate is prescribed or used.

EU drug marketing and reimbursement regulations may materially affect our ability to market and receive coverage for our products in the European member states.

We intend to seek approval to market our product candidates in both the United States and in selected foreign jurisdictions. If we obtain approval in one or more foreign jurisdictions for our product candidates, we will be subject to rules and regulations in those jurisdictions. In some foreign countries, particularly those in the EU, the pricing of biologics is subject to governmental control and other market regulations which could put pressure on the pricing and usage of our product candidates. In these countries, pricing negotiations with governmental authorities can take considerable time after obtaining marketing approval of a product candidate. In addition, market acceptance and sales of our product candidates will depend significantly on the availability of adequate coverage and reimbursement from third-party payors for our product candidates and may be affected by existing and future health care reform measures.

Much like the federal Anti-Kickback Statute prohibition in the United States, the provision of benefits or advantages to physicians to induce or encourage the prescription, recommendation, endorsement, purchase, supply, order or use of medicinal products is also prohibited in the EU. The provision of benefits or advantages to physicians is governed by the national anti-bribery laws of EU Member States, such as the UK Bribery Act 2010. Infringement of these laws could result in substantial fines and imprisonment.

Payments made to physicians in certain EU Member States must be publically disclosed. Moreover, agreements with physicians often must be the subject of prior notification and approval by the physician's employer, his or her competent professional organization and/or the regulatory authorities of the individual EU Member States. These requirements are provided in the national laws, industry codes or professional codes of conduct, applicable in the EU Member States. Failure to comply with these requirements could result in reputational risk, public reprimands, administrative penalties, fines or imprisonment.

In addition, in most foreign countries, including the European Economic Area, the proposed pricing for a drug must be approved before it may be lawfully marketed. The requirements governing drug pricing and reimbursement vary widely from country to country. For example, the EU provides options for its member states to restrict the range of medicinal products for which their national health insurance systems provide reimbursement and to control the prices of medicinal products for human use. Reference pricing used by various EU member states and parallel distribution, or arbitrage between low-priced and high-priced member states, can further reduce prices. A member state may approve a specific price for the medicinal product or it may instead adopt a system of direct or indirect controls on the profitability of the company placing the medicinal product on the market. In some countries, we may be required to conduct a clinical study or other studies that compare the cost-effectiveness of any of our product candidates to other available therapies in order to obtain or maintain reimbursement or pricing approval. There can be no

assurance that any country that has price controls or reimbursement limitations for pharmaceutical products will allow favorable reimbursement and pricing arrangements for any of our products. Historically, products launched in the EU do not follow price structures of the United States and generally prices tend to be significantly lower. Publication of discounts by third-party payors or authorities may lead to further pressure on the prices or reimbursement levels within the country of publication and other countries. If pricing is set at unsatisfactory levels or if reimbursement of our products is unavailable or limited in scope or amount, our revenues from sales by us or our strategic partners and the potential profitability of any of our product candidates in those countries would be negatively affected.

European data collection is governed by restrictive regulations governing the use, processing, and cross-border transfer of personal information.

The collection and use of personal health data in the EU is governed by the provisions of the Data Protection Directive, and as of May 2018 the General Data Protection Regulation, or GDPR. These directives impose several requirements relating to the consent of the individuals to whom the personal data relates, the information provided to the individuals, notification of data processing obligations to the competent national data protection authorities and the security and confidentiality of the personal data. The Data Protection Directive and GDPR also impose strict rules on the transfer of personal data out of the EU to the United States. Failure to comply with the requirements of the Data Protection Directive, the GDPR, and the related national data protection laws of the EU Member States may result in fines and other administrative penalties. The GDPR introduces new data protection requirements in the EU and substantial fines for breaches of the data protection rules. The GDPR regulations may impose additional responsibility and liability in relation to personal data that we process and we may be required to put in place additional mechanisms ensuring compliance with the new data protection rules. This may be onerous and adversely affect our business, financial condition, results of operations and prospects.

Additional laws and regulations governing international operations could negatively impact or restrict our operations.

If we further expand our operations outside of the United States, we must dedicate additional resources to comply with numerous laws and regulations in each jurisdiction in which we plan to operate. The FCPA prohibits any U.S. individual or business from paying, offering, authorizing payment or offering of anything of value, directly or indirectly, to any foreign official, political party or candidate for the purpose of influencing any act or decision of the foreign entity in order to assist the individual or business in obtaining or retaining business. The FCPA also obligates companies whose securities are listed in the United States to comply with certain accounting provisions requiring the company to maintain books and records that accurately and fairly reflect all transactions of the corporation, including international subsidiaries, and to devise and maintain an adequate system of internal accounting controls for international operations.

Compliance with the FCPA is expensive and difficult, particularly in countries in which corruption is a recognized problem. In addition, the FCPA presents particular challenges in the pharmaceutical industry, because, in many countries, hospitals are operated by the government, and doctors and other hospital employees are considered foreign officials. Certain payments to hospitals in connection with clinical trials and other work have been deemed to be improper payments to government officials and have led to FCPA enforcement actions.

Various laws, regulations and executive orders also restrict the use and dissemination outside of the United States, or the sharing with certain non-U.S. nationals, of information classified for national security purposes, as well as certain products and technical data relating to those products. If we expand our presence outside of the United States, it will require us to dedicate additional resources to comply with these laws, and these laws may preclude us from developing, manufacturing, or selling certain products and product candidates outside of the United States, which could limit our growth potential and increase our development costs.

The failure to comply with laws governing international business practices may result in substantial civil and criminal penalties and suspension or debarment from government contracting. The Securities and Exchange Commission, or SEC, also may suspend or bar issuers from trading securities on U.S. exchanges for violations of the FCPA's accounting provisions.

We are subject to certain U.S. and foreign anti-corruption, anti-money laundering, export control, sanctions, and other trade laws and regulations. We can face serious consequences for violations.

Among other matters, U.S. and foreign anti-corruption, anti-money laundering, export control, sanctions, and other trade laws and regulations, which are collectively referred to as Trade Laws, prohibit companies and their employees, agents, clinical research organizations, legal counsel, accountants, consultants, contractors, and other partners from authorizing, promising, offering, providing, soliciting, or receiving directly or indirectly, corrupt or improper payments or anything else of value to or from recipients in the public or private sector. Violations of Trade Laws can result in substantial criminal fines and civil penalties, imprisonment, the loss of trade privileges, debarment, tax reassessments, breach of contract and fraud litigation, reputational harm, and other consequences. We have direct or indirect interactions with officials and employees of government agencies or government-affiliated hospitals, universities, and other organizations. We also expect our non-U.S. activities to increase in time. We plan to engage third parties for clinical trials and/or to obtain necessary permits, licenses, patent registrations, and other regulatory approvals and we can be held liable for the corrupt or other illegal activities of our personnel, agents, or partners, even if we do not explicitly authorize or have prior knowledge of such activities.

Risks Related to Our Intellectual Property

Our success depends in part on our ability to protect our intellectual property. It is difficult and costly to protect our proprietary rights and technology, and we may not be able to ensure their protection.

Our commercial success will depend in large part on obtaining and maintaining patent, trademark and trade secret protection of our proprietary technologies and our product candidates, their respective components, formulations, combination therapies, methods used to manufacture them and methods of treatment, as well as successfully defending these patents against third-party challenges. Our ability to stop unauthorized third parties from making, using, selling, offering to sell or importing our product candidates is dependent upon the extent to which we have rights under valid and enforceable patents that cover these activities. If we are unable to secure and maintain patent protection for any product or technology we develop, or if the scope of the patent protection secured is not sufficiently broad, our competitors could develop and commercialize products and technology similar or identical to ours, and our ability to commercialize any product candidates we may develop may be adversely affected.

The patenting process is expensive and time-consuming, and we may not be able to file and prosecute all necessary or desirable patent applications at a reasonable cost or in a timely manner. In addition, we may not pursue or obtain patent protection in all relevant markets. It is also possible that we will fail to identify patentable aspects of our research and development output before it is too late to obtain patent protection. Moreover, in some circumstances, we may not have the right to control the preparation, filing and prosecution of patent applications, or to maintain the patents, covering technology that we license from or license to third parties and are reliant on our licensors or licensees.

The strength of patents in the biotechnology and pharmaceutical field involves complex legal and scientific questions and can be uncertain. The patent applications that we own or in-license may fail to result in issued patents with claims that cover our product candidates or uses thereof in the United States or in other foreign countries. Even if the patents do successfully issue, third parties may challenge the validity, enforceability or scope thereof, which may result in such patents being narrowed, invalidated or held unenforceable. Furthermore, even if they are unchallenged, our patents and patent applications may not adequately protect our intellectual property or prevent others from designing around our claims. If the breadth or strength of protection provided by the patent applications we hold with respect to our product

candidates is threatened, it could dissuade companies from collaborating with us to develop, and threaten our ability to commercialize, our product candidates. Further, if we encounter delays in our clinical trials, the period of time during which we could market our product candidates under patent protection would be reduced.

Since patent applications in the United States and most other countries are confidential for a period of time after filing, we cannot be certain that we were the first to file any patent application related to our product candidates. Furthermore, for United States applications in which all claims are entitled to a priority date before March 16, 2013, an interference proceeding can be provoked by a third-party or instituted by the United States patent office, or USPTO, to determine who was the first to invent any of the subject matter covered by the patent claims of our applications.

We cannot be certain that we are the first to invent the inventions covered by pending patent applications and, if we are not, we may be subject to priority disputes. We may be required to disclaim part or all of the term of certain patents or all of the term of certain patent applications. There may be prior art of which we are not aware that may affect the validity or enforceability of a patent claim. There also may be prior art of which we are aware, but which we do not believe affects the validity or enforceability of a claim, which may, nonetheless, ultimately be found to affect the validity or enforceability of a claim. No assurance can be given that if challenged, our patents would be declared by a court to be valid or enforceable or that even if found valid and enforceable, a competitor's technology or product would be found by a court to infringe our patents. We may analyze patents or patent applications of our competitors that we believe are relevant to our activities, and consider that we are free to operate in relation to our product candidates, but our competitors may achieve issued claims, including in patents we consider to be unrelated, which block our efforts or may potentially result in our product candidates or our activities infringing such claims. The possibility exists that others will develop products which have the same effect as our products on an independent basis which do not infringe our patents or other intellectual property rights, or will design around the claims of patents that we have had issued that cover our products.

Recent or future patent reform legislation could increase the uncertainties and costs surrounding the prosecution of our patent applications and the enforcement or defense of our issued patents. Under the enacted Leahy-Smith America Invents Act, or America Invents Act, enacted in 2013, the United States moved from a "first to invent" to a "first-to-file" system. Under a "first-to-file" system, assuming the other requirements for patentability are met, the first inventor to file a patent application generally will be entitled to a patent on the invention regardless of whether another inventor had made the invention earlier. The America Invents Act includes a number of other significant changes to U.S. patent law, including provisions that affect the way patent applications are prosecuted, redefine prior art and establish a new post-grant review system. The effects of these changes are currently unclear as the USPTO only recently developed new regulations and procedures in connection with the America Invents Act and many of the substantive changes to patent law, including the "first-to-file" provisions, only became effective in March 2013. In addition, the courts have yet to address many of these provisions and the applicability of the act and new regulations on specific patents discussed herein have not been determined and would need to be reviewed. However, the America Invents Act and its implementation could increase the uncertainties and costs surrounding the prosecution of our patent applications and the enforcement or defense of our issued patents, all of which could have a material adverse effect on our business and financial condition.

The degree of future protection for our proprietary rights is uncertain because legal means afford only limited protection and may not adequately protect our rights or permit us to gain or keep our competitive advantage. For example:

§ others may be able to make or use compounds or cells that are similar to the biological compositions of our product candidates but that are not covered by the claims of our patents;

- § the active biological ingredients in our current product candidates will eventually become commercially available in biosimilar drug products, and no patent protection may be available with regard to formulation or method of use;
- § we or our licensors, as the case may be, may fail to meet our obligations to the U.S. government in regards to any in-licensed patents and patent applications funded by U.S. government grants, leading to the loss of patent rights;
- § we or our licensors, as the case may be, might not have been the first to file patent applications for these inventions;
- § others may independently develop similar or alternative technologies or duplicate any of our technologies;
- § it is possible that our pending patent applications will not result in issued patents;
- § it is possible that there are prior public disclosures that could invalidate our or our licensors' patents, as the case may be, or parts of our or their patents;
- § it is possible that others may circumvent our owned or in-licensed patents;
- § it is possible that there are unpublished applications or patent applications maintained in secrecy that may later issue with claims covering our products or technology similar to ours;
- § the laws of foreign countries may not protect our or our licensors', as the case may be, proprietary rights to the same extent as the laws of the United States;
- § the claims of our owned or in-licensed issued patents or patent applications, if and when issued, may not cover our product candidates;
- § our owned or in-licensed issued patents may not provide us with any competitive advantages, may be narrowed in scope, or be held invalid or unenforceable as a result of legal challenges by third parties;
- § the inventors of our owned or in-licensed patents or patent applications may become involved with competitors, develop products or processes which design around our patents, or become hostile to us or the patents or patent applications on which they are named as inventors;
- § it is possible that our owned or in-licensed patents or patent applications omit individual(s) that should be listed as inventor(s) or include individual(s) that should not be listed as inventor(s), which may cause these patents or patents issuing from these patent applications to be held invalid or unenforceable;
- § we have engaged in scientific collaborations in the past, and will continue to do so in the future. Such collaborators may develop adjacent or competing products to ours that are outside the scope of our patents;
- § we may not develop additional proprietary technologies for which we can obtain patent protection;
- § it is possible that product candidates or diagnostic tests we develop may be covered by third parties' patents or other exclusive rights; or
- § the patents of others may have an adverse effect on our business.

We depend on intellectual property licensed from third parties and termination of any of these licenses could result in the loss of significant rights, which would harm our business.

We are dependent on patents, know-how and proprietary technology, both our own and licensed from others. Any termination of these licenses could result in the loss of significant rights and could harm our ability to commercialize our product candidates. See "Business — License Agreements" for additional information regarding our license agreements.

Disputes may also arise between us and our licensors regarding intellectual property subject to a license agreement, including:

- § the scope of rights granted under the license agreement and other interpretation-related issues;

- § whether and the extent to which our technology and processes infringe on intellectual property of the licensor that is not subject to the licensing agreement;
- § our right to sublicense patent and other rights to third parties under collaborative development relationships;
- § our diligence obligations with respect to the use of the licensed technology in relation to our development and commercialization of our product candidates, and what activities satisfy those diligence obligations; and
- § the ownership of inventions and know-how resulting from the joint creation or use of intellectual property by our licensors and us and our partners.

If disputes over intellectual property that we have licensed prevent or impair our ability to maintain our current licensing arrangements on acceptable terms, we may be unable to successfully develop and commercialize the affected product candidates.

We are generally also subject to all of the same risks with respect to protection of intellectual property that we license, as we are for intellectual property that we own, which are described below. If we or our licensors fail to adequately protect this intellectual property, our ability to commercialize products could suffer.

If we fail to comply with our obligations under our patent licenses with third parties, we could lose license rights that are important to our business.

We are a party to license agreements pursuant to which we in-license key patent and patent applications for our product candidates. These existing licenses impose various diligence, milestone payment, royalty, insurance and other obligations on us. If we fail to comply with these obligations, our licensors may have the right to terminate the license, in which event we would not be able to develop or market the products covered by such licensed intellectual property.

We may have limited control over the maintenance and prosecution of these in-licensed patents and patent applications, activities or any other intellectual property that may be related to our in-licensed intellectual property. For example, we cannot be certain that such activities by these licensors have been or will be conducted in compliance with applicable laws and regulations or will result in valid and enforceable patents and other intellectual property rights. We have limited control over the manner in which our licensors initiate an infringement proceeding against a third-party infringer of the intellectual property rights, or defend certain of the intellectual property that is licensed to us. It is possible that the licensors' infringement proceeding or defense activities may be less vigorous than had we conducted them ourselves.

If we are unable to protect the confidentiality of our trade secrets, our business and competitive position would be harmed.

In addition to patent protection, we rely heavily upon know-how and trade secret protection, as well as non-disclosure agreements and invention assignment agreements with our employees, consultants and third-parties, to protect our confidential and proprietary information, especially where we do not believe patent protection is appropriate or obtainable. In addition to contractual measures, we try to protect the confidential nature of our proprietary information using physical and technological security measures. Such measures may not, for example, in the case of misappropriation of a trade secret by an employee or third-party with authorized access, provide adequate protection for our proprietary information. Our security measures may not prevent an employee or consultant from misappropriating our trade secrets and providing them to a competitor, and recourse we take against such misconduct may not provide an adequate remedy to protect our interests fully. Enforcing a claim that a party illegally disclosed or misappropriated a trade secret can be difficult, expensive, and time-consuming, and the outcome is unpredictable. In addition, trade secrets may be independently developed by others in a manner that could prevent legal recourse by us. If any of our confidential or proprietary information, such as our trade secrets, were to be disclosed or

misappropriated, or if any such information was independently developed by a competitor, our competitive position could be harmed.

In addition, courts outside the United States are sometimes less willing to protect trade secrets. If we choose to go to court to stop a third-party from using any of our trade secrets, we may incur substantial costs. These lawsuits may consume our time and other resources even if we are successful. Although we take steps to protect our proprietary information and trade secrets, including through contractual means with our employees and consultants, third parties may independently develop substantially equivalent proprietary information and techniques or otherwise gain access to our trade secrets or disclose our technology.

Thus, we may not be able to meaningfully protect our trade secrets. It is our policy to require our employees, consultants, outside scientific collaborators, sponsored researchers and other advisors to execute confidentiality agreements upon the commencement of employment or consulting relationships with us. These agreements provide that all confidential information concerning our business or financial affairs developed or made known to the individual or entity during the course of the party's relationship with us is to be kept confidential and not disclosed to third parties except in specific circumstances. In the case of employees, the agreements provide that all inventions conceived by the individual, and which are related to our current or planned business or research and development or made during normal working hours, on our premises or using our equipment or proprietary information, are our exclusive property. In addition, we take other appropriate precautions, such as physical and technological security measures, to guard against misappropriation of our proprietary technology by third parties. We have also adopted policies and conduct training that provides guidance on our expectations, and our advice for best practices, in protecting our trade secrets.

Third-party claims of intellectual property infringement may prevent or delay our product discovery and development efforts.

Our commercial success depends in part on our ability to develop, manufacture, market and sell our product candidates and use our proprietary technologies without infringing the proprietary rights of third parties. There is a substantial amount of litigation involving patents and other intellectual property rights in the biotechnology and pharmaceutical industries, as well as administrative proceedings for challenging patents, including interference, derivation, *inter partes* review, post grant review, and reexamination proceedings before the USPTO or oppositions and other comparable proceedings in foreign jurisdictions. We may be exposed to, or threatened with, future litigation by third parties having patent or other intellectual property rights alleging that our product candidates and/or proprietary technologies infringe their intellectual property rights. Numerous U.S. and foreign issued patents and pending patent applications, which are owned by third parties, exist in the fields in which we are developing our product candidates. As the biotechnology and pharmaceutical industries expand and more patents are issued, the risk increases that our product candidates may give rise to claims of infringement of the patent rights of others. Moreover, it is not always clear to industry participants, including us, which patents cover various types of drugs, products or their methods of use or manufacture. Thus, because of the large number of patents issued and patent applications filed in our fields, there may be a risk that third parties may allege they have patent rights encompassing our product candidates, technologies or methods.

If a third-party claims that we infringe its intellectual property rights, we may face a number of issues, including, but not limited to:

- § infringement and other intellectual property claims which, regardless of merit, may be expensive and time-consuming to litigate and may divert our management's attention from our core business;
- § substantial damages for infringement, which we may have to pay if a court decides that the product candidate or technology at issue infringes on or violates the third-party's rights, and, if the court finds

that the infringement was willful, we could be ordered to pay treble damages and the patent owner's attorneys' fees;

- § a court prohibiting us from developing, manufacturing, marketing or selling our product candidates, or from using our proprietary technologies, unless the third-party licenses its product rights to us, which it is not required to do;
- § if a license is available from a third-party, we may have to pay substantial royalties, upfront fees and other amounts, and/or grant cross-licenses to intellectual property rights for our products; and
- § redesigning our product candidates or processes so they do not infringe, which may not be possible or may require substantial monetary expenditures and time.

Some of our competitors may be able to sustain the costs of complex patent litigation more effectively than we can because they have substantially greater resources. In addition, any uncertainties resulting from the initiation and continuation of any litigation could have a material adverse effect on our ability to raise the funds necessary to continue our operations or could otherwise have a material adverse effect on our business, results of operations, financial condition and prospects.

Third parties may assert that we are employing their proprietary technology without authorization. Generally, conducting clinical trials and other development activities in the United States is protected under the Safe Harbor exemption as set forth in 35 U.S.C. § 271. If and when SRK-015 or another one of our product candidates is approved by the FDA, that certain third-party may then seek to enforce its patent by filing a patent infringement lawsuit against us. While we do not believe that any claims of such patent that could otherwise materially adversely affect commercialization of our antibody candidates, if approved, are valid and enforceable, we may be incorrect in this belief, or we may not be able to prove it in a litigation. In this regard, patents issued in the U.S. by law enjoy a presumption of validity that can be rebutted only with evidence that is "clear and convincing," a heightened standard of proof. There may be third-party patents of which we are currently unaware with claims to materials, formulations, methods of manufacture or methods for treatment related to the use or manufacture of our product candidates. Because patent applications can take many years to issue, there may be currently pending patent applications which may later result in issued patents that our product candidates may infringe. In addition, third parties may obtain patents in the future and claim that use of our technologies infringes upon these patents. If any third-party patents were held by a court of competent jurisdiction to cover the manufacturing process of our product candidates, constructs or molecules used in or formed during the manufacturing process, or any final product itself, the holders of any such patents may be able to block our ability to commercialize the product candidate unless we obtained a license under the applicable patents, or until such patents expire or they are finally determined to be held invalid or unenforceable. Similarly, if any third-party patent were held by a court of competent jurisdiction to cover aspects of our formulations, processes for manufacture or methods of use, the holders of any such patent may be able to block our ability to develop and commercialize the product candidate unless we obtained a license or until such patent expires or is finally determined to be held invalid or unenforceable. In either case, such a license may not be available on commercially reasonable terms or at all. If we are unable to obtain a necessary license to a third-party patent on commercially reasonable terms, or at all, our ability to commercialize our product candidates may be impaired or delayed, which could in turn significantly harm our business. Even if we obtain a license, it may be non-exclusive, thereby giving our competitors access to the same technologies licensed to us. In addition, if the breadth or strength of protection provided by our patents and patent applications is threatened, it could dissuade companies from collaborating with us to license, develop or commercialize current or future product candidates.

Parties making claims against us may seek and obtain injunctive or other equitable relief, which could effectively block our ability to further develop and commercialize our product candidates. Defense of these claims, regardless of their merit, would involve substantial litigation expense and would be a substantial diversion of employee resources from our business. In the event of a successful claim of infringement against us, we may have to pay substantial damages, including treble damages and attorneys' fees for

willful infringement, obtain one or more licenses from third parties, pay royalties or redesign our infringing products, which may be impossible or require substantial time and monetary expenditure. We cannot predict whether any such license would be available at all or whether it would be available on commercially reasonable terms. Furthermore, even in the absence of litigation, we may need to obtain licenses from third parties to advance our research or allow commercialization of our product candidates. We may fail to obtain any of these licenses at a reasonable cost or on reasonable terms, if at all. In that event, we would be unable to further develop and commercialize our product candidates, which could harm our business significantly.

Third parties may assert that our employees or consultants have wrongfully used or disclosed confidential information or misappropriated trade secrets.

As is common in the biotechnology and pharmaceutical industries, we employ individuals who were previously employed at universities or other biopharmaceutical or pharmaceutical companies, including our competitors or potential competitors. Although no claims against us are currently pending, and although we try to ensure that our employees and consultants do not use the proprietary information or know-how of others in their work for us, we may be subject to claims that we or our employees, consultants or independent contractors have inadvertently or otherwise used or disclosed intellectual property, including trade secrets or other proprietary information, of a former employer or other third parties. Litigation may be necessary to defend against these claims. If we fail in defending any such claims, in addition to paying monetary damages, we may lose valuable intellectual property rights or personnel. Even if we are successful in defending against such claims, litigation or other legal proceedings relating to intellectual property claims may cause us to incur significant expenses, and could distract our technical and management personnel from their normal responsibilities. In addition, there could be public announcements of the results of hearings, motions or other interim proceedings or developments, and, if securities analysts or investors perceive these results to be negative, it could have a substantial adverse effect on the price of our common stock. This type of litigation or proceeding could substantially increase our operating losses and reduce our resources available for development activities. We may not have sufficient financial or other resources to adequately conduct such litigation or proceedings. Some of our competitors may be able to sustain the costs of such litigation or proceedings more effectively than we can because of their substantially greater financial resources. Uncertainties resulting from the initiation and continuation of patent litigation or other intellectual property related proceedings could adversely affect our ability to compete in the marketplace.

We may not be successful in obtaining or maintaining necessary rights to develop any future product candidates on acceptable terms.

Because our programs may involve additional product candidates that may require the use of proprietary rights held by third parties, the growth of our business may depend in part on our ability to acquire, in-license or use these proprietary rights.

Our product candidates may also require specific formulations to work effectively and efficiently and these rights may be held by others. We may develop products containing our compounds and pre-existing pharmaceutical compounds. We may be required by the FDA or comparable foreign regulatory authorities to provide a companion diagnostic test or tests with our product candidates. These diagnostic test or tests may be covered by intellectual property rights held by others. We may be unable to acquire or in-license any compositions, methods of use, processes or other third-party intellectual property rights from third parties that we identify as necessary or important to our business operations. We may fail to obtain any of these licenses at a reasonable cost or on reasonable terms, if at all, which would harm our business. We may need to cease use of the compositions or methods covered by such third-party intellectual property rights, and may need to seek to develop alternative approaches that do not infringe on such intellectual property rights which may entail additional costs and development delays, even if we were able to develop such alternatives, which may not be feasible. Even if we are able to obtain a license, it may be non-exclusive,

thereby giving our competitors access to the same technologies licensed to us. In that event, we may be required to expend significant time and resources to develop or license replacement technology.

Additionally, we sometimes collaborate with academic institutions to accelerate our preclinical research or development under written agreements with these institutions. In certain cases, these institutions provide us with an option to negotiate a license to any of the institution's rights in technology resulting from the collaboration. Regardless of such option, we may be unable to negotiate a license within the specified timeframe or under terms that are acceptable to us. If we are unable to do so, the institution may offer the intellectual property rights to others, potentially blocking our ability to pursue our program. If we are unable to successfully obtain rights to required third-party intellectual property or to maintain the existing intellectual property rights we have, we may have to abandon development of such program and our business and financial condition could suffer.

The licensing and acquisition of third-party intellectual property rights is a competitive area, and companies, which may be more established, or have greater resources than we do, may also be pursuing strategies to license or acquire third-party intellectual property rights that we may consider necessary or attractive in order to commercialize our product candidates. More established companies may have a competitive advantage over us due to their size, cash resources and greater clinical development and commercialization capabilities. There can be no assurance that we will be able to successfully complete such negotiations and ultimately acquire the rights to the intellectual property surrounding the additional product candidates that we may seek to acquire.

We may be involved in lawsuits to protect or enforce our patents or the patents of our licensors, which could be expensive, time-consuming and unsuccessful.

Competitors may infringe our patents or the patents of our licensors. To counter infringement or unauthorized use, we may be required to file infringement claims, which can be expensive and time-consuming. In addition, in an infringement proceeding, a court may decide that one or more of our patents is not valid or is unenforceable, or may refuse to stop the other party from using the technology at issue on the grounds that our patents do not cover the technology in question. An adverse result in any litigation or defense proceedings could put one or more of our patents at risk of being invalidated, held unenforceable, or interpreted narrowly and could put our patent applications at risk of not issuing. Defense of these claims, regardless of their merit, would involve substantial litigation expense and would be a substantial diversion of employee resources from our business.

We may choose to challenge the patentability of claims in a third-party's U.S. patent by requesting that the USPTO review the patent claims in an *ex-parte* re-exam, *inter partes* review or post-grant review proceedings. These proceedings are expensive and may consume our time or other resources. We may choose to challenge a third-party's patent in patent opposition proceedings in the European Patent Office, or EPO, or other foreign patent office. The costs of these opposition proceedings could be substantial, and may consume our time or other resources. If we fail to obtain a favorable result at the USPTO, EPO or other patent office then we may be exposed to litigation by a third-party alleging that the patent may be infringed by our product candidates or proprietary technologies.

In addition, because some patent applications in the United States may be maintained in secrecy until the patents are issued, patent applications in the United States and many foreign jurisdictions are typically not published until 18 months after filing, and publications in the scientific literature often lag behind actual discoveries, we cannot be certain that others have not filed patent applications for technology covered by our owned and in-licensed issued patents or our pending applications, or that we or, if applicable, a licensor were the first to invent the technology. Our competitors may have filed, and may in the future file, patent applications covering our products or technology similar to ours. Any such patent application may have priority over our owned and in-licensed patent applications or patents, which could require us to obtain rights to issued patents covering such technologies. If another party has filed a U.S. patent application on

inventions similar to those owned by or in-licensed to us, we or, in the case of in-licensed technology, the licensor may have to participate in an interference proceeding declared by the USPTO to determine priority of invention in the United States. If we or one of our licensors is a party to an interference proceeding involving a U.S. patent application on inventions owned by or in-licensed to us, we may incur substantial costs, divert management's time and expend other resources, even if we are successful.

Interference proceedings provoked by third parties or brought by the USPTO may be necessary to determine the priority of inventions with respect to our patents or patent applications or those of our licensors. An unfavorable outcome could result in a loss of our current patent rights and could require us to cease using the related technology or to attempt to license rights to it from the prevailing party. Our business could be harmed if the prevailing party does not offer us a license on commercially reasonable terms. Litigation or interference proceedings may result in a decision adverse to our interests and, even if we are successful, may result in substantial costs and distract our management and other employees. We may not be able to prevent, alone or with our licensors, misappropriation of our trade secrets or confidential information, particularly in countries where the laws may not protect those rights as fully as in the United States.

Furthermore, because of the substantial amount of discovery required in connection with intellectual property litigation, there is a risk that some of our confidential information could be compromised by disclosure during this type of litigation. In addition, there could be public announcements of the results of hearings, motions or other interim proceedings or developments. If securities analysts or investors perceive these results to be negative, it could have a substantial adverse effect on the price of our common stock.

Obtaining and maintaining our patent protection depends on compliance with various procedural, document submission, fee payment and other requirements imposed by governmental patent agencies, and our patent protection could be reduced or eliminated for non-compliance with these requirements.

Periodic maintenance fees on any issued patent are due to be paid to the USPTO and foreign patent agencies in several stages over the lifetime of the patent. The USPTO and various foreign governmental patent agencies require compliance with a number of procedural, documentary, fee payment and other provisions during the patent application process and following the issuance of a patent. While an inadvertent lapse can in many cases be cured by payment of a late fee or by other means in accordance with the applicable rules, there are situations in which noncompliance can result in abandonment or lapse of the patent or patent application, resulting in partial or complete loss of patent rights in the relevant jurisdiction. Noncompliance events that could result in abandonment or lapse of a patent or patent application include, but are not limited to, failure to respond to official actions within prescribed time limits, non-payment of fees and failure to properly legalize and submit formal documents. In such an event, our competitors might be able to enter the market, which would have a material adverse effect on our business.

Issued patents covering our product candidates could be found invalid or unenforceable if challenged in court or the USPTO.

If we or one of our licensing partners initiate legal proceedings against a third-party to enforce a patent covering one of our product candidates, the defendant could counterclaim that the patent covering our product candidate, as applicable, is invalid and/or unenforceable. In patent litigation in the United States, defendant counterclaims alleging invalidity and/or unenforceability are commonplace, and there are numerous grounds upon which a third-party can assert invalidity or unenforceability of a patent. Third parties may also raise similar claims before administrative bodies in the United States or abroad, even outside the context of litigation. Such mechanisms include re-examination, post grant review, and equivalent proceedings in foreign jurisdictions (e.g., opposition proceedings). Such proceedings could result in revocation or amendment to our patents in such a way that they no longer cover our product candidates. The outcome following legal assertions of invalidity and unenforceability is unpredictable. With respect to the validity question, for example, we cannot be certain that there is no invalidating prior art, of which we,

our patent counsel and the patent examiner were unaware during prosecution. If a defendant were to prevail on a legal assertion of invalidity and/or unenforceability, or if we are otherwise unable to adequately protect our rights, we would lose at least part, and perhaps all, of the patent protection on our product candidates. Such a loss of patent protection could have a material adverse impact on our business and our ability to commercialize or license our technology and product candidates.

Likewise, our current owned patents covering our proprietary technologies and our product candidates are expected to expire in 2034, without taking into account any possible patent term adjustments or extensions. Our earliest patents may expire before, or soon after, our first product achieves marketing approval in the United States or foreign jurisdictions. Upon the expiration of our current patents, we may lose the right to exclude others from practicing these inventions. The expiration of these patents could also have a similar material adverse effect on our business, results of operations, financial condition and prospects. We own pending patent applications covering our proprietary technologies or our product candidates that if issued as patents are expected to expire from 2033 through 2038, without taking into account any possible patent term adjustments or extensions. However, we cannot be assured that the USPTO or relevant foreign patent offices will grant any of these patent applications.

Changes in patent law in the U.S. and in ex-U.S. jurisdictions could diminish the value of patents in general, thereby impairing our ability to protect our products.

As is the case with other biopharmaceutical companies, our success is heavily dependent on intellectual property, particularly patents. Obtaining and enforcing patents in the biopharmaceutical industry involve both technological and legal complexity, and is therefore costly, time-consuming and inherently uncertain. In addition, the United States has recently enacted and is currently implementing wide-ranging patent reform legislation. Recent U.S. Supreme Court rulings have narrowed the scope of patent protection available in certain circumstances and weakened the rights of patent owners in certain situations. In addition to increasing uncertainty with regard to our ability to obtain patents in the future, this combination of events has created uncertainty with respect to the value of patents, once obtained. Depending on decisions by the U.S. Congress, the federal courts, and the USPTO, the laws and regulations governing patents could change in unpredictable ways that would weaken our ability to obtain new patents or to enforce our existing patents and patents that we might obtain in the future. For example, in the case *Amgen Inc. v. Sanofi*, the Federal Circuit held that a well characterized antigen is insufficient to satisfy the written description requirement of certain claims directed to a genus of antibodies that are solely defined by function; and in the case of *Assoc. for Molecular Pathology v. Myriad Genetics, Inc.*, the U.S. Supreme Court held that certain claims to DNA molecules are not patentable. We cannot predict how these decisions or any future decisions by the courts, the U.S. Congress or the USPTO may impact the value of our patents. Similarly, any adverse changes in the patent laws of other jurisdictions could have a material adverse effect on our business and financial condition.

We have limited foreign intellectual property rights and may not be able to protect our intellectual property rights throughout the world.

We have limited intellectual property rights outside the United States. Filing, prosecuting and defending patents on product candidates in all countries throughout the world would be prohibitively expensive, and our intellectual property rights in some countries outside the United States can be less extensive than those in the United States. In addition, the laws of some foreign countries do not protect intellectual property rights to the same extent as federal and state laws in the United States. Consequently, we may not be able to prevent third parties from practicing our inventions in all countries outside the United States, or from selling or importing products made using our inventions in and into the United States or other jurisdictions. Competitors may use our technologies in jurisdictions where we have not obtained patent protection to develop their own products and, further, may export otherwise infringing products to territories where we have patent protection but where enforcement is not as strong as that in the United States. These products may compete with our products in jurisdictions where we do not have any issued patents and our patent claims or other intellectual property rights may not be effective or sufficient to prevent them from competing.

Many companies have encountered significant problems in protecting and defending intellectual property rights in foreign jurisdictions. The legal systems of certain countries, particularly certain developing countries, do not favor the enforcement of patents, trade secrets and other intellectual property protection, particularly those relating to biopharmaceutical products, which could make it difficult for us to stop the infringement of our patents or marketing of competing products against third parties in violation of our proprietary rights generally. The initiation of proceedings by third parties to challenge the scope or validity of our patent rights in foreign jurisdictions could result in substantial cost and divert our efforts and attention from other aspects of our business. Proceedings to enforce our patent rights in foreign jurisdictions could result in substantial costs and divert our efforts and attention from other aspects of our business, could put our patents at risk of being invalidated or interpreted narrowly and our patent applications at risk of not issuing and could provoke third parties to assert claims against us. We may not prevail in any lawsuits that we initiate and the damages or other remedies awarded, if any, may not be commercially meaningful. Accordingly, our efforts to enforce our intellectual property rights around the world may be inadequate to obtain a significant commercial advantage from the intellectual property that we develop or license.

We may incur substantial costs as a result of litigation or other proceedings relating to patents, and we may be unable to protect our rights to our products and technology.

If we or our licensors choose to go to court to stop a third-party from using the inventions claimed in our owned or in-licensed patents, that third-party may ask the court to rule that the patents are invalid and/or should not be enforced against that third-party. These lawsuits are expensive and would consume time and other resources even if we or they, as the case may be, were successful in stopping the infringement of these patents. In addition, there is a risk that the court will decide that these patents are not valid and that we or they, as the case may be, do not have the right to stop others from using the inventions.

There is also the risk that, even if the validity of these patents is upheld, the court will refuse to stop the third-party on the ground that such third-party's activities do not infringe our owned or in-licensed patents. In addition, the U.S. Supreme Court has recently changed some legal principles that affect patent applications, granted patents and assessment of the eligibility or validity of these patents. As a consequence, issued patents may be found to contain invalid claims according to the newly revised eligibility and validity standards. Some of our owned or in-licensed patents may be subject to challenge and subsequent invalidation or significant narrowing of claim scope in proceedings before the USPTO, or during litigation, under the revised criteria which could also make it more difficult to obtain patents.

We, or our licensors, may not be able to detect infringement against our owned or in-licensed patents, as the case may be, which may be especially difficult for manufacturing processes or formulation patents. Even if we or our licensors detect infringement by a third-party of our owned or in-licensed patents, we or our licensors, as the case may be, may choose not to pursue litigation against or settlement with the third-party. If we, or our licensors, later sue such third-party for patent infringement, the third-party may have certain legal defenses available to it, which otherwise would not be available except for the delay between when the infringement was first detected and when the suit was brought. Such legal defenses may make it impossible for us or our licensors to enforce our owned or in-licensed patents, as the case may be, against such third-party.

If another party questions the patentability of any of our claims in our owned or in-licensed U.S. patents, the third-party can request that the USPTO review the patent claims such as in an *inter partes* review, *ex parte* re-exam or post-grant review proceedings. These proceedings are expensive and may result in a loss of scope of some claims or a loss of the entire patent. In addition to potential USPTO review proceedings, we may become a party to patent opposition proceedings at the EPO or similar proceedings in other foreign patent offices, where either our owned or in-licensed foreign patents are challenged. One of our in-licensed European patents is involved in a multi-party European opposition proceeding at the EPO. While we believe that the granted claims will ultimately be found to be valid, there is a risk that one or more of the grounds

raised by the opponents will invalidate one or more of the granted claims. This may prevent us from asserting this patent against our competitors marketing otherwise infringing products in relevant European countries where this patent has been granted.

In the future, we may be involved in similar proceedings challenging the patent rights of others, and the outcome of such proceedings is highly uncertain. An adverse determination in any such proceeding could reduce the scope of, or invalidate, our patent rights, allow third parties to commercialize our technology or products and compete directly with us, without payment to us, or result in our inability to manufacture or commercialize products without infringing third-party patent rights. The costs of these opposition or similar proceedings could be substantial, and may result in a loss of scope of some claims or a loss of the entire patent. An unfavorable result at the USPTO, EPO or other patent office may result in the loss of our right to exclude others from practicing one or more of our inventions in the relevant country or jurisdiction, which could have a material adverse effect on our business.

Patent terms may be inadequate to protect our competitive position on our product candidates for an adequate amount of time.

Patents have a limited lifespan. In the United States, if all maintenance fees are timely paid, the natural expiration of a patent is generally 20 years from its earliest U.S. non-provisional filing date. Various extensions such as patent term adjustments and/or extensions, may be available, but the life of a patent, and the protection it affords, is limited. Even if patents covering our product candidates are obtained, once the patent life has expired, we may be open to competition from competitive products, including biosimilars. Given the amount of time required for the development, testing and regulatory review of new product candidates, patents protecting such candidates might expire before or shortly after such candidates are commercialized. As a result, our owned and licensed patent portfolio may not provide us with sufficient rights to exclude others from commercializing products similar or identical to ours.

If we do not obtain patent term extension and data exclusivity for any product candidates we may develop, our business may be materially harmed.

Depending upon the timing, duration and specifics of any FDA marketing approval of any product candidates we may develop, one or more of our U.S. patents may be eligible for limited patent term extension under the Drug Price Competition and Patent Term Restoration Action of 1984 Hatch-Waxman Amendments. The Hatch-Waxman Amendments permit a patent extension term of up to five years as compensation for patent term lost during the FDA regulatory review process. A patent term extension cannot extend the remaining term of a patent beyond a total of 14 years from the date of product approval, only one patent may be extended and only those claims covering the approved drug, a method for using it, or a method for manufacturing it may be extended. However, we may not be granted an extension because of, for example, failing to exercise due diligence during the testing phase or regulatory review process, failing to apply within applicable deadlines, failing to apply prior to expiration of relevant patents, or otherwise failing to satisfy applicable requirements. Moreover, the applicable time period or the scope of patent protection afforded could be less than we request. If we are unable to obtain patent term extension or term of any such extension is less than we request, our competitors may obtain approval of competing products following our patent expiration, and our business, financial condition, results of operations, and prospects could be materially harmed.

If our trademarks and trade names are not adequately protected, then we may not be able to build name recognition in our markets of interest and our business may be adversely affected.

Our trademarks or trade names may be challenged, infringed, circumvented or declared generic or determined to be infringing on other marks. We may not be able to protect our rights to these trademarks and trade names or may be forced to stop using these names, which we need for name recognition by potential partners or customers in our markets of interest. If we are unable to establish name recognition

based on our trademarks and trade names, we may not be able to compete effectively and our business may be adversely affected.

Risks Related to Our Reliance On Third Parties

We rely on third parties to conduct certain aspects of our preclinical studies and will rely on third parties to conduct our clinical trials. If these third parties do not successfully carry out their contractual duties or meet expected deadlines or comply with regulatory requirements, we may not be able to obtain regulatory approval of or commercialize any potential product candidates.

We depend upon third parties to conduct certain aspects of our preclinical studies and will depend on third parties, including independent investigators, to conduct our clinical trials, under agreements with universities, medical institutions, CROs, strategic partners and others. We expect to have to negotiate budgets and contracts with such third parties, which may result in delays to our development timelines and increased costs.

We will rely especially heavily on third parties over the course of our clinical trials, and, as a result, will have limited control over the clinical investigators and limited visibility into their day-to-day activities, including with respect to their compliance with the approved clinical protocol. Nevertheless, we are responsible for ensuring that each of our trials is conducted in accordance with the applicable protocol, legal and regulatory requirements and scientific standards, and our reliance on third parties does not relieve us of our regulatory responsibilities. We and these third parties are required to comply with GCP requirements, which are regulations and guidelines enforced by the FDA and comparable foreign regulatory authorities for product candidates in clinical development. Regulatory authorities enforce these GCP requirements through periodic inspections of trial sponsors, clinical investigators and trial sites. If we or any of these third parties fail to comply with applicable GCP requirements, the clinical data generated in our clinical trials may be deemed unreliable and the FDA or comparable foreign regulatory authorities may require us to suspend or terminate these trials or perform additional preclinical studies or clinical trials before approving our marketing applications. We cannot be certain that, upon inspection, such regulatory authorities will determine that any of our clinical trials comply with the GCP requirements. In addition, our clinical trials must be conducted with biologic product produced under cGMP, requirements and may require a large number of patients.

Our failure or any failure by these third parties to comply with these regulations or to recruit a sufficient number of patients may require us to repeat clinical trials, which would delay the regulatory approval process. Moreover, our business may be implicated if any of these third parties violates federal or state fraud and abuse or false claims laws and regulations or healthcare privacy and security laws.

Any third parties conducting aspects of our preclinical studies or our future clinical trials will not be our employees and, except for remedies that may be available to us under our agreements with such third parties, we cannot control whether or not they devote sufficient time and resources to our preclinical studies and clinical programs. These third parties may also have relationships with other commercial entities, including our competitors, for whom they may also be conducting clinical trials or other product development activities, which could affect their performance on our behalf. If these third parties do not successfully carry out their contractual duties or obligations or meet expected deadlines, if they need to be replaced or if the quality or accuracy of the preclinical or clinical data they obtain is compromised due to the failure to adhere to our protocols or regulatory requirements or for other reasons, our development timelines, including clinical development timelines, may be extended, delayed or terminated and we may not be able to complete development of, obtain regulatory approval of or successfully commercialize our product candidates. As a result, our financial results and the commercial prospects for our product candidates would be harmed, our costs could increase and our ability to generate revenue could be delayed.

If any of our relationships with these third-party CROs or others terminate, we may not be able to enter into arrangements with alternative CROs or other third parties or to do so on commercially reasonable terms.

Switching or adding additional CROs involves additional cost and requires management time and focus. In addition, there is a natural transition period when a new CRO begins work. As a result, delays may occur, which can materially impact our ability to meet our desired development timelines. Though we carefully manage our relationships with our CROs, there can be no assurance that we will not encounter similar challenges or delays in the future or that these delays or challenges will not have a material adverse impact on our business, financial condition and prospects.

Because we rely on third-party manufacturing and supply partners, our supply of research and development, preclinical and clinical development materials may become limited or interrupted or may not be of satisfactory quantity or quality.

We rely on third-party contract manufacturers to manufacture some of our preclinical product candidate supplies and will rely on third-party contract manufacturers to manufacture all of our clinical trial product supplies. We do not own manufacturing facilities for producing any clinical trial product supplies. There can be no assurance that our preclinical and clinical development product supplies will not be limited, interrupted, or of satisfactory quality or continue to be available at acceptable prices. In particular, any replacement of our manufacturer could require significant effort and expertise because there may be a limited number of qualified replacements; this could be particularly problematic where we rely on a single-source supplier, as is currently the case for the manufacture of SRK-015.

The manufacturing process for a product candidate is subject to FDA and foreign regulatory authority review. Suppliers and manufacturers must meet applicable manufacturing requirements and undergo rigorous facility and process validation tests required by regulatory authorities in order to comply with regulatory standards, such as cGMPs. In the event that any of our manufacturers fails to comply with such requirements or to perform its obligations to us in relation to quality, timing or otherwise, or if our supply of components or other materials becomes limited or interrupted for other reasons, we may be forced to manufacture the materials ourselves, for which we currently do not have the capabilities or resources, or enter into an agreement with another third-party, which we may not be able to do on reasonable terms, if at all. In some cases, the technical skills or technology required to manufacture our product candidates may be unique or proprietary to the original manufacturer and we may have difficulty transferring such skills or technology to another third-party and a feasible alternative may not exist. These factors would increase our reliance on such manufacturer or require us to obtain a license from such manufacturer in order to have another third-party manufacture our product candidates. If we are required to change manufacturers for any reason, we will be required to verify that the new manufacturer maintains facilities and procedures that comply with quality standards and with all applicable regulations and guidelines. The delays associated with the verification of a new manufacturer could negatively affect our ability to develop product candidates in a timely manner or within budget.

We expect to continue to rely on third-party manufacturers if we receive regulatory approval for SRK-015 or any future product candidate. To the extent that we have existing, or enter into future, manufacturing arrangements with third parties, we will depend on these third parties to perform their obligations in a timely manner consistent with contractual and regulatory requirements, including those related to quality control and assurance. If we are unable to obtain or maintain third-party manufacturing for product candidates, or to do so on commercially reasonable terms, we may not be able to develop and commercialize our product candidates successfully. Our or a third-party's failure to execute on our manufacturing requirements and comply with cGMP could adversely affect our business in a number of ways, including:

- § an inability to initiate or continue clinical trials of product candidates under development;
- § delay in submitting regulatory applications, or receiving regulatory approvals, for product candidates;
- § loss of the cooperation of an existing or future collaborator;
- § subjecting third-party manufacturing facilities or our manufacturing facilities to additional inspections by regulatory authorities;

- § requirements to cease distribution or to recall batches of our product candidates; and
- § in the event of approval to market and commercialize a product candidate, an inability to meet commercial demands for our products.

In addition, we contract with fill and finishing providers with the appropriate expertise, facilities and scale to meet our needs. Failure to maintain cGMP can result in a contractor receiving FDA sanctions, which can impact our ability to operate or lead to delays in any clinical development programs. We believe that our current fill and finish contractor is operating in accordance with cGMP, but we can give no assurance that FDA, EMA or other regulatory agencies will not conclude that a lack of compliance exists. In addition, any delay in contracting for fill and finish services, or failure of the contract manufacturer to perform the services as needed, may delay any clinical trials, registration and launches, which could negatively affect our business.

Our reliance on third parties, such as manufacturers and antibody discovery vendors, may subject us to risks relating to manufacturing scale-up and may cause us to undertake substantial obligations, including financial obligations.

For example, in order to conduct clinical trials of our product candidates, we will need to manufacture them in large quantities. We, or any manufacturing partners, may be unable to successfully increase the manufacturing capacity for any of our product candidates in a timely or cost effective manner, or at all. In addition, quality issues may arise during scale-up activities. If we, or any manufacturing partners, are unable to successfully scale up the manufacture of our product candidates in sufficient quality and quantity, the development, testing, and clinical trials of that product candidate may be delayed or infeasible, and regulatory approval or commercial launch of any resulting product may be delayed or not obtained, which could significantly harm our business.

In addition, we rely, and intend to continue to rely, on third party entities to conduct antibody discovery based on criteria and specifications provided by us. Certain antibody discovery vendors may require us to enter into a license agreement with them for the right to use antibodies discovered by them in humans or for commercial purposes. While we have not executed such an agreement to date, there can be no assurance that we will not be required to execute such an agreement in the future if we select a clinical candidate that includes such an antibody and advance that clinical candidate into clinical trials. Such license agreements could include substantial milestone payments and royalties to the extent we choose to use an antibody discovered by such vendors. In addition, if we do not meet our obligations under such license agreements, the counterparties may have the ability to terminate the license agreements and we could lose the right to use the discovered antibodies, which could significantly and adversely impact our business.

Our future collaborations will be important to our business. If we are unable to enter into new collaborations, or if these collaborations are not successful, our business could be adversely affected.

A part of our strategy is to strategically evaluate and, as deemed appropriate, enter into additional partnerships in the future when strategically attractive, including potentially with major biotechnology or pharmaceutical companies. We have limited capabilities for product development and do not yet have any capability for commercialization. Accordingly, we may enter into collaborations with other companies to provide us with important technologies and funding for our programs and technology.

Any future collaborations we enter into may pose a number of risks, including the following:

- § collaborators have significant discretion in determining the efforts and resources that they will apply;
- § collaborators may not perform their obligations as expected;
- § collaborators may not pursue development and commercialization of any product candidates that achieve regulatory approval or may elect not to continue or renew development or commercialization programs or license arrangements based on clinical trial results, changes in the collaborators'

strategic focus or available funding, or external factors, such as a strategic transaction that may divert resources or create competing priorities;

- § collaborators may delay clinical trials, provide insufficient funding for a clinical trial program, stop a clinical trial or abandon a product candidate, repeat or conduct new clinical trials or require a new formulation of a product candidate for clinical testing;
- § collaborators could independently develop, or develop with third parties, products that compete directly or indirectly with our products and product candidates if the collaborators believe that the competitive products are more likely to be successfully developed or can be commercialized under terms that are more economically attractive than ours;
- § product candidates discovered in collaboration with us may be viewed by our collaborators as competitive with their own product candidates or products, which may cause collaborators to cease to devote resources to the commercialization of our product candidates;
- § collaborators may fail to comply with applicable regulatory requirements regarding the development, manufacture, distribution or marketing of a product candidate or product;
- § collaborators with marketing and distribution rights to one or more of our product candidates that achieve regulatory approval may not commit sufficient resources to the marketing and distribution of such product or products;
- § disagreements with collaborators, including disagreements over proprietary rights, contract interpretation or the preferred course of development, might cause delays or terminations of the research, development or commercialization of product candidates, might lead to additional responsibilities for us with respect to product candidates, or might result in litigation or arbitration, any of which would be time-consuming and expensive;
- § collaborators may not properly maintain or defend our intellectual property rights or may use our proprietary information in such a way as to invite litigation that could jeopardize or invalidate our intellectual property or proprietary information or expose us to potential litigation;
- § collaborators may infringe the intellectual property rights of third parties, which may expose us to litigation and potential liability;
- § if a collaborator of ours is involved in a business combination, the collaborator might deemphasize or terminate the development or commercialization of any product candidate licensed to it by us; and
- § collaborations may be terminated by the collaborator, and, if terminated, we could be required to raise additional capital to pursue further development or commercialization of the applicable product candidates.

If our collaborations do not result in the successful discovery, development and commercialization of product candidates or if one of our collaborators terminates its agreement with us, we may not receive any future research funding or milestone or royalty payments under such collaboration. All of the risks relating to product development, regulatory approval and commercialization described in this prospectus also apply to the activities of our therapeutic collaborators.

Additionally, if one of our collaborators terminates its agreement with us, we may find it more difficult to attract new collaborators and our perception in the business and financial communities could be adversely affected.

We face significant competition in seeking appropriate partners for our product candidates, and the negotiation process is time-consuming and complex. In order for us to successfully partner our product candidates, potential partners must view these product candidates as economically valuable in markets they determine to be attractive in light of the terms that we are seeking and other available products for licensing by other companies. Collaborations are complex and time-consuming to negotiate and document. In addition, there have been a significant number of recent business combinations among large pharmaceutical companies that have resulted in a reduced number of potential future collaborators. Our

ability to reach a definitive agreement for a collaboration will depend, among other things, upon our assessment of the collaborator's resources and expertise, the terms and conditions of the proposed collaboration and the proposed collaborator's evaluation of a number of factors. If we are unable to reach agreements with suitable collaborators on a timely basis, on acceptable terms, or at all, we may have to curtail the development of a product candidate, reduce or delay its development program or one or more of our other development programs, delay its potential commercialization or reduce the scope of any sales or marketing activities, or increase our expenditures and undertake development or commercialization activities at our own expense. If we elect to increase our expenditures to fund development or commercialization activities on our own, we may need to obtain additional expertise and additional capital, which may not be available to us on acceptable terms, or at all. If we fail to enter into collaborations or do not have sufficient funds or expertise to undertake the necessary development and commercialization activities, we may not be able to further develop our product candidates, bring them to market and generate revenue from sales of drugs or continue to develop our technology, and our business may be materially and adversely affected. Even if we are successful in our efforts to establish new strategic partnerships, the terms that we agree upon may not be favorable to us, and we may not be able to maintain such strategic partnerships if, for example, development or approval of a product candidate is delayed or sales of an approved product are disappointing. Any delay in entering into new strategic partnership agreements related to our product candidates could delay the development and commercialization of our product candidates and reduce their competitiveness even if they reach the market.

Risks Related to Our Common Stock and this Offering

No public market for our common stock currently exists, and we do not know whether an active, liquid and orderly trading market will develop for our common stock, or what the market price of our common stock will be, and as a result it may be difficult for you to sell your shares of our common stock.

Prior to this offering there has been no public market for shares of our common stock. Although we have applied to list our common stock on the Nasdaq Global Market, an active trading market for our shares may never develop or be sustained following this offering. You may not be able to sell your shares quickly or at the market price if trading in shares of our common stock is not active. The initial public offering price for our common stock will be determined through negotiations with the underwriters, and the negotiated price may not be indicative of the market price of the common stock after the offering. As a result of these and other factors, you may be unable to resell your shares of our common stock at or above the initial public offering price.

Further, an inactive market may also impair our ability to raise capital by selling shares of our common stock and may impair our ability to enter into strategic partnerships or acquire companies or products by using our shares of common stock as consideration.

The price of our stock may be volatile, and you could lose all or part of your investment.

The trading price of our common stock following this offering is likely to be highly volatile and could be subject to wide fluctuations in response to various factors, some of which are beyond our control, including limited trading volume. In addition to the factors discussed in this "Risk Factors" section and elsewhere in this prospectus, these factors include:

- § the commencement, enrollment or results of our planned Phase 1 clinical trial for SRK-015;
- § any delay in identifying a clinical candidate for our other development programs;
- § any delay in our regulatory filings for SRK-015 and any adverse development or perceived adverse development with respect to the applicable regulatory authority's review of such filings, including without limitation the FDA's issuance of a "refusal to file" letter or a request for additional information;

- § adverse results or delays in future clinical trials;
- § our decision to initiate a clinical trial, not to initiate a clinical trial or to terminate an existing clinical trial;
- § adverse regulatory decisions, including failure to receive regulatory approval of SRK-015 or any future product candidate;
- § changes in laws or regulations applicable to SRK-015 or any future product candidate, including but not limited to clinical trial requirements for approvals;
- § adverse developments concerning our manufacturers;
- § our inability to obtain adequate product supply for any approved product or inability to do so at acceptable prices;
- § our inability to establish collaborations, if needed;
- § our failure to commercialize our product candidates, if approved;
- § additions or departures of key scientific or management personnel;
- § unanticipated serious safety concerns related to the use of SRK-015 or any future product candidate;
- § introduction of new products or services offered by us or our competitors;
- § announcements of significant acquisitions, strategic partnerships, joint ventures or capital commitments by us or our competitors;
- § our ability to effectively manage our growth;
- § actual or anticipated variations in quarterly operating results;
- § our cash position;
- § our failure to meet the estimates and projections of the investment community or that we may otherwise provide to the public;
- § publication of research reports about us or our industry, or product candidates in particular, or positive or negative recommendations or withdrawal of research coverage by securities analysts;
- § changes in the market valuations of similar companies;
- § overall performance of the equity markets;
- § sales of our common stock by us or our stockholders in the future;
- § trading volume of our common stock;
- § changes in accounting practices;
- § ineffectiveness of our internal controls;
- § disputes or other developments relating to proprietary rights, including patents, litigation matters and our ability to obtain patent protection for our technologies;
- § significant lawsuits, including patent or stockholder litigation;
- § general political and economic conditions; and
- § other events or factors, many of which are beyond our control.

In addition, the stock market in general, and the market for biopharmaceutical companies in particular, have experienced extreme price and volume fluctuations that have often been unrelated or disproportionate to the operating performance of these companies. Broad market and industry factors may negatively affect the market price of our common stock, regardless of our actual operating performance. If the market price of our common stock after this offering does not exceed the initial public offering price, you may not realize any return on your investment in us and may lose some or all of your investment. In the past, securities class action litigation has often been instituted against companies following periods of volatility in the market price of a company's securities. This type of litigation, if instituted, could result in substantial costs and a diversion of management's attention and resources, which would harm our business, operating results or financial condition.

We do not intend to pay dividends on our common stock so any returns will be limited to the value of our stock.

We currently anticipate that we will retain future earnings for the development, operation and expansion of our business and do not anticipate declaring or paying any cash dividends for the foreseeable future. Furthermore, our ability to pay cash dividends is currently restricted by the terms of our credit facility with Silicon Valley Bank, and future debt or other financing arrangements may contain terms prohibiting or limiting the amount of dividends that may be declared or paid on our common stock. Any return to stockholders will therefore be limited to the appreciation of their stock.

Our principal stockholders and management own a significant percentage of our stock and will be able to exert significant control over matters subject to stockholder approval.

Immediately following the completion of this offering, our executive officers, directors and their affiliates will beneficially hold, in the aggregate, approximately 43.0% of our outstanding voting stock. These stockholders, acting together, would be able to significantly influence all matters requiring stockholder approval. For example, these stockholders would be able to significantly influence elections of directors, amendments of our organizational documents, or approval of any merger, sale of assets, or other major corporate transaction. This may prevent or discourage unsolicited acquisition proposals or offers for our common stock that you may feel are in your best interest as one of our stockholders.

If you purchase our common stock in this offering, you will incur immediate and substantial dilution in the book value of your shares.

The initial public offering price will be substantially higher than the pro forma as adjusted net tangible book value per share of our common stock after this offering. Investors purchasing common stock in this offering will pay a price per share that substantially exceeds the pro forma as adjusted net tangible book value per share after this offering. As a result, investors purchasing common stock in this offering will incur immediate dilution of \$9.30 per share, based on an assumed initial public offering price of \$14.00 per share, the midpoint of the price range set forth on the cover page of this prospectus, representing the difference between our pro forma as adjusted net tangible book value per share after giving effect to this offering and the assumed initial public offering price. Further, investors purchasing common stock in this offering will contribute approximately 40.7% of the total amount invested by stockholders since our inception, but will own only approximately 21.9% of the shares of common stock outstanding after this offering.

This dilution is due to our investors who purchased shares prior to this offering having paid substantially less when they purchased their shares than the price offered to the public in this offering. To the extent outstanding options are exercised, there will be further dilution to new investors. As a result of the dilution to investors purchasing shares in this offering, investors may receive significantly less than the purchase price paid in this offering, if anything, in the event of our liquidation. For a further description of the dilution that you will experience immediately after this offering, see "Dilution."

We are an emerging growth company, and we cannot be certain if the reduced reporting requirements applicable to emerging growth companies will make our common stock less attractive to investors.

We are an emerging growth company, as defined in the Jumpstart Our Business Startups Act, or the JOBS Act, enacted in April 2012. For as long as we continue to be an emerging growth company, we may take advantage of exemptions from various reporting requirements that are applicable to other public companies that are not emerging growth companies, including not being required to comply with the auditor attestation requirements of Section 404 of the Sarbanes-Oxley Act of 2002, as amended, or the Sarbanes-Oxley Act, reduced disclosure obligations regarding executive compensation in this prospectus and our periodic reports and proxy statements and exemptions from the requirements of holding nonbinding advisory votes on executive compensation and stockholder approval of any golden parachute payments not previously approved. We could be an emerging growth company for up to five years following the year in which we

complete this offering, although circumstances could cause us to lose that status earlier. We will remain an emerging growth company until the earlier of (1) the last day of the fiscal year (a) following the fifth anniversary of the completion of this offering, (b) in which we have total annual gross revenue of at least \$1.07 billion or (c) in which we are deemed to be a large accelerated filer, which requires the market value of our common stock that is held by non-affiliates to exceed \$700 million as of the prior June 30th, and (2) the date on which we have issued more than \$1 billion in non-convertible debt during the prior three-year period. We cannot predict if investors will find our common stock less attractive because we may rely on these exemptions. If some investors find our common stock less attractive as a result, there may be a less active trading market for our common stock and our stock price may be more volatile.

Under the JOBS Act, emerging growth companies can also delay adopting new or revised accounting standards until such time as those standards apply to private companies. We have irrevocably elected not to avail ourselves of delayed adoption of new or revised accounting standards and, therefore, we will be subject to the same requirements to adopt new or revised accounting standards as other public companies that are not emerging growth companies.

We will incur significant increased costs as a result of operating as a public company, and our management will be required to devote substantial time to new compliance initiatives.

As a public company, we will incur significant legal, accounting and other expenses that we did not incur as a private company. We will be subject to the reporting requirements of the Securities Exchange Act of 1934, as amended, which will require, among other things, that we file with the SEC, annual, quarterly and current reports with respect to our business and financial condition. In addition, the Sarbanes-Oxley Act, as well as rules subsequently adopted by the SEC and the Nasdaq Global Market to implement provisions of the Sarbanes-Oxley Act, impose significant requirements on public companies, including requiring establishment and maintenance of effective disclosure and financial controls and changes in corporate governance practices. Further, in July 2010, the Dodd-Frank Wall Street Reform and Consumer Protection Act, or the Dodd-Frank Act, was enacted. There are significant corporate governance and executive compensation related provisions in the Dodd-Frank Act that require the SEC to adopt additional rules and regulations in these areas such as "say on pay" and proxy access. Recent legislation permits emerging growth companies to implement many of these requirements over a longer period and up to five years from the pricing of this offering. Stockholder activism, the current political environment and the current high level of government intervention and regulatory reform may lead to substantial new regulations and disclosure obligations, which may lead to additional compliance costs and impact the manner in which we operate our business in ways we cannot currently anticipate.

We expect the rules and regulations applicable to public companies to substantially increase our legal and financial compliance costs and to make some activities more time-consuming and costly. If these requirements divert the attention of our management and personnel from other business concerns, they could have an adverse effect on our business. The increased costs will decrease our net income or increase our net loss, and may require us to reduce costs in other areas of our business or increase the prices of our products or services. For example, we expect these rules and regulations to make it more difficult and more expensive for us to obtain director and officer liability insurance and we may be required to incur substantial costs to maintain the same or similar coverage. We cannot predict or estimate the amount or timing of additional costs we may incur to respond to these requirements. The impact of these requirements could also make it more difficult for us to attract and retain qualified persons to serve on our board of directors, our board committees or as executive officers.

We may not be able to meet the internal control reporting requirements imposed by the SEC resulting in a possible decline in the price of our common stock and our inability to obtain future financing.

As directed by Section 404 of the Sarbanes-Oxley Act, the SEC adopted rules requiring each public company to include a report of management on the company's internal controls over financial reporting in its annual reports. Although the Dodd-Frank Wall Street Reform and Consumer Protection Act exempts

companies with a public float of less than \$75 million from the requirement that our independent registered public accounting firm attest to our financial controls, this exemption does not affect the requirement that we include a report of management on our internal control over financial reporting and does not affect the requirement to include the independent registered public accounting firm's attestation if our public float exceeds \$75 million.

While we expect to expend significant resources in developing the necessary documentation and testing procedures required by Section 404 of the Sarbanes-Oxley Act, there is a risk that we may not be able to comply timely with all of the requirements imposed by this rule. Regardless of whether we are required to receive a positive attestation from our independent registered public accounting firm with respect to our internal controls, if we are unable to do so, investors and others may lose confidence in the reliability of our financial statements and our stock price and ability to obtain equity or debt financing as needed could suffer.

In addition, in the event that our independent registered public accounting firm is unable to rely on our internal controls in connection with its audit of our financial statements, and in the further event that it is unable to devise alternative procedures in order to satisfy itself as to the material accuracy of our financial statements and related disclosures, it is possible that we would be unable to file our Annual Report on Form 10-K with the SEC, which could also adversely affect the market for and the market price of our common stock and our ability to secure additional financing as needed.

Sales of a substantial number of shares of our common stock by our existing stockholders in the public market could cause our stock price to fall.

If our existing stockholders sell, or indicate an intention to sell, substantial amounts of our common stock in the public market after the lock-up and other legal restrictions on resale discussed in this prospectus lapse, the trading price of our common stock could decline. Based on shares of common stock outstanding as of March 31, 2018, upon the completion of this offering we will have outstanding a total of _____ shares of common stock. Of these shares, only the shares of common stock sold in this offering by us, plus any shares sold upon exercise of the underwriters' option to purchase additional shares, will be freely tradable without restriction in the public market immediately following this offering.

The lock-up agreements pertaining to this offering will expire 180 days from the date of this prospectus, subject to earlier release of all or a portion of the shares subject to such agreements by Jefferies LLC and Cowen and Company, LLC in their sole discretion. After the lock-up agreements expire, based upon the number of shares of common stock, on an as-converted basis, outstanding as of March 31, 2018, up to an additional 19,080,536 shares of common stock will be eligible for sale in the public market. Approximately 55.1% of these additional shares are beneficially held by directors, executive officers and their affiliates and will be subject to certain limitations of Rule 144 under the Securities Act of 1933, as amended, or the Securities Act.

In addition, shares of common stock that are either subject to outstanding options or reserved for future issuance under our existing equity compensation plans will become eligible for sale in the public market to the extent permitted by the provisions of various vesting schedules, the lock-up agreements and Rule 144 and Rule 701 under the Securities Act. If these additional shares of common stock are sold, or if it is perceived that they will be sold, in the public market, the trading price of our common stock could decline. Additionally, the number of shares of our common stock reserved for issuance under 2018 Stock Option and Incentive Plan will automatically increase on January 1 of each year, beginning on January 1, 2019 and continuing through and including January 1, 2029, by 4% of the total number of shares of our capital stock outstanding on December 31 of the preceding calendar year, or a lesser number of shares determined by our board of directors or compensation committee. Unless our board of directors elects not to increase the number of shares available for future grant each year, our stockholders may experience additional dilution.

After this offering, the holders of 15,109,950 shares of our common stock as of March 31, 2018 will be entitled to rights with respect to the registration of their shares under the Securities Act as provided under the terms of an investors' rights agreement between us and the holders of our convertible preferred stock, subject to the 180-day lock-up agreements described above. See "Description of Capital Stock — Registration Rights." Registration of these shares under the Securities Act would result in the shares becoming freely tradable without restriction under the Securities Act, except for shares held by affiliates, as defined in Rule 144 under the Securities Act. Any sales of securities by these stockholders could have a material adverse effect on the trading price of our common stock.

We have broad discretion in the use of our existing cash, cash equivalents and marketable securities and the net proceeds from this offering and may not use them effectively.

Our management will have broad discretion in the application of our existing cash, cash equivalents and marketable securities and the net proceeds from this offering, including for any of the purposes described in the section titled "Use of Proceeds," and you will not have the opportunity as part of your investment decision to assess whether such proceeds are being used appropriately. Because of the number and variability of factors that will determine our use of our existing cash and cash equivalents and the net proceeds from this offering, their ultimate use may vary substantially from their currently intended use. Our management might not apply our existing cash and cash equivalents and the net proceeds from this offering in ways that ultimately increase the value of your investment. The failure by our management to apply these funds effectively could harm our business. Pending their use, we may invest the net proceeds from this offering in short-term, investment-grade, interest-bearing securities. These investments may not yield a favorable return to our stockholders. If we do not invest or apply the net proceeds from this offering in ways that enhance stockholder value, we may fail to achieve expected financial results, which could cause our stock price to decline.

Anti-takeover provisions under our charter documents and Delaware law could delay or prevent a change of control which could limit the market price of our common stock and may prevent or frustrate attempts by our stockholders to replace or remove our current management.

Our amended and restated certificate of incorporation and amended and restated bylaws, which are to become effective upon the completion of this offering, contain provisions that could delay or prevent a change of control of our company or changes in our board of directors that our stockholders might consider favorable. Some of these provisions include:

- § a board of directors divided into three classes serving staggered three-year terms, such that not all members of the board will be elected at one time;
- § a prohibition on stockholder action through written consent, which requires that all stockholder actions be taken at a meeting of our stockholders;
- § a requirement that special meetings of stockholders be called only by the chairman of the board of directors, the chief executive officer, or by a majority of the total number of authorized directors;
- § advance notice requirements for stockholder proposals and nominations for election to our board of directors;
- § a requirement that no member of our board of directors may be removed from office by our stockholders except for cause and, in addition to any other vote required by law, upon the approval of not less than two-thirds of all outstanding shares of our voting stock then entitled to vote in the election of directors;
- § a requirement of approval of not less than two-thirds of all outstanding shares of our voting stock to amend any bylaws by stockholder action or to amend specific provisions of our certificate of incorporation; and
- § the authority of the board of directors to issue convertible preferred stock on terms determined by the board of directors without stockholder approval and which convertible preferred stock may include rights superior to the rights of the holders of common stock.

In addition, because we are incorporated in Delaware, we are governed by the provisions of Section 203 of the Delaware General Corporate Law, which may prohibit certain business combinations with stockholders owning 15% or more of our outstanding voting stock. These anti-takeover provisions and other provisions in our amended and restated certificate of incorporation and amended and restated bylaws could make it more difficult for stockholders or potential acquirers to obtain control of our board of directors or initiate actions that are opposed by the then-current board of directors and could also delay or impede a merger, tender offer or proxy contest involving our company. These provisions could also discourage proxy contests and make it more difficult for you and other stockholders to elect directors of your choosing or cause us to take other corporate actions you desire. Any delay or prevention of a change of control transaction or changes in our board of directors could cause the market price of our common stock to decline.

If securities or industry analysts do not publish research or publish inaccurate or unfavorable research about our business, our stock price and trading volume could decline.

The trading market for our common stock will depend in part on the research and reports that securities or industry analysts publish about us or our business. Securities and industry analysts do not currently, and may never, publish research on our company. If no securities or industry analysts commence coverage of our company, the trading price for our stock would likely be negatively impacted. In the event securities or industry analysts initiate coverage, if one or more of the analysts who covers us downgrades our stock or publishes inaccurate or unfavorable research about our business, our stock price may decline. If one or more of these analysts ceases coverage of our company or fails to publish reports on us regularly, demand for our stock could decrease, which might cause our stock price and trading volume to decline.

Our amended and restated bylaws will contain certain exclusive forum provisions requiring that substantially all disputes between us and our stockholders be resolved in certain judicial forums, which could limit our stockholders' ability to obtain a favorable judicial forum for disputes with us or our directors, officers or employees.

Our amended and restated bylaws will provide that the Court of Chancery of the State of Delaware will be the exclusive forum for any derivative action or proceeding brought on our behalf, any action asserting a breach of fiduciary duty, any action asserting a claim against us arising pursuant to the Delaware General Corporation Law, our certificate of incorporation or our bylaws, any action to interpret, apply, enforce, or determine the validity of our certificate of incorporation or bylaws, or any action asserting a claim against us that is governed by the internal affairs doctrine. In addition, our amended and restated bylaws will contain a provision by virtue of which, unless we consent in writing to the selection of an alternative forum, the United States District Court for the District of Massachusetts will be the exclusive forum for any complaint asserting a cause of action arising under the Securities Act. In addition, our amended and restated bylaws will provide that any person or entity purchasing or otherwise acquiring any interest in shares of our common stock is deemed to have notice of and consented to the foregoing provisions. We have chosen the United States District Court for the District of Massachusetts as the exclusive forum for such causes of action because our principal executive offices are located in Cambridge, Massachusetts. Some companies that have adopted similar federal district court forum selection provisions are currently subject to a suit in the Court of Chancery of the State of Delaware brought by stockholders who assert that the federal district court forum selection provision is not enforceable. We recognize that the federal district court forum selection clause may impose additional litigation costs on stockholders who assert the provision is not enforceable and may impose more general additional litigation costs in pursuing any such claims, particularly if the stockholders do not reside in or near the Commonwealth of Massachusetts. Additionally, the choice of forum provision may limit a stockholder's ability to bring a claim in a judicial forum that it finds favorable for disputes with us or our directors, officers or other employees, which may discourage such lawsuits against us and our directors, officers and other employees. Alternatively, if a court were to find the choice of forum provision contained in our amended and restated bylaws to be inapplicable or unenforceable in an action, we may incur additional costs associated with resolving such action in other jurisdictions, which could adversely affect our business and financial condition.

SPECIAL NOTE REGARDING FORWARD-LOOKING STATEMENTS

This prospectus contains forward-looking statements that are based on management's beliefs and assumptions and on information currently available to management. Some of the statements in the sections titled "Prospectus Summary," "Risk Factors," "Management's Discussion and Analysis of Financial Condition and Results of Operations," "Business" and elsewhere in this prospectus contain forward-looking statements. In some cases, you can identify forward-looking statements by the following words: "may," "will," "could," "would," "should," "expect," "intend," "plan," "anticipate," "believe," "estimate," "predict," "project," "potential," "continue," "ongoing" or the negative of these terms or other comparable terminology, although not all forward-looking statements contain these words.

These statements involve risks, uncertainties and other factors that may cause actual results, levels of activity, performance or achievements to be materially different from the information expressed or implied by these forward-looking statements. Although we believe that we have a reasonable basis for each forward-looking statement contained in this prospectus, we caution you that these statements are based on a combination of facts and factors currently known by us and our projections of the future, about which we cannot be certain. Forward-looking statements in this prospectus include, but are not limited to, statements about:

- § the timing of initiation and completion of our planned Phase 1 clinical trial and future clinical trials for our lead product candidate, SRK-015, and the results from these trials;
- § the success, cost and timing of our other product development activities, preclinical studies and clinical trials, including statements regarding our ability to identify a clinical candidate and lead indication in our TGFb1 program, the timing of initiation and completion of preclinical studies or clinical trials and related preparatory work, and the timing of the availability of the results of these studies and trials;
- § our success in identifying and executing a development program for additional indications for SRK-015 and our TGFb1 program;
- § our ability to obtain funding for our operations, including funding necessary to complete further development and, upon successful development, if approved, commercialization of SRK-015 or any of our future product candidates;
- § the potential for our identified research priorities to advance our proprietary platform, development programs or product candidates;
- § our ability to obtain and maintain regulatory approval from the U.S. Food and Drug Administration, European Medicines Agency and other regulatory authorities for SRK-015 and any future product candidates, and any related restrictions, limitations or warnings in the label of any approved product candidate;
- § our expectations regarding our ability to obtain and maintain intellectual property protection for our product candidates and the duration of such protection;
- § our ability and the potential to successfully manufacture our product candidates for clinical trials and for commercial use, if approved;
- § the size and growth potential of the markets for our product candidates, and our ability to serve those markets, either alone or in combination with others;
- § our estimates regarding expenses, future revenue, capital requirements and needs for additional financing; and
- § our use of the proceeds from this offering.

In addition, you should refer to the section titled "Risk Factors" of this prospectus for a discussion of other important factors that may cause actual results to differ materially from those expressed or implied by the forward-looking statements. As a result of these factors, we cannot assure you that the forward-looking

statements in this prospectus will prove to be accurate. Furthermore, if the forward-looking statements prove to be inaccurate, the inaccuracy may be material. In light of the significant uncertainties in these forward-looking statements, you should not regard these statements as a representation or warranty by us or any other person that we will achieve our objectives and plans in any specified time frame, or at all. The forward-looking statements in this prospectus represent our views as of the date of this prospectus. We anticipate that subsequent events and developments will cause our views to change. However, while we may elect to update these forward-looking statements at some point in the future, we have no current intention of doing so except to the extent required by applicable law. You should, therefore, not rely on these forward-looking statements as representing our views as of any date subsequent to the date of this prospectus.

The market data and certain other statistical information used throughout this prospectus are based on independent industry publications, governmental publications, reports by market research firms or other independent sources that we believe to be reliable sources. Industry publications and third-party research, surveys and studies generally indicate that their information has been obtained from sources believed to be reliable, although they do not guarantee the accuracy or completeness of such information. We are responsible for all of the disclosure contained in this prospectus, and we believe these industry publications and third-party research, surveys and studies are reliable. While we are not aware of any misstatements regarding any third-party information presented in this prospectus, their estimates, in particular, as they relate to projections, involve numerous assumptions, are subject to risks and uncertainties and are subject to change based on various factors, including those discussed under the section titled "Risk Factors" and elsewhere in this prospectus. Some data are also based on our good faith estimates.

USE OF PROCEEDS

We estimate that the net proceeds to us from the sale of the shares of our common stock in this offering will be approximately \$67.5 million, or approximately \$78.0 million if the underwriters exercise their option to purchase additional shares in full, based upon an assumed initial public offering price of \$14.00 per share, the midpoint of the price range set forth on the cover page of this prospectus, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us.

A \$1.00 increase (decrease) in the assumed initial public offering price of \$14.00 per share, the midpoint of the price range set forth on the cover page of this prospectus, would increase (decrease) the net proceeds to us from this offering by approximately \$5.0 million, assuming the number of shares offered by us, as set forth on the cover page of this prospectus, remains the same. Similarly, an increase (decrease) of 1,000,000 shares in the number of shares offered by us would increase (decrease) the net proceeds to us from this offering by approximately \$13.0 million, assuming that the assumed initial public offering price remains the same. We do not expect that a change in the initial public offering price or the number of shares by these amounts would have a material effect on our uses of the proceeds from this offering, although a decrease in proceeds may accelerate the time at which we will need to seek additional capital.

We currently expect to use the net proceeds from this offering, together with our cash, cash equivalents and marketable securities, as follows:

- § \$35.0 million to fund research and development activities for SRK-015 through our planned Phase 2 clinical proof of concept trial;
- § \$18.0 million to fund TGFb1, BMP6 and other preclinical research and development activities; and
- § the remainder for working capital and other general corporate purposes.

Based on our current plans, we believe our existing cash, cash equivalents and marketable securities, together with the net proceeds from this offering, will be sufficient to fund our operating expenses and capital expenditure requirements into the second half of 2020.

We cannot specify with certainty all of the particular uses for the net proceeds to be received upon the completion of this offering. Due to uncertainties inherent in the product development process, it is difficult to estimate the exact amounts of the net proceeds that will be used for any particular purpose. We may use our existing cash, cash equivalents and marketable securities and the future payments, if any, generated from any future collaboration agreements to fund our operations, either of which may alter the amount of net proceeds used for a particular purpose. In addition, the amount, allocation and timing of our actual expenditures will depend upon numerous factors, including the results of our research and development efforts, the timing and success of clinical trials and the timing of regulatory submissions. Accordingly, we will have broad discretion in using these proceeds.

Pending their uses, we plan to invest the net proceeds of this offering in short-term, interest-bearing, investment-grade instruments, certificates of deposit or direct or guaranteed obligations of the U.S. government.

DIVIDEND POLICY

We have never declared or paid any cash dividends on our common stock or any other securities. We anticipate that we will retain all available funds and any future earnings, if any, for use in the operation of our business and do not anticipate paying cash dividends in the foreseeable future. In addition, our ability to pay cash dividends is currently restricted by the terms of our credit facility with Silicon Valley Bank, and future debt or other financing arrangements may contain terms prohibiting or limiting the amount of dividends that may be declared or paid on our common stock.

CAPITALIZATION

The following table sets forth our cash, cash equivalents and marketable securities and our capitalization as of March 31, 2018:

- § on an actual basis;
- § on a pro forma basis to give effect to (1) the automatic conversion of all outstanding shares of our convertible preferred stock into an aggregate of 15,109,950 shares of common stock upon the completion of this offering, (2) the automatic conversion of the outstanding warrant to purchase 21,739 shares of convertible preferred stock into a warrant to purchase 7,614 shares of common stock, resulting in the reclassification of the warrant liability to additional paid-in capital, (3) the vesting of a performance-based stock option award to purchase up to 8,757 shares of our common stock with vesting conditions contingent upon completion of this offering, and the resulting recognition of equity-based compensation expense and (4) the filing and effectiveness of our amended and restated certificate of incorporation upon the completion of this offering; and
- § on a pro forma as adjusted basis to give further effect to our issuance and sale of 5,360,000 shares of our common stock in this offering at an assumed initial public offering price of \$14.00 per share, the midpoint of the price range set forth on the cover page of this prospectus, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us.

You should read the information in this table together with our consolidated financial statements and the related notes appearing elsewhere in this prospectus, as well as the sections of this prospectus captioned "Selected Consolidated Financial Data" and "Management's Discussion and Analysis of Financial Condition and Results of Operations."

	As of March 31, 2018		
	Actual	Pro Forma	Pro Forma As Adjusted
	(in thousands, except share and per share data)		
Cash, cash equivalents and marketable securities	\$ 47,763	\$ 47,763	\$ 115,250
Convertible preferred stock, \$0.001 par value; 43,157,651 shares authorized, 43,135,911 shares issued and outstanding and aggregate liquidation preference of \$109,561, actual; no shares issued or outstanding, pro forma and pro forma as adjusted	\$ 109,232	\$ —	\$ —
Stockholders' equity (deficit):			
Preferred stock, \$0.001 par value; no shares authorized, issued or outstanding, actual; 10,000,000 shares authorized and no shares issued or outstanding, pro forma and pro forma as adjusted	—	—	—
Common stock, \$0.001 par value; 60,000,000 shares authorized, 3,970,586 shares issued and outstanding, actual; 150,000,000 shares authorized, pro forma and pro forma as adjusted; 19,080,536 shares authorized and outstanding, pro forma; 24,440,536 shares issued and outstanding, pro forma as adjusted	4	19	24
Additional paid-in capital	4,483	113,799	181,281
Accumulated other comprehensive loss	(1)	(1)	(1)
Accumulated deficit	(66,417)	(66,459)	(66,459)
Total stockholders' equity (deficit)	(61,931)	47,358	114,845
Total capitalization	\$ 47,301	\$ 47,358	\$ 114,845

The pro forma as adjusted information is illustrative only, and our capitalization following the completion of this offering will depend on the actual initial public offering price and other terms of this offering determined at pricing. A \$1.00 increase (decrease) in the assumed initial public offering price of \$14.00 per share, the midpoint of the price range set forth on the cover page of this prospectus, would increase (decrease) the pro forma as adjusted amount of each of cash, cash equivalents and marketable securities, additional paid-in capital, total stockholders' equity and total capitalization by \$5.0 million, assuming the number of shares offered by us, as set forth on the cover page of this prospectus, remains the same, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us. Similarly, each increase (decrease) of 1,000,000 shares in the number of shares offered by us in this offering, as set forth of the cover page of this prospectus, would increase (decrease) the pro forma as adjusted amount of cash and cash equivalents, additional paid-in capital, total stockholders' equity and total capitalization by \$13.0 million, assuming no change in the assumed initial public offering price per share, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us.

The actual, pro forma and pro forma as adjusted information set forth in the table excludes:

- § 7,614 shares of common stock issuable upon the exercise of a warrant outstanding as of March 31, 2018, at an exercise price of \$3.94 per share;
- § 660,319 shares of common stock issuable upon the exercise of stock options outstanding as of March 31, 2018 under our 2017 Stock Option and Incentive Plan at an exercise price of \$5.77 per share;
- § 355,539 shares of common stock issuable upon the exercise of stock options granted subsequent to March 31, 2018 under our 2017 Stock Option and Incentive Plan at an exercise price of \$7.17 per share;
- § 3,139,274 shares of our common stock that will become available for future issuance under our 2018 Stock Option and Incentive Plan upon the effectiveness of the registration statement of which this prospectus forms a part, from which we have granted options to purchase 309,925 shares of common stock, subject to and effective upon the closing of this offering, at an exercise price equal to the public offering price per share in this offering; and
- § 235,743 shares of our common stock that will become available for future issuance under our 2018 Employee Stock Purchase Plan upon the effectiveness of the registration statement of which this prospectus forms a part.

DILUTION

If you invest in our common stock in this offering, your ownership interest will be diluted immediately to the extent of the difference between the initial public offering price per share of common stock and the pro forma as adjusted net tangible book value per share of common stock immediately after this offering.

Our historical net tangible book value (deficit) as of March 31, 2018 was \$(61.9) million, or \$(15.60) per share of common stock. Our historical net tangible book value (deficit) is the amount of our total tangible assets less our total liabilities and the carrying value of our convertible preferred stock. Historical net tangible book value (deficit) per share represents historical net tangible book value (deficit) divided by the 3,970,586 shares of common stock outstanding as of March 31, 2018.

Our pro forma net tangible book value as of March 31, 2018 was \$47.4 million, or \$2.48 per share of our common stock. Pro forma net tangible book value represents the amount of our total tangible assets less our total liabilities, after giving effect to the automatic conversion immediately prior to the completion of this offering of all outstanding shares of our convertible preferred stock into an aggregate of 15,109,950 shares of common stock, the automatic conversion of the outstanding warrant to purchase 21,739 shares of convertible preferred stock into a warrant to purchase 7,614 shares of common stock, resulting in the reclassification of the warrant liability to additional paid-in capital, and the vesting of a performance-based stock option award to purchase up to 8,757 shares of our common stock with vesting conditions contingent upon completion of this offering, and the resulting recognition of equity-based compensation expense. Pro forma net tangible book value per share represents pro forma net tangible book value divided by the total number of shares outstanding as of March 31, 2018, after giving effect to the pro forma adjustments described above.

After giving further effect to the sale and issuance of 5,360,000 shares of our common stock in this offering at an assumed initial public offering price of \$14.00 per share, the midpoint of the price range set forth on the cover of this prospectus, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us, our pro forma as adjusted net tangible book value as of March 31, 2018 would have been \$114.8 million, or \$4.70 per share. This represents an immediate increase in pro forma as adjusted net tangible book value per share of \$2.22 to existing stockholders and immediate dilution per share of \$9.30 in pro forma as adjusted net tangible book value per share to new investors participating in this offering. Dilution per share to new investors is determined by subtracting pro forma as adjusted net tangible book value per share after this offering from the assumed initial public offering price per share paid by new investors.

The following table illustrates this dilution on a per-share basis to new investors:

Assumed initial public offering price per share	\$ 14.00
Historical net tangible book value (deficit) per share as of March 31, 2018	\$ (15.60)
Increase per share attributable to pro forma adjustments	<u>18.08</u>
Pro forma net tangible book value (deficit) per share as of March 31, 2018	2.48
Increase in pro forma as adjusted net tangible book value per share attributable to new investors participating in this offering	<u>2.22</u>
Pro forma as adjusted net tangible book value per share after this offering	4.70
Dilution per share to new investors participating in this offering	<u>\$ 9.30</u>

The pro forma as adjusted information discussed above is illustrative only and will depend on the actual initial price to the public and other terms of this offering determined at pricing. A \$1.00 increase

(decrease) in the assumed initial public offering price of \$14.00 per share, the midpoint of the price range set forth on the cover page of this prospectus, would increase (decrease) our pro forma as adjusted net tangible book value by \$5.0 million, or \$0.20 per share, and increase (decrease) the dilution per share to investors participating in this offering by \$0.80 per share, assuming that the number of shares offered by us, as set forth on the cover page of this prospectus, remains the same, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us. An increase of 1,000,000 in the number of shares offered by us, as set forth on the cover page of this prospectus, would increase our pro forma as adjusted net tangible book value by \$13.0 million, or \$0.33 per share, and decrease the dilution per share to investors participating in this offering by \$0.33 per share, assuming that the assumed initial public offering price remains the same, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us. A decrease of 1,000,000 in the number of shares offered by us, as set forth on the cover page of this prospectus, would decrease our pro forma as adjusted net tangible book value by \$13.0 million, or \$0.36 per share, and increase the dilution per share to investors participating in this offering by \$0.36 per share, assuming that the assumed initial public offering price remains the same, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us.

If the underwriters exercise their option in full to purchase 804,000 additional shares of common stock in this offering, our pro forma as adjusted net tangible book value after this offering would be \$125.3 million, representing an immediate increase in pro forma as adjusted net tangible book value per share of \$2.48 to existing stockholders and immediate dilution in pro forma as adjusted net tangible book value per share of \$9.04 to investors participating in this offering, assuming an initial public offering price of \$14.00 per share, the midpoint of the price range set forth on the cover page of this prospectus.

The following table summarizes, on the pro forma as adjusted basis described above as of March 31, 2018, the total number of shares of common stock purchased from us on an as converted to common stock basis, the total consideration paid or to be paid, and the average price per share paid or to be paid by existing stockholders and by investors participating in this offering at an assumed initial public offering price of \$14.00 per share, the midpoint of the price range set forth on the cover page of this prospectus, before deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us:

	Shares Purchased		Total Consideration		Average Price Per Share
	Number	Percentage	Amount (in thousands)	Percentage	
Existing stockholders	19,080,536	78.1%	\$ 109,232	59.3%	5.72
Investors participating in this offering	5,360,000	21.9	75,040	40.7	14.00
Total	24,440,536	100.0%	\$ 184,272	100.0%	

A \$1.00 increase (decrease) in the assumed initial public offering price of \$14.00 per share, the midpoint of the price range set forth on the cover page of this prospectus, would increase (decrease) the total consideration paid by new investors by \$5.4 million and, in the case of an increase, would increase the percentage of total consideration paid by new investors by 1.7 percentage points and, in the case of a decrease, would decrease the percentage of total consideration paid by new investors by 1.7 percentage points, assuming that the number of shares offered by us, as set forth on the cover page of this prospectus, remains the same. An increase (decrease) of 1,000,000 in the number of shares offered by us, as set forth on the cover page of this prospectus, would increase (decrease) the total consideration paid by new investors by \$14.0 million and, in the case of an increase, would increase the percentage of total

consideration paid by new investors by 4.2 percentage points and, in the case of a decrease, would decrease the percentage of total consideration paid by new investors by 4.8 percentage points, assuming no change in the assumed initial public offering price per share.

The table assumes no exercise of the underwriters' option to purchase additional shares in this offering. If the underwriters' option to purchase additional shares is exercised in full, the number of shares of our common stock held by existing stockholders would be reduced to 75.6% of the total number of shares of our common stock outstanding after this offering, and the number of shares of common stock held by new investors participating in the offering would be increased to 24.4% of the total number of shares outstanding after this offering.

The above discussion and tables are based on shares of common stock issued and outstanding as of March 31, 2018 and excludes:

- § 7,614 shares of common stock issuable upon the exercise of a warrant outstanding as of March 31, 2018, at an exercise price of \$3.94 per share;
- § 660,319 shares of common stock issuable upon the exercise of stock options outstanding as of March 31, 2018 under our 2017 Stock Option and Incentive Plan at an exercise price of \$5.77 per share;
- § 355,539 shares of common stock issuable upon the exercise of stock options granted subsequent to March 31, 2018 under our 2017 Stock Option and Incentive Plan at an exercise price of \$7.17 per share;
- § 3,139,274 shares of our common stock that will become available for future issuance under our 2018 Stock Option and Incentive Plan upon the effectiveness of the registration statement of which this prospectus forms a part, from which we have granted options to purchase 309,925 shares of common stock, subject to and effective upon the closing of this offering, at an exercise price equal to the public offering price per share in this offering; and
- § 235,743 shares of our common stock that will become available for future issuance under our 2018 Employee Stock Purchase Plan upon the effectiveness of the registration statement of which this prospectus forms a part.

New investors will experience further dilution if our outstanding warrant or stock options are exercised, new options or warrants are issued under our equity incentive plans or we issue additional shares of common stock, other equity securities or convertible debt securities in the future.

SELECTED CONSOLIDATED FINANCIAL DATA

We have derived the consolidated statement of operations data for the years ended December 31, 2016 and 2017 and the consolidated balance sheet data as of December 31, 2016 and 2017 from our audited consolidated financial statements included elsewhere in this prospectus. We have derived the consolidated statement of operations data for the three months ended March 31, 2017 and 2018 and the consolidated balance sheet data as of March 31, 2018 from our unaudited consolidated financial statements included elsewhere in this prospectus. In the opinion of management, the unaudited consolidated financial statements have been prepared on a basis consistent with our audited consolidated financial statements and contain all adjustments, consisting of only normal recurring adjustments, that management considers necessary for the fair presentation of the financial information set forth in those statements. You should read the following selected consolidated financial data together with our consolidated financial statements and the related notes included elsewhere in this prospectus and the "Management's Discussion and Analysis of Financial Condition and Results of Operations" section of this prospectus. The selected consolidated financial data contained in this section are not intended to replace our consolidated financial statements and the related notes. Our historical results are not necessarily indicative of the results that may be expected in the future, and the results for the three months ended March 31, 2018 are not necessarily indicative of the results to be expected for the full year ending December 31, 2018 or any other period.

	<u>Year Ended December 31,</u>		<u>Three Months Ended March 31,</u>	
	<u>2016</u>	<u>2017</u>	<u>2017</u>	<u>2018</u>
	<small>(in thousands, except unit, share, per unit and per share data)</small>			
Consolidated Statement of Operations Data:				
Collaboration revenue	\$ 379	\$ —	\$ —	\$ —
Operating expenses:				
Research and development	12,477	19,944	3,568	6,701
General and administrative	4,112	5,085	1,173	2,315
Total operating expenses	<u>16,589</u>	<u>25,029</u>	<u>4,741</u>	<u>9,016</u>
Loss from operations	(16,210)	(25,029)	(4,741)	(9,016)
Other income (expense):				
Interest income (expense), net	(19)	44	13	144
Other income (expense), net	22	(10)	(2)	(20)
Total other income	<u>3</u>	<u>34</u>	<u>11</u>	<u>124</u>
Net loss	<u>\$ (16,207)</u>	<u>\$ (24,995)</u>	<u>\$ (4,730)</u>	<u>\$ (8,892)</u>
Net loss per unit, basic and diluted ⁽¹⁾	<u>\$ (10.11)</u>		<u>\$ (2.95)</u>	
Net loss per share, basic and diluted ⁽¹⁾		<u>\$ (15.30)</u>		<u>\$ (3.18)</u>
Weighted average common units outstanding, basic and diluted ⁽¹⁾	<u>1,603,088</u>		<u>1,603,088</u>	
Weighted average common shares outstanding, basic and diluted ⁽¹⁾		<u>1,634,100</u>		<u>2,795,497</u>
Pro forma net loss per share, basic and diluted (unaudited) ⁽¹⁾		<u>\$ (2.06)</u>		<u>\$ (0.50)</u>
Pro forma weighted average common shares outstanding, basic and diluted (unaudited) ⁽¹⁾		<u>12,108,377</u>		<u>17,905,447</u>

⁽¹⁾ See Note 17 to our consolidated financial statements included elsewhere in this prospectus for further details on the calculation of basic and diluted net loss per unit and share and unaudited pro forma basic and diluted net loss per share.

	As of December 31,		As of
	2016	2017	March 31,
	(in thousands)		2018
Consolidated Balance Sheet Data:			
Cash, cash equivalents and marketable securities	\$ 29,531	\$ 57,959	\$ 47,763
Total assets	32,782	61,637	54,159
Convertible preferred units	58,057	—	—
Convertible preferred stock	—	109,232	109,232
Accumulated deficit	(32,530)	(57,525)	(66,417)
Total stockholders' deficit	(30,027)	(53,522)	(61,931)

**MANAGEMENT'S DISCUSSION AND ANALYSIS OF FINANCIAL CONDITION
AND RESULTS OF OPERATIONS**

You should read the following discussion and analysis of our financial condition and results of operations together with the "Selected Consolidated Financial Data" section of this prospectus and our consolidated financial statements and related notes appearing elsewhere in this prospectus. Some of the information contained in this discussion and analysis or set forth elsewhere in this prospectus, including information with respect to our plans and strategy for our business and related financing, includes forward-looking statements that involve risks and uncertainties. As a result of many factors, including those factors set forth in the "Risk Factors" section of this prospectus, our actual results could differ materially from the results described in or implied by the forward-looking statements contained in the following discussion and analysis.

Overview

We are a biopharmaceutical company focused on the discovery and development of innovative medicines for the treatment of serious diseases in which signaling by protein growth factors plays a fundamental role. Our newly elucidated understanding of the molecular mechanisms of growth factor activation enabled us to develop a proprietary platform for the discovery and development of monoclonal antibodies that locally and selectively target these signaling proteins at the cellular level. We believe this approach, acting in the disease microenvironment, avoids the historical challenges associated with inhibiting growth factors for therapeutic effect. We believe our focus on biologically validated growth factors may facilitate a more efficient development path. We are advancing our lead product candidate, SRK-015, a selective first-in-class inhibitor of the activation of the growth factor myostatin in skeletal muscle, into clinical development for the treatment of spinal muscular atrophy, or SMA. In March 2018, we filed an Investigational New Drug application, or IND, with the U.S. Food and Drug Administration, or FDA, for SRK-015, and in April 2018, the FDA notified us that our Phase 1 first-in-human clinical trial of SRK-015 may proceed. We plan to initiate our Phase 1 clinical trial in May 2018. The FDA has granted orphan drug designation for SRK-015 for the treatment of SMA. In addition, utilizing our proprietary platform, we are also creating a pipeline of novel product candidates with the potential to transform the lives of patients suffering from a wide range of serious diseases, including other neuromuscular disorders, cancer, fibrosis and anemia.

As more fully described in the section of this prospectus titled "Reorganization," on December 22, 2017, we completed a series of transactions pursuant to which Scholar Rock Merger Sub, LLC, a wholly owned subsidiary of Scholar Rock Holding Corporation, was merged with and into Scholar Rock, LLC. As part of the transactions, all convertible preferred units and common units of Scholar Rock, LLC issued and outstanding immediately prior to the Reorganization were exchanged for shares of Scholar Rock Holding Corporation capital stock of the same class or series on a one-for-one basis. Previously outstanding vested and unvested incentive units, irrespective of any strike price or voting rights, were exchanged for an equal number of shares of common stock or restricted common stock, respectively. The restricted common stock was issued with the same vesting terms as the unvested incentive units held immediately prior to the Reorganization. Upon consummation of the Reorganization, the historical consolidated financial statements of Scholar Rock, LLC became the historical consolidated financial statements of Scholar Rock Holding Corporation, the entity whose shares are being offered in this offering. Except as otherwise indicated or the context otherwise requires, all information included in this prospectus is presented giving effect to the Reorganization.

Since inception, our operations have focused on organizing and staffing our company, business planning, raising capital, establishing our intellectual property portfolio and performing research and development of monoclonal antibodies that selectively inhibit activation of growth factors for therapeutic effect. Revenue generation activities have been limited to research services and the issuance of a license, in each case,

pursuant to an option and license agreement with Janssen Biotech, Inc., or Janssen, a subsidiary of Johnson & Johnson. We do not have any products approved for sale and have not generated any revenue from product sales. We have funded our operations primarily through private placements of our convertible preferred stock and borrowings under a loan and security agreement, or the credit facility, with Silicon Valley Bank, or SVB. From inception through March 31, 2018, we have raised an aggregate of \$111.2 million of proceeds through the issuance of equity and debt to fund our operations, of which \$109.2 million was from the issuance of convertible preferred stock and \$2.0 million was from borrowings under the credit facility.

Since inception, we have incurred significant operating losses. Our net losses were \$16.2 million, \$25.0 million and \$8.9 million for the years ended December 31, 2016 and 2017 and the three months ended March 31, 2018, respectively. As of March 31, 2018, we had an accumulated deficit of \$66.4 million. We expect to continue to incur significant expenses and operating losses for the foreseeable future. In addition, we anticipate that our expenses will increase significantly in connection with our ongoing activities, as we:

- § continue activities in support of our planned Phase 1 first-in-human clinical trial for SRK-015, our lead product candidate, which we plan to initiate in May 2018;
- § continue to discover, validate and develop additional product candidates including from our program focused on inhibitors of the activation of transforming growth factor beta 1, or TGFb1;
- § maintain, expand and protect our intellectual property portfolio;
- § hire additional research, development and business personnel; and
- § prepare and begin to operate as a public company upon the completion of this offering.

We will not generate revenue from product sales unless and until we successfully complete clinical development and obtain regulatory approval for SRK-015 or any of our future product candidates. In addition, if we obtain regulatory approval for SRK-015 or any of our future product candidates and do not enter into a third-party commercialization partnership, we expect to incur significant expenses related to developing our commercialization capability to support product sales, marketing and distribution activities.

As a result, we will need substantial additional funding to support our continuing operations and pursue our growth strategy. Until such time as we can generate significant revenue from product sales, if ever, we expect to finance our operations through a combination of public or private equity offerings and debt financings, government funding arrangements, collaborations, strategic alliances and marketing, distribution or licensing arrangements. We may be unable to raise additional funds or enter into such other agreements or arrangements when needed on favorable terms, or at all. If we fail to raise capital or enter into such agreements as, and when, needed, we may have to significantly delay, scale back or discontinue the development and commercialization of one or more of our product candidates.

Because of the numerous risks and uncertainties associated with pharmaceutical product development, we are unable to accurately predict the timing or amount of increased expenses or when or if we will be able to achieve or maintain profitability. Even if we are able to generate product sales, we may not become profitable. If we fail to become profitable or are unable to sustain profitability on a continuing basis, then we may be unable to continue our operations at planned levels and be forced to reduce or terminate our operations.

As of March 31, 2018, we had cash, cash equivalents and marketable securities of \$47.8 million. We believe that the net proceeds from this offering, together with our existing cash, cash equivalents and marketable securities, will enable us to fund our operating expenses and capital expenditure requirements into the second half of 2020.

Financial Operations Overview

Collaboration Revenue

We do not have any products approved for sale, and as a result, we have not generated any revenue from product sales and do not expect to generate any revenue from the sale of products in the foreseeable future. We cannot predict if, when, or to what extent we will generate revenue from the commercialization and sale of SRK-015 or any of our future product candidates. We may never succeed in obtaining regulatory approval for SRK-015 or any of our future product candidates.

To date, all of our revenue has been derived from our option and license agreement with Janssen. We expect that our revenue for the next several years will be derived primarily from payments under our option and license agreement with Janssen or other collaboration and license agreements that we may enter into in the future, if any.

Expenses

Research and Development

Research and development expenses consist primarily of costs incurred for our research and development activities, including our product candidate discovery efforts, and preclinical studies and clinical trials under our research programs, which include:

- § employee-related expenses, including salaries, benefits and equity-based compensation expense for our research and development personnel;
- § costs of funding research performed by third parties that conduct research and development and preclinical activities on our behalf;
- § cost of manufacturing clinical supply related to SRK-015 and any of our future product candidates;
- § cost of conducting clinical trials of SRK-015 and any of our future product candidates;
- § consulting and professional fees related to research and development activities, including equity-based compensation to non-employees;
- § costs of purchasing laboratory supplies and non-capital equipment used in our preclinical studies;
- § costs related to compliance with clinical regulatory requirements;
- § facility costs and other allocated expenses, which include expenses for rent and maintenance of facilities, insurance, depreciation and other supplies; and
- § fees for maintaining licenses and other amounts due under our third-party licensing agreements.

Research and development costs are expensed as incurred. Costs for certain activities are recognized based on an evaluation of the progress to completion of specific tasks. Nonrefundable advance payments for research and development goods and services to be received in the future from third parties are deferred and capitalized. The capitalized amounts are expensed as the related services are performed.

The successful development of SRK-015 and any future product candidates is highly uncertain. As such, at this time, we cannot reasonably estimate or know the nature, timing and estimated costs of the efforts that will be necessary to complete the remainder of the development of SRK-015 and any future product candidates. We are also unable to predict when, if ever, material net cash inflows will commence from the sale of our product candidates, if approved. This is due to the numerous risks and uncertainties associated with developing product candidates, including the uncertainty of:

- § establishing an appropriate safety profile;
- § successful enrollment in and completion of clinical trials;
- § whether our product candidates show safety and efficacy in our clinical trials;
- § receipt of marketing approvals from applicable regulatory authorities, if any;
- § establishing commercial manufacturing capabilities or making arrangements with third-party manufacturers;

- § obtaining and maintaining patent and trade secret protection and regulatory exclusivity for our product candidates;
- § commercializing the product candidates, if and when approved, whether alone or in collaboration with others; and
- § continued acceptable safety profile of the products following any regulatory approval.

A change in the outcome of any of these variables with respect to the development of SRK-015 or any of our future product candidates would significantly change the costs and timing associated with the development of that product candidate.

Research and development activities are central to our business model. Product candidates in later stages of clinical development generally have higher development costs than those in earlier stages of clinical development, primarily due to the increased size and duration of later-stage clinical trials. We expect research and development costs to increase significantly for the foreseeable future as our product candidate development programs progress. However, we do not believe that it is possible at this time to accurately project total program-specific expenses through commercialization. There are numerous factors associated with the successful commercialization of any of our product candidates, including future trial design and various regulatory requirements, many of which cannot be determined with accuracy at this time based on our stage of development. Additionally, future commercial and regulatory factors beyond our control will impact our clinical development programs and plans.

General and Administrative

General and administrative expenses consist primarily of employee-related expenses, including salaries, benefits and equity-based compensation expenses for personnel in executive, finance, accounting, business development, legal and human resources functions. Other significant general and administrative expenses include facility costs not otherwise included in research and development expenses, legal fees relating to patent and corporate matters and fees for accounting and consulting services.

We anticipate that our general and administrative expenses will increase in the future as our business expands to support expected growth in research and development activities, including the initiation of our planned Phase 1 clinical program from SRK-015 and any future clinical programs. These increases will likely include increased costs related to the hiring of additional personnel and fees to outside consultants, among other expenses. We also anticipate increased expenses associated with being a public company, including costs for audit, legal, regulatory and tax-related services, director and officer insurance premiums and investor relations costs. In addition, if we obtain regulatory approval for any of our product candidates and do not enter into a third-party commercialization collaboration, we expect to incur significant expenses related to building a sales and marketing team to support product sales, marketing and distribution activities.

Interest Income (Expense), Net

Interest income (expense), net consists primarily of interest expense incurred on our credit facility, including amortization of debt discount and debt issuance costs, and interest income earned on our cash, cash equivalents and marketable securities.

Other Income (Expense), Net

Other income (expense), net consists primarily of non-cash changes in the fair value of warrants issued in connection with our credit facility and a gain recorded on the sale of fixed assets which was recorded in 2016.

Results of Operations

Comparison of the Three Months Ended March 31, 2017 and 2018

The following table summarizes our results of operations for the three months ended March 31, 2017 and 2018 (in thousands, except percentages):

	Three Months Ended March 31,		Change	
	2017	2018	\$	%
Operating expenses:				
Research and development	\$ 3,568	\$ 6,701	\$ 3,133	88%
General and administrative	1,173	2,315	1,142	97%
Total operating expenses	4,741	9,016	4,275	90%
Loss from operations	(4,741)	(9,016)	(4,275)	90%
Other income (expense):				
Interest income (expense), net	13	144	131	*NM
Other income (expense), net	(2)	(20)	(18)	*NM
Total other income	11	124	113	*NM
Net loss	\$ (4,730)	\$ (8,892)	\$ (4,162)	88%

* NM means not meaningful.

Research and Development

Research and development expense increased by \$3.1 million from \$3.6 million for the three months ended March 31, 2017 to \$6.7 million for the three months ended March 31, 2018, an increase of 88%. The following table summarizes our research and development expense for the three months ended March 31, 2017 and 2018 (in thousands, except percentages):

	Three Months Ended March 31,		Change	
	2017	2018	\$	%
External costs by program				
SRK-015	\$ 339	\$ 1,995	\$ 1,656	488%
Other early development candidates and unallocated costs	884	1,295	411	46%
Total external costs	1,223	3,290	2,067	169%
Internal costs:				
Employee compensation and benefits	1,415	2,138	723	51%
Facility and other	930	1,273	343	37%
Total internal costs	2,345	3,411	1,066	45%
Total research and development expense	\$ 3,568	\$ 6,701	\$ 3,133	88%

The increase in research and development expense was primarily attributable to the following:

- § The \$2.1 million increase in external costs primarily related to increased research and preclinical development and manufacturing costs associated with our lead product candidate, SRK-015, and other external research costs associated with our other early development candidates.

§ The \$1.0 million increase in internal costs was primarily driven by an increase in employee compensation and benefits costs related to increased headcount in our research and development function.

A significant portion of our research and development costs have been external costs, which we track on a program-by-program basis after a clinical product candidate has been identified. Our internal research and development costs are primarily personnel-related costs, depreciation and other indirect costs. We do not track our internal research and development expenses on a program-by-program basis as they are deployed across multiple projects under development.

General and Administrative

General and administrative expense increased by \$1.1 million from \$1.2 million for the three months ended March 31, 2017 to \$2.3 million for the three months ended March 31, 2018.

The increase in general and administrative expense was primarily attributable to an increase of \$0.3 million in employee compensation and benefits related to increased headcount and an increase of \$0.8 million in professional services and consulting fees primarily related to increases in legal fees related to business development, regulatory and patent costs, accounting and audit fees and public and investor relations fees due to ongoing business activities.

Interest Income (Expense), Net

The increase in interest income, net was attributable to increased income earned on our investment portfolio, which was significantly larger during the three months ended March 31, 2018 as compared to the three months ended March 31, 2017.

Comparison of the Years Ended December 31, 2016 and 2017

The following table summarizes our results of operations for the years ended December 31, 2016 and 2017 (in thousands, except percentages):

	Year Ended December 31,		Change	
	2016	2017	\$	%
Collaboration revenue	\$ 379	\$ —	\$ (379)	(100)%
Operating expenses:				
Research and development	12,477	19,944	7,467	60%
General and administrative	4,112	5,085	973	24%
Total operating expenses	16,589	25,029	8,440	51%
Loss from operations	(16,210)	(25,029)	(8,819)	54%
Other income (expense):				
Interest income (expense), net	(19)	44	63	332%
Other income (expense), net	22	(10)	(32)	(145)%
Total other income	3	34	31	*NM
Net loss	\$ (16,207)	\$ (24,995)	\$ (8,788)	54%

* NM means not meaningful.

Collaboration Revenue

Collaboration revenue decreased by \$0.4 million from \$0.4 million for the year ended December 31, 2016 to \$0 for the year ended December 31, 2017. We completed our performance obligations related to

conducting research services to identify molecules with either one of two pharmacological profiles under the option and license agreement with Janssen in 2016, and recorded \$0.4 million of collaboration revenue related to these activities during that period. No milestones were achieved in 2017.

Research and Development

Research and development expense increased by \$7.5 million from \$12.5 million for the year ended December 31, 2016 to \$19.9 million for the year ended December 31, 2017, an increase of 60%. The following table summarizes our research and development expense for the years ended December 31, 2016 and 2017 (in thousands, except percentages):

	Year Ended December 31,		Change	
	2016	2017	\$	%
External costs by program:				
SRK-015	\$ 3,113	\$ 6,513	\$ 3,400	109%
Other early development candidates and unallocated costs	1,361	3,025	1,664	122%
Total external costs	4,474	9,538	5,064	113%
Internal costs:				
Employee compensation and benefits	4,760	6,409	1,649	35%
Facility and other	3,243	3,997	754	23%
Total internal costs	8,003	10,406	2,403	30%
Total research and development expense	\$ 12,477	\$ 19,944	\$ 7,467	60%

The increase in research and development expense was primarily attributable to the following:

- § The \$5.1 million increase in external costs primarily related to increased research and preclinical development and manufacturing costs associated with our lead product candidate, SRK-015 and other external research costs associated with our other early development candidates.
- § The \$2.4 million increase in internal costs was primarily driven by an increase in employee compensation and benefits costs related to increased headcount in our research and development function.

A significant portion of our research and development costs have been external costs, which we track on a program-by-program basis after a clinical product candidate has been identified. Our internal research and development costs are primarily personnel-related costs, depreciation and other indirect costs. We do not track our internal research and development expenses on a program-by-program basis as they are deployed across multiple projects under development.

General and Administrative

General and administrative expense increased by \$1.0 million from \$4.1 million for the year ended December 31, 2016 to \$5.1 million for the year ended December 31, 2017.

The increase in general and administrative expense was primarily attributable to an increase of \$0.6 million in employee compensation and benefits due to increased headcount and an increase of \$0.3 million in professional services and consulting fees primarily due to increases in legal fees related to business development, regulatory and patent costs, accounting and audit fees and public and investor relations fees due to ongoing business activities.

Interest Income (Expense), Net

The increase in interest income (expense), net was attributable to increased income earned on our investment portfolio, which increased significantly year-over-year.

Liquidity and Capital Resources**Sources of Liquidity**

We have funded our operations from inception through March 31, 2018 with the net proceeds of \$109.2 million from sales of our convertible preferred stock and borrowings of \$2.0 million under our credit facility with SVB. The following table provides information regarding our total cash, cash equivalents and marketable securities at December 31, 2016 and 2017 and March 31, 2018 (in thousands):

	December 31,		March 31,
	2016	2017	2018
Cash and cash equivalents	\$ 10,033	\$ 56,461	\$ 22,822
Marketable securities	19,498	1,498	24,941
Total cash, cash equivalents and marketable securities	\$ 29,531	\$ 57,959	\$ 47,763

In August 2015, we entered into the credit facility with SVB for an equipment line of credit of up to \$2.0 million to finance the purchase of eligible equipment. Pursuant to the credit facility, SVB was obligated to make up to five equipment advances, each in an amount of at least \$100,000 during the draw period. In August 2016, we amended the credit facility to extend the draw period to December 31, 2016. We borrowed \$0.7 million against the line of credit in 2015 and \$1.3 million in 2016, which fulfilled the maximum credit line of \$2.0 million at December 31, 2016. Amounts borrowed bear interest at an annual prime rate less 0.25%. In the event of a default, and during such an event, the annual interest rate will increase by 5%. For each advance, interest-only payments were due and paid through June 2016. Principal and interest payments commenced on July 1, 2016 for a period of 36 months. A final payment fee equal to 4% of the aggregate advances is also due on June 1, 2019. We have the option to prepay the outstanding balance of the loan in full subject to a prepayment fee of 0.5% to 1.0%, depending on when the prepayment occurs. All borrowings under the credit facility mature on July 1, 2019. The loan balance at March 31, 2018 was \$0.9 million.

We granted SVB a security interest in all equipment financed under the credit facility. The credit facility contains negative covenants restricting our activities, including limitations on dispositions, change in business ownership or location, mergers or acquisitions, incurring indebtedness or liens, paying dividends or making investments and certain other business transactions.

We also issued a warrant to SVB to purchase 21,739 Series A-3 convertible preferred units at a purchase price of \$1.38 per unit, which became exercisable for 21,739 shares of Series A-3 convertible preferred stock at a purchase price of \$1.38 per share in connection with the Reorganization. The SVB warrant is exercisable immediately and expires on August 10, 2025. Following the completion of this offering, the warrant will be exercisable for 7,614 shares of our common stock at an exercise price of \$3.94 per share.

Cash Flows

The following table provides information regarding our cash flows for the years ended December 31, 2016 and 2017 and for the three months ended March 31, 2017 and 2018 (in thousands):

	Year Ended December 31,		Three Months Ended March 31,	
	2016	2017	2017	2018
Net cash used in operating activities	\$ (15,141)	\$ (21,737)	\$ (4,575)	\$ (9,884)
Net cash (used in) provided by investing activities	(20,319)	17,665	1,368	(23,588)
Net cash (used in) provided by financing activities	1,002	50,500	(167)	(167)
Net (decrease) increase in cash and cash equivalents and restricted cash	\$ (34,458)	\$ 46,428	\$ (3,374)	\$ (33,639)

Net Cash Used in Operating Activities

The use of cash in all periods resulted primarily from our net losses adjusted for non-cash charges and changes in components of working capital. Net cash used in operating activities was \$9.9 million for the three months ended March 31, 2018 compared to \$4.6 million for the three months ended March 31, 2017. The increase in cash used in operating activities was primarily due to an increase in net loss of \$4.2 million and an increase of \$1.4 million in cash used by operating assets and liabilities for the three months ended March 31, 2018 as compared to the three months ended March 31, 2017.

Net cash used in operating activities was \$21.7 million for the year ended December 31, 2017 compared to \$15.1 million for the year ended December 31, 2016. The increase in cash used in operating activities was due to an increase in net loss of \$8.8 million for the year ended December 31, 2017 as compared to the year ended December 31, 2016 partially offset by \$1.1 million of cash provided by operating assets and liabilities.

Net Cash (Used in) Provided by Investing Activities

Net cash used in investing activities was \$(23.6) million for the three months ended March 31, 2018 compared to net cash provided by investing activities of \$1.4 million for the three months ended March 31, 2017. Net cash used in investing activities for the three months ended March 31, 2018 consisted primarily of the purchase of marketable securities. Net cash provided by investing activities for the three months ended March 31, 2017 consisted primarily of the maturity of marketable securities.

Net cash provided by investing activities was \$17.7 million for the year ended December 31, 2017 compared to net cash used in investing activities of \$(20.3) million for the year ended December 31, 2016. Net cash provided by investing activities for the year ended December 31, 2017 consisted primarily of the maturity of marketable securities. Net cash used in investing activities for the year ended December 31, 2016 consisted primarily of the purchase of marketable securities.

Net Cash (Used in) Provided by Financing Activities

Net cash used in financing activities was \$(0.2) million during both the three months ended March 31, 2018 and 2017 related to principal payments made on outstanding debt.

Net cash provided by financing activities was \$50.5 million during the year ended December 31, 2017 compared to \$1.0 million during the year ended December 31, 2016. The cash provided by financing activities for the year ended December 31, 2017 was primarily the result of \$51.2 million of net proceeds received from private placements of our convertible preferred stock. The cash provided by financing activities for the year ended December 31, 2016 was primarily the result of borrowings under the loan and security agreement.

Funding Requirements

We expect our expenses to increase in connection with our ongoing activities, particularly as we continue the research and development for, initiate later stage clinical trials for, and seek marketing approval for, SRK-015 and any of our future product candidates. In addition, if we obtain marketing approval for SRK-015 or any of our future product candidates, we expect to incur significant commercialization expenses related to product sales, marketing, manufacturing and distribution, which costs we might offset through entry into collaboration agreements with third parties. Furthermore, upon the completion of this offering, we expect to incur additional costs associated with operating as a public company. Accordingly, we will need to obtain substantial additional funding in connection with our continuing operations. If we are unable to raise capital when needed or on attractive terms, we would be forced to delay, reduce or eliminate our research and development programs or future commercialization efforts.

We expect that the net proceeds from this offering, together with our existing cash, cash equivalents and marketable securities will enable us to fund our operating expenses and capital expenditure requirements into the second half of 2020.

We have based this estimate on assumptions that may prove to be wrong, and we may use our available capital resources sooner than we currently expect. Our future capital requirements will depend on many factors, including:

- § the costs of conducting future clinical trials;
- § the costs of future manufacturing;
- § the scope, progress, results and costs of discovery, preclinical development, laboratory testing and clinical trials for other potential product candidates we may develop, if any;
- § the costs, timing and outcome of regulatory review of our product candidates;
- § our ability to establish and maintain collaborations on favorable terms, if at all;
- § the achievement of milestones or occurrence of other developments that trigger payments under any collaboration agreements we might have at such time;
- § the costs and timing of future commercialization activities, including product sales, marketing, manufacturing and distribution, for any of our product candidates for which we receive marketing approval;
- § the amount of revenue, if any, received from commercial sales of our product candidates, should any of our product candidates receive marketing approval;
- § the costs of preparing, filing and prosecuting patent applications, obtaining, maintaining and enforcing our intellectual property rights and defending intellectual property-related claims;
- § our headcount growth and associated costs as we expand our business operations and research and development activities; and
- § the cost of operating as a public company.

Identifying potential product candidates and conducting preclinical studies and clinical trials is a time-consuming, expensive and uncertain process that takes years to complete, and we may never generate the necessary data or results required to obtain marketing approval and achieve product sales. In addition, our product candidates, if approved, may not achieve commercial success. Our commercial revenues, if any, will be derived from sales of products that we do not expect to be commercially available for many years, if

at all. Accordingly, we will need to continue to rely on additional financing to achieve our business objectives. Adequate additional financing may not be available to us on acceptable terms, or at all.

Until such time, if ever, as we can generate substantial product revenues, we expect to finance our cash needs through a combination of equity offerings, debt financings, collaborations, strategic alliances and licensing arrangements. To the extent that we raise additional capital through the sale of equity or convertible debt securities, your ownership interests may be diluted, and the terms of these securities may include liquidation or other preferences that could adversely affect your rights as a common stockholder. Additional debt financing, if available, may involve agreements that include restrictive covenants that limit our ability to take specific actions, such as incurring additional debt, making capital expenditures or declaring dividends, that could adversely impact our ability to conduct our business.

If we raise funds through collaborations, strategic alliances or licensing arrangements with third parties, we may have to relinquish valuable rights to our technologies, future revenue streams, research programs or product candidates or to grant licenses on terms that may not be favorable to us. If we are unable to raise additional funds through equity or debt financings when needed, we may be required to delay, limit, reduce or terminate our product development or future commercialization efforts or grant rights to develop and market product candidates that we would otherwise prefer to develop and market ourselves.

Critical Accounting Policies and Use of Estimates

This management's discussion and analysis is based on our consolidated financial statements, which have been prepared in accordance with U.S. generally accepted accounting principles. The preparation of these consolidated financial statements requires us to make judgments and estimates that affect the reported amounts of assets, liabilities, revenues and expenses and the disclosure of contingent assets and liabilities in our consolidated financial statements. We base our estimates on historical experience, known trends and events and various other factors that we believe to be reasonable under the circumstances. Actual results may differ from these estimates under different assumptions or conditions. On an ongoing basis, we evaluate our judgments and estimates in light of changes in circumstances, facts and experience. The effects of material revisions in estimates, if any, will be reflected in the consolidated financial statements prospectively from the date of change in estimates.

While our significant accounting policies are described in more detail in the notes to our consolidated financial statements appearing elsewhere in this prospectus, we believe the following accounting policies used in the preparation of our consolidated financial statements require the most significant judgments and estimates.

Accrued Research and Development Expenses

As part of the process of preparing our consolidated financial statements, we are required to estimate our accrued expenses as of each balance sheet date. This process involves reviewing open contracts and purchase orders, communicating with our personnel to identify services that have been performed on our behalf and estimating the level of service performed and the associated cost incurred for the service when we have not yet been invoiced or otherwise notified of the actual cost. The majority of our service providers invoice us monthly in arrears for services performed or when contractual milestones are met. We make estimates of our accrued expenses as of each balance sheet date based on facts and circumstances known to us at that time. We periodically confirm the accuracy of our estimates with the service providers and make adjustments if necessary. The significant estimates in our accrued research and development expenses include the costs incurred for services performed by our vendors in connection with research and development activities for which we have not yet been invoiced. In certain instances, we prepay for services to be provided in the future. These amounts are expensed as the services are performed.

We base our expenses related to research and development activities on our estimates of the services received and efforts expended pursuant to quotes and contracts with vendors that conduct research and development on our behalf. The financial terms of these agreements are subject to negotiation, vary from

contract to contract and may result in uneven payment flows. There may be instances in which payments made to our vendors will exceed the level of services provided and result in a prepayment of the research and development expense. In accruing service fees, we estimate the time period over which services will be performed and the level of effort to be expended in each period. If the actual timing of the performance of services or the level of effort varies from our estimate, we adjust the accrual or prepaid balance accordingly. Nonrefundable advance payments for goods and services that will be used in future research and development activities are expensed when the activity has been performed or when the goods have been received rather than when the payment is made.

Although we do not expect our estimates to be materially different from amounts incurred, if our estimates of the status and timing of services performed differ from the actual status and timing of services performed, it could result in us reporting amounts that are too high or too low in any particular period. To date, there have been no material differences between our estimates of such expenses and the amounts incurred.

Equity-Based Compensation

Prior to the Reorganization, our former parent company, Scholar Rock, LLC, granted incentive units, which we accounted for as equity-classified awards. As part of the Reorganization, the incentive units were exchanged for shares of our common stock and restricted common stock, which we account for as equity-classified awards. In 2018, we granted stock options, which we account for as equity-classified awards.

We measure employee equity-based compensation based on the grant date fair value of the equity-based awards and recognize equity-based compensation expense on a straight-line basis over the requisite service period of the awards, which is generally the vesting period of the respective award. For awards subject to performance conditions, we recognize equity-based compensation expense using an accelerated recognition method over the remaining period when we determine that achievement of the milestone is probable. As of January 1, 2016, we made an accounting policy election to recognize forfeitures as they occur upon adoption of guidance per Accounting Standard Update ("ASU") No. 2016-09, *Compensation — Stock Compensation*, ("ASU 2016-09"). The adoption of ASU 2016-09 did not have a material impact on our consolidated financial statements. The term "forfeitures" is distinct from "cancellations" or "expirations" and represents only the unvested portion of the surrendered equity-based award.

We recognize compensation expense for equity-based awards granted to non-employees over the related service period of the award. The fair value of the non-employee equity-based awards are subject to re-measurement at each reporting period prior to vesting, using the then-current fair value of the incentive units, common stock, and updated assumption inputs in the Black-Scholes option-pricing model, as applicable.

We classify equity-based compensation expense in our consolidated statement of operations in the same manner in which the award recipient's salary and related costs are classified or in which the award recipient's service payments are classified. In future periods, we expect equity-based compensation expense to increase, due in part to our existing unrecognized stock-based compensation expense and as we grant additional stock-based awards to continue to attract and retain our employees.

Determination of the Fair Value of Equity-Based Awards

We determine the fair value of restricted common stock awards granted based on the fair value of our common stock less any purchase price, as applicable. We estimate the fair value of stock option awards granted using the Black-Scholes option-pricing model, which uses as inputs the fair value of our common stock and subjective assumptions we make, including the expected stock price volatility, the expected term of the award, the risk-free interest rate and expected dividends. Due to the lack of a public market for the trading of our common stock and a lack of company-specific historical and implied volatility data, we base the estimate of expected volatility on the historical volatility of a representative group of publicly traded companies for which historical information is available. The historical volatility is generally calculated based

on a period of time commensurate with the expected term assumption. We use the simplified method to calculate the expected term for options granted to employees and directors. We utilize this method as we do not have sufficient historical exercise data to provide a reasonable basis upon which to estimate the expected term. For options granted to non-employees, we utilize the contractual term of the arrangement as the basis for the expected term assumption. The risk-free interest rate is based on a U.S. treasury instrument whose term is consistent with the expected term of the stock options. The expected dividend yield is assumed to be zero as we have never paid dividends and do not have current plans to pay any dividends on our common stock.

As there has been no public market for our common units or incentive units to date, the estimated fair value of our common units and incentive units has been approved by our board of directors, with input from management, as of the date of each award grant, considering our most recently available independent third-party valuations of common units and incentive units and our board of directors assessment, with input from management, of additional objective and subjective factors that we believed were relevant and which may have changed from the date of the most recent valuation through the date of the grant. In addition, there has been no public market for our common stock to date. The estimated fair value of our common stock has been determined by our board of directors as of the date of each award grant considering our most recently available independent third-party valuations of common stock and our board of directors' assessment of additional objective and subjective factors that it believed were relevant and which may have changed from the date of the most recent valuation through the date of the grant. These independent third-party valuations were performed in accordance with the guidance outlined in the American Institute of Certified Public Accountants' Accounting and Valuation Guide, *Valuation of Privately-Held-Company Equity Securities Issued as Compensation*. We estimated the value of our equity using the market approach, including the guideline public company method and a precedent transaction method which "backsolves" to a preferred price. We allocated equity value to our common units, incentive units and convertible preferred units or to our shares of common stock and shares of our convertible preferred stock, as the case may be, using either an option-pricing method, or OPM, or a hybrid method, which is a hybrid between the OPM and the probability-weighted expected return method. The OPM treats common securities and preferred securities as call options on the total equity value of a company, with exercise prices based on the value thresholds at which the allocation among the various holders of a company's securities changes. Under this method, the common units and incentive units and common stock have value only if the funds available for distribution to members exceed the value of the preferred security liquidation preference at the time of the liquidity event, such as a strategic sale or a merger. The hybrid method estimates the probability-weighted value across multiple scenarios but uses the OPM to estimate the allocation of value within at least one of the scenarios. In addition to the OPM, the hybrid method considers an initial public offering, or IPO, scenario in which the shares of convertible preferred stock are assumed to convert to common stock. The future value of the common units, incentive units and common stock in the IPO scenario is discounted back to the valuation date at an appropriate risk adjusted discount rate. In the hybrid method, the present value indicated for each scenario is probability weighted to arrive at an indication of value for the common units, incentive units and common stock.

As of December 31, 2016, our third-party valuation report estimated a valuation of our common units of \$3.29 per unit, and our incentive units (with a strike price of \$2.23) of \$2.77 per unit. As of May 18, 2017, our third-party valuation report estimated a valuation of our common units of \$3.54 per unit, and our incentive units (with a strike price of \$2.23) of \$3.00 per unit. As of September 27, 2017, our third-party valuation report estimated a valuation of our common units of \$4.89 per unit, and our incentive units (with a strike price of \$2.77) of \$3.94 per unit. As of December 22, 2017, our third-party valuation report estimated a value of our common stock of \$5.77 per share. As of March 5, 2018, our third-party valuation report estimated a value of our common stock of \$7.17 per share.

In addition to considering the results of these third-party valuations, management considered various objective and subjective factors to determine the fair value of our common units, incentive units and

common stock as of each grant date, which may be a date later than the most recent third-party valuation date, including:

- § the prices of our preferred securities sold to or exchanged between outside investors in arm's length transactions, and the rights, preferences and privileges of our preferred securities as compared to those of our common units, incentive units or common stock, including the liquidation preferences of our preferred securities;
- § the progress of our research and development efforts, including the status of preclinical studies and planned clinical trials for our product candidates;
- § the lack of liquidity of our equity as a private company;
- § our stage of development and business strategy and the material risks related to our business and industry;
- § the achievement of enterprise milestones, including entering into collaboration and license agreements;
- § the valuation of publicly traded companies in the life sciences and biotechnology sectors, as well as recently completed mergers and acquisitions of peer companies;
- § any external market conditions affecting the biotechnology industry, and trends within the biotechnology industry;
- § the likelihood of achieving a liquidity event for the holders of our common units, incentive units and common stock, such as an IPO, or a sale of our company, given prevailing market conditions; and
- § the analysis of IPOs and the market performance of similar companies in the biopharmaceutical industry.

The assumptions underlying these valuations represent management's best estimates, which involve inherent uncertainties and the application of management judgment. As a result, if factors or expected outcomes change and we use significantly different assumptions or estimates, our equity-based compensation expense could be materially different. Following the completion of this offering, the fair value of our common stock will be determined based on the quoted market price of our common stock.

The following table sets forth by grant date and type of award, the number of incentive units or stock options granted; the per unit strike price of incentive units or the per share exercise price of stock options granted between January 1, 2016 and the date of this prospectus.

<u>Date of Issuance</u>	<u>Type of Award</u>	<u>Number of Units or Shares Subject to Awards Granted</u>	<u>Per Unit Strike Price or Per Share Exercise Price</u>	<u>Fair Value per Common Unit or Share on Grant Date</u>	<u>Per Unit or Share Estimated Fair Value of Awards on Grant Date⁽¹⁾</u>
April 25, 2016	Incentive unit	280,229	\$ 2.23	\$ 3.43	\$2.80
June 3, 2016	Incentive unit	1,751	\$ 2.23	\$ 3.43	\$2.80
August 12, 2016	Incentive unit	228,597	\$ 2.23	\$ 3.40	\$2.80
November 30, 2016	Incentive unit	242,398	\$ 2.23	\$ 3.32	\$2.77
February 14, 2017	Incentive unit	141,221	\$ 2.23	\$ 3.29	\$2.77
February 21, 2017	Incentive unit	14,712	\$ 2.23	\$ 3.29	\$2.77
June 2, 2017	Incentive unit	19,616	\$ 2.23	\$ 3.54	\$3.00
September 27, 2017	Incentive unit	365,455	\$ 2.77	\$ 4.89	\$3.94
October 26, 2017	Incentive unit	80,566	\$ 2.77	\$ 4.89	\$3.94
February 20, 2018 ⁽²⁾	Stock option	660,319	\$ 5.77	\$ 5.77	\$4.20 - \$4.86
April 3, 2018	Stock option	324,014	\$ 7.17	\$ 7.17	\$5.11
April 15, 2018	Stock option	31,525	\$ 7.17	\$ 7.17	\$5.11

⁽¹⁾ For the purposes of recording equity based compensation for grants of incentive units and common stock to non-employees, we measure the fair value of the award on the service completion date (vesting date). At the end of each reporting period prior to completion of the services, we re-measure the value of any unvested portion of the award based on the then-current fair value of the award and adjust expense accordingly.

⁽²⁾ Includes 607,681 and 33,377 stock options for employee service-based awards with a per share estimated grant date fair value of \$4.20 and \$4.25, respectively, and 19,261 stock options for employee performance-based awards and non-employee awards with a per share estimated grant date fair value of \$4.86.

Determination of Estimated Offering Price

We and our underwriters determined the estimated price range set forth on the cover of this preliminary prospectus, which is \$13.00 to \$15.00 per share. In comparison, our estimate of the fair value of our common units was \$4.89 per unit at September 27, 2017 and October 26, 2017, which was determined by our board of directors with the assistance of a third-party valuation of our common units as of September 27, 2017, \$5.77 per share of common stock at February 20, 2018, which was determined by our board of directors with the assistance of a third-party valuation of our common stock as of December 22, 2017, and \$7.17 per share of common stock at April 3, 2018 and April 15, 2018, which was determined by our board of directors with the assistance of a third-party valuation of our common stock as of March 5, 2018.

These valuations utilized the hybrid method described in "—Determination of Fair Value of Equity-Based Awards." The valuation for our September 27, 2017 and October 26, 2017 incentive unit grants attributed a 10% probability to an initial public offering, or IPO, scenario and a 90% probability of remaining private, and reflected a discount for lack of marketability of 15% and 28% to the IPO and remaining private scenarios, respectively. The valuation for our February 20, 2018 option grants attributed a 25% probability to an IPO scenario and a 75% probability of remaining private and reflected a discount for lack of marketability of 18% and 24% to the IPO and remaining private scenarios, respectively. The valuation for our April 3, 2018 and April 15, 2018 option grants attributed a 40% probability to an IPO and a 60%

probability of remaining private and reflected a discount for lack of marketability of 10% and 29% to the IPO and remaining private scenarios, respectively. In addition to quantitative analysis from third-party valuations of our common stock, we also considered macro-economic and market conditions, including our subjective assessment of market conditions for initial public offerings of companies similarly situated to ours and our subjective assessment as to the likelihood of successfully executing an initial public offering in the coming months, among other factors.

We note that, as is typical in initial public offerings, the estimated price range for this offering was not derived using a formal determination of fair value, but was determined based upon discussions between us and the underwriters. Among the factors considered in setting the estimated range were prevailing market conditions, estimates of our business potential, progress in our research and development programs and developments in our business, the general condition of the securities market and the market prices of, and demand for, publicly-traded common stock of generally comparable companies.

Revenue Recognition

As of March 31, 2018, all of our revenue to date had been generated exclusively from our option and license agreement with Janssen. Effective January 1, 2018, we adopted the provisions of ASC Topic 606, *Revenue from Contracts with Customers* ("ASC 606") using the full retrospective transition method. Under this method, we revised our consolidated financial statements for the years ended December 31, 2016 and 2017, and all applicable interim periods within those years, as if ASC 606 had been effective for those periods.

Under ASC 606, we recognize revenue when our customer obtains control of promised goods or services, in an amount that reflects the consideration which we expect to receive in exchange for those goods or services. To determine the appropriate amount of revenue to be recognized for arrangements determined to be within the scope of ASC 606, we perform the following five steps: (i) identification of the contract(s) with the customer, (ii) identification of the promised goods or services in the contract and determination of whether the promised goods or services are performance obligations, (iii) measurement of the transaction price, (iv) allocation of the transaction price to the performance obligations, and (v) recognition of revenue when (or as) we satisfy each performance obligation. We only apply the five-step model to contracts when it is probable that we will collect the consideration we are entitled to in exchange for the goods or services we transfer to our customer.

Identification of the Contract(s) with the Customer

We account for a contract with a customer that is within the scope of ASC 606 when all of the following criteria are met: (i) the arrangement has been approved by the parties and the parties are committed to perform their respective obligations, (ii) each party's rights regarding the goods or services to be transferred can be identified, (iii) the payment terms for the goods or services to be transferred can be identified, (iv) the arrangement has commercial substance and (v) collection of substantially all of the consideration to which we will be entitled in exchange for the goods or services that will be transferred to the customer is probable.

Identification of the Performance Obligations

The promised goods or services in our option and license arrangement consist of license rights to our intellectual property and research and development services. The arrangement also has options for additional items (i.e., additional license rights). Options are considered to be marketing offers and are to be accounted for as separate contracts when the customer elects such options, unless we determine the option provides a material right which would not be provided without entering into the contract. Performance obligations are promised goods or services in a contract to transfer a distinct good or service to the customer. Promised goods or services are considered distinct when: (i) the customer can benefit from the good or service on its own or together with other readily available resources and (ii) the promised good or

service is separately identifiable from other promises in the contract. In assessing whether promised goods or services are distinct, we consider factors such as the stage of development of the underlying intellectual property, the capabilities of our customer to develop the intellectual property on their own and whether the required expertise is readily available.

Determination of the Transaction Price

We estimate the transaction price based on the amount of consideration we expect to receive for transferring the promised goods or services in the contract. The consideration may include both fixed consideration and variable consideration. At the inception of each arrangement that includes variable consideration, we evaluate the amount of the potential payments and the likelihood that the payments will be received. We utilize either the most likely amount method or expected value method to estimate the transaction price based on which method better predicts the amount of consideration expected to be received. If it is probable that a significant revenue reversal would not occur, the variable consideration is included in the transaction price.

Our arrangement includes development and regulatory milestone payments. We evaluate whether the milestones are considered probable of being reached and estimate the amount to be included in the transaction price using the most likely amount method. If it is probable that a significant revenue reversal would not occur, the associated milestone value is included in the transaction price. Milestone payments that are not within our control or the licensee's control, such as regulatory approvals, are not considered probable of being achieved until those approvals are received. At the end of each reporting period, we re-evaluate the probability of achievement of such milestones and any related constraint, and if necessary, adjust our estimate of the overall transaction price. Any such adjustments are recorded on a cumulative catch-up basis, which would affect collaboration revenue and earnings in the period of adjustment. To date, no milestones have been achieved under our arrangements with customers.

For sales-based royalties, including milestone payments based on the level of sales, we determine whether the sole or predominant item to which the royalties relate is a license. When the license is the sole or predominant item to which the sales-based royalty relates, we recognize revenue at the later of: (i) when the related sales occur, or (ii) when the performance obligation to which some or all of the royalty has been allocated has been satisfied (or partially satisfied). To date, we have not recognized any sales-based royalty revenue resulting from our arrangement.

Allocation of Transaction Price

We allocate the transaction price based on the estimated standalone selling price. We must develop assumptions that require judgment to determine the standalone selling price for each performance obligation identified in the contract. We utilize key assumptions to determine the standalone selling price, which may include other comparable transactions, pricing considered in negotiating the transaction and the estimated costs. Certain variable consideration is allocated specifically to one or more performance obligations in a contract when the terms of the variable consideration relate to the satisfaction of the performance obligation and the resulting amounts allocated to each performance obligation are consistent with the amounts we would expect to receive for satisfying each performance obligation.

Recognition of Revenue

For performance obligations which consist of licenses and other promises, we utilize judgment to assess the nature of the combined performance obligation in order to determine whether the combined performance obligation is satisfied over time or at a point in time. We determine the appropriate method of measuring progress of combined performance obligations satisfied over time for purposes of recognizing revenue. We evaluate the measure of progress each reporting period and, if necessary, adjust the measure of performance and related revenue recognition. If the license to our intellectual property is determined to be

distinct from the other performance obligations identified in the arrangement, we will recognize revenue from non-refundable, up-front fees allocated to the license when the license is transferred to the customer and the customer is able to use and benefit from the license.

We receive payments from customers based on billing schedules established in each contract. Up-front payments and fees are recorded as deferred revenue upon receipt or when due until we perform our obligations under these arrangements. Amounts are recorded as accounts receivable when our right to consideration is unconditional.

Off-Balance Sheet Arrangements

We did not have during the periods presented, and we do not currently have, any off-balance sheet arrangements, as defined under applicable SEC rules.

Contractual Obligations

The following table summarizes our significant contractual obligations by period presented according to the payment due date at December 31, 2017 (in thousands):

	Total	Less than 1 Year	1 to 3 Years	3 to 5 Years	More than 5 Years
Credit facility ⁽¹⁾	\$ 1,141	\$ 692	\$ 449	\$ —	\$ —
Operating lease obligations ⁽²⁾	2,009	696	1,313	—	—
Purchase obligation with a third party contract manufacturer ⁽³⁾	638	638	—	—	—
Total	\$ 3,788	\$ 2,026	\$ 1,762	\$ —	\$ —

⁽¹⁾ Consists of repayment obligation under the credit facility with SVB, including interest.

⁽²⁾ Represents future minimum repayments under our non-cancellable operating leases as of December 31, 2017, including our facility lease for the remaining term prior to the February 2018 amendment, and our equipment lease.

⁽³⁾ We are required to make certain minimum payments to a third party contract manufacturer. The amounts included in the table above represent the minimum contractual payments in excess of payments made by us as of December 31, 2017.

Under various licensing and related agreements with third parties, we have agreed to make milestone payments and pay royalties to third parties. Pursuant to an exclusive license agreement with Children's Medical Center Corporation, or CMCC, a holder of our common stock, we paid CMCC an annual license maintenance fee of \$5,000 in each of 2015 and 2016. Beginning in 2017, this obligation increased to \$10,000 per year, and continues until the agreement is terminated. We will also be responsible for up to \$1.3 million of development milestone payments through the first regulatory approval of a licensed product, tiered royalty payments of low single-digit percentages on net sales of licensed products in the event that we realize sales from products covered by the license agreement and between 10% and 20% of non-royalty income attributable to a sublicense of the CMCC rights. Products covered by the license agreement include products developed using our proprietary platform that are covered by a valid claim contained in any patent under the license agreement. Amounts paid to CMCC are recorded as research and development expense in the statements of operations.

We enter into agreements in the normal course of business with vendors for preclinical studies, preclinical and clinical supply and manufacturing services, professional consultants for expert advice and other vendors for other services for operating purposes. We have not included these payments in the table of contractual obligations above since the contracts do not contain any minimum purchase commitments and are cancelable at any time by us, generally upon 30 days prior written notice, and therefore we believe that our non-cancelable obligations under these agreements are not material.

Quantitative and Qualitative Disclosures About Market Risks

We are exposed to market risk related to changes in interest rates. Our primary exposure to market risk is interest rate sensitivity, which is affected by changes in the general level of U.S. interest rates, particularly because our cash equivalents are in the form of a money market fund, which is primarily invested in short-term U.S. Treasury obligations, and our marketable securities consist of U.S. Treasury obligations that have contractual maturities of less than one year.

Our primary exposure to market risk is interest income sensitivity, which is affected by changes in the general level of U.S. interest rates. However, because of the short-term nature of the investments in our portfolio, an immediate one percentage point change in market interest rates would not have a material impact on the fair market value of our investment portfolio or on our financial position or results of operations.

We are not currently exposed to significant market risk related to changes in foreign currency exchange rates; however, we have contracted with and may continue to contract with foreign vendors that are located in Europe. Our operations may be subject to fluctuations in foreign currency exchange rates in the future.

Inflation generally affects us by increasing our cost of labor. We do not believe that inflation had a material effect on our business, financial condition or results of operations during the years ended December 31, 2016 or 2017 or during the three months ended March 31, 2017 or 2018.

Emerging Growth Company Status

In April 2012, the Jumpstart Our Business Startups Act of 2012, or the JOBS Act, was enacted. Section 107 of the JOBS Act provides that an "emerging growth company," or EGC, can take advantage of the extended transition period provided in Section 7(a)(2)(B) of the Securities Act, for complying with new or revised accounting standards. Thus, an EGC can delay the adoption of certain accounting standards until those standards would otherwise apply to private companies. We have irrevocably elected not to avail ourselves of delayed adoption of new or revised accounting standards and, therefore, we will be subject to the same requirements to adopt new or revised accounting standards as other public companies that are not emerging growth companies.

We are in the process of evaluating the benefits of relying on other exemptions and reduced reporting requirements under the JOBS Act. Subject to certain conditions, as an EGC, we expect that:

- § we will present in this prospectus only two years of audited financial statements, in addition to any required unaudited financial statements, with correspondingly reduced Management's Discussion and Analysis of Financial Condition and Results of Operations disclosure;
- § we will avail ourselves of the exemption from providing an auditor's attestation report on our system of controls over financial reporting pursuant to Section 404(b) of the Sarbanes-Oxley Act;
- § we will avail ourselves of the exemption from complying with any requirement that may be adopted by the Public Company Accounting Oversight Board, or PCAOB, regarding mandatory audit firm rotation or a supplement to the auditor's report providing additional information about the audit and the financial statements, known as the auditor discussion and analysis; and
- § we will provide less extensive disclosure about our executive compensation arrangements

We will remain an EGC until the earlier of (i) the last day of the fiscal year in which we have total annual gross revenues of \$1.07 billion or more; (ii) the last day of 2023; (iii) the date on which we have issued more than \$1 billion in nonconvertible debt during the previous three years; or (iv) the date on which we are deemed to be a large accelerated filer under the rules of the Securities and Exchange Commission.

Recent Accounting Pronouncements

We have reviewed all recently issued standards and have determined that, other than as disclosed in Note 2 to our consolidated financial statements appearing elsewhere in this prospectus, such standards will not have a material impact on our financial statements or do not otherwise apply to our operations.

BUSINESS

Overview

We are a biopharmaceutical company focused on the discovery and development of innovative medicines for the treatment of serious diseases in which signaling by protein growth factors plays a fundamental role. Our newly elucidated understanding of the molecular mechanisms of growth factor activation enabled us to develop a proprietary platform for the discovery and development of monoclonal antibodies that locally and selectively target these signaling proteins at the cellular level. We believe this approach, acting in the disease microenvironment, avoids the historical challenges associated with inhibiting growth factors for therapeutic effect. We believe our focus on biologically validated growth factors may facilitate a more efficient development path. We are advancing our lead product candidate, SRK-015, a selective first-in-class inhibitor of the activation of the growth factor myostatin in skeletal muscle, into clinical development for the treatment of spinal muscular atrophy, or SMA. We plan to initiate our Phase 1 clinical trial in May 2018. In addition, utilizing our proprietary platform, we are also creating a pipeline of novel product candidates with the potential to transform the lives of patients suffering from a wide range of serious diseases, including other neuromuscular disorders, cancer, fibrosis and anemia.

Our proprietary platform is designed to discover and develop monoclonal antibodies that have a high degree of specificity to achieve selective modulation of growth factor signaling. Growth factors are naturally occurring proteins that typically act as signaling molecules between cells and play a fundamental role in regulating a variety of normal cellular processes, including cell growth and differentiation. Current therapeutic approaches to treating diseases in which growth factors play a fundamental role involve directly targeting an active growth factor or its receptor systemically throughout the body and have suffered from a variety of shortcomings:

- § multiple growth factors often signal through the same or overlapping sets of related receptors, making it difficult to specifically modulate one pathway over another;
- § members of the same growth factor superfamily share considerable structural similarities, making it difficult to achieve specific inhibition of the targeted growth factor; this can result in broad systemic inhibition that can cause undesirable, and in many cases toxic, side effects; and
- § systemic and non-selective inhibition of a growth factor can block the growth factor's role in the disease process, but can also simultaneously interfere with its normal physiological roles.

Our innovative approach is rooted in our structural biology insights into the mechanism by which certain growth factors are activated in close proximity to the cell surface, which we refer to as "supracellular activation." We integrate these insights with sophisticated protein expression, assay development and monoclonal antibody discovery capabilities. We believe our proprietary platform can address the challenges of current therapeutic approaches to treating diseases in which growth factors play a fundamental role by:

- § targeting the natural activation mechanism to prevent activation of the growth factor rather than attempting to inhibit the growth factor after activation;
- § achieving heightened specificity for the targeted growth factor while minimizing interactions with structurally similar and related growth factors, thereby reducing the risk of unintended systemic adverse events; and
- § targeting the disease microenvironment, where we believe we can interfere with the disease process while minimizing the effects on the normal physiological processes mediated by the same growth factors.

We are advancing our lead antibody product candidate, SRK-015, a first-in-class inhibitor of the activation of myostatin, into clinical development for the treatment of SMA. Myostatin is a negative regulator of muscle mass expressed primarily in skeletal muscle tissue, and a member of the transforming growth factor beta, or TGF β , superfamily, a group of more than 30 related growth factors that mediate diverse biological

processes. Vertebrate animals that lack the myostatin gene display increased muscle mass and strength relative to their normal counterparts, but are otherwise healthy. We believe inhibition of the activation of myostatin may promote a clinically meaningful increase in muscle mass and strength. As a result, we have focused our initial development efforts for SRK-015 on the treatment of SMA. SMA is a rare, and often fatal, genetic disorder arising from a deficiency of a protein known as "survival of motor neuron," or SMN. This disease typically manifests in young children and is characterized by atrophy of the voluntary muscles of the limbs and trunk and dramatically reduced normal neuromuscular function. An estimated 30,000 to 35,000 patients suffer from SMA in the United States and Europe. In preclinical studies, we observed that SRK-015 promoted increased muscle mass and strength, and *in vitro* studies have shown that the antibody selectively avoids interaction with other closely related growth factors that play distinctly different physiological roles. We believe that SRK-015 has the potential to be the first muscle-directed therapy to reverse or prevent muscle atrophy in SMA patients and could be used both as a monotherapy or in conjunction with therapies that upregulate the expression of SMN. In March 2018, we filed an Investigational New Drug application, or IND, with the U.S. Food and Drug Administration, or FDA for SRK-015. In April 2018, the FDA notified us that our Phase 1 first-in-human clinical trial of SRK-015 may proceed, and we plan to initiate our Phase 1 clinical trial in May 2018.

Our second antibody program is focused on the discovery and development of highly specific inhibitors of the activation of TGFb1. TGFb1 is also a member of the TGFb superfamily, and increased signaling by TGFb1 is a key driver of a number of disease-relevant processes, including tissue and organ fibrosis, immune system evasion by cancer cells, and bone marrow fibrosis associated with hematological disorders. Historically, selectively targeting TGFb1 signaling has been challenging due to the inability of both small molecule inhibitors and antibodies to avoid off-target inhibition of other, closely related growth factors, TGFb2 and TGFb3. Treatment of animals with these pan-TGFb inhibitors has been associated with a range of toxicities, most notably cardiac toxicity. In preclinical studies of our antibodies, we have observed specific inhibition of TGFb1 activation *in vitro* and immunomodulatory and antifibrotic activity in multiple disease models *in vivo*. In addition, we have completed a 28-day pilot toxicology study of our leading antibody and, to date, we have not observed any drug-related toxicity up to the highest doses tested in the study. In the same study, we tested pan-TGFb inhibitors and observed the toxicities, including cardiac toxicity, that have been observed by others. We are actively evaluating a limited number of our selective inhibitors of the activation of TGFb1 in multiple disease models, and we intend to nominate a clinical candidate to initially pursue in one or more of our currently targeted indications of oncology, immuno-oncology and fibrosis by the first half of 2019.

Our third antibody program targets the signaling of bone morphogenetic protein 6, or BMP6, another member of the TGFb superfamily, which is involved in a diverse set of biological processes in various parts of the body. For example, in the liver, BMP6 signaling is a key controller of the body's ability to regulate iron levels. Given BMP6's important role in iron metabolism, we believe that targeting BMP6 signaling in a liver-selective fashion presents the potential to address both iron-restricted anemias and iron overload conditions. In preclinical studies of our antibodies that target BMP6 signaling in the liver, we have observed increased iron levels in the bloodstream of healthy animals and we are now evaluating a limited number of these antibodies in disease models of iron restricted anemia.

We have worldwide rights to our proprietary platform and all of our product candidates and antibodies with the exception of certain early-stage antibodies that specifically inhibit the activation of TGFb1 in the context of regulatory T cells, which we licensed to Janssen Biotech, Inc., or Janssen, a subsidiary of Johnson & Johnson.

We have assembled an experienced management team, board of directors, scientific founders and advisory board who bring extensive industry experience to our company. The members of our team have deep experience in discovering, developing and commercializing therapeutics with a particular focus on rare diseases, having worked at companies such as Alnylam Pharmaceuticals, Inc., Avila Therapeutics, Inc.,

Biogen, Inc. and Dyax Corp. We were founded by internationally respected scientists, Drs. Timothy A. Springer and Leonard I. Zon of Harvard Medical School and Boston Children's Hospital.

Since our inception in 2012, we have raised over \$100 million through convertible preferred stock financings. Our investors include ARCH Venture Partners, Cormorant Asset Management, EcoR1 Capital, Fidelity Management and Research Company, Invus, The Kraft Group, Polaris Partners, Redmile Group and Timothy A. Springer, Ph.D.

Our Approach and Proprietary Platform

Our innovative approach is rooted in our newly elucidated understanding of the molecular mechanisms of growth factor activation and signaling and is designed to discover and develop monoclonal antibody product candidates that can inhibit the activation of a growth factor with an unprecedented degree of selectivity. Our proprietary platform is designed to generate product candidates that target the growth factor's latent precursor form prior to its activation within the disease microenvironment, or tissue where it is localized, and would normally signal upon activation.

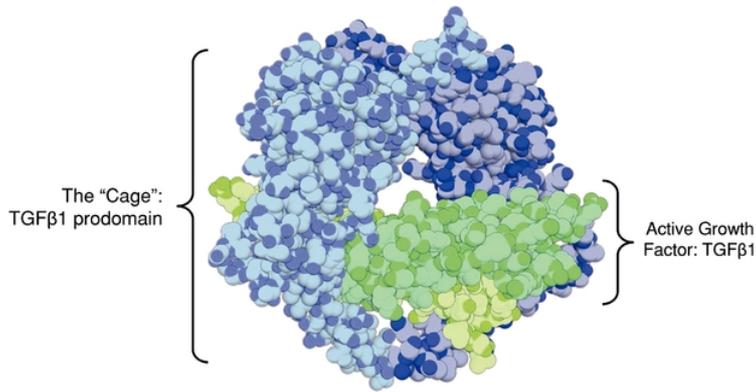
Growth factors are naturally occurring proteins that typically act as signaling molecules between cells and play a fundamental role in regulating a variety of normal cellular processes. Members of the TGF β superfamily of growth factors, for example, can mediate diverse biological functions, including cell growth and differentiation, tissue homeostasis, immune modulation and extracellular matrix remodeling. Growth factors, including members of the TGF β superfamily, such as myostatin, TGF β 1 and BMP6, have also been shown to play a fundamental role in a variety of disease processes, including neuromuscular disorders, cancer, fibrosis and anemia. Because of the importance of growth factors in multiple diseases, the pharmaceutical industry has made many attempts to inhibit growth factors in a variety of therapeutic settings. However, products utilizing conventional approaches have seen only limited success. Current therapeutic approaches to treating diseases in which growth factors play a fundamental role involve directly targeting an activated growth factor or its receptor systemically throughout the body and have suffered from a variety of shortcomings:

- § multiple growth factors often signal through the same or overlapping sets of related receptors, making it difficult to specifically modulate one pathway over another;
- § members of the same growth factor superfamily share considerable similarities (for example, myostatin and GDF11 are approximately 90% identical in the growth factor domains) making it difficult to achieve selective inhibition of the targeted growth factor. Inhibiting both the intended growth factor target and other closely related targets can result in unintentionally broad systemic inhibition that can cause undesirable, and in many cases toxic, side effects; and
- § systemic and nonselective inhibition of a growth factor can block the growth factor's role in the disease process, but can also simultaneously interfere with its normal physiological roles.

Our approach to the discovery and development of growth factor-targeted drugs is fundamentally new and different from traditional approaches. Our approach is based on the breakthrough discovery by the laboratory of our co-founder, Timothy A. Springer Ph.D. of Harvard Medical School and Boston Children's Hospital, of the supracellular activation mechanism by which growth factors in the TGF β superfamily are locally activated by a variety of specific stimuli in close proximity to the cell surface.

Unlike many other proteins that are produced and secreted by cells in a mature, or active, form, many growth factors are expressed by cells in a precursor, or latent, form. For example, TGF β 1 is produced by cells as a single protein which is then enzymatically processed by the cells into two distinct and physically separated domains — the mature growth factor and the remaining portion of the original protein, referred to as the prodomain — which remain associated as part of a complex. This secreted complex is latent, or inactive, and must first be activated to carry out its normal function in a highly localized tissue or disease microenvironment. In a seminal peer-reviewed publication in 2011, Dr. Springer elucidated a new understanding of the mechanism of supracellular activation as it applies to members of the TGF β

superfamily, by solving a high resolution x-ray crystal structure of this latent form of TGFβ1, as illustrated in the graphic below.



Structural representation of the latent form of TGFβ1 wherein the prodomain wraps around the active growth factor.

This research explained at a molecular level why the secreted form of TGFβ1 is inactive. The prodomain, though physically separated from the mature growth factor domain, forms a "cage" around the active form of TGFβ1, blocking the growth factor from signaling through its receptor. Only when the cage is "unlocked" by a supracellular activation event can the growth factor be released and mediate its effects in the local microenvironment. Dr. Springer further hypothesized that this phenomenon likely holds true for most members of the TGFβ superfamily, though the exact nature of the activation event, such as integrin binding or enzymatic cleavage, may differ among members of the superfamily. Importantly, while many growth factors are structurally very similar, their cages are structurally diverse, and this provides the basis for our approach to improved selectivity.

To enable our novel approach, we have built a proprietary platform that is rooted in our structural biology insights into supracellular activation. We integrate these insights with sophisticated protein expression, assay development and monoclonal antibody discovery capabilities. In addition to such know-how, our proprietary platform is covered by two patent families, with issued patents projected to expire in 2034. The key elements of our proprietary platform include the following:

- § focusing on growth factor targets with a high degree of evidence implicating them in a disease process or processes;
- § generating recombinant versions of the latent forms of targeted growth factors, as well as versions of closely related growth factors utilizing proprietary technology and in-house expertise;
- § developing proprietary assays in which we are able to recapitulate the natural supracellular activation mechanism that these growth factors undergo in the human body;
- § designing sophisticated selection strategies utilizing recombinant antibody libraries such as phage and yeast display that allow us to identify monoclonal antibodies, a well-established therapeutic modality, that can modulate the supracellular activation of these growth factors without having an effect on the activation of other closely related growth factors; and

§ optimizing the output of such selections to ensure that our product candidates have the appropriate characteristics for manufacturability and further development.

Using our innovative approach and proprietary platform, we are creating a pipeline of novel product candidates that selectively modulate the activation of growth factors implicated in a variety of serious diseases.

We believe there are several important advantages to our approach over conventional therapeutic approaches, which inhibit the growth factors or their receptors systemically throughout the body:

- § targeting the latent precursor allows intervention at the site of action, within the microenvironment of the diseased tissue. Because our antibodies specifically bind the latent forms of the growth factors, we can prevent the activation of the growth factors. Given that many growth factors act primarily within the microenvironment where they are activated, as opposed to exerting their effects systemically, we believe that prevention of activation is a preferred mode of action for achieving improved outcomes. In contrast, traditional approaches to targeting growth factor signaling are focused on inhibiting the growth factor after it has been activated and released systemically;
- § targeting the latent precursor allows heightened selectivity among structurally related growth factors, which we believe could limit off-target effects. For example, two members of the TGF β superfamily, myostatin and GDF11, are 90% identical in their growth factor domains. Therefore, many of the traditional inhibitors that target myostatin also inadvertently inhibit GDF11. Similarly, most of the known inhibitors of TGF β are pan-inhibitors, meaning they do not distinguish among the three isoforms of TGF β , namely, TGF β 1, TGF β 2 and TGF β 3. Despite the sequence similarities of the active forms of these growth factors, their cages are structurally diverse. We have been able to harness this diversity to generate antibodies that specifically bind the inactive growth factor precursors and inhibit activation of a particular growth factor of interest, but not others that are closely related; and
- § targeting these precursor forms in the disease microenvironment, we believe we can interfere with the disease process while minimizing the effects on the normal physiological processes mediated by growth factors.

Our Strategy

Using our proprietary platform to unlock the therapeutic potential of targeting growth factor signaling in the disease microenvironment, our goal is to deliver novel therapies to underserved patients suffering from a wide range of serious diseases, including neuromuscular disorders, cancer, fibrosis and anemia. To achieve this goal we plan to:

- § **Rapidly advance our lead product candidate, SRK-015, through clinical proof-of-concept.** We are currently developing our lead product candidate, SRK-015, for the treatment of patients with SMA. By targeting the latent form of myostatin and specifically inhibiting its activation in muscle, we believe SRK-015 holds considerable promise in addressing the atrophy of skeletal muscle in patients with SMA. In March 2018, we filed an IND for SRK-015 and, in April 2018, the FDA notified us that our first-in-human clinical trial of SRK-015 may proceed. We plan to initiate our Phase 1 clinical trial in May 2018. Assuming successful results and subject to regulatory feedback, we intend to conduct a Phase 2 clinical proof-of-concept trial to evaluate the efficacy and safety of SRK-015 in patients with later-onset SMA, including those patients who are being treated with a currently approved SMA therapy. We plan to commence our Phase 2 trial in the first quarter of 2019 and expect to report top-line results in the second half of 2019.
- § **Advance our TGF β 1 program into clinical development.** Our second antibody program is focused on the discovery and development of highly specific inhibitors of the activation of TGF β 1. We believe that the selectivity of our antibodies is a significant differentiator in our efforts to address the historical challenges with inhibiting the TGF β signaling pathway. In preclinical studies of our antibodies, we have observed inhibition of TGF β 1 activation *in vitro*, and immunomodulatory and antifibrotic activity in multiple *in vivo* disease models. We intend to nominate a clinical candidate in one or more of our currently targeted indications of oncology, immuno-oncology and fibrosis in our TGF β 1 program by the first half of 2019.

- § **Explore additional indications for our existing and emerging product candidates.** Given the multiple physiological roles played by the distinct targets in our lead programs, we believe that there is potential for us to address multiple additional indications beyond those already selected. For example, we believe that SRK-015 may have a role in treating other muscle-wasting diseases and we believe that our TGFb1 program has the potential to address multiple disorders associated with increased TGFb1 signaling, such as tissue and organ fibrosis, immune system evasion by cancer cells, and bone marrow fibrosis associated with hematological disorders. Our goal is to maximize the value of our existing programs by exploring their potential in additional indications.
- § **Continue to leverage our proprietary platform to expand our pipeline beyond current lead programs.** We will continue to leverage our proprietary platform to selectively target the activation of additional growth factors, both within and beyond the TGFb superfamily. Given the established role of signaling by protein growth factors in numerous diseases, we believe that these efforts could result in multiple new opportunities to treat diseases with high unmet medical need. In order to support our pipeline expansion and intention to be the leader in the field of growth factor-targeted drug development, we are investing in the technologies supporting our proprietary platform, including a focus on tools and assays to enhance and accelerate our drug discovery process.
- § **Selectively seek strategic collaborations to maximize the value of our proprietary platform and pipeline.** Given the potential of our proprietary platform to generate novel product candidates that could treat a wide variety of diseases, we believe that we can maintain in-house discipline with respect to our key development and commercialization efforts, while at the same time maximizing the full potential of our proprietary platform for other disease areas and indications. As a result, we may seek to form strategic collaborations around certain targets, product candidates or disease areas that we believe could benefit from the resources of either larger biopharmaceutical companies or those specialized in a particular area of relevance.
- § **Attract and retain people that share our commitment to scientific excellence and a focus on patients.** We are focused on developing novel medicines that make a significant difference in the lives of patients suffering from devastating and life-threatening diseases. In addition to building a team of people with deep experience in biology, protein sciences, antibody drug discovery, and development and operations, we believe our focus on a patient-centric collaborative and passionate workplace culture is critical to the success of our mission. We will continue to emphasize this focus as we grow and build our company.

Our Pipeline Programs

Using our innovative approach and proprietary platform, we are creating a pipeline of novel product candidates that selectively inhibit the supracellular activation of growth factors believed to be important drivers in a variety of diseases, including neuromuscular disorders, cancer, fibrosis and anemia. Our proprietary platform includes (i) our know-how expressing and purifying latent protein growth factor complexes in quantity and quality sufficient to enable antibody discovery; (ii) strategies to identify rare antibodies that selectively bind targeted latent protein growth factor complexes; and (iii) assays developed by us in which to test the highly selective antibodies' ability to modulate the activation of specific latent growth factors. We have worldwide rights to our proprietary platform and all of our product candidates, with the exception of certain early-stage antibodies that specifically inhibit the activation of TGFb1 in the context of regulatory T cells, which we licensed to Janssen.

Our Lead Product Candidate and Additional Programs

SRK-015 — Our Inhibitor of Myostatin Activation

We are developing SRK-015, a selective first-in-class inhibitor of the activation of the growth factor myostatin in skeletal muscle, for the treatment of SMA. Myostatin, a member of the TGFb superfamily of growth factors, is expressed primarily in skeletal muscle cells and the absence of its gene is associated with an increase in muscle mass and strength in multiple animal species. We believe that inhibition of the

activation of myostatin may promote a clinically meaningful increase in muscle mass and strength. In preclinical studies, treatment with SRK-015 resulted in an increase in muscle mass and strength in healthy animals as well as maintenance of muscle in multiple models of muscle atrophy. In March 2018, we filed an IND for SRK-015 and, in April 2018, the FDA notified us that our Phase 1 first-in-human clinical trial of SRK-015 may proceed. We plan to initiate our Phase 1 clinical trial in May 2018.

Background on SMA

SMA is a rare, and often fatal, genetic disorder that typically manifests in young children. It is characterized by the loss of motor neurons, atrophy of the voluntary muscles of the limbs and trunk and progressive muscle weakness. Disease severity in SMA can range from patients who die soon after birth to patients who live into adulthood with varying degrees of morbidity. The underlying pathology of SMA is caused by insufficient production of a protein known as "survival of motor neuron," or SMN. The SMN protein, essential for the survival of motor neurons, is encoded by two genes, SMN1 and SMN2.

- § SMN1 genes produce the majority of functional SMN protein; healthy individuals have one or two functional copies of SMN1, while patients with SMA have mutations in or deletions of both copies of the gene.
- § SMN2 genes produce only 10% to 20% of functional SMN protein and an individual's copy number of the SMN2 gene can range from zero to eight. In SMA patients, the number of SMN2 genes present in their genome is correlated with disease onset and severity; patients who have a lower number of SMN2 gene copies generally develop earlier and more severe SMA, because they produce less SMN protein.

SMA Natural History and Epidemiology

SMA, the most common monogenic cause of death in infants, is a rare neuromuscular disorder. An estimated 30,000 to 35,000 patients suffer from SMA in the United States and Europe. Patients with SMA can be categorized as one of four types, Type 1 through Type 4. More than 80% of SMA patients currently living are categorized as having Type 2 or Type 3 disease, sometimes referred to as later-onset SMA, and represent our initially targeted patient population.

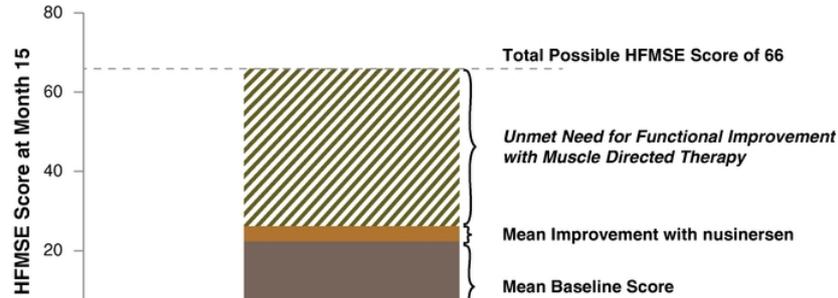
- § Type 1 disease is the most severe form, with clinical signs emerging at or shortly following birth. Patients with Type 1 SMA suffer from respiratory compromise and often require mechanical ventilation shortly after birth. Type 1 infants are never able to sit without support. Type 1 SMA is the most common form of the disease, and accounts for 58% of patients born with SMA. Historically, only 1% of patients with Type 1 disease survive beyond two years of age without mechanical respiratory support. Type 1 SMA represents only 14% of patients with SMA, although recent therapies may extend patient lifespans. Type 1 patients begin to lose motor neurons and muscle mass before birth.
- § Type 2 disease manifests in early childhood and is less severe than Type 1 disease, although patients exhibit profound deficits in motor function. Patients with Type 2 disease may be able to sit independently but they will never walk without aid. While only 29% of the incident population, patients with Type 2 disease account for 51% of the patients living with SMA today.
- § Type 3 disease manifests usually in childhood and accounts for about 13% of patients born with SMA, although patients in this category account for 35% of all patients with SMA. While Type 3 SMA patients usually learn to walk unaided, the majority lose that ability over time. Ambulatory Type 3 SMA patients commonly suffer from substantial motor functional impairment, as evidenced by Hammersmith Functional Motor Scale Expanded, or HFMSSE, scores and Six-Minute Walk Test distances, two commonly used measures of motor function.
- § Type 4 disease is the mildest form of SMA, and its population is not well characterized. After symptom onset, which is most commonly reported between 20 and 30 years of age, patients

experience mild to moderate muscle weakness and increasing disabilities. Patients are ambulatory and their life expectancy is normal.

Unmet Medical Need in SMA

Patients with SMA continue to have a high unmet medical need despite recent approval of nusinersen, an SMN upregulator. Nusinersen was approved by the FDA in December 2016 and the European Medicines Agency, or EMA, in June 2017 for the treatment of patients with SMA. Nusinersen is an antisense oligonucleotide directed against SMN2 that aims to increase functional SMN protein expression. SMN upregulator therapies act primarily to preserve motor neurons. While this approach may improve motor function, it does not act directly on the muscle to reverse or prevent atrophy. We believe that SRK-015 has the potential to be the first muscle-directed therapy to reverse or prevent muscle atrophy in patients with all types of SMA and further improve patient outcomes as a monotherapy or when used in conjunction with SMN upregulator therapies such as nusinersen.

The CHERISH pivotal trial of nusinersen in later-onset SMA patients made use of the HFMSE, a validated outcome measure specifically designed for evaluation of Type 2 and 3 SMA patients that is often used in clinical practice and studies. This examination assesses 33 individual items of motor activity, each scored from 0 to 2 points (lower score indicates worse motor function), with a maximum possible score of 66. The HFMSE evaluates a patient's ability to perform basic tasks such as sitting, reaching one's hand to one's head, changing body positions (e.g. sitting to lying position), crawling, standing, kneeling, squatting, jumping and ascending/descending stairs. These tasks are viewed by SMA patients and caregivers as meaningful and relevant to conducting activities of daily living. In this trial, as illustrated in the figure below, treatment with nusinersen improved motor function, but the HFSME scores of treated patients remained well below those of healthy children. Patients who received nusinersen achieved a 3.9-point mean improvement at Month 15 from a mean baseline of 22.4. Compared to control patients, there was a statistically significant difference of 4.9 points in the mean change from baseline to Month 15 in the HFMSE score. The percentage of nusinersen-treated patients achieving a > 3-point increase was 57%. Although this trial met its primary endpoint and demonstrated a clinically meaningful benefit overall, these results also indicate that most of the gap in attaining normal HFMSE performance has not been adequately addressed by nusinersen therapy and significant unmet need remains.



Mean improvement in HFMSE score experienced by patients with later-onset SMA in the Phase 3 CHERISH clinical trial of nusinersen.

While nusinersen has shown improvement in motor function in patients with SMA, there remains a significant unmet need for an effective muscle-directed therapy that can reverse or prevent muscle atrophy, thereby improving muscle strength and motor function in patients with SMA. Given the novel mechanism of SRK-015, we believe this therapy has the potential to provide a clinically meaningful improvement in motor function in a broad population of SMA patients who may or may not be on background SMN upregulator therapies, such as nusinersen. Accordingly, we believe SRK-015 has the potential to fulfill a significant unmet medical need for SMA patients both as a monotherapy and in conjunction with standard of care.

Myostatin in SMA and Challenges with Traditional Approaches

Our lead product candidate, SRK-015, is a selective inhibitor of the activation of latent myostatin that acts locally within skeletal muscle. Myostatin, also known as growth differentiation factor 8, or GDF8, is a member of the TGF β superfamily and is produced by skeletal muscle cells. As with other tissues and organs in the human body, healthy muscle homeostasis is maintained by a proper balance of growth signals, or anabolic stimuli, and breakdown signals, or catabolic stimuli. In humans, the anabolic stimuli that drive muscle growth are proteins such as human growth hormone and insulin-like growth factor 1. In contrast, myostatin is a catabolic agent that functions as a negative regulator of muscle mass.

Skeletal muscle fibers are generally classified as fast-twitch or slow-twitch. Fast-twitch fibers play a key role in motor activities such as those involving quick bursts of strength, sprinting or eccentric contraction. In contrast, slow-twitch fibers are important for endurance activities. Animals lacking functional myostatin genes, or its receptor, have larger muscles and increased strength compared to normal animals. While the absence of myostatin does lead to overall increases in muscle mass, a preferential effect on muscles enriched for fast-twitch muscle fibers has been observed in animals. Such animals are otherwise healthy and live a normal life-span.

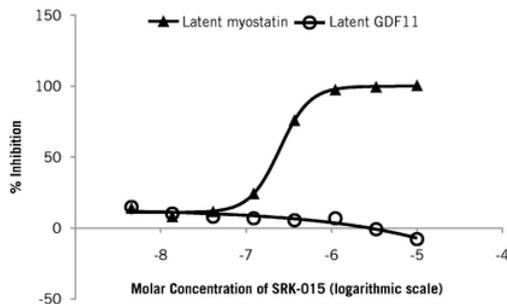
Because of its role in regulating muscle mass, myostatin has been a popular target for a variety of drug development programs. There have been two general approaches to trying to inhibit the signaling of myostatin in humans. The first is to develop an antibody, or an antibody-like molecule, that binds to mature myostatin in circulation and prevents its ability to signal through its receptor, the ActRIIb receptor. The second is to develop an antibody to the ActRIIb receptor itself, or a soluble decoy of the ActRIIb receptor, with a goal of preventing myostatin signaling through its receptor. Both of these approaches, however, have significant limitations.

As a member of the TGF β superfamily, mature myostatin shares considerable structural similarity with other family members. For example, the active form of myostatin and its most closely related family member, GDF11, are 90% identical in the growth factor domains, making it extremely challenging to identify antibodies that are truly specific for myostatin and do not interfere with other targets. Moreover, attempts to interrupt myostatin signaling through its receptor are complicated by the fact that the ActRIIb receptor, in addition to being the receptor for myostatin, is also the receptor for a number of related family members, including GDF11, activin and other growth factors. Attempts to block the signaling of myostatin by targeting its receptor therefore inevitably interfere with the signaling of these other growth factors, many of which are involved in normal biological processes unrelated to muscle.

There are multiple examples of clinical trials demonstrating the risk of non-selective inhibition of myostatin. For example, in a Phase 2 trial in Duchenne Muscular Dystrophy reported in 2017, a soluble decoy of the ActRIIb receptor resulted in bleeding side effects believed by the sponsor to be unrelated to inhibition of myostatin signaling, but instead related to the inhibition of signaling by certain other members of the TGF β superfamily known to be important in the maintenance of vascular integrity. These side effects resulted in termination of the clinical program. More recently, results from a clinical trial were reported showing that treatment of patients with an antibody to the ActRIIb receptor resulted in suppression of the levels of follicle stimulating hormone, an important reproductive hormone. In this trial, the sponsor believed that these effects were likely related to inhibition of signaling through the ActRIIb receptor.

Our Solution

Utilizing our proprietary platform, we targeted the precursor form of myostatin and generated SRK-015, a selective first-in-class inhibitor of the activation of myostatin from its inactive precursor in skeletal muscle where myostatin resides and signals upon activation. While mature myostatin is 90% identical in the growth factor domain to its most closely related TGF β superfamily member, GDF11, the prodomain that cages mature myostatin and keeps it in its latent precursor form is only 52% identical to the GDF11 prodomain. As a result, in preclinical studies, we observed that SRK-015 bound to latent myostatin with a high level of selectivity, while having no binding to, and no effect on, the activation of related TGF β family members.



SRK-015 showed dose-dependent inhibition of the activation of latent myostatin in an *in vitro* activation assay and had no effect on latent GDF11 activation.

We believe that the pathophysiologic and clinical characteristics of SMA are well-aligned with the optimal setting for observing therapeutic benefit from inhibition of myostatin activation. These characteristics are summarized in the figure below. Since myostatin regulates muscle catabolism rather than anabolism, we believe that having a background of anabolic capacity is important to drive muscle growth in the setting of myostatin inhibition. Anabolic capacity is most robust in younger individuals and diminishes as one ages. Furthermore, in SMA, there is a significant but incomplete loss of motor neurons, ensuring at least some intact connectivity between muscle and nerve, also known as innervation. This partial loss of motor neurons causes substantial atrophy of fast-twitch muscle fibers that in turn leads to many of the motor function impairments. Validated outcome measures are available for SMA clinical trials that are relevant to fast-twitch fiber activity. These outcome measures, such as the HFMSE, assess a large number of motor

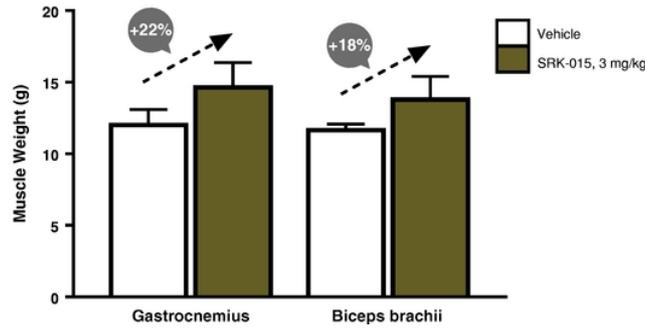
activities that involve short-term bursts of strength, which are driven by fast-twitch muscle fibers. These endpoints therefore measure an outcome that may be more likely to be directly affected by SRK-015.

Optimal Setting for Myostatin Inhibition	Key Characteristics of SMA
Younger population	Genetic disorder with onset in childhood
Muscle disease with at least partially intact innervation	Incomplete loss of motor neurons
Need for increase in fast-twitch muscle fibers	Substantial deficit in fast-twitch fibers
Clinical trial endpoint driven by fast-twitch fiber function	Fast-twitch fiber function: prominent role in SMA outcome measures

Table summarizing why the pathophysiologic and clinical characteristics of SMA are well aligned with the optimal setting for observing therapeutic benefit from inhibition of myostatin activation.

SRK-015 Preclinical Results

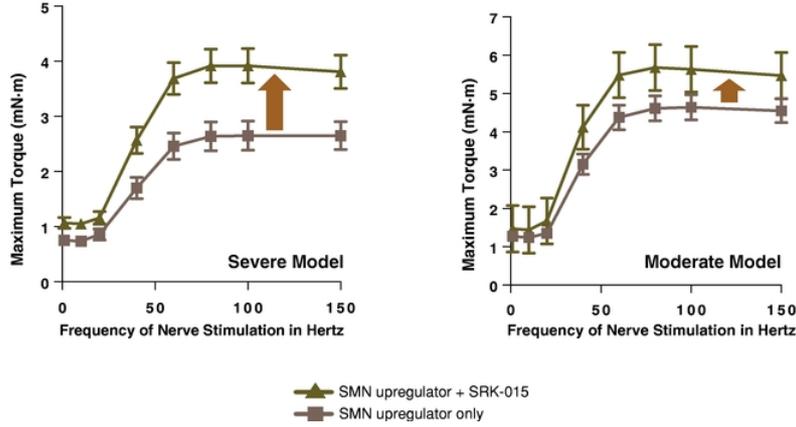
In our earliest pharmacology work, we observed that treatment with SRK-015 robustly increased muscle mass and strength in healthy mice and rats. In addition to increasing muscle mass, we also observed that treatment with SRK-015 resulted in gains in muscle function. The increase in muscle mass was replicated in non-human primates. As shown in the figure below, in this study the gastrocnemius (a calf muscle) and biceps brachii (an arm muscle), two muscles containing a higher proportion of fast-twitch fibers than slow-twitch fibers, increased in size by 18% and 22%, respectively, in cynomolgus monkeys treated with SRK-015 as compared to the vehicle control group.



SRK-015 treatment increased muscle mass, as measured by muscle weight in the gastrocnemius (a calf muscle) and biceps brachii (an arm muscle) of cynomolgus monkeys as compared to monkeys treated with the vehicle only.

We next assessed the ability of SRK-015 to improve muscle function in the D7 mouse model, a genetic model of SMA wherein the SMN1 gene has been deleted and copies of the human SMN2 gene have been introduced, thus mimicking the genetics of the human disease. SMN D7 mice are extremely fragile if not treated with a drug that upregulates the underlying deficiency in SMN. Accordingly, this model is best suited for determining the effect of a product candidate such as SRK-015 when administered in conjunction with an SMN upregulator. In this study, we used a small molecule SMN2 splice modulator, SMN-C1, as the SMN upregulator.

We evaluated the ability of SRK-015 to improve muscle force generation in two versions of the D7 mouse model: one designed to emulate a more severe form of SMA and the other a more moderate form of SMA. As shown in the figure below, in both models animals treated with SRK-015 in conjunction with the SMN upregulator experienced a significant increase in maximum muscle torque generation in the leg compared to animals treated with the SMN upregulator alone.



SRK-015, in combination with an SMN upregulator, improved in vivo muscle force generation in versions of the D7 mouse model designed to emulate either a severe (left side) or moderate (right side) form of SMA, as compared to SMN upregulator therapy alone. Muscle force was assessed by maximum torque generation following nerve stimulation, at a range of frequencies, in the plantarflexor muscle group in the leg. The arrows indicate the increase in muscle force generation due to SRK-015 treatment.

We have filed patent applications seeking to cover SRK-015 as well as other antibodies with the same mechanism. In September 2017, we announced the issuance of U.S. Patent 9,758,576, which covers monoclonal antibodies that selectively inhibit myostatin signaling by blocking the proteolytic activation of latent myostatin, providing protection for our lead antibody SRK-015, as well as any other monoclonal antibodies that work by this unique mechanism of action. This patent expires in May 2034, not including any potential patent term extension.

Clinical Development Overview

We are currently advancing SRK-015 into a clinical development program for the treatment of SMA. Our IND-enabling toxicology studies in rat and cynomolgus monkeys have been completed and, in March 2018,

we filed an IND for SRK-015. In April 2018, the FDA notified us that our Phase 1 first-in-human clinical trial of SRK-015 may proceed. We plan to initiate our Phase 1 clinical trial in May 2018. Our Phase 1 trial is designed to assess the safety, tolerability, pharmacokinetics, immunogenicity and pharmacodynamics of single- and multiple-ascending doses of intravenous SRK-015 in healthy adult volunteers.

Assuming the successful completion of our Phase 1 trial, we plan to conduct a Phase 2 proof-of-concept trial to evaluate the efficacy and safety of SRK-015 in patients with later-onset SMA. This includes Type 2 and non-ambulatory Type 3 SMA patients who may already be receiving an approved SMN upregulator therapy like nusinersen as background standard of care, as well as ambulatory Type 3 patients who will be administered SRK-015 as a monotherapy. We expect top-line results from the Phase 2 proof-of-concept trial to be available in the second half of 2019.

Beyond the initial proof-of-concept trials in Type 2 and Type 3 SMA patients, we believe that SRK-015 has the potential to contribute an important therapeutic benefit to patients with both more and less severe forms of SMA.

On March 22, 2018, the FDA granted orphan drug designation for SRK-015 for the treatment of SMA.

Other Myostatin Indications

We believe that SRK-015 has therapeutic potential to improve muscle function in multiple other muscle-wasting disorders, including muscle atrophy due to partial denervation, incomplete spinal cord injury, amyotrophic lateral sclerosis, glucocorticoid-induced muscle-wasting and Duchenne muscular dystrophy. These disorders bear many of the characteristics relevant to the optimal setting in which we believe that therapeutic benefit from myostatin inhibition may be observed. In addition to conducting a proof of concept study in SMA, we are actively considering the investigation of SRK-015 in multiple other indications.

Inhibitor of TGFb1 Activation Programs

TGFb1 is also a member of the TGFb superfamily, and increased signaling by TGFb1 is a key driver of a number of disease-relevant processes, including tissue and organ fibrosis, immune system evasion by cancer cells and bone marrow fibrosis associated with hematological disorders. Historically, selectively targeting TGFb1 signaling has been challenging due to the inability of both small molecule inhibitors and antibodies to avoid off-target inhibition of other, closely related growth factors, TGFb2 and TGFb3. Treatment of animals with these pan-TGFb inhibitors has been associated with a range of toxicities, most notably cardiac toxicity. Furthermore, since each of these growth factors signals through the same TGFb receptor, ALK5, inhibitors of the TGFb receptor suffer from similar dose-limiting toxicities. Using our proprietary platform, we have generated highly specific and local inhibitors of the activation of TGFb1 that, in our preclinical studies, showed no detectable inhibition of the activation of TGFb2 or TGFb3.

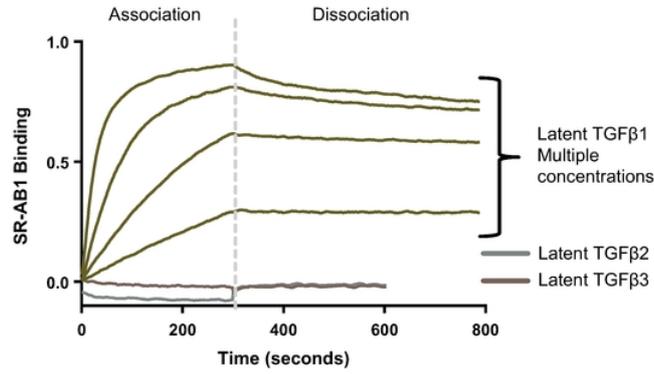
Identification of Selective Inhibitors of TGFb1 Activation

TGFb1 is produced by cells as a single protein and is then enzymatically processed by the cells into two distinct and physically separated domains — the mature, active growth factor and the remaining portion of the original protein, referred to as the prodomain — which remain associated and inactive. This complex also includes one of a number of "presenting molecules" which when secreted serve to tether the latent precursor in specific locations in the body. TGFb1 is produced by a variety of cell types, including fibroblasts, which deposit latent TGFb1 in connective tissue, as well as regulatory T cells and macrophages, which display latent TGFb1 on their cell surfaces.

In a seminal peer-reviewed publication in 2011, Dr. Springer elucidated a new understanding of the mechanism of supracellular activation as it applies to members of the TGFb superfamily, by solving a high resolution x-ray crystal structure of the latent form of TGFb1. This research explained at a molecular level why the secreted form of TGFb1 is inactive. The prodomain, though physically separated from the mature

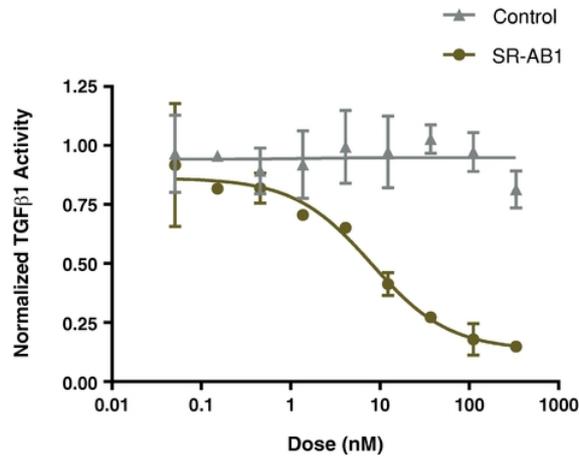
growth factor domain, forms a "cage" around the active form of TGF β 1, blocking the ability of the growth factor to signal through its receptor. Integrin proteins are able to unlock the "cage" by binding to the prodomain of the latent TGF β 1 complex and applying force to pull the complex open, allowing the mature growth factor to be released and signal in its microenvironment. While mature TGF β 1 shares a high degree of structural similarity with its closely related family members, TGF β 2 and TGF β 3, their respective cages are structurally diverse. By taking advantage of the differences among the prodomains, together with our understanding of the activation mechanism and ability to recapitulate the activation mechanism *in vitro*, we were able to identify multiple highly selective inhibitors of the activation of latent TGF β 1.

We have conducted *in vitro* and *in vivo* studies to characterize our selective TGF β 1 activation inhibitors. An example of the selectivity and *in vitro* inhibitory activity observed for one of our selective TGF β 1 activation inhibitors, SR-AB1, is shown in the figure below. This inhibitor bound to latent TGF β 1 with high affinity and showed no detectable binding to latent TGF β 2 or latent TGF β 3. Given the *in vitro* and *in vivo* activity observed with SR-AB1 and at least three additional TGF β 1 activation inhibitors, we believe that one or more of these may be advanced as a clinical candidate, and upcoming comparative *in vivo* studies may enable selection of the best candidate. We intend to nominate a clinical candidate in one or more of our currently targeted indications of oncology, immuno-oncology and fibrosis in our TGF β 1 program by the first half of 2019.



SR-AB1, one of our selective TGF β 1 activation inhibitors, showed dose-dependent binding of latent TGF β 1 with no detectable binding to latent TGF β 2 or latent TGF β 3 *in vitro*.

We have also observed potent inhibitory activity for this inhibitor in an *in vitro* latent TGF β 1 activation assay, as shown in the figure below.



SR-AB1 showed dose-dependent inhibition of TGF β 1 activity in an *in vitro* cell based assay of latent TGF β 1 activation.

We have also completed 7- and 28-day pilot toxicology studies for our leading selective inhibitor of the activation of latent TGF β 1, and have identified no drug-related toxicities up to 100 mg/kg dosed weekly, the highest dose tested. This is in contrast to the cardiac pathology we observed after up to one week of dosing with an ALK5 inhibitor or an inhibitor of all three forms of mature TGF β .

TGF β 1 in Fibrosis

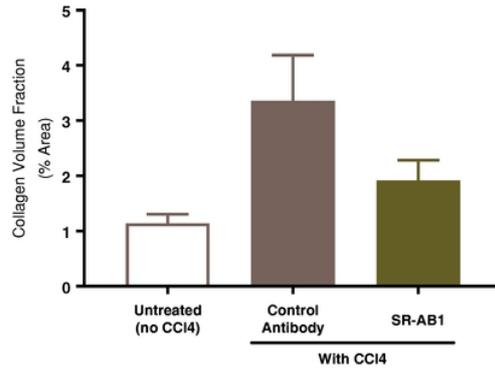
Based on our preclinical results, we believe that specific inhibition of TGF β 1 alone may be sufficient to suppress profibrotic signaling in multiple organs, and holds the promise of better tolerated and more effective therapies for a variety of fibrotic diseases than historical approaches. We are currently evaluating our selective inhibitors of latent TGF β 1 activation in a variety of translational models of organ fibrosis.

Fibrosis is a pathological feature of disease in virtually all organs, characterized by excessive accumulation of extracellular matrix in the affected tissue, and accounts for substantial morbidity and mortality. Multiple peer-reviewed studies have implicated TGF β signaling as a central regulator of fibrosis. TGF β is upregulated in many animal models of fibrosis, and overexpression of TGF β *in vivo* induces fibrotic changes. Furthermore, TGF β inhibition in animal models has been shown to reduce fibrosis in models of hepatic, renal and cardiac fibrosis. In humans, in an open-label trial of fresolimumab, an inhibitor of all three forms of TGF β , in systemic sclerosis, a fibrotic connective tissue disease, improved clinical skin disease as measured by the modified Rodnan skin score, a commonly used measure of skin thickness, was observed, although bleeding episodes were also reported in this trial. These data suggest that novel approaches to targeting TGF β signaling may have broad applicability to the treatment of fibrotic disease.

We observed that our selective inhibitors of the activation of latent TGF β 1 resulted in the inhibition of TGF β signaling, measured by the phosphorylation of SMAD2/3, a direct downstream target of TGF β 1, in a

progressive, genetic mouse model of kidney fibrosis known as the Alport Syndrome model. In this model, we observed that our selective inhibitor of the activation of latent TGF β 1 completely suppressed phosphorylation of SMAD 2/3, and was as effective as 1D11, an inhibitor of all three mature forms of TGF β . Based on these observations, we believe that TGF β 1 is the primary driver of TGF β signaling in this disease model.

We have also observed in the unilateral ureteral obstruction model, a well-characterized model of renal fibrosis, that a number of our selective TGF β 1 activation inhibitors resulted in robust suppression of TGF β 1 target genes and downstream fibrotic markers. Furthermore, in the CCl $_4$ model of liver fibrosis, we have observed that a number of our selective antibodies inhibited fibrotic progression in the liver, reducing collagen content both as assessed by a pathologist and, as shown in the figure below for one of our antibodies, SR-AB1, by quantitative histopathological staining.



SR-AB1, a selective inhibitor of TGF β 1 activation, inhibited carbon tetrachloride (CCl $_4$) induced liver fibrosis, as compared to a negative control antibody. Fibrosis was identified by increased collagen deposition, as assessed by quantitative histopathological staining.

TGF β 1 in Cancer Immunotherapy

We believe that our preclinical and safety data suggest that specific inhibition of the activation of latent TGF β 1 in combination with checkpoint inhibitors may have a significant impact in treating innate resistance to checkpoint immunotherapies. We are continuing to evaluate combinations of checkpoint inhibitors and our selective inhibitors of the activation of latent TGF β 1 in preclinical models of cancer immunotherapy.

Immune checkpoints are cellular mechanisms that act as a brake on the immune system, and tumors express these proteins in the tumor microenvironment to create an immunosuppressive environment to evade being killed by the immune system. Immune checkpoint proteins, such as PD-1/PD-L1, have therefore become key therapeutic targets in the tumor microenvironment. By inhibiting these proteins, the brakes on the immune system are released, allowing the T cells to kill the cancer cells. There are currently multiple approved immunotherapies that target the PD-1/PD-L1 pathway, including pembrolizumab, marketed as Keytruda, nivolumab, marketed as Opdivo, and atezolizumab, marketed as Tecentriq. A significant proportion of patients fail to respond to checkpoint inhibition because they have an innate resistance to immunotherapy or initially respond but subsequently progress.

Multiple peer-reviewed studies have implicated TGF β signaling in innate resistance to checkpoint inhibition. Whole-exome sequencing of pre-treatment melanoma tumors identified multiple TGF β -related signaling signatures associated with innate resistance to anti-PD-1 therapy. It has also been reported that retrospective pathway analysis of the atezolizumab bladder cancer trial identified the TGF β pathway as a major determinant of resistance to atezolizumab. The combination of atezolizumab with an anti-TGF β antibody in a tumor model, known as the EMT6 syngeneic tumor model, increased the number of complete responses to 70%, from 10% and 0% with treatment by atezolizumab or the anti-TGF β antibody alone, respectively. Our analysis of publicly available human tumor data has identified TGF β 1 as the predominant TGF β isoform in many human tumors, in particular for those cancers, such as bladder, lung and melanoma, where checkpoint therapies are already approved.

We have conducted both *in vitro* and *in vivo* mechanistic studies with our antibodies in order to evaluate whether our inhibitors of the activation of latent TGF β 1 may be effective in cancer immunotherapy. *In vitro*, we have observed that, by inhibiting the activation of latent TGF β 1, our antibodies suppressed the effect that human regulatory T cells have on the proliferation of human effector T cells. Moreover, in an *in vivo* model of colitis, we have observed that treatment with our antibodies increased immune system activity, a desired outcome in cancer immunotherapy.

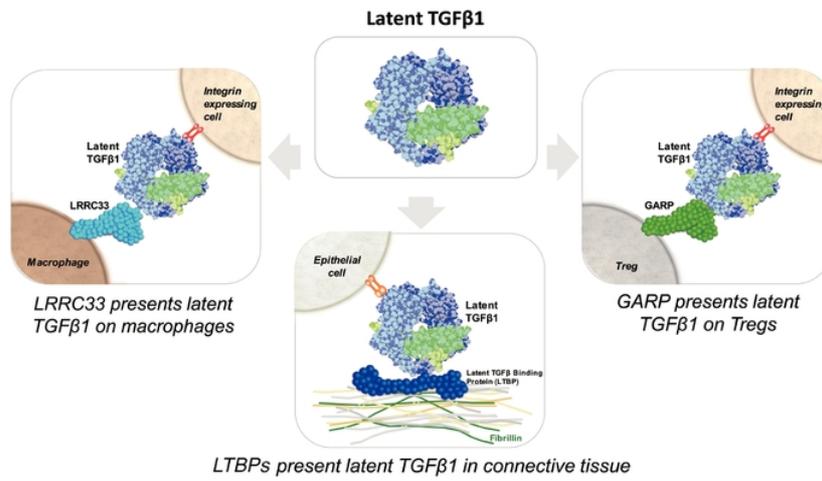
TGF β 1 in Myelofibrosis

Multiple peer-reviewed studies implicate TGF β 1 as a driver of fibrotic progression in myelofibrosis. We are currently evaluating our selective inhibitors of the activation of TGF β 1 in models of myelofibrosis, a hematological disorder characterized by fibrosis of the bone marrow. Myelofibrosis affects between 17,000 and 18,000 patients in the United States with significant morbidity and mortality. The only currently approved treatment for myelofibrosis, a JAK2 inhibitor, provides symptomatic benefit, but only modest reductions in bone marrow fibrosis. Therefore, we believe that significant unmet need remains for new therapeutic options.

TGF β 1 is produced by multiple cell types in the bone marrow microenvironment, including myofibroblasts, megakaryocytes and myeloid cells, and it has been shown to be upregulated in both human patient samples and preclinical mouse models of myelofibrosis. Inhibition of TGF β signaling with an ALK5 inhibitor reduced splenomegaly, collagen deposition and bone marrow fibrosis in a preclinical model of myelofibrosis. Furthermore, reconstitution of bone marrow with TGF β 1 knockout bone marrow stem cells in a model of hematological disease protected animals from bone marrow fibrosis, suggesting that TGF β 1 expression is necessary for disease pathogenesis.

Context-dependent inhibition of TGF β 1

When latent TGF β 1 is secreted from cells, it is further associated with a third protein, referred to as a presenting molecule. The presenting molecules are covalently bound to the prodomain, and serve to tether the latent TGF β 1 complex in a particular microenvironment. Unlike TGF β 1, a given presenting molecule's expression pattern is restricted to particular cellular and tissue environments. For example, the presenting molecule GARP is found primarily on regulatory T cells, or Tregs, the presenting molecules LTBP1 and LTBP3 are localized to the connective tissue in the extracellular matrix, and the presenting molecule LRRC33 is found primarily on certain myeloid lineage cells such as macrophages.



Latent TGFβ1 is associated with distinct presenting molecules in particular cellular and tissue environments.

Using our proprietary platform, we are able to identify antibodies that selectively inhibit the activation of latent TGFβ1 in the context of specific presenting molecules, which we refer to as context-dependent inhibition. For example, we have identified antibodies that specifically bind to and inhibit the activation of GARP-presented latent TGFβ1 on regulatory T cells with no detectable binding to latent TGFβ1 associated with other presenting molecules. These antibodies are the subject of our license agreement with Janssen.

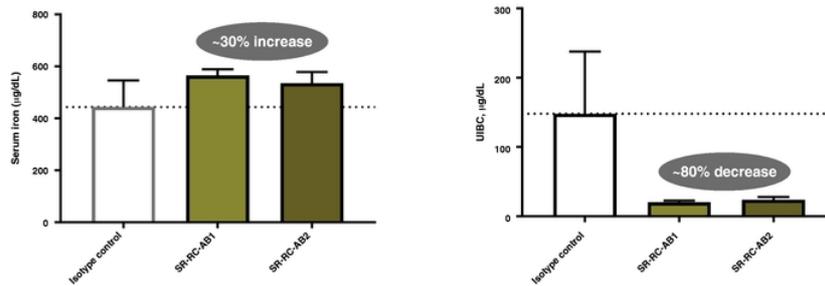
We have an active discovery program to identify antibodies that specifically bind to and inhibit the activation of LTBP1- and LTBP3-presented latent TGFβ1 with no cross-reactivity to GARP- or LRRC33-presented latent TGFβ1. We believe that such antibodies may have therapeutic potential for the treatment of organ fibrosis by inhibiting TGFβ1 function in connective tissue while having no impact on the activation or signaling of TGFβ1 in the immune system. We have identified antibodies with the desired binding specificity and *in vitro* inhibitory activity, and these are currently undergoing further optimization and characterization.

We also have an active discovery program to identify antibodies that specifically bind to and inhibit the activation of LRRC33-presented latent TGFβ1 with no cross-reactivity to LTBP1-, LTBP3- or GARP- presented latent TGFβ1. We believe that such antibodies may have therapeutic potential for specific oncology and cancer immunotherapy applications where selective modulation of myeloid lineage cells is desirable, for example inhibition of tumor-associated macrophages. We also have a related program to identify antibodies that specifically inhibit the activation of both LRRC33- and GARP-presented latent TGFβ1, with no cross-reactivity to LTBP1- or LTBP3-presented latent TGFβ1. We believe such antibodies could have broad inhibitory activity against TGFβ1 in the immune system for cancer immunotherapy, while avoiding inhibition of TGFβ1 in other tissues. We have identified antibodies that potentially meet the desired binding specificities, and these are currently undergoing characterization and further optimization.

BMP6 Signaling Program

We believe that liver-selective inhibition of BMP6 signaling could provide a way to target a variety of iron-restricted anemias, including anemia of chronic kidney disease, anemia of cancer and anemia of chronic inflammation. We are currently evaluating a limited number of our liver-selective inhibitors of BMP6 signaling in preclinical disease models of iron-restricted anemia.

BMPs are a broad subfamily of growth factors in the TGF β superfamily originally discovered by their ability to induce the formation of bone and cartilage. Beyond their association with bone, like many other growth factors, the BMPs are involved in a diverse set of biological processes. For example, while BMP6 plays roles in many different biologies, including fat metabolism and ovarian physiology, in the liver it functions as a critical control point in iron modulation in humans via regulation of hepcidin, a central regulator of iron homeostasis. Traditional approaches to inhibiting the signaling of BMP6 systemically would likely perturb the numerous different physiological processes in which BMP6 is involved. While the details of BMP6 activation are different from myostatin and TGF β 1, activation of BMP6 is a localized phenomenon, driven by a co-receptor molecule, RGMc, also known as hemojuvelin, which is required for BMP6 signaling upon binding to its receptor. RGMc is a member of a small family of proteins that include RGMa and RGMb. While each of these family members shares significant structural homology, particularly across their BMP binding domains, their physiological roles are quite different. RGMa and RGMb are reported to have roles in nervous system biology, immunity, inflammation, angiogenesis, and growth. Unlike RGMa and RGMb, RGMc's known function is localized to hepatocytes. As such identification of RGMc selective-antibodies that do not bind to RGMa or RGMb could provide the potential for liver-specific modulation of BMP6 biology. Utilizing our structural biology insights into BMP6 and its co-receptors, we have identified highly specific inhibitors of RGMc's interaction with BMP6 and, as shown in the figure below, in a preclinical study in rats we have shown proof-of-principle that our antibodies can modulate iron levels *in vivo*.



SR-RC-AB1 and SR-RC-AB2, two of our selective BMP6 signaling inhibitors, increased serum iron in rats as compared to control (left side) and reduced unsaturated iron binding capacity, or UIBC, in healthy rats (right side).

License Agreements

License Agreement with Janssen

On December 17, 2013 we entered into an option and license agreement with Janssen, or the Janssen Agreement. Pursuant to the Janssen Agreement, Janssen funded our drug discovery research to identify molecules with either one or two pharmacological profiles, over a two-year period beginning on December 17, 2013, or the collaboration period. During the collaboration period, we granted Janssen a non-exclusive license to research, develop, and use the collaboration molecule(s) and/or lead molecule(s). Janssen was not granted a license to commercialize any collaboration molecule, lead molecule or a product

that is derived from an optioned molecule, or a licensed product, unless and until Janssen exercised its license option in accordance with the Janssen Agreement. We received funding from Janssen based on a set rate per annual full-time equivalent personnel working on the research plus actual external costs incurred by us up to a maximum dollar amount as specified in the Janssen Agreement, with costs approximating the funding provided. During the collaboration period, we billed Janssen quarterly, in arrears, based on time and actual costs incurred, and Janssen was not entitled to any refunds.

The activities under the Janssen Agreement were governed by a program committee, consisting of three members from each of our company and Janssen, with all decisions being by unanimous vote or written consent, subject to an escalating dispute resolution procedure in the event any disputes could be not resolved by the program committee.

We also granted Janssen an option to exclusively license molecules identified during the collaboration period that meet either one or both pharmacological profiles by providing us with written notice and paying an option exercise fee of \$1.0 million per option exercised (up to two). If Janssen failed to exercise its license option by the end of the collaboration period, the term could be extended for up to one additional year by mutual written agreement of the parties. Once Janssen exercised its option, our obligations under the program plan for the molecule and related pharmacological profile ceased and Janssen assumed full responsibility for further development of the molecules at its sole cost, and we were obligated to transfer any and all manufacturing related activities for such molecule to Janssen at Janssen's sole cost. In December 2015, Janssen exercised its option for collaboration molecules for one pharmacological profile, the selective inhibition of TGF β 1 in the context of regulatory T cells. In addition, the parties agreed to extend the collaboration period for the second pharmacological profile through March 31, 2016. The option exercise period for this profile expired unexercised on March 31, 2016, and all rights with respect to molecules generated during the collaboration period with respect to this second pharmacological profile were retained by us.

After Janssen exercised its option, it became obligated to pay us up to \$25 million upon the achievement of specified development milestones and up to \$97 million upon the achievement of specified regulatory milestones. In addition for any licensed product, Janssen is required to pay to us up to \$130 million upon the achievement of specified annual net sales thresholds. For a period commencing on the first commercial sale of a product, on a product-by-product and country-by-country basis, until the latest to occur of (i) the expiration date of the last valid claim within the licensed patent rights covering the licensed product, (ii) the tenth anniversary date of the first commercial sale of a licensed product, or (iii) the termination or expiration of regulatory exclusivity for a licensed product, such period the royalty period, Janssen is required to pay to us, single digit percentage tiered royalties based on annual net sales thresholds.

The Janssen Agreement will expire on a country-by-country basis on the expiration of the last royalty period for a licensed product within such country. Janssen has the right to terminate the Janssen Agreement, in whole or in part, without cause upon 90 days written notice to us. In addition, either we, or Janssen may terminate the Janssen Agreement if the other party commits a material breach of the agreement and fails to cure such breach within 60 days (or 30 days in the case of a failure to make any payment) after written notice is provided, or, upon the other party's bankruptcy, insolvency, dissolution or winding up. Upon termination, any licensed product reverts to us and if Janssen has commenced clinical trials for such licensed product, upon commercialization of such licensed product, we will be required to pay Janssen single digit percentage tiered royalties on such licensed product based on annual net sales thresholds.

License Agreement with Children's Medical Corporation

On December 17, 2013, we entered into an exclusive license agreement with Children's Medical Center Corporation, or CMCC, or the CMCC Agreement, to gain exclusive control over co-owned patent rights related to our platform technology. Under the CMCC Agreement, we received an exclusive worldwide license to CMCC's rights in certain patent rights jointly owned by us and CMCC, to develop and commercialize any product or process that but for the licenses granted to us under the CMCC Agreement would infringe such

patent rights, a licensed product and licensed process, respectively, for any use. We are entitled to sublicense the rights granted to us under the CMCC Agreement. These licenses and rights are subject to certain limitations and retained rights, including retained rights to practice and use the patent rights for research, educational, clinical and charitable purposes. In addition, the CMCC Agreement obligates us to meeting certain diligence milestones, including obligations to raise funds, seek collaborations and initiate discovery efforts.

As consideration for the license, we paid CMCC a non-refundable license fee of \$5,000 and issued to CMCC common units, which were exchanged for shares of common stock in connection with the Reorganization and subject to a subsequent 2.8548-to-one reverse split for an aggregate of 26,796 issued shares. We must pay CMCC annual license maintenance fees, which were \$5,000 through 2016 and increased to \$10,000 for 2017 and each year thereafter. We will also be responsible for up to \$1.3 million of development and regulatory milestone payments through the first regulatory approval of a licensed product, tiered royalty payments of low single-digit percentages on net sales of licensed products in the event that we realize sales from products covered by the license agreement, and between 10% and 20% of non-royalty income attributable to a sublicense of the CMCC rights. Such products include products developed using our proprietary platform that are covered by a valid claim contained in any patent under the license agreement. Amounts paid to CMCC are recorded as research and development expense in the statement of operations. The royalty term will terminate on the expiration date of the last valid claim within the licensed patent rights.

CMCC may terminate the CMCC Agreement if we commit a breach of the agreement, and fail to cure such breach within 60 days (or 30 days in the case of our failure to make any payment) after written notice is provided, or immediately upon our bankruptcy, insolvency, dissolution or winding up, or upon 30 days' notice if we bring patent challenges relating to any patent families licensed by us under the CMCC Agreement. In addition, we may terminate the CMCC Agreement for convenience upon three months prior written notice to CMCC. Upon expiration of the CMCC Agreement, we will have a worldwide, perpetual, irrevocable, sublicensable license to the intellectual property previously covered by the CMCC Agreement.

Intellectual Property

Our commercial success depends in part on our ability to protect intellectual property for our product candidates, including our lead product candidate SRK-015, and related methods, as well as our novel approach and proprietary platform for generating monoclonal antibodies; to secure freedom to operate to enable commercialization of our product candidates, if approved; and to prevent others from infringing upon our patent rights. Our policy is to seek to protect our intellectual property position by filing patent applications in key jurisdictions, including the United States, Europe, Canada, Japan and Australia, covering our proprietary technology, inventions and improvements that are important to innovate, develop, sustain and implement our business.

We file patent applications directed to compositions comprising our antibodies, classes of antibodies covering our product candidates, use of such antibodies for treating diseases, as well as related manufacturing methods. As of May 7, 2018, we have 12 international patent families (PCT filings) pending. Among the pending families, six have been nationalized, in which five applications have matured into U.S. issued patents, two granted in Australia, one granted in Singapore, and one granted in South Africa. Collectively, there are 41 national utility applications pending. In addition, there are three patent family filings which are in the priority year. We continue to review new inventions for new patent filings. As of May 7, 2018, for our BMP6 program, we had no issued patents and one pending application for a method of use patent directed to the therapeutic use of certain inhibitors.

We have no contested proceedings or third-party claims relating to any patents at this time, but we can not provide any assurances that we will not have such proceedings or third-party claims at a later date.

Ownership and IP Rights

Our earliest patent family, PCT/US2013/068613 (published as WO 2014/074532), is jointly owned by us and CMCC. CMCC is the assignee of the intellectual property rights transferred from two of our co-founders, Drs. Timothy A. Springer and Leonard I. Zon. The portion of rights owned by CMCC is exclusively licensed to us. We are the sole legal owner of all subsequent patent families we have to date.

As described, a portion of our TGFb technology is out-licensed to Janssen. This is carved out as PCT/US2017/042162 (published as WO 2018/013939). The licensee takes lead in the prosecution of this patent family. The licensee also has a non-exclusive license to our platform technology to enable their development in the licensed field.

Brief descriptions of our patent families are provided below, with projected patent terms excluding any possible patent term adjustments or extensions.

Platform

Our novel approach to generating selective modulators of supracellular activation of growth factors is broadly embodied in our two earliest patent families, PCT/US2013/068613 (published as WO 2014/074532) and PCT/US2014/036933 (published as WO 2014/182676). These patent families are directed to methods for modulating the activation of the TGFb superfamily of growth factors by using a monoclonal antibody that specifically targets an inactive form of the growth factor, thereby preventing release of mature growth factor from its latent complex. The TGFb superfamily is a group of more than 30 related growth factors that mediate diverse biological processes and includes TGFb1 and myostatin (also known as GDF-8). Issued U.S. patents include: U.S. Patents Nos. 9,573,995 (issued 02/21/2017); 9,758,576 (issued 09/12/2017); 9,580,500 (issued 02/28/2017); 9,399,676 (issued 07/26/2016) and 9,758,577 (issued 09/12/2017). These patents are projected to expire in 2034.

Specifically, U.S. Patent No. 9,573,995 has issued composition of matter claims directed to an antibody that specifically binds to GARP associated with a human TGFb1 LAP complex.

U.S. Patent No. 9,758,576 has issued composition of matter claims directed to an isolated monoclonal antibody, or a fragment thereof, that specifically binds the prodomain of a pro/latent GDF-8 complex, thereby preventing proteolytic cleavage between residues Arg 75 and Asp 76 of GDF-8 prodomain, so as to inhibit the release of mature GDF-8 growth factor from the complex.

U.S. Patent No. 9,580,500 has issued claims directed to phage display library-based antibody production methods for identifying an antibody that binds a GARP/proTGFb1 complex.

U.S. Patent No. 9,399,676 has issued claims directed to phage display library-based antibody production methods for identifying an antibody that binds a pro/latent GDF-8 complex that has been subjected to enzymatic cleavage. Related product-by-process claims are included in issued U.S. Patent No. 9,758,577.

Myostatin Activation Inhibitors

Five patent families have been filed to date to cover proprietary myostatin inhibitors and their use in the treatment of various muscle diseases. Patent prosecution of these five pending patent families is in the early stages, and no patents have issued to date.

Two families are directed to composition of matter claims that cover our proprietary antibodies. PCT/US2015/059468 filed November 6, 2015, broadly covers a class of monoclonal antibodies that specifically bind inactive precursors thereby preventing activation of myostatin. This patent family is projected to expire in November 2035. A second family, PCT/US2016/052014 filed September 15, 2016, discloses the specific amino acid sequence of SRK-015 and is projected to expire in September 2036.

In addition, the following three patent families are directed to therapeutic use/methods. PCT/US2017/012606 (published as WO 2017/120523) broadly covers treatment methods for a number of muscle and neuromuscular disease and disorders with the use of an antibody that specifically blocks the

activation step of myostatin. This patent family is projected to expire in January 2037. PCT/US2017/037332 (published as WO 2017/218592) is directed to methods for treating neuromuscular diseases and selecting patient populations that are likely to respond to myostatin inhibition. This filing includes the treatment of SMA in patients who are on an SMN upregulator therapy. This patent family is projected to expire in June 2037. Finally, PCT/US2018/012686 (expected to publish in July 2018) relates to the treatment of metabolic diseases with the use of a myostatin activation inhibitor and is projected to expire in January 2038.

In addition to the five pending patent families listed above, the issued claims of U.S. Patent 9,758,576 from the platform patents discussed in detail above cover monoclonal antibodies that selectively inhibit myostatin signaling by blocking the proteolytic activation of latent myostatin. These issued composition of matter claims provide protection for our lead antibody SRK-015, as well as any other monoclonal antibodies that work by this unique mechanism of action. This patent expires in May 2034, not including any potential patent term extension.

TGFb1 Activation Inhibitors

Five patent families have been filed to date, covering various aspects of our TGFb program. Patent prosecution of these five pending patent families is in the early stages, and no patents have issued to date. Isoform-specific inhibitors of TGFb1 and related methods are described in PCT/US2017/021972 (published as WO 2017/156500). This family is projected to expire in March 2037. Among TGFb1 inhibitors, one of our leading context-independent antibodies is separately claimed and related preclinical data are described in PCT/US2018/012601. This patent application is expected to publish in July 2018 and is projected to expire in January 2038.

PCT/US2017/042162 (published as WO 2018/013939) is a collaboration patent family exclusively licensed to Janssen. This patent family covers antibodies that specifically inhibit GARP-associated TGFb1, and is projected to expire in July 2037. Janssen takes prosecution lead in this case.

Two additional patent families related to the TGFb program have been filed and are still in the priority year. These provisional applications will be converted to international patent applications (PCT) in May and July 2018, respectively.

Intellectual Property Protection

We cannot predict whether the patent applications we pursue will issue as patents in any particular jurisdiction or whether the claims of any issued patents will provide any proprietary protection from competitors. Even if our pending patent applications are granted as issued patents, those patents, as well as any patents we license from third parties, may be challenged, circumvented or invalidated by third parties. While there are currently no contested proceedings or third-party claims relating to any of the patents described above, we cannot provide any assurances that we will not have such proceedings or third-party claims at a later date.

The term of individual patents depends upon the legal term of the patents in the countries in which they are obtained. In most countries in which we file, the patent term is 20 years from the earliest date of filing a non-provisional patent application. In the United States, the patent term of a patent that covers an FDA-approved drug or biologic may also be eligible for patent term extension, which permits patent term restoration as compensation for the patent term lost during FDA regulatory review process. The Hatch-Waxman Amendments permit a patent term extension of up to five years beyond the expiration of the patent. The length of the patent term extension is related to the length of time the drug or biologic is under regulatory review. Patent term extension cannot extend the remaining term of a patent beyond a total of 14 years from the date of product approval and only one patent applicable to an approved drug or biologic may be extended. Similar provisions are available in Europe and other foreign jurisdictions to extend the term of a patent that covers an approved drug or biologic or provide an additional period of protection for the approved pharmaceutical product following expiry of the patent. In the future, if our products receive

FDA approval, we expect to apply for patent term extensions on patents covering those products. We plan to seek patent term extensions to any of our issued patents in any jurisdiction where these are available, however there is no guarantee that the applicable authorities, including the U.S. Patent and Trademark Office in the United States and the national patent offices in Europe, will agree with our assessment of whether such extensions should be granted, and if granted, the length of such extensions.

In addition to our reliance on patent protection for our inventions, product candidates and research programs, we also rely on trade secret protection for our confidential and proprietary information. For example, certain elements of our proprietary platform may be based on unpatented trade secrets that are not publicly disclosed. Although we take steps to protect our proprietary information and trade secrets, including through contractual means with our employees and consultants, third parties may independently develop substantially equivalent proprietary information and techniques or otherwise gain access to our trade secrets or disclose our technology. Thus, we may not be able to meaningfully protect our trade secrets. It is our policy to require our employees, consultants, outside scientific collaborators, sponsored researchers and other advisors to execute confidentiality agreements upon the commencement of employment or consulting relationships with us. These agreements provide that all confidential information concerning our business or financial affairs developed or made known to the individual or entity during the course of the party's relationship with us is to be kept confidential and not disclosed to third parties except in specific circumstances. In the case of employees, the agreements provide that all inventions conceived by the individual, and which are related to our current or planned business or research and development or made during normal working hours, on our premises or using our equipment or proprietary information, are our exclusive property. In addition, we take other appropriate precautions, such as physical and technological security measures, to guard against misappropriation of our proprietary technology by third parties. We have also adopted policies and conduct training that provides guidance on our expectations, and our advice for best practices, in protecting our trade secrets.

Manufacturing

We do not own or operate facilities for clinical drug manufacturing, storage, distribution or quality testing. Currently, all of our clinical manufacturing is outsourced to third-party manufacturers. As our development programs expand and we build new process efficiencies, we expect to continually evaluate this strategy with the objective of satisfying demand for registration trials and, if approved, the manufacture, sale and distribution of commercial products.

Antibody Discovery

We rely on third party entities to conduct antibody discovery based on criteria and specifications provided by us. Certain antibody discovery vendors require us to enter into a license agreement with them for the right to use antibodies discovered by them in humans or for commercial purposes. Such license agreement could include substantial milestone payments and royalties to the extent we choose to use an antibody discovered by such vendor. While we have not executed such an agreement to date, there can be no assurance that we will not be required to execute such a license agreement at a later date if we select a clinical candidate that includes such an antibody and advance that clinical candidate into clinical trials.

Competition

The biotechnology and pharmaceutical industries are characterized by rapid evolution of technologies, fierce competition and strong defense of intellectual property. While we believe that our product candidates, discovery programs, technology, knowledge, experience and scientific resources provide us with competitive advantages, we face competition from major pharmaceutical and biotechnology companies, academic institutions, governmental agencies and public and private research institutions, among others.

Any product candidates that we successfully develop and commercialize will compete with currently approved therapies and new therapies that may become available in the future. Key product features that would affect our ability to effectively compete with other therapeutics include the efficacy, safety and convenience of our products.

At this time, there are no FDA- or EMA-approved muscle-directed treatments for SMA. We believe SRK-015 may be used in conjunction with SMN upregulators. Biogen markets nusinersen, the only currently marketed SMN upregulator. AveXis, Inc., Génethon, Novartis and Roche have SMN upregulators in various stages of preclinical or clinical development. In addition, Catalyst Pharmaceuticals, Inc., Cytokinetics Incorporated and Roche are developing investigational agents with other mechanisms of action for the treatment of SMA.

Acceleron Pharma, Inc., Novartis, Pfizer, Regeneron Pharmaceuticals, Inc. and Roche are developing therapies for muscle-wasting diseases, other than SMA, that are intended to work, at least in part, through inhibition of the myostatin signaling pathway.

Our competitors may also include companies that are or will be developing therapies for the same therapeutic areas that we are targeting within our early pipeline, including other neuromuscular disorders, cancer, fibrosis and anemia.

Many of the companies against which we may compete have significantly greater financial resources and expertise in research and development, manufacturing, preclinical testing, conducting clinical trials, obtaining regulatory approvals and marketing approved products than we do. These competitors also compete with us in recruiting and retaining qualified scientific and management personnel and establishing clinical trial sites and patient registration for clinical trials, as well as in acquiring technologies complementary to, or necessary for, our programs.

The availability of reimbursement from government and other third-party payors will also significantly affect the pricing and competitiveness of our products. Our competitors also may obtain FDA or other regulatory approval for their products more rapidly than we may obtain approval for ours, which could result in our competitors establishing a strong market position before we are able to enter the market.

Government Regulation

Government authorities in the United States at the federal, state and local level and in other countries regulate, among other things, the research, development, testing, manufacture, quality control, approval, labeling, packaging, storage, record-keeping, promotion, advertising, distribution, post-approval monitoring and reporting, marketing and export and import of drug and biological products, such as SRK-015 and any future product candidates. Generally, before a new drug or biologic can be marketed, considerable data demonstrating its quality, safety and efficacy must be obtained, organized into a format specific for each regulatory authority, submitted for review and approved by the regulatory authority.

U.S. Biological Product Development

In the United States, the FDA regulates drugs under the Federal Food, Drug, and Cosmetic Act, or FDCA, and its implementing regulations and biologics under the FDCA, the Public Health Service Act, or PHSA, and their implementing regulations. Both drugs and biologics also are subject to other federal, state and local statutes and regulations. The process of obtaining regulatory approvals and the subsequent compliance with appropriate federal, state and local statutes and regulations requires the expenditure of substantial time and financial resources. Failure to comply with the applicable U.S. requirements at any time during the product development process, approval process or post-market may subject an applicant to administrative or judicial sanctions. These sanctions could include, among other actions, the FDA's refusal to approve pending applications, withdrawal of an approval, a clinical hold, untitled or warning letters, product recalls or market withdrawals, product seizures, total or partial suspension of production or distribution, injunctions, fines, refusals of government contracts, restitution, disgorgement and civil or criminal penalties. Any agency or judicial enforcement action could have a material adverse effect on us.

SRK-015 and any future product candidates must be approved by the FDA through a Biologics License Application, or BLA, process before they may be legally marketed in the United States. The process generally involves the following:

- § Completion of extensive preclinical studies in accordance with applicable regulations, including studies conducted in accordance with good laboratory practice, or GLP, requirements;
- § Submission to the FDA of an IND application, which must become effective before human clinical trials may begin;
- § Approval by an institutional review board, or IRB, or independent ethics committee at each clinical trial site before each trial may be initiated;
- § Performance of adequate and well-controlled human clinical trials in accordance with applicable IND regulations, good clinical practice, or GCP, requirements and other clinical trial-related regulations to establish the safety and efficacy of the investigational product for each proposed indication;
- § Submission to the FDA of a BLA;
- § A determination by the FDA within 60 days of its receipt of a BLA to accept the filing for review;
- § Satisfactory completion of an FDA pre-approval inspection of the manufacturing facility or facilities where the biologic will be produced to assess compliance with cGMP requirements to assure that the facilities, methods and controls are adequate to preserve the biologic's identity, strength, quality and purity;
- § Potential FDA audit of the clinical trial sites that generated the data in support of the BLA; and
- § FDA review and approval of the BLA, including consideration of the views of any FDA advisory committee, prior to any commercial marketing or sale of the biologic in the United States.

The preclinical and clinical testing and approval process requires substantial time, effort and financial resources, and we cannot be certain that any approvals for SRK-015 and any future product candidates will be granted on a timely basis, or at all.

Preclinical Studies and IND

Preclinical studies include laboratory evaluation of product chemistry and formulation, as well as *in vitro* and animal studies to assess the potential for adverse events and in some cases to establish a rationale for therapeutic use. The conduct of preclinical studies is subject to federal regulations and requirements, including GLP regulations for safety/toxicology studies.

An IND sponsor must submit the results of the preclinical tests, together with manufacturing information, analytical data, any available clinical data or literature and plans for clinical studies, among other things, to the FDA as part of an IND. An IND is a request for authorization from the FDA to administer an investigational product to humans, and must become effective before human clinical trials may begin. Some long-term preclinical testing may continue after the IND is submitted. An IND automatically becomes effective 30 days after receipt by the FDA, unless before that time, the FDA raises concerns or questions related to one or more proposed clinical trials and places the trial on clinical hold. In such a case, the IND sponsor and the FDA must resolve any outstanding concerns before the clinical trial can begin. As a result, submission of an IND may not result in the FDA allowing clinical trials to commence.

Clinical Trials

The clinical stage of development involves the administration of the investigational product to healthy volunteers or patients under the supervision of qualified investigators, generally physicians not employed by or under the trial sponsor's control, in accordance with GCP requirements, which include the requirement that all patients provide their informed consent for their participation in any clinical trial. Clinical trials are conducted under protocols detailing, among other things, the objectives of the clinical trial, dosing procedures, subject selection and exclusion criteria and the parameters to be used to monitor subject safety and assess efficacy. Each protocol, and any subsequent amendments to the protocol, must be submitted to

the FDA as part of the IND. Furthermore, each clinical trial must be reviewed and approved by an IRB for each institution at which the clinical trial will be conducted to ensure that the risks to individuals participating in the clinical trials are minimized and are reasonable in relation to anticipated benefits. The IRB also approves the informed consent form that must be provided to each clinical trial subject or his or her legal representative, and must monitor the clinical trial until completed. There also are requirements governing the reporting of ongoing clinical trials and completed clinical trial results to public registries.

A sponsor who wishes to conduct a clinical trial outside of the United States may, but need not, obtain FDA authorization to conduct the clinical trial under an IND. If a foreign clinical trial is not conducted under an IND, the sponsor may submit data from the clinical trial to the FDA in support of a BLA. The FDA will accept a well-designed and well-conducted foreign clinical study not conducted under an IND if the study was conducted in accordance with GCP requirements, and the FDA is able to validate the data through an onsite inspection if deemed necessary.

Clinical trials generally are conducted in three sequential phases, known as Phase 1, Phase 2 and Phase 3, and may overlap.

- § Phase 1 clinical trials generally involve a small number of healthy volunteers or disease-affected patients who are initially exposed to a single dose and then multiple doses of the product candidate. The primary purpose of these clinical trials is to assess the metabolism, pharmacologic action, side effect tolerability and safety of the product candidate.
- § Phase 2 clinical trials generally involve studies in disease-affected patients to evaluate proof of concept and/or determine the dosing regimen(s) for subsequent investigations. At the same time, safety and further pharmacokinetic and pharmacodynamic information is collected, possible adverse effects and safety risks are identified and a preliminary evaluation of efficacy is conducted.
- § Phase 3 clinical trials generally involve a large number of patients at multiple sites and are designed to provide the data necessary to demonstrate the effectiveness of the product for its intended use, its safety in use and to establish the overall benefit/risk relationship of the product and provide an adequate basis for product labeling.

Post-approval trials, sometimes referred to as Phase 4 clinical trials, may be conducted after initial marketing approval. These trials are used to gain additional experience from the treatment of patients in the intended therapeutic indication. In certain instances, the FDA may mandate the performance of Phase 4 clinical trials as a condition of approval of a BLA.

Progress reports detailing the results of the clinical trials, among other information, must be submitted at least annually to the FDA and written IND safety reports must be submitted to the FDA and the investigators for serious and unexpected suspected adverse events, findings from other studies or animal or *in vitro* testing that suggest a significant risk for human subjects and any clinically important increase in the rate of a serious suspected adverse reaction over that listed in the protocol or investigator brochure.

Phase 1, Phase 2 and Phase 3 clinical trials may not be completed successfully within any specified period, if at all. The FDA or the sponsor may suspend or terminate a clinical trial at any time on various grounds, including a finding that the patients are being exposed to an unacceptable health risk. Similarly, an IRB can suspend or terminate approval of a clinical trial at its institution if the clinical trial is not being conducted in accordance with the IRB's requirements or if the drug or biologic has been associated with unexpected serious harm to patients. Additionally, some clinical trials are overseen by an independent group of qualified experts organized by the clinical trial sponsor, known as a data safety monitoring board or committee. This group provides authorization for whether a trial may move forward at designated check points based on access to certain data from the trial. Concurrent with clinical trials, companies usually complete additional animal studies and also must develop additional information about the chemistry and physical characteristics of the drug or biologic as well as finalize a process for manufacturing the product in commercial quantities in accordance with cGMP requirements. The manufacturing process must be capable

of consistently producing quality batches of the product and, among other things, companies must develop methods for testing the identity, strength, quality and purity of the final product. Additionally, appropriate packaging must be selected and tested and stability studies must be conducted to demonstrate that the product candidates do not undergo unacceptable deterioration over their shelf life.

FDA Review Process

Following completion of the clinical trials, data are analyzed to assess whether the investigational product is safe and effective for the proposed indicated use or uses. The results of preclinical studies and clinical trials are then submitted to the FDA as part of a BLA, along with proposed labeling, chemistry and manufacturing information to ensure product quality and other relevant data. The BLA is a request for approval to market the biologic for one or more specified indications and must contain proof of safety and efficacy for a drug or safety, purity and potency for a biologic. The application may include both negative and ambiguous results of preclinical studies and clinical trials, as well as positive findings. Data may come from company-sponsored clinical trials intended to test the safety and efficacy of a product's use or from a number of alternative sources, including studies initiated by investigators. To support marketing approval, the data submitted must be sufficient in quality and quantity to establish the safety and efficacy of the investigational product to the satisfaction of the FDA. FDA approval of a BLA must be obtained before a biologic may be marketed in the United States.

Under the Prescription Drug User Fee Act, or PDUFA, as amended, each BLA must be accompanied by a user fee. The FDA adjusts the PDUFA user fees on an annual basis. Fee waivers or reductions are available in certain circumstances, including a waiver of the application fee for the first application filed by a small business. Additionally, no user fees are assessed on BLAs for products designated as orphan drugs, unless the product also includes a non-orphan indication.

The FDA reviews all submitted BLAs before it accepts them for filing, and may request additional information rather than accepting the BLA for filing. The FDA must make a decision on accepting a BLA for filing within 60 days of receipt, and such decision could include a refusal to file (RTF) by the FDA. Once the submission is accepted for filing, the FDA begins an in-depth review of the BLA. Under the goals and policies agreed to by the FDA under PDUFA, the FDA has 10 months, from the filing date, in which to complete its initial review of an original BLA and respond to the applicant, and six months from the filing date of an original BLA designated for priority review. The FDA does not always meet its PDUFA goal dates for standard and priority BLAs, and the review process is often extended by FDA requests for additional information or clarification.

Before approving a BLA, the FDA will conduct a pre-approval inspection of the manufacturing facilities for the new product to determine whether they comply with cGMP requirements. The FDA will not approve the product unless it determines that the manufacturing processes and facilities are in compliance with cGMP requirements and adequate to assure consistent production of the product within required specifications. The FDA also may audit data from clinical trials to ensure compliance with GCP requirements. Additionally, the FDA may refer applications for novel products or products which present difficult questions of safety or efficacy to an advisory committee, typically a panel that includes clinicians and other experts, for review, evaluation and a recommendation as to whether the application should be approved and under what conditions, if any. The FDA is not bound by recommendations of an advisory committee, but it considers such recommendations when making decisions on approval. The FDA likely will reanalyze the clinical trial data, which could result in extensive discussions between the FDA and the applicant during the review process. After the FDA evaluates a BLA, it will issue an approval letter or a Complete Response Letter. An approval letter authorizes commercial marketing of the biologic with specific prescribing information for specific indications. A Complete Response Letter indicates that the review cycle of the application is complete and the application will not be approved in its present form. A Complete Response Letter usually describes all of the specific deficiencies in the BLA identified by the FDA. The Complete Response Letter may require additional clinical data, additional pivotal Phase 3 clinical trial(s) and/or other significant and time-consuming requirements related to clinical trials, preclinical studies or manufacturing. If a Complete

Response Letter is issued, the applicant may either resubmit the BLA, addressing all of the deficiencies identified in the letter, or withdraw the application. Even if such data and information are submitted, the FDA may decide that the BLA does not satisfy the criteria for approval. Data obtained from clinical trials are not always conclusive and the FDA may interpret data differently than we interpret the same data.

Orphan Drug Designation

Under the Orphan Drug Act, the FDA may grant orphan designation to a drug or biological product intended to treat a rare disease or condition, which is generally a disease or condition that affects fewer than 200,000 individuals in the United States, or more than 200,000 individuals in the United States and for which there is no reasonable expectation that the cost of developing and making the product available in the United States for this type of disease or condition will be recovered from sales of the product. On March 22, 2018, the FDA granted orphan drug designation for SRK-015 for the treatment of SMA.

Orphan drug designation must be requested before submitting a BLA. After the FDA grants orphan drug designation, the identity of the therapeutic agent and its potential orphan use are disclosed publicly by the FDA. Orphan drug designation does not convey any advantage in or shorten the duration of the regulatory review and approval process.

If a product that has orphan designation subsequently receives the first FDA approval for the disease or condition for which it has such designation, the product is entitled to orphan drug exclusivity, which means that the FDA may not approve any other applications to market the same drug for the same indication for seven years from the date of such approval, except in limited circumstances, such as a showing of clinical superiority to the product with orphan exclusivity by means of greater effectiveness, greater safety or providing a major contribution to patient care or in instances of drug supply issues. Competitors, however, may receive approval of either a different product for the same indication or the same product for a different indication but that could be used off-label in the orphan indication. Orphan drug exclusivity also could block the approval of one of our products for seven years if a competitor obtains approval before we do for the same product, as defined by the FDA, for the same indication we are seeking approval, or if our product is determined to be contained within the scope of the competitor's product for the same indication or disease. If one of our products designated as an orphan drug receives marketing approval for an indication broader than that which is designated, it may not be entitled to orphan drug exclusivity. Orphan drug status in the European Union has similar, but not identical, requirements and benefits.

Expedited Development and Review Programs

The FDA has a fast track program that is intended to expedite or facilitate the process for reviewing new drugs and biologics that meet certain criteria. Specifically, new drugs and biologics are eligible for fast track designation if they are intended to treat a serious or life threatening condition and preclinical or clinical data demonstrate the potential to address unmet medical needs for the condition. Fast track designation applies to both the product and the specific indication for which it is being studied. The sponsor can request the FDA to designate the product for fast track status any time before receiving BLA approval, but ideally no later than the pre-BLA meeting. Any product submitted to the FDA for marketing, including under a fast track program, may be eligible for other types of FDA programs intended to expedite development and review, such as priority review and accelerated approval. Any product is eligible for priority review if it treats a serious or life-threatening condition and, if approved, would provide a significant improvement in safety and effectiveness compared to available therapies. The FDA will attempt to direct additional resources to the evaluation of an application for a new drug or biologic designated for priority review in an effort to facilitate the review.

A product may also be eligible for accelerated approval, if it treats a serious or life-threatening condition and generally provides a meaningful advantage over available therapies. In addition, it must demonstrate an effect on a surrogate endpoint that is reasonably likely to predict clinical benefit or on a clinical endpoint that can be measured earlier than irreversible morbidity or mortality, or IMM, that is reasonably likely to predict an effect on IMM or other clinical benefit. As a condition of approval, the FDA may require that a

sponsor of a drug or biologic receiving accelerated approval perform adequate and well-controlled post-marketing clinical trials. If the FDA concludes that a drug or biologic shown to be effective can be safely used only if distribution or use is restricted, it will require such post-marketing restrictions, as it deems necessary to assure safe use of the product. If the FDA determines that the conditions of approval are not being met, the FDA can withdraw its accelerated approval for such drug or biologic.

Additionally, a drug or biologic may be eligible for designation as a breakthrough therapy if the product is intended, alone or in combination with one or more other drugs or biologics, to treat a serious or life-threatening condition and preliminary clinical evidence indicates that the product may demonstrate substantial improvement over currently approved therapies on one or more clinically significant endpoints. The benefits of breakthrough therapy designation include the same benefits as fast track designation, plus intensive guidance from the FDA to ensure an efficient drug development program.

Fast track designation, priority review, accelerated approval and breakthrough therapy designation do not change the standards for approval, but may expedite the development or approval process.

Pediatric Information

Under the Pediatric Research Equity Act, or PREA, a BLA or supplement to a BLA must contain data to assess the safety and efficacy of the drug for the claimed indications in all relevant pediatric subpopulations and to support dosing and administration for each pediatric subpopulation for which the product is safe and effective. The FDA may grant deferrals for submission of pediatric data or full or partial waivers. A sponsor who is planning to submit a marketing application for a drug that includes a new active ingredient, new indication, new dosage form, new dosing regimen or new route of administration submit an initial Pediatric Study Plan, or PSP, within 60 days of an end-of-Phase 2 meeting or, if there is no such meeting, as early as practicable before the initiation of the Phase 3 or Phase 2/3 study. The initial PSP must include an outline of the pediatric study or studies that the sponsor plans to conduct, including study objectives and design, age groups, relevant endpoints and statistical approach, or a justification for not including such detailed information, and any request for a deferral of pediatric assessments or a full or partial waiver of the requirement to provide data from pediatric studies along with supporting information. The FDA and the sponsor must reach an agreement on the PSP. A sponsor can submit amendments to an agreed-upon initial PSP at any time if changes to the pediatric plan need to be considered based on data collected from preclinical studies, early phase clinical trials and/or other clinical development programs.

Post-marketing Requirements

Following approval of a new product, the manufacturer and the approved product are subject to continuing regulation by the FDA, including, among other things, monitoring and record-keeping activities, reporting of adverse experiences, complying with promotion and advertising requirements, which include restrictions on promoting products for unapproved uses or patient populations (known as "off-label use") and limitations on industry-sponsored scientific and educational activities. Although physicians may prescribe legally available products for off-label uses, manufacturers may not market or promote such uses. Prescription drug and biologic promotional materials must be submitted to the FDA in conjunction with their first use. Further, if there are any modifications to the drug or biologic, including changes in indications, labeling or manufacturing processes or facilities, the applicant may be required to submit and obtain FDA approval of a new BLA or BLA supplement, which may require the development of additional data or preclinical studies and clinical trials.

The FDA may also place other conditions on approvals including the requirement for a Risk Evaluation and Mitigation Strategy, or REMS, to assure the safe use of the product. If the FDA concludes a REMS is needed, the sponsor of the BLA must submit a proposed REMS. The FDA will not approve the BLA without an approved REMS, if required. A REMS could include medication guides, physician communication plans or elements to assure safe use, such as restricted distribution methods, patient registries and other risk minimization tools. Any of these limitations on approval or marketing could restrict the commercial

promotion, distribution, prescription or dispensing of products. Product approvals may be withdrawn for non-compliance with regulatory standards or if problems occur following initial marketing.

FDA regulations require that products be manufactured in specific approved facilities and in accordance with cGMP regulations. We rely, and expect to continue to rely, on third parties for the production of clinical and commercial quantities of our products in accordance with cGMP regulations. These manufacturers must comply with cGMP regulations that require, among other things, quality control and quality assurance, the maintenance of records and documentation and the obligation to investigate and correct any deviations from cGMP. Manufacturers and other entities involved in the manufacture and distribution of approved drugs or biologics are required to register their establishments with the FDA and certain state agencies, and are subject to periodic unannounced inspections by the FDA and certain state agencies for compliance with cGMP requirements and other laws. Accordingly, manufacturers must continue to expend time, money and effort in the area of production and quality control to maintain cGMP compliance. The discovery of violative conditions, including failure to conform to cGMP regulations, could result in enforcement actions, and the discovery of problems with a product after approval may result in restrictions on a product, manufacturer or holder of an approved BLA, including recall.

Other Regulatory Matters

Manufacturing, sales, promotion and other activities following product approval are also subject to regulation by numerous regulatory authorities in the United States in addition to the FDA, including the Centers for Medicare & Medicaid Services, or CMS, other divisions of the Department of Health and Human Services, or DHHS, the Department of Justice, the Drug Enforcement Administration, the Consumer Product Safety Commission, the Federal Trade Commission, the Occupational Safety & Health Administration, the Environmental Protection Agency and state and local governments.

Other Healthcare Laws

Healthcare providers, physicians, and third-party payors will play a primary role in the recommendation and prescription of any products for which we obtain marketing approval. Our future arrangements with third-party payors, healthcare providers and physicians may expose us to broadly applicable fraud and abuse and other healthcare laws and regulations that may constrain the business or financial arrangements and relationships through which we market, sell and distribute any drugs for which we obtain marketing approval. In particular, the research of our product candidates, as well as the promotion, sales and marketing of healthcare items and services, as well as certain business arrangements in the healthcare industry, are subject to extensive laws designed to prevent fraud, kickbacks, self-dealing and other abusive practices. These laws and regulations may restrict or prohibit a wide range of pricing, discounting, marketing and promotion, structuring and commission(s), certain customer incentive programs and other business arrangements generally. Activities subject to these laws also involve the improper use of information obtained in the course of patient recruitment for clinical trials. In the United States, these laws include, without limitation, state and federal anti-kickback, false claims, physician transparency, and patient data privacy and security laws and regulations, including but not limited to those described below.

- § The Anti Kickback Statute, or AKS, which makes it illegal for among other things, any person or entity, including a prescription drug manufacturer (or a party acting on its behalf), to knowingly and willfully solicit, receive, offer or pay any remuneration, directly or indirectly, overtly or covertly, in cash or in kind, that is intended to induce or reward referrals, including the purchase, recommendation, order or prescription of a particular drug, for which payment may be made under a federal healthcare program, such as Medicare or Medicaid. Violations of this law are punishable by up to five years in prison, criminal fines, administrative civil money penalties and exclusion from participation in federal healthcare programs. In addition, a person or entity does not need to have actual knowledge of the statute or specific intent to violate it to have committed a violation.

- § The federal civil and criminal false claims laws, including the False Claims Act, which prohibits individuals or entities (including prescription drug manufacturers) from knowingly presenting, or causing to be presented false or fraudulent claims for payment by a federal healthcare program or making a false statement or record material to payment of a false claim or avoiding, decreasing or concealing an obligation to pay money to the federal government. The government may deem manufacturers to have "caused" the submission of false or fraudulent claims by, for example, providing inaccurate billing or coding information to customers or promoting a product off label. Claims which include items or services resulting from a violation of the federal Anti Kickback Statute are false or fraudulent claims for purposes of the False Claims Act. Our future marketing and activities relating to the reporting of wholesaler or estimated retail prices for our products, the reporting of prices used to calculate Medicaid rebate information and other information affecting federal, state and third-party reimbursement for our products, and the sale and marketing of our product and any future product candidates, are subject to scrutiny under these laws.
- § The Health Insurance Portability and Accountability Act of 1996, or HIPAA, which created additional federal criminal statutes that prohibit among other things, knowingly and willfully executing a scheme, or attempting to execute a scheme, to defraud any healthcare benefit program, including private payors, or falsifying, concealing or covering up a material fact or making any materially false statements in connection with the delivery of or payment for healthcare benefits, items or services.
- § HIPAA, as amended by the Health Information Technology for Economic and Clinical Health Act of 2009, or HITECH, and their respective implementing regulations, which impose, among other things, specified requirements on covered entities, which include certain healthcare providers, health plans and healthcare clearinghouses, and their business associates, which include individuals or entities that perform services for covered entities involving the creation, use, maintenance or disclosure of, individually identifiable health information, relating to the privacy and security of individually identifiable health information including mandatory contractual terms and required implementation of technical safeguards of such information. HITECH also created new tiers of civil monetary penalties, amended HIPAA to make civil and criminal penalties directly applicable to business associates, and gave state attorneys general new authority to file civil actions for damages or injunctions in federal courts to enforce the federal HIPAA laws and seek attorneys' fees and costs associated with pursuing federal civil actions.
- § The Physician Payments Sunshine Act, enacted as part of the Patient Protection and Affordable Care Act, as amended by the Health Care and Education Reconciliation Act of 2010, or collectively, the ACA, which impose new annual reporting requirements for certain manufacturers of drugs, devices, biologics, and medical supplies for which payment is available under Medicare, Medicaid, or the Children's Health Insurance Program, for certain payments and "transfers of value" provided to physicians and teaching hospitals, as well as ownership and investment interests held by physicians and their immediate family members.
- § Analogous state and foreign fraud and abuse laws and regulations, such as state anti kickback and false claims laws, which may be broader in scope and apply regardless of payor. Such laws are enforced by various state agencies and through private actions. Some state laws require pharmaceutical companies to comply with the pharmaceutical industry's voluntary compliance guidelines and the relevant federal government compliance guidance, require drug manufacturers to report information related to payments and other transfers of value to physicians and other healthcare providers, and restrict marketing practices or require disclosure of marketing expenditures. Some state and local laws require the registration of pharmaceutical sales representatives. State and foreign laws also govern the privacy and security of health information in some circumstances. Such data privacy and security laws may differ from each other in significant ways and often are not preempted by HIPAA, thus complicating compliance efforts.

The scope and enforcement of each of these laws is uncertain and subject to rapid change in the current environment of healthcare reform, especially in light of the lack of applicable precedent and regulations. Federal and state enforcement bodies have recently increased their scrutiny of interactions between healthcare companies and healthcare providers, which has led to a number of investigations, prosecutions, convictions and settlements in the healthcare industry. It is possible that governmental authorities will conclude that our business practices do not comply with current or future statutes, regulations or case law involving applicable fraud and abuse or other healthcare laws and regulations. If our operations are found to be in violation of any of these laws or any other related governmental regulations that may apply to us, we may be subject to significant civil, criminal and administrative penalties, damages, fines, imprisonment, disgorgement, exclusion of drugs from participation in state and federal healthcare programs, such as Medicare and Medicaid, reputational harm, additional oversight and reporting obligations if we become subject to a corporate integrity agreement or similar settlement to resolve allegations of non compliance with these laws and the curtailment or restructuring of our operations. If any of the physicians or other healthcare providers or entities with whom we expect to do business is found to be not in compliance with applicable laws, they may be subject to similar actions, penalties and sanctions. Ensuring business arrangements comply with applicable healthcare laws, as well as responding to possible investigations by government authorities, can be time and resource consuming and can divert a company's attention from the business.

Current and Future Healthcare Reform Legislation

In the United States and foreign jurisdictions, there have been a number of legislative and regulatory changes and proposed changes regarding the healthcare system that could prevent or delay marketing approval of our product candidates, restrict or regulate post-approval activities and affect our ability to profitably sell any product candidates for which we obtain marketing approval. We expect that current laws, as well as other healthcare reform measures that may be adopted in the future, may result in more rigorous coverage criteria and in additional downward pressure on the price that we, or any collaborators, may receive for any approved products.

The ACA, for example, contains provisions that subject biological products to potential competition by lower cost biosimilars and may reduce the profitability of drug products through increased rebates for drugs reimbursed by Medicaid programs, extension of Medicaid rebates to Medicaid managed care plans, mandatory discounts for certain Medicare Part D beneficiaries and annual fees based on pharmaceutical companies' share of sales to federal health care programs. The Trump Administration and Congress have taken steps to make administrative or legislative changes, including modification, repeal, or replacement of all, or certain provisions of, the ACA, which may impact reimbursement for drugs and biologics. On January 20, 2017, President Trump signed an Executive Order directing federal agencies with authorities and responsibilities under the ACA to waive, defer, grant exemptions from, or delay the implementation of any provision of the ACA that would impose a fiscal or regulatory burden on states, individuals, healthcare providers, health insurers, or manufacturers of pharmaceuticals or medical devices. On October 13, 2017, President Trump signed an Executive Order terminating the cost sharing subsidies that reimburse insurers under the ACA. Several state Attorneys General filed suit to stop the administration from terminating the subsidies, but their request for a restraining order was denied by a federal judge in California on October 25, 2017. In addition, the Centers for Medicare & Medicaid Services, or CMS has recently proposed regulations that would give states greater flexibility in setting benchmarks for insurers in the individual and small group marketplaces, which may have the effect of relaxing the essential health benefits required under the ACA for plans sold through such marketplaces. Further, each chamber of Congress has put forth multiple bills designed to repeal or repeal and replace portions of the ACA. While Congress has not passed repeal legislation, two bills affecting the implementation of certain taxes under the ACA have been signed into law. The Tax Reform Act includes a provision repealing, effective January 1, 2019, the tax based shared responsibility payment imposed by the ACA on certain individuals who fail to maintain qualifying health coverage for all or part of a year that is commonly referred to as the "individual mandate." Additionally, on January 22, 2018, President Trump signed a continuing resolution on appropriations for

fiscal year 2018 that delayed the implementation of certain ACA-mandated fees, including the "Cadillac" tax on certain high cost employer-sponsored insurance plans, the annual fee imposed on certain health insurance providers based on market share, and the medical device excise tax on non-exempt medical devices. Further, the Bipartisan Budget Act of 2018, or the BBA, among other things, amends the ACA, effective January 1, 2019, to increase from 50 percent to 70 percent the point-of-sale discount that is owed by pharmaceutical manufacturers who participate in Medicare Part D and to close the coverage gap in most Medicare drug plans, commonly referred to as the "donut hole." Congress may consider additional legislation to repeal or replace other elements of the ACA. Litigation and legislation over the ACA are likely to continue, with unpredictable and uncertain results.

Additionally, other federal health reform measures have been proposed and adopted in the United States since the ACA was enacted:

- § The Budget Control Act of 2011, among other things, created measures for spending reductions by Congress. A Joint Select Committee on Deficit Reduction, tasked with recommending a targeted deficit reduction of at least \$1.2 trillion for the years 2013 through 2027, was unable to reach required goals, thereby triggering the legislation's automatic reduction to several government programs. These changes included aggregate reductions to Medicare payments to providers of up to 2% per fiscal year, which went into effect in April 2013 and, due to subsequent legislative amendments, including the BBA, will remain in effect through 2025 unless additional Congressional action is taken.
- § The American Taxpayer Relief Act of 2012, among other things, reduced Medicare payments to several providers, and increased the statute of limitations period for the government to recover overpayments to providers from three to five years.
- § The Middle Class Tax Relief and Job Creation Act of 2012 required that the CMS, reduce the Medicare clinical laboratory fee schedule by 2% in 2013, which served as a base for 2014 and subsequent years. In addition, effective January 1, 2014, CMS also began bundling the Medicare payments for certain laboratory tests ordered while a patient received services in a hospital outpatient setting.

Further, there has been heightened governmental scrutiny over the manner in which manufacturers set prices for their marketed products, which have resulted in several recent Congressional inquiries and proposed bills and enacted federal and state legislation designed to, among other things, bring more transparency to product pricing, review the relationship between pricing and manufacturer patient programs, and reform government program reimbursement methodologies for products. In addition, the United States government, state legislatures, and foreign governments have shown significant interest in implementing cost containment programs, including price controls, restrictions on reimbursement and requirements for substitution of generic products for branded prescription drugs to limit the growth of government paid health care costs. For example, the United States government has passed legislation requiring pharmaceutical manufacturers to provide rebates and discounts to certain entities and governmental payors to participate in federal healthcare programs. Additionally, the Trump Administration's budget proposal for fiscal year 2019 contains further drug price control measures that could be enacted during the 2019 budget process or in other future legislation, including, for example, measures to permit Medicare Part D plans to negotiate the price of certain drugs under Medicare Part B, to allow some states to negotiate drug prices under Medicaid, and to eliminate cost sharing for generic drugs for low-income patients. While any proposed measures will require authorization through additional legislation to become effective, Congress and the Trump Administration have each indicated that it will continue to seek new legislative and/or administrative measures to control drug costs. Individual states in the United States have also become increasingly aggressive in passing legislation and implementing regulations designed to control pharmaceutical and biological product pricing, including price or patient reimbursement constraints, discounts, restrictions on certain product access and marketing cost disclosure and transparency measures, and, in some cases, designed to encourage importation from other countries and bulk purchasing.

Packaging and Distribution in the United States

If our products are made available to authorized users of the Federal Supply Schedule of the General Services Administration, additional laws and requirements apply. Products must meet applicable child resistant packaging requirements under the U.S. Poison Prevention Packaging Act. Manufacturing, sales, promotion and other activities also are potentially subject to federal and state consumer protection and unfair competition laws.

The distribution of pharmaceutical products is subject to additional requirements and regulations, including extensive record keeping, licensing, storage and security requirements intended to prevent the unauthorized sale of pharmaceutical products.

The failure to comply with any of these laws or regulatory requirements subjects firms to possible legal or regulatory action. Depending on the circumstances, failure to meet applicable regulatory requirements can result in criminal prosecution, fines or other penalties, injunctions, exclusion from federal healthcare programs, requests for recall, seizure of products, total or partial suspension of production, denial or withdrawal of product approvals, or refusal to allow a firm to enter into supply contracts, including government contracts. Any action against us for violation of these laws, even if we successfully defend against it, could cause us to incur significant legal expenses and divert our management's attention from the operation of our business. Prohibitions or restrictions on sales or withdrawal of future products marketed by us could materially affect our business in an adverse way.

Changes in regulations, statutes or the interpretation of existing regulations could impact our business in the future by requiring, for example: (i) changes to our manufacturing arrangements; (ii) additions or modifications to product labeling; (iii) the recall or discontinuation of our products; or (iv) additional record keeping requirements. If any such changes were to be imposed, they could adversely affect the operation of our business.

Other U.S. Environmental, Health and Safety Laws and Regulations

We may be subject to numerous environmental, health and safety laws and regulations, including those governing laboratory procedures and the handling, use, storage, treatment and disposal of hazardous materials and wastes. From time to time and in the future, our operations may involve the use of hazardous and flammable materials, including chemicals and biological materials, and may also produce hazardous waste products. Even if we contract with third parties for the disposal of these materials and waste products, we cannot completely eliminate the risk of contamination or injury resulting from these materials. In the event of contamination or injury resulting from the use or disposal of our hazardous materials, we could be held liable for any resulting damages, and any liability could exceed our resources. We also could incur significant costs associated with civil or criminal fines and penalties for failure to comply with such laws and regulations.

We maintain workers' compensation insurance to cover us for costs and expenses we may incur due to injuries to our employees, but this insurance may not provide adequate coverage against potential liabilities. However, we do not maintain insurance for environmental liability or toxic tort claims that may be asserted against us.

In addition, we may incur substantial costs in order to comply with current or future environmental, health and safety laws and regulations. Current or future environmental laws and regulations may impair our research, development or production efforts. In addition, failure to comply with these laws and regulations may result in substantial fines, penalties or other sanctions.

U.S. Patent Term Restoration and Marketing Exclusivity

Depending upon the timing, duration and specifics of FDA approval of SRK-015 and any future product candidates, some of our U.S. patents may be eligible for limited patent term extension under the Drug Price Competition and Patent Term Restoration Act of 1984, commonly referred to as the Hatch Waxman Amendments. The Hatch Waxman Amendments permit restoration of the patent term of up to five years as

compensation for patent term lost during product development and FDA regulatory review process. Patent term restoration, however, cannot extend the remaining term of a patent beyond a total of 14 years from the product's approval date. The patent term restoration period is generally one half the time between the effective date of an IND and the submission date of a BLA plus the time between the submission date of a BLA and the approval of that application, except that the review period is reduced by any time during which the applicant failed to exercise due diligence. Only one patent applicable to an approved drug is eligible for the extension and the application for the extension must be submitted prior to the expiration of the patent. The U.S. PTO, in consultation with the FDA, reviews and approves the application for any patent term extension or restoration. In the future, we may apply for restoration of patent term for our currently owned or licensed patents to add patent life beyond its current expiration date, depending on the expected length of the clinical trials and other factors involved in the filing of the relevant BLA.

An abbreviated approval pathway for biological products shown to be similar to, or interchangeable with, an FDA licensed reference biological product was created by the Biologics Price Competition and Innovation Act of 2009, or BPCI Act. This amendment to the PHS Act, in part, attempts to minimize duplicative testing. Biosimilarity, which requires that the biological product be highly similar to the reference product notwithstanding minor differences in clinically inactive components and that there be no clinically meaningful differences between the product and the reference product in terms of safety, purity and potency, can be shown through analytical studies, animal studies and a clinical trial or trials. Interchangeability requires that a biological product be biosimilar to the reference product and that the product can be expected to produce the same clinical results as the reference product in any given patient and, for products administered multiple times to an individual, that the product and the reference product may be alternated or switched after one has been previously administered without increasing safety risks or risks of diminished efficacy relative to exclusive use of the reference biological product without such alternation or switch.

A reference biological product is granted 12 years of data exclusivity from the time of first licensure of the product, and the FDA will not accept an application for a biosimilar or interchangeable product based on the reference biological product until four years after the date of first licensure of the reference product. "First licensure" typically means the initial date the particular product at issue was licensed in the United States. Date of first licensure does not include the date of licensure of (and a new period of exclusivity is not available for) a biological product if the licensure is for a supplement for the biological product or for a subsequent application by the same sponsor or manufacturer of the biological product (or licensor, predecessor in interest, or other related entity) for a change (not including a modification to the structure of the biological product) that results in a new indication, route of administration, dosing schedule, dosage form, delivery system, delivery device or strength, or for a modification to the structure of the biological product that does not result in a change in safety, purity, or potency.

Pediatric exclusivity is another type of regulatory market exclusivity in the United States. Pediatric exclusivity, if granted, adds six months to existing regulatory exclusivity periods. This six month exclusivity may be granted based on the voluntary completion of a pediatric trial in accordance with an FDA issued "Written Request" for such a trial.

European Union Drug Development

In the European Union, or EU, our future products also may be subject to extensive regulatory requirements. As in the United States, medicinal products can be marketed only if a marketing authorization from the competent regulatory agencies has been obtained.

Similar to the United States, the various phases of preclinical and clinical research in the European Union are subject to significant regulatory controls. Although the EU Clinical Trials Directive 2001/20/EC has sought to harmonize the EU clinical trials regulatory framework, setting out common rules for the control and authorization of clinical trials in the EU, the EU Member States have transposed and applied the

provisions of the Directive differently. This has led to significant variations in the member state regimes. Under the current regime, before a clinical trial can be initiated it must be approved in each of the EU countries where the trial is to be conducted by two distinct bodies: the National Competent Authority, or NCA, and one or more Ethics Committees, or ECs. Under the current regime all suspected unexpected serious adverse reactions to the investigated drug that occur during the clinical trial have to be reported to the NCA and ECs of the Member State where they occurred.

The EU clinical trials legislation currently is undergoing a transition process mainly aimed at harmonizing and streamlining clinical trial authorization, simplifying adverse event reporting procedures, improving the supervision of clinical trials and increasing their transparency. Recently enacted Clinical Trials Regulation EU No 536/2014 ensures that the rules for conducting clinical trials in the EU will be identical.

European Union Drug Marketing

Much like the Anti Kickback Statute prohibition in the United States, the provision of benefits or advantages to physicians to induce or encourage the prescription, recommendation, endorsement, purchase, supply, order or use of medicinal products is also prohibited in the EU. The provision of benefits or advantages to physicians is governed by the national anti bribery laws of European Union Member States, such as the U.K. Bribery Act 2010. Infringement of these laws could result in substantial fines and imprisonment.

Payments made to physicians in certain EU Member States must be publicly disclosed. Moreover, agreements with physicians often must be the subject of prior notification and approval by the physician's employer, his or her competent professional organization and/or the regulatory authorities of the individual EU Member States. These requirements are provided in the national laws, industry codes or professional codes of conduct, applicable in the EU Member States. Failure to comply with these requirements could result in reputational risk, public reprimands, administrative penalties, fines or imprisonment.

European Union Drug Review and Approval

In the European Economic Area, or EEA, which is comprised of the 27 Member States of the EU (including Norway and excluding Croatia), Iceland and Liechtenstein, medicinal products can only be commercialized after obtaining a Marketing Authorization, or MA. There are two types of marketing authorizations.

The Community MA is issued by the European Commission through the Centralized Procedure, based on the opinion of the Committee for Medicinal Products for Human Use, or CHMP, of the European Medicines Agency, or EMA, and is valid throughout the entire territory of the EEA. The Centralized Procedure is mandatory for certain types of products, such as biotechnology medicinal products, orphan medicinal products, advanced therapy medicines such as gene therapy, somatic cell therapy or tissue engineered medicines and medicinal products containing a new active substance indicated for the treatment of HIV, AIDS, cancer, neurodegenerative disorders, diabetes, autoimmune and other immune dysfunctions and viral diseases. The Centralized Procedure is optional for products containing a new active substance not yet authorized in the EEA, or for products that constitute a significant therapeutic, scientific or technical innovation or which are in the interest of public health in the EU.

National MAs, which are issued by the competent authorities of the Member States of the EEA and only cover their respective territory, are available for products not falling within the mandatory scope of the Centralized Procedure. Where a product has already been authorized for marketing in a Member State of the EEA, this National MA can be recognized in another Member States through the Mutual Recognition Procedure. If the product has not received a National MA in any Member State at the time of application, it can be approved simultaneously in various Member States through the Decentralized Procedure. Under the Decentralized Procedure an identical dossier is submitted to the competent authorities of each of the Member States in which the MA is sought, one of which is selected by the applicant as the Reference Member State, or RMS. The competent authority of the RMS prepares a draft assessment report, a draft summary of the product characteristics, or SPC, and a draft of the labeling and package leaflet, which are

sent to the other Member States (referred to as the Member States Concerned) for their approval. If the Member States Concerned raise no objections, based on a potential serious risk to public health, to the assessment, SPC, labeling, or packaging proposed by the RMS, the product is subsequently granted a national MA in all the Member States (i.e., in the RMS and the Member States Concerned).

Under the above described procedures, before granting the MA, the EMA or the competent authorities of the Member States of the EEA make an assessment of the risk benefit balance of the product on the basis of scientific criteria concerning its quality, safety and efficacy.

European Union New Chemical Entity Exclusivity

In the EU, new chemical entities, sometimes referred to as new active substances, qualify for eight years of data exclusivity upon marketing authorization and an additional two years of market exclusivity. The data exclusivity, if granted, prevents regulatory authorities in the EU from referencing the innovator's data to assess a generic application for eight years, after which generic marketing authorization can be submitted, and the innovator's data may be referenced, but not approved for two years. The overall 10 year period will be extended to a maximum of 11 years if, during the first eight years of those 10 years, the marketing authorization holder obtains an authorization for one or more new therapeutic indications which, during the scientific evaluation prior to their authorization, are determined to bring a significant clinical benefit in comparison with currently approved therapies.

European Union Orphan Designation and Exclusivity

In the EU, the EMA's Committee for Orphan Medicinal Products grants orphan drug designation to promote the development of products that are intended for the diagnosis, prevention or treatment of life threatening or chronically debilitating conditions affecting not more than five in 10,000 persons in the EU community (or where it is unlikely that the development of the medicine would generate sufficient return to justify the investment) and for which no satisfactory method of diagnosis, prevention or treatment has been authorized (or, if a method exists, the product would be a significant benefit to those affected).

In the EU, orphan drug designation entitles a party to financial incentives such as reduction of fees or fee waivers and 10 years of market exclusivity is granted following medicinal product approval. This period may be reduced to six years if the orphan drug designation criteria are no longer met, including where it is shown that the product is sufficiently profitable not to justify maintenance of market exclusivity. Orphan drug designation must be requested before submitting an application for marketing approval. Orphan drug designation does not convey any advantage in, or shorten the duration of, the regulatory review and approval process.

European Data Collection

The collection and use of personal health data in the European Union is governed by the provisions of the Data Protection Directive, and as of May 2018 the General Data Protection Regulation, or GDPR. This directive imposes several requirements relating to the consent of the individuals to whom the personal data relates, the information provided to the individuals, notification of data processing obligations to the competent national data protection authorities and the security and confidentiality of the personal data. The Data Protection Directive and GDPR also impose strict rules on the transfer of personal data out of the EU to the United States. Failure to comply with the requirements of the Data Protection Directive, the GDPR, and the related national data protection laws of the EU Member States may result in fines and other administrative penalties. The GDPR introduces new data protection requirements in the EU and substantial fines for breaches of the data protection rules. The GDPR regulations may impose additional responsibility and liability in relation to personal data that we process and we may be required to put in place additional mechanisms ensuring compliance with the new data protection rules. This may be onerous and adversely affect our business, financial condition, results of operations and prospects.

Rest of the World Regulation

For other countries outside of the EU and the United States, such as countries in Eastern Europe, Latin America or Asia, the requirements governing the conduct of clinical trials, product licensing, pricing and reimbursement vary from country to country. Additionally, the clinical trials must be conducted in accordance with GCP requirements and the applicable regulatory requirements and the ethical principles that have their origin in the Declaration of Helsinki.

If we fail to comply with applicable foreign regulatory requirements, we may be subject to, among other things, fines, suspension or withdrawal of regulatory approvals, product recalls, seizure of products, operating restrictions and criminal prosecution.

Additional Laws and Regulations Governing International Operations

If we further expand our operations outside of the United States, we must dedicate additional resources to comply with numerous laws and regulations in each jurisdiction in which we plan to operate. The Foreign Corrupt Practices Act, or FCPA, prohibits any U.S. individual or business from paying, offering, authorizing payment or offering of anything of value, directly or indirectly, to any foreign official, political party or candidate for the purpose of influencing any act or decision of the foreign entity in order to assist the individual or business in obtaining or retaining business. The FCPA also obligates companies whose securities are listed in the United States to comply with certain accounting provisions requiring the company to maintain books and records that accurately and fairly reflect all transactions of the corporation, including international subsidiaries, and to devise and maintain an adequate system of internal accounting controls for international operations.

Compliance with the FCPA is expensive and difficult, particularly in countries in which corruption is a recognized problem. In addition, the FCPA presents particular challenges in the pharmaceutical industry, because, in many countries, hospitals are operated by the government, and doctors and other hospital employees are considered foreign officials. Certain payments to hospitals in connection with clinical trials and other work have been deemed to be improper payments to government officials and have led to FCPA enforcement actions.

Various laws, regulations and executive orders also restrict the use and dissemination outside of the United States, or the sharing with certain non U.S. nationals, of information classified for national security purposes, as well as certain products and technical data relating to those products. If we expand our presence outside of the United States, it will require us to dedicate additional resources to comply with these laws, and these laws may preclude us from developing, manufacturing, or selling certain products and product candidates outside of the United States, which could limit our growth potential and increase our development costs.

The failure to comply with laws governing international business practices may result in substantial civil and criminal penalties and suspension or debarment from government contracting. The U.S. Securities and Exchange Commission, or SEC, also may suspend or bar issuers from trading securities on U.S. exchanges for violations of the FCPA's accounting provisions.

Reimbursement

Sales of our products will depend, in part, on the extent to which our products, if approved, will be covered by third-party payors, such as government health programs, commercial insurers and managed healthcare organizations, as well as the level of reimbursement such third-party payors provide for our products. Patients and providers are unlikely to use our products unless coverage is provided and reimbursement is adequate to cover a significant portion of the cost of our products. In the United States no uniform policy of coverage and reimbursement for drugs or biological products exists, and one payor's determination to provide coverage and adequate reimbursement for a product does not assure that other payors will make a

similar determination. Accordingly, decisions regarding the extent of coverage and amount of reimbursement to be provided for any of our products candidates, if approved, will be made on a payor by payor basis. As a result, the coverage determination process may be a time consuming and costly process that will require us to provide scientific and clinical support for the use of our products to each payor separately, with no assurance that coverage and adequate reimbursement will be obtained.

The Medicaid Drug Rebate Program requires pharmaceutical manufacturers to enter into and have in effect a national rebate agreement with the Secretary of the DHHS as a condition for states to receive federal matching funds for the manufacturer's outpatient drugs furnished to Medicaid patients. The ACA made several changes to the Medicaid Drug Rebate Program, including increasing pharmaceutical manufacturers' rebate liability by raising the minimum basic Medicaid rebate on most branded prescription drugs from 15.1% of average manufacturer price, or AMP, to 23.1% of AMP and adding a new rebate calculation for "line extensions" (i.e., new formulations, such as extended release formulations) of solid oral dosage forms of branded products, creating a new method by which rebates owed by are calculated for drugs that are inhaled, infused, instilled, implanted or injected, as well as potentially impacting their rebate liability by modifying the statutory definition of AMP. The ACA also expanded the universe of Medicaid utilization subject to drug rebates by requiring pharmaceutical manufacturers to pay rebates on Medicaid managed care utilization and by enlarging the population potentially eligible for Medicaid drug benefits. Pricing and rebate programs must also comply with the Medicaid rebate requirements of the U.S. Omnibus Budget Reconciliation Act of 1990.

The Medicare Prescription Drug, Improvement, and Modernization Act of 2003, or the MMA, established the Medicare Part D program to provide a voluntary prescription drug benefit to Medicare beneficiaries. Under Part D, Medicare beneficiaries may enroll in prescription drug plans offered by private entities that provide coverage of outpatient prescription drugs. Unlike Medicare Part A and B, Part D coverage is not standardized. While all Medicare drug plans must give at least a standard level of coverage set by Medicare, Part D prescription drug plan sponsors are not required to pay for all covered Part D drugs, and each drug plan can develop its own drug formulary that identifies which drugs it will cover and at what tier or level. However, Part D prescription drug formularies must include drugs within each therapeutic category and class of covered Part D drugs, though not necessarily all the drugs in each category or class. Any formulary used by a Part D prescription drug plan must be developed and reviewed by a pharmacy and therapeutic committee. Government payment for some of the costs of prescription drugs may increase demand for products for which we receive marketing approval. However, any negotiated prices for our products covered by a Part D prescription drug plan likely will be lower than the prices we might otherwise obtain. Moreover, while the MMA applies only to drug benefits for Medicare beneficiaries, private payors often follow Medicare coverage policy and payment limitations in setting their own payment rates. Any reduction in payment that results from the MMA may result in a similar reduction in payments from non governmental payors.

For a drug product to receive federal reimbursement under the Medicaid or Medicare Part B programs or to be sold directly to U.S. government agencies, the manufacturer must extend discounts to entities eligible to participate in the 340B drug pricing program. The required 340B discount on a given product is calculated based on the AMP and Medicaid rebate amounts reported by the manufacturer. As of 2010, the ACA expanded the types of entities eligible to receive discounted 340B pricing, although, under the current state of the law, with the exception of children's hospitals, these newly eligible entities will not be eligible to receive discounted 340B pricing on orphan drugs. In addition, as 340B drug pricing is determined based on AMP and Medicaid rebate data, the revisions to the Medicaid rebate formula and AMP definition described above could cause the required 340B discount to increase.

As noted above, the marketability of any products for which we receive regulatory approval for commercial sale may suffer if the government and third-party payors fail to provide coverage and adequate reimbursement. An increasing emphasis on cost containment measures in the United States has increased and we expect will continue to increase the pressure on pharmaceutical pricing. Coverage policies and

third-party reimbursement rates may change at any time. Even if favorable coverage and reimbursement status is attained for one or more products for which we receive regulatory approval, less favorable coverage policies and reimbursement rates may be implemented in the future.

These laws, and future state and federal healthcare reform measures may be adopted in the future, any of which may result in additional reductions in Medicare and other healthcare funding and otherwise affect the prices we may obtain for any of our product candidates for which we may obtain regulatory approval or the frequency with which any such product candidate is prescribed or used.

In addition, in most foreign countries, the proposed pricing for a drug must be approved before it may be lawfully marketed. The requirements governing drug pricing and reimbursement vary widely from country to country. For example, the EU provides options for its member states to restrict the range of medicinal products for which their national health insurance systems provide reimbursement and to control the prices of medicinal products for human use. Reference pricing used by various EU Member States and parallel distribution, or arbitrage between low priced and high priced member states, can further reduce prices. A member state may approve a specific price for the medicinal product or it may instead adopt a system of direct or indirect controls on the profitability of the company placing the medicinal product on the market. In some countries, we may be required to conduct a clinical study or other studies that compare the cost effectiveness of any of our product candidates to other available therapies in order to obtain or maintain reimbursement or pricing approval. There can be no assurance that any country that has price controls or reimbursement limitations for pharmaceutical products will allow favorable reimbursement and pricing arrangements for any of our products. Historically, products launched in the EU do not follow price structures of the United States and generally prices tend to be significantly lower. Publication of discounts by third-party payors or authorities may lead to further pressure on the prices or reimbursement levels within the country of publication and other countries.

Employees

As of May 7, 2018, we had 50 full-time employees, including 22 employees with M.D. or Ph.D. degrees. Of these full-time employees, 40 employees are engaged in research and development activities and ten are engaged in general and administrative activities. None of our employees is represented by a labor union or covered by a collective bargaining agreement.

Facilities

Our facility comprises 21,000 square feet (including over 9,000 square feet of expansion space which we have not yet occupied) of office and laboratory space in Cambridge, Massachusetts. We executed a lease amendment on February 22, 2018 for the additional expansion space and expect to occupy the expansion space in the third quarter of 2018. The lease expires five years after our landlord delivers the expansion space to us. We have an option to extend the lease term for five additional years. We believe that our existing facilities, including our expansion space, are adequate to meet our current needs, and that suitable additional space will be available as and when needed.

Legal Proceedings

From time to time, we may be involved in various claims and legal proceedings relating to claims arising out of our operations. We are not currently a party to any material legal proceedings.

MANAGEMENT

The following table sets forth the name and position of each of our executive officers and directors and each of their ages as of May 7, 2018:

Name	Age	Position
Executive Officers		
Nagesh K. Mahanthappa, Ph.D.	53	President, Chief Executive Officer and Director
Alan J. Buckler, Ph.D.	58	Chief Scientific Officer
Rhonda M. Chicko, C.P.A.	52	Chief Financial Officer
Yung H. Chyung, M.D.	42	Chief Medical Officer
Elan Z. Ezickson	54	Chief Operating Officer & Head of Corporate Development
Non-Employee Directors		
David Hallal ⁽²⁾⁽³⁾	51	Chairman of the Board of Directors
Kristina Burow ⁽¹⁾⁽²⁾	44	Director
Jeffrey S. Flier, M.D. ⁽³⁾	70	Director
Michael Gilman, Ph.D. ⁽¹⁾	63	Director
Amir Nashat, Sc.D. ⁽¹⁾⁽²⁾	45	Director
Timothy A. Springer, Ph.D. ⁽³⁾	70	Director

⁽¹⁾ Member of the audit committee

⁽²⁾ Member of the compensation committee

⁽³⁾ Member of the nominating and corporate governance committee

Executive Officers

Nagesh K. Mahanthappa, Ph.D. is the founding employee of Scholar Rock and has served as a director and our President and Chief Executive Officer since October 2012. Prior to joining us, from February 2007 to May 2012, Dr. Mahanthappa was a founding employee and Vice President, Corporate Development & Operations at Avila Therapeutics, Inc. (acquired by Celgene Corporation in March 2012). Previously, from August 2002 to February 2007, he served in roles of increasing responsibility at Alnylam Pharmaceuticals, Inc., most recently as Vice President, Scientific & Strategic Development. He was also a founder of TwistDx, Inc. a DNA diagnostics company acquired by Inverness Medical Innovations, Inc. (now Alere, Inc.) in 2010. Dr. Mahanthappa received his Ph.D. in Neurobiology from the California Institute of Technology, and completed his post-doctoral training at the E.K. Shriver Center for Mental Retardation (then affiliated with Massachusetts General Hospital) and Harvard Medical School. He received his M.B.A. from the F.W. Olin Graduate School of Management at Babson College and his B.A. in Biology and Chemistry from the University of Colorado, Boulder. Our board of directors believes that Dr. Mahanthappa's extensive experience in the pharmaceutical industry qualifies him to serve on our board of directors.

Alan J. Buckler, Ph.D. has served as our Chief Scientific Officer since November 2016. Prior to joining us, Dr. Buckler served as Vice President, Cell and Protein Sciences, at Biogen Inc. from 2014 to 2016. From 2005 to 2014, Dr. Buckler served as Director, Developmental and Molecular Pathways in the Novartis Institutes for Biomedical Research. Prior to Novartis, Dr. Buckler served as the Chief Scientific Officer of Ardaís Corporation from 1999 to 2004 and as Vice President of Molecular Genetics at Sequana Therapeutics/Axys Pharmaceuticals from 1996 to 1999. Prior to joining the private sector, Dr. Buckler served on the Neurology faculty of Massachusetts General Hospital and Harvard Medical School from 1991 to 1996. Dr. Buckler received his A.B. in Biology from the University of Chicago, Ph.D. in Microbiology from the Boston University School of Medicine, and completed his post-doctoral training at the Center for Cancer Research, Massachusetts Institute of Technology.

Rhonda M. Chicko, C.P.A. has served as our Chief Financial Officer since April 2018. Prior to joining us, she served as Vice President, Finance at Editas Medicine, Inc. where she worked from September 2015 to March 2018. From 2005 to 2015, Ms. Chicko worked at Ironwood Pharmaceuticals, Inc. in financial roles of increasing responsibility, culminating as Senior Director, Finance and Tax. Earlier in her career, Ms. Chicko held a range of positions at investment management and accounting firms, including Wellington Management Company, LLP and PricewaterhouseCoopers, LLP. Ms. Chicko holds a B.S. in accounting from Le Moyne College and an M.S.T. from Bentley University.

Yung H. Chyung, M.D. has served as our Chief Medical Officer since February 2016. Prior to joining us, Dr. Chyung served in roles of increasing responsibility at Dyax Corp. (acquired by Shire Plc in January 2016) from 2011 to February 2016, most recently as Vice President of Medical Research, where he was responsible for clinical research and medical affairs. From 2010 to 2011, Dr. Chyung worked at Genzyme Corporation where he was responsible for medical affairs efforts globally for multiple rare disease programs. Dr. Chyung earned his M.D. from Harvard Medical School and completed his internal medicine residency and allergy and immunology fellowship at Massachusetts General Hospital. Dr. Chyung also holds an A.B. in Biochemical Sciences from Harvard College.

Elan Z. Ezickson has served as our Chief Operating Officer & Head of Corporate Development since August 2014. Prior to joining us, Mr. Ezickson served most recently as Executive Vice President and Chief Operating Officer of Aveo Pharmaceuticals, Inc., where he worked from 2003 to July 2013. From 1994 to 2003, he worked at Biogen Inc. in roles that included President of Biogen Canada, Program Executive and Associate General Counsel. Mr. Ezickson holds a B.A. in Political Science from Yale University and a J.D. from the Columbia University School of Law.

Non-Employee Directors

David Hallal has served as the Chairman of our board of directors since July 2017. Most recently, from June 2006 to December 2016, Mr. Hallal served in executive roles of increasing responsibility at Alexion Pharmaceuticals, Inc., most recently serving as Chief Executive Officer and a board member. Prior to his role as CEO, Mr. Hallal served Alexion as COO and Director as well as Chief Commercial Officer and Head of Commercial Operations. Prior to Alexion from 2004 to 2006, Mr. Hallal served as Vice President of Sales for OSI Eyetech, Inc. From 2002 to 2004, Mr. Hallal served as Head of Sales at Biogen Inc. From 1992 to 2002, Mr. Hallal held various leadership roles at Amgen Inc. From 1988 to 1992, Mr. Hallal began his pharmaceutical career at The Upjohn Company as a sales representative. Mr. Hallal holds a B.A. in psychology from the University of New Hampshire. Mr. Hallal also currently serves as an independent director at Seer Biosciences, Inc. Our board of directors believes that Mr. Hallal's experience as an executive at numerous pharmaceutical companies qualifies him to serve as our Chairman of the board of directors.

Kristina Burow has served as a member of our board of directors since August 2014. Ms. Burow has served as Managing Director of ARCH Venture Partners, or ARCH, since November 2011 and previously held roles of increasing responsibility at ARCH from August 2002 to November 2011. Ms. Burow currently serves on the board of directors of several biopharmaceutical and biotechnology companies, including Vividion Therapeutics, Inc., Lycera Corp., BlackThorn Therapeutics, Inc., Metacrine, Inc., Unity Biotechnology, Inc., AgBiome Inc., Vir Biotechnology Inc., and AgTech Accelerator, an agricultural technology startup accelerator. Ms. Burow also serves on the board of directors of Sienna. She previously was a co-founder and member of the board of directors of Receptos, Inc., a public pharmaceutical company, until its acquisition by Celgene Corporation, a public biopharmaceutical company, and of Sapphire Energy, Inc., energy company. Ms. Burow has participated in a number of other ARCH portfolio companies including KYTHERA, Siluria Technologies, Inc., an energy company, and Ikaria, Inc., a biotechnology company, acquired by Madison Dearborn Partners, a private equity firm. Prior to joining ARCH, Ms. Burow was an Associate with the Novartis BioVenture Fund in San Diego and an early employee at the Genomics Institute of the Novartis Research Foundation. Ms. Burow received a B.A. in Chemistry from the University of California, Berkeley, an M.A. in Chemistry from Columbia University, and an M.B.A. from the University of Chicago. We believe that

Ms. Burow is qualified to serve on our board of directors due to her extensive experience investing in biopharmaceutical and biotechnology companies and her experience on boards of directors in the medical industry.

Jeffrey S. Flier, M.D. has served as member of our board of directors since October 2016. Since August 2016, Dr. Flier has served as the Higginson Professor of Physiology and Medicine and Harvard University Distinguished Service Professor, and from 2007 to August 2016 served as the twenty-first Dean of the Faculty of Medicine at Harvard University. Previously, from 2002 to 2007, Dr. Flier served as Chief Academic Officer of Beth Israel Deaconess Medical Center and served as Harvard Medical School Faculty Dean for Academic Programs. An elected member of the National Academy of Medicine and a fellow of the American Academy of Arts and Sciences, his many honors include the Eli Lilly Award of the American Diabetes Association, and the Berson Lecture of the American Physiological Society. He was the recipient of the 2005 Banting Medal from the American Diabetes Association, its highest scientific honor. Dr. Flier received his B.S. from City College of New York and his M.D. from Mount Sinai School of Medicine with highest academic honors, and he completed his residency training at Mount Sinai School of Medicine. Our board of directors believes that Dr. Flier's extensive medical and scientific experience and his leadership skills qualify him to serve on our board of directors.

Michael Gilman, Ph.D. has served as a member of our board of directors since November 2013. Dr. Gilman is currently Chairman and Chief Executive Officer for Arrakis Therapeutics, Inc., a role he has served in since 2016, and Chief Executive Officer and Director for Obsidian Therapeutics, Inc., a role he has served in since 2016. Previously, from 2014 to 2016 Dr. Gilman was Founder and Chief Executive Officer of Padlock Therapeutics, Inc. Prior to Padlock, Dr. Gilman served as Senior Vice President, Early-Stage Pipeline, at Biogen Idec Inc. from 2012 to 2013. He joined Biogen Idec Inc. in 2012 following its acquisition of Stromedix, Inc., where he was Founder and Chief Executive Officer. Prior to founding Stromedix in 2006, from 1999 to 2005, Dr. Gilman served in a variety of capacities, most recently as Executive Vice President, Research at Biogen Idec. From 1994 to 1999, Dr. Gilman was at ARIAD Pharmaceuticals, Inc., where he was Executive Vice President and Chief Scientific Officer. From 1986 to 1994, Dr. Gilman was on the scientific staff of Cold Spring Harbor Laboratory in New York. He also serves on the Board of Directors of X4 Pharmaceuticals, Inc. and the Scientific Advisory Board of FutuRx, an Israeli biotech accelerator. Dr. Gilman was a postdoctoral fellow with Dr. Robert Weinberg at the Whitehead Institute. He holds a Ph.D. in Biochemistry from University of California, Berkeley, and an S.B. in Life Sciences from Massachusetts Institute of Technology. Our board of directors believes that Dr. Gilman's extensive experience in the pharmaceuticals industry qualifies him to serve on our board of directors.

Amir Nashat, Sc.D. has served as a member of our board of directors since October 2012. Dr. Nashat is a managing partner at Polaris Partners, a venture capital firm, where he has worked since 2002. Dr. Nashat was also the founding Chief Executive Officer of Living Proof, Inc. and Sun Catalytix Corporation. Dr. Nashat currently represents Polaris as a Director of Agbiome, Inc., aTyr Pharmaceuticals, Inc., Fate Therapeutics, Inc., Jnana Therapeutics, where he also serves as the CEO, CAMP4, Metacine, Inc., Morphic Therapeutic, Inc., Olivo Labs, Promedior, Inc., Selecta Biosciences Inc., Syros Pharmaceuticals, Inc., and Taris Biomedical, LLC. Dr. Nashat also serves on the Partners Innovation Fund, the Investment Advisory Committee for The Engine at MIT, and helped launch the MIT Sandbox Innovation Fund as its active president. Dr. Nashat previously served on the Board of the New England Venture Capital Association. Dr. Nashat received an M.S. and B.S. in materials science and mechanical engineering from the University of California, Berkeley and a Sc.D. as a Hertz Fellow in Chemical Engineering at the Massachusetts Institute of Technology with a minor in Biology under Dr. Robert Langer. Our board of directors believes that Dr. Nashat's biotechnology investment experience qualifies him to serve on our board of directors.

Timothy A. Springer, Ph.D. is a co-founder and investor in Scholar Rock and has served as a member of our board of directors since October 2012. Since 1989, Dr. Springer has served as the Latham Family Professor of Pathology at Harvard Medical School. He has also served as Senior Investigator in the Program in Cellular and Molecular Medicine at Boston Children's Hospital since 2012 and as Professor of Biological Chemistry

and Molecular Pharmacology at Harvard Medical School and Professor of Medicine at Boston Children's Hospital since 2011. Dr. Springer was the Founder and Chairman of the Scientific Advisory Board of LeukoSite, Inc., a biotechnology company acquired by Millennium Pharmaceuticals, Inc. in 1999. He is a founder, investor and board member of Morpnic Therapeutic, Inc. and an investor and board member of Selecta Biosciences Inc. Dr. Springer is the Chairman of the Institute for Protein Innovation and is a member of the National Academy of Sciences. His honors include the Crafoord Prize, the American Association of Immunologists Meritorious Career Award, the Stratton Medal from the American Society of Hematology, and the Basic Research Prize from the American Heart Association. Dr. Springer received a B.A. from the University of California, Berkeley, and a Ph.D. from Harvard University. Our board of directors believes that Dr. Springer's extensive knowledge of our business and the biotechnology field qualifies him to serve on our board of directors.

Composition of Our Board of Directors

As of May 7, 2018, our board of directors consisted of seven members, each of whom are members pursuant to the board composition provisions of our certificate of incorporation and agreements with our stockholders. These board composition provisions will terminate upon the completion of this offering. Upon the termination of these provisions, there will be no further contractual obligations regarding the election of our directors. Our nominating and corporate governance committee and our board of directors may therefore consider a broad range of factors relating to the qualifications and background of nominees. Our nominating and corporate governance committee's and our board of directors' priority in selecting board members is the identification of persons who will further the interests of our stockholders through their established record of professional accomplishment, the ability to contribute positively to the collaborative culture among board members, knowledge of our business, understanding of the competitive landscape, and professional and personal experiences and expertise relevant to our growth strategy. Our directors hold office until their successors have been elected and qualified or until the earlier of their resignation or removal. Our amended and restated certificate of incorporation and amended and restated by-laws that will become effective upon the completion of this offering also provide that our directors may be removed only for cause by the affirmative vote of the holders of at least two-thirds of the votes that all our stockholders would be entitled to cast in an annual election of directors, and that any vacancy on our board of directors, including a vacancy resulting from an enlargement of our board of directors, may be filled only by vote of a majority of our directors then in office.

Director Independence

Our board of directors has determined that all members of the board of directors, except Dr. Mahanthappa, are independent directors, including for purposes of the rules of The Nasdaq Global Market and the Securities and Exchange Commission, or SEC. In making such independence determination, our board of directors considered the relationships that each non-employee director has with us and all other facts and circumstances that our board of directors deemed relevant in determining their independence, including the beneficial ownership of our capital stock by each non-employee director. In considering the independence of the directors listed above, our board of directors considered the association of our directors with the holders of more than 5% of our common stock. Upon the completion of this offering, we expect that the composition and functioning of our board of directors and each of our committees will comply with all applicable requirements of The Nasdaq Global Market and the rules and regulations of the SEC. There are no family relationships among any of our directors or executive officers. Dr. Mahanthappa is not an independent director under these rules because he is an executive officer of our company.

Staggered Board

In accordance with the terms of our amended and restated certificate of incorporation and amended and restated by-laws that will become effective upon the completion of this offering, our board of directors will be divided into three staggered classes of directors and each director will be assigned to one of the three classes. At each annual meeting of the stockholders, a class of directors will be elected for a three-year term to succeed the directors of the same class whose terms are then expiring. The terms of the directors will expire upon the election and qualification of successor directors at the annual meeting of stockholders

to be held during the years 2019 for Class I directors, 2020 for Class II directors and 2021 for Class III directors.

- § Our Class I directors will be Dr. Springer and Dr. Mahanthappa;
- § Our Class II directors will be Ms. Burow, Mr. Hallal and Dr. Gilman; and
- § Our Class III directors will be Dr. Flier and Dr. Nashat.

Our amended and restated certificate of incorporation and amended and restated by-laws that will become effective upon the completion of this offering will provide that the number of directors shall be fixed from time to time by a resolution of the majority of our board of directors.

The division of our board of directors into three classes with staggered three-year terms may delay or prevent stockholder efforts to effect a change of our management or a change in control.

Board Leadership Structure and Board's Role in Risk Oversight

Mr. Hallal is the current chairman of our board of directors and Dr. Mahanthappa is our current Chief Executive Officer, hence the roles of chairman of our board of directors and Chief Executive Officer are separated. We believe that separating these positions allows our Chief Executive Officer to focus on our day-to-day business, while allowing our chairman of the board to lead the board of directors in its fundamental role of providing advice to and independent oversight of management. Our board of directors recognizes the time, effort and energy that the Chief Executive Officer is required to devote to his position in the current business environment, as well as the commitment required to serve as our chairman, particularly as the board of directors' oversight responsibilities continue to grow. While our amended and restated by-laws and corporate governance guidelines do not require that our chairman and Chief Executive Officer positions be separate, our board of directors believes that having separate positions is the appropriate leadership structure for us at this time and demonstrates our commitment to good corporate governance.

Risk is inherent with every business, and how well a business manages risk can ultimately determine its success. We face a number of risks, including risks relating to our financial condition, development and commercialization activities, operations, strategic direction and intellectual property as more fully discussed in the section entitled "Risk Factors" appearing elsewhere in this prospectus. Management is responsible for the day-to-day management of risks we face, while our board of directors, as a whole and through its committees, has responsibility for the oversight of risk management. In its risk oversight role, our board of directors has the responsibility to satisfy itself that the risk management processes designed and implemented by management are adequate and functioning as designed.

The role of the board of directors in overseeing the management of our risks is conducted primarily through committees of the board of directors, as disclosed in the descriptions of each of the committees below and in the charters of each of the committees. The full board of directors (or the appropriate board committee in the case of risks that are under the purview of a particular committee) discusses with management our major risk exposures, their potential impact on us, and the steps we take to manage them. When a board committee is responsible for evaluating and overseeing the management of a particular risk or risks, the chairman of the relevant committee reports on the discussion to the full board of directors during the committee reports portion of the next board meeting. This enables the board of directors and its committees to coordinate the risk oversight role, particularly with respect to risk interrelationships.

Committees of Our Board of Directors

Our board of directors has established an audit committee, a compensation committee, a nominating and corporate governance committee and a science, innovation and technology committee, each of which will operate pursuant to a charter adopted by our board of directors and will be effective upon the effectiveness of the registration statement of which this prospectus is a part. Upon the effectiveness of the registration statement of which this prospectus is a part, the composition and functioning of all of our committees will

comply with all applicable requirements of the Sarbanes-Oxley Act of 2002, Nasdaq and SEC rules and regulations.

Audit Committee

Effective upon effectiveness of the registration statement of which this prospectus forms a part, Dr. Gilman, Ms. Burow and Dr. Nashat will serve on the audit committee, which will be chaired by Dr. Gilman. Our board of directors has determined that each member of the audit committee is "independent" for audit committee purposes as that term is defined in the rules of the SEC and the applicable Nasdaq rules, and each has sufficient knowledge in financial and auditing matters to serve on the audit committee. Our board of directors has designated Ms. Burow as an "audit committee financial expert," as defined under the applicable rules of the SEC. The audit committee's responsibilities include:

- § appointing, approving the compensation of, and assessing the independence of our independent registered public accounting firm;
- § pre-approving auditing and permissible non-audit services, and the terms of such services, to be provided by our independent registered public accounting firm;
- § reviewing the overall audit plan with our independent registered public accounting firm and members of management responsible for preparing our financial statements;
- § reviewing and discussing with management and our independent registered public accounting firm our annual and quarterly financial statements and related disclosures as well as critical accounting policies and practices used by us;
- § coordinating the oversight and reviewing the adequacy of our internal control over financial reporting;
- § establishing policies and procedures for the receipt and retention of accounting-related complaints and concerns;
- § recommending based upon the audit committee's review and discussions with management and our independent registered public accounting firm whether our audited financial statements shall be included in our Annual Report on Form 10-K;
- § monitoring the integrity of our financial statements and our compliance with legal and regulatory requirements as they relate to our financial statements and accounting matters;
- § preparing the audit committee report required by SEC rules to be included in our annual proxy statement;
- § reviewing all related person transactions for potential conflict of interest situations and approving all such transactions; and
- § reviewing quarterly earnings releases.

Compensation Committee

Effective upon effectiveness of the registration statement of which this prospectus forms a part, Ms. Burow, Dr. Nashat and Mr. Hallal will serve on the compensation committee, which will be chaired by Ms. Burow. Our board of directors has determined that each member of the compensation committee is "independent" as defined in the applicable Nasdaq rules. The compensation committee's responsibilities include:

- § annually reviewing and recommending to our board of directors for approval, the corporate goals and objectives relevant to the compensation of our Chief Executive Officer;
- § evaluating the performance of our Chief Executive Officer in light of such corporate goals and objectives and based on such evaluation: (i) determining the cash compensation of our Chief Executive Officer and (ii) reviewing and approving grants and awards to our Chief Executive Officer under equity-based plans;
- § reviewing and approving the cash compensation of, and any grants and awards under equity-based plans for, our other executive officers, senior vice presidents and vice presidents;
- § reviewing and establishing our overall management compensation, philosophy and policy;
- § overseeing and administering our compensation and similar plans;

- § evaluating and assessing potential and current compensation advisors in accordance with the independence standards identified in the applicable Nasdaq rules;
- § reviewing and approving our policies and procedures for the grant of equity-based awards;
- § reviewing and recommending to the board of directors the compensation of our directors;
- § preparing the compensation committee report required by SEC rules, if and when required, to be included in our annual proxy statement; and
- § reviewing and approving the retention, termination or compensation of any consulting firm or outside advisor to assist in the evaluation of compensation matters.

Nominating and Corporate Governance Committee

Effective upon effectiveness of the registration statement of which this prospectus forms a part, Mr. Hallal, Dr. Flier and Dr. Springer will serve on the nominating and corporate governance committee, which will be chaired by Mr. Hallal. Our board of directors has determined that each member of the nominating and corporate governance committee is "independent" as defined in the applicable Nasdaq rules. The nominating and corporate governance committee's responsibilities include:

- § developing and recommending to the board of directors criteria for board and committee membership;
- § establishing procedures for identifying and evaluating board of director candidates, including nominees recommended by stockholders;
- § reviewing the composition of the board of directors to ensure that it is composed of members containing the appropriate skills and expertise to advise us;
- § identifying individuals qualified to become members of the board of directors;
- § recommending to the board of directors the persons to be nominated for election as directors and to each of the board's committees;
- § developing and recommending to the board of directors a code of business conduct and ethics and a set of corporate governance guidelines; and
- § overseeing the evaluation of our board of directors and management.

Science, Innovation and Technology Committee

Effective upon effectiveness of the registration statement of which this prospectus forms a part, our science, innovation and technology committee will be composed of Dr. Flier, Dr. Gilman and Dr. Springer, with Dr. Flier serving as chairman of the committee. The science, innovation and technology committee's responsibilities upon completion of this offering will include:

- § providing a general oversight function regarding our research and development activities;
- § providing recommendations to our board of directors and us regarding our long-term strategic goals and objectives related to our research and development programs;
- § providing recommendations regarding intellectual property strategies;
- § providing recommendations regarding key discovery and development strategies to align with our business needs; and
- § providing feedback to the board of directors and to our research and development group.

Our board of directors may from time to time establish other committees.

Compensation Committee Interlocks and Insider Participation

None of the members of our compensation committee has at any time during the prior three years been one of our officers or employees. None of our executive officers currently serves, or in the past fiscal year has served, as a member of the board of directors or compensation committee of any entity that has one or more executive officers serving on our board of directors or compensation committee.

Corporate Governance

We have adopted a written code of business conduct and ethics, effective upon the effectiveness of the registration statement of which this prospectus is a part, that applies to our directors, officers and employees, including our principal executive officer, principal financial officer, principal accounting officer or controller, or persons performing similar functions. Following the effectiveness of the registration statement of which this prospectus is a part, a current copy of the code will be posted on the investor relations section of our website, which is located at <http://www.scholarrock.com>. If we make any substantive amendments to, or grant any waivers from, the code of business conduct and ethics for any officer or director, we will disclose the nature of such amendment or waiver on our website or in a current report on Form 8-K.

EXECUTIVE COMPENSATION

Executive Compensation Overview

Our executive compensation program has reflected our growth and development-oriented corporate culture. To date, the compensation of the individuals listed below, whom we refer to as our named executive officers, has primarily consisted of a combination of base salary, bonuses and long-term incentive compensation. Our named executive officers, like all of our full-time employees, are eligible to participate in our health and welfare benefit plans. As we transition from a private company to a publicly traded company, we will evaluate our compensation values and philosophy and compensation plans and arrangements as circumstances require. At a minimum, we expect to review executive compensation annually with input from a compensation consultant. As part of this review process, we expect the board of directors and the compensation committee to apply our values and philosophy, while considering the compensation levels needed to ensure our executive compensation program remains competitive. We will also review whether we are meeting our retention objectives and the potential cost of replacing a key employee.

Summary Compensation Table — 2017

The following table presents information regarding the total compensation awarded to, earned by, and paid to our named executive officer for services rendered to us in all capacities for the year ended December 31, 2017.

Name and Principal Position	Year	Salary (\$)	Stock Awards (\$)	Non-Equity Incentive Plan Compensation (\$) ⁽¹⁾	Total (\$)
Nagesh K. Mahanthappa Ph.D., President and Chief Executive Officer	2017	382,454	590,810 ⁽²⁾	147,245	1,120,509
Yung H. Chyung M.D., Chief Medical Officer	2017	359,660	165,200 ⁽³⁾	118,688	643,548
Elan Z. Ezickson, Chief Operating Officer & Head of Corporate Development	2017	354,447	108,643 ⁽⁴⁾	116,968	580,058
		1,096,561	864,653	\$ 382,901	2,344,115

⁽¹⁾ Amounts reflect the cash incentive bonuses received by our named executive officers in 2018 for performance of services in 2017, which were based upon achievement of corporate performance goals.

⁽²⁾ \$391,065 of this amount reflects the aggregate grant date fair value of a stock award granted during the year calculated in accordance with the provisions of Financial Accounting Standards Board Accounting Standard Codification Topic 718, *Compensation — Stock Compensation*. For information regarding assumptions underlying the valuation of this stock award, see Note 12 to our financial statements appearing at the end of this prospectus. The remaining \$199,745 of this amount represents the incremental fair value resulting from the exchange of incentive units of Scholar Rock, LLC into shares of our common stock and restricted common stock in connection with the Reorganization, as further described in the section titled "Reorganization."

⁽³⁾ The amount reported for Dr. Chyung represents the incremental fair value resulting from the exchange of incentive units of Scholar Rock, LLC into shares of our common stock and restricted common stock in connection with the Reorganization. Dr. Chyung did not receive a stock award in 2017.

⁽⁴⁾ The amount reported for Mr. Ezickson represents the incremental fair value resulting from the exchange of incentive units of Scholar Rock, LLC into shares of our common stock and restricted common stock in connection with the Reorganization. Mr. Ezickson did not receive a stock award in 2017.

Narrative Disclosure to 2017 Summary Compensation Table

Base Salary

Each named executive officer's base salary is a fixed component of annual compensation for performing specific duties and functions, and has been established by our board of directors taking into account each individual's role, responsibilities, skills, and experience.

Non-Equity Incentive Plan Compensation

Our annual bonus program is intended to reward our named executive officers for meeting objective or subjective individual and/or company-wide performance goals for a fiscal year. For 2017, our named executive officers received incentive compensation based upon achievement of corporate objectives.

Long-Term Equity Incentives

Our equity grant program is intended to align the interests of our named executive officers with those of our stockholders and to motivate them to make important contributions to our performance.

Employment Arrangements and Severance Agreements with Our Named Executive Officers

Nagesh K. Mahanthappa, Ph.D.

For the year ended December 31, 2017, the annual base salary for Dr. Mahanthappa was \$382,454. For 2017, Dr. Mahanthappa was eligible to earn an annual cash incentive bonus targeted at 35% of his base salary, with the actual cash incentive bonus determined by the board of directors based on the achievement of specified corporate goals. Dr. Mahanthappa is also eligible to participate in the employee benefit plans available to our employees, subject to the terms of those plans.

Pursuant to Dr. Mahanthappa's offer letter, dated October 10, 2012, in the event that he is terminated by us without "cause" or he resigns for "good reason," subject to his execution of a separation agreement and general release, he will be entitled to (1) continuation of his base salary for a period of six successive months plus one additional successive month for each full year of service to us, up to a maximum of 12 total months following his termination of employment; provided, however, that such continuation of his base salary is subject to reduction in the event of his employment or self-employment after the initial three-month period, and (2) payment of health insurance premiums provided under COBRA following his termination of employment at the same rate as was in effect on the date of termination for a period of six successive months plus one additional successive month for each full year of service to us, up to a maximum of 12 total months. Additionally, in the event Dr. Mahanthappa is terminated without "cause" or he resigns for "good reason" within 18 months following a "sale event" (each as defined in his offer letter), he will be entitled to full acceleration of any unvested equity awards.

We intend to enter into a new employment agreement with Dr. Mahanthappa, effective upon the effectiveness of the registration statement of which this prospectus forms a part, pursuant to which Dr. Mahanthappa is entitled to receive an annual base salary of \$475,000 and an annual target bonus equal to 50% of his annual base salary based upon our board of directors' assessment of his performance and our attainment of targeted goals as set by our board of directors in its sole discretion. The new employment agreement provides that, in the event that his employment is terminated by us without "cause" or by him for "good reason," subject to the execution and effectiveness of a separation agreement and release, he will be entitled to receive (i) an amount equal to (x) 12 months of base salary, payable on our normal payroll cycle if such termination is not in connection with a "change in control" or (y) 18 months if such termination is in connection with a "change in control" within 18 months of such "change in control," plus 1.5 times his annual target bonus he would have been entitled to receive in the fiscal year of such termination and (ii) reimbursement of COBRA premiums for health benefit coverage for him and his immediate family in an amount equal to the monthly employer contribution that we would have made to provide health insurance to Dr. Mahanthappa had he remained employed with us for up to (x) 12 months following termination if such termination is not in connection with a "change in control" or (y) 18 months if

such termination is in connection with a "change in control". In addition, if within 18 months following a "change in control," Dr. Mahanthappa is terminated by us without "cause" or he resigns for "good reason," all time-based stock options and other time-based stock-based awards held by Dr. Mahanthappa will accelerate and vest immediately.

This new employment agreement incorporates the terms of the employee confidentiality, inventions assignment, non-solicitation and non-competition agreement previously entered into between us and Dr. Mahanthappa as described below under "— Employment Agreements and Severance Agreements with Our Named Executive Officers — Other Agreements."

Yung H. Chyung, M.D.

For the year ended December 31, 2017, the annual base salary for Dr. Chyung was \$359,660. For 2017, Dr. Chyung was eligible to earn an annual cash incentive bonus targeted at 30% of his base salary, with the actual cash incentive bonus determined by the board of directors based on the achievement of specified corporate goals. Dr. Chyung is also eligible to participate in the employee benefit plans available to our employees, subject to the terms of those plans.

Pursuant to Dr. Chyung's offer letter, dated February 2, 2016, in the event that he is terminated by us without "cause" or he resigns for "good reason," subject to his execution of a separation agreement and general release, he will be entitled to (1) continuation of his base salary for a period of six successive months plus one additional successive month for each full year of service to us, up to a maximum of nine total months following his termination of employment; provided, however, that such continuation of his base salary is subject to reduction in the event of his employment or self-employment after the initial three-month period, and (2) payment of health insurance premiums provided under COBRA following his termination of employment at the same rate as was in effect on the date of termination for a period of six successive months plus one additional successive month for each full year of service to us, up to a maximum of nine total months. Additionally, in the event Dr. Chyung is terminated without "cause" or he resigns for "good reason" within 18 months following a "sale event" (each as defined in his offer letter), he will be entitled to full acceleration of any unvested equity awards.

We intend to enter into a new employment agreement with Dr. Chyung, effective upon the effectiveness of the registration statement of which this prospectus forms a part, pursuant to which Dr. Chyung is entitled to receive an annual base salary of \$400,000 and an annual target bonus equal to 35% of his annual base salary based upon our board of directors' assessment of his performance and our attainment of targeted goals as set by our board of directors in its sole discretion. The new employment agreement provides that, in the event that his employment is terminated by us without "cause" or by him for "good reason," subject to the execution and effectiveness of a separation agreement and release, he will be entitled to receive (i) an amount equal to (x) 9 months of base salary, payable on our normal payroll cycle if such termination is not in connection with a "change in control" or (y) 12 months if such termination is in connection with a "change in control" within 18 months of such "change in control," plus 1.0 times his annual target bonus he would have been entitled to receive in the fiscal year of such termination and (ii) reimbursement of COBRA premiums for health benefit coverage for him and his immediate family in an amount equal to the monthly employer contribution that we would have made to provide health insurance to Dr. Chyung had he remained employed with us for up to (x) 9 months following termination if such termination is not in connection with a "change in control" or (y) 12 months if such termination is in connection with a "change in control". In addition, if within 18 months following a "change in control," Dr. Chyung is terminated by us without "cause" or he resigns for "good reason," all time-based stock options and other time-based stock-based awards held by Dr. Chyung will accelerate and vest immediately.

This new employment agreement incorporates the terms of the employee confidentiality, inventions assignment, non-solicitation and non-competition agreement previously entered into between us and Dr. Chyung as described below under "— Employment Agreements and Severance Agreements with Our Named Executive Officers — Other Agreements."

Elan Z. Ezickson

For the year ended December 31, 2017, the annual base salary for Mr. Ezickson was \$354,447. For 2017, Mr. Ezickson was eligible to earn an annual cash incentive bonus targeted at 30% of his base salary, with the actual cash incentive bonus determined by the board of directors based on the achievement of specified corporate goals. Mr. Ezickson is also eligible to participate in the employee benefit plans available to our employees, subject to the terms of those plans.

Pursuant to Mr. Ezickson's offer letter, dated July 17, 2014, in the event that he is terminated by us without "cause" or he resigns for "good reason," subject to his execution of a separation agreement and general release, he will be entitled to (1) continuation of his base salary for a period of six successive months plus one additional successive month for each full year of service to us, up to a maximum of nine total months following his termination of employment and (2) payment of health insurance premiums provided under COBRA following his termination of employment at the same rate as was in effect on the date of termination for a period of six successive months plus one additional successive month for each full year of service to us, up to a maximum of nine total months. Additionally, in the event Mr. Ezickson is terminated without "cause" or he resigns for "good reason" within 18 months following a "sale event" (each as defined in his offer letter), he will be entitled to full acceleration of any unvested equity awards.

We intend to enter into a new employment agreement with Mr. Ezickson, effective upon the effectiveness of the registration statement of which this prospectus forms a part, pursuant to which Mr. Ezickson is entitled to receive an annual base salary of \$380,000 and an annual target bonus equal to 35% of his annual base salary based upon our board of directors' assessment of his performance and our attainment of targeted goals as set by our board of directors in its sole discretion. The new employment agreement provides that, in the event that his employment is terminated by us without "cause" or by him for "good reason," subject to the execution and effectiveness of a separation agreement and release, he will be entitled to receive (i) an amount equal to (x) 9 months of base salary, payable on our normal payroll cycle if such termination is not in connection with a "change in control" or (y) 12 months if such termination is in connection with a "change in control" within 18 months of such "change in control," plus 1.0 times his annual target bonus he would have been entitled to receive in the fiscal year of such termination and (ii) reimbursement of COBRA premiums for health benefit coverage for him and his immediate family in an amount equal to the monthly employer contribution that we would have made to provide health insurance to Mr. Ezickson had he remained employed with us for up to (x) 9 months following termination if such termination is not in connection with a "change in control" or (y) 12 months if such termination is in connection with a "change in control". In addition, if within 18 months following a "change in control," Mr. Ezickson is terminated by us without "cause" or he resigns for "good reason," all time-based stock options and other time-based stock-based awards held by Mr. Ezickson will accelerate and vest immediately.

This new employment agreement incorporates the terms of the employee confidentiality, inventions assignment, non-solicitation and non-competition agreement previously entered into between us and Mr. Ezickson as described below under "— Employment Agreements and Severance Agreements with Our Named Executive Officers — Other Agreements."

Other Agreements

We have also entered into employee confidentiality, inventions assignment, non-solicitation and non-competition agreements with each of our named executive officers. Under such agreements, each named executive officer has agreed (1) not to compete with us during his or her employment and for a period of one year after the termination of such employment, (2) not to solicit our employees during his or her employment and for a period of one year after the termination of such employment, (3) to protect our confidential and proprietary information and (4) to assign to us related intellectual property developed during the course of his or her employment.

Outstanding Equity Awards as of December 31, 2017

The following table sets forth information concerning outstanding equity awards held by our named executive officers as of December 31, 2017:

Name and Principal Position	Number of Shares That Have Not Vested (#)⁽¹⁾	Market Value of Shares That Have Not Vested (\$)⁽²⁾
Nagesh K. Mahanthappa Ph.D., President and Chief Executive Officer ⁽³⁾	180,598	1,041,464
Yung H. Chyung M.D., Chief Medical Officer ⁽⁴⁾	116,250	670,388
Elan Z. Ezickson, Chief Operating Officer & Head of Corporate Development ⁽⁵⁾	133,547	770,129

⁽¹⁾ Stock award totals include shares of our restricted common stock received by the applicable named executive officer upon the exchange of incentive units of Scholar Rock, LLC in connection with the Reorganization.

⁽²⁾ There was no public market for our common stock as of December 31, 2017. The fair market value of our common stock as of December 31, 2017, as determined by our board of directors, was \$5.77 per share.

⁽³⁾ Represents incentive units that were exchanged for restricted common stock in connection with the Reorganization from the following grants: (1) 216,127 units granted on November 12, 2014, which vest as follows: 20% vested on November 12, 2015 and the remainder vesting in equal quarterly installments for a period of 16 quarters thereafter, and (2) 141,221 units granted on February 14, 2017, which vest in equal monthly installments over a period of four years beginning on August 12, 2016.

⁽⁴⁾ Represents incentive units that were exchanged for restricted common stock in connection with the Reorganization from the following grants: (1) 172,341 units granted on April 25, 2016, which shares vest as follows: 25% vested on February 25, 2017, with the remainder vesting in equal quarterly installments over a three year period thereafter, and (2) 34,328 units granted on August 12, 2016, which shares vest as follows: 25% vested on February 25, 2017, with the remainder vesting in equal quarterly installments over a three year period thereafter.

⁽⁵⁾ Represents incentive units that were exchanged for restricted common stock in connection with the Reorganization from the following grants: (1) 261,152 units granted on November 12, 2014, which vest as follows: 20% vested on August 1, 2015, with the remainder vesting in equal quarterly installments for a period of 16 quarters thereafter, and (2) 61,300 units granted on August 12, 2016, which vest in equal quarterly installments over a period of four years beginning on August 12, 2016.

Compensation Risk Assessment

We believe that although a portion of the compensation provided to our executive officers and other employees is performance-based, our executive compensation program does not encourage excessive or unnecessary risk taking. This is primarily due to the fact that our compensation programs are designed to encourage our executive officers and other employees to remain focused on both short-term and long-term strategic goals. As a result, we do not believe that our compensation programs are reasonably likely to have a material adverse effect on us.

Employee Benefit and Equity Compensation Plans**2018 Stock Option and Incentive Plan**

Our 2018 Stock Option and Incentive Plan, or 2018 Plan, was adopted by our board of directors on May 2, 2018, and approved by our stockholders on May 11, 2018, and will become effective upon effectiveness of the registration statement of which this prospectus is part. The 2018 Plan will replace our 2017 Stock Option and Grant Plan as our board of directors has determined not to make additional awards under that plan following the consummation of our initial public offering. The 2018 Plan allows the board of directors'

compensation committee to make equity-based incentive awards to our officers, employees, directors and other key persons (including consultants).

We have initially reserved 3,139,274 shares of our common stock for the issuance of awards under the 2018 Plan, or the Initial Limit (which includes 352,204 unused shares reserved for issuance under our 2017 Stock Option and Incentive Plan that will become available under our 2018 Plan upon the completion of this offering). The 2018 Plan provides that the number of shares reserved and available for issuance under the plan will automatically increase each January 1, beginning on January 1, 2019, by 4% of the outstanding number of shares of our common stock on the immediately preceding December 31 or such lesser number of shares as determined by our board of directors or compensation committee, or the Annual Increase. These limits are subject to adjustment in the event of a stock split, stock dividend or other change in our capitalization.

The shares we issue under the 2018 Plan will be authorized but unissued shares or shares we reacquire. The shares of common stock underlying any awards that are forfeited, cancelled, held back upon exercise or settlement of an award to satisfy the exercise price or tax withholding, reacquired by us prior to vesting, satisfied without the issuance of stock, expire or are otherwise terminated (other than by exercise) under the 2018 Plan and the 2017 Plan will be added back to the shares of common stock available for issuance under the 2018 Plan.

The maximum aggregate number of shares that may be issued in the form of incentive stock options shall not exceed the Initial Limit cumulatively increased on January 1, 2019 and on each January 1 thereafter by the lesser of the Annual Increase for such year or 1,885,946 shares of common stock.

The 2018 Plan will be administered by our compensation committee. Our compensation committee has full power to select, from among the individuals eligible for awards, the individuals to whom awards will be granted, to make any combination of awards to participants, and to determine the specific terms and conditions of each award, subject to the provisions of the 2018 Plan. Persons eligible to participate in the 2018 Plan will be those full or part-time officers, employees, non-employee directors and other key persons (including consultants) as selected from time to time by our compensation committee in its discretion. Our compensation committee may, in its discretion, delegate to a committee consisting of one or more officers the authority to make certain awards described in the 2018 Plan.

The 2018 Plan permits the granting of both options to purchase common stock intended to qualify as incentive stock options under Section 422 of the Code and options that do not so qualify. The option exercise price of each option will be determined by our compensation committee but may not be less than 100% of the fair market value of our common stock on the date of grant. The term of each option will be fixed by our compensation committee and may not exceed ten years from the date of grant. Our compensation committee will determine at what time or times each option may be exercised.

Our compensation committee may award stock appreciation rights subject to such conditions and restrictions as it may determine. Stock appreciation rights entitle the recipient to shares of common stock, or cash, equal to the value of the appreciation in our stock price over the exercise price. The exercise price may not be less than 100% of the fair market value of our common stock on the date of grant. The term of each stock appreciation right will be fixed by our compensation committee and may not exceed ten years from the date of grant. Our compensation committee will determine at what time or times each stock appreciation right may be exercised.

Our compensation committee may award restricted shares of common stock and restricted stock units to participants subject to such conditions and restrictions as it may determine. These conditions and restrictions may include the achievement of certain performance goals and/or continued employment with us through a specified vesting period. Our compensation committee may also grant shares of common stock that are free from any restrictions under the 2018 Plan. Unrestricted stock may be granted to participants

in recognition of past services or for other valid consideration and may be issued in lieu of cash compensation due to such participant. Our compensation committee may grant dividend equivalent rights to participants that entitle the recipient to receive credits for dividends that would be paid if the recipient had held a specified number of shares of common stock.

Our compensation committee may grant cash bonuses under the 2018 Plan to participants, subject to the achievement of certain performance goals.

The 2018 Plan provides that upon the effectiveness of a "sale event," as defined in the 2018 Plan, an acquirer or successor entity may assume, continue or substitute outstanding awards under the 2018 Plan. To the extent that awards granted under the 2018 Plan are not assumed or continued or substituted by the successor entity, upon the effective time of the sale event, such awards under the 2018 Plan shall terminate. In the event of such termination, individuals holding options and stock appreciation rights will be permitted to exercise such options and stock appreciation rights (to the extent exercisable) within a specified period of time prior to the sale event. In addition, in connection with the termination of the 2018 Plan upon a sale event, we may make or provide for a cash payment to participants holding vested and exercisable options and stock appreciation rights equal to the difference between the per share cash consideration payable to stockholders in the sale event and the exercise price of the options or stock appreciation rights and we may make or provide for a cash payment to participants holding other vested awards.

Our board of directors may amend or discontinue the 2018 Plan and our compensation committee may amend or cancel outstanding awards for purposes of satisfying changes in law or any other lawful purpose, but no such action may adversely affect rights under an award without the holder's consent. Certain amendments to the 2018 Plan require the approval of our stockholders.

No awards may be granted under the 2018 Plan after the date that is ten years from the date of stockholder approval of the 2018 Plan. No awards under the 2018 Plan have been made prior to the date hereof.

2017 Stock Option and Incentive Plan

Our 2017 Stock Option and Incentive Plan, or 2017 Plan, was approved and adopted by our board of directors on December 22, 2017, and approved by our stockholders on that same day. Under the 2017 Plan we reserved for issuance an aggregate of 3,455,330 shares of our common stock, subject to adjustment in the event of a stock split, reverse stock split, stock dividend, recapitalization, reclassification of shares, reorganization, or other similar change in our capitalization.

The shares of common stock underlying awards that are forfeited, cancelled, terminated, reacquired prior to vesting, satisfied without the issuance of shares of common stock, or withheld to cover the exercise price or tax withholding are added back to the shares of common stock available for issuance under the 2017 Plan.

Our board of directors has acted as administrator of the 2017 Plan. The administrator has full power to select, from among the individuals eligible for awards, the individuals to whom awards will be granted, and to determine the specific terms and conditions of each award, subject to the provisions of the 2017 Plan. Persons eligible to participate in the 2017 Plan are those full or part-time employees, key persons, officers and directors of, and consultants to, our company as selected from time to time by the administrator in its discretion.

The 2017 Plan permits the granting of (1) options to purchase common stock intended to qualify as incentive stock options under Section 422 of the Internal Revenue Code of 1986, as amended, (2) options that do not so qualify, (3) restricted stock, (4) unrestricted stock, or (5) restricted stock units. For stock options, the administrator will determine the per share option exercise price and at what time or times each option may be exercised.

The 2017 Plan provides that upon the occurrence of a merger, reorganization, consolidation, liquidation, dissolution, sale of all or substantially all of the Company's assets, acquisition of a majority of our voting stock, or any other transaction that our board determines to be an acquisition of the business, or a Sale Event, all outstanding stock options shall terminate and all outstanding restricted stock and restricted stock units shall be forfeited if not assumed, continued, or substituted with comparable awards by the successor entity. In the event of such termination, holders will be permitted to exercise any vested options (including those that will become vested as a result of the Sale Event) or we may, in our sole discretion, cancel such options in exchange for a cash payment equal to the value payable per share of stock in the Sale Event multiplied by the number of shares subject to the vested portion of the option, less the aggregate exercise price. In the event that restricted stock is forfeited in connection with a Sale Event, the 2017 Plan provides that the holder shall be paid the lower of the purchase price paid for the restricted stock or the fair market value of the stock at the time of the sale event. The Company may, but is not required to, cancel the restricted stock in exchange for a price per share equal to the value payable per share of stock in the Sale Event.

Our board of directors may amend or discontinue the 2017 Plan at any time, subject to stockholder approval where such approval is required by applicable law. Our board of directors may also amend or cancel any outstanding award, provided that no amendment to an award may adversely affect a participant's rights without his or her consent.

The 2017 Plan will terminate automatically on December 22, 2027; however awards previously granted may extend beyond that date. As of May 7, 2018, 2,367,498 shares of common stock and restricted common stock and options to purchase 1,015,858 shares of common stock were outstanding under the 2017 Plan. Our board of directors has determined not to make any further awards under the 2017 Plan following the completion of this offering.

2018 Employee Stock Purchase Plan

On May 2, 2018, our board of directors adopted the 2018 Employee Stock Purchase Plan, or 2018 ESPP, and on May 11, 2018, our stockholders approved the 2018 ESPP. The 2018 ESPP is intended to qualify as an "employee stock purchase plan" within the meaning of Section 423 of the Code. The 2018 ESPP initially reserves and authorizes the issuance of up to a total of 235,743 shares of common stock to participating employees. The ESPP provides that the number of shares reserved and available for issuance will automatically increase each January 1, beginning on January 1, 2019, by the lesser of (i) 353,614 shares of common stock, (ii) 1% of the outstanding number of shares of our common stock on the immediately preceding December 31 or (iii) such lesser number of shares as determined by the 2018 ESPP administrator. The number of shares reserved under the 2018 ESPP is subject to adjustment in the event of a stock split, stock dividend or other change in our capitalization.

All employees whose customary employment is for more than 20 hours per week are eligible to participate in the ESPP. However, any participating employee who would own 5% or more of the total combined voting power or value of all classes of stock after an option were granted under the ESPP would not be eligible to purchase shares under the 2018 ESPP.

We will make one or more offerings each year to our employees to purchase shares under the ESPP. Offerings will usually begin on each December 1 and June 1 and will continue for six-month periods, referred to as offering periods. Each eligible employee may elect to participate in any offering by submitting an enrollment form at least 15 business days before the relevant offering date.

Each employee who is a participant in the 2018 ESPP may purchase shares by authorizing payroll deductions of up to 15% of his or her base compensation during an offering period. Unless the participating employee has previously withdrawn from the offering, his or her accumulated payroll deductions will be used to purchase shares on the last business day of the offering period at a price equal to 85% of the fair market value of the shares on the first business day or the last business day of the

offering period, whichever is lower. Under applicable tax rules, an employee may purchase no more than \$25,000 worth of shares of common stock, valued at the start of the purchase period, under the ESPP in any calendar year.

The accumulated payroll deductions of any employee who is not a participant on the last day of an offering period will be refunded. An employee's rights under the 2018 ESPP terminate upon voluntary withdrawal from the plan or when the employee ceases employment with us for any reason.

The 2018 ESPP may be terminated or amended by our board of directors at any time. An amendment that increases the number of shares of common stock authorized under the ESPP and certain other amendments require the approval of our stockholders.

Senior Executive Cash Incentive Bonus Plan

On May 2, 2018, our board of directors adopted the Senior Executive Cash Incentive Bonus Plan, or the Bonus Plan. The Bonus Plan provides for cash bonus payments based upon the attainment of performance targets established by our compensation committee. The payment targets will be related to financial and operational measures or objectives with respect to our company, or Corporate Performance Goals, as well as individual performance objectives.

Our compensation committee may select Corporate Performance Goals from among the following: cash flow (including, but not limited to, operating cash flow and free cash flow); sales or revenue; corporate revenue; earnings before interest, taxes, depreciation and amortization; net income (loss) (either before or after interest, taxes, depreciation and/or amortization); changes in the market price of our common stock; economic value-added; development, clinical, regulatory or commercial milestones; acquisitions or strategic transactions, partnerships or joint ventures; operating income (loss); return on capital, assets, equity, or investment; stockholder returns; return on sales; gross or net profit levels; productivity; expense efficiency; margins; operating efficiency; customer satisfaction; working capital; earnings (loss) per share of our common stock; sales or market shares; number of customers; operating income and/or other strategic, financial or operational objectives, any of which may be measured in absolute terms, as compared to any incremental increase, in terms of growth, as compared to results of a peer group, against the market as a whole, compared to applicable market indices and/or measured on a pre-tax or post-tax basis.

Each executive officer who is selected to participate in the Bonus Plan will have a target bonus opportunity set for each performance period. The bonus formulas will be adopted in each performance period by the compensation committee and communicated to each executive. The Corporate Performance Goals will be measured at the end of each performance period after our financial reports have been published or such other appropriate time as the compensation committee determines. If the Corporate Performance Goals and individual performance objectives are met, payments will be made as soon as practicable following the end of each performance period. The Bonus Plan also permits the compensation committee to approve additional bonuses to executive officers in its sole discretion.

401(k) Plan

We maintain the Scholar Rock Holding Corporation 401(k) Plan, a tax-qualified retirement plan for our employees. Our 401(k) plan is intended to qualify under Section 401(k) of the Code so that contributions to our 401(k) plan by employees or by us, and the investment earnings thereon, are not taxable to the employees until withdrawn from our 401(k) plan, and so that contributions by us, if any, will be deductible by us when made. Under our 401(k) plan, employees may elect to reduce their current compensation by up to the statutorily prescribed annual limit and to have the amount of such reduction contributed to our 401(k) plan. We have not historically made any discretionary matching contributions under our 401(k) plan but may do so in the future.

Limitations on Liability and Indemnification Matters

Our amended and restated certificate of incorporation contains provisions that limit the liability of our directors for monetary damages to the fullest extent permitted by the Delaware General Corporation Law, or DGCL. Consequently, our directors are not personally liable to us or our stockholders for monetary damages for any breach of fiduciary duties as directors, except liability for:

- § any breach of the director's duty of loyalty to us or our stockholders;
- § any act or omission not in good faith or that involves intentional misconduct or a knowing violation of law;
- § unlawful payments of dividends or unlawful stock repurchases or redemptions as provided in Section 174 of the DGCL; or
- § any transaction from which the director derived an improper personal benefit.

Our amended and restated bylaws require us to indemnify our directors and officers to the maximum extent not prohibited by the DGCL and allow us to indemnify other employees and agents as set forth in the DGCL. Subject to certain limitations, our amended and restated bylaws also require us to advance expenses incurred by our directors and officers for the defense of any action for which indemnification is required or permitted.

We have entered, and intend to continue to enter, into separate indemnification agreements with our directors, officers and certain of our key employees, in addition to the indemnification provided for in our amended and restated certificate of incorporation and amended and restated bylaws. These agreements, among other things, require us to indemnify our directors, officers and key employees for certain expenses, including attorneys' fees, judgments, penalties, fines and settlement amounts actually incurred by these individuals in any action or proceeding arising out of their service to us or any of our subsidiaries or any other company or enterprise to which these individuals provide services at our request. Subject to certain limitations, our indemnification agreements also require us to advance expenses incurred by our directors, officers and key employees for the defense of any action for which indemnification is required or permitted.

We believe that provisions of our amended and restated certificate of incorporation, amended and restated bylaws and indemnification agreements are necessary to attract and retain qualified directors, officers and key employees. We also maintain directors' and officers' liability insurance.

The limitation of liability and indemnification provisions in our amended and restated certificate of incorporation and amended and restated bylaws may discourage stockholders from bringing a lawsuit against our directors and officers for breach of their fiduciary duty. They may also reduce the likelihood of derivative litigation against our directors and officers, even though an action, if successful, might benefit us and other stockholders. Further, a stockholder's investment may be adversely affected to the extent that we pay the costs of settlement and damage awards against directors and officers as required by these indemnification provisions.

At present, there is no pending litigation or proceeding involving any of our directors or executive officers as to which indemnification is required or permitted, and we are not aware of any threatened litigation or proceeding that may result in a claim for indemnification.

Insofar as indemnification for liabilities arising under the Securities Act may be permitted to directors, executive officers or persons controlling us, we have been informed that, in the opinion of the SEC, such indemnification is against public policy as expressed in the Securities Act and is therefore unenforceable.

DIRECTOR COMPENSATION

The following table presents the total compensation for each person who served as a non-employee member of our board of directors and received compensation for such service during the year ended December 31, 2017. Other than as set forth in the table and described more fully below, we did not pay any compensation, make any additional equity awards or non-equity awards to, or pay any other compensation to any of the non-employee members of our board of directors in 2017. We reimburse non-employee members of our board of directors for reasonable travel and out-of-pocket expenses incurred in attending meetings of our board of directors and committees of our board of directors.

We also do not, and do not expect to, provide separate compensation to our directors who are also our employees, such as Dr. Mahanthappa, our President and Chief Executive Officer. Dr. Mahanthappa's compensation as an executive officer is reported above in "Executive Compensation — Summary Compensation Table."

Name	Fees Earned or Paid in Cash (\$)	Stock Awards (\$) ⁽¹⁾	Total (\$)
David Hallal	75,000 ⁽³⁾	1,453,425 ⁽²⁾⁽³⁾	1,528,425
Katrine Bosley ⁽⁴⁾	—	61,594 ⁽⁵⁾	61,594
Kristina Burow ⁽⁶⁾	—	—	—
Jeffrey Flier ⁽¹¹⁾	—	27,440 ⁽⁷⁾	27,440
Michael Gilman ⁽¹¹⁾	—	11,780 ⁽⁸⁾	11,780
Amir Nashat, Sc.D. ⁽⁹⁾	—	—	—
Timothy A. Springer, Ph.D. ⁽¹¹⁾	—	—	—

⁽¹⁾ As of December 31, 2017, our directors held the following number of shares of restricted common stock: David Hallal (256,021 shares), Jeffrey Flier (25,746 shares) and Michael Gilman (10,650 shares). Katrine Bosley, Kristina Burow, Amir Nashat, Sc.D. and Timothy A. Springer, Ph.D. did not hold any unvested shares as of December 31, 2017.

⁽²⁾ \$1,152,717 of this amount reflects the aggregate grant date fair value of stock awards granted during 2017 calculated in accordance with the provisions of Financial Accounting Standards Board Accounting Standard Codification Topic 718, *Compensation — Stock Compensation*. For information regarding assumptions underlying the valuation of stock awards, see Note 12 to our financial statements appearing at the end of this prospectus. The amount reported for Mr. Hallal includes the incremental fair value of \$300,708 resulting from the exchange of incentive units of Scholar Rock, LLC into shares of restricted and unrestricted common stock of Scholar Rock Holding Corporation in connection with the Reorganization. Pursuant to the Hallal Letter Agreement, Mr. Hallal's equity is subject to (1) an additional six months of vesting if Mr. Hallal's service is terminated without "cause" (as defined in the Hallal Letter Agreement) and Mr. Hallal has provided at least six months of service as of that date and (2) full acceleration upon the consummation of a "sale event" (as defined in the Hallal Letter agreement).

⁽³⁾ Pursuant to a letter agreement entered into between Mr. Hallal and the Company (the "Hallal Letter Agreement"), Mr. Hallal is eligible to receive an annual retainer equal to \$150,000, payable on a quarterly basis. The amounts reported in this column represent Mr. Hallal's annual retainer, from July 1, 2017, the date he joined the Board.

⁽⁴⁾ Ms. Bosley resigned from our board of directors on February 21, 2017. Any unvested portion of her stock award accelerated and became fully vested as of the date of her resignation.

⁽⁵⁾ The amount reported for Ms. Bosley includes the incremental fair value of \$11,780 resulting from the exchange of incentive units of Scholar Rock, LLC into shares of common stock of Scholar Rock Holding Corporation in connection with the Reorganization.

⁽⁶⁾ Ms. Burow did not receive compensation in connection with serving as a director.

- (7) The amount reported for Mr. Flier represents the incremental fair value resulting from the exchange of incentive units of the Scholar Rock, LLC into shares of restricted and unrestricted common stock of Scholar Rock Holding Corporation in connection with the Reorganization.
- (8) The amount reported for Mr. Gilman represents the incremental fair value resulting from the exchange of incentive units of Scholar Rock, LLC into shares of restricted and unrestricted common stock of Scholar Rock Holding Corporation in connection with the Reorganization.
- (9) Dr. Nashat did not receive compensation in connection with serving as a director.
- (10) Dr. Springer did not receive compensation in connection with serving as a director.
- (11) Drs. Flier, Gilman and Springer each received cash compensation in connection with consulting services provided by each individual to us. In addition, Drs. Flier and Gilman also received equity awards for the consulting services provided by each of them to us, which will fully accelerate and become fully vested immediately prior to any initial public offering of our securities. See the section titled "Certain Relationships and Related Party Transactions" for more details on the consulting services provided to us by Dr. Springer.

Non-Employee Director Compensation Policy

Our board of directors intends to adopt a non-employee director compensation policy, to be effective upon effectiveness of the registration statement of which this prospectus forms a part, that is designed to enable us to attract and retain, on a long-term basis, highly qualified non-employee directors. Under the policy, each director who is not an employee will be paid cash compensation from and after the completion of this offering, as set forth below:

	Member Annual Fee (\$)	Chairman Additional Annual Fee (\$)
Board of Directors	35,000	115,000
Audit Committee	7,500	7,500
Compensation Committee	5,000	5,000
Nominating and Corporate Governance Committee	4,000	4,000
Science, Innovation and Technology Committee	4,000	4,000

In addition, each non-employee director serving on our board of directors upon completion of this offering and each non-employee director elected or appointed to our board of directors following the completing of this offering will be granted a one-time equity award of 20,316 shares on the date of such director's election or appointment to the board of directors, which will vest monthly over three years, subject to continued service through such vesting dates. On the date of each annual meeting of stockholders of our company, each non-employee director (other than a director receiving an appointment equity award within eight months prior to such annual meeting) will be granted an annual equity award of 10,158 shares, which will vest in full of the earlier to occur of the first anniversary of the date of grant or the next scheduled annual meeting, subject to continued service as a director through such vesting date.

CERTAIN RELATIONSHIPS AND RELATED PARTY TRANSACTIONS

Other than the compensation agreements and other arrangements described under "Executive Compensation" and "Director Compensation" in this prospectus and the transactions described below, since December 31, 2014, there has not been and there is not currently proposed, any transaction or series of similar transactions to which we were, or will be, a party in which the amount involved exceeded, or will exceed, \$120,000 and in which any director, executive officer, holder of 5% or more of any class of our capital stock or any member of the immediate family of, or entities affiliated with, any of the foregoing persons, had, or will have, a direct or indirect material interest.

Series A-3 Convertible Preferred Unit Financing

On February 12, 2015, we sold an aggregate of 5,579,709 Series A-3 convertible preferred units at a purchase price of \$1.38 per unit, pursuant to a unit purchase agreement entered into with certain of our investors. Certain investors holding bridge units, originally issued in 2014, exchanged such bridge units for shares of our Series A-3 convertible preferred units. The following table summarizes purchases of our Series A-3 convertible preferred units by related persons:

<u>5% Stockholder</u>	<u>Series A-3 Convertible Preferred Units⁽¹⁾ (#)</u>	<u>Total Purchase Price (\$)</u>
Entities Affiliated with Polaris Venture Partners VI, L.P. ⁽²⁾	1,449,275	2,000,000
ARCH Venture Fund VIII, L.P. ⁽³⁾	1,811,595	2,500,001
Timothy A. Springer, Ph.D. ⁽⁴⁾	1,449,275	2,000,000
Entities Affiliated with EcoRI Capital Fund Qualified, L.P. ⁽⁵⁾	513,043	707,999

(1) All outstanding Series A-3 convertible preferred units were exchanged for our shares of Series A-3 convertible preferred stock on a one-for-one basis on December 22, 2017 in connection with the Reorganization. Following the reverse split of our common stock in May 2018, every 2.8548 shares of Series A-3 convertible preferred stock became convertible into one share of common stock.

(2) Polaris Venture Partners VI, L.P. is an affiliate fund of Polaris Venture Partners Founders' Fund VI, L.P. and is a holder of 5% or more of our capital stock. The amount set forth in the table consists of (1) 1,369,259 Series A-3 convertible preferred units purchased by Polaris Venture Partners VI, L.P. and (2) 80,016 Series A-3 convertible preferred units purchased by Polaris Venture Partners Founders' Fund VI, L.P. Amir Nashat, Sc.D., a partner at Polaris Venture Partners VI, is a member of our board of directors.

(3) Kristina Burow is a managing director of ARCH Venture Fund VIII, L.P. ARCH Venture Fund VIII, L.P. is a holder of 5% or more of our capital stock.

(4) Timothy A. Springer, Ph.D. is holder of 5% or more of our capital stock and is a member of our board of directors.

(5) EcoRI Capital Fund Qualified, L.P. is an affiliate fund of EcoRI Capital Fund, L.P., and is a holder of 5% or more of our capital stock. The amount set forth in the table consists of (1) 318,076 Series A-3 convertible preferred units purchased by EcoRI Capital Fund Qualified, L.P. and (2) 194,967 Series A-3 convertible preferred units purchased by EcoRI Capital Fund, L.P.

Series A-4 Convertible Preferred Unit Financing

On September 21, 2015, we sold an aggregate of 3,906,738 Series A-4 convertible preferred units at a purchase price of \$1.587 per unit, pursuant to a unit purchase agreement entered into with certain of our investors. The following table summarizes purchases of our Series A-4 convertible preferred units by related persons:

<u>5% Stockholder</u>	<u>Series A-4 Convertible Preferred Units⁽¹⁾ (#)</u>	<u>Total Purchase Price (\$)</u>
Entities Affiliated with Polaris Venture Partners VI, L.P. ⁽²⁾	787,649	1,249,999
ARCH Venture Fund VIII, L.P. ⁽³⁾	1,575,299	2,500,000
Timothy A. Springer, Ph.D. ⁽⁴⁾	787,649	1,249,999
Entities Affiliated with EcoRI Capital Fund Qualified, L.P. ⁽⁵⁾	446,123	707,997

- (1) All outstanding Series A-4 convertible preferred units were exchanged for our shares of Series A-4 convertible preferred stock on a one-for-one basis on December 22, 2017 in connection with the Reorganization. Following the reverse split of our common stock in May 2018, every 2.8548 shares of Series A-4 convertible preferred stock became convertible into one share of common stock.
- (2) Polaris Venture Partners VI, L.P. is an affiliate fund of Polaris Venture Partners Founders' Fund VI, L.P. and is a holder of 5% or more of our capital stock. The amount set forth in the table consists of (1) 744,162 Series A-4 convertible preferred units purchased by Polaris Venture Partners VI, L.P. and (2) 43,487 Series A-4 convertible preferred units purchased by Polaris Venture Partners Founders' Fund VI, L.P. Dr. Nashat, Sc.D., a partner at Polaris Venture Partners VI, is a member of our board of directors.
- (3) Kristina Burow is a managing director of ARCH Venture Fund VIII, L.P. ARCH Venture Fund VIII, L.P. is a holder of 5% or more of our capital stock.
- (4) Timothy A. Springer, Ph.D. is holder of 5% or more of our capital stock and is a member of our board of directors.
- (5) EcoRI Capital Fund Qualified, L.P. is an affiliate fund of EcoRI Capital Fund, L.P., and is a holder of 5% or more of capital stock. The amount set forth in the table consists of (1) 276,587 Series A-4 convertible preferred units purchased by EcoRI Capital Fund Qualified, L.P. and (2) 169,536 Series A-4 convertible preferred units purchased by EcoRI Capital Fund, L.P.

Series B Convertible Preferred Unit Financing

On December 17, 2015, and May 18, 2017, we sold an aggregate of 13,526,994 Series B convertible preferred units at a purchase price of \$3.00 per unit, pursuant to a unit purchase agreement entered into with certain of our investors. The following table summarizes purchases of our Series B convertible preferred units by related persons:

<u>5% Stockholder</u>	<u>Series B Convertible Preferred Units⁽¹⁾ (#)</u>	<u>Total Purchase Price (\$)</u>
Entities Affiliated with Polaris Venture Partners VI, L.P. ⁽²⁾	2,094,875	6,284,625
ARCH Venture Fund VIII, L.P. ⁽³⁾	2,054,197	6,162,591
Timothy A. Springer, Ph.D. ⁽⁴⁾	2,094,875	6,284,625
Entities Affiliated with EcoRI Capital Fund Qualified, L.P. ⁽⁵⁾	976,068	2,928,204
Entities Affiliated with Fidelity Management and Research Company ⁽⁶⁾	5,360,334	16,081,002

- (1) All outstanding Series B convertible preferred units were exchanged for our shares of our Series B convertible preferred stock on a one-for-one basis on December 22, 2017 in connection with the Reorganization. Following the reverse split of our common stock in May 2018, every 2.5848 shares of Series B convertible preferred stock became convertible into one share of common stock.

- (2) Polaris Venture Partners VI, L.P. is an affiliate fund of Polaris Venture Partners Founders' Fund VI, L.P. and is a holder of 5% or more of our capital stock. The amount set forth in the table consists of (1) 1,979,216 Series B convertible preferred units purchased by Polaris Venture Partners VI, L.P. and (2) 115,659 Series B convertible preferred units purchased by Polaris Venture Partners Founders' Fund VI, L.P. Dr. Nashat, Sc.D., a partner at Polaris Venture Partners VI, is a member of our board of directors.
- (3) Kristina Burow is a managing director of ARCH Venture Fund VIII, L.P. ARCH Venture Fund VIII, L.P. is a holder of 5% or more of our capital stock.
- (4) Timothy A. Springer, Ph.D. controls TAS Partners, LLC. Dr. Springer is holder of 5% or more of our capital stock and a member of our board of directors. The amount set forth in the table consists of (1) 666,666 shares of Series B convertible preferred units purchased by TAS Partners, LLC and (2) 1,428,209 shares of Series B convertible preferred units purchased by Dr. Springer.
- (5) EcoRI Capital Fund Qualified, L.P. is an affiliate fund of EcoRI Capital Fund, L.P. and is a holder of 5% or more of our capital stock. The amount set forth in the table consists of (1) 687,152 shares of Series B convertible preferred units purchased by EcoRI Capital Fund Qualified, L.P. and (2) 288,916 shares of Series B convertible preferred units purchased by EcoRI Capital Fund, L.P.
- (6) Fidelity Management and Research Company is an affiliate of Fidelity Advisor Series VII: Biotechnology Fund and Fidelity Select Portfolios: Biotechnology Portfolio and is a beneficial owner of 5% or more of our capital stock. The amount set forth in the table consists of (1) 1,083,994 shares of Series B convertible preferred units held by Fidelity Advisor Series VII: Biotechnology Fund and (2) 4,276,340 shares of Series B convertible preferred units purchased by Fidelity Select Portfolios: Biotechnology Portfolio.

Series C Convertible Preferred Stock Financing

On December 22, 2017, immediately following the Reorganization, we sold an aggregate of 13,055,555 shares of our Series C convertible preferred stock at a purchase price of \$3.60 per share, pursuant to a stock purchase agreement entered into with certain of our investors. The following table summarizes purchases of our Series C convertible preferred stock by related persons:

5% Stockholder	Series C Preferred Stock (#)⁽¹⁾	Total Purchase Price (\$)
Entities Affiliated with Polaris Venture Partners VI, L.P. ⁽²⁾	855,391	3,079,408
ARCH Venture Fund VIII, L.P. ⁽³⁾	838,780	3,019,608
Timothy A. Springer, Ph.D. ⁽⁴⁾	855,391	3,079,408
Entities Affiliated with EcoRI Capital Fund Qualified, L.P. ⁽⁵⁾	415,343	1,495,235
Entities Affiliated with Fidelity Management and Research Company ⁽⁶⁾	2,777,778	10,000,001
Artal International SCA ⁽⁷⁾	5,555,556	20,000,002

(1) Following the reverse split of our common stock in May 2018, every 2.5848 shares of Series C convertible preferred stock became convertible into one share of common stock.

(2) Polaris Venture Partners VI, L.P. is an affiliate fund of Polaris Venture Partners Founders' Fund VI, L.P. and is a holder of 5% or more of our capital stock. The amount set forth in the table consists of (1) 808,166 shares of Series C convertible preferred stock purchased by Polaris Venture Partners VI, L.P. and (2) 47,225 shares of Series C convertible preferred stock purchased by Polaris Venture Partners Founders' Fund VI, L.P. Dr. Amir Nashat, Sc.D., a partner at Polaris Venture Partners VI, is a member of our board of directors.

(3) Kristina Burow is a managing director of ARCH Venture Fund VIII, L.P. ARCH Venture Fund VIII, L.P. is a holder of 5% or more of our capital stock.

(4) Timothy A. Springer, Ph.D. is holder of 5% or more of our capital stock and a member of our board of directors.

- ⁽⁵⁾ EcoRI Capital Fund Qualified, L.P. is an affiliate fund of EcoRI Capital Fund, L.P., and is a holder of 5% or more of our capital stock. The amount set forth in the table consists of (1) 328,036 shares of Series C convertible preferred stock held by EcoRI Capital Fund Qualified, L.P. and (2) 87,307 shares of Series C convertible preferred stock held by EcoRI Capital Fund, L.P.
- ⁽⁶⁾ Fidelity Management and Research Company is an affiliate of Fidelity Mt. Vernon Street Trust: Fidelity Series Growth Company Fund State Street Bank & Trust, Fidelity Growth Company Commingled Pool and Fidelity Mt. Vernon Street Trust: Fidelity Series Growth Company Fund BNY Mellon, and is a beneficial owner of 5% or more of our capital stock. The amount set forth in the table consists of (1) 267,635 shares of Series C convertible preferred stock purchased by Fidelity Mt. Vernon Street Trust: Fidelity Series Growth Company Fund State Street Bank & Trust, (2) 1,166,154 shares of Series C convertible preferred stock purchased by Fidelity Growth Company Commingled Pool, and (3) 1,343,989 shares of Series C convertible preferred stock purchased by Fidelity Mt. Vernon Street Trust: Fidelity Series Growth Company Fund BNY Mellon.
- ⁽⁷⁾ Artal International SCA is a holder of 5% or more of our capital stock.

Consulting Agreements

The Company entered into a consulting agreement on October 10, 2012 with Timothy A. Springer, Ph.D. to provide services related to the advancement of the research and development platform of the company.

The consulting arrangement is on a fixed-fee basis, paid quarterly. The initial contract term was four years and terminated on October 10, 2016. The contract was extended for an additional four year period. The Company incurred \$80,000 of consulting expense related to this contract, in each year, for the years ended December 31, 2016 and 2017.

Indemnification Agreements

In connection with this offering, we intend to enter into agreements to indemnify our directors and executive officers. These agreements will, among other things, require us to indemnify these individuals for certain expenses (including attorneys' fees), judgments, fines and settlement amounts reasonably incurred by such person in any action or proceeding, including any action by or in our right, on account of any services undertaken by such person on behalf of our company or that person's status as a member of our board of directors to the maximum extent allowed under Delaware law.

Investors' Rights Agreement

In connection with our Series C convertible preferred stock financing, we entered into an investors' rights agreement with certain of our stockholders, including related persons. The investors' rights agreement among other things:

- § grants such stockholders certain registration rights with respect to shares of our common stock, including shares of common stock issued or issuable upon conversion of our convertible preferred stock;
- § obligates us to deliver periodic financial statements to any stockholder who holds at least 1,000,000 shares of our convertible preferred stock, which we refer to as a "Majority Investors;"
- § grants a right of first offer with respect to sales of our shares by us, subject to specified exclusions (which exclusions include the sale of the shares in connection with this offering), to qualified holders; and
- § requires us to reimburse certain legal expenses of the investors in connection with future financings or a liquidation event.

For more information regarding the registration rights provided in this agreement, please refer to the section of this prospectus titled "Description of Capital Stock — Registration Rights."

Certain provisions of this agreement, including the covenants described above other than registration rights, will terminate automatically upon completion of this offering. This is not a complete description of the

investors' rights agreement and is qualified by the full text of the investors' rights agreement filed as an exhibit to the registration statement of which this prospectus is a part.

Voting Agreement

In connection with our Series C convertible preferred stock financing, we entered into a voting agreement with certain of our stockholders, including related persons. The voting agreement among other things: provides the terms for the voting of shares with respect to the constituency of our board of directors.

This voting agreement will terminate automatically upon completion of this offering. This is not a complete description of the voting agreement and is qualified by the full text of the voting agreement filed as an exhibit to the registration statement of which this prospectus is a part.

Right of First Refusal and Co-Sale Agreement

In connection with our Series C convertible preferred stock financing, we entered into a right of first refusal and co-sale agreement with certain of our stockholders, including related persons. The right of first refusal and co-sale agreement, among other things:

§ grants our investors certain rights of first refusal and co-sale with respect to proposed transfers of our securities by certain stockholders; and

§ grants us certain rights of first refusal with respect to proposed transfers of our securities by certain stockholders.

The right of first refusal and co-sale agreement will terminate automatically upon completion of this offering. This is not a complete description of the right of first refusal and co-sale agreement and is qualified by the full text of right of first refusal and co-sale agreement filed as an exhibit to the registration statement of which this prospectus is a part.

Policies for Approval of Related Party Transactions

Our board of directors reviews and approves transactions with directors, officers and holders of five percent or more of our voting securities and their affiliates, each a related party. Prior to this offering, the material facts as to the related party's relationship or interest in the transaction are disclosed to our board of directors prior to their consideration of such transaction, and the transaction is not considered approved by our board of directors unless a majority of the directors who are not interested in the transaction approve the transaction. Further, when stockholders are entitled to vote on a transaction with a related party, the material facts of the related party's relationship or interest in the transaction are disclosed to the stockholders, who must approve the transaction in good faith.

In connection with this offering, we have adopted a written related party transactions policy that such transactions must be approved by our audit committee. This policy will become effective on the date on which the registration statement of which this prospectus is part is declared effective by the SEC. Pursuant to this policy, the audit committee has the primary responsibility for reviewing and approving or disapproving "related party transactions," which are transactions between us and related persons in which the aggregate amount involved exceeds or may be expected to exceed \$120,000 and in which a related person has or will have a direct or indirect material interest. For purposes of this policy, a related person will be defined as a director, executive officer, nominee for director, or greater than 5% beneficial owner of our common stock, in each case since the beginning of the most recently completed year, and their immediate family members.

PRINCIPAL STOCKHOLDERS

The following table sets forth certain information known to us regarding beneficial ownership of our capital stock as of March 31, 2018, as adjusted to reflect the sale of common stock offered by us in this offering, for:

- § each person or group of affiliated persons known by us to be the beneficial owner of more than 5% of our capital stock;
- § each of our named executive officers;
- § each of our directors; and
- § all of our executive officers and directors as a group.

Beneficial ownership is determined in accordance with the rules of the SEC and generally includes voting or investment power with respect to securities. Under those rules, beneficial ownership includes any shares as to which the individual or entity has sole or shared voting power or investment power, and includes securities that the individual or entity has the right to acquire, such as through the exercise of stock options, within 60 days of March 31, 2018. Except as noted by footnote, and subject to community property laws where applicable, we believe, based on the information provided to us, that the persons and entities named in the table below have sole voting and investment power with respect to all common stock shown as beneficially owned by them.

The percentage of beneficial ownership prior to this offering in the table below is based on shares of common stock deemed to be outstanding as of March 31, 2018, assuming the conversion of all outstanding shares of our convertible preferred stock upon the completion of this offering into an aggregate of 15,109,950 shares of common stock upon the completion of this offering, and the percentage of beneficial ownership after this offering in the table below is based on shares of common stock assumed to be outstanding after the completion of the offering.

Name of Beneficial Owner ⁽¹⁾	Number of Shares Beneficially Owned Prior to Offering	Percentage of Shares Beneficially Owned	
		Before Offering	After Offering
5% or greater stockholders:			
Timothy A. Springer, Ph.D. ⁽²⁾	3,477,994	18.2%	14.2%
Affiliates of Fidelity Management and Research Company ⁽³⁾	2,850,675	14.9	11.7
Entities Affiliated with Polaris Venture Partners VI, L.P. ⁽⁴⁾	2,689,849	14.1	11.0
ARCH Venture Fund VIII, L.P. ⁽⁵⁾	2,637,617	13.8	10.8
Artal International SCA	1,946,040	10.2	8.0
EcoRI Capital Fund Qualified, LP ⁽⁶⁾	995,600	5.2	4.1
Named executive officers and directors:			
Nagesh K. Mahanthappa, Ph.D. ⁽⁷⁾	633,197	3.3	2.6
Yung H. Chyung, M.D. ⁽⁸⁾	209,294	1.1	*
Elan Z. Ezickson ⁽⁹⁾	322,451	1.7	1.4
David Halla ⁽¹⁰⁾	299,527	1.6	1.2
Kristina Burow ⁽¹¹⁾	2,637,617	13.8	10.8
Jeffrey S. Flier, M.D. ⁽¹²⁾	44,085	*	*
Michael Gilman, Ph.D. ⁽¹³⁾	44,259	*	*
Amir Nashat, Sc.D. ⁽¹⁴⁾	2,689,849	14.1	11.0
Timothy A. Springer, Ph.D. ⁽²⁾	3,477,994	18.2	14.2
All executive officers and directors as a group (11 persons)⁽¹⁵⁾	10,518,747	55.1%	43.0%

* Represents less than 1%.

- (1) Address is c/o Scholar Rock Holding Corporation, 620 Memorial Dr., 2nd Floor, Cambridge, MA, unless otherwise indicated.
- (2) Consists of 3,202,091 shares of common stock and shares of common stock issuable upon conversion of shares of convertible preferred stock held by Timothy A. Springer and 275,903 shares of common stock issuable upon conversion of shares of convertible preferred stock held by TAS Partners LLC.
- (3) Consists of (i) 379,709 shares of common stock issuable upon conversion of shares of convertible preferred stock held by Fidelity Advisor Series VII: Biotechnology Fund, (ii) 1,497,947 shares of common stock issuable upon conversion of shares of convertible preferred stock held by Fidelity Select Portfolios: Biotechnology Portfolio, (iii) 93,749 shares of common stock issuable upon conversion of shares of convertible preferred stock held by Fidelity Mt. Vernon Street Trust: Fidelity Series Growth Company Fund State Street Bank & Trust, (iv) 408,488 shares of common stock issuable upon conversion of shares of convertible preferred stock held by Fidelity Growth Company Commingled Pool, and (v) 470,782 shares of common stock issuable upon conversion of shares of convertible preferred stock held by Fidelity Mt. Vernon Street Trust: Fidelity Growth Company Fund. The Funds that hold shares of our capital stock are managed by direct or indirect subsidiaries of Fidelity Management & Research, LLC, or FMR LLC. Edward C. Johnson 3rd is a Director and the Chairman of FMR LLC and Abigail P. Johnson is a Director, the Vice Chairman and the President of FMR LLC. Members of the family of Edward C. Johnson 3rd, including Abigail P. Johnson, are the predominant owners, directly or through trusts, of Series B voting common shares of FMR LLC, representing 49% of the voting power of FMR LLC. The Johnson family group and all other Series B shareholders have entered into a shareholders' voting agreement under which all Series B voting common shares will be voted in accordance with the majority vote of Series B voting common shares. Accordingly, through their ownership of voting common shares and the execution of the shareholders' voting agreement, members of the Johnson family may be deemed, under the Investment Company Act of 1940, to form a controlling group with respect to FMR LLC. Neither FMR LLC nor Edward C. Johnson 3rd nor Abigail P. Johnson has the sole power to vote or direct the voting of the shares owned directly by the various investment companies registered under the Investment Company Act, or Fidelity Funds, advised by Fidelity Management & Research Company, or FMR Co, a wholly owned subsidiary of FMR LLC, which power resides with the Fidelity Funds' Boards of Trustees. Fidelity Management & Research Company carries out the voting of the shares under written guidelines established by the Fidelity Funds' Boards of Trustees.
- (4) Consists of (i) 2,541,341 shares of common stock issuable upon conversion of shares of convertible preferred stock held by Polaris Venture Partners VI, L.P. and (ii) 148,508 shares of common stock issuable upon conversion of shares of convertible preferred stock held by Polaris Venture Partners Founders Fund VI, L.P. Polaris Venture Management Co. VI, L.L.C. is a general partner of each of the funds that hold shares of our capital stock and may be deemed to have the sole voting and dispositive power over the shares held by these funds. Dr. Amir Nashat, a member of our board of directors, is a Managing Partner of Polaris Partners and may be deemed to share voting and dispositive power over the shares held by these funds. The address of the Polaris funds is Polaris Funds is One Marina Park Drive, 10th Floor, Boston, Massachusetts 02210.
- (5) The sole general partner of this fund is ARCH Venture Partners VIII, L.P. or ARCH Partners VIII, which may be deemed to beneficially own the shares held by this fund. The sole general partner of ARCH Partners VIII is ARCH Venture Partners VIII, LLC or ARCH VIII LLC, which may be deemed to beneficially own the shares held by this fund. ARCH Partners VIII and ARCH VIII LLC disclaim beneficial ownership of such shares, except to the extent of any pecuniary interest therein. The managing directors of ARCH VIII LLC are Keith L. Crandell, Clinton Bybee and Robert Nelsen, and they may be deemed to beneficially own the shares held by this fund. Messrs. Crandell, Bybee and Nelsen disclaim beneficial ownership of such shares, except to the extent of any pecuniary interest therein.
- (6) Consists of (i) 324,915 shares of common stock issuable upon conversion of shares of convertible preferred stock held by EcoR1 Capital Fund, LP and (ii) 670,685 shares of common stock issuable upon conversion of shares of convertible preferred stock held by EcoR1 Capital Fund Qualified, LP. EcoR1 Capital, LLC, as the sole general partner of EcoR1 Capital Fund, L.P. and EcoR1 Capital Fund Qualified, L.P., may be deemed to beneficially own the shares held of record by EcoR1 Capital Fund, L.P. and EcoR1 Capital Fund Qualified, L.P. The address of the EcoR1 funds is 409 Illinois Street, San Francisco, CA 94158.
- (7) Consists of 620,062 shares of common stock and 13,135 shares of common stock issuable upon exercise of options to purchase common stock exercisable within 60 days of March 31, 2018.
- (8) Consists of 206,667 shares of common stock and 2,627 shares of common stock issuable upon exercise of options to purchase common stock exercisable within 60 days of March 31, 2018.
- (9) Consists of 322,451 shares of common stock and, with respect to beneficial ownership following this offering, an additional 8,757 shares of common stock issuable upon exercise of options to purchase common stock vesting upon the closing of this offering.
- (10) Consists of 292,595 shares of common stock and 6,932 shares of common stock issuable upon exercise of options to purchase common stock exercisable within 60 days of March 31, 2018.

- (11) Consists of the shares described in footnote (5) above. Kristina Burow one of our directors, is a managing director at ARCH Venture Partners. Ms. Burow owns an interest in ARCH Partners VIII but does not have voting or investment control over the shares held by the fund, and disclaims beneficial ownership of such shares, except to the extent of any pecuniary interest therein. The address of the fund is 8755 West Higgins Road, Suite 1025, Chicago, Illinois 60631.
- (12) Consists of 43,785 shares of common stock and 300 shares of common stock issuable upon exercise of options to purchase common stock exercisable within 60 days of March 31, 2018.
- (13) Consists of 43,959 shares of common stock and 300 shares of common stock issuable upon exercise of options to purchase common stock exercisable within 60 days of March 31, 2018.
- (14) Consists of the shares described in footnote (4) above. Dr. Nashat is a partner at Polaris Venture Partners VI, L.P. and shares voting and investment control with respect to these shares. Dr. Nashat disclaims beneficial ownership of all shares held by Polaris Venture Partners VI, L.P. except to the extent of any pecuniary interest therein.
- (15) Consists of an aggregate of 10,492,607 shares of common stock and common stock issuable upon conversion of convertible preferred stock, an aggregate of 26,140 shares of common stock issuable upon exercise of options to purchase common stock exercisable within 60 days of March 31, 2018, and with respect to beneficial ownership following this offering, an additional 8,757 shares of common stock issuable upon exercise of options to purchase common stock vesting upon the closing of this offering.

DESCRIPTION OF CAPITAL STOCK

The following descriptions are summaries of the material terms of our amended and restated certificate of incorporation, which will be effective upon the completion of this offering and amended and restated by-laws, which will be effective upon the effectiveness of the registration statement of which this prospectus is a part. The descriptions of the common stock and convertible preferred stock give effect to changes to our capital structure that will occur immediately prior to the completion of this offering.

General

Upon completion of this offering, our authorized capital stock will consist of 150,000,000 shares of common stock, par value \$0.001 per share, and 10,000,000 shares of convertible preferred stock, par value \$0.001 per share, all of which shares of convertible preferred stock will be undesignated.

As of March 31, 2018, 3,970,586 shares of our common stock and 43,135,911 shares of convertible preferred stock were outstanding and held by 83 stockholders of record. These amounts do not give effect to the conversion of all outstanding shares of our convertible preferred stock into common stock upon the completion of this offering.

Common Stock

The holders of our common stock are entitled to one vote for each share held on all matters submitted to a vote of the stockholders. The holders of our common stock do not have any cumulative voting rights. Holders of our common stock are entitled to receive ratably any dividends declared by our board of directors out of funds legally available for that purpose, subject to any preferential dividend rights of any outstanding convertible preferred stock. Our common stock has no preemptive rights, conversion rights or other subscription rights or redemption or sinking fund provisions.

In the event of our liquidation, dissolution or winding up, holders of our common stock will be entitled to share ratably in all assets remaining after payment of all debts and other liabilities and any liquidation preference of any outstanding convertible preferred stock. The shares to be issued by us in this offering will be, when issued and paid for, validly issued, fully paid and non-assessable.

Preferred Stock

Upon the completion of this offering, all outstanding shares of our convertible preferred stock will be converted into shares of our common stock. Upon the consummation of this offering, our board of directors will have the authority, without further action by our stockholders, to issue up to 10,000,000 shares of convertible preferred stock in one or more series and to fix the rights, preferences, privileges and restrictions thereof. These rights, preferences and privileges could include dividend rights, conversion rights, voting rights, terms of redemption, liquidation preferences, sinking fund terms and the number of shares constituting, or the designation of, such series, any or all of which may be greater than the rights of common stock. The issuance of our convertible preferred stock could adversely affect the voting power of holders of common stock and the likelihood that such holders will receive dividend payments and payments upon our liquidation. In addition, the issuance of convertible preferred stock could have the effect of delaying, deferring or preventing a change in control of our company or other corporate action. Immediately after consummation of this offering, no shares of convertible preferred stock will be outstanding, and we have no present plan to issue any shares of convertible preferred stock.

Registration Rights

Upon the completion of this offering, the holders of 15,109,950 shares of our common stock, including those issuable upon the conversion of convertible preferred stock will be entitled to rights with respect to the registration of these securities under the Securities Act. These rights are provided under the terms of an

investors' rights agreement between us and holders of our convertible preferred stock. The investors' rights agreement includes demand registration rights, short-form registration rights and piggyback registration rights. All fees, costs and expenses of underwritten registrations under this agreement will be borne by us and all selling expenses, including underwriting discounts and selling commissions, will be borne by the holders of the shares being registered.

Demand Registration Rights

Beginning 180 days after the effective date of this registration statement, the holders of 15,109,950 shares of our common stock, including those issuable upon the conversion of convertible preferred stock upon completion of this offering, are entitled to demand registration rights. Under the terms of the investors' rights agreement, we will be required, upon the written request of holders of at least 25% of these securities that would result in an aggregate offering price of at least \$3.0 million, to file a registration statement and use best efforts to effect the registration of all or a portion of these shares for public resale. We are required to effect only two registrations pursuant to this provision of the investors' rights agreement.

Short-Form Registration Rights

Pursuant to the investors' rights agreement, if we are eligible to file a registration statement on Form S-3, upon the written request of at least 10% of these holders to sell registrable securities at an aggregate price of at least \$1.0 million, we will be required to use commercially reasonable efforts to effect a registration of such shares. We are required to effect only two registrations in any twelve month period pursuant to this provision of the investors' rights agreement. The right to have such shares registered on Form S-3 is further subject to other specified conditions and limitations.

Piggyback Registration Rights

Pursuant to the investors' rights agreement, if we register any of our securities either for our own account or for the account of other security holders, the holders of these shares are entitled to include their shares in the registration. Subject to certain exceptions contained in the investors' rights agreement, we and the underwriters may limit the number of shares included in the underwritten offering to the number of shares which we and the underwriters determine in our sole discretion will not jeopardize the success of the offering.

Indemnification

Our investors' rights agreement contains customary cross-indemnification provisions, under which we are obligated to indemnify holders of registrable securities in the event of material misstatements or omissions in the registration statement attributable to us, and they are obligated to indemnify us for material misstatements or omissions attributable to them.

Expiration of Registration Rights

The demand registration rights and short form registration rights granted under the investors' rights agreement will terminate on the fifth anniversary of the completion of this offering or at such time after this offering when the holders' shares may be sold without restriction pursuant to Rule 144 within a three month period.

Anti-Takeover Effects of Our Amended and Restated Certificate of Incorporation and Amended and Restated By-laws and Delaware Law

Our amended and certificate of incorporation and amended and restated by-laws include a number of provisions that may have the effect of delaying, deferring or preventing another party from acquiring control of us and encouraging persons considering unsolicited tender offers or other unilateral takeover proposals to negotiate with our board of directors rather than pursue non-negotiated takeover attempts. These provisions include the items described below.

Board Composition and Filling Vacancies

Our amended and restated certificate of incorporation provides for the division of our board of directors into three classes serving staggered three-year terms, with one class being elected each year. Our amended and restated certificate of incorporation also provides that directors may be removed only for cause and then only by the affirmative vote of the holders of two-thirds or more of the shares then entitled to vote at an election of directors. Furthermore, any vacancy on our board of directors, however occurring, including a vacancy resulting from an increase in the size of our board, may only be filled by the affirmative vote of a majority of our directors then in office even if less than a quorum. The classification of directors, together with the limitations on removal of directors and treatment of vacancies, has the effect of making it more difficult for stockholders to change the composition of our board of directors.

No Written Consent of Stockholders

Our amended and restated certificate of incorporation provides that all stockholder actions are required to be taken by a vote of the stockholders at an annual or special meeting, and that stockholders may not take any action by written consent in lieu of a meeting. This limit may lengthen the amount of time required to take stockholder actions and would prevent the amendment of our amended and restated bylaws or removal of directors by our stockholders without holding a meeting of stockholders.

Meetings of Stockholders

Our amended and restated certificate of incorporation and amended and restated by-laws provide that only a majority of the members of our board of directors then in office may call special meetings of stockholders and only those matters set forth in the notice of the special meeting may be considered or acted upon at a special meeting of stockholders. Our amended and restated by-laws limit the business that may be conducted at an annual meeting of stockholders to those matters properly brought before the meeting.

Advance Notice Requirements

Our amended and restated by-laws establish advance notice procedures with regard to stockholder proposals relating to the nomination of candidates for election as directors or new business to be brought before meetings of our stockholders. These procedures provide that notice of stockholder proposals must be timely given in writing to our corporate secretary prior to the meeting at which the action is to be taken. Generally, to be timely, notice must be received at our principal executive offices not less than 90 days nor more than 120 days prior to the first anniversary date of the annual meeting for the preceding year. Our by-laws specify the requirements as to form and content of all stockholders' notices. These requirements may preclude stockholders from bringing matters before the stockholders at an annual or special meeting.

Amendment to Amended and Restated Certificate of Incorporation and Amended and Restated By-laws

Any amendment of our amended and restated certificate of incorporation must first be approved by a majority of our board of directors, and if required by law or our amended and restated certificate of incorporation, must thereafter be approved by a majority of the outstanding shares entitled to vote on the amendment and a majority of the outstanding shares of each class entitled to vote thereon as a class, except that the amendment of the provisions relating to stockholder action, board composition, limitation of liability and the amendment of our by-laws and certificate of incorporation must be approved by not less than two-thirds of the outstanding shares entitled to vote on the amendment, and not less than two-thirds of the outstanding shares of each class entitled to vote thereon as a class. Our amended and restated by-laws may be amended by the affirmative vote of a majority of the directors then in office, subject to any limitations set forth in the amended and restated by-laws, and may also be amended by the affirmative vote of at least two-thirds of the outstanding shares entitled to vote on the amendment, or, if our board of directors recommends that the stockholders approve the amendment, by the affirmative vote of the majority of the outstanding shares entitled to vote on the amendment, in each case voting together as a single class.

Undesignated Preferred Stock

Our amended and restated certificate of incorporation provides for 10,000,000 authorized shares of convertible preferred stock. The existence of authorized but unissued shares of convertible preferred stock

may enable our board of directors to discourage an attempt to obtain control of us by means of a merger, tender offer, proxy contest or otherwise. For example, if in the due exercise of its fiduciary obligations, our board of directors were to determine that a takeover proposal is not in the best interests of our stockholders, our board of directors could cause shares of convertible preferred stock to be issued without stockholder approval in one or more private offerings or other transactions that might dilute the voting or other rights of the proposed acquirer or insurgent stockholder or stockholder group. In this regard, our amended and restated certificate of incorporation grants our board of directors broad power to establish the rights and preferences of authorized and unissued shares of convertible preferred stock. The issuance of shares of convertible preferred stock could decrease the amount of earnings and assets available for distribution to holders of shares of common stock. The issuance may also adversely affect the rights and powers, including voting rights, of these holders and may have the effect of delaying, deterring or preventing a change in control of us.

Choice of Forum

Our amended and restated bylaws provide that, unless we consent in writing to the selection of an alternative forum, the Court of Chancery of the State of Delaware will be the sole and exclusive forum for: (1) any derivative action or proceeding brought on our behalf; (2) any action asserting a claim of breach of a fiduciary duty or other wrongdoing by any of our directors, officers, employees or agents to us or our stockholders; (3) any action asserting a claim against us arising pursuant to any provision of the General Corporation Law of the State of Delaware or our certificate of incorporation or by-laws; (4) any action to interpret, apply, enforce or determine the validity of our certificate of incorporation or bylaws; or (5) any action asserting a claim governed by the internal affairs doctrine.

In addition, our amended and restated bylaws that will become effective upon the closing of this offering will contain a provision by virtue of which, unless we consent in writing to the selection of an alternative forum, the United States District Court for the District of Massachusetts will be the exclusive forum for any private action asserting violations by us or any of our directors or officers of the Securities Act, or the rules and regulations promulgated thereunder, and of all suits in equity and actions at law brought to enforce any liability or duty created by those statutes or the rules and regulations under such statutes. If any action the subject matter of which is within the scope of the preceding sentence is filed in a court other than the United States District Court for the District of Massachusetts, the plaintiff or plaintiffs shall be deemed by this provision of our amended and restated bylaws (i) to have consented to removal of the action by us to the United States District Court for the District of Massachusetts, in the case of an action filed in a state court, and (ii) to have consented to transfer of the action to the United States District Court for the District of Massachusetts.

Our amended and restated bylaws also provide that any person or entity purchasing or otherwise acquiring any interest in shares of our capital stock will be deemed to have notice of and to have consented to this choice of forum provision. It is possible that a court of law could rule that the choice of forum provisions contained in our amended and restated bylaws are inapplicable or unenforceable if they are challenged in a proceeding or otherwise. The enforceability of similar choice of forum provisions in other companies' certificates of incorporation and bylaws has been challenged in legal proceedings.

Section 203 of the Delaware General Corporation Law

Upon completion of this offering, we will be subject to the provisions of Section 203 of the Delaware General Corporation Law. In general, Section 203 prohibits a publicly held Delaware corporation from engaging in a "business combination" with an "interested stockholder" for a three-year period following the time that this stockholder becomes an interested stockholder, unless the business combination is approved in a prescribed manner. Under Section 203, a business combination between a corporation and an interested stockholder is prohibited unless it satisfies one of the following conditions:

- § before the stockholder became interested, our board of directors approved either the business combination or the transaction which resulted in the stockholder becoming an interested stockholder;
- § upon consummation of the transaction which resulted in the stockholder becoming an interested stockholder, the interested stockholder owned at least 85% of the voting stock of the corporation outstanding at the time the transaction commenced, excluding for purposes of determining the voting stock outstanding, shares owned by persons who are directors and also officers, and employee stock plans, in some instances, but not the outstanding voting stock owned by the interested stockholder; or
- § at or after the time the stockholder became interested, the business combination was approved by our board of directors and authorized at an annual or special meeting of the stockholders by the affirmative vote of at least two-thirds of the outstanding voting stock which is not owned by the interested stockholder.

Section 203 defines a business combination to include:

- § any merger or consolidation involving the corporation and the interested stockholder;
- § any sale, transfer, lease, pledge or other disposition involving the interested stockholder of 10% or more of the assets of the corporation;
- § subject to exceptions, any transaction that results in the issuance or transfer by the corporation of any stock of the corporation to the interested stockholder;
- § subject to exceptions, any transaction involving the corporation that has the effect of increasing the proportionate share of the stock of any class or series of the corporation beneficially owned by the interested stockholder; and
- § the receipt by the interested stockholder of the benefit of any loans, advances, guarantees, pledges or other financial benefits provided by or through the corporation.

In general, Section 203 defines an interested stockholder as any entity or person beneficially owning 15% or more of the outstanding voting stock of the corporation and any entity or person affiliated with or controlling or controlled by the entity or person.

Nasdaq Global Market Listing

We have applied to list our common stock on The Nasdaq Global Market under the trading symbol "SRRK."

Transfer Agent and Registrar

The transfer agent and registrar for our common stock will be Computershare Trust Company, N.A. The transfer agent and registrar's address is 250 Royall Street, Canton, Massachusetts 02021, and its telephone number is (800) 962-4284.

SHARES ELIGIBLE FOR FUTURE SALE

Prior to this offering, there has been no public market for our shares. Future sales of our common stock in the public market, or the availability of such shares for sale in the public market, could adversely affect market prices prevailing from time to time. As described below, only a limited number of shares will be available for sale shortly after this offering due to contractual and legal restrictions on resale. Nevertheless, sales of our common stock in the public market after such restrictions lapse, or the perception that those sales may occur, could adversely affect the prevailing market price at such time and our ability to raise equity capital in the future.

Based on the number of shares outstanding as of March 31, 2018 upon the completion of this offering, 24,440,536 shares of our common stock will be outstanding. Of the outstanding shares, all of the shares sold in this offering will be freely tradable, except that any shares held by our affiliates, as that term is defined in Rule 144 under the Securities Act, may only be sold in compliance with the limitations described below. All remaining shares of common stock held by existing stockholders immediately prior to the completion of this offering will be "restricted securities" as such term is defined in Rule 144. These restricted securities were issued and sold by us, or will be issued and sold by us, in private transactions and are eligible for public sale only if registered under the Securities Act or if they qualify for an exemption from registration under the Securities Act, including the exemptions provided by Rule 144 or Rule 701, summarized below.

Rule 144

In general, a person who has beneficially owned restricted stock for at least six months would be entitled to sell their securities provided that (i) such person is not deemed to have been one of our affiliates at the time of, or at any time during the 90 days preceding, a sale and (ii) we are subject to the Securities Exchange Act of 1934, as amended, or the Exchange Act, periodic reporting requirements for at least 90 days before the sale. Persons who have beneficially owned restricted shares for at least six months but who are our affiliates at the time of, or any time during the 90 days preceding, a sale, would be subject to additional restrictions, by which such person would be entitled to sell within any three-month period only a number of securities that does not exceed the greater of the following:

- § 1% of the number of shares then outstanding, which will equal approximately 244,405 shares immediately after this offering, assuming no exercise of the underwriters' option to purchase additional shares, based on the number of shares outstanding as of March 31, 2018; or
- § the average weekly trading volume of our common stock on The Nasdaq Global Market during the four calendar weeks preceding the filing of a notice on Form 144 with respect to the sale;

provided, in each case, that we are subject to the Exchange Act periodic reporting requirements for at least 90 days before the sale. Such sales both by affiliates and by non-affiliates must also comply with the manner of sale, current public information and notice provisions of Rule 144.

Rule 701

Rule 701 under the Securities Act, as in effect on the date of this prospectus, permits resales of shares in reliance upon Rule 144 but without compliance with certain restrictions of Rule 144, including the holding period requirement. Most of our employees, executive officers or directors who purchased shares under a written compensatory plan or contract may be entitled to rely on the resale provisions of Rule 701, but all holders of Rule 701 shares are required to wait until 90 days after the date of this prospectus before selling their shares.

However, substantially all Rule 701 shares are subject to lock-up agreements as described below and under "Underwriting" included elsewhere in this prospectus and will become eligible for sale upon the expiration of the restrictions set forth in those agreements.

Lock-Up Agreements

We, our directors and executive officers and holders of substantially all of our common stock have signed a lock-up agreement that prevent us and them from selling any of our common stock or any securities convertible into or exercisable or exchangeable for common stock for a period of not less than 180 days from the date of this prospectus without the prior written consent of the Underwriters, subject to certain exceptions. See the section entitled "Underwriters" appearing elsewhere in this prospectus for more information.

Registration Rights

Upon completion of this offering, certain holders of our securities will be entitled to various rights with respect to registration of their shares under the Securities Act. Registration of these shares under the Securities Act would result in these shares becoming fully tradable without restriction under the Securities Act immediately upon the effectiveness of the registration. See the section entitled "Description of Capital Stock — Registration Rights" appearing elsewhere in this prospectus for more information.

Equity Incentive Plans

We intend to file one or more registration statements on Form S-8 under the Securities Act to register our shares issued or reserved for issuance under our equity incentive plans. The first such registration statement is expected to be filed soon after the date of this prospectus and will automatically become effective upon filing with the SEC. Accordingly, shares registered under such registration statement will be available for sale in the open market, unless such shares are subject to vesting restrictions with us or the lock-up restrictions described above. We estimate that such registration statement on Form S-8 will cover approximately 4,400,000 shares.

**CERTAIN MATERIAL U.S. FEDERAL INCOME TAX CONSIDERATIONS FOR
NON-U.S. HOLDERS**

The following discussion is a summary of the material U.S. federal income tax considerations applicable to non-U.S. holders (as defined below) with respect to their ownership and disposition of shares of our common stock issued pursuant to this offering. For purposes of this discussion, a non-U.S. holder means a beneficial owner of our common stock that is for U.S. federal income tax purposes:

- § a non-resident alien individual;
- § a foreign corporation or any other foreign organization taxable as a corporation for U.S. federal income tax purposes; or
- § a foreign estate or trust, the income of which is not subject to U.S. federal income tax on a net income basis.

This discussion does not address the tax treatment of partnerships or other entities that are pass-through entities for U.S. federal income tax purposes or persons that hold their common stock through partnerships or other pass-through entities. A partner in a partnership or other pass-through entity that will hold our common stock should consult his, her or its tax advisor regarding the tax consequences of acquiring, holding and disposing of our common stock through a partnership or other pass-through entity, as applicable.

This discussion is based on current provisions of the U.S. Internal Revenue Code of 1986, as amended, which we refer to as the Code, existing and proposed U.S. Treasury Regulations promulgated thereunder, current administrative rulings and judicial decisions, all as in effect as of the date of this prospectus and, all of which are subject to change or to differing interpretation, possibly with retroactive effect. Any such change or differing interpretation could alter the tax consequences to non-U.S. holders described in this prospectus. There can be no assurance that the Internal Revenue Service, which we refer to as the IRS, will not challenge one or more of the tax consequences described herein. We assume in this discussion that a non-U.S. holder holds shares of our common stock as a capital asset within the meaning of Section 1221 of the Code, generally property held for investment.

This discussion does not address all aspects of U.S. federal income taxation that may be relevant to a particular non-U.S. holder in light of that non-U.S. holder's individual circumstances nor does it address any aspects of any U.S. federal tax other than the income tax, U.S. state, local or non-U.S. taxes, the alternative minimum tax, the rules regarding qualified small business stock within the meaning of Section 1202 of the Code or the Medicare tax on net investment income. This discussion also does not consider any specific facts or circumstances that may apply to a non-U.S. holder and does not address the special tax rules applicable to particular non-U.S. holders, such as:

- § insurance companies;
- § tax-exempt or governmental organizations;
- § financial institutions;
- § brokers or dealers in securities;
- § regulated investment companies;
- § pension plans;
- § "controlled foreign corporations," "passive foreign investment companies," and corporations that accumulate earnings to avoid U.S. federal income tax;
- § "qualified foreign pension funds," or entities wholly owned by a "qualified foreign pension fund";
- § partnerships or other entities or arrangements treated as partnerships for U.S. federal income tax purposes (and partners and investors therein);
- § persons deemed to sell our common stock under the constructive sale provisions of the Code;

- § persons that hold our common stock as part of a straddle, hedge, conversion transaction, synthetic security or other integrated investment;
- § persons who hold or receive our common stock pursuant to the exercise of any employee stock option or otherwise as compensation; and
- § certain U.S. expatriates.

This discussion is for general information only and is not tax advice. Accordingly, all prospective non-U.S. holders of our common stock should consult their tax advisors with respect to the U.S. federal, state, local and non-U.S. tax consequences of the purchase, ownership and disposition of our common stock.

Distributions on Our Common Stock

Distributions, if any, on our common stock will constitute dividends for U.S. federal income tax purposes to the extent paid from our current or accumulated earnings and profits, as determined under U.S. federal income tax principles. If a distribution exceeds our current and accumulated earnings and profits, the excess will be treated as a tax-free return of the non-U.S. holder's investment, up to such holder's tax basis in the common stock. Any remaining excess will be treated as capital gain, subject to the tax treatment described below in "Gain on Sale or Other Taxable Disposition of Our Common Stock." Any such distributions will also be subject to the discussions below under the sections titled "Backup Withholding and Information Reporting" and "Withholding and Information Reporting Requirements — FATCA."

Subject to the discussion in the following two paragraphs in this section, dividends paid to a non-U.S. holder generally will be subject to withholding of U.S. federal income tax at a 30% rate or such lower rate as may be specified by an applicable income tax treaty between the United States and such holder's country of residence.

Dividends that are treated as effectively connected with a trade or business conducted by a non-U.S. holder within the United States and, if an applicable income tax treaty so provides, that are attributable to a permanent establishment or a fixed base maintained by the non-U.S. holder within the United States, are generally exempt from the 30% withholding tax if the non-U.S. holder satisfies applicable certification and disclosure requirements. However, such U.S. effectively connected income, net of specified deductions and credits, is taxed at the same graduated U.S. federal income tax rates applicable to United States persons (as defined in the Code). Any U.S. effectively connected income received by a non-U.S. holder that is a corporation may also, under certain circumstances, be subject to an additional "branch profits tax" at a 30% rate or such lower rate as may be specified by an applicable income tax treaty between the United States and such holder's country of residence.

A non-U.S. holder of our common stock who claims the benefit of an applicable income tax treaty between the United States and such holder's country of residence generally will be required to provide a properly executed IRS Form W-8BEN or W-8BEN-E (or successor form) to the applicable withholding agent and satisfy applicable certification and other requirements. Non-U.S. holders are urged to consult their tax advisors regarding their entitlement to benefits under a relevant income tax treaty. A non-U.S. holder that is eligible for a reduced rate of U.S. withholding tax under an income tax treaty may obtain a refund or credit of any excess amounts withheld by timely filing a U.S. tax return with the IRS.

Gain on Sale or Other Taxable Disposition of Our Common Stock

Subject to the discussions below under "Backup Withholding and Information Reporting" and "Withholding and Information Reporting Requirements — FATCA," a non-U.S. holder generally will not be subject to any U.S. federal income tax on any gain realized upon such holder's sale or other taxable disposition of shares of our common stock unless:

- § the gain is effectively connected with the non-U.S. holder's conduct of a U.S. trade or business and, if an applicable income tax treaty so provides, is attributable to a permanent establishment or a fixed-base maintained by such non-U.S. holder in the United States, in which case the non-U.S. holder generally will be taxed on a net income basis at the graduated U.S. federal income tax rates applicable to United States persons (as defined in the Code) and, if the non-U.S. holder is a foreign corporation, the branch profits tax described above in "Distributions on Our Common Stock" also may apply;
- § the non-U.S. holder is a nonresident alien individual who is present in the United States for 183 days or more in the taxable year of the disposition and certain other conditions are met, in which case the non-U.S. holder will be subject to a 30% tax (or such lower rate as may be specified by an applicable income tax treaty between the United States and such holder's country of residence) on the net gain derived from the disposition, which may be offset by certain U.S. source capital losses of the non-U.S. holder, if any (even though the individual is not considered a resident of the United States), provided that the non-U.S. holder has timely filed U.S. federal income tax returns with respect to such losses; or
- § we are, or have been, at any time during the five-year period preceding such sale or other taxable disposition (or the non-U.S. holder's holding period, if shorter) a "U.S. real property holding corporation," unless our common stock is regularly traded on an established securities market and the non-U.S. holder holds no more than 5% of our outstanding common stock, directly or indirectly, actually or constructively, during the shorter of the 5-year period ending on the date of the disposition or the period that the non-U.S. holder held our common stock. Generally, a corporation is a U.S. real property holding corporation only if the fair market value of its U.S. real property interests equals or exceeds 50% of the sum of the fair market value of its worldwide real property interests plus its other assets used or held for use in a trade or business. Although there can be no assurance, we do not believe that we are, or have been, a U.S. real property holding corporation, or that we are likely to become one in the future. No assurance can be provided that our common stock will be regularly traded on an established securities market for purposes of the rules described above. If we are a U.S. real property holding corporation and either our common stock is not regularly traded on an established securities market or a non-U.S. holder holds, or is treated as holding, more than 5% of our outstanding common stock, directly or indirectly, during the applicable testing period, any gain recognized by such non-U.S. holder will generally be subject to U.S. federal income tax rates in the same manner as if the non-U.S. holder were a resident of the United States. If we are a U.S. real property holding corporation and our common stock is not regularly traded on an established securities market, such non-U.S. holder's proceeds received on the disposition of shares will also generally be subject to withholding at a rate of 15%. Prospective investors are encouraged to consult their own tax advisors regarding the possible consequences to them if we are, or were to become, a U.S. real property holding corporation.

Backup Withholding and Information Reporting

We must report annually to the IRS and to each non-U.S. holder the gross amount of the distributions on our common stock paid to such holder and the tax withheld, if any, with respect to such distributions. Non-U.S. holders may have to comply with specific certification procedures to establish that the holder is not a United States person (as defined in the Code) in order to avoid backup withholding at the applicable rate with respect to dividends on our common stock. Dividends paid to non-U.S. holders subject to

withholding of U.S. federal income tax, as described above in "Distributions on Our Common Stock," generally will be exempt from U.S. backup withholding.

Information reporting and backup withholding will generally apply to the proceeds of a disposition of our common stock by a non-U.S. holder effected by or through the U.S. office of any broker, U.S. or foreign, unless the holder certifies its status as a non-U.S. holder and satisfies certain other requirements, or otherwise establishes an exemption. Generally, information reporting and backup withholding will not apply to a payment of disposition proceeds to a non-U.S. holder where the transaction is effected outside the United States through a non-U.S. office of a broker. However, for information reporting purposes, dispositions effected through a non-U.S. office of a broker with substantial U.S. ownership or operations generally will be treated in a manner similar to dispositions effected through a U.S. office of a broker. Non-U.S. holders should consult their tax advisors regarding the application of the information reporting and backup withholding rules to them. Copies of information returns may be made available to the tax authorities of the country in which the non-U.S. holder resides or is incorporated under the provisions of a specific treaty or agreement. Backup withholding is not an additional tax. Any amounts withheld under the backup withholding rules from a payment to a non-U.S. holder can be refunded or credited against the non-U.S. holder's U.S. federal income tax liability, if any, provided that an appropriate claim is filed with the IRS in a timely manner.

Withholding and Information Reporting Requirements — FATCA

The Foreign Account Tax Compliance Act, or FATCA, generally imposes a U.S. federal withholding tax at a rate of 30% on payments of dividends on, or gross proceeds from the sale or other disposition of, our common stock paid to a foreign entity unless (i) if the foreign entity is a "foreign financial institution," such foreign entity undertakes certain due diligence, reporting, withholding, and certification obligations, (ii) if the foreign entity is not a "foreign financial institution," such foreign entity identifies certain of its U.S. investors, if any, or (iii) the foreign entity is otherwise exempt under FATCA. Under applicable U.S. Treasury regulations, withholding under FATCA currently applies to payments of dividends on our common stock, but will only apply to payments of gross proceeds from a sale or other disposition of our common stock made after December 31, 2018. Under certain circumstances, a non-U.S. holder may be eligible for refunds or credits of this withholding tax. An intergovernmental agreement between the United States and an applicable foreign country may modify the requirements described in this paragraph. Non-U.S. holders should consult their tax advisors regarding the possible implications of this legislation on their investment in our common stock and the entities through which they hold our common stock, including, without limitation, the process and deadlines for meeting the applicable requirements to prevent the imposition of the 30% withholding tax under FATCA.

UNDERWRITING

Subject to the terms and conditions set forth in the underwriting agreement, dated _____, 2018, by and among us and Jefferies LLC, Cowen and Company, LLC and BMO Capital Markets Corp., as the representatives of the underwriters named below, we have agreed to sell to the underwriters, and each of the underwriters has agreed, severally and not jointly, to purchase from us, the respective number of shares of common stock shown opposite its name below:

<u>Underwriter</u>	<u>Number of Shares</u>
Jefferies LLC	
Cowen and Company, LLC	
BMO Capital Markets Corp.	
Wedbush Securities Inc.	
Total	<u>5,360,000</u>

The underwriting agreement provides that the obligations of the several underwriters are subject to certain conditions precedent such as the receipt by the underwriters of officers' certificates and legal opinions and approval of certain legal matters by their counsel. The underwriting agreement provides that the underwriters will purchase all of the shares of common stock if any of them are purchased. If an underwriter defaults, the underwriting agreement provides that the purchase commitments of the nondefaulting underwriters may be increased or the underwriting agreement may be terminated. We have agreed to indemnify the underwriters and certain of their controlling persons against certain liabilities, including liabilities under the Securities Act, and to contribute to payments that the underwriters may be required to make in respect of those liabilities.

The underwriters have advised us that, following the completion of this offering, they currently intend to make a market in the common stock as permitted by applicable laws and regulations. However, the underwriters are not obligated to do so, and the underwriters may discontinue any market-making activities at any time without notice in their sole discretion. Accordingly, no assurance can be given as to the liquidity of the trading market for the common stock, that you will be able to sell any of the common stock held by you at a particular time or that the prices that you receive when you sell will be favorable.

The underwriters are offering the shares of common stock subject to their acceptance of the shares of common stock from us and subject to prior sale. The underwriters reserve the right to withdraw, cancel or modify offers to the public and to reject orders in whole or in part.

Commission and Expenses

The underwriters have advised us that they propose to offer the shares of common stock to the public at the initial public offering price set forth on the cover page of this prospectus and to certain dealers, which may include the underwriters, at that price less a concession not in excess of \$ _____ per share of common stock. After the offering, the initial public offering price and concession to dealers may be reduced by the representatives. No such reduction will change the amount of proceeds to be received by us as set forth on the cover page of this prospectus.

The following table shows the public offering price, the underwriting discounts and commissions that we are to pay the underwriters and the proceeds, before expenses, to us in connection with this offering. Such amounts are shown assuming both no exercise and full exercise of the underwriters' option to purchase additional shares.

	Per Share		Total	
	Without Option to Purchase Additional Shares	With Option to Purchase Additional Shares	Without Option to Purchase Additional Shares	With Option to Purchase Additional Shares
Public offering price	\$	\$	\$	\$
Underwriting discounts and commissions paid by us	\$	\$	\$	\$
Proceeds to us, before expenses	\$	\$	\$	\$

We estimate expenses payable by us in connection with this offering, other than the underwriting discounts and commissions referred to above, will be approximately \$2.3 million. We have also agreed to reimburse the underwriters for certain of their expenses in an amount up to \$40,000.

Determination of Offering Price

Prior to this offering, there has not been a public market for our common stock. Consequently, the initial public offering price for our common stock will be determined by negotiations between us and the representatives. Among the factors to be considered in these negotiations will be prevailing market conditions, our financial information, market valuations of other companies that we and the underwriters believe to be comparable to us, estimates of our business potential, the present state of our development and other factors deemed relevant.

We offer no assurances that the initial public offering price will correspond to the price at which the common stock will trade in the public market subsequent to the offering or that an active trading market for the common stock will develop and continue after the offering.

Listing

We have applied to have our common stock approved for listing on The Nasdaq Global Market under the trading symbol "SRRK."

Stamp Taxes

If you purchase shares of common stock offered in this prospectus, you may be required to pay stamp taxes and other charges under the laws and practices of the country of purchase, in addition to the offering price listed on the cover page of this prospectus.

Option to Purchase Additional Shares

We have granted to the underwriters an option, exercisable for 30 days from the date of this prospectus, to purchase, from time to time, in whole or in part, up to an aggregate of 804,000 shares from us at the public offering price set forth on the cover page of this prospectus, less underwriting discounts and commissions. If the underwriters exercise this option, each underwriter will be obligated, subject to specified conditions, to purchase a number of additional shares proportionate to that underwriter's initial purchase commitment as indicated in the table above. This option may be exercised only if the underwriters sell more shares than the total number set forth on the cover page of this prospectus.

No Sales of Similar Securities

We, our officers, directors and holders of all or substantially all our outstanding capital stock and other securities have agreed, subject to specified exceptions, not to directly or indirectly:

- § sell, offer, contract or grant any option to sell (including any short sale), pledge, transfer, establish an open "put equivalent position" within the meaning of Rule 16a-1(h) under the Securities Exchange Act of 1934, as amended, or
- § otherwise dispose of any shares of common stock, options or warrants to acquire shares of common stock, or securities exchangeable or exercisable for or convertible into shares of common stock currently or hereafter owned either of record or beneficially, or
- § publicly announce an intention to do any of the foregoing for a period of 180 days after the date of this prospectus without the prior written consent of Jefferies LLC and Cowen and Company, LLC

This restriction terminates after the close of trading of the common stock on and including the 180th day after the date of this prospectus.

Jefferies LLC and Cowen and Company, LLC may, in their sole discretion and at any time or from time to time before the termination of the 180-day period release all or any portion of the securities subject to lock-up agreements. There are no existing agreements between the underwriters and any of our shareholders who will execute a lock-up agreement, providing consent to the sale of shares prior to the expiration of the lock-up period.

Stabilization

The underwriters have advised us that they, pursuant to Regulation M under the Securities Exchange Act of 1934, as amended, certain persons participating in the offering may engage in short sale transactions, stabilizing transactions, syndicate covering transactions or the imposition of penalty bids in connection with this offering. These activities may have the effect of stabilizing or maintaining the market price of the common stock at a level above that which might otherwise prevail in the open market. Establishing short sales positions may involve either "covered" short sales or "naked" short sales.

"Covered" short sales are sales made in an amount not greater than the underwriters' option to purchase additional shares of our common stock in this offering. The underwriters may close out any covered short position by either exercising their option to purchase additional shares of our common stock or purchasing shares of our common stock in the open market. In determining the source of shares to close out the covered short position, the underwriters will consider, among other things, the price of shares available for purchase in the open market as compared to the price at which they may purchase shares through the option to purchase additional shares.

"Naked" short sales are sales in excess of the option to purchase additional shares of our common stock. The underwriters must close out any naked short position by purchasing shares in the open market. A naked short position is more likely to be created if the underwriters are concerned that there may be downward pressure on the price of the shares of our common stock in the open market after pricing that could adversely affect investors who purchase in this offering.

A stabilizing bid is a bid for the purchase of shares of common stock on behalf of the underwriters for the purpose of fixing or maintaining the price of the common stock. A syndicate covering transaction is the bid for or the purchase of shares of common stock on behalf of the underwriters to reduce a short position incurred by the underwriters in connection with the offering. Similar to other purchase transactions, the underwriter's purchases to cover the syndicate short sales may have the effect of raising or maintaining the market price of our common stock or preventing or retarding a decline in the market price of our common stock. As a result, the price of our common stock may be higher than the price that might otherwise exist in the open market. A penalty bid is an arrangement permitting the underwriters to reclaim the selling concession otherwise accruing to a syndicate member in connection with the offering if the common stock

originally sold by such syndicate member are purchased in a syndicate covering transaction and therefore have not been effectively placed by such syndicate member.

Neither we nor any of the underwriters make any representation or prediction as to the direction or magnitude of any effect that the transactions described above may have on the price of our common stock. The underwriters are not obligated to engage in these activities and, if commenced, any of the activities may be discontinued at any time.

The underwriters may also engage in passive market making transactions in our common stock on The Nasdaq Global Market in accordance with Rule 103 of Regulation M during a period before the commencement of offers or sales of shares of our common stock in this offering and extending through the completion of distribution. A passive market maker must display its bid at a price not in excess of the highest independent bid of that security. However, if all independent bids are lowered below the passive market maker's bid, that bid must then be lowered when specified purchase limits are exceeded.

Electronic Distribution

A prospectus in electronic format may be made available by e-mail or on the web sites or through online services maintained by one or more of the underwriters or their affiliates. In those cases, prospective investors may view offering terms online and may be allowed to place orders online. The underwriters may agree with us to allocate a specific number of shares of common stock for sale to online brokerage account holders. Any such allocation for online distributions will be made by the underwriters on the same basis as other allocations. Other than the prospectus in electronic format, the information on the underwriters' web sites and any information contained in any other web site maintained by any of the underwriters is not part of this prospectus, has not been approved and/or endorsed by us or the underwriters and should not be relied upon by investors.

Other Activities and Relationships

The underwriters and certain of their affiliates are full service financial institutions engaged in various activities, which may include securities trading, commercial and investment banking, financial advisory, investment management, investment research, principal investment, hedging, financing and brokerage activities. The underwriters and certain of their affiliates have, from time to time, performed, and may in the future perform, various commercial and investment banking and financial advisory services for us and our affiliates, for which they received or will receive customary fees and expenses.

In the ordinary course of their various business activities, the underwriters and certain of their affiliates may make or hold a broad array of investments and actively trade debt and equity securities (or related derivative securities) and financial instruments (including bank loans) for their own account and for the accounts of their customers, and such investment and securities activities may involve securities and/or instruments issued by us and our affiliates. If the underwriters or their respective affiliates have a lending relationship with us, they routinely hedge their credit exposure to us consistent with their customary risk management policies. The underwriters and their respective affiliates may hedge such exposure by entering into transactions which consist of either the purchase of credit default swaps or the creation of short positions in our securities or the securities of our affiliates, including potentially the common stock offered hereby. Any such short positions could adversely affect future trading prices of the common stock offered hereby. The underwriters and certain of their respective affiliates may also communicate independent investment recommendations, market color or trading ideas and/or publish or express independent research views in respect of such securities or instruments and may at any time hold, or recommend to clients that they acquire, long and/or short positions in such securities and instruments.

Disclaimers About Non-U.S. Jurisdictions

Australia

This prospectus is not a disclosure document for the purposes of Australia's Corporations Act 2001 (Cth) of Australia, or Corporations Act, has not been lodged with the Australian Securities & Investments Commission and is only directed to the categories of exempt persons set out below. Accordingly, if you receive this prospectus in Australia:

You confirm and warrant that you are either:

- § a "sophisticated investor" under section 708(8)(a) or (b) of the Corporations Act;
- § a "sophisticated investor" under section 708(8)(c) or (d) of the Corporations Act and that you have provided an accountant's certificate to the Company which complies with the requirements of section 708(8)(c)(i) or (ii) of the Corporations Act and related regulations before the offer has been made;
- § a person associated with the Company under Section 708(12) of the Corporations Act; or
- § a "professional investor" within the meaning of section 708(11)(a) or (b) of the Corporations Act.

To the extent that you are unable to confirm or warrant that you are an exempt sophisticated investor, associated person or professional investor under the Corporations Act any offer made to you under this prospectus is void and incapable of acceptance.

You warrant and agree that you will not offer any of the securities issued to you pursuant to this prospectus for resale in Australia within 12 months of those securities being issued unless any such resale offer is exempt from the requirement to issue a disclosure document under section 708 of the Corporations Act.

Canada

- (A) **Resale Restrictions.** The distribution of common stock in Canada is being made only in the provinces of Ontario, Quebec, Alberta and British Columbia on a private placement basis exempt from the requirement that we prepare and file a prospectus with the securities regulatory authorities in each province where trades of these securities are made. Any resale of the common stock in Canada must be made under applicable securities laws which may vary depending on the relevant jurisdiction, and which may require resales to be made under available statutory exemptions or under a discretionary exemption granted by the applicable Canadian securities regulatory authority. Purchasers are advised to seek legal advice prior to any resale of the securities.
- (B) **Representations of Canadian Purchasers.** By purchasing common stock in Canada and accepting delivery of a purchase confirmation, a purchaser is representing to us and the dealer from whom the purchase confirmation is received that:
 - § the purchaser is entitled under applicable provincial securities laws to purchase the common stock without the benefit of a prospectus qualified under those securities laws as it is an "accredited investor" as defined under National Instrument 45-106 — *Prospectus Exemptions*,
 - § the purchaser is a "permitted client" as defined in National Instrument 31-103 — *Registration Requirements, Exemptions and Ongoing Registrant Obligations*,
 - § where required by law, the purchaser is purchasing as principal and not as agent, and
 - § the purchaser has reviewed the text above under Resale Restrictions.
- (C) **Conflicts of Interest.** Canadian purchasers are hereby notified that the representatives are relying on the exemption set out in section 3A.3 or 3A.4, if applicable, of National Instrument 33-105 — *Underwriting Conflicts* from having to provide certain conflict of interest disclosure in this document.
- (D) **Statutory Rights of Action.** Securities legislation in certain provinces or territories of Canada may provide a purchaser with remedies for rescission or damages if the offering memorandum (including any amendment thereto) such as this document contains a misrepresentation, provided that the remedies

for rescission or damages are exercised by the purchaser within the time limit prescribed by the securities legislation of the purchaser's province or territory. The purchaser of these securities in Canada should refer to any applicable provisions of the securities legislation of the purchaser's province or territory for particulars of these rights or consult with a legal advisor.

- (E) **Enforcement of Legal Rights.** All of our directors and officers as well as the experts named herein may be located outside of Canada and, as a result, it may not be possible for Canadian purchasers to effect service of process within Canada upon us or those persons. All or a substantial portion of our assets and the assets of those persons may be located outside of Canada and, as a result, it may not be possible to satisfy a judgment against us or those persons in Canada or to enforce a judgment obtained in Canadian courts against us or those persons outside of Canada.
- (F) **Taxation and Eligibility for Investment.** Canadian purchasers of common stock should consult their own legal and tax advisors with respect to the tax consequences of an investment in the common stock in their particular circumstances and about the eligibility of the common stock for investment by the purchaser under relevant Canadian legislation.

European Economic Area

Any distributor subject to MiFID II that is offering, selling or recommending the common stock is responsible for undertaking its own target market assessment in respect of the common stock and determining its own distribution channels for the purposes of the MiFID product governance rules under Commission Delegated Directive (EU) 2017/593, or the Delegated Directive. Neither we nor the underwriters make any representations or warranties as to a distributor's compliance with the Delegated Directive.

In relation to each member state of the European Economic Area which has implemented the Prospectus Directive, or a Relevant Member State, an offer to the public of any shares of common stock which are the subject of the offering contemplated by this prospectus may not be made in that Relevant Member State except that an offer to the public in that Relevant Member State of any shares of common stock may be made at any time under the following exemptions under the Prospectus Directive, if they have been implemented in that Relevant Member State:

- § to any legal entity which is a "qualified investor" as defined in the Prospectus Directive;
- § to fewer than 100 or, if the Relevant Member State has implemented the relevant provision of the 2010 PD Amending Directive, 150, natural or legal persons (other than qualified investors as defined in the Prospectus Directive), as permitted under the Prospectus Directive, subject to obtaining the prior consent of the underwriters or the underwriters nominated by us for any such offer; or
- § in any other circumstances falling within Article 3(2) of the Prospectus Directive,

provided that no such offer of shares of common stock shall require us or any of the underwriters to publish a prospectus pursuant to Article 3 of the Prospectus Directive or supplement a prospectus pursuant to Article 16 of the Prospectus Directive.

For the purposes of this provision, the expression an "offer shares of common stock to the public" in relation to the shares of common stock in any Relevant Member State means the communication in any form and by any means of sufficient information on the terms of the offer and the shares of common stock to be offered so as to enable an investor to decide to purchase or subscribe to the shares of common stock, as the same may be varied in that Relevant Member State by any measure implementing the Prospectus Directive in that Relevant Member State and the expression "Prospectus Directive" means Directive 2003/71/EC (and amendments thereto, including the 2010 PD Amending Directive, to the extent implemented in the Relevant Member State), and includes any relevant implementing measure in the Relevant Member State and the expression "2010 PD Amending Directive" means Directive 2010/73/EU.

Hong Kong

No securities have been offered or sold, and no securities may be offered or sold, in Hong Kong, by means of any document, other than to persons whose ordinary business is to buy or sell shares or debentures, whether as principal or agent; or to "professional investors" as defined in the Securities and Futures Ordinance (Cap. 571) of Hong Kong, or SFO, and any rules made under that Ordinance; or in other circumstances which do not result in the document being a "prospectus" as defined in the Companies Ordinance (Cap. 32) of Hong Kong, or CO, or which do not constitute an offer or invitation to the public for the purpose of the CO or the SFO. No document, invitation or advertisement relating to the securities has been issued or may be issued or may be in the possession of any person for the purpose of issue (in each case whether in Hong Kong or elsewhere), which is directed at, or the contents of which are likely to be accessed or read by, the public of Hong Kong (except if permitted under the securities laws of Hong Kong) other than with respect to securities which are or are intended to be disposed of only to persons outside Hong Kong or only to "professional investors" as defined in the SFO and any rules made under that Ordinance.

This prospectus has not been registered with the Registrar of Companies in Hong Kong. Accordingly, this prospectus may not be issued, circulated or distributed in Hong Kong, and the securities may not be offered for subscription to members of the public in Hong Kong. Each person acquiring the securities will be required, and is deemed by the acquisition of the securities, to confirm that he is aware of the restriction on offers of the securities described in this prospectus and the relevant offering documents and that he is not acquiring, and has not been offered any securities in circumstances that contravene any such restrictions.

Israel

This document does not constitute a prospectus under the Israeli Securities Law, 5728-1968, or the Securities Law, and has not been filed with or approved by the Israel Securities Authority. In Israel, this prospectus is being distributed only to, and is directed only at, and any offer of the common stock is directed only at, (i) a limited number of persons in accordance with the Israeli Securities Law and (ii) investors listed in the first addendum, or the Addendum, to the Israeli Securities Law, consisting primarily of joint investment in trust funds, provident funds, insurance companies, banks, portfolio managers, investment advisors, members of the Tel Aviv Stock Exchange, underwriters, venture capital funds, entities with equity in excess of NIS 50 million and "qualified individuals," each as defined in the Addendum (as it may be amended from time to time), collectively referred to as qualified investors (in each case, purchasing for their own account or, where permitted under the Addendum, for the accounts of their clients who are investors listed in the Addendum). Qualified investors are required to submit written confirmation that they fall within the scope of the Addendum, are aware of the meaning of same and agree to it.

Japan

The offering has not been and will not be registered under the Financial Instruments and Exchange Law of Japan (Law No. 25 of 1948 of Japan, as amended), or FIEL, and the underwriters will not offer or sell any securities, directly or indirectly, in Japan or to, or for the benefit of, any resident of Japan (which term as used herein means any person resident in Japan, including any corporation or other entity organized under the laws of Japan), or to others for re-offering or resale, directly or indirectly, in Japan or to, or for the benefit of, any resident of Japan, except pursuant to an exemption from the registration requirements of, and otherwise in compliance with, the FIEL and any other applicable laws, regulations and ministerial guidelines of Japan.

Singapore

This prospectus has not been and will not be lodged or registered as a prospectus with the Monetary Authority of Singapore. Accordingly, this prospectus and any other document or material in connection with the offer or sale, or invitation for subscription or purchase, of the common stock may not be circulated or distributed, nor may the common stock be offered or sold, or be made the subject of an invitation for

subscription or purchase, whether directly or indirectly, to persons in Singapore other than (i) to an institutional investor under Section 274 of the Securities and Futures Act, Chapter 289 of Singapore, or SFA, (ii) to a relevant person pursuant to Section 275(1), or any person pursuant to Section 275(1A), and in accordance with the conditions specified in Section 275, of the SFA, or (iii) otherwise pursuant to, and in accordance with the conditions of, any other applicable provision of the SFA.

Where the securities are subscribed or purchased under Section 275 of the SFA by a relevant person which is:

- § a corporation (which is not an accredited investor (as defined in Section 4A of the SFA)) the sole business of which is to hold investments and the entire share capital of which is owned by one or more individuals, each of whom is an accredited investor; or
- § a trust (where the trustee is not an accredited investor) whose sole purpose is to hold investments and each beneficiary of the trust is an individual who is an accredited investor,

securities (as defined in Section 239(1) of the SFA) of that corporation or the beneficiaries' rights and interest (howsoever described) in that trust shall not be transferred within six months after that corporation or that trust has acquired the securities pursuant to an offer made under Section 275 of the SFA except:

- § to an institutional investor or to a relevant person defined in Section 275(2) of the SFA, or to any person arising from an offer referred to in Section 275(1A) or Section 276(4)(i)(B) of the SFA;
- § where no consideration is or will be given for the transfer;
- § where the transfer is by operation of law;
- § as specified in Section 276(7) of the SFA; or
- § as specified in Regulation 32 of the Securities and Futures (Offers of Investments) (Shares and Debentures) Regulations 2005 of Singapore.

Switzerland

The securities may not be publicly offered in Switzerland and will not be listed on the SIX Swiss Exchange, or SIX, or on any other stock exchange or regulated trading facility in Switzerland. This prospectus has been prepared without regard to the disclosure standards for issuance prospectuses under art. 652a or art. 1156 of the Swiss Code of Obligations or the disclosure standards for listing prospectuses under art. 27 ff. of the SIX Listing Rules or the listing rules of any other stock exchange or regulated trading facility in Switzerland. Neither this prospectus nor any other offering or marketing material relating to the securities or the offering may be publicly distributed or otherwise made publicly available in Switzerland.

Neither this prospectus nor any other offering or marketing material relating to the offering, the Company or the securities have been or will be filed with or approved by any Swiss regulatory authority. In particular, this prospectus will not be filed with, and the offer of securities will not be supervised by, the Swiss Financial Market Supervisory Authority FINMA, and the offer of securities has not been and will not be authorized under the Swiss Federal Act on Collective Investment Schemes, or CISA. The investor protection afforded to acquirers of interests in collective investment schemes under the CISA does not extend to acquirers of securities.

United Kingdom

This prospectus is only being distributed to, and is only directed at, persons in the United Kingdom that are qualified investors within the meaning of Article 2(1)(e) of the Prospectus Directive that are also (i) investment professionals falling within Article 19(5) of the Financial Services and Markets Act 2000 (Financial Promotion) Order 2005, as amended, or Order, and/or (ii) high net worth entities falling within Article 49(2)(a) to (d) of the Order and other persons to whom it may lawfully be communicated, or a Relevant Person.

This prospectus and its contents are confidential and should not be distributed, published or reproduced (in whole or in part) or disclosed by recipients to any other persons in the United Kingdom. Any person in the United Kingdom that is not a Relevant Person should not act or rely on this document or any of its content.

LEGAL MATTERS

The validity of the shares of common stock offered by this prospectus will be passed upon for us by Goodwin Procter LLP, Boston, Massachusetts. Certain legal matters related to this offering will be passed upon for the underwriters by Cooley LLP, Boston, Massachusetts.

EXPERTS

The consolidated financial statements of Scholar Rock Holding Corporation at December 31, 2016 and 2017, and for the years then ended, appearing in this prospectus and registration statement have been audited by Ernst & Young LLP, independent registered accounting firm, as set forth in their report thereon (which contains an explanatory paragraph describing conditions that raise substantial doubt about our ability to continue as a going concern as described in Note 1 to the consolidated financial statements) appearing elsewhere herein and are included in reliance upon such report given on the authority of such firm as experts in accounting and auditing.

WHERE YOU CAN FIND MORE INFORMATION

We have filed with the SEC a registration statement on Form S-1 (File Number 333-224493) under the Securities Act with respect to the common stock we are offering by this prospectus. This prospectus does not contain all of the information included in the registration statement. For further information pertaining to us and our common stock, you should refer to the registration statement and to its exhibits. Whenever we make reference in this prospectus to any of our contracts, agreements or other documents, the references are not necessarily complete, and you should refer to the exhibits attached to the registration statement for copies of the actual contract, agreement or other document.

Upon the completion of the offering, we will be subject to the informational requirements of the Exchange Act and will file annual, quarterly and current reports, proxy statements and other information with the SEC. You can read our SEC filings, including the registration statement, at the SEC's website at www.sec.gov. You may also read and copy any document we file with the SEC at its public reference facility at 100 F Street, N.E., Room 1580, Washington, D.C. 20549. We also maintain a website at <http://www.scholarrock.com>. Upon completion of the offering, you may access, free of charge, our annual reports on Form 10-K, quarterly reports on Form 10-Q, current reports on Form 8-K and amendment to those reports filed or furnished pursuant to Section 13(a) or 15(d) of the Exchange Act as soon as reasonably practicable after such material is electronically filed with, or furnished to, the SEC.

You may also obtain copies of the documents at prescribed rates by writing to the Public Reference Section of the SEC at 100 F Street, N.E., Washington, D.C. 20549. Please call the SEC at 1-800-SEC-0330 for further information on the operation of the public reference facilities.

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REPORT OF INDEPENDENT REGISTERED PUBLIC ACCOUNTING FIRM

To the Stockholders and the Board of Directors of Scholar Rock Holding Corporation

Opinion on the Financial Statements

We have audited the accompanying consolidated balance sheets of Scholar Rock Holding Corporation (the Company) as of December 31, 2017 and 2016, the related consolidated statements of operations and comprehensive loss, convertible preferred stock and stockholders' equity (deficit), and cash flows for the years then ended, and the related notes (collectively referred to as the "consolidated financial statements"). In our opinion, the consolidated financial statements present fairly, in all material respects, the financial position of the Company at December 31, 2017 and 2016, and the results of its operations and its cash flows for the years then ended in conformity with U.S. generally accepted accounting principles.

The Company's Ability to Continue as a Going Concern

The accompanying consolidated financial statements have been prepared assuming that the Company will continue as a going concern. As discussed in Note 1 to the consolidated financial statements, the Company has suffered recurring losses from operations, will require substantial additional capital to fund operations, and has stated that substantial doubt exists about the Company's ability to continue as a going concern. Management's evaluation of the events and conditions and management's plans regarding these matters are also described in Note 1. The consolidated financial statements do not include any adjustments that might result from the outcome of this uncertainty.

Adoption of New Accounting Standards

As discussed in Note 1 to the consolidated financial statements, the Company changed its method for accounting for revenue from contracts with customers in 2017 and 2016.

Basis for Opinion

These financial statements are the responsibility of the Company's management. Our responsibility is to express an opinion on the Company's financial statements based on our audits. We are a public accounting firm registered with the Public Company Accounting Oversight Board (United States) (PCAOB) and are required to be independent with respect to the Company in accordance with the U.S. federal securities laws and the applicable rules and regulations of the Securities and Exchange Commission and the PCAOB.

We conducted our audits in accordance with the standards of the PCAOB. Those standards require that we plan and perform the audit to obtain reasonable assurance about whether the financial statements are free of material misstatement, whether due to error or fraud. The Company is not required to have, nor were we engaged to perform, an audit of its internal control over financial reporting. As part of our audits we are required to obtain an understanding of internal control over financial reporting but not for the purpose of expressing an opinion on the effectiveness of the Company's internal control over financial reporting. Accordingly, we express no such opinion.

Our audits included performing procedures to assess the risks of material misstatement of the financial statements, whether due to error or fraud, and performing procedures that respond to those risks. Such procedures included examining, on a test basis, evidence regarding the amounts and disclosures in the financial statements. Our audits also included evaluating the accounting principles used and significant estimates made by management, as well as evaluating the overall presentation of the financial statements. We believe that our audits provide a reasonable basis for our opinion.

/s/ Ernst & Young LLP

We have served as the Company's auditor since 2015.
Boston, Massachusetts
May 8, 2018,
except for Note 22, as to which the date is May 14, 2018

SCHOLAR ROCK HOLDING CORPORATION
CONSOLIDATED BALANCE SHEETS
(In thousands, except unit, share and per share data)

	<u>December 31,</u>		<u>March 31,</u>	<u>Pro Forma</u>
	<u>2016</u>	<u>2017</u>	<u>2018</u>	<u>March 31,</u>
			(unaudited)	2018
				(unaudited)
Assets				
Current assets:				
Cash and cash equivalents	\$ 10,033	\$ 56,461	\$ 22,822	\$ 22,822
Marketable securities	18,002	1,498	24,941	24,941
Prepaid expenses and other current assets	461	1,242	3,173	3,173
Total current assets	28,496	59,201	50,936	50,936
Marketable securities	1,496	—	—	—
Property and equipment, net	2,430	2,181	2,033	2,033
Restricted cash	205	205	205	205
Deferred offering costs	—	—	985	985
Other long-term assets	155	50	—	—
Total assets	<u>\$ 32,782</u>	<u>\$ 61,637</u>	<u>\$ 54,159</u>	<u>\$ 54,159</u>
Liabilities, Convertible Preferred Stock and Stockholders' Equity (Deficit)				
Current liabilities:				
Accounts payable	\$ 663	\$ 1,359	\$ 3,254	\$ 3,254
Accrued expenses	1,487	2,796	1,998	1,998
Deferred rent	209	228	—	—
Loan payable	647	641	641	641
Total current liabilities	3,006	5,024	5,893	5,893
Long-term portion of deferred rent	696	468	664	664
Long-term portion of loan payable	1,023	398	244	244
Warrant to purchase redeemable security	27	37	57	—
Total liabilities	<u>4,752</u>	<u>5,927</u>	<u>6,858</u>	<u>6,801</u>
Commitments and contingencies (Note 14)				
Convertible preferred units (Series A-1, A-2, A-3, A-4 and B), 30,102,095 units authorized and 28,652,147 units issued and outstanding at December 31, 2016; (aggregate liquidation preference of \$58,277 at December 31, 2016); no units authorized, issued or outstanding as of December 31, 2017, March 31, 2018 (unaudited) or pro forma as of March 31, 2018 (unaudited)				
	58,057	—	—	—
Convertible preferred stock (Series A-1, A-2, A-3, A-4, B and C), \$0.001 par value; no shares authorized, issued or outstanding as of December 31, 2016; 43,157,651 shares authorized and 43,135,911 shares issued and outstanding as of December 31, 2017 and March 31, 2018 (unaudited); (aggregate liquidation preference of \$109,561 as of December 31, 2017 and March 31, 2018 (unaudited)); no shares issued or outstanding, pro forma as of March 31, 2018 (unaudited)				
	—	109,232	109,232	—
Stockholders' equity (deficit):				
Common units, 21,130,140 units authorized and 1,603,088 units issued and outstanding at December 31, 2016; no units authorized, issued or outstanding as of December 31, 2017, March 31, 2018 (unaudited) or pro forma as of March 31, 2018 (unaudited)				
	1,471	—	—	—
Common stock, \$0.001 par value; no shares authorized, issued or outstanding as of December 31, 2016; 60,000,000 shares authorized and 3,970,586 shares issued and outstanding as of December 31, 2017 and March 31, 2018 (unaudited); 19,080,536 shares issued and outstanding, pro forma as of March 31, 2018 (unaudited)				
	—	4	4	19
Additional paid-in capital	1,052	4,001	4,483	113,799
Accumulated other comprehensive loss	(20)	(2)	(1)	(1)
Accumulated deficit	(32,530)	(57,525)	(66,417)	(66,459)
Total stockholders' equity (deficit)	<u>(30,027)</u>	<u>(53,522)</u>	<u>(61,931)</u>	<u>47,358</u>
Total liabilities, convertible preferred stock and stockholders' equity (deficit)	<u>\$ 32,782</u>	<u>\$ 61,637</u>	<u>\$ 54,159</u>	<u>\$ 54,159</u>

The accompanying notes are an integral part of these consolidated financial statements.

SCHOLAR ROCK HOLDING CORPORATION
CONSOLIDATED STATEMENTS OF OPERATIONS AND COMPREHENSIVE LOSS
(In thousands, except unit, share, per unit and per share data)

	Year Ended December 31,		Three Months Ended March 31,	
	2016	2017	2017 (unaudited)	2018
Collaboration revenue	\$ 379	\$ —	\$ —	\$ —
Operating expenses:				
Research and development	12,477	19,944	3,568	6,701
General and administrative	4,112	5,085	1,173	2,315
Total operating expenses	16,589	25,029	4,741	9,016
Loss from operations	(16,210)	(25,029)	(4,741)	(9,016)
Other income (expense):				
Interest income (expense), net	(19)	44	13	144
Other income (expense), net	22	(10)	(2)	(20)
Total other income	3	34	11	124
Net loss	\$ (16,207)	\$ (24,995)	\$ (4,730)	\$ (8,892)
Net loss per unit, basic and diluted	\$ (10.11)		\$ (2.95)	
Net loss per share, basic and diluted		\$ (15.30)		\$ (3.18)
Weighted average common units outstanding, basic and diluted	1,603,088		1,603,088	
Weighted average common shares outstanding, basic and diluted		1,634,100		2,795,497
Pro forma net loss per share, basic and diluted (unaudited)		\$ (2.06)		\$ (0.50)
Pro forma weighted average common shares outstanding, basic and diluted (unaudited)		12,108,377		17,905,447
Comprehensive loss:				
Net loss	\$ (16,207)	\$ (24,995)	\$ (4,730)	\$ (8,892)
Other comprehensive income (loss):				
Unrealized gain (loss) on marketable securities	(20)	18	(3)	1
Total other comprehensive income (loss)	(20)	18	(3)	1
Comprehensive loss	\$ (16,227)	\$ (24,977)	\$ (4,733)	\$ (8,891)

The accompanying notes are an integral part of these consolidated financial statements.

SCHOLAR ROCK HOLDING CORPORATION

CONSOLIDATED STATEMENTS OF CONVERTIBLE PREFERRED STOCK AND STOCKHOLDERS' EQUITY (DEFICIT)

(In thousands, except unit and share data)

	Convertible Preferred Units		Convertible Preferred Stock		Common Units		Common Stock		Additional Paid-in Capital	Accumulated Other Comprehensive Loss	Accumulated Deficit	Total Stockholders' Equity (Deficit)
	Units	Amount	Shares	Amount	Units	Amount	Shares	Amount				
Balance at December 31, 2015	28,652,147	\$ 58,057	—	\$ —	1,603,088	\$ 1,471	—	\$ —	\$ 496	\$ —	\$ (16,323)	\$ (14,356)
Unrealized loss on marketable securities	—	—	—	—	—	—	—	—	—	(20)	—	(20)
Equity-based compensation expense	—	—	—	—	—	—	—	—	556	—	—	556
Net loss	—	—	—	—	—	—	—	—	—	—	(16,207)	(16,207)
Balance at December 31, 2016	28,652,147	58,057	—	—	1,603,088	1,471	—	—	1,052	(20)	(32,530)	(30,027)
Unrealized gain on marketable securities	—	—	—	—	—	—	—	—	—	18	—	18
Issuance of Series B convertible preferred units	1,428,209	4,285	—	—	—	—	—	—	—	—	—	—
Effect of Reorganization	(30,080,356)	(62,342)	30,080,356	62,342	(1,603,088)	(1,471)	1,603,088	2	1,469	—	—	—
Issuance of common stock and restricted common stock as part of Reorganization	—	—	—	—	—	—	2,367,498	2	(2)	—	—	—
Issuance of Series C convertible preferred stock, net of issuance costs of \$109	—	—	13,055,555	46,890	—	—	—	—	—	—	—	—
Equity-based compensation expense	—	—	—	—	—	—	—	—	1,482	—	—	1,482
Net loss	—	—	—	—	—	—	—	—	—	—	(24,995)	(24,995)
Balance at December 31, 2017	—	—	43,135,911	109,232	—	—	3,970,586	4	4,001	(2)	(57,525)	(53,522)
Unrealized gain on marketable securities (unaudited)	—	—	—	—	—	—	—	—	—	1	—	1
Equity-based compensation expense (unaudited)	—	—	—	—	—	—	—	—	482	—	—	482
Net Loss (unaudited)	—	—	—	—	—	—	—	—	—	—	(8,892)	(8,892)
Balance at March 31, 2018 (unaudited)	—	—	43,135,911	109,232	—	—	3,970,586	4	4,483	(1)	(66,417)	(61,931)
Conversion of convertible preferred stock into common stock (unaudited)	—	—	(43,135,911)	(109,232)	—	—	15,109,950	15	109,217	—	—	109,232
Reclassification of warrant to purchase convertible preferred stock to stockholders' equity (deficit) (unaudited)	—	—	—	—	—	—	—	—	57	—	—	57
Equity-based compensation expense for stock-based award with vesting conditions contingent upon initial public offering (unaudited)	—	—	—	—	—	—	—	—	42	—	(42)	—
Pro forma balance at March 31, 2018 (unaudited)	—	\$ —	—	\$ —	—	\$ —	19,080,536	\$ 19	\$ 113,799	\$ (1)	\$ (66,459)	\$ 47,358

The accompanying notes are an integral part of these consolidated financial statements.

SCHOLAR ROCK HOLDING CORPORATION
CONSOLIDATED STATEMENTS OF CASH FLOWS
(In thousands)

	Year Ended December 31,		Three Months Ended March 31,	
	2016	2017	2017	2018
	(unaudited)			
Cash flows from operating activities:				
Net loss	\$ (16,207)	\$ (24,995)	\$ (4,730)	\$ (8,892)
Adjustments to reconcile net loss to net cash used in operating activities:				
Depreciation and amortization	575	669	164	179
Equity-based compensation	556	1,482	257	482
Amortization of deferred rent	(184)	(209)	(51)	(32)
Deferred payroll tax credit	(250)	(199)	—	42
Other	22	47	19	2
Change in operating assets and liabilities:				
Accounts receivable	380	—	—	—
Prepaid expenses and other current assets	(141)	(477)	(265)	(1,923)
Accounts payable	91	636	523	1,529
Accrued expenses	17	1,309	(492)	(1,271)
Net cash used in operating activities	<u>(15,141)</u>	<u>(21,737)</u>	<u>(4,575)</u>	<u>(9,884)</u>
Cash flows from investing activities:				
Purchase of property and equipment	(794)	(361)	(132)	(176)
Purchases of marketable securities	(19,525)	—	—	(23,412)
Maturities of marketable securities	—	18,026	1,500	—
Net cash (used in) provided by investing activities	<u>(20,319)</u>	<u>17,665</u>	<u>1,368</u>	<u>(23,588)</u>
Cash flows from financing activities:				
Proceeds from loan payable and warrants, net of issuance costs	1,318	—	—	—
Principal payments on loan payable	(302)	(667)	(167)	(167)
Proceeds from issuance of Series B convertible preferred units	—	4,285	—	—
Proceeds from issuance of Series C convertible preferred shares, net of issuance costs	—	46,890	—	—
Principal payments on capital lease obligation and other	(14)	(8)	—	—
Net cash (used in) provided by financing activities	<u>1,002</u>	<u>50,500</u>	<u>(167)</u>	<u>(167)</u>
Net (decrease) increase in cash and cash equivalents and restricted cash	<u>(34,458)</u>	<u>46,428</u>	<u>(3,374)</u>	<u>(33,639)</u>
Cash and cash equivalents and restricted cash, beginning of period	44,696	10,238	10,238	56,666
Cash and cash equivalents and restricted cash, end of period	<u>\$ 10,238</u>	<u>\$ 56,666</u>	<u>\$ 6,864</u>	<u>\$ 23,027</u>
Supplemental disclosure of non-cash items:				
Property and equipment purchases in accounts payable	\$ 98	\$ 159	\$ 64	\$ 13
Deferred offering costs in accounts payable and accrued expenses	\$ —	\$ —	\$ —	\$ 985
Supplemental cash flow information:				
Cash paid for interest	<u>\$ 42</u>	<u>\$ 53</u>	<u>\$ 14</u>	<u>\$ 10</u>

The accompanying notes are an integral part of these consolidated financial statements.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

1. Nature of the Business and Basis of Presentation

Organization

Scholar Rock, LLC was formed on May 22, 2012, as a Delaware limited liability company to discover and develop a new class of biologics.

On December 22, 2017, a series of transactions were completed pursuant to which Scholar Rock Merger Sub, LLC, a wholly owned subsidiary of Scholar Rock Holding Corporation, a Delaware corporation, was merged with and into Scholar Rock, LLC (the "Reorganization"). As part of the Reorganization, each issued and outstanding convertible preferred and common unit of Scholar Rock, LLC outstanding immediately prior to the Reorganization was exchanged for shares of Scholar Rock Holding Corporation capital stock of the same class and/or series on a one-for-one basis. Previously outstanding vested and unvested incentive units, irrespective of any strike price or voting rights, were exchanged for an equal number of shares of common stock or restricted common stock, respectively. The restricted common stock was issued with the same vesting terms as the unvested incentive units held immediately prior to the Reorganization.

Upon consummation of the Reorganization, the historical consolidated financial statements of Scholar Rock, LLC became the historical consolidated financial statements of Scholar Rock Holding Corporation, the entity whose shares are being offered in this offering. For purposes of these consolidated financial statements, "the Company" refers to Scholar Rock, LLC prior to the Reorganization and Scholar Rock Holding Corporation after the Reorganization. The Company is based in Cambridge, Massachusetts.

The Company is a biopharmaceutical company focused on the discovery and development of innovative medicines for the treatment of serious diseases in which signaling by protein growth factors plays a fundamental role. Since its inception, the Company's operations have focused on organizing and staffing, business planning, raising capital, establishing the Company's intellectual property portfolio and performing research and development of monoclonal antibodies that selectively inhibit activation of growth factors for therapeutic effect. Revenue generation activities have been limited to research services and the issuance of a license, in each case, pursuant to an option and license agreement with Janssen Biotech, Inc. ("Janssen"), a subsidiary of Johnson & Johnson. No revenues have been recorded from the sale of any commercial product.

Basis of Presentation

The consolidated financial statements prior to the Reorganization include the accounts of Scholar Rock, LLC and its wholly owned subsidiary, Scholar Rock, Inc. The consolidated financial statements subsequent to the Reorganization include the accounts of Scholar Rock Holding Corporation and its wholly owned subsidiaries. All intercompany balances have been eliminated in consolidation.

These consolidated financial statements have been prepared in conformity with accounting principles generally accepted in the United States ("GAAP"). Any reference in these notes to applicable guidance is meant to refer to the authoritative United States generally accepted accounting principles as found in the Accounting Standards Codification ("ASC") and Accounting Standards Update ("ASU") of the Financial Accounting Standards Board ("FASB").

Effective January 1, 2018, the Company adopted the requirements of ASC Topic 606, *Revenue from Contracts with Customers* ("ASC 606") using the full retrospective method as discussed below in "Note 2. Summary of Significant Accounting Policies." All amounts and disclosures set forth in these financial

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

1. Nature of the Business and Basis of Presentation (Continued)

statements have been revised for all periods presented to reflect this change. There was no impact to the Company's balance sheet, statement of operations, stockholders' equity (deficit) or cash flows as a result of adopting this standard.

Effective January 1, 2018, the Company adopted the requirements of ASU 2016-18, *Restricted Cash*, using a retrospective method as discussed below in "Note 2: Summary of Significant Accounting Policies." All amounts and disclosures set forth in these financial statements have been revised for all periods presented to reflect this change. The adoption of this standard did not affect revenue, net loss, or any other financial statement line item in any period.

Going Concern

In accordance with ASU 2014-15, *Disclosure of Uncertainties about an Entity's Ability to Continue as a Going Concern (Subtopic 205-40)*, the Company has evaluated whether there are conditions and events considered in the aggregate that raise substantial doubt about the Company's ability to continue as a going concern.

Since inception, the Company has primarily funded its operations with proceeds from sale of convertible preferred units and convertible preferred stock and a credit facility with Silicon Valley Bank ("SVB"). From inception through March 31, 2018, the Company has raised an aggregate of \$111.2 million, of which \$109.2 million was from the issuance of convertible preferred units and convertible preferred stock and \$2.0 million was from the Company's credit facility. The Company has incurred recurring losses and negative cash flows from operations since inception, including net losses of \$16.2 million, \$25.0 million and \$8.9 million for the years ended December 31, 2016 and 2017 and three months ended March 31, 2018, respectively, and negative cash flows from operating activities of \$15.1 million, \$21.7 million and \$9.9 million for the years ended December 31, 2016 and 2017 and three months ended March 31, 2018, respectively. As of March 31, 2018, the Company had an accumulated deficit of \$66.4 million.

The Company is also subject to a number of risks similar to other life science companies, including, but not limited to, successful discovery and development of its drug candidates, raising additional capital, development by its competitors of new technological innovations, protection of proprietary technology and regulatory approval and market acceptance of the Company's products. The Company anticipates that it will continue to incur significant operating losses for the next several years as it continues to develop its product candidates.

The Company expects its operating losses and negative cash flows to continue into the foreseeable future as it continues to develop, manufacture, and commercialize its products. As of March 31, 2018, the Company had cash, cash equivalents and marketable securities of \$47.8 million. Based on its current operating plan, the Company expects its cash, cash equivalents and marketable securities will be sufficient to fund operating expenses and capital expenditure requirements into the first quarter of 2019. Based on the Company's available cash resources, the Company does not have sufficient cash on hand to support current operations for at least the next twelve months from the date that these financial statements are issued. This condition results in substantial doubt about the Company's ability to continue as a going concern.

The Company is seeking to complete an initial public offering ("IPO") of its common stock. Upon the closing of a qualified public offering on specified terms, the Company's outstanding convertible preferred

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

1. Nature of the Business and Basis of Presentation (Continued)

stock will automatically convert to common stock. In the event the Company does not complete an IPO, the Company expects to seek additional funding through private or public equity financings, debt financings, government funding arrangements, collaborations, strategic alliances and marketing, distribution or licensing agreements. The Company may not be able to obtain funding on acceptable terms, or at all. The terms of any financing may adversely affect the holdings or rights of the Company's shareholders.

If the Company is unable to obtain financing, the Company could be forced to delay, reduce or eliminate some or all of its research and development programs, product portfolio expansion or commercialization efforts, which could adversely affect its business prospects, or the Company may be unable to continue operations. Although management continues to pursue these plans, there is no assurance that the Company will be successful in obtaining sufficient funding on terms acceptable to the Company to fund continuing operations, if at all.

The accompanying consolidated financial statements have been prepared on a going concern basis, which contemplates the realization of assets and satisfaction of liabilities in the ordinary course of business. The consolidated financial statements do not include any adjustments relating to the recoverability and classification of recorded asset amounts or the amounts and classification of liabilities that might result from the outcome of the uncertainties described above.

2. Summary of Significant Accounting Policies

Use of Estimates

The preparation of financial statements in accordance with GAAP requires management to make estimates and judgments that may affect the reported amounts of assets and liabilities and related disclosures of contingent assets and liabilities at the date of the financial statements and the related reporting of revenues and expenses during the reporting period. Significant estimates of accounting reflected in these consolidated financial statements include, but are not limited to, estimates related to accrued expenses, the valuation of equity-based compensation, including incentive units, common stock, restricted common stock and stock options, and income taxes. Actual results could differ from those estimates.

The Company utilizes significant estimates and assumptions in determining the fair value of its equity-based compensation, including incentive units, common stock, restricted common stock and stock options. The Company utilized various valuation methodologies in accordance with the framework of the 2013 American Institute of Certified Public Accountants Technical Practice Aid, *Valuation of Privately-Held Company Equity Securities Issued as Compensation*, to estimate the fair value of its equity awards. Each valuation methodology includes estimates and assumptions that require the Company's judgment. These estimates and assumptions include a number of objective and subjective factors, including external market conditions, guideline public company information, the prices at which the Company sold convertible preferred units and convertible preferred stock, the superior rights and preferences of securities senior to the Company's common units and common stock at the time and the likelihood of achieving a liquidity event such as an initial public offering or sale. Significant changes to the assumptions used in the valuations could result in different fair values of common units, incentive units, common stock, restricted common stock and stock options at each valuation date, as applicable.

SCHOLAR ROCK HOLDING CORPORATION**Notes to Consolidated Financial Statements (Continued)**

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

2. Summary of Significant Accounting Policies (Continued)***Unaudited Interim Financial Information***

The accompanying consolidated balance sheet as of March 31, 2018, the consolidated statements of operations and comprehensive loss and cash flows for the three months ended March 31, 2017 and 2018, and the consolidated statement of convertible preferred stock and stockholders' equity (deficit) for the three months ended March 31, 2018 are unaudited. The unaudited interim financial statements have been prepared on the same basis as the audited annual financial statements and, in the opinion of management, reflect all adjustments, which include only normal recurring adjustments, necessary for the fair presentation of the Company's financial position as of March 31, 2018 and the results of its operations and its cash flows for the three months ended March 31, 2017 and 2018. The financial data and other information disclosed in these notes related to the three months ended March 31, 2017 and 2018 are also unaudited. The results for the three months ended March 31, 2018 are not necessarily indicative of results to be expected for the year ended December 31, 2018, any other interim periods, or any future year or period.

Unaudited Pro Forma Information

On January 26, 2018, the Company's board of directors authorized the management of the Company to file a registration statement with the Securities and Exchange Commission to sell shares of its common stock to the public. Upon the closing of a qualified initial public offering (as defined in the Company's Certificate of Incorporation), all of the Company's outstanding shares of convertible preferred stock will automatically convert into shares of common stock, the outstanding warrant for the purchase of shares of convertible preferred stock will automatically convert into a warrant for the purchase of shares of common stock and the Company's performance-based stock option award with vesting conditions contingent upon the closing of an IPO will vest. The accompanying unaudited pro forma consolidated balance sheet and consolidated statement of convertible preferred stock and stockholders' equity (deficit) as of March 31, 2018 have been prepared to give effect to (1) the automatic conversion of all outstanding shares of convertible preferred stock into 15,109,950 shares of common stock, (2) the automatic conversion of the outstanding warrant to purchase 21,739 shares of convertible preferred stock into a warrant to purchase 7,614 shares of common stock, resulting in the reclassification of the warrant liability to additional paid-in capital and (3) the vesting of a performance-based stock option award to purchase up to 8,757 shares of the Company's common stock with vesting conditions contingent upon the closing of an IPO, resulting in the recognition of equity-based compensation expense, as if the Company's proposed IPO had occurred on March 31, 2018. The shares of common stock issuable and the proceeds expected to be received in the proposed IPO are excluded from such pro forma financial information.

The unaudited pro forma basic and diluted net loss per share in the accompanying consolidated statements of operations and comprehensive loss for the year ended December 31, 2017 and for the three months ended March 31, 2018 have been computed to give effect to the automatic conversion of all outstanding shares of convertible preferred stock into shares of common stock and the automatic conversion of the warrant to purchase shares of convertible preferred stock into a warrant to purchase shares of common stock. In addition, the unaudited pro forma basic and diluted net loss per share in the accompanying consolidated statement of operations and comprehensive loss for the three months ended March 31, 2018 has been computed to also give effect to the vesting of a performance-based stock option award with vesting conditions contingent upon the closing of an IPO. The unaudited pro forma basic and diluted net loss per share for the year ended December 31, 2017 and three months ended March 31, 2018 were computed using the weighted average number of shares of common stock outstanding, including the pro

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

2. Summary of Significant Accounting Policies (Continued)

forma effect of the conversion of all outstanding shares of convertible preferred stock into shares of common stock, as if the Company's proposed IPO had occurred on the later of January 1, 2017 or the original issuance dates of the convertible preferred units or convertible preferred stock. The unaudited pro forma net loss used in the calculation of unaudited basic and diluted pro forma net loss per share for the year ended December 31, 2017 and the three months ended March 31, 2018 excludes the impact of the change in fair value of the warrant liability that was recorded by the Company during such periods. In addition, the unaudited pro forma net loss used in the calculation of unaudited basic and diluted pro forma net loss per share for the three months ended March 31, 2018 also includes additional equity-based compensation expense related to the vesting of the Company's performance-based stock option award with vesting conditions contingent upon the closing of an IPO. The unaudited pro forma net loss per share does not include the shares expected to be sold or related proceeds to be received in the proposed IPO.

Concentration of Credit Risk and Off-Balance Sheet Risk

The Company has no off-balance sheet risk, such as foreign exchange contracts, option contracts or other foreign-hedging arrangements.

The primary objectives for the Company's investment portfolio are the preservation of capital and maintenance of liquidity. In 2016, the Company expanded its investment policy to allow funds to be held outside bank accounts, but to be invested only in fixed income instruments denominated and payable in U.S. dollars including obligations of the U.S. government and its agencies and money market funds registered according to SEC Rule 2a-7 of the Investment Company Act of 1940. Investments in the money market fund shall be consistent with approved instruments and assets under management must be at least \$10 billion.

All securities must have a readily ascertainable market value, must be readily marketable and be U.S. dollar denominated.

Cash and Cash Equivalents and Restricted Cash

The Company considers highly liquid investments with a maturity of three months or less when purchased to be cash equivalents. At December 31, 2016 and 2017 and March 31, 2018, cash and cash equivalents include bank demand deposits and money market funds that invest primarily in U.S. government-backed securities and treasuries. Cash equivalents are stated at cost, which approximates market value.

Restricted cash consists of a letter of credit in the amount of \$205,000 issued to the landlord of the Company's facility lease. The terms of the letter of credit extend beyond one year. The following table reconciles cash and cash equivalents and restricted cash per the balance sheet to the statement of cash flows:

	As of December 31,		As of March 31,	
	2016	2017	2017	2018
Cash and cash equivalents	\$ 10,033	\$ 56,461	\$ 6,659	\$ 22,822
Restricted cash	205	205	205	205
	<u>\$ 10,238</u>	<u>\$ 56,666</u>	<u>\$ 6,864</u>	<u>\$ 23,027</u>

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

2. Summary of Significant Accounting Policies (Continued)

Property and Equipment

Property and equipment are recorded at cost. Expenditures for major renewals or betterments that extend the useful lives of property and equipment are capitalized; expenditures for maintenance and repairs are charged to expense as incurred. Depreciation is calculated on a straight-line basis over the estimated useful lives of the related asset. Property and equipment are depreciated as follows:

	Estimated Useful Life (in Years)
Laboratory equipment	3 - 5
Machinery and equipment	3 - 5
Furniture and fixtures	5

Leasehold improvements are amortized over the shorter of the useful life or remaining term of the lease.

Impairment of Long-Lived Assets

Long-lived assets consist of property and equipment. Long-lived assets to be held and used are tested for recoverability whenever events or changes in business circumstances indicate that the carrying amount of the assets may not be fully recoverable. Factors that the Company considers in deciding when to perform an impairment review include significant underperformance of the business in relation to expectations, significant negative industry or economic trends and significant changes or planned changes in the use of the assets. If an impairment review is performed to evaluate a long-lived asset group for recoverability, the Company compares forecasts of undiscounted cash flows expected to result from the use and eventual disposition of the long-lived asset group to its carrying value. An impairment loss would be recognized when estimated undiscounted future cash flows expected to result from the use of an asset group are less than its carrying amount. The impairment loss would be based on the excess of the carrying value of the impaired asset group over its fair value, determined based on discounted cash flows. The Company did not record any impairment losses on long-lived assets during the years ended December 31, 2016 or 2017 or during the three months ended March 31, 2017 or 2018.

Fair Value Measurements

ASC Topic 820, *Fair Value Measurement* ("ASC 820"), establishes a fair value hierarchy for instruments measured at fair value that distinguishes between assumptions based on market data (observable inputs) and the Company's own assumptions (unobservable inputs). Observable inputs are inputs that market participants would use in pricing the asset or liability based on market data obtained from sources independent of the Company. Unobservable inputs are inputs that reflect the Company's assumptions about the inputs that market participants would use in pricing the asset or liability and are developed based on the best information available in the circumstances. ASC 820 identifies fair value as the exchange price, or exit price, representing the amount that would be received to sell an asset or paid to transfer a liability in an orderly transaction between market participants. As a basis for considering market participant

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

2. Summary of Significant Accounting Policies (Continued)

assumptions in fair value measurements, ASC 820 establishes a three-tier fair value hierarchy that distinguishes between the following:

Level 1 — Quoted market prices in active markets for identical assets or liabilities.

Level 2 — Inputs other than Level 1 inputs that are either directly or indirectly observable, such as quoted market prices, interest rates and yield curves.

Level 3 — Unobservable inputs developed using estimates of assumptions developed by the Company, which reflect those that a market participant would use.

To the extent the valuation is based on models or inputs that are less observable or unobservable in the market, the determination of fair values requires more judgment. Accordingly, the degree of judgment exercised by the Company in determining fair value is greatest for instruments categorized as Level 3. A financial instrument's level within the fair value hierarchy is based on the lowest level of any input that is significant to the fair value measurement.

Segment Information

Operating segments are defined as components of an entity about which separate discrete information is available for evaluation by the chief operating decision maker, or decision-making group, in deciding how to allocate resources and in assessing performance. The Company views its operations and manages its business in one operating segment operating exclusively in the United States.

Revenue Recognition

Effective January 1, 2018, the Company adopted the provisions of ASC 606 using the full retrospective transition method. Under this method, the Company revised its consolidated financial statements for the years ended December 31, 2016 and 2017, and all applicable interim periods within those years, as if ASC 606 had been effective for those periods. This standard applies to all contracts with customers, except for contracts that are within the scope of other standards, such as leases, insurance, collaboration arrangements and financial instruments.

To date all revenue has been generated from an agreement with Janssen.

Under ASC 606, an entity recognizes revenue when its customer obtains control of promised goods or services, in an amount that reflects the consideration which the entity expects to receive in exchange for those goods or services. To determine the appropriate amount of revenue to be recognized for arrangements determined to be within the scope of ASC 606, the Company performs the following five steps: (i) identification of the contract(s) with the customer, (ii) identification of the promised goods or services in the contract and determination of whether the promised goods or services are performance obligations, (iii) measurement of the transaction price, (iv) allocation of the transaction price to the performance obligations, and (v) recognition of revenue when (or as) the Company satisfies each performance obligation. The Company only applies the five-step model to contracts when it is probable that the entity will collect consideration it is entitled to in exchange for the goods or services it transfers to the customer.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

2. Summary of Significant Accounting Policies (Continued)

The Company accounts for a contract with a customer that is within the scope of ASC 606 when all of the following criteria are met: (i) the arrangement has been approved by the parties and the parties are committed to perform their respective obligations, (ii) each party's rights regarding the goods or services to be transferred can be identified, (iii) the payment terms for the goods or services to be transferred can be identified, (iv) the arrangement has commercial substance and (v) collection of substantially all of the consideration to which the Company will be entitled in exchange for the goods or services that will be transferred to the customer is probable.

The promised goods or services in the Company's arrangement consist of license rights to the Company's intellectual property and research and development services. The arrangement also has options for additional items (i.e., additional license rights). Options are considered to be marketing offers and are to be accounted for as separate contracts when the customer elects such options, unless the Company determines the option provides a material right which would not be provided without entering into the contract. Performance obligations are promised goods or services in a contract to transfer a distinct good or service to the customer. Promised goods or services are considered distinct when: (i) the customer can benefit from the good or service on its own or together with other readily available resources and (ii) the promised good or service is separately identifiable from other promises in the contract. In assessing whether promised goods or services are distinct, the Company considers factors such as the stage of development of the underlying intellectual property, the capabilities of the customer to develop the intellectual property on their own and whether the required expertise is readily available.

The Company estimates the transaction price based on the amount of consideration the Company expects to be received for transferring the promised goods or services in the contract. The consideration may include both fixed consideration and variable consideration. At the inception of each arrangement that includes variable consideration, the Company evaluates the amount of the potential payments and the likelihood that the payments will be received. The Company utilizes either the most likely amount method or expected value method to estimate the transaction price based on which method better predicts the amount of consideration expected to be received. If it is probable that a significant revenue reversal would not occur, the variable consideration is included in the transaction price.

The Company's arrangement includes development and regulatory milestone payments. The Company evaluates whether the milestones are considered probable of being reached and estimates the amount to be included in the transaction price using the most likely amount method. If it is probable that a significant revenue reversal would not occur, the associated milestone value is included in the transaction price. Milestone payments that are not within the Company's control or the licensee's control, such as regulatory approvals, are not considered probable of being achieved until those approvals are received. At the end of each reporting period, the Company re-evaluates the probability of achievement of such milestones and any related constraint, and if necessary, adjusts the estimate of the overall transaction price. Any such adjustments are recorded on a cumulative catch-up basis, which would affect collaboration revenue and net income (loss) in the period of adjustment. To date, no milestones have been achieved under the Company's arrangement.

For sales-based royalties, including milestone payments based on the level of sales, the Company determines whether the sole or predominant item to which the royalties relate is a license. When the license

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

2. Summary of Significant Accounting Policies (Continued)

is the sole or predominant item to which the sales-based royalty relates, the Company recognizes revenue at the later of: (i) when the related sales occur, or (ii) when the performance obligation to which some or all of the royalty has been allocated has been satisfied (or partially satisfied). To date, the Company has not recognized any sales-based royalty revenue resulting from the Company's arrangement.

The Company allocates the transaction price based on the estimated standalone selling price. The Company must develop assumptions that require judgment to determine the standalone selling price for each performance obligation identified in the contract. The Company utilizes key assumptions to determine the standalone selling price, which may include other comparable transactions, pricing considered in negotiating the transaction and the estimated costs. Certain variable consideration is allocated specifically to one or more performance obligations in a contract when the terms of the variable consideration relate to the satisfaction of the performance obligation and the resulting amounts allocated to each performance obligation are consistent with the amounts the Company would expect to receive for each performance obligation.

For performance obligations which consist of licenses and other promises, the Company utilizes judgment to assess the nature of the combined performance obligation in order to determine whether the combined performance obligation is satisfied over time or at a point in time. The Company determines the appropriate method of measuring progress of combined performance obligations satisfied over time for purposes of recognizing revenue. The Company evaluates the measure of progress each reporting period and, if necessary, adjusts the measure of performance and related revenue recognition. If the license to the Company's intellectual property is determined to be distinct from the other performance obligations identified in the arrangement, the Company will recognize revenue from non-refundable, up-front fees allocated to the license when the license is transferred to the customer and the customer is able to use and benefit from the license.

The Company receives payments from customers based on billing schedules established in each contract. Up-front payments and fees are recorded as deferred revenue upon receipt or when due until the Company performs its obligations under these arrangements. Amounts are recorded as accounts receivable when the Company's right to consideration is unconditional.

Research and Development Expenses

Research and development expenses are expensed as incurred and consist of costs incurred in performing research and development activities, including compensation related expenses for research and development personnel, preclinical and clinical activities including cost of supply, overhead expenses including facilities expenses, materials and supplies, amounts paid to consultants and outside service providers, and depreciation of equipment. Upfront license payments related to acquired technologies which have not yet reached technological feasibility and have no alternative future use are also included in research and development expense.

Research Contract Costs and Accruals

The Company has entered into various research service arrangements under which vendors perform various services. The Company records accrued expenses for estimated costs incurred under the arrangements. When evaluating the adequacy of the accrued expenses, the Company analyzed the progress of the studies,

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

2. Summary of Significant Accounting Policies (Continued)

trials or other services performed, including invoices received and contracted costs. Significant judgments and estimates are made in determining the accrued expense balances at the end of each reporting period.

Equity-Based Compensation

The Company accounts for equity awards, including incentive units, common stock, restricted common stock and common stock options, granted to employees as equity award compensation in accordance with ASC Topic 718, *Compensation — Stock Compensation* ("ASC 718"). ASC 718 requires all stock-based payments to employees, which includes grants of employee equity awards, to be recognized as expense in the statements of operations based on their grant date fair values. The Company estimates the fair value of incentive units and common stock using an appropriate valuation methodology, based on the guideline public company (GPC) method or the precedent transaction method which "backsolves" to a preferred price. The use of these valuation approaches requires management to make assumptions with respect to the expected volatility of its common units and common stock, time until a liquidity event and risk-free interest rates. The fair value of each restricted common stock award is based on the fair value of the Company's common stock less any purchase price, if applicable. The fair value of each stock option award is estimated using the Black-Scholes option-pricing model, which uses as inputs the fair value of the Company's common stock and certain subjective assumptions, including the expected stock price volatility, the expected term of the award, the risk-free rate, and expected dividends. Expected volatility is calculated based on reported volatility data for a representative group of publicly traded companies for which historical information was available. The historical volatility is generally calculated based on a period of time commensurate with the expected term assumptions. The risk-free interest rate is based on the U.S. Treasury yield curve in effect at the time of grant commensurate with the expected term assumption. The Company uses the simplified method, under which the expected term is presumed to be the midpoint between the vesting date and the end of the contractual term. The Company utilizes this method due to lack of historical exercise data and the plain nature of its stock-based awards. The Company uses the remaining contractual term for the expected life of non-employee awards. The expected dividend yield is assumed to be zero as the Company has never paid dividends and has no current plans to pay any dividends on common stock.

Compensation expense related to equity awards to employees that are subject to graded vesting is recognized on a straight-line basis, based on the grant date fair value, over the requisite service period of the award, which is generally the vesting term. For awards subject to performance conditions, the Company recognizes equity award compensation expense using an accelerated recognition method over the remaining service period when management determines that achievement of the milestone is probable. Management evaluates when the achievement of a performance-based milestone is probable based on the relative satisfaction of the performance conditions as of the reporting date.

For equity awards granted to non-employees, the Company accounts for the related equity award compensation in accordance with the provisions of ASC 718 and ASC Topic 505, *Equity*, and recognizes equity award compensation expense over the related service period of the non-employee award. Equity awards issued to non-employees are recorded at their fair values, using the then-current fair value of the incentive units, common stock and updated assumption inputs in the Black-Scholes option-pricing model, as applicable, and are periodically revalued as the equity instruments vest.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

2. Summary of Significant Accounting Policies (Continued)

The Company classifies equity-based compensation expense in its consolidated statements of operations in the same manner in which the award recipient's salary and related costs are classified or in which the award recipient's service payments are classified.

Convertible Preferred Units and Convertible Preferred Stock

The Company records all convertible preferred units or convertible preferred shares at their respective fair values on the dates of issuance less issuance costs. The Company classifies its convertible preferred units and convertible preferred shares outside of stockholders' equity (deficit) when the redemption of such units or shares is outside the Company's control. The Company does not adjust the carrying values of the convertible preferred units or convertible preferred stock to the liquidation preferences of such units or shares until such time as a deemed liquidation event is probable of occurring.

Comprehensive Loss

Comprehensive loss is the change in equity of a business enterprise during a period from transactions and other events and circumstances from non-owner sources. Comprehensive loss includes net loss and the change in accumulated other comprehensive loss for the period. Accumulated other comprehensive loss consisted entirely of unrealized gains and losses on available-for-sale marketable securities.

Net Loss per Unit and Share

The Company applies the two-class method to compute basic and diluted net loss per unit and net loss per share because it has issued units and shares that meet the definition of participating securities. The two-class method determines net income (loss) per unit and share for each class of common and participating securities according to dividends declared or accumulated and participation rights in undistributed earnings. The two-class method requires income (losses) available to common unit holders and common stockholders for the period to be allocated between common and participating securities based upon their respective rights to share in the earnings as if all income (losses) for the period had been distributed. During periods of loss, there is no allocation required under the two-class method since the participating securities do not have a contractual obligation to fund the losses of the Company.

Prior to the Reorganization, the Company calculates basic net loss per unit by dividing net loss by the weighted average number of common units outstanding. Subsequent to the Reorganization, the Company calculates basic net loss per share by dividing net loss by the weighted average number of common shares outstanding, excluding restricted common stock. The Company calculates diluted net loss per unit and diluted net loss per share by dividing net loss by the weighted average number of common units outstanding or weighted average number of common shares outstanding, as applicable, after giving consideration to the dilutive effect of convertible preferred units, convertible preferred stock, incentive units, restricted common stock, warrants and stock options that are outstanding during the period. The Company has generated a net loss in all periods presented, so the basic and diluted net loss per unit and net loss per share are the same, as the inclusion of the potentially dilutive securities would be anti-dilutive.

Income Taxes

Prior to the Reorganization, Scholar Rock, LLC elected to be treated under the partnership provisions of the Internal Revenue Service code. Accordingly, all income and deductions of Scholar Rock, LLC were recorded on the members' individual tax returns and no taxes were recorded by Scholar Rock, LLC. Scholar Rock, Inc., the wholly-owned subsidiary of Scholar Rock, LLC, was taxed as a C-corporation for federal

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

2. Summary of Significant Accounting Policies (Continued)

income tax purposes and filed a separate corporate income tax return from the LLC entity. All operations were recorded at Scholar Rock, Inc. Subsequent to the Reorganization, Scholar Rock Holding Corporation became the 100% owner of Scholar Rock, LLC, creating a new ultimate parent company, and a consolidated group for income tax reporting. The Reorganization and change in tax status of the reporting entity did not have an impact on the consolidated tax provision.

Income taxes for Scholar Rock Holding Corporation and Scholar Rock, Inc. are recorded in accordance with ASC Topic 740, *Income Taxes ("ASC 740")*, which provides for deferred taxes using an asset and liability approach. Under this method, deferred income tax assets and liabilities are recognized based on future income tax consequences attributable to differences between the financial statement carrying amount of existing assets and liabilities, and their respective income tax basis. Deferred income tax assets and liabilities are measured using enacted income tax rates expected to apply to taxable income in the years in which those temporary differences are expected to be recovered or settled. The effect of changes in income tax rates on deferred income tax assets and liabilities is recognized as income or expense in the period that a valuation allowance for any income tax benefits of which future realization is not more likely than not.

The Company provides reserves for potential payments of tax to various tax authorities related to uncertain tax positions. The tax benefits recorded are based on a determination of whether and how much of a tax benefit taken by the Company in its tax filings or positions is "more likely than not" to be realized following resolution of any uncertainty related to the tax benefit, assuming that the matter in question will be raised by the tax authorities.

The Company is open to examination by the Internal Revenue Service for the tax years ended December 31, 2012 to December 31, 2017. The Company is currently not under examination by the Internal Revenue Service or any other jurisdictions for any tax years. The Company has not recorded any interest or penalties on any unrecognized tax benefits since its inception.

Marketable Securities

The Company classifies its marketable securities as available-for-sale. Marketable securities with a remaining maturity date greater than one year are classified as non-current. Marketable securities are maintained by an investment manager and consist of U.S. treasury securities. Marketable securities are carried at fair value with the unrealized gains and losses included in accumulated other comprehensive loss as a component of stockholders' equity (deficit) until realized. Any premium or discount arising at purchase is amortized and/or accreted to interest income and/or expense over the life of the underlying marketable security.

Although available to be sold to meet operating needs or otherwise, securities are generally held through maturity. The cost of securities sold is determined on a specific identification basis, and realized gains and losses are included in interest income (expense) within the statement of operations and comprehensive loss. During 2016, 2017 and the three months ended March 31, 2017 and 2018, there were no realized gains or losses on sales of marketable securities and no marketable securities were adjusted for other than temporary declines in fair value.

The Company evaluates its marketable securities with unrealized losses for other-than-temporary impairment. When assessing marketable securities for other-than-temporary declines in value, the Company

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

2. Summary of Significant Accounting Policies (Continued)

considers such factors as, among other things, how significant the decline in value is as a percentage of the original cost, how long the market value of the investment has been less than its original cost, the Company's ability and intent to retain the investment for a period of time sufficient to allow for any anticipated recovery in fair value and market conditions in general. If any adjustment to fair value reflects a decline in the value of the investment that the Company considers to be "other than temporary," the Company would reduce the investment to fair value through a charge to the statement of operations and comprehensive loss. No such adjustments were necessary during the periods presented.

Warrant Liability

The Company accounts for warrant instruments that either conditionally or unconditionally obligate the issuer to transfer assets or issue equity shares as a liability, if it permits the holder to purchase redeemable shares, including shares that are contingently redeemable outside the control of the Company and the warrant itself is indexed to an underlying share that embodies the issuer's obligation to repurchase the share and the issuer has a conditional obligation to transfer the assets if the shares are put back. These warrants are subject to revaluation at each balance sheet date, and any change in fair value is recorded as a component of other expense until the earlier of their exercise or expiration or upon the completion of a liquidation event.

Deferred Offering Costs

The Company capitalizes certain legal, professional accounting and other third-party fees that are directly associated with in-process equity financings as deferred offering costs until such financings are consummated. After consummation of the equity financing, these costs are recorded in stockholders' equity (deficit) as a reduction of additional paid-in capital generated as a result of the offering. Should the planned equity financing be abandoned, the deferred offering costs will be expensed immediately as a charge to operating expenses in the consolidated statement of operations and comprehensive loss.

Subsequent Events

The Company considers events or transactions that occur after the balance sheet date but prior to the issuance of the consolidated financial statements for potential recognition or disclosure in the consolidated financial statements. Subsequent events have been evaluated through the date these consolidated financial statements were issued for potential recognition or disclosure in the consolidated financial statements.

Recently Adopted Accounting Pronouncements

In March 2016, the FASB issued ASU No. 2016-09, *Improvements to Employee Share-Based Payment Accounting* ("ASU 2016-09"). ASU 2016-09 simplified several aspects of the accounting for share-based payment transactions, including the income tax consequences, classification of awards as either equity or liabilities and classification on the statement of cash flows. The Company elected to early adopt ASU 2016-09 effective for the year ended December 31, 2016 and has elected to account for forfeitures when they occur instead of estimating the number of awards that are expected to vest. The adoption of ASU 2016-09 did not have a material impact on the Company's financial statements. The Company did not have any excess tax benefits associated with equity exercises and therefore there was no deferred tax asset recorded upon adoption.

In January 2017, the FASB issued ASU No. 2017-01, *Business Combinations* ("ASU 2017-01"), which clarified the definition of a business and provides a screen to determine when an integrated set of assets

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

2. Summary of Significant Accounting Policies (Continued)

and activities is not a business. The screen requires that when substantially all of the fair value of the gross assets acquired, or disposed of, is concentrated in a single identifiable asset or a group of similar identifiable assets, the set is not a business. The Company adopted this new standard as of January 1, 2017, with prospective application to any business development transactions.

In May 2014, the FASB issued ASC 606, which superseded nearly all existing revenue recognition guidance, including most industry-specific guidance. In 2015 and 2016, the FASB issued additional ASUs related to ASC 606 that delayed the effective date of the guidance and clarified various aspects of the new revenue guidance, including principal versus agent considerations, identifying performance obligations and licensing, and other improvements and practical expedients. The Company adopted this new standard on January 1, 2018 using the full retrospective transition method, and has elected to use the contract modification practical expedient as permitted under the rules of adoption. Under the contract modification practical expedient, the impact of adoption upon initial application of the standard reflects the aggregate effect of all modifications that occur prior to initial date of adoption when: (i) identifying the satisfied and unsatisfied performance obligations, (ii) determining the transaction price and (iii) allocating the transaction price to the satisfied and unsatisfied performance obligations. The adoption of the new revenue recognition guidance did not have any impact on the consolidated financial statements as of the date of adoption or for any period presented.

In August 2016, the FASB issued ASU No. 2016-15, *Statement of Cash Flows: Classification of Certain Cash Receipts and Cash Payments*. The new standard clarifies certain aspects of the statement of cash flows, including several aspects that are not currently applicable to the Company. The new standard also clarifies that an entity should determine each separately identifiable source or use within the cash receipts and cash payments on the basis of the nature of the underlying cash flows. In situations in which cash receipts and payments have aspects of more than one class of cash flows and cannot be separated by source or use, the appropriate classification should depend on the activity that is likely to be the predominant source or use of cash flows for the item. The Company adopted this new standard as of January 1, 2018, with the retrospective transition method to each period presented. The adoption of the new cash flow guidance did not have any impact the consolidated financial statements as of the date of adoption or for any period presented.

In October 2016, the FASB issued ASU No. 2016-18, *Restricted Cash*, which requires that a statement of cash flows explain the change during the period in the total of cash, cash equivalents, and amounts generally described as restricted cash or restricted cash equivalents. The amounts generally described as restricted cash and restricted cash equivalents should be included with cash and cash equivalents when reconciling the beginning-of-period and end-of period total amounts shown on the statement of cash flows. ASU No. 2016-18 is effective for fiscal years beginning after December 15, 2017, and interim periods within those years, using a retrospective transition method to each period presented. Early adoption was permitted. The Company adopted this new standard as of January 1, 2018, with retrospective application to each period presented. As a result of the adoption, the Company have added restricted cash to the reconciliation of beginning and ending cash and cash equivalents and included a reconciliation of total cash, cash equivalents and restricted cash to the balance sheet for each period presented in the consolidated statements of cash flows.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

2. Summary of Significant Accounting Policies (Continued)

In May 2017, the FASB issued ASU No. 2017-09, *Compensation — Stock Compensation* ("ASU 2017-09"), which provided guidance about which changes to the terms or conditions of a share-based payment award require an entity to apply modification accounting. The new guidance requires modification accounting if the vesting condition, fair value or award classification is not the same both before and after a change to the terms and conditions of the award. The Company adopted this new standard as of January 1, 2018, with prospective application to any awards modifications on or after the adoption date.

Recently Issued Accounting Pronouncements

From time to time, new accounting pronouncements are issued by the FASB or other standard setting bodies and adopted by the Company as of the specified effective date.

In February 2016, the FASB issued ASU No. 2016-02, *Leases (Topic 842)*, which supersedes all existing lease guidance. The new standard requires a company to recognize lease assets and liabilities for leases previously classified as operating leases. The new standard will be effective for the Company on January 1, 2019. The Company is currently evaluating the potential impact that this update may have on the Company's financial position and results of operations.

3. Fair Value of Financial Assets and Liabilities

The following tables summarize the assets and liabilities measured at fair value on a recurring basis at December 31, 2016 and 2017 and March 31, 2018 (in thousands):

	Fair Value Measurements at December 31,			
	2016			
	Total	Level 1	Level 2	Level 3
Assets:				
Money market funds, included in cash and cash equivalents	\$ 8,166	\$ 8,166	\$ —	\$ —
Marketable securities:				
U.S. Treasury obligations	19,498	19,498	—	—
Total assets	\$ 27,664	\$ 27,664	\$ —	\$ —
Liabilities:				
Warrant to purchase redeemable security	\$ 27	\$ —	\$ —	\$ 27
Total liabilities	\$ 27	\$ —	\$ —	\$ 27

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

3. Fair Value of Financial Assets and Liabilities (Continued)

	Fair Value Measurements at December 31, 2017			
	Total	Level 1	Level 2	Level 3
Assets:				
Money market funds, included in cash and cash equivalents	\$ 55,291	\$ 55,291	\$ —	\$ —
Marketable securities:				
U.S. Treasury obligations	1,498	1,498	—	—
Total assets	\$ 56,789	\$ 56,789	\$ —	\$ —
Liabilities:				
Warrant to purchase redeemable security	\$ 37	\$ —	\$ —	\$ 37
Total liabilities	\$ 37	\$ —	\$ —	\$ 37

	Fair Value Measurements at March 31, 2018			
	Total	Level 1	Level 2	Level 3
Assets:				
Money market funds, included in cash and cash equivalents	\$ 21,242	\$ 21,242	\$ —	\$ —
Marketable securities:				
U.S. Treasury obligations	24,941	24,941	—	—
Total assets	\$ 46,183	\$ 46,183	\$ —	\$ —
Liabilities:				
Warrant to purchase redeemable security	\$ 57	\$ —	\$ —	\$ 57
Total liabilities	\$ 57	\$ —	\$ —	\$ 57

Cash and cash equivalents and marketable securities include investments in money market funds that invest in U.S. government securities that are valued using quoted market prices. Accordingly, money market funds and government funds are categorized as Level 1 as of December 31, 2016 and 2017 and March 31, 2018.

The carrying amounts reflected in the balance sheets for accounts receivable, prepaid expenses and other current assets, accounts payable, and accrued expenses approximate their fair values at December 31, 2016 and 2017 and March 31, 2018, due to their short-term nature. The Company believes the terms of the loan payable reflect current market conditions for an instrument with similar terms and maturity, therefore the carrying value of the Company's debt approximates its fair value based on Level 3 of the fair value hierarchy.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

3. Fair Value of Financial Assets and Liabilities (Continued)

Warrants to Purchase Convertible Preferred Units or Stock Subject to Redemption

In conjunction with the loan and security agreement, the Company issued a warrant to SVB to purchase 21,739 Series A-3 Convertible Preferred Units at a purchase price of \$1.38 per unit. In connection with the Reorganization, the warrant was converted to or exchanged for a warrant to purchase 21,739 shares of Series A-3 Convertible Preferred Stock at the same purchase price. The warrant is exercisable immediately and expires on August 10, 2025. The Company determined that the warrant represents an instrument to purchase a redeemable security, and, therefore, is required to be classified as a liability on the balance sheet. Because the warrant is classified as a liability, the liability is re-measured at its fair value at each balance sheet date.

The fair value of the warrant is estimated using the Black-Scholes option-pricing model. The estimates in the Black-Scholes option-pricing model are based, in part, on subjective assumptions, including stock price volatility, term of the warrants, risk free interest rate, dividend yield and fair value of the preferred stock underlying the warrants. Such assumptions could differ materially in the future. The warrant was initially valued at \$20,300 and was included in issuance costs incurred in connection with the loan payable, recorded as a discount to the debt and amortized over the term of the loan as interest expense. The change in fair value of the warrant is recorded as other expense in the consolidated statement of operations.

The following assumptions were used in valuing the warrant at December 31, 2016 and 2017 and March 31, 2018:

	December 31,		March 31,
	2016	2017	2018
Interest rate	2.35%	2.33%	2.68%
Expected dividend yield	0.00%	0.00%	0.00%
Expected term (years)	8.61	7.61	7.37
Expected volatility	60.40%	77.40%	84.02%

The following table sets forth a summary of changes in the fair value of the warrant, which represented a recurring measurement classified within Level 3 of the fair value hierarchy, wherein fair value was estimated using significant unobservable inputs (in thousands):

Balance at December 31, 2015	\$ (28)
Change in fair value of warrant included in other income (expense), net	1
Balance at December 31, 2016	(27)
Change in fair value of warrant included in other income (expense), net	(10)
Balance at December 31, 2017	(37)
Change in fair value of warrant included in other income (expense), net	(20)
Balance at March 31, 2018	\$ (57)

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

4. Marketable Securities

The following table summarizes the Company's investments as of December 31, 2016 (in thousands):

	Amortized Cost	Gross Unrealized		Estimated Fair Value
		Gains	Losses	
Marketable securities available-for-sale:				
U.S. Treasury obligations	\$ 19,518	\$ —	\$ (20)	\$ 19,498
Total available-for-sale securities	<u>\$ 19,518</u>	<u>\$ —</u>	<u>\$ (20)</u>	<u>\$ 19,498</u>

The aggregate fair value of marketable securities with unrealized losses was \$19.5 million at December 31, 2016. At December 31, 2016, 13 investments were in an unrealized loss position. All such investments have been in an unrealized loss position for less than a year and these losses are considered temporary. The Company has the ability and intent to hold these investments until a recovery of their amortized cost. As of December 31, 2016, the Company held one investment with a fair value of \$1.5 million with a maturity greater than one year. This investment matures in March 2018.

The following table summarizes the Company's investments as of December 31, 2017 (in thousands):

	Amortized Cost	Gross Unrealized		Estimated Fair Value
		Gains	Losses	
Marketable securities available-for-sale:				
U.S. Treasury obligations	\$ 1,500	\$ —	\$ (2)	\$ 1,498
Total available-for-sale securities	<u>\$ 1,500</u>	<u>\$ —</u>	<u>\$ (2)</u>	<u>\$ 1,498</u>

The Company did not have any marketable securities in an unrealized loss position at December 31, 2017.

The following table summarizes the Company's investments as of March 31, 2018 (in thousands):

	Amortized Cost	Gross Unrealized		Estimated Fair Value
		Gains	Losses	
Marketable securities available-for-sale:				
U.S. Treasury obligations	\$ 24,942	\$ —	\$ (1)	\$ 24,941
Total available-for-sale securities	<u>\$ 24,942</u>	<u>\$ —</u>	<u>\$ (1)</u>	<u>\$ 24,941</u>

The Company did not have any marketable securities in an unrealized loss position at March 31, 2018.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

5. Property and Equipment, Net

At December 31, 2016 and 2017 and March 31, 2018, property and equipment consists of the following (in thousands):

	As of December 31,		As of
	2016	2017	March 31, 2018
Laboratory equipment	\$ 1,673	\$ 2,074	\$ 2,097
Furniture & fixtures	132	151	159
Machinery & equipment	7	7	7
Leasehold improvements	1,498	1,498	1,498
	<u>3,310</u>	<u>3,730</u>	<u>3,761</u>
Less: Accumulated depreciation and amortization	(880)	(1,549)	(1,728)
	<u>\$ 2,430</u>	<u>\$ 2,181</u>	<u>\$ 2,033</u>

Depreciation and amortization expense was \$575,000, \$669,000, \$164,000 and \$179,000 for the years ended December 31, 2016 and 2017 and the three months ended March 31, 2017 and 2018, respectively.

6. Accrued Expenses

At December 31, 2016 and 2017 and March 31, 2018, accrued expenses consist of the following (in thousands):

	As of December 31,		As of March 31,
	2016	2017	2018
Accrued external research and development expense	\$ 451	\$ 1,225	\$ 463
Accrued payroll and related expenses	817	1,174	621
Accrued professional and consulting expense	201	382	414
Accrued initial public offering costs	—	—	473
Accrued other	18	15	27
	<u>\$ 1,487</u>	<u>\$ 2,796</u>	<u>\$ 1,998</u>

7. Member Units

Prior to the Reorganization, all interests of members in distributions and other amounts were represented by their units of membership in the Company as specified in its operating agreement. There were two classes of units, capital units and incentive units. Capital units were comprised of common units and convertible preferred units, which represent a capital interest in the Company, while incentive units represent profits

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

7. Member Units (Continued)

interests within the meaning of IRS Revenue Procedures 93-27 and 2001-43. The various classes of capital units are described below.

8. Common Units

As of December 31, 2015, the Company had outstanding 1,603,088 common units. There were no additional common units issued during the years ended December 31, 2016 and 2017 or the three months ended March 31, 2018.

Holders of common units were entitled to one vote per unit and receive dividends, when and if declared by the Board of Directors. No common unit dividends were declared. The voting, dividend, and liquidation rights of the holders of the common units were subject to, and qualified by, the rights of the holders of the Series A Convertible Preferred Units and the Series B Convertible Preferred Units.

9. Convertible Preferred Units

Prior to 2016, the Company had sold Series A-1, A-2, A-3 and A-4 Convertible Preferred Units. The Series A-1, A-2, A-3 and A-4 Convertible Preferred Units are collectively referred to as the Series A Convertible Preferred Units. Additionally, the Company sold 12,098,785 Series B Convertible Preferred Units. The Series A Convertible Preferred Units and Series B Convertible Preferred Units are collectively considered the Convertible Preferred Units.

As of December 31, 2016, the Convertible Preferred Units consisted of the following (in thousands, except unit amounts):

	As of December 31, 2016				Common Units Issuable Upon Conversion
	Preferred Units Authorized	Preferred Units Issued and Outstanding	Carrying Value	Liquidation Preference	
Series A-1 convertible preferred units	2,000,000	2,000,000	\$ 1,942	\$ 2,000	700,575
Series A-2 convertible preferred units	5,066,915	5,066,915	6,004	6,080	1,774,876
Series A-3 convertible preferred units	5,601,448	5,579,709	7,698	7,700	1,954,501
Series A-4 convertible preferred units	3,906,738	3,906,738	6,198	6,200	1,368,480
Series B convertible preferred units	13,526,994	12,098,785	36,215	36,297	4,238,049
	<u>30,102,095</u>	<u>28,652,147</u>	<u>\$ 58,057</u>	<u>\$ 58,277</u>	<u>10,036,481</u>

In May 2017, the Company issued 1,428,209 shares of Series B Convertible Preferred Units for net proceeds of \$4.3 million.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

9. Convertible Preferred Units (Continued)

The rights, preferences and privileges of the Convertible Preferred Units were as follows:

Liquidation

In the event of any liquidation, dissolution or winding up of affairs of the Company (including a change of control), distributions would have first been made to holders of the Series B Convertible Preferred Units equal to their original issue price plus any declared but unpaid dividends. The Series B Convertible Preferred Units were issued at \$3.00 per unit. After distribution to the Series B Convertible Preferred Unit holders, the holders of the Series A Convertible Preferred Units as a class would have received a distribution equal to their original issue price plus any declared but unpaid dividends. The Series A-1, A-2, A-3 and A-4 Convertible Preferred Units were issued at \$1.00, \$1.20, \$1.38 and \$1.587, respectively. Next, the common unit holders and incentive unit holders would have received a distribution in proportion to the respective aggregate number of common units and incentive units held by each member until each common and incentive unit holder had received, on a per unit basis, the sum of the amounts distributed to the holders of the Series A Convertible Preferred Units less any amounts distributed for dividends. Next, the Series A Convertible Preferred Unit holders, the common unit holders, and incentive unit holders would have received a distribution in proportion to the respective aggregate number of Series A Convertible Preferred Units, common units, and incentive units held by each member until each Series A Convertible Preferred Unit holder, common unit holder, and incentive unit holder had received, on a per unit basis, the sum of the amounts distributed to the holders of the Series B Convertible Preferred Units less any amounts distributed for dividends. Any remaining amount available for distribution would have been made to holders of the Convertible Preferred Units, on an as converted basis, common units and incentive units, in proportion to the respective number of units held by each member.

Incentive unit holders would have participated in distributions as described above only after the distribution met the strike price with respect to such unit. The strike price is an amount per incentive unit determined by the Board of Directors based on the amount of distributions that the holders of a common unit would have been entitled to receive in a hypothetical liquidation of the Company on the date of issuance of the incentive unit in which the Company sold its assets at fair market value, satisfied its liabilities and distributed the net proceeds to the holders of units in liquidation of the Company. Incentive unit holders would have participated in distributions only after the distribution met the strike price with respect to such unit. The Board of Directors had the discretion to determine the extent to which an incentive unit would have been excluded from participating in distributions.

Conversion

Upon either the closing of a firm-commitment underwritten public offering pursuant to an effective registration statement under the Securities Act of 1933, as amended, in which the price per common unit was at least \$3.00 per share and the aggregate gross proceeds were at least \$40 million or the occurrence of an event, specified by vote or written consent of the supermajority interest (members holding at least 70% of the outstanding Convertible Preferred Units, voting or consenting together as a single class on an as-converted basis), all outstanding Convertible Preferred Units would have been automatically converted into common units. Further, the automatic conversion of the Series B Convertible Preferred Units would not have been automatically converted into common units without the affirmative vote or written consent of the members holding at least 61% of the outstanding Series B Convertible Preferred Units.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

9. Convertible Preferred Units (Continued)

Dividends

Dividends, if declared, were payable to each Convertible Preferred Unit holder as follows:

Convertible Preferred Unit	Dividend Per Unit
B	\$ 0.240
A-4	0.127
A-3	0.110
A-2	0.096
A-1	0.080

Dividends were subject to appropriate adjustment in the event of any unit dividend, unit split, combination or other similar recapitalization with respect to the units. Dividends were payable when and as declared by the Board of Directors, were not cumulative and did not accrue to unit holders by reason of the fact that they are not declared or paid in any calendar year. No dividends have been declared by the Board of Directors since inception.

Voting

On any matter to be approved by the unit holders, holders of Convertible Preferred Units had the right to cast a number of votes equal to the number of common units into which the Convertible Preferred Units held by such holder would have converted.

Redemption

The Company's Convertible Preferred Units have been classified as temporary equity on the accompanying consolidated balance sheets in accordance with authoritative guidance for the classification and measurement of redeemable securities as the Convertible Preferred Units are redeemable upon the occurrence of a liquidation event. The carrying value of the Company's Convertible Preferred Units was not adjusted because a liquidation event was not probable and did not occur.

The Company has evaluated the Convertible Preferred Units and determined that they should be considered an "equity host" and not a "debt host." The evaluation was necessary to determine if any embedded features required bifurcation and separate accounting as a derivative financial instrument. The Company's analysis was based on a consideration of the economic characteristics and risks and more specifically, evaluated all the stated and implied substantive terms and features including (i) whether the Convertible Preferred Unit included redemption features, (ii) how and when any redemption features could have been exercised, (iii) whether the Convertible Preferred Units were entitled to dividends, (iv) the voting rights of the Convertible Preferred Unit and (v) the existence and nature of any conversion rights. As a result of its evaluation that the Convertible Preferred Unit is an "equity host," the various embedded conversion options are not considered a separate, embedded derivative.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

10. Reorganization and Convertible Preferred Stock

In connection with the Reorganization:

- § Holders of Scholar Rock, LLC Series B Convertible Preferred Units received one share of Scholar Rock Holding Corporation Series B Convertible Preferred Stock for each outstanding Series B Convertible Preferred Units held immediately prior to the Reorganization, with an aggregate of 13,526,994 shares of Scholar Rock Holding Corporation Series B Convertible Preferred Stock issued in the Reorganization;
- § Holders of Scholar Rock, LLC Series A-4 Convertible Preferred Units received one share of Scholar Rock Holding Corporation Series A-4 Convertible Preferred Stock for each outstanding Series A-4 Convertible Preferred Units held immediately prior to the Reorganization, with an aggregate of 3,906,738 shares of Scholar Rock Holding Corporation Series A-4 Convertible Preferred Stock issued in the Reorganization;
- § Holders of Scholar Rock, LLC Series A-3 Convertible Preferred Units received one share of Scholar Rock Holding Corporation Series A-3 Convertible Preferred Stock for each outstanding Series A-3 Convertible Preferred Units held immediately prior to the Reorganization, with an aggregate of 5,579,709 shares of Scholar Rock Holding Corporation Series A-3 Convertible Preferred Stock issued in the Reorganization;
- § Holders of Scholar Rock, LLC Series A-2 Convertible Preferred Units received one share of Scholar Rock Holding Corporation Series A-2 Convertible Preferred Stock for each outstanding Series A-2 Convertible Preferred Units held immediately prior to the Reorganization, with an aggregate of 5,066,915 shares of Scholar Rock Holding Corporation Series A-2 Convertible Preferred Stock issued in the Reorganization;
- § Holders of Scholar Rock, LLC Series A-1 Convertible Preferred Units received one share of Scholar Rock Holding Corporation Series A-1 Convertible Preferred Stock for each outstanding Series A-1 Convertible Preferred Units held immediately prior to the Reorganization, with an aggregate of 2,000,000 shares of Scholar Rock Holding Corporation Series A-1 Convertible Preferred Stock issued in the Reorganization;
- § Holders of Scholar Rock, LLC Common Units received one share of Scholar Rock Holding Corporation common stock for each outstanding Common Unit held immediately prior to the Reorganization, with an aggregate of 1,603,088 shares of common stock issued;
- § Holders of Scholar Rock, LLC vested and unvested incentive units, irrespective of any strike price or voting rights on any such outstanding incentive units, exchanged one incentive unit for one share of common stock or restricted common stock, respectively. The restricted common stock was issued with the same vesting terms as the unvested incentive units held immediately prior to the Reorganization. An aggregate of 2,367,498 shares of common stock and restricted common stock were issued and
- § The outstanding warrant to purchase 21,739 shares of Series A-3 Convertible Preferred Units at a purchase price of \$1.38 per share was converted to a warrant to purchase 21,739 shares of Series A-3 Convertible Preferred Stock at the same purchase price.

In evaluating the Reorganization, the Company considered that (i) with the exception of holders of Incentive Units, there were no changes in ownership interest held by each stockholder as a result of the Reorganization, (ii) the changes in the overall ownership interest of the Company resulting from the changes

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

10. Reorganization and Convertible Preferred Stock (Continued)

in ownership interest related to the holders of Incentive Units as a result of the Reorganization is not significant and (iii) the Reorganization occurred between a parent and wholly owned subsidiary, where the parent, Scholar Rock, LLC, had no substantive operations. Based on this evaluation, the Company determined that the Reorganization lacked economic substance and should be accounted for in a manner consistent with a common control transaction. Similarly, there was no change in fair value between the stockholders, individually or as a class, the Company determined that the exchange of shares occurring in the Reorganization should be accounted for as a modification of equity securities.

11. Convertible Preferred Stock and Common Stock

Convertible Preferred Stock

The Series A-1, A-2, A-3 and A-4 Convertible Preferred Stock issued in connection with the Reorganization are collectively referred to as Series A Convertible Preferred Stock.

On December 22, 2017, the Company entered into the Series C Preferred Stock Purchase Agreement pursuant to which the Company issued 13,055,555 shares of Series C Convertible Preferred Stock for net proceeds of \$46.9 million.

The Series A Convertible Preferred Stock, Series B Convertible Preferred Stock and the Series C Convertible Preferred Stock are collectively referred to as Convertible Preferred Stock.

As of December 31, 2017 and March 31, 2018, the Convertible Preferred Stock consisted of the following (in thousands, except share amounts):

	As of December 31, 2017 and March 31, 2018				
	Preferred Shares Authorized	Preferred Shares Issued and Outstanding	Carrying Value	Liquidation Preference	Common Shares Issuable Upon Conversion
Series A-1 convertible preferred stock	2,000,000	2,000,000	\$ 1,942	\$ 2,000	700,575
Series A-2 convertible preferred stock	5,066,915	5,066,915	6,004	6,080	1,774,876
Series A-3 convertible preferred stock	5,601,448	5,579,709	7,698	7,700	1,954,501
Series A-4 convertible preferred stock	3,906,739	3,906,738	6,198	6,200	1,368,480
Series B convertible preferred stock	13,526,994	13,526,994	40,500	40,581	4,738,333
Series C convertible preferred stock	13,055,555	13,055,555	46,890	47,000	4,573,185
	<u>43,157,651</u>	<u>43,135,911</u>	<u>\$ 109,232</u>	<u>\$ 109,561</u>	<u>15,109,950</u>

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

11. Convertible Preferred Stock and Common Stock (Continued)

The terms of the Convertible Preferred Stock are as follows:

Liquidation

In the event of any liquidation, dissolution or winding up of affairs of the Company (or upon a deemed liquidation event), distributions are first made to holders of the Series C Convertible Preferred Stock equal to the greater of (i) their original issuance price, plus any declared but unpaid dividends or (ii) the amount that the holder would be entitled upon conversion into common stock. The original issue price of the Series C Convertible Preferred Stock was \$3.60 per share. After distribution to the Series C Convertible Preferred Stockholders, distributions are made to holders of the Series B Convertible Preferred Stock equal to the greater of (i) their original issue price plus any declared but unpaid dividends or (ii) the amount that the holder would be entitled upon conversion into common stock. The original issue price of Series B Convertible Preferred Stock is \$3.00 per share. After distribution to the Series B Convertible Preferred Stockholders, the holders of the Series A Convertible Preferred Stock, as a class, will receive a distribution equal to the greater of (i) their original issue price plus any declared but unpaid dividends or (ii) the amount that the holder would be entitled upon conversion into common stock. The original issue price of the Series A-1, A-2, A-3 and A-4 Convertible Preferred Stock is \$1.00, \$1.20, \$1.38 and \$1.587, respectively. Upon completion of the preferential payments to holders of Convertible Preferred Stock, all of the remaining assets shall be distributed among the holders of Convertible Preferred Stock and common stock pro rata based on the number of shares of common stock held by each, assuming conversion of all outstanding shares of Convertible Preferred Stock.

A deemed liquidation event is defined as a merger (unless the shares of capital stock prior to the transaction represent a majority of the post merger voting rights) or the sale or transfer of substantially all of the assets of the Company unless the holders of at least (i) 80% of the Convertible Preferred Stock, (ii) 61% of the Series B Convertible Preferred Stock and (iii) a majority of the Series C Convertible Preferred Stock elect otherwise.

Conversion

Shares of Convertible Preferred Stock may be converted by the holder at any time into a number of common shares. The conversion price is equal to the original issue price for all Convertible Preferred Stock as of December 31, 2017 and March 31, 2018. The conversion price is subject to adjustments in the event of any stock dividend, stock split, combination or other similar recapitalization and other adjustments as set forth in the Company's certificate of incorporation. Upon either the closing of a firm-commitment underwritten public offering pursuant to an effective registration statement under the Securities Act of 1933, as amended, in which the aggregate gross proceeds are at least \$55 million or the occurrence of an event, specified by vote or written consent of the supermajority interest (members holding at least 80% of the outstanding Convertible Preferred Stock, voting or consenting together as a single class on an as-converted basis), all outstanding Convertible Preferred Stock will be automatically converted into common shares.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

11. Convertible Preferred Stock and Common Stock (Continued)

Dividends

Dividends, if declared, are payable to each holder of Convertible Preferred Stock as follows:

Convertible Preferred Stock	Dividend Per Share
C	\$ 0.288
B	0.240
A-4	0.127
A-3	0.110
A-2	0.096
A-1	0.080

Dividends are subject to appropriate adjustment in the event of any stock split, stock dividend, combination or other similar recapitalization with respect to the shares. Dividends are payable when and as declared by the Board of Directors, are not cumulative and do not accrue to shareholders by reason of the fact that they are not declared or paid in any calendar year. No dividends have been declared or paid by the Company to the holders of Convertible Preferred Stock since issuance of the Convertible Preferred Stock.

Voting

On any matter to be approved by the stockholders, holders of Convertible Preferred Stock have the right to cast a number of votes equal to the number of shares of common stock into which the shares of Convertible Preferred Stock held by such holder convert.

Redemption

The Company's Convertible Preferred Stock has been classified as temporary equity on the accompanying consolidated balance sheets in accordance with authoritative guidance for the classification and measurement of redeemable securities as the Convertible Preferred Stock is redeemable upon the occurrence of a deemed liquidation event. The carrying value of the Company's Convertible Preferred Stock is not being adjusted because a deemed liquidation event is not probable.

The Company has evaluated the Convertible Preferred Stock and determined that they should be considered an "equity host" and not a "debt host." The evaluation was necessary to determine if any embedded features require bifurcation and separate accounting as a derivative financial instrument. The Company's analysis was based on a consideration of the economic characteristics and risks and more specifically, evaluated all the stated and implied substantive terms and features including (i) whether the Convertible Preferred Stock includes redemption features, (ii) how and when any redemption features could be exercised, (iii) whether the Convertible Preferred Stock is entitled to dividends, (iv) the voting rights of the Convertible Preferred Stock and (v) the existence and nature of any conversion rights. As a result of its evaluation that the Convertible Preferred Stock is an "equity host," the various embedded conversion options are not considered a separate, embedded derivative.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

11. Convertible Preferred Stock and Common Stock (Continued)

Common Stock

The voting, dividend and liquidation rights of the holders of common stock are subject to and qualified by the rights, powers and preferences of the holders of Convertible Preferred Stock. The common stock has the following characteristics:

Voting

The holders of shares of common stock are entitled to one vote for each share of common stock held at any meeting of stockholders and at the time of any written action in lieu of a meeting.

Dividends

The holders of shares of common stock are entitled to receive dividends, if and when declared by the Company's board of directors. Cash dividends may not be declared or paid to holders of shares of common stock until all unpaid dividends on Convertible Preferred Stock have been paid in accordance with their terms. No dividends have been declared or paid by the company to the holders of common stock since the issuance of the common stock.

Liquidation

After payment of the respective liquidation preferences to the holders of Convertible Preferred Stock, the holders of common stock are entitled to share ratably in the Company's remaining assets available for distribution to its stockholders in the event of any voluntary or involuntary liquidation, dissolution or winding up of the Company or upon the occurrence of a deemed liquidation event.

Shares Reserved For Future Issuance

As of December 31, 2017 and March 31, 2018, the Company had reserved common shares for the conversion of outstanding Convertible Preferred Stock and for future issuance under the 2017 Stock Option and Incentive Plan (the "2017 Plan") as follows:

	As of December 31, 2017	As of March 31, 2018
Common shares reserved for conversion of convertible preferred stock outstanding	15,109,950	15,109,950
Common shares reserved for conversion of convertible preferred shares issuable upon exercise of a warrant	7,614	7,614
Common shares reserved for exercise of outstanding stock options under the 2017 Plan	—	660,319
Common shares reserved for future issuance under the 2017 Plan	1,087,832	427,513
	<u>16,205,396</u>	<u>16,205,396</u>

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

12. Equity-Based Compensation

Prior to the Reorganization, the Company's operating agreement, as amended and restated, provided for the granting of incentive units to employees, officers, directors and consultants under the 2013 Equity Incentive Plan (the "2013 Plan"), as determined by the Board of Directors. At December 31, 2016, 6,086,500 incentive units were authorized to be granted under the 2013 Plan.

2013 Equity Incentive Plan

The terms of the incentive units granted prior to the Reorganization were determined by the Board of Directors and included vesting, forfeiture, repurchase and other provisions. The Board of Directors also determined whether each incentive unit granted was a voting incentive unit, having the right to vote on any matter the common units had the right to vote on, or a non-voting incentive unit and not carry the right to vote. At December 31, 2016, there were 1,319,101 incentive units that were voting, and 439,151 incentive units that were non-voting, respectively. Incentive units had no rights to dividends and were entitled to distributions. Incentive unit holders were not required to purchase or "exercise" their incentive units in order to receive such distributions. However, distributions to incentive unit holders began only after the cumulative amount distributed to common unit holders exceeded the strike price with respect to such incentive unit. Distributions were entitled to be made to incentive unit holders whether vested or unvested. Unvested distributions were to be held by the Company until the incentive units vest, at which time they would be released to the incentive unit holder. The Board of Directors had the discretion to determine the extent to which an incentive unit would be excluded from participating in distributions. Unless otherwise approved by the Board of Directors, the incentive units generally vested over a four year period with the first 25% vesting following 12 months of employment or service and the remaining incentive units vesting in equal quarterly installments over the following 36 months. The incentive units had no contractual term.

In connection with the issuance of each incentive unit, the Board of Directors set a strike price based on the amount of distributions that the holders of a common unit would be entitled to receive in a hypothetical liquidation of the Company on the date of issuance of the incentive unit in which the Company sold its assets at fair market value, satisfied its liabilities and distributed the net proceeds to the holders of units in liquidation of the Company.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

12. Equity-Based Compensation (Continued)

A summary of the Company's incentive unit activity under the 2013 Plan and related information is as follows:

	Number of Units	Weighted Average Fair Value per Unit	Weighted Average Strike Price per Unit
Outstanding at December 31, 2016	1,758,252	\$ 2.03	\$ 1.09
Granted	621,497	3.63	2.63
Forfeited	(12,251)	2.86	2.23
Exchanged for common stock and restricted common stock pursuant to the Reorganization	(2,367,498)	2.43	1.49
Outstanding at December 31, 2017	<u> </u>		

The weighted average grant date fair value for incentive units granted in 2016 was \$2.80 per unit.

A summary of vested incentive units is as follows:

	Number of Units
Vested at December 31, 2016	635,898
Vesting through the date of the Reorganization	491,479
Vested as of the Reorganization	<u>1,127,377</u>

The total fair value of employee incentive units vested during 2016 and during 2017 through the date of the Reorganization was \$391,000 and \$1.1 million, respectively.

Incentive Unit Compensation Expense Assumptions

In 2017, the fair value of incentive units granted to both employees and non-employees was determined using the market approach, including the guideline public company method and a precedent transaction method which "backsolves" to a preferred price. Equity value was allocated to the common units, incentive units and convertible preferred units using either an option-pricing method ("OPM"), or a hybrid method, which is a hybrid between the OPM and the probability-weighted expected return method. The OPM treats common securities and preferred securities as call options on the total equity value of a company, with exercise prices based on the value thresholds at which the allocation among the various holders of a company's securities changes. Under this method, the common and incentive units have value only if the funds available for distribution to members exceed the value of the preferred security liquidation preference at the time of the liquidity event, such as a strategic sale or a merger. The hybrid method estimates the probability-weighted value across multiple scenarios but uses the OPM to estimate the allocation of value.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

12. Equity-Based Compensation (Continued)

within at least one of the scenarios. In addition to the OPM, the hybrid method considers an IPO scenario in which preferred shares are assumed to convert to common stock. The future value of the incentive units in the IPO scenario is discounted back to the valuation date at an appropriate risk adjusted discount rate. In the hybrid method, the present value indicated for each scenario is probability weighted to arrive at an indication of value for the common and incentive units.

The following assumptions were used in determining the fair value of incentive units granted to both employees and non-employees during 2016 and 2017:

	2016	2017
Risk-free interest rate	1.61%	1.48% - 1.97%
Expected dividend yield	—	—
Expected term (years to liquidity)	3.61	2.63 - 3.23
Expected volatility	69%	71% - 77%

The Company determined the risk-free rate based on constant maturing U.S. Treasury rates commensurate with the expected term, not seasonally adjusted. The Company has never paid, and does not anticipate paying, any cash dividends in the foreseeable future, and therefore uses an expected dividend yield of zero. Incentive units do not have an explicit contractual term. The Company, therefore, based its assumption on a weighted average of various liquidation scenarios, which would require a distribution to the incentive units. Since the Company was privately held as of the date of these financial statements, it does not have relevant historical data to support its expected volatility. As such, the Company has used a weighted-average of expected volatility based on the volatilities of a representative group of publicly traded biopharmaceutical companies. For purposes of identifying representative companies, the Company considered characteristics such as stage of development and area of therapeutic focus. Forfeitures of incentive units are accounted for when they occur, pursuant to the Company's early adoption of ASU 2016-09. In 2016 and 2017, the Company recorded a reduction in compensation expense for forfeitures of incentive units of \$18,000 and \$15,000, respectively, in accordance with ASU 2016-09. No prior periods were retrospectively adjusted as the impact of this change in accounting was immaterial to the financial statements.

Incentive Units Granted to Non-Employees

During the years ended December 31, 2016 and 2017, the Company granted incentive units to non-employees. The Company valued these incentive units based on their fair value on the date of grant.

The unvested incentive units granted to non-employees have been re-measured using the Company's estimate of fair value at each vesting date through the remaining vesting period. Equity-based compensation expense of \$22,000 and \$60,000 was recorded for the years ended December 31, 2016 and 2017, respectively relating to non-employee incentive unit awards.

Compensation Expense related to Incentive Units

The Company recorded equity-based compensation expense for incentive units granted to employees, directors and non-employees of \$556,000 and \$1.1 million for the years ended December 31, 2016 and

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

12. Equity-Based Compensation (Continued)

2017, respectively and \$257,000 for the three months ended March 31, 2017. There is no comparable amount recognized for the three months ended March 31, 2018 as the incentive units converted to common stock and restricted stock upon the Reorganization.

2017 Stock Option and Incentive Plan

The 2017 Plan provides for the grant of incentive stock options, non-qualified stock options, restricted stock awards, unrestricted stock awards and restricted stock units. The 2017 Plan is administered by the Board of Directors, or at the discretion of the Board of Directors, by a committee of the board comprised of not less than two directors. The exercise prices, vesting and other restrictions are determined at the discretion of the Board of Directors, or their committee if so delegated, except that the exercise price per share of stock options may not be less than 100% of the fair market value of the share of common stock on the date of grant and the term of stock option may not be greater than ten years. Stock options granted under the 2017 Plan to employees generally vest over four years. The number of shares initially reserved for issuance under the 2017 Plan was 3,455,330 shares of common stock. The shares of common stock underlying any awards that are forfeited, cancelled, repurchased or are otherwise terminated by the Company under the 2017 Plan will be added back to the shares of common stock available for issuance under the 2017 Plan.

As of December 31, 2017 and March 31, 2018, 3,455,330 shares were authorized for issuance under the 2017 Plan. As of December 31, 2017 and March 31, 2018, there were 1,087,832 and 427,513 shares, respectively, available for future issuance under the 2017 Plan.

Reorganization

As part of the Reorganization, the Company terminated the 2013 Plan and instituted the 2017 Plan.

Pursuant to the Reorganization, all vested and unvested incentive units granted under the 2013 Plan which were outstanding immediately prior to the Reorganization, irrespective of any strike price or voting rights on any such outstanding incentive units, were exchanged for an equal number of shares of common stock or restricted common stock, respectively, under the 2017 Plan. The restricted common stock was issued with the same vesting terms as the unvested incentive units held immediately prior to the Reorganization.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

12. Equity-Based Compensation (Continued)

The following table summarizes the common stock and restricted common stock activity under the 2017 Plan:

	Number of Shares	Weighted Average Fair Value per Share at Issuance
Common stock and restricted common stock issued as part of the Reorganization	2,367,498	\$ 5.77
Vested as of and after the Reorganization	1,147,413	\$ 5.77
Restricted common stock as of December 31, 2017	1,220,085	\$ 5.77
Granted	—	\$ 5.77
Vested	105,996	\$ 5.77
Forfeited	—	\$ 5.77
Restricted common stock as of March 31, 2018	1,114,089	\$ 5.77

The Company accounted for the exchange of incentive units in Scholar Rock, LLC for common stock and restricted common stock of Scholar Rock Holding Corporation, as a modification in accordance with the requirements of ASC 718. The Company determined the fair value of the common stock and restricted common stock using the market approach, including the guideline public company method and the precedent transaction method which "backsolves" to a preferred price. Accordingly, the Company determined there was an excess fair value of the replacement awards over the fair value of the incentive units exchanged in connection with the Reorganization, which resulted in incremental compensation expense of \$1.4 million of which \$423,000 was recognized in 2017 and \$84,000 was recognized during the three months ending March 31, 2018. The incremental fair value related to vested awards was recognized immediately as compensation expense. The incremental fair value of unvested awards and any remaining unrecognized compensation of the original awards will be recognized as compensation expense over the remaining vesting period.

The aggregate fair value of restricted stock awards that vested subsequent to the Reorganization during the year ended December 31, 2017 and during the three months ended March 31, 2018, based on estimated fair values of stock underlying the restricted stock awards on the date of vesting was \$116,000 and \$653,000, respectively.

Restricted Stock Granted to Non-Employees

The unvested incentive units granted to non-employees that converted into restricted stock upon the Reorganization have been re-measured using the Company's estimate of fair value at each vesting date through the remaining vesting period. The Company recorded equity-based compensation expense of \$26,000 for the three months ended March 31, 2018, for the non-employee restricted stock awards. There

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

12. Equity-Based Compensation (Continued)

were no comparable amounts recognized for the years ended December 31, 2016, 2017 or for the three months ended March 31, 2017.

Compensation Expense related to Restricted Stock

The Company recorded equity-based compensation expense for restricted stock granted to employees, directors and non-employees of \$423,000 and \$423,000 for the year ended December 31, 2017 and for the three months ended March 31, 2018, which is comprised of the original grant date fair value associated with the incentive units that converted to restricted stock upon Reorganization and the incremental compensation expense associated with the exchange of incentive units for restricted stock awards. There were no comparable amounts recognized for the year ended December 31, 2016 or for the three months ended March 31, 2017.

As of March 31, 2018, the Company had unrecognized equity-based compensation expense of \$4.0 million related to restricted stock issued to employees and directors, which is expected to be recognized over a period of 2.7 years.

Stock Options

The Company granted stock option awards under the 2017 Plan during the three months ended March 31, 2018. The following table summarizes the Company's stock option activity under the 2017 Plan:

	Number of Shares	Weighted Average Exercise Price	Weighted Average Remaining Contractual Term (in years)	Aggregate Intrinsic Value (in thousands)
Outstanding as of December 31, 2017	—	\$ —	—	\$ —
Granted	660,319	5.77	—	—
Exercised	—	—	—	—
Forfeited	—	—	—	—
Outstanding as of March 31, 2018	<u>660,319</u>	<u>5.77</u>	<u>9.9</u>	<u>\$ 924</u>
Options exercisable as of March 31, 2018	—	—	—	—

The weighted average grant-date fair value per share of stock options granted to employees and directors for stock option awards with service-based vesting conditions during the three months ended March 31, 2018 was \$4.20 per share. The weighted average grant-date fair value per share of stock options granted to employees and directors with performance-based vesting conditions and to non-employees with service-based vesting conditions during the three months ended March 31, 2018 was \$4.85 per share. The performance condition underlying the performance-based award is not probable of achievement as of March 31, 2018, therefore no compensation expense has been recorded for this award. The aggregate intrinsic value of stock options is calculated as the difference between the exercise price of the stock options and the fair value of the Company's common stock for those stock options that had exercise prices

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

12. Equity-Based Compensation (Continued)

lower than the fair value of the Company's common stock. There were no stock options exercised during the three months ended March 31, 2018.

Stock Option Valuation

The weighted average assumptions that the Company used to determine the fair value of the stock options granted to employees and directors during the three months ended March 31, 2018 were as follows:

Risk-free interest rate	2.73%
Expected dividend yield	0.00%
Expected term (in years)	6.13
Expected Volatility	84.66%

Stock Options Granted to Non-Employees

During the three months ended March 31, 2018, the Company granted stock options to non-employees. The Company valued these stock options based on their fair value on the date of grant. The unvested stock options granted to non-employees have been re-measured using the Company's estimate of fair value as of March 31, 2018, based on the following assumptions, presented on a weighted average basis:

Risk-free interest rate	2.74%
Expected dividend yield	0.00%
Expected term (in years)	9.89
Expected Volatility	84.23%

Equity-based compensation expense of \$2,000 was recorded for the three months ended March 31, 2018 relating to non-employee stock options, with no comparable amount for the years ended December 31, 2016 or 2017 or the three months ended March 31, 2017.

Compensation Expense related to Stock Options

The Company recorded equity-based compensation expense for stock options granted to employees, directors and non-employees of \$59,000 for the three months ended March 31, 2018, with no comparable amount in the years ended December 31, 2016 or 2017 or the three months ended March 31, 2017.

As of March 31, 2018, the Company had unrecognized equity-based compensation expense of \$2.6 million related to stock options issued to employees and directors, excluding performance-based awards for which vesting is not considered probable, which is expected to be recognized over a weighted average period of 3.9 years.

Performance-based Awards

The Company had granted incentive units, which contain both performance-based and service-based vesting criteria. Consistent with all incentive units, as part of the Reorganization, the incentive units granted with performance-based and service-based vesting criteria were exchanged for restricted common stock with the

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

12. Equity-Based Compensation (Continued)

same vesting terms. Equity-based compensation expense associated with performance-based awards is recognized if the performance condition is considered probable of achievement using management's best estimates. As of the Reorganization, all performance conditions had been achieved for incentive units granted with performance-based vesting criteria which converted into restricted stock upon Reorganization. The Company recorded expense related to the performance-based awards of \$101,000, \$91,000, \$15,000 and \$7,000 for the years ended December 31, 2016 and 2017 and the three months ended March 31, 2017 and 2018, respectively.

During the three months ended March 31, 2018, the Company granted a stock option award with a performance-based vesting criteria. The performance-based vesting criteria for this award is subject to milestone events specific to the Company's closing of an IPO. No equity-based compensation expense has been recognized for this award because the performance condition is not considered probable of achievement as of March 31, 2018.

Total Equity-based Compensation Expense

The Company recorded equity-based compensation expense related to all equity-based awards for employees and non-employees, which was allocated as follows in the consolidated statements of operations (in thousands):

	Year Ended December 31,		Three Months Ended March 31,	
	2016	2017	2017	2018
Research and development expense	\$ 264	\$ 704	\$ 108	\$ 245
General and administrative expense	292	778	149	237
	<u>\$ 556</u>	<u>\$ 1,482</u>	<u>\$ 257</u>	<u>\$ 482</u>

13. Income Taxes

The Company has not recorded a current or deferred tax provision for the years ended December 31, 2016 and 2017 or the three months ended March 31, 2017 or 2018.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

13. Income Taxes (Continued)

The effective income tax rate differed from the amount computed by applying the federal statutory rate to our loss before income taxes as follows:

	For Year Ended December 31,	
	2016	2017
Tax effected at statutory rate	34.0%	34.0%
State taxes	6.0	8.1
Stock compensation	(1.2)	(2.0)
Non-taxable income	0.5	0.3
Non deductible expenses	0.0	0.1
Impact of federal rate change on net deferred taxes	0.0	(27.0)
Federal research and development credits	1.2	2.3
Change in valuation allowance	(40.5)	(15.8)
	<u>0.0%</u>	<u>0.0%</u>

Deferred tax assets (liabilities) consist of the following at December 31, 2016 and 2017 (in thousands):

	As of December 31,	
	2016	2017
Long-term net deferred tax assets:		
Reserve and accruals	\$ 341	\$ 340
Net operating loss carryforwards	10,532	13,734
Deferred rent	355	190
Tax credits	847	1,665
Fixed and intangible assets	(235)	(128)
Total long-term net deferred tax assets:	<u>11,840</u>	<u>15,801</u>
Valuation allowance	(11,840)	(15,801)
Total net deferred tax assets	<u>\$ —</u>	<u>\$ —</u>

Total Net Deferred Tax Assets

Deferred tax assets are reduced by a valuation allowance if, based on the weight of available positive and negative evidence, it is more likely than not that some portion or all of the deferred tax assets will not be realized. Accordingly, a full valuation allowance has been established against the net deferred tax assets as of December 31, 2016 and 2017. For the years ended December 31, 2016 and 2017, the valuation allowance for deferred tax assets increased by \$6.6 million and \$4.0 million, respectively. This increase

SCHOLAR ROCK HOLDING CORPORATION**Notes to Consolidated Financial Statements (Continued)****(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)****13. Income Taxes (Continued)**

mainly relates to the establishment of valuation allowance against the Company's net domestic deferred tax assets in connection with net operating losses generated in each year, and the recording of additional net operating losses and credit carryforwards, partially offset by a revaluation of the federal deferred tax assets in 2017 based on the tax law change discussed below. As of December 31, 2017, the Company had \$50.4 million and \$49.8 million of Federal and state operating loss carryforwards respectively, which begin to expire in 2034. These loss carryforwards are available to reduce future taxable income, if any. In addition, as of December 31, 2017, the Company had \$1.1 million and \$0.7 million of Federal and state credit carryovers which begin to expire in 2034 and 2020, respectively. These loss and credit carryforwards are subject to review and possible adjustment by the appropriate taxing authorities. The amount of loss and credit carryforwards that may be utilized in any future period may be limited based upon changes in the ownership of the company's ultimate parent.

On December 22, 2017, the Tax Cuts and Jobs Act ("TCJA") was signed into law. The TCJA significantly revises the U.S. corporate income tax by, among other things, lowering corporate income tax rates, implementing a hybrid territorial tax system, and imposing a one-time repatriation tax on foreign cash and earnings. The Company has assessed the impact of this law change on the realization of its deferred tax assets and the remeasurement of the deferred tax assets and liabilities to the lower statutory federal US tax rate. The remeasurement of the deferred tax assets and liabilities did not result in a change to the current year income tax provision or balance sheet, as an offsetting adjustment was also recorded to the valuation allowance maintained on these accounts. The Company also had no investments in specified foreign corporations as of December 31, 2017. The Company's assessment of the TCJA is ongoing and assumptions and estimates may need to be revised based on new information available and as additional transition guidance is related by the IRS. The assessment is expected to be completed no later than the fourth quarter of calendar year 2018.

The Company follows the provisions of ASC Topic 740-10, "Accounting for Uncertainty in Income Taxes," which specifies how tax benefits for uncertain tax positions are to be recognized, measured, and recorded in financial statements; requires certain disclosures of uncertain tax matters; specifies how reserves for uncertain tax positions should be classified on the balance sheet; and provides transition and interim period guidance, among other provisions. As of December 31, 2016 and 2017 and March 31, 2018, the Company has not recorded any amounts for uncertain tax positions. The Company's policy is to recognize interest and penalties accrued on any uncertain tax positions as a component of income tax expense, if any, in its statements of income. For the years ended December 31, 2016 and 2017 and the three months ended March 31, 2017 and 2018, no estimated interest or penalties were recognized on uncertain tax positions. The Company has not yet conducted a study of its research and development credit carry forwards. Such a study may result in an adjustment to the Company's research and development credit carryforwards; however, until a study is completed and any adjustment is known, no amount is being presented as an uncertain tax position. A full valuation allowance has been provided against the Company's research and development credits, and, if an adjustment is required, this adjustment would be offset by an adjustment to the valuation allowance. Thus, there would be no impact to the balance sheet or statement of operations and comprehensive loss if an adjustment were required.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

14. Commitments and Contingencies

Operating LeasesOffice Equipment

The Company leases certain office equipment under non-cancelable operating leases. Total costs for such leases was \$11,000, \$11,000, \$3,000 and \$3,000 for the year ended December 31, 2016 and 2017 and three months ended March 31, 2017 and 2018, respectively. The future minimum lease payments for these leases as of December 31, 2017 and March 31, 2018 are as follows (in thousands):

Year ending December 31, 2018	Total Minimum Lease Payments	
	December 31, 2017	March 31, 2018
Total minimum lease payments	\$ 7	\$ 4
	\$ 7	\$ 4

Facility Lease

In March 2015, the Company entered into a 5-year facility lease for approximately 11,600 square feet of space at 620 Memorial Drive, Cambridge, Massachusetts, as amended in February 2016 (the "lease"). The lease was amended in February 2018, to add an additional 9,132 square feet (the "expansion space") at the current location and to extend the lease term (the "amended lease"). The amended lease is set to expire 5 years from the date the landlord delivers the expansion space which is expected to occur in the third quarter of 2018. Rent for the expansion space increases from \$667,000 a year to \$750,000 a year over the term of the lease. The Company is entitled to free rent for delays in the delivery of the expansion space beyond May 1, 2018. Based on the current expected delivery of the expansion space, the Company is entitled to approximately two months of free rent for the expansion space. The rent for the original space will increase to an amount based on the rental amount per square foot of the expansion space for the period beginning November 1, 2018 through the extended term of the lease. The landlord provided the Company with a tenant improvement allowance of \$91,000 for costs to perform alterations of the expansion space. Tenant improvement allowances are capitalized and recorded as deferred rent and amortized as a reduction to rent expense over the lease term. The Company can elect to receive an additional \$137,000 of tenant improvement allowances to be repaid as rent expense over the lease term at an interest rate of 8% per annum. The Company has the option to extend the term of the amended lease for one additional term of 5 years commencing after the amended lease expires.

The Company recognizes rent expense for the space it currently occupies and records a deferred rent obligation representing the cumulative difference between actual rent payments and rent expense recognized ratably over the lease period, which is included in the Company's consolidated balance sheets as of December 31, 2016 and 2017 and the three months ended March 31, 2018. The Company will begin recognizing expense for the expansion space, which the Company did not yet occupy as of March 31, 2018, upon taking possession of the space.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

14. Commitments and Contingencies (Continued)

Minimum annual rent payments under this lease for the remaining term of the amended lease, excluding operating expenses and taxes which are not fixed for future periods as of December 31, 2017 and March 31, 2018, are as follows (in thousands):

Year ending December 31,	Total Minimum Lease Payments	
	December 31, 2017	March 31, 2018
2018	\$ 689	\$ 740
2019	709	1,382
2020	604	1,447
2021	—	1,623
2022	—	1,672
2023	—	1,137
Total minimum lease payments	\$ 2,002	\$ 8,001

In accordance with the lease, the Company entered into a cash-collateralized irrevocable standby letter of credit in the amount of \$205,000 naming the landlord as beneficiary and the amount is included in restricted cash in the consolidated balance sheets. The Company has a single option to extend the lease by an additional five years at the greater of market rental rates or the base rent for the last rent year of the prior term.

The Company recorded approximately \$461,000, \$461,000, \$115,000 and \$140,000 in rent expense for the years ended December 31, 2016 and 2017 and the three months ended March 31, 2017 and 2018, respectively.

Legal Proceedings

The Company is not currently a party to any material legal proceedings.

15. Loan Payable

In August 2015, the Company entered into a Loan and Security Agreement with Silicon Valley Bank ("SVB"), which provided the Company an equipment line of credit of up to \$2.0 million to finance the purchase of eligible equipment. Pursuant to the agreement, SVB was obligated to make up to five equipment advances, each in an amount of at least \$100,000 during the draw period which began on the effective date, August 11, 2015, and ended on June 30, 2016. The Company borrowed \$677,000 against the line of credit as of December 31, 2015.

In August 2016, the Company entered into the First Amendment to the Loan and Security Agreement ("First Amendment"), which provided the Company an extension of the draw period to December 31, 2016. The Company borrowed \$1.3 million in 2016 against the line of credit, which fulfilled the maximum credit

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

15. Loan Payable (Continued)

line of \$2.0 million at December 31, 2016. The loan balance at December 31, 2017 and March 31, 2018 was \$1.0 million and \$0.9 million, respectively.

In addition, the First Amendment expanded the operating accounts provision to require the Company to, at all times, have on deposit in operating, depository and securities accounts maintained with SVB or SVB's affiliates, unrestricted and unencumbered cash in an amount equal to the lesser of (a) 105% of the then outstanding obligations or (b) 100% of the dollar value of the Company's accounts at all financial institutions. The Company was in compliance with this covenant at December 31, 2017 and March 31, 2018, which required the Company to hold a minimum of \$1.1 million and \$0.9 million in accounts with SVB, respectively.

Amounts borrowed bear interest at an annual prime rate as published in the Wall Street Journal less 0.25%, which, at December 31, 2016 and 2015 was 3.50% and 3.25%, respectively. For each advance, interest-only payments were due and paid through June 2016. Principal and interest payments commenced on July 1, 2016 for a period of 36 months. A final payment fee equal to 4% of the aggregate advances is also due on June 1, 2019. The final payment is being accrued over the term of the loan and is being recorded as interest expense.

Future principal payments on this loan as of December 31, 2017 and March 31, 2018 are as follows (in thousands):

Year ending December 31,	Total Future Principal Payments	
	December 31, 2017	March 31, 2018
2018	\$ 667	\$ 500
2019	365	365
	<u>\$ 1,032</u>	<u>\$ 865</u>

The Company incurred costs on behalf of the lender recorded as a debt discount of \$53,000 and incurred issuance costs recorded as deferred financing costs of \$19,000, both of which are recorded as a deduction from the carrying amount of the loan and are being amortized as interest expense over the term of the loan.

The Company has granted SVB a security interest in the equipment financed under the agreement. The Loan and Security Agreement contains negative covenants restricting the Company's activities, including limitations on dispositions, change in business ownership or location, mergers or acquisitions, incurring indebtedness or liens, paying dividends or making investments and certain other business transactions.

The Company has the option to prepay the loan and upon prepayment will pay the outstanding principal and interest, the final payment fee and the prepayment premium. The prepayment premium is equal to 1% of the then outstanding principal if made on or prior to the second anniversary and 0.5% of the principal balance if made after August 11, 2017. The Company evaluated the prepayment option ("Call Option") to

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

15. Loan Payable (Continued)

determine if the features should be separated from the loan and recognized as a derivative under ASC Topic 815, *Derivatives and Hedging*, ("ASC 815"), concluding that the Call Option is clearly and closely related to the Loan and does not meet the criteria for bifurcation from the loan.

In the event of a default, and during such an event, the interest rate will increase by 5% per year. The Company evaluated this increase to determine if it should be separated from the loan and recognized as a derivative. The Company determined that it met the requirements of ASC 815 in that a default could occur due to non-credit related matters. Therefore, the economic characteristics and risks are not closely related to that of the debt, and the interest rate feature requires bifurcation from the loan, however, the value associated with this embedded feature is *de minimis*, and thus not recorded at the issuance date through March 31, 2018.

The Company recorded total interest expense for this loan of \$42,000, \$101,000, \$26,000 and \$23,000 for the years ended December 31, 2016 and 2017 and three months ended March 31, 2017 and 2018, respectively.

16. Option and License Agreement

Overview

On December 17, 2013, the Company entered into an option and license agreement with Janssen. Under this agreement, the Company conducted drug discovery research to identify molecules with either one or two pharmacological profiles, and Janssen funded such research during a two-year period beginning on December 17, 2013. During the two-year period, the Company granted Janssen a research license to research, develop and use the collaboration molecule(s) and/or lead molecule(s) for use in the field and in the territory. Janssen was not granted a license to commercialize any collaboration molecule, lead molecule or licensed product unless and until Janssen exercised its license option in accordance with the agreement. The license option in the agreement provides Janssen an option to exclusively license molecules identified during the term that meet either one or both pharmacological profiles. If Janssen were not to exercise its license option by the end of this period, the term could be extended for up to one additional year by mutual written agreement of the parties. The activities under the agreement were governed by a program committee, which met quarterly and consisted of three members each from the Company and Janssen.

The Company received funding from Janssen based on a set rate per annual full-time equivalent personnel working on the research plus actual external costs incurred by the Company up to a maximum dollar amount as defined in the agreement. Costs approximated the funding provided. All amounts billed by the Company to Janssen were made quarterly, in arrears, based on time and actual costs incurred. There are no refund provisions in the agreement. Pursuant to the contract, if a molecule was not identified under either pharmacological profile, or Janssen did not exercise its option to the molecule(s) identified, the agreement would expire at the end of the term, December 17, 2015, unless extended by the parties.

At any time during the two-year collaboration period, Janssen held the right to exercise its license option for molecules with either or both pharmacological profiles by providing written notice to the Company and paying an option exercise fee of \$1.0 million per option exercised (up to two). Once Janssen exercises its option, the Company's obligations under the program plan for the molecule and related pharmacological profile cease and Janssen assumes full responsibility for further development of the molecules at its sole

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

16. Option and License Agreement (Continued)

cost. The Company is obligated to transfer any and all manufacturing related activities to Janssen at Janssen's cost. In addition, after Janssen exercises its option, it is obligated to pay the Company certain development milestones totaling up to \$25.0 million and regulatory milestones totaling up to \$97.0 million for each pharmacological profile as detailed in the agreement during the development period and through successful regulatory approval. Development milestones are triggered upon the achievement of specified development criteria or dosing of a specified number of patients in phases of clinical trials. Regulatory milestones are triggered upon approval to market a product candidate by the United States Food and Drug Administration ("FDA") or other global regulatory authorities. Additionally commercial milestone payments totaling up to \$130.0 million for each pharmacological profile are eligible to be earned as certain sales thresholds are achieved by Janssen and royalties are also required to be paid by Janssen to the Company based on annual net sales thresholds, based on Janssen's sales of a product derived from the collaboration molecule(s). The next potential milestone the Company may be entitled to receive under the agreement is a milestone payment of \$2.0 million upon achievement of a development milestone.

On December 15, 2015, Janssen delivered notice to the Company of its exercise of the license option for collaboration molecules for one of the pharmacological profiles, upon which the Company received \$1.0 million. In addition, the Company and Janssen also agreed to extend the collaboration period for the second pharmacological profile through March 31, 2016. The option exercise period for this profile expired unexercised on March 31, 2016.

Accounting Analysis

The Company has accounted for the agreement under ASC 606, which was adopted using the full retrospective transition method and the contract modification practical expedient. In applying the contract modification practical expedient, the Company has aggregated the effect of all modifications through the initial date of application of ASC 606 for the purposes of i) identifying the satisfied and unsatisfied performance obligations, ii) determining the transaction price and iii) allocating the transaction price to the satisfied and unsatisfied performance obligations.

The Company has concluded the performance obligations in the agreement include i) a non-exclusive research and development license and the related research services and ii) an exclusive license to commercialize any collaboration molecule, lead molecule, or licensed product for one of the pharmacological profiles. The services to be provided related to the program committee were determined to be immaterial to the overall arrangement and were not included as a performance obligation under the arrangement. In addition, the Company has concluded the unexercised license option at the date of adoption was a marketing offer as the option did not provide any discounts or other rights that would be considered a material right in the arrangement.

The Company determined that the research and development license was not distinct from the related research services to be provided by the Company as Janssen cannot obtain the benefit of the license without the related research services. The exclusive commercialization license is distinct from the research and development license and related services as the ability of Janssen to obtain the benefit of the exclusive commercialization license is not dependent upon the research and development license or research services. The research services for the pharmacological profile being licensed would be completed at the time of the exercise of the option.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

16. Option and License Agreement (Continued)

As of the date of adoption, the total estimated transaction price was \$4.9 million, which consisted of i) \$3.9 million of estimated cost reimbursement payments related to reimbursement of full time equivalents at the established rate and external costs incurred and ii) \$1.0 million of the option exercise fee related to the option exercised on December 15, 2015. The Company utilized the expected value approach to estimate the remaining amount of cost reimbursement payments. The final costs were incurred by March 31, 2016 upon completion of the research services and were consistent with the amount estimated at adoption. The Company utilizes the most likely amount approach to estimate any development and regulatory milestones. There are no milestone payments included in the estimated transaction price. The Company considered the stage of development required to achieve the milestones, as well as whether the achievement of the milestones is outside the control of the Company or Janssen. The milestone payments were fully constrained, as a result of the uncertainty whether any of the associated milestones would be achieved. The Company has determined that any commercial milestones and sales-based royalties will be recognized when the related sales occur as they were determined to relate predominantly to the license granted and therefore have been excluded from the transaction price. The Company will re-evaluate the transaction price in each reporting period and as uncertain events are resolved or other changes in circumstances occur. There have been no changes to the transaction price since the completion of the research services in the quarter ended March 31, 2016.

The amount of the transaction price related to the cost reimbursement payments was allocated solely to the research license and related services, as the amount of the payments is related to the satisfaction of the performance obligation and the amount allocated to the research and development license and related services is consistent with the amounts the Company would expect to receive for that performance obligation. Similarly, the amount of the transaction price related to the option exercise fee has been solely allocated to the exclusive commercialization license which was granted and recognized as revenue upon exercise of the option. Accordingly, the Company allocated the \$3.9 million of the transaction price to the research license and related services and \$1.0 million to the exclusive commercialization license.

During the year ended December 31, 2016, the Company recognized revenue of \$379,000 which was solely attributable to the research and development license and related research services performance obligation which was recognized as revenue as the associated services were performed based on the costs incurred using an input method of recognition. There was no revenue recognized in 2017 or 2018.

The Company had receivables due from Janssen on January 1, 2016 of \$380,000. There were no contract assets or contract liabilities as of January 1, 2016 or for any period presented.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

17. Net Loss per Unit and Share and Unaudited Pro Forma Net Loss per Share

Basic and diluted net loss per unit is calculated as follows (in thousands, except unit and per unit data):

	Year Ended December 31, 2016	Three Months Ended March 31, 2017
Net loss	\$ (16,207)	\$ (4,730)
Weighted average common units outstanding, basic and diluted	1,603,088	1,603,088
Net loss per unit, basic and diluted	\$ (10.11)	\$ (2.95)

Following the Reorganization, the Company calculates net loss per share based on its outstanding shares of common stock. For the year ended December 31, 2017, the weighted average number of common shares outstanding includes the weighted average number of common units outstanding prior to the Reorganization.

Basic and diluted net loss per share is calculated as follows (in thousands, except share and per share data):

	Year Ended December 31, 2017	Three Months Ended March 31, 2018
Net loss	\$ (24,995)	\$ (8,892)
Weighted average common shares outstanding, basic and diluted	1,634,100	2,795,497
Net loss per share, basic and diluted	\$ (15.30)	\$ (3.18)

The following table sets forth the outstanding common unit or common stock equivalents, presented based on amounts outstanding at each period end, that have been excluded from the calculation of diluted net

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

17. Net Loss per Unit and Share and Unaudited Pro Forma Net Loss per Share (Continued)

loss per unit or share for the periods indicated because their inclusion would have been anti-dilutive (in common unit or common stock equivalent shares, as applicable):

	Year Ended December 31,		Three Months Ended March 31,	
	2016	2017	2017	2018
Convertible preferred units	10,036,481	—	10,036,481	—
Convertible preferred stock	—	15,109,950	—	15,109,950
Incentive units	1,758,252	—	1,911,033	—
Restricted common stock	—	1,220,085	—	1,114,089
Warrant	7,614	7,614	7,614	7,614
Stock options	—	—	—	660,319
	<u>11,802,347</u>	<u>16,337,649</u>	<u>11,955,128</u>	<u>16,891,972</u>

Unaudited pro forma basic and diluted net loss per share is calculated as follows (in thousands, except share and per share data):

	Year Ended	Three Months
	December 31, 2017	Ended March 31, 2018
Numerator:		
Net loss	\$ (24,995)	\$ (8,892)
Change in fair value of warrant liability	10	20
Equity-based compensation expense for stock-based award with vesting conditions contingent upon initial public offering	—	(42)
Pro forma net loss	<u>\$ (24,985)</u>	<u>\$ (8,914)</u>
Denominator:		
Weighted average common shares outstanding, basic and diluted	1,634,100	2,795,497
Pro forma adjustment for the automatic conversion of all outstanding shares of convertible preferred stock into shares of common stock	10,474,277	15,109,950
Pro forma weighted average common shares outstanding, basic and diluted	<u>12,108,377</u>	<u>17,905,447</u>
Pro forma net loss per share, basic and diluted	<u>\$ (2.06)</u>	<u>\$ (0.50)</u>

18. Retirement Plan

The Company sponsors a 401(K) retirement plan, in which substantially all of its full-time employees are eligible to participate. Participants may contribute a percentage of their annual compensation to this plan.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

18. Retirement Plan (Continued)

subject to statutory limitations. The Company did not provide any contributions to this plan during the years ended December 31, 2016 and 2017 or during the three months ended March 31, 2017 and 2018.

19. Deferred Payroll Tax Credit

In December 2015, the Protecting Americans from Tax Hikes (PATH) Act of 2015 was signed into law, which created several new research and development ("R&D") tax credit provisions, including allowing qualified small businesses to utilize the R&D tax credit against the employer's portion of payroll tax up to a maximum of \$250,000 per year. This provision is available for R&D tax credits generated in tax years beginning after 2015. The Company qualified as a small business under PATH for both 2016 and 2017, and has elected to apply the maximum \$250,000 for each of the 2016 R&D tax credit and the 2017 R&D tax credit generated against future employer payroll tax liabilities. The \$250,000 benefit was recorded as a reduction of research and development costs for both of the years ended December 31, 2016 and 2017. The R&D tax credit of \$95,000, \$399,000 and \$406,000 is recorded in prepaid expenses and other current assets as of December 31, 2016 and 2017 and March 31, 2018, respectively. The R&D tax credit of \$155,000 and \$50,000 is recorded in other long term assets as of December 31, 2016 and 2017, respectively. No comparable amount is recorded in other long term assets as of March 31, 2018.

20. Related Party Transactions

Licensing Agreement

Pursuant to a license agreement with Children's Medical Center Corporation ("CMCC"), a common share holder, the Company paid CMCC an annual license maintenance fee of \$5,000 in 2016. Beginning in 2017, this obligation increased to \$10,000 per year, and continues until the agreement is terminated. The Company will also be responsible for up to \$1.3 million of development milestone payments through the first regulatory approval of a licensed product, tiered royalty payments of low single-digit percentages on net sales of licensed product in the event that the Company realizes sales from products covered by the license agreement, which are products that the Company develops using its proprietary platform, and between 10% to 20% of non-royalty income attributable to a sublicense of the CMCC rights. The Company recorded research and development expense in the statements of operations of \$5,000 and \$10,000 for the years ended December 31, 2016 and 2017, respectively. There are no comparable expense amounts for the three months ended March 31, 2017 and 2018. There are no amounts due at December 31, 2016, 2017 or March 31, 2018.

Consulting Agreements

The Company entered into consulting agreements on October 10, 2012 with its two scientific co-founders to provide services related to the advancement of the research and development platform of the company.

The consulting arrangements are on a fixed-fee basis, paid quarterly. The initial contract terms were four years and terminated on October 10, 2016. The contracts were extended for an additional four year period. The Company incurred \$160,000 of consulting expense related to these contracts, in each year, for the years ended December 31, 2016 and 2017. The Company incurred \$40,000 of consulting expense related to these contracts, for the three months ended March 31, 2017 and 2018. There are no amounts due at December 31, 2016, December 31, 2017 or March 31, 2018.

SCHOLAR ROCK HOLDING CORPORATION

Notes to Consolidated Financial Statements (Continued)

(Information as of March 31, 2018 and for the three months ended March 31, 2017 and 2018 is unaudited)

21. Subsequent Events

On April 3, 2018, the Company's Board of Directors authorized 280,230 additional shares of common stock for future issuance under the 2017 Plan.

On April 3, 2018, the Company granted stock options to purchase 324,014 shares of common stock at an exercise price of \$7.17 per share under the 2017 Plan.

On April 15, 2018, the Company granted stock options to purchase 31,525 shares of common stock at an exercise price of \$7.17 per share under the 2017 Plan.

22. Subsequent Event — Reverse Stock Split

The Company's Board of Directors and stockholders approved a 2.8548-to-one reverse stock split of the Company's issued and outstanding shares of common stock that became effective on May 11, 2018. All unit, per unit, share and per share amounts in the consolidated financial statements and notes thereto have been retrospectively adjusted for all periods presented to give effect of the reverse stock split.

5,360,000 Shares



SCHOLAR ROCK
Scholar Rock Holding Corporation

Common Stock

PRELIMINARY PROSPECTUS

Joint Book-Running Managers

Jefferies
Cowen
BMO Capital Markets

Co-Manager

Wedbush PacGrow

Until _____, 2018 (25 days after the date of this prospectus), all dealers that effect transactions in these securities, whether or not participating in this offering, may be required to deliver a prospectus. This is in addition to the dealers' obligation to deliver a prospectus when acting as underwriters and with respect to their unsold allotments or subscriptions.

PART II

Information Not Required in Prospectus

Item 13. Other Expenses of Issuance and Distribution.

The following table sets forth the fees and expenses, other than underwriting discounts and commissions, payable in connection with the registration of the common stock hereunder. All amounts are estimates except the SEC registration fee.

	Amount to be Paid
SEC registration fee	\$ 11,511
FINRA filing fee	14,369
Nasdaq Global Market listing fee	125,000
Printing and mailing	195,000
Legal fees and expenses	1,000,000
Accounting fees and expenses	800,000
Transfer agent and registrar fees and expenses	3,500
Miscellaneous	150,620
Total	\$ 2,300,000

Item 14. Indemnification of Directors and Officers.

Section 145 of the Delaware General Corporation Law or the DGCL, authorizes a corporation to indemnify its directors and officers against liabilities arising out of actions, suits and proceedings to which they are made or threatened to be made a party by reason of the fact that they have served or are currently serving as a director or officer to a corporation. The indemnity may cover expenses (including attorneys' fees) judgments, fines and amounts paid in settlement actually and reasonably incurred by the director or officer in connection with any such action, suit or proceeding. Section 145 permits corporations to pay expenses (including attorneys' fees) incurred by directors and officers in advance of the final disposition of such action, suit or proceeding. In addition, Section 145 provides that a corporation has the power to purchase and maintain insurance on behalf of its directors and officers against any liability asserted against them and incurred by them in their capacity as a director or officer, or arising out of their status as such, whether or not the corporation would have the power to indemnify the director or officer against such liability under Section 145.

We have adopted provisions in our certificate of incorporation to be in effect upon the completion of this offering and by-laws to be in effect upon the effectiveness of this registration statement that limit or eliminate the personal liability of our directors to the fullest extent permitted by the DGCL, as it now exists or may in the future be amended. Consequently, a director will not be personally liable to us or our stockholders for monetary damages or breach of fiduciary duty as a director, except for liability for:

- § any breach of the director's duty of loyalty to us or our stockholders;
- § any act or omission not in good faith or that involves intentional misconduct or a knowing violation of law;
- § any unlawful payments related to dividends or unlawful stock purchases, redemptions or other distributions; or
- § any transaction from which the director derived an improper personal benefit.

These limitations of liability do not alter director liability under the federal securities laws and do not affect the availability of equitable remedies such as an injunction or rescission.

In addition, our by-laws provide that:

- § we will indemnify our directors, officers and, in the discretion of our board of directors, certain employees to the fullest extent permitted by the DGCL, as it now exists or may in the future be amended; and
- § we will advance reasonable expenses, including attorneys' fees, to our directors and, in the discretion of our board of directors, to our officers and certain employees, in connection with legal proceedings relating to their service for or on behalf of us, subject to limited exceptions.

We have entered into indemnification agreements with each of our directors and intend to enter into such agreements with certain of our executive officers. These agreements provide that we will indemnify each of our directors, certain of our executive officers and, at times, their affiliates to the fullest extent permitted by Delaware law. We will advance expenses, including attorneys' fees (but excluding judgments, fines and settlement amounts), to each indemnified director, executive officer or affiliate in connection with any proceeding in which indemnification is available and we will indemnify our directors and officers for any action or proceeding arising out of that person's services as a director or officer brought on behalf of us or in furtherance of our rights. Additionally, certain of our directors or officers may have certain rights to indemnification, advancement of expenses or insurance provided by their affiliates or other third parties, which indemnification relates to and might apply to the same proceedings arising out of such director's or officer's services as a director referenced herein. Nonetheless, we have agreed in the indemnification agreements that our obligations to those same directors or officers are primary and any obligation of such affiliates or other third parties to advance expenses or to provide indemnification for the expenses or liabilities incurred by those directors are secondary.

We also maintain general liability insurance which covers certain liabilities of our directors and officers arising out of claims based on acts or omissions in their capacities as directors or officers, including liabilities under the Securities Act of 1933, as amended, or the Securities Act.

The underwriting agreement filed as Exhibit 1.1 to this registration statement provides for indemnification of us and our directors and officers by the underwriters against certain liabilities under the Securities Act and the Securities Exchange Act of 1934.

Item 15. Recent Sales of Unregistered Securities.

In the three years preceding the filing of this registration statement, we have issued the following securities that were not registered under the Securities Act:

(a) Reorganization and Issuance of Convertible Preferred Stock

Reorganization

In connection with the Reorganization:

- § Holders of Scholar Rock, LLC outstanding Series B convertible preferred units received one share of Scholar Rock Holding Corporation Series B convertible preferred stock for each Series B convertible preferred unit held immediately prior to the Reorganization, with an aggregate of 13,526,994 shares of Scholar Rock Holding Corporation Series B convertible Preferred stock issued in the Reorganization;
- § Holders of Scholar Rock, LLC outstanding Series A-4 convertible preferred units received one share of Scholar Rock Holding Corporation Series A-4 convertible preferred stock for each Series A-4 convertible preferred unit held immediately prior to the Reorganization, with an aggregate of 3,906,738 shares of Scholar Rock Holding Corporation Series A-4 convertible preferred stock issued in the Reorganization;

- § Holders of Scholar Rock, LLC outstanding Series A-3 convertible preferred units received one share of Scholar Rock Holding Corporation Series A-3 convertible preferred stock for each Series A-3 convertible preferred unit held immediately prior to the Reorganization, with an aggregate of 5,579,709 shares of Scholar Rock Holding Corporation Series A-3 convertible preferred stock issued in the Reorganization;
- § Holders of Scholar Rock, LLC outstanding Series A-2 convertible preferred units received one share of Scholar Rock Holding Corporation Series A-2 convertible preferred stock for each Series A-2 convertible preferred unit held immediately prior to the Reorganization, with an aggregate of 5,066,915 shares of Scholar Rock Holding Corporation Series A-2 convertible preferred stock issued in the Reorganization;
- § Holders of Scholar Rock, LLC outstanding Series A-1 convertible preferred units received one share of Scholar Rock Holding Corporation Series A-1 convertible preferred stock for each Series A-1 convertible preferred unit held immediately prior to the Reorganization, with an aggregate of 2,000,000 shares of Scholar Rock Holding Corporation Series A-1 convertible preferred stock issued in the Reorganization;
- § Holders of Scholar Rock, LLC outstanding common units received one share of Scholar Rock Holding Corporation common stock for each outstanding common unit held immediately prior to the Reorganization, which were subject to a subsequent 2.8548-to-one reverse split with an aggregate of 1,603,088 shares of common stock; and
- § Holders of Scholar Rock, LLC vested and unvested incentive units, irrespective of any strike price or voting rights on any such outstanding incentive units, exchanged one unit for one share of common stock or restricted common stock, respectively, which were subject to a subsequent 2.8548-to-one reverse split. The restricted common stock was issued with the same vesting terms as the unvested incentive units held immediately prior to the Reorganization. An aggregate of 2,367,498 shares of restricted common stock and common stock were issued to the prior holders of incentive units. The restricted common stock was issued with the same vesting terms as the incentive units held immediately prior the Reorganization.

Issuances of Capital Stock

In December 2017, we issued and sold an aggregate of 13,055,555 shares of Series C convertible preferred stock at a purchase price of \$3.60 per share, for an aggregate purchase price of \$47.0 million to Artal International SCA, Redmile Capital Fund, LP, Redmile Biopharma Investments I, L.P. (and affiliates), Fidelity Mt. Vernon Street Trust: Fidelity Series Growth Company Fund (and affiliates), Cormorant Private Healthcare Fund I, LP (and affiliates), ARCH Venture Fund VIII, L.P., Polaris Venture Partners VI, L.P. (and affiliates), Timothy A. Springer, Ph.D, EcoR1 Capital Fund, LP (and affiliates), KPC Venture Capital, LLC, and JAKII, LLC.

In May 2017, we issued and sold an aggregate of 1,428,209 shares of Series B convertible preferred units at a purchase price of \$3.00 per unit, for an aggregate purchase price of \$4.3 million to Timothy A. Springer, Ph.D. Series B convertible preferred units converted on a one-to-one basis for Series B convertible preferred stock in connection with the Reorganization.

In December 2015, we issued and sold an aggregate of 12,098,785 Series B convertible preferred units at a purchase price of \$3.00 per unit, for an aggregate purchase price of \$36.3 million to Fidelity Advisor Biotechnology Fund and affiliates, Cormorant Global Healthcare Master Fund, LP, Arch Venture Fund VII, L.P., Polaris Venture Partners VI, L.P. (and affiliates), TAS Partners, LLC, EcoR1 Capital Fund, LP (and affiliates), KPC Venture Capital, LLC, and JAKII, LLC. Series B convertible preferred units converted on a one-to-one basis for Series B convertible preferred stock in connection with the Reorganization.

No underwriters were involved in the foregoing sales of securities. Unless otherwise stated, the sales of securities described above were deemed to be exempt from registration pursuant to Section 4(a)(2) of the Securities Act, including Regulation D and Rule 506 promulgated thereunder, as transactions by an issuer

not involving a public offering. All of the purchasers in these transactions represented to us in connection with their purchase that they were acquiring the securities for investment and not distribution, that they could bear the risks of the investment and could hold the securities for an indefinite period of time. Such purchasers received written disclosures that the securities had not been registered under the Securities Act and that any resale must be made pursuant to a registration or an available exemption from such registration. All of the foregoing securities are deemed restricted securities for the purposes of the Securities Act.

(b) Grants and Exercises of Stock Options

We have granted stock options to purchase an aggregate of 1,015,858 shares of our common stock, of which 660,319 have an exercise price of \$5.77 per share, and 355,539 have an exercise price of \$7.17 per share, to employees, directors and consultants pursuant to the 2017 Plan. No shares of common stock have been issued upon the exercise of stock options pursuant to the 2017 Plan.

The issuances of the securities described above were deemed to be exempt from registration pursuant to Section 4(a)(2) of the Securities Act or Rule 701 promulgated under the Securities Act as transactions pursuant to compensatory benefit plans. The shares of common stock issued upon the exercise of options are deemed to be restricted securities for purposes of the Securities Act.

Item 16. Exhibits and Financial Statement Schedules.

(a) Exhibits

Exhibit No.	Exhibit Index
1.1*	Form of Underwriting Agreement
3.1**	Amended and Restated Certificate of Incorporation of the Registrant, as currently in effect
3.1.1*	Amendment to Amended and Restated Certificate of Incorporation of the Registrant
3.2**	Form of Amended and Restated Certificate of Incorporation of the Registrant (to be effective upon the completion of this offering)
3.3**	By-laws of the Registrant, as currently in effect
3.4**	Form of Amended and Restated By-laws (to be effective upon the completion of this offering)
4.1**	Investors' Rights Agreement among the Registrant and certain of its stockholders, dated December 22, 2017
4.2*	Specimen Stock Certificate evidencing shares of common stock
4.3**	Amended and Restated Warrant to Purchase Stock, by and between Silicon Valley Bank and the Registrant, dated December 22, 2017
5.1*	Opinion of Goodwin Procter LLP
10.1#**	2017 Stock Option and Incentive Plan and forms of award agreements thereunder
10.2#*	2018 Stock Option and Incentive Plan and forms of award agreements thereunder
10.3#**	Senior Executive Cash Incentive Bonus Plan
10.4#*	Employee Stock Purchase Plan
10.5#*	Form of Indemnification Agreement
10.6†**	Exclusive License Agreement by and between the Registrant, and Children's Medical Center, dated as December 16, 2013
10.7#**	Offer Letter by and between Nagesh K. Mahanthappa, Ph.D. and the Registrant, dated October 10, 2012
10.8#**	Offer Letter by and between Yung H. Chyung, M.D. and the Registrant, dated February 2, 2016
10.9#**	Offer Letter by and between Elan Z. Ezickson and the Registrant, dated July 17, 2014
10.10#**	Non-Competition, Non-Solicitation, Confidentiality and Assignment Agreement, by Nagesh K. Mahanthappa, dated October 10, 2012
10.11#**	Non-Competition, Non-Solicitation, Confidentiality and Assignment Agreement, by Yung H. Chyung, M.D., dated February 2, 2016
10.12#**	Non-Competition, Non-Solicitation, Confidentiality and Assignment Agreement, by Elan Z. Ezickson, dated July 17, 2014
10.13†**	Option and License Agreement by and between the Registrant and Janssen Biotech, Inc., dated as of December 17, 2013

Exhibit No.	Exhibit Index
10.14**	Lease Agreement by and between 620 Memorial Leasehold LLC and the Registrant, dated March 5, 2015, as amended by the First Amendment dated February 22, 2016 and the Second Amendment dated February 22, 2018
10.15#*	Form of Employment Agreement to be entered into by and between Nagesh K. Mahanthappa, Ph.D., and the Registrant.
10.16#*	Form of Employment Agreement to be entered into by and between Rhonda M. Chicks, C.P.A., and the Registrant.
10.17#*	Form of Employment Agreement to be entered into by and between Yung H. Chyung, M.D., and the Registrant.
10.18#*	Form of Employment Agreement to be entered into by and between Efan Z. Ezickson, and the Registrant.
21.1**	Subsidiaries of the Registrant
23.1*	Consent of Ernst and Young LLP, Independent Registered Public Accounting Firm
23.2*	Consent of Goodwin Procter LLP (included in Exhibit 5.1)
24.1**	Power of Attorney (included on the signature page hereto)

* Filed herewith.

** Previously filed.

† Application has been made to the Securities and Exchange Commission for confidential treatment of certain provisions. Omitted material for which confidential treatment has been requested has been filed separately with the Securities and Exchange Commission.

Indicates a management contract or any compensatory plan, contract or arrangement.

(b) Financial Statements Schedules:

Schedules have been omitted because the information required to be set forth therein is not applicable or is shown in the financial statements or notes thereto.

Item 17. Undertakings.

Insofar as indemnification for liabilities arising under the Securities Act of 1933, as amended, or the Act, may be permitted to directors, officers and controlling persons of the Registrant pursuant to the foregoing provisions, or otherwise, the Registrant has been advised that in the opinion of the Securities and Exchange Commission such indemnification is against public policy as expressed in the Act and is therefore unenforceable. In the event that a claim for indemnification against such liabilities (other than the payment by the Registrant of expenses incurred or paid by a director, officer or controlling person of the Registrant in the successful defense of any action, suit or proceeding) is asserted by such director, officer or controlling person in connection with the securities being registered, the Registrant will, unless in the opinion of its counsel the matter has been settled by controlling precedent, submit to a court of appropriate jurisdiction the question whether such indemnification by it is against public policy as expressed in the Act and will be governed by the final adjudication of such issue.

The Registrant hereby undertakes that:

- (a) The Registrant will provide to the underwriter at the closing as specified in the underwriting agreement, certificates in such denominations and registered in such names as required by the underwriter to permit prompt delivery to each purchaser.
- (b) For purposes of determining any liability under the Act, as amended, the information omitted from a form of prospectus filed as part of this registration statement in reliance upon Rule 430A and contained in the form of prospectus filed by the Registrant pursuant to Rule 424(b)(1) or (4) or 497(h) under the Act, as amended, shall be deemed to be part of this registration statement as of the time it was declared effective.
- (c) For the purpose of determining any liability under the Act, as amended, each post-effective amendment that contains a form of prospectus shall be deemed to be a new registration statement relating to the securities offered therein, and the offering of such securities at that time shall be deemed to be the initial *bona fide* offering thereof.

Name

Title

Date

*

Timothy A. Springer, Ph.D.

Director

May 14, 2018

*By: /s/ NAGESH K. MAHANTHAPPA

Name: Nagesh K. Mahanthappa
Title: *Attorney-in-fact*

[-] Shares

Scholar Rock Holding Corporation

UNDERWRITING AGREEMENT

[-], 2018

JEFFERIES LLC
COWEN AND COMPANY, LLC
BMO CAPITAL MARKETS CORP.
As Representatives of the several Underwriters

c/o JEFFERIES LLC
520 Madison Avenue
New York, New York 10022

c/o COWEN AND COMPANY, LLC
599 Lexington Avenue
New York, New York 10022

c/o BMO CAPITAL MARKETS CORP.
3 Times Square, 25th Floor
New York, New York 10036

Ladies and Gentlemen:

Introductory. Scholar Rock Holding Corporation, a Delaware corporation (the “Company”), proposes to issue and sell to the several underwriters named in Schedule A (the “Underwriters”) an aggregate of [-] shares of its common stock, par value \$0.001 per share (the “Shares”). The [-] Shares to be sold by the Company are called the “Firm Shares.” In addition, the Company has granted to the Underwriters an option to purchase up to an additional [-] Shares as provided in Section 2. The additional [-] Shares to be sold by the Company pursuant to such option are collectively called the “Optional Shares.” The Firm Shares and, if and to the extent such option is exercised, the Optional Shares are collectively called the “Offered Shares.” Jefferies LLC (“Jefferies”), Cowen and Company, LLC (“Cowen”) and BMO Capital Markets Corp. (“BMO”) have agreed to act as representatives of the several Underwriters (in such capacity, the “Representatives”) in connection with the offering and sale of the Offered Shares. To the extent there are no additional underwriters listed on Schedule A, the term “Representatives” as used herein shall mean you, as Underwriters, and the term “Underwriters” shall mean either the singular or the plural, as the context requires.

The Company has prepared and filed with the Securities and Exchange Commission (the “Commission”) a registration statement on Form S-1, File No. 333-[-] which contains a form of prospectus to be used in connection with the public offering and sale of the Offered Shares. Such registration statement, as amended, including the financial statements, exhibits and schedules thereto, in the form in which it became effective under the Securities Act of 1933, as amended, and the rules and regulations promulgated thereunder (collectively, the “Securities Act”), including any information deemed to be a part thereof at the time of effectiveness pursuant to Rule 430A under the Securities Act, is called the “Registration Statement.” Any registration statement filed by the Company pursuant to Rule 462(b) under the Securities Act in connection with the offer and sale of the Offered Shares is called the “Rule

462(b) Registration Statement,” and from and after the date and time of filing of any such Rule 462(b) Registration Statement the term “Registration Statement” shall include the Rule 462(b) Registration Statement. The prospectus, in the form first used by the Underwriters to confirm sales of the Offered Shares or in the form first made available to the Underwriters by the Company to meet requests of purchasers pursuant to Rule 173 under the Securities Act, is called the “Prospectus.” The preliminary prospectus dated [-], 2018 describing the Offered Shares and the offering thereof is called the “Preliminary Prospectus,” and the Preliminary Prospectus and any other prospectus in preliminary form that describes the Offered Shares and the offering thereof and is used prior to the filing of the Prospectus is called a “preliminary prospectus.” As used herein, “Applicable Time” is [-] a.m. / [-] p.m. (New York City time) on [-], 2018. As used herein, “free writing prospectus” has the meaning set forth in Rule 405 under the Securities Act, and “Time of Sale Prospectus” means the Preliminary Prospectus together with the free writing prospectuses, if any, identified in Schedule B hereto. As used herein, “Road Show” means a “road show” (as defined in Rule 433 under the Securities Act) relating to the offering of the Offered Shares contemplated hereby that is a “written communication” (as defined in Rule 405 under the Securities Act). As used herein, “Section 5(d) Written Communication” means each written communication (within the meaning of Rule 405 under the Securities Act) that is made in reliance on Section 5(d) of the Securities Act by the Company or any person authorized to act on behalf of the Company to one or more potential investors that are qualified institutional buyers (“QIBs”) and/or institutions that are accredited investors (“IAIs”), as such terms are respectively defined in Rule 144A and Rule 501(a) under the Securities Act, to determine whether such investors might have an interest in the offering of the Offered Shares; “Section 5(d) Oral Communication” means each oral communication, if any, made in reliance on Section 5(d) of the Securities Act by the Company or any person authorized to act on behalf of the Company made to one or more QIBs and/or one or more IAIs to determine whether such investors might have an interest in the offering of the Offered Shares; “Marketing Materials” means any materials or information provided to investors by, or with the approval of, the Company in connection with the marketing of the offering of the Offered Shares, including any roadshow or investor presentations made to investors by the Company (whether in person or electronically); and “Permitted Section 5(d) Communication” means the Section 5(d) Written Communication(s) and Marketing Materials listed on Schedule C attached hereto.

All references in this Agreement to (i) the Registration Statement, any preliminary prospectus (including the Preliminary Prospectus), or the Prospectus, or any amendments or supplements to any of the foregoing, or any free writing prospectus, shall include any copy thereof filed with the Commission pursuant to its Electronic Data Gathering, Analysis and Retrieval System (“EDGAR”) and (ii) the Prospectus shall be deemed to include any “electronic Prospectus” provided for use in connection with the offering of the Offered Shares as contemplated by Section 3(n) of this Agreement.

In the event that the Company has only one subsidiary, then all references herein to “subsidiaries” of the Company shall be deemed to refer to such single subsidiary, *mutatis mutandis*.

The Company hereby confirms its agreements with the Underwriters as follows:

Section 1. Representations and Warranties. The Company hereby represents, warrants and covenants to each Underwriter, as of the date of this Agreement, as of the First Closing Date (as defined in Section 2) and as of each Option Closing Date (as defined in Section 2), if any, as follows:

(a) **Compliance with Registration Requirements.** The Registration Statement has become effective under the Securities Act. The Company has complied, to the Commission’s satisfaction with all requests of the Commission for additional or supplemental information, if any. No stop order suspending the effectiveness of the Registration Statement is in effect and no proceedings for such purpose have been

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instituted or are pending or, to the best knowledge of the Company, are contemplated or threatened by the Commission.

(b) **Disclosure.** Each preliminary prospectus and the Prospectus when filed complied in all material respects with the Securities Act and, if filed by electronic transmission pursuant to EDGAR, was identical (except as may be permitted by Regulation S-T under the Securities Act) to the copy thereof delivered to the Underwriters for use in connection with the offer and sale of the Offered Shares. Each of the Registration Statement and any post-effective amendment thereto, at the time it became or becomes effective, complied and will comply in all material respects with the Securities Act and did not and will not contain any untrue statement of a material fact or omit to state a material fact required to be stated therein or necessary to make the statements therein not misleading. As of the Applicable Time, the Time of Sale Prospectus (including any preliminary prospectus wrapper) did not, and at the First Closing Date and at each applicable Option Closing Date, will not, contain any untrue statement of a material fact or omit to state a material fact necessary to make the statements therein, in the light of the circumstances under which they were made, not misleading. The Prospectus (including any Prospectus wrapper), as of its date, did not, and at the First Closing Date and at each applicable Option Closing Date, will not, contain any untrue statement of a material fact or omit to state a material fact necessary in order to make the statements therein, in the light of the circumstances under which they were made, not misleading. The representations and warranties set forth in the three immediately preceding sentences do not apply to statements in or omissions from the Registration Statement or any post-effective amendment thereto, or the Prospectus or the Time of Sale Prospectus, or any amendments or supplements thereto, made in reliance upon and in conformity with written information relating to any Underwriter furnished to the Company in writing by the Representatives expressly for use therein, it being understood and agreed that the only such information consists of the information described in Section 9(b) below. There are no contracts or other documents required to be described in the Time of Sale Prospectus or the Prospectus or to be filed as an exhibit to the Registration Statement which have not been described or filed as required.

(c) **Free Writing Prospectuses; Road Show.** As of the determination date referenced in Rule 164(h) under the Securities Act, the Company was not, is not or will not be (as applicable) an “ineligible issuer” in connection with the offering of the Offered Shares pursuant to Rules 164, 405 and 433 under the Securities Act. Each free writing prospectus that the Company is required to file pursuant to Rule 433(d) under the Securities Act has been, or will be, filed with the Commission in accordance with the requirements of the Securities Act. Each free writing prospectus that the Company has filed, or is required to file, pursuant to Rule 433(d) under the Securities Act or that was prepared by or on behalf of or used or referred to by the Company complies or will comply in all material respects with the requirements of Rule 433 under the Securities Act, including timely filing with the Commission or retention where required and legending, and each such free writing prospectus, as of its issue date and at all subsequent times through the completion of the public offer and sale of the Offered Shares did not, does not and will not include any information that conflicted, conflicts or will conflict with the information contained in the Registration Statement, the Prospectus or any preliminary prospectus and not superseded or modified. Except for the free writing prospectuses, if any, identified in Schedule B, and electronic road shows, if any, furnished to you before first use, the Company has not prepared, used or referred to, and will not, without your prior written consent, which consent shall not be unreasonably withheld, prepare, use or refer to, any free writing prospectus. Each Road Show, when considered together with the Time of Sale Prospectus, did not, as of the Applicable Time, contain any untrue statement of a material fact or omit to state a material fact necessary to make the statements therein, in the light of the circumstances under which they were made, not misleading.

(d) **Distribution of Offering Material By the Company.** Prior to the latest of (i) the expiration or termination of the option granted to the several Underwriters in Section 2, (ii) the completion of the Underwriters’ distribution of the Offered Shares and (iii) the expiration of twenty-five (25) days

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after the date of the Prospectus, the Company has not distributed and will not distribute any offering material in connection with the offering and sale of the Offered Shares other than the Registration Statement, the Time of Sale Prospectus, the Prospectus or any free writing prospectus reviewed and consented to by the Representatives, which consent shall not be unreasonably withheld, the free writing prospectuses, if any, identified on Schedule B hereto and any Permitted Section 5(d) Communications.

(e) **The Underwriting Agreement.** This Agreement has been duly authorized, executed and delivered by the Company.

(f) **Authorization of the Offered Shares.** The Offered Shares have been duly authorized for issuance and sale pursuant to this Agreement and, when issued and delivered by the Company against payment therefor pursuant to this Agreement, will be validly issued, fully paid and nonassessable, and the issuance and sale of the Offered Shares is not subject to any preemptive rights, rights of first refusal or other similar rights to subscribe for or purchase the Offered Shares.

(g) **No Applicable Registration or Other Similar Rights.** There are no persons with registration or other similar rights to have any equity or debt securities registered for sale under the Registration Statement or included in the offering contemplated by this Agreement, except for such rights as have been duly waived.

(h) **No Material Adverse Change.** Except as otherwise disclosed in the Registration Statement, the Time of Sale Prospectus and the Prospectus, subsequent to the respective dates as of which information is given in the Registration Statement, the Time of Sale Prospectus and the Prospectus: (i) there has been no material adverse change, or any development that would reasonably be expected to result in a material adverse change, in the condition, financial or otherwise, or in the earnings, business, properties, operations, assets, liabilities or prospects, whether or not arising from transactions in the ordinary course of business, of the Company and its subsidiaries, considered as one entity (any such change being referred to herein as a “**Material Adverse Change**”); (ii) the Company and its subsidiaries, considered as one entity, have not incurred any material liability or obligation, indirect, direct or contingent, including without limitation any losses or interference with its business from fire, explosion, flood, earthquakes, accident or other calamity, whether or not covered by insurance, or from any strike, labor dispute or court or governmental action, order or decree, that are material, individually or in the aggregate, to the Company and its subsidiaries, considered as one entity, or has entered into any transactions not in the ordinary course of business; and (iii) there has not been any material decrease in the capital stock or any material increase in any short-term or long-term indebtedness of the Company or its subsidiaries and there has been no dividend or distribution of any kind declared, paid or made by the Company or, except for dividends paid to the Company or other subsidiaries, by any of the Company’s subsidiaries on any class of capital stock, or any repurchase or redemption by the Company or any of its subsidiaries of any class of capital stock.

(i) **Independent Accountants.** Ernst & Young LLP, which has expressed its opinion with respect to the financial statements (which term as used in this Agreement includes the related notes thereto) filed with the Commission as a part of the Registration Statement, the Time of Sale Prospectus and the Prospectus, is (i) an independent registered public accounting firm as required by the Securities Act, the Securities Exchange Act of 1934, as amended, and the rules and regulations promulgated thereunder (collectively, the “**Exchange Act**”), and the rules of the Public Company Accounting Oversight Board (“**PCAOB**”), (ii) in compliance with the applicable requirements relating to the qualification of accountants under Rule 2-01 of Regulation S-X under the Securities Act and (iii) a registered public accounting firm as defined by the PCAOB whose registration has not been suspended or revoked and who has not requested such registration to be withdrawn.

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(j) **Financial Statements.** The financial statements filed with the Commission as a part of the Registration Statement, the Time of Sale Prospectus and the Prospectus present fairly, in all material respects, the consolidated financial position of the Company and its subsidiaries as of the dates indicated and the results of their operations, changes in stockholders’ equity and cash flows for the periods specified. Such financial statements have been prepared in conformity with generally accepted accounting principles as applied in the United States (“**U.S. GAAP**”), applied on a consistent basis throughout the periods involved, except as may be expressly stated in the related notes thereto and except in the case of unaudited financial statements, which are subject to normal and recurring year-end adjustments and do not contain all footnotes as permitted by the applicable rules of the Commission. No other financial statements or supporting schedules are required to be included in the Registration Statement, the Time of Sale Prospectus or the Prospectus. The financial data set forth in each of the Registration Statement, the Time of Sale Prospectus and the Prospectus under the captions “**Prospectus Summary—Summary Consolidated Financial Data**,” “**Capitalization**” and “**Selected Consolidated Financial Data**” fairly present, in all material respects, the information set forth therein on a basis consistent with that of the audited financial statements contained in the Registration Statement, the Time of Sale Prospectus and the Prospectus. All disclosures contained in the Registration Statement, any preliminary prospectus or the Prospectus and any free writing prospectus, that constitute non-GAAP financial measures (as defined by the rules and regulations under the Securities Act and the Exchange Act) comply with Regulation G under the Exchange Act and Item 10 of Regulation S-K under the Securities Act, as applicable. To the Company’s knowledge, no person who has been suspended or barred from being associated with a registered public accounting firm, or who has failed to comply with any sanction pursuant to Rule 5300 promulgated by the PCAOB, has participated in or otherwise aided the preparation of, or audited, the financial statements, supporting schedules or other financial data filed with the Commission as a part of the Registration Statement, the Time of Sale Prospectus and the Prospectus.

(k) **Company’s Accounting System.** The Company and each of its subsidiaries make and keep accurate books and records and maintain a system of internal accounting controls sufficient to provide reasonable assurance that: (i) transactions are executed in accordance with management’s general or specific authorization; (ii) transactions are recorded as necessary to permit preparation of financial statements in conformity with U.S. GAAP and to maintain accountability for assets; (iii) access to assets is permitted only in accordance with management’s general or specific authorization; and (iv) the recorded accountability for assets is compared with existing assets at reasonable intervals and appropriate action is taken with respect to any differences.

(l) **Disclosure Controls and Procedures; Deficiencies in or Changes to Internal Control Over Financial Reporting.** Except as described in the Registration Statement, the Time of Sale Prospectus and the Prospectus, the Company has established and maintains disclosure controls and procedures (as defined in Rules 13a-15 and 15d-15 under the Exchange Act), which (i) are designed to ensure that material information relating to the Company, including its consolidated subsidiaries, is made known to the Company’s principal executive officer and its principal financial officer by others within those entities, particularly during the periods in which the periodic reports required under the Exchange Act are being prepared; (ii) have been evaluated by management of the Company for effectiveness as of the end of the Company’s most recent fiscal quarter; and (iii) are effective in all material respects to perform the functions for which they were established. Since the end of the Company’s most recent audited fiscal year, there have been no significant deficiencies or material weakness in the Company’s internal control over financial reporting (whether or not remediated) and no change in the Company’s internal control over financial reporting that has materially affected, or is reasonably likely to materially affect, the Company’s internal control over financial reporting. The Company is not aware of any change in its internal control over financial reporting that has occurred during its most recent fiscal quarter that has materially affected, or is reasonably likely to materially affect, the Company’s internal control over financial reporting.

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(m) **Incorporation and Good Standing of the Company.** The Company has been duly incorporated and is validly existing as a corporation in good standing under the laws of the State of Delaware and has the corporate power and authority to own, lease and operate its properties and to conduct its business as described in the Registration Statement, the Time of Sale Prospectus and the Prospectus and to enter into and perform its obligations under this Agreement. The Company is duly qualified as a foreign corporation to transact business and is in good standing in each jurisdiction in which such qualification is required, whether by reason of the ownership or leasing of property or the conduct of business, except where the failure to be so qualified or in good standing, as the case may be, or to have such power or authority would not, individually or in the aggregate, have a Material Adverse Effect (defined below).

(n) **Subsidiaries.** Each of the Company’s “subsidiaries” (for purposes of this Agreement, as defined in Rule 405 under the Securities Act) has been duly incorporated or organized, as the case may be, and is validly existing and in good standing (where such concept is recognized) under the laws of the jurisdiction of its incorporation or organization and has the power and authority (corporate or other) to own, lease and operate its properties and to conduct its business as described in the Registration Statement, the Time of Sale Prospectus and the Prospectus. Each of the Company’s subsidiaries is duly qualified to transact business and is in good standing (where such concept is recognized) in each jurisdiction in which such qualification is required, whether by reason of the ownership or leasing of property or the conduct of business, except where the failure to be so qualified or in good standing, as the case may be, or have such power or authority would not, individually or in the aggregate, have a Material Adverse Effect. All of the issued and outstanding capital stock or other equity or ownership interests of each of the Company’s subsidiaries have been duly authorized and validly issued, are fully paid and nonassessable and are owned by the Company, directly or through subsidiaries, free and clear of any security interest, mortgage, pledge, lien, encumbrance or adverse claim. The Company does not own or control, directly or indirectly, any corporation, association or other entity other than the subsidiaries listed in Exhibit 21 to the Registration Statement.

(o) **Capitalization and Other Capital Stock Matters.** The authorized, issued and outstanding capital stock of the Company is as set forth in the Registration Statement, the Time of Sale Prospectus and the Prospectus under the caption “**Capitalization**” (other than for subsequent issuances, if any, pursuant to employee benefit plans, or upon the exercise of outstanding options or warrants, in each case described in the Registration Statement, the Time of Sale Prospectus and the Prospectus). The Shares (including the Offered Shares, when issued pursuant to the terms of this Agreement) conform, in all material respects, to the description thereof contained in the Time of Sale Prospectus. All of the issued and outstanding Shares have been duly authorized and validly issued, are fully paid and nonassessable and have been issued in compliance with all applicable federal and state securities laws. None of the outstanding Shares was issued in violation of any preemptive rights, rights of first refusal or other similar rights to subscribe for or purchase securities of the Company. There are no authorized or outstanding options, warrants, preemptive rights, rights of first refusal or other rights to purchase, or equity or debt securities convertible into or exchangeable or exercisable for, any capital stock of the Company or any of its subsidiaries other than those described in the Registration Statement, the Time of Sale Prospectus and the Prospectus. The descriptions of the Company’s stock option, stock bonus and other stock plans or arrangements, and the options or other rights granted thereunder, set forth in the Registration Statement, the Time of Sale Prospectus and the Prospectus accurately and fairly presents, in all material respects, the information required to be shown with respect to such plans, arrangements, options and rights.

(p) **Stock Exchange Listing.** The Offered Shares have been approved for listing on The Nasdaq Global Market (the “**Nasdaq**”), subject only to official notice of issuance.

(q) **Non-Contravention of Existing Instruments; No Further Authorizations or Approvals Required.** Neither the Company nor any of its subsidiaries is in violation of its charter or by-laws,

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partnership agreement or operating agreement or similar organizational documents, as applicable, or is in default (or, with the giving of notice or lapse of time, would be in default) (“**Default**”) under any indenture, loan, credit agreement, note, lease, license agreement, contract, franchise or other instrument (including, without limitation, any pledge agreement, security agreement, mortgage or other instrument or agreement evidencing, guaranteeing, securing or relating to indebtedness) to which the Company or any of its subsidiaries is a party or by which it or any of them may be bound, or to which any of their respective properties or assets are subject (each, an “**Existing Instrument**”), except for such Defaults as could not be expected, individually or in the aggregate, to have a material adverse effect on the condition (financial or other), earnings, business, properties, operations, assets, liabilities or prospects of the Company and its subsidiaries, considered as one entity (a “**Material Adverse Effect**”). The Company’s execution, delivery and performance of this Agreement, consummation of the transactions contemplated hereby and by the Registration Statement, the Time of Sale Prospectus and the Prospectus and the issuance and sale of the Offered Shares (including the use of proceeds from the sale of the Offered Shares as described in the Registration Statement, the Time of Sale Prospectus and the Prospectus under the caption “**Use of Proceeds**”) (i) have been duly authorized by all necessary corporate action and will not result in any violation of the provisions of the charter or by-laws, partnership agreement or operating agreement or similar organizational documents, as applicable, of the Company or any subsidiary (ii) will not conflict with or constitute a breach of, or Default or a Debt Repayment Triggering Event (as defined below) under, or result in the creation or imposition of any lien, charge or encumbrance upon any property or assets of the Company or any of its subsidiaries pursuant to, or require the consent of any other party to, any Existing Instrument, except as could not be expected, individually or in the aggregate, to have a Material Adverse Effect and (iii) will not result in any violation of any law, administrative regulation or administrative or court decree applicable to the Company or any of its subsidiaries, except for such violations as would not be expected, individually or in the aggregate, to have a Material Adverse Effect. No consent, approval, authorization or other order of, or registration or filing with, any court or other governmental or regulatory authority or agency, is required for the Company’s execution, delivery and performance of this Agreement and consummation of the transactions contemplated hereby and by the Registration Statement, the Time of Sale Prospectus and the Prospectus, except such as have been obtained or made or will be made by the Company under the Securities Act and such as may be required under applicable state securities or blue sky laws or the Financial Industry Regulatory Authority, Inc. (“**FINRA**”). As used herein, a “**Debt Repayment Triggering Event**” means any event or condition which gives, or with the giving of notice or lapse of time would give, the holder of any note, debenture or other evidence of indebtedness (or any person acting on such holder’s behalf) the right to require the repurchase, redemption or repayment of all or a portion of such indebtedness by the Company or any of its subsidiaries.

(r) **Compliance with Laws.** The Company and its subsidiaries have been and are in compliance with all applicable laws, rules and regulations, except where failure to be so in compliance could not be expected, individually or in the aggregate, to have a Material Adverse Effect.

(s) **No Material Actions or Proceedings.** There is no action, suit, proceeding, inquiry or investigation brought by or before any governmental entity now pending or, to the knowledge of the Company, threatened, against or affecting the Company or any of its subsidiaries, which if determined adversely to the Company or any of its subsidiaries would reasonably be expected, individually or in the aggregate, to have a Material Adverse Effect or materially and adversely affect the consummation of the transactions contemplated by this Agreement or the performance by the Company of its obligations hereunder; and the aggregate of all pending legal or governmental proceedings to which the Company or any such subsidiary is a party or of which any of their respective properties or assets is the subject, including ordinary routine litigation incidental to the business, if determined adversely to the Company, would not reasonably be expected, individually or in the aggregate, to have a Material Adverse Effect. No material labor dispute with the employees of the Company or any of its subsidiaries exists or, to the

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knowledge of the Company, is threatened or imminent. To the knowledge of the Company, no material labor dispute with the employees of any principal supplier, manufacturer, customer or contractor of the Company, exists, or is threatened or imminent.

(t) **Intellectual Property Rights.** The Company and its subsidiaries own, or have obtained valid and enforceable licenses for, the inventions, patent applications, patents, trademarks, trade names, service names, copyrights, trade secrets and other intellectual property described in the Registration Statement, the Time of Sale Prospectus and the Prospectus as being owned or licensed by them or which are necessary for the conduct of their respective businesses as currently conducted or as currently proposed to be conducted (with respect to the commercialization of the product candidates described in the Registration Statement, the Time of Sale Prospectus and the Prospectus, except where the failure to own or license such rights would not, individually or in the aggregate, have a Material Adverse Effect) (collectively, “**Intellectual Property**”). There are no third parties who have rights to any Intellectual Property, except for: (i) the exclusive license granted to Janssen Biotech, Inc., a subsidiary of Johnson & Johnson (pursuant to Option and License Agreement dated December 17, 2013, as amended); (ii) certain rights retained by Children’s Medical Center Corporation (CMCC), including rights to practice and use the co-owned patent rights for research, educational, clinical and charitable purposes; and (iii) any customary reversionary rights of third-party licensors with respect to Intellectual Property that is disclosed in the Registration Statement, the Time of Sale Prospectus and the Prospectus as licensed to the Company or one or more of its subsidiaries; and, to the Company’s knowledge, there is no infringement by third parties of any Intellectual Property. There is no pending or, to the Company’s knowledge, threatened action, suit, proceeding or claim by others: (A) challenging the Company’s rights in or to any Intellectual Property, and the Company is unaware of any facts which would form a reasonable basis for any such action, suit, proceeding or claim; (B) challenging the validity, enforceability or scope of any Intellectual Property, and the Company is unaware of any facts which would form a reasonable basis for any such action, suit, proceeding or claim; or (C) asserting that the Company or any of its subsidiaries infringes or otherwise violates, or would, upon the commercialization of any product or service described in the Registration Statement, the Time of Sale Prospectus or the Prospectus as under development, infringe or violate, any patent, trademark, trade name, service name, copyright, trade secret or other proprietary rights of others, and the Company is unaware of any facts which would form a reasonable basis for any such action, suit, proceeding or claim. The Company and its subsidiaries have complied with the terms of each agreement pursuant to which Intellectual Property has been licensed to the Company or any subsidiary, and all such agreements are in full force and effect. The product candidates described in the Registration Statement, the Time of Sale Prospectus and the Prospectus as under development by the Company or any subsidiary fall within the scope of the claims of one or more patents or pending patent applications owned by, or exclusively licensed to, the Company or any subsidiary.

(u) **All Necessary Permits, etc.** The Company and its subsidiaries possess such valid and current certificates, authorizations or permits required by state, federal or foreign regulatory agencies or bodies to conduct their respective businesses as currently conducted and as described in the Registration Statement, the Time of Sale Prospectus or the Prospectus (“**Permits**”), except where the failure to possess the same or so qualify would not reasonably be expected, individually or in the aggregate, to have a Material Adverse Effect. Neither the Company nor any of its subsidiaries is in violation of, or in default under, any of the Permits or has received any notice of proceedings relating to the revocation or modification of, or non-compliance with the Permits, except for such violations, defaults or proceedings if resolved unfavorably, would not reasonably be expected, individually or in the aggregate, to result in a Material Adverse Effect.

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(v) **Title to Properties.** The Company and its subsidiaries have good and marketable title to all of the real and personal property and other assets reflected as owned in the financial statements referred to in Section 1(j) above (or elsewhere in the Registration Statement, the Time of Sale Prospectus or the Prospectus), in each case free and clear of any security interests, mortgages, liens, encumbrances, equities, adverse claims and other defects, except such as do not materially and adversely affect the value of such property and do not materially interfere with the use made or proposed to be made of such property by the Company. The real property, improvements, equipment and personal property held under lease by the Company or any of its subsidiaries are held under valid and enforceable leases, with such exceptions as are not material and do not materially interfere with the use made or proposed to be made of such real property, improvements, equipment or personal property by the Company or such subsidiary.

(w) **Tax Law Compliance.** Except in any case in which failure to pay or file (as applicable) would not, individually or in the aggregate, have a Material Adverse Effect, the Company and its subsidiaries have filed all necessary federal, state and foreign income and franchise tax returns or have properly requested extensions thereof and have paid all taxes required to be paid by any of them and, if due and payable, any related or similar assessment, fine or penalty levied against any of them except as may be being contested in good faith and by appropriate proceedings. Except to the extent of any inadequacy that would not, individually or in the aggregate, result in a Material Adverse Effect, the Company has made adequate charges, accruals and reserves in the applicable financial statements referred to in Section 1(j) above in respect of all federal, state and foreign income and franchise taxes for all periods as to which the tax liability of the Company or any of its subsidiaries has not been finally determined.

(x) **Insurance.** Each of the Company and its subsidiaries are insured by recognized, financially sound and reputable institutions with policies in such amounts and with such deductibles and covering such risks as are reasonably deemed adequate and customary for their businesses including, but not limited to, policies covering real and personal property owned or leased by the Company and its subsidiaries against theft, damage, destruction, acts of vandalism and [earthquakes] and policies covering the Company and its subsidiaries for product liability claims and clinical trial liability claims. The Company has no reason to believe that it or any of its subsidiaries will not be able (i) to renew its existing insurance coverage as and when such policies expire or (ii) to obtain comparable coverage from similar institutions as may be necessary or appropriate to conduct its business as now conducted and at a cost that would not reasonably be expected, individually or in the aggregate, to have a Material Adverse Effect. Neither the Company nor any of its subsidiaries has been denied any insurance coverage which it has sought or for which it has applied.

(y) **Compliance with Environmental Laws.** Except as would not reasonably be expected, individually or in the aggregate, to have a Material Adverse Effect: (i) neither the Company nor any of its subsidiaries is in violation of any federal, state, local or foreign statute, law, rule, regulation, ordinance, code, policy or rule of common law or any judicial or administrative interpretation thereof, including any judicial or administrative order, consent, decree or judgment, relating to pollution or protection of human health, the environment (including, without limitation, ambient air, surface water, groundwater, land surface or subsurface strata) or wildlife, including, without limitation, laws and regulations relating to the release or threatened release of chemicals, pollutants, contaminants, wastes, toxic substances, hazardous substances, petroleum or petroleum products (collectively, “**Hazardous Materials**”) or to the manufacture, processing, distribution, use, treatment, storage, disposal, transport or handling of Hazardous Materials (collectively, “**Environmental Laws**”); (ii) the Company and its subsidiaries have all permits, authorizations and approvals required under any applicable Environmental Laws and are each in compliance with their requirements; (iii) there are no pending or, to the knowledge of the Company, threatened administrative, regulatory or judicial actions, suits, demands, demand letters, claims, liens, notices of noncompliance or violation, investigation or proceedings relating to any Environmental Law against the Company or any of its subsidiaries; and (iv) there are no events or circumstances that might

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reasonably be expected to form the basis of an order for clean-up or remediation, or an action, suit or proceeding by any private party or governmental body or agency, against or affecting the Company or any of its subsidiaries relating to Hazardous Materials or any Environmental Laws.

(z) **No Rated Debt or Preferred Securities.** There are no debt or preferred securities issued, or guaranteed, by the Company or its subsidiaries that are rated by a “nationally recognized statistical rating organization,” as such term is defined in Section 3(a)(62) of the Exchange Act.

(aa) **ERISA Compliance.** The Company and its subsidiaries and any “employee benefit plan” (as defined under the Employee Retirement Income Security Act of 1974, as amended, and the regulations and published interpretations thereunder (collectively, “**ERISA**”)) established or maintained by the Company, its subsidiaries or their “ERISA Affiliates” (as defined below) are in compliance in all material respects with ERISA. “**ERISA Affiliate**” means, with respect to the Company or any of its subsidiaries, any member of any group of organizations described in Sections 414(b), (c), (m) or (o) of the Internal Revenue Code of 1986, as amended, and the regulations and published interpretations thereunder (the “**Code**”) of which the Company or such subsidiary is a member. No “reportable event” (as defined under ERISA), for which notice has not been waived, has occurred or is reasonably expected to occur with respect to any “employee benefit plan” established or maintained by the Company, its subsidiaries or any of their ERISA Affiliates. No “employee benefit plan” established or maintained by the Company, its subsidiaries or any of their ERISA Affiliates, if such “employee benefit plan” were terminated, would have any “amount of unfunded benefit liabilities” (as defined under ERISA). Neither the Company, its subsidiaries nor any of their ERISA Affiliates has incurred or reasonably expects to incur any liability under (i) Title IV of ERISA with respect to termination of, or withdrawal from, any “employee benefit plan” or (ii) Sections 412, 4971, 4975 or 4980B of the Code. Each employee benefit plan established or maintained by the Company, its subsidiaries or any of their ERISA Affiliates that is intended to be qualified under Section 401(a) of the Code is so qualified and, to the knowledge of the Company, nothing has occurred, whether by action or failure to act, which would cause the loss of such qualification.

(bb) **Company Not an “Investment Company.”** The Company is not, and will not be, either after receipt of payment for the Offered Shares or after the application of the proceeds therefrom as described under “Use of Proceeds” in the Registration Statement, the Time of Sale Prospectus or the Prospectus, required to register as an “investment company” under the Investment Company Act of 1940, as amended (the “**Investment Company Act**”).

(cc) **No Price Stabilization or Manipulation; Compliance with Regulation M.** Neither the Company nor any of its subsidiaries has taken, directly or indirectly, without giving effect to activities by the Underwriters, any action designed to or that might reasonably be expected to cause or result in stabilization or manipulation of the price of the Shares or of any “reference security” (as defined in Rule 100 of Regulation M under the Exchange Act (“**Regulation M**”)) with respect to the Shares, whether to facilitate the sale or resale of the Offered Shares or otherwise, and has taken no action which would directly or indirectly violate Regulation M.

(dd) **Related-Party Transactions.** There are no business relationships or related-party transactions involving the Company or any of its subsidiaries or any other person required to be described in the Registration Statement, the Time of Sale Prospectus or the Prospectus that have not been described as required.

(ee) **FINRA Matters.** All of the information provided to the Underwriters or to counsel for the Underwriters by the Company, its counsel, its officers and directors and, to the Company’s knowledge, the holders of any securities (debt or equity) or options to acquire any securities of the Company in connection with the offering of the Offered Shares is true, complete, correct and compliant with FINRA’s rules and

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any letters, filings or other supplemental information provided to FINRA pursuant to FINRA Rules or NASD Conduct Rules is true, complete and correct.

(ff) **Parties to Lock-Up Agreements.** The Company has furnished to the Underwriters a letter agreement in the form attached hereto as **Exhibit D** (the “**Lock-Up Agreement**”) from each of the persons listed on **Exhibit E**. Such **Exhibit E** lists under an appropriate caption the directors and officers of the Company. If any additional persons shall become directors or officers of the Company prior to the end of the Company Lock-up Period (as defined below), the Company shall cause each such person, prior to or contemporaneously with their appointment or election as a director or officer of the Company, to execute and deliver to Jefferies and Cowen a Lock-up Agreement.

(gg) **Statistical and Market-Related Data.** All statistical, demographic and market-related data included in the Registration Statement, the Time of Sale Prospectus or the Prospectus are based on or derived from sources that the Company believes, after reasonable inquiry, to be reliable and accurate. To the extent required, the Company has obtained the written consent to the use of such data from such sources.

(hh) **No Unlawful Contributions or Other Payments.** Neither the Company nor any of its subsidiaries nor, to the best knowledge of the Company, any employee or agent of the Company or any of its subsidiaries, has made any contribution or other payment to any official of, or candidate for, any federal, state or foreign office in violation of any applicable law or of the character required to be disclosed in the Registration Statement, the Time of Sale Prospectus or the Prospectus.

(ii) **Foreign Corrupt Practices Act.** Neither the Company nor any of its subsidiaries nor, to the knowledge of the Company, any director, officer, agent, employee, controlled affiliate or other person acting on behalf of the Company or any of its subsidiaries has, in the course of its actions for, or on behalf of, the Company or any of its subsidiaries (i) used any corporate funds for any unlawful contribution, gift, entertainment or other unlawful expenses relating to political activity; (ii) made any direct or indirect unlawful payment to any domestic government official, “foreign official” (as defined in the U.S. Foreign Corrupt Practices Act of 1977, as amended, and the rules and regulations thereunder, collectively, the “**FCPA**”) or employee from corporate funds; (iii) violated or is in violation of any provision of the FCPA or any applicable non-U.S. anti-bribery statute or regulation; or (iv) made any unlawful bribe, rebate, payoff, influence payment, kickback or other unlawful payment to any domestic government official, such foreign official or employee; and the Company and its subsidiaries and, to the knowledge of the Company, the Company’s controlled affiliates have conducted their respective businesses in compliance with the FCPA and have instituted and maintain policies and procedures designed to ensure, and which are reasonably expected to continue to ensure, continued compliance therewith.

(jj) **Money Laundering Laws.** The operations of the Company and its subsidiaries are, and have been conducted at all times, in compliance with applicable financial recordkeeping and reporting requirements of the Currency and Foreign Transactions Reporting Act of 1970, as amended, the money laundering statutes of all applicable jurisdictions, the rules and regulations thereunder and any related or similar applicable rules, regulations or guidelines, issued, administered or enforced by any governmental agency (collectively, the “**Money Laundering Laws**”) and no action, suit or proceeding by or before any court or governmental agency, authority or body or any arbitrator involving the Company or any of its subsidiaries with respect to the Money Laundering Laws is pending or, to the best knowledge of the Company, threatened.

(kk) **OFAC.** Neither the Company nor any of its subsidiaries nor, to the knowledge of the Company, after due inquiry, any director, officer, agent, employee, controlled affiliate or person acting on behalf of the Company or any of its subsidiaries is currently subject to any U.S. sanctions administered by

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the Office of Foreign Assets Control of the U.S. Treasury Department (“**OFAC**”); and the Company will not directly or indirectly use the proceeds of this offering, or lend, contribute or otherwise make available such proceeds to any subsidiary, or any joint venture partner or other person or entity, for the purpose of financing the activities of or business with any person, or in any country or territory, that currently is the subject to any U.S. sanctions administered by OFAC or in any other manner that will result in a violation by any person (including any person participating in the transaction whether as underwriter, advisor, investor or otherwise) of U.S. sanctions administered by OFAC.

(ll) **Brokers.** Except pursuant to this Agreement, there is no broker, finder or other party that is entitled to receive from the Company any brokerage or finder’s fee or other fee or commission as a result of any transactions contemplated by this Agreement.

(mm) **Forward-Looking Statements.** Each financial or operational projection or other “forward-looking statement” (as defined by Section 27A of the Securities Act or Section 21E of the Exchange Act) contained in the Registration Statement, the Time of Sale Prospectus or the Prospectus (i) was so included by the Company in good faith and with reasonable basis after due consideration by the Company of the underlying assumptions, estimates and other applicable facts and circumstances and (ii) is accompanied by meaningful cautionary statements identifying those factors that could cause actual results to differ materially from those in such forward-looking statement. No such statement was made with the knowledge of an executive officer or director of the Company that it was false or misleading.

(nn) **Emerging Growth Company Status.** From the time of initial confidential submission of the Registration Statement to the Commission (or, if earlier, the first date on which the Company engaged in any Section 5(d) Written Communication or any Section 5(d) Oral Communication) through the date hereof, the Company has been and is an “emerging growth company,” as defined in Section 2(a) of the Securities Act (an “**Emerging Growth Company**”).

(oo) **Communications.** The Company (i) has not alone engaged in communications with potential investors in reliance on Section 5(d) of the Securities Act other than Permitted Section 5(d) Communications with the consent of the Representatives with entities that are QIBs or IAs and (ii) has not authorized anyone other than the Representatives to engage in such communications; the Company reconfirms that the Representatives have been authorized to act on its behalf in undertaking Marketing Materials, Section 5(d) Oral Communications and Section 5(d) Written Communications; as of the Applicable Time, each Permitted Section 5(d) Communication, when considered together with the Time of Sale Prospectus, did not, as of the Applicable Time, include an untrue statement of a material fact or omit to state a material fact necessary in order to make the statements therein, in the light of the circumstances under which they were made, not misleading; and each Permitted Section 5(d) Communication, if any, does not, as of the date hereof, conflict with the information contained in the Registration Statement, the Preliminary Prospectus and the Prospectus; and the Company has filed publicly on EDGAR at least fifteen (15) calendar days prior to any “road show” (as defined in Rule 433 under the Act), any confidentially submitted registration statement and registration statement amendments relating to the offer and sale of the Offered Shares.

(pp) **Clinical Data and Regulatory Compliance.** The preclinical tests and clinical trials, if any, and other studies (collectively, “studies”) conducted by or on behalf of or sponsored by the Company that are described in, or the results of which are referred to in, the Registration Statement, the Time of Sale Prospectus or the Prospectus were and, if still pending, are being conducted in all material respects in accordance with the protocols, procedures and controls designed and approved for such studies and with standard medical and scientific research procedures; each description of the results of such studies is accurate and complete in all material respects and fairly presents the data derived from such studies, and the Company and its subsidiaries have no knowledge of any other studies the results of which are

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inconsistent with, or otherwise call into question, the results described or referred to in the Registration Statement, the Time of Sale Prospectus or the Prospectus; the Company and its subsidiaries have made all such filings and obtained all such approvals as may be required by the Food and Drug Administration of the U.S. Department of Health and Human Services or any committee thereof or from any other U.S. or foreign government or drug or medical device regulatory agency, or health care facility Institutional Review Board (collectively, the “**Regulatory Agencies**”); neither the Company nor any of its subsidiaries has received any notice of, or correspondence from, any Regulatory Agency requiring the termination, suspension or material modification of any clinical trials that are described or referred to in the Registration Statement, the Time of Sale Prospectus or the Prospectus; and the Company and its subsidiaries have each operated and currently are in compliance in all material respects with all applicable rules, regulations and policies of the Regulatory Agencies.

(qq) **Compliance with Health Care Laws.** Except as described in the Registration Statement, the Time of Sale Prospectus or the Prospectus, and except as would not, individually or in the aggregate, have or may reasonably be expected to have a Material Adverse Effect: (i) the Company’s and each of its subsidiaries’ business practices have been structured in a manner designed to comply with state, federal and foreign laws applicable to the Company and its subsidiaries respective businesses, and the Company and its subsidiaries are in compliance with such laws including, without limitation, applicable provisions of: (A) the Federal Food, Drug, and Cosmetic Act (21 U.S.C. § 301 et seq.) and the Public Health Service Act (42 U.S.C. § 201 et seq.), and the regulations promulgated thereunder; (B) all applicable federal, state, local and all applicable foreign health care related fraud and abuse laws, including, without limitation, the U.S. Anti-Kickback Statute (42 U.S.C. § 1320a-7b(b)), the U.S. civil False Claims Act (31 U.S.C. § 3729 et seq.), the federal criminal false claims law (42 U.S.C. § 1320a-7b(a)), the federal civil monetary penalties law (42 U.S.C. § 1320a-7a), the Stark Law (42 U.S.C. § 1395nn), the U.S. Physician Payments Sunshine Act (42 U.S.C. § 1320a-7h), 18 U.S.C. §§ 286, 287, 1347, and 1349 and the health care fraud criminal provisions under the U.S. Health Insurance Portability and Accountability Act of 1996 (“**HIPAA**”) (42 U.S.C. § 1320d et seq.), the exclusion law (42 U.S.C. § 1320a-7), Medicare, Title XVIII of the Social Security Act, and Medicaid, Title XIX of the Social Security Act, and the regulations promulgated pursuant to such statutes; and (C) HIPAA, as amended by the Health Information Technology for Economic and Clinical Health Act (42 U.S.C. § 17921 et seq.), and the regulations promulgated thereunder and any state or non-U.S. counterpart thereof or other law or regulation the purpose of which is to protect the privacy of individuals or prescribers (collectively, “**Health care Laws**”); (ii) the Company and its subsidiaries have not engaged in activities which are cause for false claims liability, civil penalties, or mandatory or permissive exclusion from Medicare, Medicaid, or any other state health care program or federal health care program; (iii) neither the Company nor its subsidiaries have received written notice of any claim, action, suit, proceeding, hearing, enforcement, investigation, arbitration or other action from any court or arbitrator or governmental or regulatory authority or third party alleging that any product, operation or activity is in violation of any Health Care Laws nor, to the knowledge of the Company, is any, such claim, action, suit, proceeding, hearing, enforcement, investigation, arbitration or other action threatened; (iv) neither the Company nor any subsidiary has received written notice that any court or arbitrator or governmental or regulatory authority has taken, is taking or intends to take action to limit, suspend, modify or revoke applicable Permits or has any knowledge that any such court or arbitrator or governmental or regulatory authority is considering such action; (v) the Company and its subsidiaries have filed, maintained or submitted all material reports, documents, forms, notices, applications, records, claims, submissions and supplements or amendments as required by any Health Care Laws, and all such reports, documents, forms, notices, applications, records, claims, submissions and supplements or amendments were complete and accurate on the date filed in all material respects (or were corrected or supplemented by a subsequent submission); (vi) neither the Company nor its subsidiaries are a party to any corporate integrity agreements, monitoring agreements, consent decrees, plans of correction, settlement orders, or similar agreements with or imposed by any governmental or regulatory authority; and (vii) neither the Company, its subsidiaries nor any of their respective officers, directors, employees, or agents

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have been excluded, suspended or debarred from participation in any U.S. federal health care program or human clinical research or, to the knowledge of the Company, is subject to a governmental inquiry, investigation, proceeding, or other similar action that could reasonably be expected to result in debarment, suspension, or exclusion.

(rr) **No Rights to Purchase Preferred Stock.** The issuance and sale of the Shares as contemplated hereby will not cause any holder of any shares of capital stock, securities convertible into or exchangeable or exercisable for capital stock or options, warrants or other rights to purchase capital stock or any other securities of the Company to have any right to acquire any shares of preferred stock of the Company.

(ss) **No Contract Terminations.** Neither the Company nor any of its subsidiaries has sent or received any communication regarding termination of, or intent not to renew, any of the contracts or agreements referred to or described in any preliminary prospectus, the Prospectus or any free writing prospectus, or referred to or described in, or filed as an exhibit to, the Registration Statement, and no such termination or non-renewal has been threatened by the Company or any of its subsidiaries or, to the Company’s knowledge, any other party to any such contract or agreement, which threat of termination or non-renewal has not been rescinded as of the date hereof.

(tt) **Dividend Restrictions.** No subsidiary of the Company is prohibited or restricted, directly or indirectly, from paying dividends to the Company, or from making any other distribution with respect to such subsidiary’s equity securities or from repaying to the Company or any other subsidiary of the Company any amounts that may from time to time become due under any loans or advances to such subsidiary from the Company or from transferring any property or assets to the Company or to any other subsidiary.

Any certificate signed by any officer of the Company or any of its subsidiaries and delivered to any Underwriter or to counsel for the Underwriters in connection with the offering, or the purchase and sale, of the Offered Shares shall be deemed a representation and warranty by the Company (and not by such officer in his or her personal capacity) to each Underwriter as to the matters covered thereby.

The Company has a reasonable basis for making each of the representations set forth in this Section 1. The Company acknowledges that the Underwriters and, for purposes of the opinions to be delivered pursuant to Section 6 hereof, counsel to the Company and counsel to the Underwriters, will rely upon the accuracy and truthfulness of the foregoing representations and hereby consents to such reliance.

Section 2. Purchase, Sale and Delivery of the Offered Shares.

(a) **The Firm Shares.** Upon the terms herein set forth, the Company agrees to issue and sell to the several Underwriters an aggregate of [-] Firm Shares. On the basis of the representations, warranties and agreements herein contained, and upon the terms but subject to the conditions herein set forth, the Underwriters agree, severally and not jointly, to purchase from the Company the respective number of Firm Shares set forth opposite their names on Schedule A. The purchase price per Firm Share to be paid by the several Underwriters to the Company shall be \$[-] per share.

(b) **The First Closing Date.** Delivery of certificates for the Firm Shares to be purchased by the Underwriters and payment therefor shall be made at the offices of Cooley LLP, 500 Boylston Street, Boston, MA 02116 (or such other place as may be agreed to by the Company and the Representatives) at 9:00 a.m. New York City time, on [-], 2018, or such other time and date not later than 1:30 p.m. New York City time, on [-], 2018 as the Representatives shall designate by notice to the Company (the time and date of such closing are called the “**First Closing Date**”). The Company hereby acknowledges that

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circumstances under which the Representatives may provide notice to postpone the First Closing Date as originally scheduled include, but are not limited to, any determination by the Company or the Representatives to recirculate to the public copies of an amended or supplemented Prospectus or a delay as contemplated by the provisions of Section 11.

(c) **The Optional Shares; Option Closing Date.** In addition, on the basis of the representations, warranties and agreements herein contained, and upon the terms but subject to the conditions herein set forth, the Company hereby grants an option to the several Underwriters to purchase, severally and not jointly, up to an aggregate of [-] Optional Shares from the Company at the purchase price per share to be paid by the Underwriters for the Firm Shares. The

option granted hereunder may be exercised at any time and from time to time in whole or in part upon notice by the Representatives to the Company, which notice may be given at any time within thirty (30) days from the date of this Agreement. Such notice shall set forth (i) the aggregate number of Optional Shares as to which the Underwriters are exercising the option and (ii) the time, date and place at which certificates for the Optional Shares will be delivered (which time and date may be simultaneous with, but not earlier than, the First Closing Date; and in the event that such time and date are simultaneous with the First Closing Date, the term "First Closing Date" shall refer to the time and date of delivery of certificates for the Firm Shares and such Optional Shares). Any such time and date of delivery, if subsequent to the First Closing Date, is called an "Option Closing Date," shall be determined by the Representatives and shall not be earlier than three or later than five (5) full business days after delivery of such notice of exercise. If any Optional Shares are to be purchased, each Underwriter agrees, severally and not jointly, to purchase the number of Optional Shares (subject to such adjustments to eliminate fractional shares as the Representatives may determine) that bears the same proportion to the total number of Optional Shares to be purchased as the number of Firm Shares set forth on Schedule A opposite the name of such Underwriter bears to the total number of Firm Shares. The Representatives may cancel the option at any time prior to its expiration by giving written notice of such cancellation to the Company.

(d) **Public Offering of the Offered Shares.** The Representatives hereby advise the Company that the Underwriters intend to offer for sale to the public, initially on the terms set forth in the Registration Statement, the Time of Sale Prospectus and the Prospectus, their respective portions of the Offered Shares as soon after this Agreement has been executed and the Registration Statement has been declared effective as the Representatives, in their sole judgment, have determined is advisable and practicable.

(e) **Payment for the Offered Shares.** (i) Payment for the Offered Shares shall be made at the First Closing Date (and, if applicable, at each Option Closing Date) by wire transfer of immediately available funds to the order of the Company.

(ii) It is understood that the Representatives have been authorized, for their own account and the accounts of the several Underwriters, to accept delivery of and receipt for, and make payment of the purchase price for, the Firm Shares and any Optional Shares the Underwriters have agreed to purchase. Each of Jefferies, Cowen and BMO, individually and not as the Representatives of the Underwriters, may (but shall not be obligated to) make payment for any Offered Shares to be purchased by any Underwriter whose funds shall not have been received by the Representatives by the First Closing Date or the applicable Option Closing Date, as the case may be, for the account of such Underwriter, but any such payment shall not relieve such Underwriter from any of its obligations under this Agreement.

(f) **Delivery of the Offered Shares.** The Company shall deliver, or cause to be delivered to the Representatives for the accounts of the several Underwriters certificates for the Firm Shares at the First Closing Date, against release of a wire transfer of immediately available funds for the amount of the purchase price therefor. The Company shall also deliver, or cause to be delivered to the Representatives for the accounts of the several Underwriters, certificates for the Optional Shares the Underwriters have

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agreed to purchase at the First Closing Date or the applicable Option Closing Date, as the case may be, against the release of a wire transfer of immediately available funds for the amount of the purchase price therefor. The certificates for the Offered Shares shall be registered in such names and denominations as the Representatives shall have requested at least two (2) full business days prior to the First Closing Date (or the applicable Option Closing Date, as the case may be) and shall be made available for inspection on the business day preceding the First Closing Date (or the applicable Option Closing Date, as the case may be) at a location in New York City as the Representatives may designate. Time shall be of the essence, and delivery at the time and place specified in this Agreement is a further condition to the obligations of the Underwriters.

Section 3. Additional Covenants. The Company further covenants and agrees with each Underwriter as follows:

(a) **Delivery of Registration Statement, Time of Sale Prospectus and Prospectus.** The Company shall furnish to you in New York City, without charge, prior to 10:00 a.m. New York City time on the second business day next succeeding the date of this Agreement and during the period when a prospectus relating to the Offered Shares is required by the Securities Act to be delivered (whether physically or through compliance with Rule 172 under the Securities Act or any similar rule) in connection with sales of the Offered Shares, as many copies of the Time of Sale Prospectus, the Prospectus and any supplements and amendments thereto or to the Registration Statement as you may reasonably request.

(b) **Representatives' Review of Proposed Amendments and Supplements.** During the period when a prospectus relating to the Offered Shares is required by the Securities Act to be delivered (whether physically or through compliance with Rule 172 under the Securities Act or any similar rule), the Company (i) will furnish to the Representatives for review, a reasonable period of time prior to the proposed time of filing of any proposed amendment or supplement to the Registration Statement, a copy of each such amendment or supplement and (ii) will not amend or supplement the Registration Statement without the Representatives' prior written consent, which consent shall not be unreasonably withheld. Prior to amending or supplementing any preliminary prospectus, the Time of Sale Prospectus or the Prospectus, the Company shall furnish to the Representatives for review, a reasonable amount of time prior to the time of filing or use of the proposed amendment or supplement, a copy of each such proposed amendment or supplement. The Company shall not file or use any such proposed amendment or supplement without the Representatives' prior written consent, which consent shall not be unreasonably withheld. The Company shall file with the Commission within the applicable period specified in Rule 424(b) under the Securities Act any prospectus required to be filed pursuant to such Rule.

(c) **Free Writing Prospectuses.** The Company shall furnish to the Representatives for review, a reasonable amount of time prior to the proposed time of filing or use thereof, a copy of each proposed free writing prospectus or any amendment or supplement thereto prepared by or on behalf of, used by, or referred to by the Company, and the Company shall not file, use or refer to any proposed free writing prospectus or any amendment or supplement thereto without the Representatives' prior written consent, which consent shall not be unreasonably withheld. The Company shall furnish to each Underwriter, without charge, as many copies of any free writing prospectus prepared by or on behalf of, used by or referred to by the Company as such Underwriter may reasonably request. If at any time when a prospectus is required by the Securities Act to be delivered (whether physically or through compliance with Rule 172 under the Securities Act or any similar rule) in connection with sales of the Offered Shares (but in any event at any time through and including the First Closing Date) there occurred or occurs an event or development as a result of which any free writing prospectus prepared by or on behalf of, used by, or referred to by the Company conflicted or would conflict with the information contained in the Registration Statement or included or would include an untrue statement of a material fact or omitted or would omit to state a material fact necessary in order to make the statements therein, in the light of the circumstances

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prevailing at such time, not misleading, the Company shall promptly amend or supplement such free writing prospectus to eliminate or correct such conflict so that the statements in such free writing prospectus as so amended or supplemented will not include an untrue statement of a material fact or omit to state a material fact necessary in order to make the statements therein, in the light of the circumstances prevailing at such time, not misleading, as the case may be; provided, however, that prior to amending or supplementing any such free writing prospectus, the Company shall furnish to the Representatives for review, a reasonable amount of time prior to the proposed time of filing or use thereof, a copy of such proposed amended or supplemented free writing prospectus, and the Company shall not file, use or refer to any such amended or supplemented free writing prospectus without the Representatives' prior written consent, which consent shall not be unreasonably withheld.

(d) **Filing of Underwriter Free Writing Prospectuses.** The Company shall not take any action that would result in an Underwriter or the Company being required to file with the Commission pursuant to Rule 433(d) under the Securities Act a free writing prospectus prepared by or on behalf of such Underwriter that such Underwriter otherwise would not have been required to file thereunder.

(e) **Amendments and Supplements to Time of Sale Prospectus.** If the Time of Sale Prospectus is being used to solicit offers to buy the Offered Shares at a time when the Prospectus is not yet available to prospective purchasers, and any event shall occur or condition exist as a result of which it is necessary to amend or supplement the Time of Sale Prospectus so that the Time of Sale Prospectus does not include an untrue statement of a material fact or omit to state a material fact necessary in order to make the statements therein, in the light of the circumstances when delivered to a prospective purchaser, not misleading, or if any event shall occur or condition exist as a result of which the Time of Sale Prospectus conflicts with the information contained in the Registration Statement, or if, in the opinion of the Company, counsel for the Company, the Representatives or counsel for the Underwriters, it is necessary to amend or supplement the Time of Sale Prospectus to comply with applicable law, the Company shall (subject to Section 3(b) and Section 3(c) hereof) promptly prepare, file with the Commission and furnish, at its own expense, to the Underwriters and to any dealer upon request, either amendments or supplements to the Time of Sale Prospectus so that the statements in the Time of Sale Prospectus as so amended or supplemented will not include an untrue statement of a material fact or omit to state a material fact necessary in order to make the statements therein, in the light of the circumstances when delivered to a prospective purchaser, not misleading or so that the Time of Sale Prospectus, as amended or supplemented, will no longer conflict with the information contained in the Registration Statement, or so that the Time of Sale Prospectus, as amended or supplemented, will comply with applicable law.

(f) **Certain Notifications and Required Actions.** After the date of this Agreement, the Company shall promptly advise the Representatives in writing of: (i) the receipt of any comments of, or requests for additional or supplemental information from, the Commission; (ii) the time and date of any filing of any post-effective amendment to the Registration Statement or any amendment or supplement to any preliminary prospectus, the Time of Sale Prospectus, any free writing prospectus or the Prospectus; (iii) the time and date that any post-effective amendment to the Registration Statement becomes effective; and (iv) the issuance by the Commission of any stop order suspending the effectiveness of the Registration Statement or any post-effective amendment thereto or any amendment or supplement to any preliminary prospectus, the Time of Sale Prospectus or the Prospectus or of any order preventing or suspending the use of any preliminary prospectus, the Time of Sale Prospectus, any free writing prospectus or the Prospectus, or of any proceedings to remove, suspend or terminate from listing or quotation the Shares from any securities exchange upon which they are listed for trading or included or designated for quotation, or of the threatening or initiation of any proceedings for any of such purposes. If the Commission shall enter any such stop order at any time, the Company will use its best efforts to obtain the lifting of such order at the earliest possible moment. Additionally, the Company agrees that it shall comply with all applicable provisions of Rule 424(b), Rule 433 and Rule 430A under the Securities Act and will use its reasonable

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efforts to confirm that any filings made by the Company under Rule 424(b) or Rule 433 were received in a timely manner by the Commission.

(g) **Amendments and Supplements to the Prospectus and Other Securities Act Matters.** If, during the Prospective Delivery Period (as defined below), any event shall occur or condition exist as a result of which it is necessary to amend or supplement the Prospectus so that the Prospectus does not include an untrue statement of a material fact or omit to state a material fact necessary in order to make the statements therein, in the light of the circumstances when the Prospectus is delivered (whether physically or through compliance with Rule 172 under the Securities Act or any similar rule) to a purchaser, not misleading, or if in the opinion of the Company, counsel for the Company, the Representatives or counsel for the Underwriters it is otherwise necessary to amend or supplement the Prospectus to comply with applicable law, the Company agrees (subject to Section 3(b) and Section 3(c) hereof) to promptly prepare, file with the Commission and furnish, at its own expense, to the Underwriters and to any dealer upon request, amendments or supplements to the Prospectus so that the statements in the Prospectus as so amended or supplemented will not include an untrue statement of a material fact or omit to state a material fact necessary in order to make the statements therein, in the light of the circumstances when the Prospectus is delivered (whether physically or through compliance with Rule 172 under the Securities Act or any similar rule) to a purchaser, not misleading or so that the Prospectus, as amended or supplemented, will comply with applicable law. Neither the Representatives' consent to, nor delivery of, any such amendment or supplement shall constitute a waiver of any of the Company's obligations under Section 3(b) or Section 3(c). As used herein, the term "Prospectus Delivery Period" means such period of time after the first date of the public offering of the Shares as in the opinion of counsel for Underwriters a prospectus relating to the Shares is required by law to be delivered (or required to be delivered but for Rule 172 under the Securities Act) in connection with the sales of Shares by any Underwriter or dealer.

(h) **Blue Sky Compliance.** The Company shall cooperate with the Representatives and counsel for the Underwriters to qualify or register the Offered Shares for sale under (or obtain exemptions from the application of) the state securities or blue sky laws or Canadian provincial securities laws (or other foreign laws) of those jurisdictions reasonably designated by the Representatives, shall comply with such laws and shall continue such qualifications, registrations and exemptions in effect so long as required for the distribution of the Offered Shares; provided that the Company shall not be required to qualify as a foreign corporation or to take any action that would subject it to general service of process in any such jurisdiction where it is not presently qualified or where it would be subject to taxation as a foreign corporation. The Company will advise the Representatives promptly of the suspension of the qualification or registration of (or any such exemption relating to) the Offered Shares for offering, sale or trading in any jurisdiction or any initiation or threat of any proceeding for any such purpose, and in the event of the issuance of any order suspending such qualification, registration or exemption, the Company shall use its reasonable best efforts to obtain the withdrawal thereof at the earliest possible moment.

(i) **Use of Proceeds.** The Company shall apply the net proceeds from the sale of the Offered Shares sold by it in the manner described under the caption "Use of Proceeds" in the Registration Statement, the Time of Sale Prospectus and the Prospectus.

(j) **Transfer Agent.** The Company shall engage and maintain, at its expense, a registrar and transfer agent for the Shares.

(k) **Earnings Statement.** The Company will make generally available to its security holders and to the Representatives as soon as practicable an earnings statement (which need not be audited) covering a period of at least twelve (12) months beginning with the first fiscal quarter of the Company commencing after the date of this Agreement that will satisfy the provisions of Section 11(a) of the Securities Act and the rules and regulations of the Commission thereunder.

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(l) **Continued Compliance with Securities Laws.** The Company will comply with the Securities Act and the Exchange Act so as to permit the completion of the distribution of the Offered Shares as contemplated by this Agreement, the Registration Statement, the Time of Sale Prospectus and the Prospectus. Without limiting the generality of the foregoing, the Company will, during the period when a prospectus relating to the Offered Shares is required by the Securities Act to be delivered (whether physically or through compliance with Rule 172 under the Securities Act or any similar rule), file on a timely basis with the Commission and the Nasdaq all reports and documents required to be filed under the Exchange Act. Additionally, the Company shall report the use of proceeds from the issuance of the Offered Shares as may be required under Rule 463 under the Securities Act.

(m) **Listing.** The Company will use its best efforts to list, subject to notice of issuance, the Offered Shares on the Nasdaq.

(n) **Company to Provide Copy of the Prospectus in Form That May be Downloaded from the Internet.** If requested by the Representatives, the Company shall cause to be prepared and delivered, at its expense, within one business day from the effective date of this Agreement, to the Representatives a "electronic Prospectus" to be used by the Underwriters in connection with the offering and sale of the Offered Shares. As used herein, the term "electronic Prospectus" means a form of Time of Sale Prospectus, and any amendment or supplement thereto, that meets each of the following conditions: (i) it shall be encoded in an electronic format, satisfactory to the Representatives, that may be transmitted electronically by the Representatives and the other Underwriters to offerees and purchasers of the Offered Shares; (ii) it shall disclose the same information as the paper Time of Sale Prospectus, except to the extent that graphic and image material cannot be disseminated electronically, in which case such graphic and image material shall be replaced in the electronic Prospectus with a fair and accurate narrative description or tabular representation of such material, as appropriate; and (iii) it shall be in or convertible into a paper format or an electronic format, satisfactory to the Representatives, that will allow investors to store and have continuously ready access to the Time of Sale Prospectus at any future time, without charge to investors (other than any fee charged for subscription to the Internet as a whole and for on-line time). The Company hereby confirms that it has included or will include in the Prospectus filed pursuant to EDGAR or otherwise with the Commission and in the Registration Statement at the time it was declared effective an undertaking that, upon receipt of a request by an investor or his or her representative, the Company shall transmit or cause to be transmitted promptly, without charge, a paper copy of the Time of Sale Prospectus.

(o) **Agreement Not to Offer or Sell Additional Shares.** During the period commencing on and including the date hereof and continuing through and including the 180th day following the date of the Prospectus (such period, being referred to herein as the "Lock-up Period"), the Company will not, without the prior written consent of Jefferies and Cowen (which consent may be withheld in their sole discretion), directly or indirectly: (i) sell, offer to sell, contract to sell or lend any Shares or Related Securities (as defined below); (ii) effect any short sale, or establish or increase any "put equivalent position" (as defined in Rule 16a-1(h) under the Exchange Act) or liquidate or decrease any "call equivalent position" (as defined in Rule 16a-1(b) under the Exchange Act) of any Shares or Related Securities; (iii) pledge, hypothecate or grant any security interest in any Shares or Related Securities; (iv) in any other way transfer or dispose of any Shares or Related Securities; (v) enter into any swap, hedge or similar arrangement or agreement that transfers, in whole or in part, the economic risk of ownership of any Shares or Related Securities, regardless of whether any such transaction is to be settled in securities, in cash or otherwise; (vi) announce the offering of any Shares or Related Securities; (vii) file any registration statement under the Securities Act in respect of any Shares or Related Securities (other than as contemplated by this Agreement with respect to the Offered Shares); or (viii) publicly announce the intention to do any of the foregoing; *provided, however*, that the Company may (A) effect the transactions contemplated hereby, (B) issue Shares upon exercise of options or warrants outstanding on the date hereof

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or issue Shares or awards or options to purchase Shares pursuant to any stock option, stock bonus, or other stock plan or arrangement described in the Registration Statement, the Time of Sale Prospectus and the Prospectus, (C) file a registration statement on Form S-8 with respect to any Shares or Related Securities issued or issuable pursuant to any stock option, stock bonus, or other stock plan or arrangement described in the Registration Statement, the Time of Sale Prospectus and the Prospectus, (D) issue shares of Common Stock in connection with the acquisition by the Company of the securities, business, property or other assets of another person or business entity or pursuant to any employee benefit plan assumed by the Company in connection with any such acquisition or (E) issue shares of Common Stock or Related Securities in connection with joint ventures, commercial relationships or other strategic transactions; *provided that*, in the case of immediately preceding clauses (D) and (E), (x) the aggregate number of shares of Common Stock issued or underlying such Related Securities issued in connection with all such acquisitions and other transactions does not exceed 10% of the aggregate number of shares of Common Stock outstanding immediately following the consummation of the offering of the Offered Shares pursuant to this Agreement and (y) the recipients of the shares of Common Stock or Related Securities agrees in writing to be bound by the same terms described in the agreement attached hereto as Exhibit D. For purposes of the foregoing, "Related Securities" shall mean any options or warrants or other rights to acquire Shares or any securities exchangeable or exercisable for or convertible into Shares, or to acquire other securities or rights ultimately exchangeable or exercisable for, or convertible into, Shares.

(p) **Future Reports to the Representatives.** During the period of three years hereafter for so long as the Company is subject to the reporting requirements of the Exchange Act during that time, the Company will furnish or make available to the Representatives: c/o Jefferies, at 520 Madison Avenue, New York, New York 10022, Attention: Global Head of Syndicate; c/o Cowen, Attention: Head of Equity Capital Markets, Fax 646-562-1249 with a copy to the General Counsel, Fax 646-562-1124; and c/o BMO, 3 Times Square, New York, New York 10036, Attention: Legal Department, Fax: 212-702-1205: (i) as soon as practicable after the end of each fiscal year, copies of the Annual Report of the Company containing the balance sheet of the Company as of the close of such fiscal year and statements of income, stockholders' equity and cash flows for the year then ended and the opinion thereon of the Company's independent public or certified public accountants; (ii) as soon as practicable after the filing or furnishing thereof, copies of each proxy statement, Annual Report on Form 10-K, Quarterly Report on Form 10-Q, Current Report on Form 8-K or other report filed or furnished by the Company with the Commission or any securities exchange; and (iii) as soon as available, copies of any report or communication of the Company furnished or made available generally to holders of its capital stock; *provided, however*, that the requirements of this Section 3(p) shall be satisfied to the extent that such reports, statement, communications, financial statements or other documents are available on EDGAR.

(q) **Investment Limitation.** The Company shall not invest or otherwise use the proceeds received by the Company from its sale of the Offered Shares in such a manner as would require the Company or any of its subsidiaries to register as an investment company under the Investment Company Act.

(r) **No Stabilization or Manipulation; Compliance with Regulation M.** The Company will not take, and will ensure that no controlled affiliate or other person acting on behalf of the Company will take, directly or indirectly, without giving effect to the activities by the Underwriters, any action designed to or that might reasonably be expected to cause or result in stabilization or manipulation of the price of the Shares or any reference security with respect to the Shares, whether to facilitate the sale or resale of the Offered Shares or otherwise, and the Company will, and shall cause each of its affiliates to, comply with all applicable provisions of Regulation M.

(s) **Enforce Lock-Up Agreements.** During the Lock-up Period, the Company will enforce all agreements between the Company and any of its security holders that restrict or prohibit, expressly or in

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operation, the offer, sale or transfer of Shares or Related Securities or any of the other actions restricted or prohibited under the terms of the form of Lock-up Agreement. In addition, the Company will direct the transfer agent to place stop transfer restrictions upon any such securities of the Company that are bound by such "lock-up" agreements for the duration of the periods contemplated in such agreements, including, without limitation, "lock-up" agreements entered into by the Company's directors, officers and stockholders pursuant to Section 6(j) hereof.

(t) **Company to Provide Interim Financial Statements.** Prior to the First Closing Date and each applicable Option Closing Date, the Company will furnish the Underwriters, as soon as they have been prepared by or are available to the Company, a copy of any unaudited interim financial statements of the Company for any period subsequent to the period covered by the most recent financial statements appearing in the Registration Statement and the Prospectus.

(u) **Amendments and Supplements to Permitted Section 5(d) Communications.** If at any time following the distribution of any Permitted Section 5(d) Communication during the Prospectus Delivery Period, there occurred or occurs an event or development as a result of which such Permitted Section 5(d) Communication included or would include an untrue statement of a material fact or omitted or would omit to state a material fact necessary in order to make the statements therein, in the light of the circumstances existing at that subsequent time, not misleading, the Company will promptly notify the Representatives and will promptly amend or supplement, at its own expense, such Permitted Section 5(d) Communication to eliminate or correct such untrue statement or omission.

(v) **Emerging Growth Company Status.** The Company will promptly notify the Representatives if the Company ceases to be an Emerging Growth Company at any time prior to the later of (i) the time when a prospectus relating to the Offered Shares is not required by the Securities Act to be delivered (whether physically or through compliance with Rule 172 under the Securities Act or any similar rule) and (ii) the expiration of the Lock-Up Period (as defined herein).

(w) **Announcement Regarding Lock-ups.** The Company agrees to announce the Underwriters' intention to release any director or "officer" (within the meaning of Rule 16a-1(f) under the Exchange Act) of the Company from any of the restrictions imposed by any Lock-Up Agreement, by issuing, through a major news service, a press release in form and substance satisfactory to the Representatives promptly following the Company's receipt of any notification from the Representatives in which such intention is indicated, but in any case not later than the close of the third business day prior to the date on which such release or waiver is to become effective; *provided, however*, that nothing shall prevent the Representatives, on behalf of the Underwriters, from announcing the same through a major news service, irrespective of whether the Company has made the required announcement; and *provided, further*, that no such announcement shall be made of any release or waiver granted solely to permit a transfer of securities that is not for consideration and where the transferee has agreed in writing to be bound by the terms of a Lock-Up Agreement in the form set forth as Exhibit D hereto.

Section 4. Payment of Expenses. The Company agrees to pay all costs, fees and expenses incurred in connection with the performance of its obligations hereunder and in connection with the transactions contemplated hereby, including without limitation (i) all expenses incident to the issuance and delivery of the Offered Shares (including all printing and engraving costs), (ii) all fees and expenses of the registrar and transfer agent of the Shares, (iii) all necessary issue, transfer and other stamp taxes in connection with the issuance and sale of the Offered Shares to the Underwriters, (iv) all fees and expenses of the Company's counsel, independent public or certified public accountants and other advisors, (v) all costs and expenses incurred in connection with the preparation, printing, filing, shipping and distribution of the Registration Statement (including financial statements, exhibits, schedules, consents and certificates of experts), the Time of Sale Prospectus, the Prospectus, each free writing prospectus prepared by or on

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behalf of, used by, or referred to by the Company, and each preliminary prospectus, each Permitted Section 5(d) Communication, and all amendments and supplements thereto, and this Agreement, (vi) all filing fees, reasonable attorneys' fees and expenses incurred by the Company or the Underwriters in connection with qualifying or registering (or obtaining exemptions from the qualification or registration of) all or any part of the Offered Shares for offer and sale under the state securities or blue sky laws or the provincial securities laws of Canada, and, if requested by the Representatives, preparing and printing a "Blue Sky Survey" or memorandum and a "Canadian wrapper", and any supplements thereto, advising the Underwriters of such qualifications, registrations and exemptions (not to exceed \$10,000 with respect to this clause (vi)), (vii) the costs, fees and expenses incurred by the Underwriters in connection with determining their compliance with the rules and regulations of FINRA related to the Underwriters' participation in the offering and distribution of the Offered Shares, including any related filing fees and the legal fees of (not to exceed \$30,000 with respect to this clause (vii)), and disbursements by, counsel to the Underwriters, (viii) the costs and expenses of the Company relating to investor presentations on any "road show", any Permitted Section 5(d) Communication or any Section 5(d) Oral Communication undertaken in connection with the offering of the Offered Shares, including, without limitation, expenses associated with the preparation or dissemination of any electronic road show, expenses associated with the production of road show slides and graphics, fees and expenses of any consultants engaged in connection with the road show presentations with the prior approval of the Company, travel and lodging expenses of the representatives, employees and officers of the Company and any such consultants, and 50% of the cost of any aircraft chartered in connection with the road show, with the other 50% being paid by the Underwriters, (ix) the fees and expenses associated

with listing the Offered Shares on the Nasdaq, and (x) all other fees, costs and expenses of the nature referred to in Item 13 of Part II of the Registration Statement. Except as provided in this Section 4 or in Section 7, Section 9 or Section 10 hereof, the Underwriters shall pay their own expenses, including the fees and disbursements of their counsel and their own travel and lodging expenses.

Section 5. Covenant of the Underwriters. Each Underwriter severally and not jointly covenants with the Company not to take any action that would result in the Company being required to file with the Commission pursuant to Rule 433(d) under the Securities Act a free writing prospectus prepared by or on behalf of such Underwriter that otherwise would not, but for such actions, be required to be filed by the Company under Rule 433(d).

Section 6. Conditions of the Obligations of the Underwriters. The respective obligations of the several Underwriters hereunder to purchase and pay for the Offered Shares as provided herein on the First Closing Date and, with respect to the Optional Shares, each Option Closing Date, shall be subject to the accuracy of the representations and warranties on the part of the Company set forth in Section 1 hereof as of the date hereof and as of the First Closing Date as though then made and, with respect to the Optional Shares, as of each Option Closing Date as though then made, to the timely performance by the Company of its covenants and other obligations hereunder, and to each of the following additional conditions:

(a) **Comfort Letter.** On the date hereof, the Representatives shall have received from Ernst & Young LLP, independent registered public accountants for the Company, a letter dated the date hereof addressed to the Underwriters, in form and substance satisfactory to the Representatives, containing statements and information of the type ordinarily included in accountant's "comfort letters" to underwriters, delivered according to Statement of Auditing Standards No. 72 (or any successor bulletin), with respect to the audited and unaudited financial statements and certain financial information contained in the Registration Statement, the Time of Sale Prospectus, and each free writing prospectus, if any.

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(b) **Compliance with Registration Requirements; No Stop Order; No Objection from FINRA.**

(i) The Company shall have filed the Prospectus with the Commission (including the information required by Rule 430A under the Securities Act) in the manner and within the time period required by Rule 424(b) under the Securities Act; or the Company shall have filed a post-effective amendment to the Registration Statement containing the information required by such Rule 430A, and such post-effective amendment shall have become effective.

(ii) No stop order suspending the effectiveness of the Registration Statement or any post-effective amendment to the Registration Statement shall be in effect, and no proceedings for such purpose shall have been instituted or, to the knowledge of the Company, threatened by the Commission.

(iii) FINRA shall have raised no objection to the fairness and reasonableness of the underwriting terms and arrangements.

(c) **No Material Adverse Change.** For the period from and after the date of this Agreement and through and including the First Closing Date and, with respect to any Optional Shares purchased after the First Closing Date, each Option Closing Date, in the judgment of the Representatives there shall not have occurred any Material Adverse Change.

(d) **Opinion of Counsel for the Company.** On each of the First Closing Date and each Option Closing Date the Representatives shall have received the opinion of Goodwin Procter LLP, counsel for the Company, dated as of such date, in form and substance previously agreed to with the Representatives and counsel for the Underwriters.

(e) **Opinion of Intellectual Property Counsel for the Company.** On each of the First Closing Date and each Option Closing Date, the Representatives shall have received the opinion of McCarter & English LLP, counsel for the Company with respect to Intellectual Property matters, dated as of such date, in form and substance previously agreed to with the Representatives and counsel for the Underwriters.

(f) **Opinion of Counsel for the Underwriters.** On each of the First Closing Date and each Option Closing Date the Representatives shall have received the opinion of Cooley LLP, counsel for the Underwriters in connection with the offer and sale of the Offered Shares, dated as of such date, in form and substance satisfactory to the Underwriters.

(g) **CFO Certificate.** On the date hereof and each of the First Closing Date and each Option Closing Date, the Representatives shall have received a certificate executed by the Chief Financial Officer of the Company, on behalf of the Company and not in her individual capacity dated as of such date, in form and substance satisfactory to the Underwriters.

(h) **Officers' Certificate.** On each of the First Closing Date and each Option Closing Date, the Representatives shall have received a certificate executed by the Chief Executive Officer or President of the Company and the Chief Financial Officer of the Company, on behalf of the Company and not in their individual capacities, dated as of such date, to the effect set forth in Section 6(b)(ii) and further to the effect that:

(i) for the period from and including the date of this Agreement through and including such date, there has not occurred any Material Adverse Change;

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(ii) the representations, warranties and covenants of the Company set forth in Section 1 of this Agreement are true and correct with the same force and effect as though expressly made on and as of such date; and

(iii) the Company has complied with all the agreements hereunder and satisfied all the conditions on its part to be performed or satisfied hereunder at or prior to such date.

(i) **Bring-down Comfort Letter.** On each of the First Closing Date and each Option Closing Date the Representatives shall have received from Ernst & Young LLP, independent registered public accountants for the Company, a letter dated such date, in form and substance satisfactory to the Representatives, which letter shall: (i) reaffirm the statements made in the letter furnished by them pursuant to Section 6(a), except that the specified date referred to therein for the carrying out of procedures shall be no more than three (3) business days prior to the First Closing Date or the applicable Option Closing Date, as the case may be; and (ii) cover certain financial information contained in the Prospectus as applicable and agreed to by Ernst & Young LLP.

(j) **Lock-Up Agreements.** On or prior to the date hereof, the Company shall have furnished to the Representatives an agreement in the form of **Exhibit D** hereto from each of the persons listed on **Exhibit E** hereto, and each such agreement shall be in full force and effect on each of the First Closing Date and each Option Closing Date.

(k) **Rule 462(b) Registration Statement.** In the event that a Rule 462(b) Registration Statement is filed in connection with the offering contemplated by this Agreement, such Rule 462(b) Registration Statement shall have been filed with the Commission on the date of this Agreement and shall have become effective automatically upon such filing.

(l) **Approval of Listing.** At the First Closing Date, the Offered Shares shall have been approved for listing on the Nasdaq, subject only to official notice of issuance.

(m) **Additional Documents.** On or before each of the First Closing Date and each Option Closing Date, the Representatives and counsel for the Underwriters shall have received such information, documents and opinions as they may reasonably request for the purposes of enabling them to pass upon the issuance and sale of the Offered Shares as contemplated herein, or in order to evidence the accuracy of any of the representations and warranties, or the satisfaction of any of the conditions or agreements, herein contained; and all proceedings taken by the Company in connection with the issuance and sale of the Offered Shares as contemplated herein and in connection with the other transactions contemplated by this Agreement shall be reasonably satisfactory in form and substance to the Representatives and counsel for the Underwriters.

If any condition specified in this Section 6 is not satisfied when and as required to be satisfied, this Agreement may be terminated by the Representatives by notice from Jefferies, Cowen and BMO to the Company at any time on or prior to the First Closing Date and, with respect to the Optional Shares, at any time on or prior to the applicable Option Closing Date, which termination shall be without liability on the part of any party to any other party, except that Section 4, Section 7, Section 9 and Section 10 shall at all times be effective and shall survive such termination.

Section 7. Reimbursement of Underwriters' Expenses. If this Agreement is terminated by the Representatives pursuant to Section 6, Section 11 or Section 12, or if the sale to the Underwriters of the Offered Shares on the First Closing Date is not consummated because of any refusal, inability or failure on the part of the Company to perform any agreement herein or to comply with any provision hereof, the Company agrees to reimburse the Representatives and the other Underwriters (or

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such Underwriters as have terminated this Agreement with respect to themselves), severally, upon demand for all out-of-pocket expenses that shall have been reasonably incurred by the Representatives and the Underwriters in connection with the proposed purchase and the offering and sale of the Offered Shares, including, but not limited to, fees and disbursements of counsel, printing expenses, travel expenses, postage, facsimile and telephone charges; *provided, however*, that, in the event any such termination is effected after the First Closing Date but prior to any Option Closing Date with respect to the purchase of any Optional Shares, the Company shall only reimburse the Underwriters for all of their out-of-pocket expenses, including the reasonable fees and disbursements of counsel for the Underwriters, incurred after the First Closing Date and in connection with the proposed purchase of any Optional Shares.

Section 8. Effectiveness of this Agreement. This Agreement shall become effective upon the execution and delivery hereof by the parties hereto.

Section 9. Indemnification.

(a) **Indemnification of the Underwriters.** The Company agrees to indemnify and hold harmless each Underwriter, its affiliates, directors, officers, employees and agents, and each person, if any, who controls any Underwriter within the meaning of the Securities Act or the Exchange Act against any loss, claim, damage, liability or expense, as incurred, to which such Underwriter or such affiliate, director, officer, employee, agent or controlling person may become subject, under the Securities Act, the Exchange Act, other federal or state statutory law or regulation, or the laws or regulations of foreign jurisdictions where Offered Shares have been offered or sold or at common law or otherwise (including in settlement of any litigation, if such settlement is effected with the written consent of the Company), insofar as such loss, claim, damage, liability or expense (or actions in respect thereof as contemplated below) arises out of or is based upon (i) any untrue statement or alleged untrue statement of a material fact contained in the Registration Statement, or any amendment thereto, or the omission or alleged omission to state therein a material fact required to be stated therein or necessary to make the statements therein not misleading; or (ii) any untrue statement or alleged untrue statement of a material fact included in any preliminary prospectus, the Time of Sale Prospectus, any free writing prospectus that the Company has used, referred to or filed, or is required to file, pursuant to Rule 433(d) of the Securities Act, any Marketing Material, any Section 5(d) Written Communication or the Prospectus (or any amendment or supplement to the foregoing), or the omission or alleged omission to state therein a material fact necessary in order to make the statements, in the light of the circumstances under which they were made, not misleading; and to reimburse each Underwriter and each such affiliate, director, officer, employee, agent and controlling person for any and all expenses (including reasonable fees and disbursements of counsel) as such expenses are incurred by such Underwriter or such affiliate, director, officer, employee, agent or controlling person in connection with investigating, defending, settling, compromising or paying any such loss, claim, damage, liability, expense or action; *provided, however*, that the foregoing indemnity agreement shall not apply to any loss, claim, damage, liability or expense to the extent, but only to the extent, arising out of or based upon any untrue statement or alleged untrue statement or omission or alleged omission made in reliance upon and in conformity with information relating to any Underwriter furnished to the Company by the Representatives in writing expressly for use in the Registration Statement, any preliminary prospectus, the Time of Sale Prospectus, any such free writing prospectus, any Marketing Material, any Section 5(d) Written Communication or the Prospectus (or any amendment or supplement thereto), it being understood and agreed that the only such information consists of the information described in Section 9(b) below. The indemnity agreement set forth in this Section 9(a) shall be in addition to any liabilities that the Company may otherwise have.

(b) **Indemnification of the Company and its Directors and Officers.** Each Underwriter agrees, severally and not jointly, to indemnify and hold harmless the Company, each of its directors, each of its officers who signed the Registration Statement and each person, if any, who controls the Company

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within the meaning of the Securities Act or the Exchange Act, against any loss, claim, damage, liability or expense, as incurred, to which the Company, or any such director, officer or controlling person may become subject, under the Securities Act, the Exchange Act, or other federal or state statutory law or regulation, or at common law or otherwise (including in settlement of any litigation, if such settlement is effected with the written consent of such Underwriter), insofar as such loss, claim, damage, liability or expense (or actions in respect thereof as contemplated below) arises out of or is based upon (i) any untrue statement or alleged untrue statement of a material fact contained in the Registration Statement, or any amendment thereto, or any omission or alleged omission to state therein a material fact required to be stated therein or necessary to make the statements therein not misleading or (ii) any untrue statement or alleged untrue statement of a material fact included in any preliminary prospectus, the Time of Sale Prospectus, any free writing prospectus, that the Company has used, referred to or filed, or is required to file, pursuant to Rule 433 of the Securities Act, any Marketing Materials, any Section 5(d) Written Communication or the Prospectus (or any such amendment or supplement) or the omission or alleged omission to state therein a material fact necessary in order to make the statements therein, in the light of the circumstances under which they were made, not misleading, in each case to the extent, but only to the extent, that such untrue statement or alleged untrue statement or omission or alleged omission was made in the Registration Statement, such preliminary prospectus, the Time of Sale Prospectus, such free writing prospectus, such Marketing Materials, such Section 5(d) Written Communication or the Prospectus (or any such amendment or supplement), in reliance upon and in conformity with information relating to such Underwriter furnished to the Company by the Representatives in writing expressly for use therein; and to reimburse the Company, or any such director, officer or controlling person for any and all expenses (including the fees and disbursements of counsel) as such expenses are incurred by the Company, or any such director, officer or controlling person in connection with investigating, defending, settling, compromising or paying any such loss, claim, damage, liability, expense or action. The Company hereby acknowledges that the only information that the Representatives have furnished to the Company expressly for use in the Registration Statement, any preliminary prospectus, the Time of Sale Prospectus, any free writing prospectus that the Company has filed, or is required to file, pursuant to Rule 433(d) of the Securities Act, any Marketing Materials, any Section 5(d) Written Communication or the Prospectus (or any amendment or supplement to the foregoing) are the statements set forth in the first sentence of the third paragraph in the section titled "Underwriting," the first two sentences of the first paragraph in the section titled "Underwriting—Commissions and Expenses," and the first sentence of the section titled "Underwriting—Stabilization" in the Preliminary Prospectus and the Prospectus. The indemnity agreement set forth in this Section 9(b) shall be in addition to any liabilities that each Underwriter may otherwise have.

(c) **Notifications and Other Indemnification Procedures.** Promptly after receipt by an indemnified party under this Section 9 of notice of the commencement of any action, such indemnified party will, if a claim in respect thereof is to be made against an indemnifying party under this Section 9, notify the indemnifying party in writing of the commencement thereof, but the omission so to notify the indemnifying party will not relieve the indemnifying party from any liability which it may have to any indemnified party to the extent the indemnifying party is not materially prejudiced as a proximate result of such failure and shall not in any event relieve the indemnifying party from any liability that it may have otherwise than on account of this indemnity agreement. In case any such action is brought against any indemnified party and such indemnified party seeks or intends to seek indemnity from an indemnifying party, the indemnifying party will be entitled to participate in, and, to the extent that it shall elect, jointly with all other indemnifying parties similarly notified, by written notice delivered to the indemnified party promptly after receiving the aforesaid notice from such indemnified party, to assume the defense thereof with counsel reasonably satisfactory to such indemnified party; *provided, however*, that if the defendants in any such action include both the indemnified party and the indemnifying party and the indemnified party shall have reasonably concluded that a conflict may arise between the positions of the indemnifying party and the indemnified party in conducting the defense of any such action or that there may be legal defenses

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available to it and/or other indemnified parties which are different from or additional to those available to the indemnifying party, the indemnified party or parties shall have the right to select separate counsel to assume such legal defenses and to otherwise participate in the defense of such action on behalf of such indemnified party or parties. Upon receipt of notice from the indemnifying party to such indemnified party of such indemnifying party's election so to assume the defense of such action and approval by the indemnified party of counsel, such approval not to be unreasonably withheld or delayed, the indemnifying party will not be liable to such indemnified party under this Section 9 for any legal or other expenses subsequently incurred by such indemnified party in connection with the defense thereof unless (i) the indemnified party shall have employed separate counsel in accordance with the proviso to the preceding sentence (it being understood, however, that the indemnifying party shall not be liable for the fees and expenses of more than one separate counsel (together with one local counsel in each relevant jurisdiction), representing the indemnified parties who are parties to such action), which counsel (together with one local counsel in each relevant jurisdiction) for the indemnified parties shall be selected by the Representatives (in the case of counsel for the indemnified parties referred to in Section 9(a) above) or by the Company (in the case of counsel for the indemnified parties referred to in Section 9(b) above) or (ii) the indemnifying party shall not have employed counsel satisfactory to the indemnified party to represent the indemnified party within a reasonable time after notice of commencement of the action or (iii) the indemnifying party has authorized in writing the employment of counsel for the indemnified party at the expense of the indemnifying party, in each of which cases the fees and expenses of counsel shall be at the expense of the indemnifying party and shall be paid as they are incurred.

(d) **Settlements.** The indemnifying party under this Section 9 shall not be liable for any settlement of any proceeding effected without its written consent, but if settled with such consent or if there be a final judgment for the plaintiff, the indemnifying party agrees to indemnify the indemnified party against any loss, claim, damage, liability or expense by reason of such settlement or judgment. Notwithstanding the foregoing sentence, if at any time an indemnified party shall have requested an indemnifying party to reimburse the indemnified party for fees and expenses of counsel as contemplated by Section 9(c) hereof, the indemnifying party shall be liable for any settlement of any proceeding effected without its written consent if (i) such settlement is entered into more than thirty (30) days after receipt by such indemnifying party of the aforesaid request and (ii) such indemnifying party shall not have reimbursed the indemnified party in accordance with such request prior to the date of such settlement. No indemnifying party shall, without the prior written consent of the indemnified party, effect any settlement, compromise or consent to the entry of judgment in any pending or threatened action, suit or proceeding in respect of which any indemnified party is or could have been a party and indemnity was or could have been sought hereunder by such indemnified party, unless such settlement, compromise or consent includes an unconditional release of such indemnified party from all liability on claims that are the subject matter of such action, suit or proceeding and does not include an admission of fault or culpability or a failure to act by or on behalf of such indemnified party.

Section 10. Contribution. If the indemnification provided for in Section 9 is for any reason held to be unavailable to or otherwise insufficient to hold harmless an indemnified party in respect of any losses, claims, damages, liabilities or expenses referred to therein, then each indemnifying party shall contribute to the aggregate amount paid or payable by such indemnified party, as incurred, as a result of any losses, claims, damages, liabilities or expenses referred to therein (i) in such proportion as is appropriate to reflect the relative benefits received by the Company, on the one hand, and the Underwriters, on the other hand, from the offering of the Offered Shares pursuant to this Agreement or (ii) if the allocation provided by clause (i) above is not permitted by applicable law, in such proportion as is appropriate to reflect not only the relative benefits referred to in clause (i) above but also the relative fault of the Company, on the one hand, and the Underwriters, on the other hand, in connection with the statements or omissions which resulted in such losses, claims, damages, liabilities or expenses, as well as any other relevant equitable considerations. The relative benefits received by the Company, on the one

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hand, and the Underwriters, on the other hand, in connection with the offering of the Offered Shares pursuant to this Agreement shall be deemed to be in the same respective proportions as the total proceeds from the offering of the Offered Shares pursuant to this Agreement (before deducting expenses) received by the Company, and the total underwriting discounts and commissions received by the Underwriters, in each case as set forth on the front cover page of the Prospectus, bear to the aggregate initial public offering price of the Offered Shares as set forth on such cover. The relative fault of the Company, on the one hand, and the Underwriters, on the other hand, shall be determined by reference to, among other things, whether any such untrue or alleged untrue statement of a material fact or omission or alleged omission to state a material fact relates to information supplied by the Company, on the one hand, or the Underwriters, on the other hand, and the parties' relative intent, knowledge, access to information and opportunity to correct or prevent such statement or omission.

The amount paid or payable by a party as a result of the losses, claims, damages, liabilities and expenses referred to above shall be deemed to include, subject to the limitations set forth in Section 9(c), any legal or other fees or expenses reasonably incurred by such party in connection with investigating or defending any action or claim. The provisions set forth in Section 9(c) with respect to notice of commencement of any action shall apply if a claim for contribution is to be made under this Section 10; *provided, however*, that no additional notice shall be required with respect to any action for which notice has been given under Section 9(c) for purposes of indemnification.

The Company and the Underwriters agree that it would not be just and equitable if contribution pursuant to this Section 10 were determined by pro rata allocation (even if the Underwriters were treated as one entity for such purpose) or by any other method of allocation which does not take account of the equitable considerations referred to in this Section 10.

Notwithstanding the provisions of this Section 10, no Underwriter shall be required to contribute any amount in excess of the underwriting discounts and commissions received by such Underwriter in connection with the Offered Shares underwritten by it and distributed to the public. No person guilty of fraudulent misrepresentation (within the meaning of Section 11(f) of the Securities Act) shall be entitled to contribution from any person who was not guilty of such fraudulent misrepresentation. The Underwriters' obligations to contribute pursuant to this Section 10 are several, and not joint, in proportion to their respective underwriting commitments as set forth opposite their respective names on Schedule A. For purposes of this Section 10, each affiliate, director, officer, and employee of an Underwriter and each person, if any, who controls an Underwriter within the meaning of the Securities Act or the Exchange Act shall have the same rights to contribution as such Underwriter, and each director of the Company, each officer of the Company who signed the Registration Statement, and each person, if any, who controls the Company within the meaning of the Securities Act and the Exchange Act shall have the same rights to contribution as the Company.

Section 11. Default of One or More of the Several Underwriters. If, on the First Closing Date or any Option Closing Date any one or more of the several Underwriters shall fail or refuse to purchase Offered Shares that it or they have agreed to purchase hereunder on such date, and the aggregate number of Offered Shares which such defaulting Underwriter or Underwriters agreed but failed or refused to purchase does not exceed 10% of the aggregate number of the Offered Shares to be purchased on such date, the Representatives may make arrangements satisfactory to the Company for the purchase of such Offered Shares by other persons, including any of the Underwriters, but if no such arrangements are made by such date, the other Underwriters shall be obligated, severally and not jointly, in the proportions that the number of Firm Shares set forth opposite their respective names on Schedule A bears to the aggregate number of Firm Shares set forth opposite the names of all such non-defaulting Underwriters, or in such other proportions as may be specified by the Representatives with the consent of the non-defaulting Underwriters, to purchase the Offered Shares which such defaulting Underwriter or Underwriters agreed

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but failed or refused to purchase on such date. If, on the First Closing Date or any Option Closing Date any one or more of the Underwriters shall fail or refuse to purchase Offered Shares and the aggregate number of Offered Shares with respect to which such default occurs exceeds 10% of the aggregate number of Offered Shares to be purchased on such date, and arrangements satisfactory to the Representatives and the Company for the purchase of such Offered Shares are not made within 48 hours after such default, this Agreement shall terminate without liability of any party to any other party except that the provisions of Section 4, Section 7, Section 9 and Section 10 shall at all times be effective and shall survive such termination. In any such case either the Representatives or the Company shall have the right to postpone the First Closing Date or the applicable Option Closing Date, as the case may be, but in no event for longer than seven (7) days in order that the required changes, if any, to the Registration Statement and the Prospectus or any other documents or arrangements may be effected.

As used in this Agreement, the term "Underwriter" shall be deemed to include any person substituted for a defaulting Underwriter under this Section 11. Any action taken under this Section 11 shall not relieve any defaulting Underwriter from liability in respect of any default of such Underwriter under this Agreement.

Section 12. Termination of this Agreement. Prior to the purchase of the Firm Shares by the Underwriters on the First Closing Date, this Agreement may be terminated by the Representatives by notice given to the Company if at any time: (i) trading or quotation in any of the Company's securities shall have been suspended or limited by the Commission or by the Nasdaq, or trading in securities generally on either the Nasdaq or the New York Stock Exchange shall have been suspended or limited, or minimum or maximum prices shall have been generally established on any of such stock exchanges; (ii) a general banking moratorium shall have been declared by any of federal or New York authorities; (iii) there shall have occurred any outbreak or escalation of national or international hostilities or any crisis or calamity, or any change in the United States or international financial markets, or any substantial change or development involving a prospective substantial change in United States' or international political, financial or economic conditions, as in the judgment of the Representatives is material and adverse and makes it impracticable to market the Offered Shares in the manner and on the terms described in the Time of Sale Prospectus or the Prospectus or to enforce contracts for the sale of securities; (iv) in the judgment of the Representatives there shall have occurred any Material Adverse Change; or (v) the Company shall have sustained a loss by strike, fire, flood, earthquake, accident or other calamity of such character as in the judgment of the Representatives may interfere materially with the conduct of

the business and operations of the Company regardless of whether or not such loss shall have been insured. Any termination pursuant to this Section 12 shall be without liability on the part of (a) the Company to any Underwriter, except that the Company shall be obligated to reimburse the expenses of the Representatives and the Underwriters pursuant to Section 4 or Section 7 hereof or (b) any Underwriter to the Company; *provided, however*, that the provisions of Section 9 and Section 10 shall at all times be effective and shall survive such termination.

Section 13. No Advisory or Fiduciary Relationship. The Company acknowledges and agrees that (a) the purchase and sale of the Offered Shares pursuant to this Agreement, including the determination of the public offering price of the Offered Shares and any related discounts and commissions, is an arm's-length commercial transaction between the Company, on the one hand, and the several Underwriters, on the other hand, (b) in connection with the offering contemplated hereby and the process leading to such transaction, each Underwriter is and has been acting solely as a principal and is not the agent or fiduciary of the Company, or its stockholders, creditors, employees or any other party, (c) no Underwriter has assumed or will assume an advisory or fiduciary responsibility in favor of the Company with respect to the offering contemplated hereby or the process leading thereto (irrespective of whether such Underwriter has advised or is currently advising the Company on other matters) and no Underwriter has any obligation to the Company with respect to the offering contemplated hereby except the obligations

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expressly set forth in this Agreement, (d) the Underwriters and their respective affiliates may be engaged in a broad range of transactions that involve interests that differ from those of the Company, and (e) the Underwriters have not provided any legal, accounting, regulatory or tax advice with respect to the offering contemplated hereby and the Company has consulted its own legal, accounting, regulatory and tax advisors to the extent it deemed appropriate.

Section 14. Representations and Indemnities to Survive Delivery. The respective indemnities, agreements, representations, warranties and other statements of the Company, of its officers and of the several Underwriters set forth in or made pursuant to this Agreement will remain in full force and effect, regardless of any investigation made by or on behalf of any Underwriter or the Company or any of its or their partners, officers or directors or any controlling person, as the case may be, and, anything herein to the contrary notwithstanding, will survive delivery of and payment for the Offered Shares sold hereunder and any termination of this Agreement.

Section 15. Notices. All communications hereunder shall be in writing and shall be mailed, hand delivered or telecopied and confirmed to the parties hereto as follows:

If to the Representatives:

Jefferies LLC
520 Madison Avenue
New York, New York 10022
Facsimile: (646) 619-4437
Attention: General Counsel

Cowen and Company, LLC
599 Lexington Avenue
New York, New York 10022
Facsimile: 646-562-1249; 646-562-1124
Attention: Head of Equity Capital Markets; General Counsel

BMO Capital Markets Corp.
3 Times Square, 25th Floor
New York, New York 10036
Facsimile: 212-702-1205
Attention: Legal Department

with a copy (which shall not constitute notice) to:

Cooley LLP
500 Boylston Street
Boston, Massachusetts 02116
Attention: Marc Recht

If to the Company:

Scholar Rock
620 Memorial Drive, 2nd Floor
Cambridge, Massachusetts 02139
Attention: Junlin Ho

with a copy (which shall not constitute notice) to:

Goodwin Procter LLP
100 Northern Avenue
Boston, Massachusetts 02210
Attention: Kingsley L. Taft and Laurie A. Burlingame

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Any party hereto may change the address for receipt of communications by giving written notice to the others.

Section 16. Successors. This Agreement will inure to the benefit of and be binding upon the parties hereto, including any substitute Underwriters pursuant to Section 11 hereof, and to the benefit of the affiliates, directors, officers, employees, agents and controlling persons referred to in Section 9 and Section 10, and in each case their respective successors, and personal representatives, and no other person will have any right or obligation hereunder. The term "successors" shall not include any purchaser of the Offered Shares as such from any of the Underwriters merely by reason of such purchase.

Section 17. Partial Unenforceability. The invalidity or unenforceability of any section, paragraph or provision of this Agreement shall not affect the validity or enforceability of any other section, paragraph or provision hereof. If any section, paragraph or provision of this Agreement is for any reason determined to be invalid or unenforceable, there shall be deemed to be made such minor changes (and only such minor changes) as are necessary to make it valid and enforceable.

Section 18. Governing Law Provisions. This Agreement shall be governed by and construed in accordance with the internal laws of the State of New York applicable to agreements made and to be performed in such state. Any legal suit, action or proceeding arising out of or based upon this Agreement or the transactions contemplated hereby ("**Related Proceedings**") may be instituted in the federal courts of the United States of America located in the Borough of Manhattan in the City of New York or the courts of the State of New York in each case located in the Borough of Manhattan in the City of New York (collectively, the "**Specified Courts**"), and each party irrevocably submits to the exclusive jurisdiction (except for proceedings instituted in regard to the enforcement of a judgment of any such court (a "**Related Judgment**"), as to which such jurisdiction is non-exclusive) of such courts in any such suit, action or proceeding. Service of any process, summons, notice or document by mail to such party's address set forth above shall be effective service of process for any suit, action or other proceeding brought in any such court. The parties irrevocably and unconditionally waive any objection to the laying of venue of any suit, action or other proceeding in the Specified Courts and irrevocably and unconditionally waive and agree not to plead or claim in any such court that any such suit, action or other proceeding brought in any such court has been brought in an inconvenient forum.

Section 19. General Provisions. This Agreement constitutes the entire agreement of the parties to this Agreement and supersedes all prior written or oral and all contemporaneous oral agreements, understandings and negotiations with respect to the subject matter hereof. This Agreement may be executed in two or more counterparts, each one of which shall be an original, with the same effect as if the signatures thereto and hereto were upon the same instrument. This Agreement may not be amended or modified unless in writing by all of the parties hereto, and no condition herein (express or implied) may be waived unless waived in writing by each party whom the condition is meant to benefit. The section headings herein are for the convenience of the parties only and shall not affect the construction or interpretation of this Agreement.

Each of the parties hereto acknowledges that it is a sophisticated business person who was adequately represented by counsel during negotiations regarding the provisions hereof, including, without limitation, the indemnification provisions of Section 9 and the contribution provisions of Section 10, and is fully informed regarding said provisions. Each of the parties hereto further acknowledges that the provisions of Section 9 and Section 10 hereof fairly allocate the risks in light of the ability of the parties to investigate the Company, its affairs and its business in order to assure that adequate disclosure has been made in the Registration Statement, any preliminary prospectus, the Time of Sale Prospectus, each free writing prospectus and the Prospectus (and any amendments and supplements to the foregoing), as contemplated by the Securities Act and the Exchange Act.

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If the foregoing is in accordance with your understanding of our agreement, kindly sign and return to the Company the enclosed copies hereof, whereupon this instrument, along with all counterparts hereof, shall become a binding agreement in accordance with its terms.

Very truly yours,

SCHOLAR ROCK HOLDING CORPORATION

By: _____

Name:
Title:

The foregoing Underwriting Agreement is hereby confirmed and accepted by the Representatives in New York, New York as of the date first above written.

JEFFERIES LLC
COWEN AND COMPANY, LLC
BMO CAPITAL MARKETS CORP.
Acting individually and as Representatives
of the several Underwriters named in
the attached Schedule A.

JEFFERIES LLC

By: _____
Name:
Title:

COWEN AND COMPANY, LLC

By: _____
Name:
Title:

BMO CAPITAL MARKETS CORP.

By: _____
Name:
Title:

[Signature page to Scholar Rock Holding Corporation—Underwriting Agreement]

Schedule A

Underwriters	Number of Firm Shares to be Purchased
Jefferies LLC	[•]
Cowen and Company, LLC	[•]
BMO Capital Markets Corp.	[•]
Wedbush Securities Inc.	[•]
Total	[•]

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Schedule B

Free Writing Prospectuses Included in the Time of Sale Prospectus

[to be added]

B-1

Schedule C

Permitted Section 5(d) Communications

[to be added]

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Exhibit D

Form of Lock-up Agreement

, 2018

JEFFERIES LLC
COWEN AND COMPANY, LLC
As Representatives of the Several Underwriters

c/o Jefferies LLC
520 Madison Avenue
New York, New York 10022

and

c/o Cowen and Company, LLC
599 Lexington Avenue
New York, New York 10022

RE: Scholar Rock Holding Corporation (the "Company")

Ladies & Gentlemen:

The undersigned is an owner of shares of common stock, par value \$0.001 per share, of the Company ("Shares") or of securities convertible into or exchangeable or exercisable for Shares. The Company proposes to conduct a public offering of Shares (the "Offering") for which Jefferies LLC ("Jefferies") and Cowen and Company, LLC ("Cowen") will act as the representatives of the underwriters. The undersigned recognizes that the Offering will benefit each of the Company and the undersigned. The undersigned acknowledges that the underwriters are relying on the representations and agreements of the undersigned contained in this letter agreement in conducting the Offering and, at a subsequent date, in entering into an underwriting agreement (the "Underwriting Agreement") and other underwriting arrangements with the Company with respect to the Offering.

Annex A sets forth definitions for capitalized terms used in this letter agreement that are not defined in the body of this letter agreement. Those definitions are a part of this letter agreement.

In consideration of the foregoing, and for other good and valuable consideration, the receipt and sufficiency of which are hereby acknowledged, the undersigned hereby agrees that, during the Lock-up Period, the undersigned will not (and will cause any Family Member not to), without the prior written consent of Jefferies and Cowen, which may withhold their consent in their sole discretion:

- Sell or Offer to Sell any Shares or Related Securities currently or hereafter owned either of record or beneficially (as defined in Rule 13d-3 under the Exchange Act) by the undersigned or such Family Member,
- enter into any Swap,
- make any demand for, or exercise any right with respect to, the registration under the Securities Act of the offer and sale of any Shares or Related Securities, or cause to be filed a

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registration statement, prospectus or prospectus supplement (or an amendment or supplement thereto) with respect to any such registration, or

- publicly announce any intention to do any of the foregoing.

The foregoing will not apply to:

- (a) the registration of the offer and sale of the Shares, and the sale of the Shares to the underwriters, in each case as contemplated by the Underwriting Agreement;
- (b) the transfer or disposition of Shares or Related Securities (i) by gift, (ii) by will or intestate succession, (iii) to a Family Member or to a trust whose beneficiaries consist exclusively of one or more of the undersigned and/or a Family Member, (iv) if the undersigned is a corporation, limited liability company, partnership or other business entity, (x) to another corporation, limited liability company, partnership or other business entity that controls, is controlled by or is under common control with the undersigned, or (y) in the case of an investment fund, that is managed by, or is under common management with, the undersigned (including, for the avoidance of doubt, a fund managed by the same manager or managing member or general partner or management company or by an entity controlling, controlled by, or under common control with such manager or managing member or general partner or management company as the undersigned or who shares a common investment advisor with the undersigned), to limited partners, members, stockholders or other equityholders of the undersigned, *provided* that, in each case, any such transfer or distribution shall not involve a disposition for value, or (v) pursuant to a qualified domestic order or in connection with a divorce settlement,
- provided, however*, that in any such case, it shall be a condition to such transfer or disposition that:
- each donee, transferee or distributee executes and delivers to Jefferies and Cowen an agreement in form and substance satisfactory to Jefferies and Cowen stating that such donee, transferee or distributee is receiving and holding such Shares and/or Related Securities subject to the provisions of this letter agreement and agrees not to Sell or Offer to Sell such Shares and/or Related Securities, engage in any Swap or engage in any other activities restricted under this letter agreement except in accordance with this letter agreement (as if such donee, transferee or distributee had been an original signatory hereto), and
 - prior to the expiration of the Lock-up Period, no public disclosure or filing under the Exchange Act by any party to the transfer (donor, donee, transferor, transferee, distributor or distributee) shall be required, or made voluntarily, reporting a reduction in beneficial ownership of Shares in connection with such transfer or distribution;
- (c) transactions relating to Shares or Related Securities acquired in open market transactions after the completion of the Offering, *provided* that no public disclosure or filing under the Exchange Act shall be required, or made voluntarily, during the Lock-up Period in connection with any subsequent sales of such Shares or Related Securities acquired in such open market transactions;
- (d) the transfer of Shares or Related Securities upon a vesting event of the Company's securities or upon the exercise of options or warrants to purchase the Company's securities, in each case on a "cashless" or "net exercise" basis or to cover tax withholding obligations of the undersigned in connection with such vesting or exercise, *provided* that such securities were granted pursuant to the Company's stock incentive plan or stock purchase plan described in the Company's registration statement related to the Offering prior to the date of the Underwriting Agreement, *provided further*, that the underlying Shares continue to be subject to the restrictions set forth in this letter agreement and, *provided further*, that no public disclosure or filing under the Exchange

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Act reporting a disposition of Shares shall be required, or made voluntarily, in connection with such vesting or exercise during the Lock-up Period, other than a filing on a Form 4 that reports such disposition under the transaction code "F" and includes a footnote noting the circumstances described in this clause;

- (e) the establishment of a trading plan pursuant to Rule 10b5-1 under the Exchange Act for the transfer of Shares, *provided* that (i) such plan does not provide for the transfer of Shares during the Lock-up Period and (ii) no public disclosure or filing under the Exchange Act regarding the entry into such plan shall be required, or made voluntarily, during the Lock-up Period;
- (f) the conversion of the outstanding preferred stock of the Company into Shares as described in the Company's registration statement related to the Offering, *provided* that such Shares remain subject to the terms of this letter agreement;
- (g) the transfer of Shares or Related Securities to the Company in connection with the termination of the undersigned's employment or other service relationship with the Company, pursuant to agreements under which the Company has the option to repurchase such Shares or Related Securities or a right of first refusal with respect to transfers of such Shares or Related Securities, *provided* that no public disclosure or filing under the Exchange Act reporting a disposition of Shares shall be required, or made voluntarily, during the Lock-up Period, other than a filing on a Form 4 that includes a footnote noting the circumstances described in this clause; or
- (h) the transfer of Shares or Related Securities pursuant to a bona fide third party tender offer for all outstanding Shares of the Company, merger, consolidation or other similar transaction made to all holders of the Shares involving a change of control of the Company (including, without limitation, entering into any lock-up, voting or similar agreement pursuant to which the undersigned may agree to transfer, sell, tender or otherwise dispose of Shares or such other securities in connection with any such transaction, or vote any securities in favor of any such transaction), *provided* that if the tender offer, merger, consolidation or other such transaction is not completed, the Shares owned by the undersigned shall remain subject to the restrictions contained in this letter agreement.

If the undersigned is an officer or director of the Company, the undersigned further agrees that the foregoing provisions shall be equally applicable to any Company-directed Shares the undersigned may purchase or otherwise receive in the Offering (including pursuant to a directed share program).

In addition, if the undersigned is an officer or director of the Company, (i) Jefferies and Cowen agree that, at least three business days before the effective date of any release or waiver of the foregoing restrictions in connection with a transfer of Shares, Jefferies and Cowen will notify the Company of the impending release or waiver, and (ii) the Company (in accordance with the provisions of the Underwriting Agreement) will announce the impending release or waiver by press release through a major news service at least two business days before the effective date of the release or waiver. Any release or waiver granted by Jefferies and Cowen hereunder to any such officer or director shall only be effective two business days after the publication date of such press release. The provisions of this paragraph will not apply if both (a) the release or waiver is effected solely to permit a transfer not for consideration and (b) the transferee has agreed in writing to be bound by the same terms described in this letter agreement that are applicable to the transferor to the extent and for the duration that such terms remain in effect at the time of the transfer.

The undersigned also agrees and consents to the entry of stop transfer instructions with the Company's transfer agent and registrar against the transfer of Shares or Related Securities held by the undersigned and the undersigned's Family Members, if any, except in compliance with the foregoing restrictions.

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With respect to the Offering only, the undersigned waives any registration rights relating to registration under the Securities Act of the offer and sale of any Shares and/or any Related Securities owned either of record or beneficially by the undersigned, including any rights to receive notice of the Offering.

The undersigned confirms that the undersigned has not, and has no knowledge that any Family Member has, directly or indirectly, taken any action designed to or that might reasonably be expected to cause or result in the stabilization or manipulation of the price of any security of the Company to facilitate the sale of the Shares. The undersigned will not, and will cause any Family Member not to take, directly or indirectly, any such action.

Whether or not the Offering occurs as currently contemplated or at all depends on market conditions and other factors. The Offering will only be made pursuant to the Underwriting Agreement, the terms of which are subject to negotiation between the Company and the underwriters.

If (i) the Company, on the one hand, or Jefferies and Cowen, on the other hand, notifies the other in writing that it does not intend to proceed with the Offering, (ii) the Company files an application to withdraw the registration statement related to the Offering, (iii) the Underwriting Agreement is not executed on or before December 31, 2018, or (iv) the Underwriting Agreement (other than the provisions thereof that survive termination) terminates or is terminated prior to the First Closing Date (as defined in the Underwriting Agreement), then in each case, this letter agreement shall automatically, and without any action on the part of any other party, terminate and be of no further force and effect, and the undersigned shall automatically be released from the obligations under this letter agreement.

The undersigned hereby represents and warrants that the undersigned has full power, capacity and authority to enter into this letter agreement. This letter agreement is irrevocable and will be binding on the undersigned and the successors, heirs, personal representatives and assigns of the undersigned. This letter agreement and any claim, controversy or dispute arising under or related to this letter shall be governed by, and construed in accordance with, the laws of the State of New York, without regard to the conflict of laws principles thereof.

Signature

Printed Name of Person Signing

(Indicate capacity of person signing if signing as custodian or trustee, or on behalf of an entity)

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**Certain Defined Terms
Used in Lock-up Agreement**

For purposes of the letter agreement to which this Annex A is attached and of which it is made a part:

- "Call Equivalent Position" shall have the meaning set forth in Rule 16a-1(b) under the Exchange Act.
- "Exchange Act" shall mean the Securities Exchange Act of 1934, as amended.
- "Family Member" shall mean the spouse of the undersigned, an immediate family member of the undersigned or an immediate family member of the undersigned's spouse, in each case living in the undersigned's household or whose principal residence is the undersigned's household (regardless of whether such spouse or family member may at the time be living elsewhere due to educational activities, health care treatment, military service, temporary internship or employment or otherwise). "Immediate family member" as used above shall have the meaning set forth in Rule 16a-1(e) under the Exchange Act.
- "Lock-up Period" shall mean the period beginning on the date hereof and continuing through the close of trading on the date that is 180 days after the date of the prospectus (as defined in the Underwriting Agreement); *provided*, that if (i) during the last 17 days of the 180-day initial lock-up period, the Company issues an earnings release or discloses material news or a material event relating to the Company occurs or (ii) prior to the expiration of such period, the Company announces that it will release earnings results during the 16-day period beginning on the last day of such period, then, in each case, the Lock-up Period will be extended until the

expiration of the 18-day period beginning on the date of the issuance of the earnings release or the disclosure of the material news or occurrence of the material event, as applicable, unless Jefferies and Cowen waive, in writing, such extension. If the initial lock-up period is extended pursuant to the provisions above, "Lock-up Period" shall mean the period described in the first clause of this paragraph, as so extended.

- "Put Equivalent Position" shall have the meaning set forth in Rule 16a-1(h) under the Exchange Act.
- "Related Securities" shall mean any options or warrants or other rights to acquire Shares or any securities exchangeable or exercisable for or convertible into Shares, or to acquire other securities or rights ultimately exchangeable or exercisable for or convertible into Shares.
- "Securities Act" shall mean the Securities Act of 1933, as amended.
- "Sell" or "Offer to Sell" shall mean to:
 - sell, offer to sell, contract to sell or lend,
 - effect any short sale or establish or increase a Put Equivalent Position or liquidate or decrease any Call Equivalent Position,
 - pledge, hypothecate or grant any security interest in, or
 - in any other way transfer or dispose of, in each case whether affected directly or indirectly.

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- "Swap" shall mean any swap, hedge or similar arrangement or agreement that transfers, in whole or in part, the economic risk of ownership of Shares or Related Securities, regardless of whether any such transaction is to be settled in securities, in cash or otherwise.

Capitalized terms not defined in this Annex A shall have the meanings given to them in the body of this lock-up agreement.

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Exhibit E

Directors, Officers and Others Signing Lock-up Agreement

Directors:

David Hallal

Kristina Burow

Jeffrey S. Flier, MD

Michael Gilman, PhD

Amir Nashat, PhD

Timothy A. Springer, PhD

Officers:

Nagesh K. Mahanthappa (Director, President and CEO)

Rhonda M. Chicko (CFO)

Elan Z. Ezickson (COO & Head of Corporate Development)

Yung H. Chyung, M.D. (CMO)

Alan J. Buckler, Ph.D. (CSO)

Others:

Leonard Zon

Boston Children's Medical Center

Polaris Venture Partners VI, L.P.

Polaris Venture Partners Founders Fund VI, L.P.

ARCH Venture Fund VIII, L.P.

TAS Partners, LLC

EcoRI Capital Fund, LP

EcoRI Capital Fund Qualified, LP

KPC Venture Capital, LLC

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JAKII LLC

Fidelity Advisor Series VII: Biotechnology Fund

Fidelity Select Portfolios: Biotechnology Portfolio

Fidelity Mt. Vernon Street Trust: Fidelity Series Growth Company Fund State Street Bank & Trust

Fidelity Growth Company Commingled Pool

Fidelity Mt. Vernon Street Trust: Fidelity Growth Company Fund

Cormorant Global Healthcare Master Fund, LP

Cormorant Private Healthcare Fund I, LP

CRMA SPV, LP

Artal International SCA

Redmile Capital Fund, LP

Redmile Capital Offshore Fund, Ltd.

Redmile Capital Offshore Fund II, Ltd.

**CERTIFICATE OF AMENDMENT
TO THE
AMENDED AND RESTATED
CERTIFICATE OF INCORPORATION
OF
SCHOLAR ROCK HOLDING CORPORATION**

(Pursuant to Section 242 of the
General Corporation Law of the State of Delaware)

Scholar Rock Holding Corporation (the "Corporation"), a corporation organized and existing under and by virtue of the General Corporation Law of the State of Delaware (the "DGCL"), does hereby certify that:

1. The Corporation was originally incorporated on December 18, 2017, and an Amended and Restated Certificate of Incorporation was filed with the Secretary of State of Delaware on December 22, 2017 (the "Charter"). Pursuant to Section 242 of the DGCL, this Certificate of Amendment (this "Amendment") amends certain provisions of the Charter.
2. This Amendment has been approved and duly adopted by the Board of Directors of the Corporation.
3. This Amendment has been duly adopted in accordance with the provisions of Section 242 of the DGCL by written consent of the stockholders holding the requisite number of shares, with written notice to be given as required by Section 228 of the DGCL.
4. The Charter is hereby amended as follows:

The following is hereby inserted into Article FOURTH immediately before the first sentence therein:

"Effective upon the filing of this Certificate of Amendment to the Amended and Restated Certificate of Incorporation with the Secretary of State of the State of Delaware (the "Effective Time"), every 2.8548 shares of Common Stock then issued and outstanding or held in the treasury of the Corporation immediately prior to the Effective Time shall automatically be combined into one (1) share of Common Stock, without any further action by the holders of such shares (the "Reverse Stock Split"). The Reverse Stock Split will be effected on a certificate-by-certificate basis, and any fractional shares resulting from such combination shall be rounded down to the nearest whole share on a certificate-by-certificate basis. No fractional shares shall be issued in connection with the Reverse Stock Split. In lieu of any fractional shares to which a holder would otherwise be entitled, the Corporation shall pay cash equal to such fraction multiplied by the fair market value of a share of Common Stock as determined in good faith by the Corporation's Board of Directors. The Reverse Stock Split shall occur automatically without any further action by the holders of the shares of Common Stock and Preferred Stock affected thereby. All rights, preferences and privileges of the Common Stock and

the Preferred Stock shall be appropriately adjusted to reflect the Reverse Stock Split in accordance with this Amended and Restated Certificate of Incorporation."

[Remainder of page intentionally left blank]

IN WITNESS WHEREOF, this Amendment, having been duly adopted in accordance with Section 242 of the DGCL, has been duly executed by a duly authorized officer of the corporation on this 11th day of May, 2018.

By: /s/ Nagesh K. Mahanthappa
Name: Nagesh K. Mahanthappa
Title: President and Chief Executive Officer

SCHOLAR ROCK HOLDING CORPORATION

THE COMPANY WILL FURNISH WITHOUT CHARGE TO EACH SHAREHOLDER WHO SO REQUESTS, A SUMMARY OF THE POWERS, DESIGNATIONS, PREFERENCES AND RELATIVE, PARTICIPATING, OPTIONAL OR OTHER SPECIAL RIGHTS OF EACH CLASS OF STOCK OF THE COMPANY AND THE QUALIFICATIONS, LIMITATIONS OR RESTRICTIONS OF SUCH PREFERENCES AND RIGHTS, AND THE VARIATIONS IN RIGHTS, PREFERENCES AND LIMITATIONS DETERMINED FOR EACH SERIES, WHICH ARE FIXED BY THE CERTIFICATE OF INCORPORATION OF THE COMPANY, AS AMENDED, AND THE RESOLUTIONS OF THE BOARD OF DIRECTORS OF THE COMPANY, AND THE AUTHORITY OF THE BOARD OF DIRECTORS TO DETERMINE VARIATIONS FOR FUTURE SERIES. SUCH REQUEST MAY BE MADE TO THE OFFICE OF THE SECRETARY OF THE COMPANY OR TO THE TRANSFER AGENT. THE BOARD OF DIRECTORS MAY REQUIRE THE OWNER OF A LOST OR DESTROYED STOCK CERTIFICATE, OR HIS LEGAL REPRESENTATIVES, TO GIVE THE COMPANY A BOND TO INDEMNIFY IT AND ITS TRANSFER AGENTS AND REGISTRARS AGAINST ANY CLAIM THAT MAY BE MADE AGAINST THEM ON ACCOUNT OF THE ALLEGED LOSS OR DESTRUCTION OF ANY SUCH CERTIFICATE.

The following abbreviations, when used in the inscription on the face of this certificate, shall be construed as though they were written out in full according to applicable laws or regulations:

TEN COM - as tenants in common	UNIF GIFT MIN ACTCustodian.....(Minor).....
TEN ENT - as tenants by the entireties		under Uniform Gifts to Minors Act(State).....
JT TEN - as joint tenants with right of survivorship and not as tenants in common	UNIF TRF MIN ACTCustodian (until age.....)(State).....
	(Cust).....(Minor).....under Uniform Transfers to Minors Act.....

Additional abbreviations may also be used though not in the above list.

For value received, _____ hereby sell, assign and transfer unto _____ PLEASE INSERT SOCIAL SECURITY OR OTHER IDENTIFYING NUMBER OF ASSIGNEE

(PLEASE PRINT OR TYPEWRITE NAME AND ADDRESS, INCLUDING POSTAL ZIP CODE, OF ASSIGNEE)

_____ Shares
 of the common stock represented by the within Certificate, and do hereby irrevocably constitute and appoint _____ Attorney
 to transfer the said stock on the books of the within-named Company with full power of substitution in the premises.

Dated: _____ 20____
 Signature: _____
 Signature: _____

Signature(s) Guaranteed: Medallion Guarantee Stamp
THE SIGNATURE(S) SHOULD BE GUARANTEED BY AN ELIGIBLE GUARANTOR INSTITUTION (Banks, Stockbrokers, Savings and Loan Associations and Credit Unions) WITH MEMBERSHIP IN AN APPROVED SIGNATURE GUARANTEE MEDALLION PROGRAM, PURSUANT TO S.E.C. RULE 17A-15.

Notice: The signature to this assignment must correspond with the name as written upon the face of the certificate, in every particular, without alteration or enlargement, or any change whatever.

SECURITY INSTRUCTIONS
 THIS IS WATERMARKED PAPER. DO NOT ACCEPT WITHOUT NOTING WATERMARK. HOLD TO LIGHT TO VERIFY WATERMARK.



The IRS requires that the named transfer agent ("we") report the cost basis of certain shares or units acquired after January 1, 2011. If your shares or units are covered by the legislation, and you requested to sell or transfer the shares or units using a specific cost basis calculation method, then we have processed as you requested. If you did not specify a cost basis calculation method, then we have defaulted to the first in, first out (FIFO) method. Please consult your tax advisor if you need additional information about cost basis.
 If you do not keep in contact with the issuer or do not have any activity in your account for the time period specified by state law, your property may become subject to state unclaimed property laws and transferred to the appropriate state.

1534201

May 14, 2018

Scholar Rock Holding Corporation
620 Memorial Drive, 2nd Floor
Cambridge, MA 02139

Re: Securities Registered under Registration Statement on Form S-1

Ladies and Gentlemen:

We have acted as counsel to you in connection with your filing of a Registration Statement on Form S-1 (File No. 333-224493) (as amended or supplemented, the "Registration Statement") pursuant to the Securities Act of 1933, as amended (the "Securities Act"), relating to the registration of the offering by Scholar Rock Holding Corporation, a Delaware corporation (the "Company") of up to 6,164,000 shares (the "Shares") of the Company's Common Stock, \$0.001 par value per share, including Shares purchasable by the underwriters upon their exercise of an over-allotment option granted to the underwriters by the Company. The Shares are being sold to the several underwriters named in, and pursuant to, an underwriting agreement among the Company and such underwriters (the "Underwriting Agreement").

We have reviewed such documents and made such examination of law as we have deemed appropriate to give the opinions set forth below. We have relied, without independent verification, on certificates of public officials and, as to matters of fact material to the opinions set forth below, on certificates of officers of the Company.

The opinion set forth below is limited to the Delaware General Corporation Law.

Based on the foregoing, we are of the opinion that the Shares have been duly authorized and, upon issuance and delivery against payment therefor in accordance with the terms of the Underwriting Agreement, the Shares will be validly issued, fully paid and non-assessable.

We hereby consent to the inclusion of this opinion as Exhibit 5.1 to the Registration Statement and to the references to our firm under the caption "Legal Matters" in the Registration Statement. In giving our consent, we do not admit that we are in the category of persons whose consent is required under Section 7 of the Securities Act or the rules and regulations thereunder.

Very truly yours,

/s/Goodwin Procter LLP

GOODWIN PROCTER LLP

SCHOLAR ROCK HOLDING CORPORATION

2018 STOCK OPTION AND INCENTIVE PLAN

SECTION 1. GENERAL PURPOSE OF THE PLAN; DEFINITIONS

The name of the plan is the Scholar Rock Holding Corporation 2018 Stock Option and Incentive Plan (the “Plan”). The purpose of the Plan is to encourage and enable the officers, employees, Non-Employee Directors and Consultants of Scholar Rock Holding Corporation (the “Company”) and its Subsidiaries upon whose judgment, initiative and efforts the Company largely depends for the successful conduct of its businesses to acquire a proprietary interest in the Company. It is anticipated that providing such persons with a direct stake in the Company’s welfare will assure a closer identification of their interests with those of the Company and its stockholders, thereby stimulating their efforts on the Company’s behalf and strengthening their desire to remain with the Company.

The following terms shall be defined as set forth below:

“Act” means the Securities Act of 1933, as amended, and the rules and regulations thereunder.

“Administrator” means either the Board or the compensation committee of the Board or a similar committee performing the functions of the compensation committee and which is comprised of not less than two Non-Employee Directors who are independent.

“Award” or “Awards” except where referring to a particular category of grant under the Plan, shall include Incentive Stock Options, Non-Qualified Stock Options, Stock Appreciation Rights, Restricted Stock Units, Restricted Stock Awards, Unrestricted Stock Awards, Cash-Based Awards, and Dividend Equivalent Rights.

“Award Certificate” means a written or electronic document setting forth the terms and provisions applicable to an Award granted under the Plan. Each Award Certificate is subject to the terms and conditions of the Plan.

“Board” means the Board of Directors of the Company.

“Cash-Based Award” means an Award entitling the recipient to receive a cash-denominated payment.

“Code” means the Internal Revenue Code of 1986, as amended, and any successor Code, and related rules, regulations and interpretations.

“Consultant” means any natural person that provides bona fide services to the Company, and such services are not in connection with the offer or sale of securities in a capital-raising transaction and do not directly or indirectly promote or maintain a market for the Company’s securities.

“Dividend Equivalent Right” means an Award entitling the grantee to receive credits based on cash dividends that would have been paid on the shares of Stock specified in the Dividend Equivalent Right (or other award to which it relates) if such shares had been issued to and held by the grantee.

“Effective Date” means the date on which the Plan becomes effective as set forth in Section 19.

“Exchange Act” means the Securities Exchange Act of 1934, as amended, and the rules and regulations thereunder.

“Fair Market Value” of the Stock on any given date means the fair market value of the Stock determined in good faith by the Administrator; provided, however, that if the Stock is admitted to quotation on the National Association of Securities Dealers Automated Quotation System (“Nasdaq”), Nasdaq Global Market, The New York Stock Exchange or another national securities exchange, the determination shall be made by reference to market quotations. If there are no market quotations for such date, the determination shall be made by reference to the last date preceding such date for which there are market quotations; provided further, however, that if the date for which Fair Market Value is determined is the Registration Date, the Fair Market Value shall be the “Price to the Public” (or equivalent) set forth on the cover page for the final prospectus relating to the Company’s Initial Public Offering.

“Incentive Stock Option” means any Stock Option designated and qualified as an “incentive stock option” as defined in Section 422 of the Code.

“Initial Public Offering” means the first underwritten, firm commitment public offering pursuant to an effective registration statement under the Act covering the offer and sale by the Company of its equity securities, or such other event as a result of or following which the Stock shall be publicly held.

“Non-Employee Director” means a member of the Board who is not also an employee of the Company or any Subsidiary.

“Non-Qualified Stock Option” means any Stock Option that is not an Incentive Stock Option.

“Option” or “Stock Option” means any option to purchase shares of Stock granted pursuant to Section 5.

“Registration Date” means the date upon which the registration statement on Form S-1 that is filed by the Company with respect to the Initial Public Offering is declared effective by the Securities and Exchange Commission.

“Restricted Shares” means the shares of Stock underlying a Restricted Stock Award that remain subject to a risk of forfeiture or the Company’s right of repurchase.

“Restricted Stock Award” means an Award of Restricted Shares subject to such restrictions and conditions as the Administrator may determine at the time of grant.

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“Restricted Stock Units” means an Award of stock units subject to such restrictions and conditions as the Administrator may determine at the time of grant.

“Sale Event” shall mean (i) the sale of all or substantially all of the assets of the Company on a consolidated basis to an unrelated person or entity, (ii) a merger, reorganization or consolidation pursuant to which the holders of the Company’s outstanding voting power and outstanding stock immediately prior to such transaction do not own a majority of the outstanding voting power and outstanding stock or other equity interests of the resulting or successor entity (or its ultimate parent, if applicable) immediately upon completion of such transaction, (iii) the sale of all of the Stock of the Company to an unrelated person, entity or group thereof acting in concert, or (iv) any other transaction in which the owners of the Company’s outstanding voting power immediately prior to such transaction do not own at least a majority of the outstanding voting power of the Company or any successor entity immediately upon completion of the transaction other than as a result of the acquisition of securities directly from the Company.

“Sale Price” means the value as determined by the Administrator of the consideration payable, or otherwise to be received by stockholders, per share of Stock pursuant to a Sale Event.

“Section 409A” means Section 409A of the Code and the regulations and other guidance promulgated thereunder.

“Service Relationship” means any relationship as a full-time employee, part-time employee, director or other key person (including Consultants) of the Company or any Subsidiary or any successor entity (e.g., a Service Relationship shall be deemed to continue without interruption in the event an individual’s status changes from full-time employee to part-time employee or Consultant).

“Stock” means the Common Stock, par value \$0.001 per share, of the Company, subject to adjustments pursuant to Section 3.

“Stock Appreciation Right” means an Award entitling the recipient to receive shares of Stock (or cash, to the extent explicitly provided for in the applicable Award Certificate) having a value equal to the excess of the Fair Market Value of the Stock on the date of exercise over the exercise price of the Stock Appreciation Right multiplied by the number of shares of Stock with respect to which the Stock Appreciation Right shall have been exercised.

“Subsidiary” means any corporation or other entity (other than the Company) in which the Company has at least a fifty percent (50%) interest, either directly or indirectly.

“Ten Percent Owner” means an employee who owns or is deemed to own (by reason of the attribution rules of Section 424(d) of the Code) more than ten percent (10%) of the combined voting power of all classes of stock of the Company or any parent or subsidiary corporation.

“Unrestricted Stock Award” means an Award of shares of Stock free of any restrictions.

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SECTION 2. ADMINISTRATION OF PLAN; ADMINISTRATOR AUTHORITY TO SELECT GRANTEEES AND DETERMINE AWARDS

(a) Administration of Plan. The Plan shall be administered by the Administrator.

(b) Powers of Administrator. The Administrator shall have the power and authority to grant Awards consistent with the terms of the Plan, including the power and authority:

(i) to select the individuals to whom Awards may from time to time be granted;

(ii) to determine the time or times of grant, and the extent, if any, of Incentive Stock Options, Non-Qualified Stock Options, Stock Appreciation Rights, Restricted Stock Awards, Restricted Stock Units, Unrestricted Stock Awards, Cash-Based Awards, and Dividend Equivalent Rights, or any combination of the foregoing, granted to any one or more grantees;

(iii) to determine the number of shares of Stock to be covered by any Award;

(iv) to determine and modify from time to time the terms and conditions, including restrictions, not inconsistent with the terms of the Plan, of any Award, which terms and conditions may differ among individual Awards and grantees, and to approve the forms of Award Certificates;

(v) to accelerate at any time the exercisability or vesting of all or any portion of any Award;

(vi) subject to the provisions of Section 5(c), to extend at any time the period in which Stock Options may be exercised; and

(vii) at any time to adopt, alter and repeal such rules, guidelines and practices for administration of the Plan and for its own acts and proceedings as it shall deem advisable; to interpret the terms and provisions of the Plan and any Award (including related written instruments); to make all determinations it deems advisable for the administration of the Plan; to decide all disputes arising in connection with the Plan; and to otherwise supervise the administration of the Plan.

All decisions and interpretations of the Administrator shall be binding on all persons, including the Company and Plan grantees.

(c) Delegation of Authority to Grant Awards. Subject to applicable law, the Administrator, in its discretion, may delegate to a committee consisting of one or more officers of the Company including the Chief Executive Officer of the Company all or part of the Administrator's authority and duties with respect to the granting of Awards to individuals who are (i) not subject to the reporting and other provisions of Section 16 of the Exchange Act and (ii) members of the delegated committee. Any such delegation by the Administrator shall include a limitation as to the amount of Stock underlying Awards that may be granted during the period of the delegation and shall contain guidelines as to the determination of the exercise price and the vesting criteria. The Administrator may revoke or amend the terms of a delegation at

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any time but such action shall not invalidate any prior actions of the Administrator's delegate or delegates that were consistent with the terms of the Plan.

(d) Award Certificate. Awards under the Plan shall be evidenced by Award Certificates that set forth the terms, conditions and limitations for each Award which may include, without limitation, the term of an Award and the provisions applicable in the event employment or service terminates.

(e) Indemnification. Neither the Board nor the Administrator, nor any member of either or any delegate thereof, shall be liable for any act, omission, interpretation, construction or determination made in good faith in connection with the Plan, and the members of the Board and the Administrator (and any delegate thereof) shall be entitled in all cases to indemnification and reimbursement by the Company in respect of any claim, loss, damage or expense (including, without limitation, reasonable attorneys' fees) arising or resulting therefrom to the fullest extent permitted by law and/or under the Company's articles or bylaws or any directors' and officers' liability insurance coverage which may be in effect from time to time and/or any indemnification agreement between such individual and the Company.

(f) Foreign Award Recipients. Notwithstanding any provision of the Plan to the contrary, in order to comply with the laws in other countries in which the Company and its Subsidiaries operate or have employees or other individuals eligible for Awards, the Administrator, in its sole discretion, shall have the power and authority to: (i) determine which Subsidiaries shall be covered by the Plan; (ii) determine which individuals outside the United States are eligible to participate in the Plan; (iii) modify the terms and conditions of any Award granted to individuals outside the United States to comply with applicable foreign laws; (iv) establish subplans and modify exercise procedures and other terms and procedures, to the extent the Administrator determines such actions to be necessary or advisable (and such subplans and/or modifications shall be attached to this Plan as appendices); provided, however, that no such subplans and/or modifications shall increase the share limitations contained in Section 3(a) hereof; and (v) take any action, before or after an Award is made, that the Administrator determines to be necessary or advisable to obtain approval or comply with any local governmental regulatory exemptions or approvals. Notwithstanding the foregoing, the Administrator may not take any actions hereunder, and no Awards shall be granted, that would violate the Exchange Act or any other applicable United States securities law, the Code, or any other applicable United States governing statute or law.

SECTION 3. STOCK ISSUABLE UNDER THE PLAN; MERGERS; SUBSTITUTION

(a) Stock Issuable. The maximum number of shares of Stock reserved and available for issuance under the Plan shall be 3,139,274 shares (the "Initial Limit"), subject to adjustment as provided in Section 3(c), plus on January 1, 2019 and each January 1 thereafter, the number of shares of Stock reserved and available for issuance under the Plan shall be cumulatively increased by 4 percent of the number of shares of Stock issued and outstanding on the immediately preceding December 31, or such lesser increase as determined by the Administrator on or before the immediately preceding December 31 (the "Annual Increase"). Subject to such overall limitation, the maximum aggregate number of shares of Stock that may be issued in the form of Incentive Stock Options shall not exceed the Initial Limit cumulatively increased on

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January 1, 2019 and on each January 1 thereafter by the lesser of the Annual Increase for such year or 1,885,946 shares of Stock, subject in all cases to adjustment as provided in Section 3(c). For purposes of this limitation, the shares of Stock underlying any Awards under the Plan and the Company's 2017 Stock Option and Incentive Plan that are forfeited, canceled, held back upon exercise of an Option or settlement of an Award to cover the exercise price or tax withholding, reacquired by the Company prior to vesting, satisfied without the issuance of Stock or otherwise terminated (other than by exercise) shall be added back to the shares of Stock available for issuance under the Plan. In the event the Company repurchases shares of Stock on the open market, such shares shall not be added to the shares of Stock available for issuance under the Plan. Subject to such overall limitations, shares of Stock may be issued up to such maximum number pursuant to any type or types of Award. The shares available for issuance under the Plan may be authorized but unissued shares of Stock or shares of Stock reacquired by the Company.

(b) Reserved.

(c) Changes in Stock. Subject to Section 3(d) hereof, if, as a result of any reorganization, recapitalization, reclassification, stock dividend, stock split, reverse stock split or other similar change in the Company's capital stock, the outstanding shares of Stock are increased or decreased or are exchanged for a different number or kind of shares or other securities of the Company, or additional shares or new or different shares or other securities of the Company or other non-cash assets are distributed with respect to such shares of Stock or other securities, or, if, as a result of any merger or consolidation, sale of all or substantially all of the assets of the Company, the outstanding shares of Stock are converted into or exchanged for securities of the Company or any successor entity (or a parent or subsidiary thereof), the Administrator shall make an appropriate or proportionate adjustment in (i) the maximum number of shares reserved for issuance under the Plan, including the maximum number of shares that may be issued in the form of Incentive Stock Options, (ii) the number and kind of shares or other securities subject to any then outstanding Awards under the Plan, (iii) the repurchase price, if any, per share subject to each outstanding Restricted Stock Award, and (iv) the exercise price for each share subject to any then outstanding Stock Options and Stock Appreciation Rights under the Plan, without changing the aggregate exercise price (i.e., the exercise price multiplied by the number of Stock Options and Stock Appreciation Rights) as to which such Stock Options and Stock Appreciation Rights remain exercisable. The Administrator shall also make equitable or proportionate adjustments in the number of shares subject to outstanding Awards and the exercise price and the terms of outstanding Awards to take into consideration cash dividends paid other than in the ordinary course or any other extraordinary corporate event. The adjustment by the Administrator shall be final, binding and conclusive. No fractional shares of Stock shall be issued under the Plan resulting from any such adjustment, but the Administrator in its discretion may make a cash payment in lieu of fractional shares.

(d) Mergers and Other Transactions. In the case of and subject to the consummation of a Sale Event, the parties thereto may cause the assumption or continuation of Awards theretofore granted by the successor entity, or the substitution of such Awards with new Awards of the successor entity or parent thereof, with appropriate adjustment as to the number and kind of shares and, if appropriate, the per share exercise prices, as such parties shall agree. To the extent the parties to such Sale Event do not provide for the assumption, continuation or

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substitution of Awards, upon the effective time of the Sale Event, the Plan and all outstanding Awards granted hereunder shall terminate. In such case, except as may be otherwise provided in the relevant Award Certificate, all Options and Stock Appreciation Rights that are not exercisable immediately prior to the effective time of the Sale Event shall become fully exercisable as of the effective time of the Sale Event, all other Awards with time-based vesting, conditions or restrictions shall become fully vested and nonforfeitable as of the effective time of the Sale Event, and all Awards with conditions and restrictions relating to the attainment of performance goals may become vested and nonforfeitable in connection with a Sale Event in the Administrator's discretion or to the extent specified in the relevant Award Certificate. In the event of such termination, (i) the Company shall have the option (in its sole discretion) to make or provide for a payment, in cash or in kind, to the grantees holding Options and Stock Appreciation Rights, in exchange for the cancellation thereof, in an amount equal to the difference between (A) the Sale Price multiplied by the number of shares of Stock subject to outstanding Options and Stock Appreciation Rights (to the extent then exercisable at prices not in excess of the Sale Price) and (B) the aggregate exercise price of all such outstanding Options and Stock Appreciation Rights (provided that, in the case of an Option or Stock Appreciation Right with an exercise price equal to or less than the Sale Price, such Option or Stock Appreciation Right shall be cancelled for no consideration); or (ii) each grantee shall be permitted, within a specified period of time prior to the consummation of the Sale Event as determined by the Administrator, to exercise all outstanding Options and Stock Appreciation Rights (to the extent then exercisable) held by such grantee. The Company shall also have the option (in its sole discretion) to make or provide for a payment, in cash or in kind, to the grantees holding other Awards in an amount equal to the Sale Price multiplied by the number of vested shares of Stock under such Awards.

SECTION 4. ELIGIBILITY

Grantees under the Plan will be such full or part-time officers and other employees, Non-Employee Directors and Consultants of the Company and its Subsidiaries as are selected from time to time by the Administrator in its sole discretion.

SECTION 5. STOCK OPTIONS

(a) Award of Stock Options. The Administrator may grant Stock Options under the Plan. Any Stock Option granted under the Plan shall be in such form as the Administrator may from time to time approve.

Stock Options granted under the Plan may be either Incentive Stock Options or Non-Qualified Stock Options. Incentive Stock Options may be granted only to employees of the Company or any Subsidiary that is a "subsidiary corporation" within the meaning of Section 424(f) of the Code. To the extent that any Option does not qualify as an Incentive Stock Option, it shall be deemed a Non-Qualified Stock Option.

Stock Options granted pursuant to this Section 5 shall be subject to the following terms and conditions and shall contain such additional terms and conditions, not inconsistent with the terms of the Plan, as the Administrator shall deem desirable. If the Administrator so determines,

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Stock Options may be granted in lieu of cash compensation at the optionee's election, subject to such terms and conditions as the Administrator may establish.

(b) **Exercise Price.** The exercise price per share for the Stock covered by a Stock Option granted pursuant to this Section 5 shall be determined by the Administrator at the time of grant but shall not be less than one hundred percent (100%) of the Fair Market Value on the date of grant. In the case of an Incentive Stock Option that is granted to a Ten Percent Owner, the option price of such Incentive Stock Option shall be not less than one hundred ten percent (110%) of the Fair Market Value on the grant date. Notwithstanding the foregoing, Stock Options may be granted with an exercise price per share that is less than one hundred percent (100%) of the Fair Market Value on the date of grant pursuant to a transaction described in, and in a manner consistent with, Section 424(a) of the Code.

(c) **Option Term.** The term of each Stock Option shall be fixed by the Administrator, but no Stock Option shall be exercisable more than ten years after the date the Stock Option is granted. In the case of an Incentive Stock Option that is granted to a Ten Percent Owner, the term of such Stock Option shall be no more than five years from the date of grant.

(d) **Exercisability; Rights of a Stockholder.** Stock Options shall become exercisable at such time or times, whether or not in installments, as shall be determined by the Administrator at or after the grant date. The Administrator may at any time accelerate the exercisability of all or any portion of any Stock Option. An optionee shall have the rights of a stockholder only as to shares acquired upon the exercise of a Stock Option and not as to unexercised Stock Options.

(e) **Method of Exercise.** Stock Options may be exercised in whole or in part, by giving written or electronic notice of exercise to the Company, specifying the number of shares to be purchased. Payment of the purchase price may be made by one or more of the following methods except to the extent otherwise provided in the Option Award Certificate:

(i) In cash, by certified or bank check or other instrument acceptable to the Administrator;

(ii) Through the delivery (or attestation to the ownership following such procedures as the Company may prescribe) of shares of Stock that are not then subject to restrictions under any Company plan. Such surrendered shares shall be valued at Fair Market Value on the exercise date;

(iii) By the optionee delivering to the Company a properly executed exercise notice together with irrevocable instructions to a broker to promptly deliver to the Company cash or a check payable and acceptable to the Company for the purchase price; provided that in the event the optionee chooses to pay the purchase price as so provided, the optionee and the broker shall comply with such procedures and enter into such agreements of indemnity and other agreements as the Company shall prescribe as a condition of such payment procedure; or

(iv) With respect to Stock Options that are not Incentive Stock Options, by a "net exercise" arrangement pursuant to which the Company will reduce the number of shares of Stock issuable upon exercise by the largest whole number of shares with a Fair Market Value that does not exceed the aggregate exercise price.

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Payment instruments will be received subject to collection. The transfer to the optionee on the records of the Company or of the transfer agent of the shares of Stock to be purchased pursuant to the exercise of a Stock Option will be contingent upon receipt from the optionee (or a purchaser acting in his stead in accordance with the provisions of the Stock Option) by the Company of the full purchase price for such shares and the fulfillment of any other requirements contained in the Option Award Certificate or applicable provisions of laws (including the satisfaction of any withholding taxes that the Company is obligated to withhold with respect to the optionee). In the event an optionee chooses to pay the purchase price by previously-owned shares of Stock through the attestation method, the number of shares of Stock transferred to the optionee upon the exercise of the Stock Option shall be net of the number of attested shares. In the event that the Company establishes, for itself or using the services of a third party, an automated system for the exercise of Stock Options, such as a system using an internet website or interactive voice response, then the paperless exercise of Stock Options may be permitted through the use of such an automated system.

(f) **Annual Limit on Incentive Stock Options.** To the extent required for "incentive stock option" treatment under Section 422 of the Code, the aggregate Fair Market Value (determined as of the time of grant) of the shares of Stock with respect to which Incentive Stock Options granted under this Plan and any other plan of the Company or its parent and subsidiary corporations become exercisable for the first time by an optionee during any calendar year shall not exceed \$100,000. To the extent that any Stock Option exceeds this limit, it shall constitute a Non-Qualified Stock Option.

SECTION 6. STOCK APPRECIATION RIGHTS

(a) **Award of Stock Appreciation Rights.** The Administrator may grant Stock Appreciation Rights under the Plan. A Stock Appreciation Right is an Award entitling the recipient to receive shares of Stock (or cash, to the extent explicitly provided for in the applicable Award Certificate) having a value equal to the excess of the Fair Market Value of a share of Stock on the date of exercise over the exercise price of the Stock Appreciation Right multiplied by the number of shares of Stock with respect to which the Stock Appreciation Right shall have been exercised.

(b) **Exercise Price of Stock Appreciation Rights.** The exercise price of a Stock Appreciation Right shall not be less than one hundred percent (100%) of the Fair Market Value of the Stock on the date of grant.

(c) **Grant and Exercise of Stock Appreciation Rights.** Stock Appreciation Rights may be granted by the Administrator independently of any Stock Option granted pursuant to Section 5 of the Plan.

(d) **Terms and Conditions of Stock Appreciation Rights.** Stock Appreciation Rights shall be subject to such terms and conditions as shall be determined on the date of grant by the Administrator. The term of a Stock Appreciation Right may not exceed ten years. The terms and conditions of each such Award shall be determined by the Administrator, and such terms and conditions may differ among individual Awards and grantees.

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SECTION 7. RESTRICTED STOCK AWARDS

(a) **Nature of Restricted Stock Awards.** The Administrator may grant Restricted Stock Awards under the Plan. A Restricted Stock Award is any Award of Restricted Shares subject to such restrictions and conditions as the Administrator may determine at the time of grant. Conditions may be based on continuing employment (or other Service Relationship) and/or achievement of pre-established performance goals and objectives.

(b) **Rights as a Stockholder.** Upon the grant of the Restricted Stock Award and payment of any applicable purchase price, a grantee shall have the rights of a stockholder with respect to the voting of the Restricted Shares and receipt of dividends; provided that if the lapse of restrictions with respect to the Restricted Stock Award is tied to the attainment of performance goals, any dividends paid by the Company during the performance period shall accrue and shall not be paid to the grantee until and to the extent the performance goals are met with respect to the Restricted Stock Award. Unless the Administrator shall otherwise determine, (i) uncertificated Restricted Shares shall be accompanied by a notation on the records of the Company or the transfer agent to the effect that they are subject to forfeiture until such Restricted Shares are vested as provided in Section 7(d) below, and (ii) certificated Restricted Shares shall remain in the possession of the Company until such Restricted Shares are vested as provided in Section 7(d) below, and the grantee shall be required, as a condition of the grant, to deliver to the Company such instruments of transfer as the Administrator may prescribe.

(c) **Restrictions.** Restricted Shares may not be sold, assigned, transferred, pledged or otherwise encumbered or disposed of except as specifically provided herein or in the Restricted Stock Award Certificate. Except as may otherwise be provided by the Administrator either in the Award Certificate or, subject to Section 15 below, in writing after the Award is issued, if a grantee's employment (or other service relationship) with the Company and its Subsidiaries terminates for any reason, any Restricted Shares that have not vested at the time of termination shall automatically and without any requirement of notice to such grantee from or other action by or on behalf of, the Company be deemed to have been reacquired by the Company at its original purchase price (if any) from such grantee or such grantee's legal representative simultaneously with such termination of employment (or other service relationship), and thereafter shall cease to represent any ownership of the Company by the grantee or rights of the grantee as a stockholder. Following such deemed reacquisition of Restricted Shares that are represented by physical certificates, a grantee shall surrender such certificates to the Company upon request without consideration.

(d) **Vesting of Restricted Shares.** The Administrator at the time of grant shall specify the date or dates and/or the attainment of pre-established performance goals, objectives and other conditions on which the non-transferability of the Restricted Shares and the Company's right of repurchase or forfeiture shall lapse. Subsequent to such date or dates and/or the attainment of such pre-established performance goals, objectives and other conditions, the shares on which all restrictions have lapsed shall no longer be Restricted Shares and shall be deemed "vested."

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SECTION 8. RESTRICTED STOCK UNITS

(a) **Nature of Restricted Stock Units.** The Administrator may grant Restricted Stock Units under the Plan. A Restricted Stock Unit is an Award of stock units that may be settled in shares of Stock (or cash, to the extent explicitly provided for in the Award Certificate) upon the satisfaction of such restrictions and conditions at the time of grant. Conditions may be based on continuing employment (or other service relationship) and/or achievement of pre-established performance goals and objectives. The terms and conditions of each such Award shall be determined by the Administrator, and such terms and conditions may differ among individual Awards and grantees. Except in the case of Restricted Stock Units with a deferred settlement date that complies with Section 409A, at the end of the vesting period, the Restricted Stock Units, to the extent vested, shall be settled in the form of shares of Stock. Restricted Stock Units with deferred settlement dates are subject to Section 409A and shall contain such additional terms and conditions as the Administrator shall determine in its sole discretion in order to comply with the requirements of Section 409A.

(b) **Election to Receive Restricted Stock Units in Lieu of Compensation.** The Administrator may, in its sole discretion, permit a grantee to elect to receive a portion of future cash compensation otherwise due to such grantee in the form of an award of Restricted Stock Units. Any such election shall be made in writing and shall be delivered to the Company no later than the date specified by the Administrator and in accordance with Section 409A and such other rules and procedures established by the Administrator. Any such future cash compensation that the grantee elects to defer shall be converted to a fixed number of Restricted Stock Units based on the Fair Market Value of Stock on the date the compensation would otherwise have been paid to the grantee if such payment had not been deferred as provided herein. The Administrator shall have the sole right to determine whether and under what circumstances to permit such elections and to impose such limitations and other terms and conditions thereon as the Administrator deems appropriate. Any Restricted Stock Units that are elected to be received in lieu of cash compensation shall be fully vested, unless otherwise provided in the Award Certificate.

(c) **Rights as a Stockholder.** A grantee shall have the rights as a stockholder only as to shares of Stock acquired by the grantee upon settlement of Restricted Stock Units; provided, however, that the grantee may be credited with Dividend Equivalent Rights with respect to the stock units underlying his Restricted Stock Units, subject to the provisions of Section 11 and such terms and conditions as the Administrator may determine.

(d) **Termination.** Except as may otherwise be provided by the Administrator either in the Award Certificate or, subject to Section 15 below, in writing after the Award is issued, a grantee's right in all Restricted Stock Units that have not vested shall automatically terminate upon the grantee's termination of employment (or cessation of service relationship) with the Company and its Subsidiaries for any reason.

SECTION 9. UNRESTRICTED STOCK AWARDS

Grant or Sale of Unrestricted Stock. The Administrator may grant (or sell at par value or such higher purchase price determined by the Administrator) an Unrestricted Stock Award under the Plan. An Unrestricted Stock Award is an Award pursuant to which the grantee may receive

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shares of Stock free of any restrictions under the Plan. Unrestricted Stock Awards may be granted in respect of past services or other valid consideration, or in lieu of cash compensation due to such grantee.

SECTION 10. CASH-BASED AWARDS

Grant of Cash-Based Awards. The Administrator may grant Cash-Based Awards under the Plan. A Cash-Based Award is an Award that entitles the grantee to a payment in cash upon the attainment of specified performance goals. The Administrator shall determine the maximum duration of the Cash-Based Award, the amount of cash to which the Cash-Based Award pertains, the conditions upon which the Cash-Based Award shall become vested or payable, and such other provisions as the Administrator shall determine. Each Cash-Based Award shall specify a cash-denominated payment amount, formula or payment ranges as determined by the Administrator. Payment, if any, with respect to a Cash-Based Award shall be made in accordance with the terms of the Award and may be made in cash.

SECTION 11. DIVIDEND EQUIVALENT RIGHTS

(a) **Dividend Equivalent Rights.** The Administrator may grant Dividend Equivalent Rights under the Plan. A Dividend Equivalent Right is an Award entitling the grantee to receive credits based on cash dividends that would have been paid on the shares of Stock specified in the Dividend Equivalent Right (or other Award to which it relates) if such shares had been issued to the grantee. A Dividend Equivalent Right may be granted hereunder to any grantee as a component of an award of Restricted Stock Units or as a freestanding award. The terms and conditions of Dividend Equivalent Rights shall be specified in the Award Certificate. Dividend equivalents credited to the holder of a Dividend Equivalent Right may be paid currently or may be deemed to be reinvested in additional shares of Stock, which may thereafter accrue additional equivalents. Any such reinvestment shall be at Fair Market Value on the date of reinvestment or such other price as may then apply under a dividend reinvestment plan sponsored by the Company, if any. Dividend Equivalent Rights may be settled in cash or shares of Stock or a combination thereof, in a single installment or installments. A Dividend Equivalent Right granted as a component of an Award of Restricted Stock Units shall provide that such Dividend Equivalent Right shall be settled only upon settlement or payment of, or lapse of restrictions on, such other Award, and that such Dividend Equivalent Right shall expire or be forfeited or annulled under the same conditions as such other Award.

(b) **Termination.** Except as may otherwise be provided by the Administrator either in the Award Certificate or, subject to Section 15 below, in writing after the Award is issued, a grantee's rights in all Dividend Equivalent Rights shall automatically terminate upon the grantee's termination of employment (or cessation of service relationship) with the Company and its Subsidiaries for any reason.

SECTION 12. TRANSFERABILITY OF AWARDS

(a) **Transferability.** Except as provided in Section 12(b) below, during a grantee's lifetime, his or her Awards shall be exercisable only by the grantee, or by the grantee's legal representative or guardian in the event of the grantee's incapacity. No Awards shall be sold,

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assigned, transferred or otherwise encumbered or disposed of by a grantee other than by will or by the laws of descent and distribution or pursuant to a domestic relations order. No Awards shall be subject, in whole or in part, to attachment, execution, or levy of any kind, and any purported transfer in violation hereof shall be null and void.

(b) **Administrator Action.** Notwithstanding Section 12(a), the Administrator, in its discretion, may provide either in the Award Certificate regarding a given Award or by subsequent written approval that the grantee (who is an employee or director) may transfer his or her Non-Qualified Stock Options to his or her immediate family members, to trusts for the benefit of such family members, or to partnerships in which such family members are the only partners, provided that the transferee agrees in writing with the Company to be bound by all of the terms and conditions of this Plan and the applicable Award. In no event may an Award be transferred by a grantee for value.

(c) **Family Member.** For purposes of Section 12(b), "family member" shall mean a grantee's child, stepchild, grandchild, parent, stepparent, grandparent, spouse, former spouse, sibling, niece, nephew, mother-in-law, father-in-law, son-in-law, daughter-in-law, brother-in-law, or sister-in-law, including adoptive relationships, any person sharing the grantee's household (other than a tenant of the grantee), a trust in which these persons (or the grantee) have more than fifty percent (50%) of the beneficial interest, a foundation in which these persons (or the grantee) control the management of assets, and any other entity in which these persons (or the grantee) own more than fifty percent (50%) of the voting interests.

(d) **Designation of Beneficiary.** To the extent permitted by the Company, each grantee to whom an Award has been made under the Plan may designate a beneficiary or beneficiaries to exercise any Award or receive any payment under any Award payable on or after the grantee's death. Any such designation shall be on a form provided for that purpose by the Administrator and shall not be effective until received by the Administrator. If no beneficiary has been designated by a deceased grantee, or if the designated beneficiaries have predeceased the grantee, the beneficiary shall be the grantee's estate.

SECTION 13. TAX WITHHOLDING

(a) **Payment by Grantee.** Each grantee shall, no later than the date as of which the value of an Award or of any Stock or other amounts received thereunder first becomes includable in the gross income of the grantee for Federal income tax purposes, pay to the Company, or make arrangements satisfactory to the Administrator regarding payment of, any Federal, state, or local taxes of any kind required by law to be withheld by the Company with respect to such income. The Company and its Subsidiaries shall, to the extent permitted by law, have the right to deduct any such taxes from any payment of any kind otherwise due to the grantee. The Company's obligation to deliver evidence of book entry (or stock certificates) to any grantee is subject to and conditioned on tax withholding obligations being satisfied by the grantee.

(b) **Payment in Stock.** Subject to approval by the Administrator, a grantee may elect to have the Company's required tax withholding obligation satisfied, in whole or in part, by authorizing the Company to withhold from shares of Stock to be issued pursuant to any Award a

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number of shares with an aggregate Fair Market Value (as of the date the withholding is effected) that would satisfy the withholding amount due; provided, however, that the amount withheld does not exceed the maximum statutory tax rate or such lesser amount as is necessary to avoid liability accounting treatment. The Administrator may also require Awards to be subject to mandatory share withholding up to the required withholding amount. For purposes of share withholding, the Fair Market Value of withheld shares shall be determined in the same manner as the value of Stock includable in income of the Participants. The required tax withholding obligation may also be satisfied, in whole or in part, by an arrangement whereby a certain number of shares of Stock issued pursuant to any Award are immediately sold and proceeds from such sale are remitted to the Company in an amount that would satisfy the withholding amount due.

SECTION 14. SECTION 409A AWARDS

To the extent that any Award is determined to constitute "nonqualified deferred compensation" within the meaning of Section 409A (a "409A Award"), the Award shall be subject to such additional rules and requirements as specified by the Administrator from time to time in order to comply with Section 409A. In this regard, if any amount under a 409A Award is payable upon a "separation from service" (within the meaning of Section 409A) to a grantee who is then considered a "specified employee" (within the meaning of Section 409A), then no such payment shall be made prior to the date that is the earlier of (i) six months and one day after the grantee's separation from service, or (ii) the grantee's death, but only to the extent such delay is necessary to prevent such payment from being subject to interest, penalties and/or additional tax imposed pursuant to Section 409A. Further, the settlement of any 409A Award may not be accelerated except to the extent permitted by Section 409A.

SECTION 15. TERMINATION OF EMPLOYMENT, TRANSFER, LEAVE OF ABSENCE, ETC.

(a) **Termination of Employment.** If the grantee's Service Relationship is with a Subsidiary and such Subsidiary ceases to be a Subsidiary, the grantee shall be deemed to have terminated his or her Service Relationship for purposes of the Plan.

(b) For purposes of the Plan, the following events shall not be deemed a termination of employment:

(i) a transfer to the employment of the Company from a Subsidiary or from the Company to a Subsidiary, or from one Subsidiary to another; or

(ii) an approved leave of absence for military service or sickness, or for any other purpose approved by the Company, if the employee's right to re-employment is guaranteed either by a statute or by contract or under the policy pursuant to which the leave of absence was granted or if the Administrator otherwise so provides in writing.

SECTION 16. AMENDMENTS AND TERMINATION

The Board may, at any time, amend or discontinue the Plan and the Administrator may, at any time, amend or cancel any outstanding Award for the purpose of satisfying changes in law or

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for any other lawful purpose, but no such action shall adversely affect rights under any outstanding Award without the holder's consent. Except as provided in Section 3(c) or 3(d), without prior stockholder approval, in no event may the Administrator exercise its discretion to reduce the exercise price of outstanding Stock Options or Stock Appreciation Rights or effect repricing through cancellation and re-grants or cancellation of Stock Options or Stock Appreciation Rights in exchange for cash or other Awards. To the extent required under the rules of any securities exchange or market system on which the Stock is listed, to the extent determined by the Administrator to be required by the Code to ensure that Incentive Stock Options granted under the Plan are qualified under Section 422 of the Code, Plan amendments shall be subject to approval by the Company stockholders entitled to vote at a meeting of stockholders. Nothing in this Section 16 shall limit the Administrator's authority to take any action permitted pursuant to Section 3(c) or 3(d).

SECTION 17. STATUS OF PLAN

With respect to the portion of any Award that has not been exercised and any payments in cash, Stock or other consideration not received by a grantee, a grantee shall have no rights greater than those of a general creditor of the Company unless the Administrator shall otherwise expressly determine in connection with any Award or Awards. In its sole discretion, the Administrator may authorize the creation of trusts or other arrangements to meet the Company's obligations to deliver Stock or make payments with respect to Awards hereunder, provided that the existence of such trusts or other arrangements is consistent with the foregoing sentence.

SECTION 18. GENERAL PROVISIONS

(a) **No Distribution.** The Administrator may require each person acquiring Stock pursuant to an Award to represent to and agree with the Company in writing that such person is acquiring the shares without a view to distribution thereof.

(b) Issuance of Stock. To the extent certificated, stock certificates to grantees under this Plan shall be deemed delivered for all purposes when the Company or a stock transfer agent of the Company shall have mailed such certificates in the United States mail, addressed to the grantee, at the grantee's last known address on file with the Company. Uncertificated Stock shall be deemed delivered for all purposes when the Company or a Stock transfer agent of the Company shall have given to the grantee by electronic mail (with proof of receipt) or by United States mail, addressed to the grantee, at the grantee's last known address on file with the Company, notice of issuance and recorded the issuance in its records (which may include electronic "book entry" records). Notwithstanding anything herein to the contrary, the Company shall not be required to issue or deliver any evidence of book entry or certificates evidencing shares of Stock pursuant to the exercise or settlement of any Award, unless and until the Administrator has determined, with advice of counsel (to the extent the Administrator deems such advice necessary or advisable), that the issuance and delivery is in compliance with all applicable laws, regulations of governmental authorities and, if applicable, the requirements of any exchange on which the shares of Stock are listed, quoted or traded. Any Stock issued pursuant to the Plan shall be subject to any stop-transfer orders and other restrictions as the Administrator deems necessary or advisable to comply with federal, state or foreign jurisdiction, securities or other laws, rules and quotation system on which the Stock is listed, quoted or

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traded. The Administrator may place legends on any Stock certificate or notations on any book entry to reference restrictions applicable to the Stock. In addition to the terms and conditions provided herein, the Administrator may require that an individual make such reasonable covenants, agreements, and representations as the Administrator, in its discretion, deems necessary or advisable in order to comply with any such laws, regulations, or requirements. The Administrator shall have the right to require any individual to comply with any timing or other restrictions with respect to the settlement or exercise of any Award, including a window-period limitation, as may be imposed in the discretion of the Administrator.

(c) Stockholder Rights. Until Stock is deemed delivered in accordance with Section 18(b), no right to vote or receive dividends or any other rights of a stockholder will exist with respect to shares of Stock to be issued in connection with an Award, notwithstanding the exercise of a Stock Option or any other action by the grantee with respect to an Award.

(d) Other Compensation Arrangements; No Employment Rights. Nothing contained in this Plan shall prevent the Board from adopting other or additional compensation arrangements, including trusts, and such arrangements may be either generally applicable or applicable only in specific cases. The adoption of this Plan and the grant of Awards do not confer upon any employee any right to continued employment with the Company or any Subsidiary.

(e) Trading Policy Restrictions. Option exercises and other Awards under the Plan shall be subject to the Company's insider trading policies and procedures, as in effect from time to time.

(f) Clawback Policy. Awards under the Plan shall be subject to the Company's clawback policy, as in effect from time to time.

SECTION 19. EFFECTIVE DATE OF PLAN

This Plan shall become effective upon the date immediately preceding the Registration Date following stockholder approval in accordance with applicable state law, the Company's bylaws and articles of incorporation, and applicable stock exchange rules. No grants of Stock Options and other Awards may be made hereunder after the tenth anniversary of the Effective Date and no grants of Incentive Stock Options may be made hereunder after the tenth anniversary of the date the Plan is approved by the Board.

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SECTION 20. GOVERNING LAW

This Plan and all Awards and actions taken thereunder shall be governed by, and construed in accordance with the General Corporation Law of the State of Delaware as to matters within the scope thereof, and as to all other matters shall be governed by and construed in accordance with the internal laws of the Commonwealth of Massachusetts, applied without regard to conflict of law principles.

DATE APPROVED BY BOARD OF DIRECTORS: May 2, 2018

DATE APPROVED BY STOCKHOLDERS: May 11, 2018

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INCENTIVE STOCK OPTION AGREEMENT UNDER THE SCHOLAR ROCK HOLDING CORPORATION 2018 STOCK OPTION AND INCENTIVE PLAN

Name of Optionee:

No. of Option Shares:

Option Exercise Price per Share:

\$

[FMV on Grant Date (110% of FMV if a 10% owner)]

Grant Date:

Expiration Date:

[up to 10 years (5 if a 10% owner)]

Pursuant to the Scholar Rock Holding Corporation 2018 Stock Option and Incentive Plan as amended through the date hereof (the "Plan"), Scholar Rock Holding Corporation (the "Company") hereby grants to the Optionee named above an option (the "Stock Option") to purchase on or prior to the Expiration Date specified above all or part of the number of shares of Common Stock, par value \$0.001 per share (the "Stock"), of the Company specified above at the Option Exercise Price per Share specified above subject to the terms and conditions set forth herein and in the Plan.

1. Exercisability Schedule. No portion of this Stock Option may be exercised until such portion shall have become exercisable. Except as set forth below, and subject to the discretion of the Administrator (as defined in Section 2 of the Plan) to accelerate the exercisability schedule hereunder, this Stock Option shall be exercisable with respect to the following number of Option Shares on the dates indicated so long as the Optionee remains an employee of the Company or a Subsidiary on such dates:

Incremental Number of Option Shares Exercisable*	Exercisability Date
()%	
()%	
()%	
()%	
()%	

* Max. of \$100,000 per yr.

Once exercisable, this Stock Option shall continue to be exercisable at any time or times prior to the close of business on the Expiration Date, subject to the provisions hereof and of the Plan.

2. Manner of Exercise.

(a) The Optionee may exercise this Stock Option only in the following manner: from time to time on or prior to the Expiration Date of this Stock Option, the Optionee may give written notice to the Administrator of his or her election to purchase some or all of the Option Shares purchasable at the time of such notice. This notice shall specify the number of Option Shares to be purchased.

Payment of the purchase price for the Option Shares may be made by one or more of the following methods: (i) in cash, by certified or bank check or other instrument acceptable to the Administrator; (ii) through the delivery (or attestation to the ownership) of shares of Stock that have been purchased by the Optionee on the open market or that are beneficially owned by the Optionee and are not then subject to any restrictions under any Company plan and that otherwise satisfy any holding periods as may be required by the Administrator; or (iii) by the Optionee delivering to the Company a properly executed exercise notice together with irrevocable instructions to a broker to promptly deliver to the Company cash or a check payable and acceptable to the Company to pay the option purchase price, provided that in the event the Optionee chooses to pay the option purchase price as so provided, the Optionee and the broker shall comply with such procedures and enter into such agreements of indemnity and other agreements as the Administrator shall prescribe as a condition of such payment procedure; or (iv) a combination of (i), (ii) and (iii) above. Payment instruments will be received subject to collection.

The transfer to the Optionee on the records of the Company or of the transfer agent of the Option Shares will be contingent upon (i) the Company's receipt from the Optionee of the full purchase price for the Option Shares, as set forth above, (ii) the fulfillment of any other requirements contained herein or in the Plan or in any other agreement or provision of laws, and (iii) the receipt by the Company of any agreement, statement or other evidence that the Company may require to satisfy itself that the issuance of Stock to be purchased pursuant to the exercise of Stock Options under the Plan and any subsequent resale of the shares of Stock will be in compliance with applicable laws and regulations. In the event the Optionee chooses to pay the purchase price by previously-owned shares of Stock through the attestation method, the number of shares of Stock transferred to the Optionee upon the exercise of the Stock Option shall be net of the Shares attested to.

(b) The shares of Stock purchased upon exercise of this Stock Option shall be transferred to the Optionee on the records of the Company or of the transfer agent upon compliance to the satisfaction of the Administrator with all requirements under applicable laws or regulations in connection with such transfer and with the requirements hereof and of the Plan. The determination of the Administrator as to such compliance shall be final and binding on the Optionee. The Optionee shall not be deemed to be the holder of, or to have any of the rights of a holder with respect to, any shares of Stock subject to this Stock Option unless and until this Stock Option shall have been exercised pursuant to the terms hereof, the Company or the transfer agent shall have transferred the shares to the Optionee, and the Optionee's name shall have been entered as the stockholder of record on the books of the Company. Thereupon, the Optionee shall have full voting, dividend and other ownership rights with respect to such shares of Stock.

(c) The minimum number of shares with respect to which this Stock Option may be exercised at any one time shall be 100 shares, unless the number of shares with respect to which this Stock Option is being exercised is the total number of shares subject to exercise under this Stock Option at the time.

(d) Notwithstanding any other provision hereof or of the Plan, no portion of this Stock Option shall be exercisable after the Expiration Date hereof.

3. **Termination of Service Relationship.** If the Optionee's Service Relationship by the Company or a Subsidiary (as defined in the Plan) is terminated, the period within which to exercise the Stock Option may be subject to earlier termination as set forth below.

(a) **Termination Due to Death.** If the Optionee's Service Relationship terminates by reason of the Optionee's death, any portion of this Stock Option outstanding on such date, to the extent exercisable on the date of death, may thereafter be exercised by the Optionee's legal representative or legatee for a period of 12 months from the date of death or until the Expiration Date, if earlier. Any portion of this Stock Option that is not exercisable on the date of death shall terminate immediately and be of no further force or effect.

(b) **Termination Due to Disability.** If the Optionee's Service Relationship terminates by reason of the Optionee's disability (as determined by the Administrator), any portion of this Stock Option outstanding on such date, to the extent exercisable on the date of such termination of Service Relationship, may thereafter be exercised by the Optionee for a period of 12 months from the date of disability or until the Expiration Date, if earlier. Any portion of this Stock Option that is not exercisable on the date of disability shall terminate immediately and be of no further force or effect.

(c) **Termination for Cause.** If the Optionee's Service Relationship terminates for Cause, any portion of this Stock Option outstanding on such date shall terminate immediately and be of no further force and effect. For purposes hereof, "Cause" shall mean, unless otherwise provided in an employment agreement (or similar services agreements) between the Company and the Optionee, a determination by the Administrator that the Optionee shall be dismissed as a result of (i) any material breach by the Optionee of any agreement between the Optionee and the Company; (ii) the conviction of, indictment for or plea of nolo contendere by the Optionee to a felony or a crime involving moral turpitude; or (iii) any material misconduct or willful and deliberate non-performance (other than by reason of disability) by the Optionee of the Optionee's duties to the Company.

(d) **Other Termination.** If the Optionee's Service Relationship terminates for any reason other than the Optionee's death, the Optionee's disability, or Cause, and unless otherwise determined by the Administrator, any portion of this Stock Option outstanding on such date may be exercised, to the extent exercisable on the date of termination, for a period of three months from the date of termination or until the Expiration Date, if earlier. Any portion of this Stock Option that is not exercisable on the date of termination shall terminate immediately and be of no further force or effect.

The Administrator's determination of the reason for termination of the Optionee's Service Relationship shall be conclusive and binding on the Optionee and his or her representatives or legatees.

4. **Incorporation of Plan.** Notwithstanding anything herein to the contrary, this Stock Option shall be subject to and governed by all the terms and conditions of the Plan, including the powers of the Administrator set forth in Section 2(b) of the Plan. Capitalized terms in this Agreement shall have the meaning specified in the Plan, unless a different meaning is specified herein.

5. **Transferability.** This Agreement is personal to the Optionee, is non-assignable and is not transferable in any manner, by operation of law or otherwise, other than by will or the laws of descent and distribution. This Stock Option is exercisable, during the Optionee's lifetime, only by the Optionee, and thereafter, only by the Optionee's legal representative or legatee.

6. **Status of the Stock Option.** This Stock Option is intended to qualify as an "incentive stock option" under Section 422 of the Internal Revenue Code of 1986, as amended (the "Code"), but the Company does not represent or warrant that this Stock Option qualifies as such. The Optionee should consult with his or her own tax advisors regarding the tax effects of this Stock Option and the requirements necessary to obtain favorable income tax treatment under Section 422 of the Code, including, but not limited to, holding period requirements. To the extent any portion of this Stock Option does not so qualify as an "incentive stock option," such portion shall be deemed to be a non-qualified stock option. If the Optionee intends to dispose or does dispose (whether by sale, gift, transfer or otherwise) of any Option Shares within the one-year period beginning on the date after the transfer of such shares to him or her, or within the two-year period beginning on the day after the grant of this Stock Option, he or she will so notify the Company within 30 days after such disposition.

7. **Tax Withholding.** The Optionee shall, not later than the date as of which the exercise of this Stock Option becomes a taxable event for Federal income tax purposes, pay to the Company or make arrangements satisfactory to the Administrator for payment of any Federal, state, and local taxes required by law to be withheld on account of such taxable event. The Company shall have the authority to cause the required tax withholding obligation to be satisfied, in whole or in part, by withholding from shares of Stock to be issued to the Optionee a number of shares of Stock with an aggregate Fair Market Value that would satisfy the withholding amount due; provided, however, that the amount withheld does not exceed the maximum statutory tax rate or such lesser amount as is necessary to avoid adverse accounting treatment or as determined by the Administrator.

8. **No Obligation to Continue Service Relationship.** Neither the Company nor any Subsidiary is obligated by or as a result of the Plan or this Agreement to continue the Optionee's Service Relationship and neither the Plan nor this Agreement shall interfere in any way with the right of the Company or any Subsidiary to terminate the Service Relationship of the Optionee at any time.

9. **Integration.** This Agreement constitutes the entire agreement between the parties with respect to this Stock Option and supersedes all prior agreements and discussions between the parties concerning such subject matter.

10. **Data Privacy Consent.** In order to administer the Plan and this Agreement and to implement or structure future equity grants, the Company, its subsidiaries and affiliates and certain agents thereof (together, the "Relevant Companies") may process any and all personal or professional data, including but not limited to Social Security or other identification number, home address and telephone number, date of birth and other information that is necessary or desirable for the administration of the Plan and/or this Agreement (the "Relevant Information"). By entering into this Agreement, the Optionee (i) authorizes the Company to collect, process, register and transfer to the Relevant Companies all Relevant Information; (ii) waives any privacy rights the Optionee may have with respect to the Relevant Information; (iii) authorizes the Relevant Companies to store and transmit such information in electronic form; and (iv) authorizes the transfer of the Relevant Information to any jurisdiction in which the Relevant Companies consider appropriate. The Optionee shall have access to, and the right to change, the Relevant Information. Relevant Information will only be used in accordance with applicable law.

11. **Notices.** Notices hereunder shall be mailed or delivered to the Company at its principal place of business and shall be mailed or delivered to the Optionee at the address on file

with the Company or, in either case, at such other address as one party may subsequently furnish to the other party in writing.

SCHOLAR ROCK HOLDING CORPORATION

By: _____
Title:

The foregoing Agreement is hereby accepted and the terms and conditions thereof hereby agreed to by the undersigned. Electronic acceptance of this Agreement pursuant to the Company's instructions to the Optionee (including through an online acceptance process) is acceptable.

Dated: _____

Optionee's Signature

Optionee's name and address:

**NON-QUALIFIED STOCK OPTION AGREEMENT
FOR NON-EMPLOYEE DIRECTORS
UNDER THE SCHOLAR ROCK HOLDING CORPORATION
2018 STOCK OPTION AND INCENTIVE PLAN**

Name of Optionee:

No. of Option Shares:

Option Exercise Price per Share: \$

[FMV on Grant Date]

Grant Date:

Expiration Date:

Pursuant to the Scholar Rock Holding Corporation 2018 Stock Option and Incentive Plan as amended through the date hereof (the "Plan"), Scholar Rock Holding Corporation (the "Company") hereby grants to the Optionee named above, who is a Director of the Company but is not an employee of the Company, an option (the "Stock Option") to purchase on or prior to the Expiration Date specified above all or part of the number of shares of Common Stock, par value \$0.001 per share (the "Stock"), of the Company specified above at the Option Exercise Price per Share specified above subject to the terms and conditions set forth herein and in the Plan. This Stock Option is not intended to be an "incentive stock option" under Section 422 of the Internal Revenue Code of 1986, as amended.

1. **Exercisability Schedule.** No portion of this Stock Option may be exercised until such portion shall have become exercisable. Except as set forth below, and subject to the discretion of the Administrator (as defined in Section 2 of the Plan) to accelerate the exercisability schedule hereunder, this Stock Option shall be exercisable with respect to the following number of Option Shares on the dates indicated so long as the Optionee remains in service as a member of the Board on such dates:

Incremental Number of Option Shares Exercisable	Exercisability Date
()%	
()%	
()%	
()%	
()%	

Once exercisable, this Stock Option shall continue to be exercisable at any time or times prior to the close of business on the Expiration Date, subject to the provisions hereof and of the Plan.

2. **Manner of Exercise.**

(a) The Optionee may exercise this Stock Option only in the following manner: from time to time on or prior to the Expiration Date of this Stock Option, the Optionee may give written notice to the Administrator of his or her election to purchase some or all of the Option Shares purchasable at the time of such notice. This notice shall specify the number of Option Shares to be purchased.

Payment of the purchase price for the Option Shares may be made by one or more of the following methods: (i) in cash, by certified or bank check or other instrument acceptable to the Administrator; (ii) through the delivery (or attestation to the ownership) of shares of Stock that have been purchased by the Optionee on the open market or that are beneficially owned by the Optionee and are not then subject to any restrictions under any Company plan and that otherwise satisfy any holding periods as may be required by the Administrator; (iii) by the Optionee delivering to the Company a properly executed exercise notice together with irrevocable instructions to a broker to promptly deliver to the Company cash or a check payable and acceptable to the Company to pay the option purchase price, provided that in the event the Optionee chooses to pay the option purchase price as so provided, the Optionee and the broker shall comply with such procedures and enter into such agreements of indemnity and other agreements as the Administrator shall prescribe as a condition of such payment procedure; (iv) by a "net exercise" arrangement pursuant to which the Company will reduce the number of shares of Stock issuable upon exercise by the largest whole number of shares with a Fair Market Value that does not exceed the aggregate exercise price; or (v) a combination of (i), (ii), (iii) and (iv) above. Payment instruments will be received subject to collection.

The transfer to the Optionee on the records of the Company or of the transfer agent of the Option Shares will be contingent upon (i) the Company's receipt from the Optionee of the full purchase price for the Option Shares, as set forth above, (ii) the fulfillment of any other requirements contained herein or in the Plan or in any other agreement or provision of laws, and (iii) the receipt by the Company of any agreement, statement or other evidence that the Company may require to satisfy itself that the issuance of Stock to be purchased pursuant to the exercise of Stock Options under the Plan and any subsequent resale of the shares of Stock will be in compliance with applicable laws and regulations. In the event the Optionee chooses to pay the purchase price by previously-owned shares of Stock through the attestation method, the number of shares of Stock transferred to the Optionee upon the exercise of the Stock Option shall be net of the Shares attested to.

(b) The shares of Stock purchased upon exercise of this Stock Option shall be transferred to the Optionee on the records of the Company or of the transfer agent upon compliance to the satisfaction of the Administrator with all requirements under applicable laws or regulations in connection with such transfer and with the requirements hereof and of the Plan. The determination of the Administrator as to such compliance shall be final and binding on the Optionee. The Optionee shall not be deemed to be the holder of, or to have any of the rights of a

holder with respect to, any shares of Stock subject to this Stock Option unless and until this Stock Option shall have been exercised pursuant to the terms hereof, the Company or the transfer agent shall have transferred the shares to the Optionee, and the Optionee's name shall have been entered as the stockholder of record on the books of the Company. Thereupon, the Optionee shall have full voting, dividend and other ownership rights with respect to such shares of Stock.

(c) The minimum number of shares with respect to which this Stock Option may be exercised at any one time shall be 100 shares, unless the number of shares with respect to which this Stock Option is being exercised is the total number of shares subject to exercise under this Stock Option at the time.

(d) Notwithstanding any other provision hereof or of the Plan, no portion of this Stock Option shall be exercisable after the Expiration Date hereof.

3. **Termination as Director.** If the Optionee ceases to be a Director of the Company, the period within which to exercise the Stock Option may be subject to earlier termination as set forth below.

(a) **Termination Due to Death.** If the Optionee's service as a Director terminates by reason of the Optionee's death, any portion of this Stock Option outstanding on such date, to the extent exercisable on the date of death, may thereafter be exercised by the Optionee's legal representative or legatee for a period of 12 months from the date of death or until the Expiration Date, if earlier. Any portion of this Stock Option that is not exercisable on the date of death shall terminate immediately and be of no further force or effect.

(b) **Other Termination.** If the Optionee ceases to be a Director for any reason other than the Optionee's death, any portion of this Stock Option outstanding on such date may be exercised, to the extent exercisable on the date the Optionee ceased to be a Director, for a period of six months from the date the Optionee ceased to be a Director or until the Expiration Date, if earlier. Any portion of this Stock Option that is not exercisable on the date the Optionee ceases to be a Director shall terminate immediately and be of no further force or effect.

4. **Incorporation of Plan.** Notwithstanding anything herein to the contrary, this Stock Option shall be subject to and governed by all the terms and conditions of the Plan, including the powers of the Administrator set forth in Section 2(b) of the Plan. Capitalized terms in this Agreement shall have the meaning specified in the Plan, unless a different meaning is specified herein.

5. **Transferability.** This Agreement is personal to the Optionee, is non-assignable and is not transferable in any manner, by operation of law or otherwise, other than by will or the laws of descent and distribution. This Stock Option is exercisable, during the Optionee's lifetime, only by the Optionee, and thereafter, only by the Optionee's legal representative or legatee.

6. **No Obligation to Continue as a Director.** Neither the Plan nor this Stock Option confers upon the Optionee any rights with respect to continuance as a Director.

7. **Integration.** This Agreement constitutes the entire agreement between the parties with respect to this Stock Option and supersedes all prior agreements and discussions between the parties concerning such subject matter.

8. **Data Privacy Consent.** In order to administer the Plan and this Agreement and to implement or structure future equity grants, the Company, its subsidiaries and affiliates and certain agents thereof (together, the "Relevant Companies") may process any and all personal or professional data, including but not limited to Social Security or other identification number, home address and telephone number, date of birth and other information that is necessary or desirable for the administration of the Plan and/or this Agreement (the "Relevant Information"). By entering into this Agreement, the Optionee (i) authorizes the Company to collect, process, register and transfer to the Relevant Companies all Relevant Information; (ii) waives any privacy rights the Optionee may have with respect to the Relevant Information; (iii) authorizes the Relevant Companies to store and transmit such information in electronic form; and (iv) authorizes the transfer of the Relevant Information to any jurisdiction in which the Relevant Companies consider appropriate. The Optionee shall have access to, and the right to change, the Relevant Information. Relevant Information will only be used in accordance with applicable law.

9. **Notices.** Notices hereunder shall be mailed or delivered to the Company at its principal place of business and shall be mailed or delivered to the Optionee at the address on file

with the Company or, in either case, at such other address as one party may subsequently furnish to the other party in writing.

SCHOLAR ROCK HOLDING CORPORATION

By: _____
Title:

The foregoing Agreement is hereby accepted and the terms and conditions thereof hereby agreed to by the undersigned. Electronic acceptance of this Agreement pursuant to the Company's instructions to the Optionee (including through an online acceptance process) is acceptable.

Dated: _____

Optionee's Signature _____

Optionee's name and address:

**NON-QUALIFIED STOCK OPTION AGREEMENT
FOR COMPANY EMPLOYEES
UNDER THE SCHOLAR ROCK HOLDING CORPORATION
2018 STOCK OPTION AND INCENTIVE PLAN**

Name of Optionee:

No. of Option Shares:

Option Exercise Price per Share:

\$

[FMV on Grant Date]

Grant Date:

Expiration Date:

[No more than 10 years]

Pursuant to the Scholar Rock Holding Corporation 2018 Stock Option and Incentive Plan as amended through the date hereof (the "Plan"), Scholar Rock Holding Corporation (the "Company") hereby grants to the Optionee named above an option (the "Stock Option") to purchase on or prior to the Expiration Date specified above all or part of the number of shares of Common Stock, par value \$0.001 per share (the "Stock") of the Company specified above at the Option Exercise Price per Share specified above subject to the terms and conditions set forth herein and in the Plan. This Stock Option is not intended to be an "incentive stock option" under Section 422 of the Internal Revenue Code of 1986, as amended.

1. Exercisability Schedule. No portion of this Stock Option may be exercised until such portion shall have become exercisable. Except as set forth below, and subject to the discretion of the Administrator (as defined in Section 2 of the Plan) to accelerate the exercisability schedule hereunder, this Stock Option shall be exercisable with respect to the following number of Option Shares on the dates indicated so long as Optionee continues to have a Service Relationship with the Company or a Subsidiary on such dates:

Incremental Number of Option Shares Exercisable	Exercisability Date
()%	
()%	
()%	
()%	
()%	

Once exercisable, this Stock Option shall continue to be exercisable at any time or times prior to the close of business on the Expiration Date, subject to the provisions hereof and of the Plan.

2. Manner of Exercise.

(a) The Optionee may exercise this Stock Option only in the following manner: from time to time on or prior to the Expiration Date of this Stock Option, the Optionee may give written notice to the Administrator of his or her election to purchase some or all of the Option Shares purchasable at the time of such notice. This notice shall specify the number of Option Shares to be purchased.

Payment of the purchase price for the Option Shares may be made by one or more of the following methods: (i) in cash, by certified or bank check or other instrument acceptable to the Administrator; (ii) through the delivery (or attestation to the ownership) of shares of Stock that have been purchased by the Optionee on the open market or that are beneficially owned by the Optionee and are not then subject to any restrictions under any Company plan and that otherwise satisfy any holding periods as may be required by the Administrator; (iii) by the Optionee delivering to the Company a properly executed exercise notice together with irrevocable instructions to a broker to promptly deliver to the Company cash or a check payable and acceptable to the Company to pay the option purchase price, provided that in the event the Optionee chooses to pay the option purchase price as so provided, the Optionee and the broker shall comply with such procedures and enter into such agreements of indemnity and other agreements as the Administrator shall prescribe as a condition of such payment procedure; (iv) by a "net exercise" arrangement pursuant to which the Company will reduce the number of shares of Stock issuable upon exercise by the largest whole number of shares with a Fair Market Value that does not exceed the aggregate exercise price; or (v) a combination of (i), (ii), (iii) and (iv) above. Payment instruments will be received subject to collection.

The transfer to the Optionee on the records of the Company or of the transfer agent of the Option Shares will be contingent upon (i) the Company's receipt from the Optionee of the full purchase price for the Option Shares, as set forth above, (ii) the fulfillment of any other requirements contained herein or in the Plan or in any other agreement or provision of laws, and (iii) the receipt by the Company of any agreement, statement or other evidence that the Company may require to satisfy itself that the issuance of Stock to be purchased pursuant to the exercise of Stock Options under the Plan and any subsequent resale of the shares of Stock will be in compliance with applicable laws and regulations. In the event the Optionee chooses to pay the purchase price by previously-owned shares of Stock through the attestation method, the number of shares of Stock transferred to the Optionee upon the exercise of the Stock Option shall be net of the Shares attested to.

(b) The shares of Stock purchased upon exercise of this Stock Option shall be transferred to the Optionee on the records of the Company or of the transfer agent upon compliance to the satisfaction of the Administrator with all requirements under applicable laws or regulations in connection with such transfer and with the requirements hereof and of the Plan. The determination of the Administrator as to such compliance shall be final and binding on the Optionee. The Optionee shall not be deemed to be the holder of, or to have any of the rights of a

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holder with respect to, any shares of Stock subject to this Stock Option unless and until this Stock Option shall have been exercised pursuant to the terms hereof, the Company or the transfer agent shall have transferred the shares to the Optionee, and the Optionee's name shall have been entered as the stockholder of record on the books of the Company. Thereupon, the Optionee shall have full voting, dividend and other ownership rights with respect to such shares of Stock.

(c) The minimum number of shares with respect to which this Stock Option may be exercised at any one time shall be 100 shares, unless the number of shares with respect to which this Stock Option is being exercised is the total number of shares subject to exercise under this Stock Option at the time.

(d) Notwithstanding any other provision hereof or of the Plan, no portion of this Stock Option shall be exercisable after the Expiration Date hereof.

3. Termination of Service Relationship. If the Optionee's Service Relationship by the Company or a Subsidiary (as defined in the Plan) is terminated, the period within which to exercise the Stock Option may be subject to earlier termination as set forth below.

(a) Termination Due to Death. If the Optionee's Service Relationship terminates by reason of the Optionee's death, any portion of this Stock Option outstanding on such date, to the extent exercisable on the date of death, may thereafter be exercised by the Optionee's legal representative or legatee for a period of 12 months from the date of death or until the Expiration Date, if earlier. Any portion of this Stock Option that is not exercisable on the date of death shall terminate immediately and be of no further force or effect.

(b) Termination Due to Disability. If the Optionee's Service Relationship terminates by reason of the Optionee's disability (as determined by the Administrator), any portion of this Stock Option outstanding on such date, to the extent exercisable on the date of such termination of Service Relationship, may thereafter be exercised by the Optionee for a period of 12 months from the date of disability or until the Expiration Date, if earlier. Any portion of this Stock Option that is not exercisable on the date of disability shall terminate immediately and be of no further force or effect.

(c) Termination for Cause. If the Optionee's Service Relationship terminates for Cause, any portion of this Stock Option outstanding on such date shall terminate immediately and be of no further force and effect. For purposes hereof, "Cause" shall mean, unless otherwise provided in an employment agreement (or similar services agreements) between the Company and the Optionee, a determination by the Administrator that the Optionee shall be dismissed as a result of (i) any material breach by the Optionee of any agreement between the Optionee and the Company; (ii) the conviction of, indictment for or plea of nolo contendere by the Optionee to a felony or a crime involving moral turpitude; or (iii) any material misconduct or willful and deliberate non-performance (other than by reason of disability) by the Optionee of the Optionee's duties to the Company.

(d) Other Termination. If the Optionee's Service Relationship terminates for any reason other than the Optionee's death, the Optionee's disability or Cause, and unless otherwise determined by the Administrator, any portion of this Stock Option outstanding on such

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date may be exercised, to the extent exercisable on the date of termination, for a period of three months from the date of termination or until the Expiration Date, if earlier. Any portion of this Stock Option that is not exercisable on the date of termination shall terminate immediately and be of no further force or effect.

The Administrator's determination of the reason for termination of the Optionee's Service Relationship shall be conclusive and binding on the Optionee and his or her representatives or legatees.

4. Incorporation of Plan. Notwithstanding anything herein to the contrary, this Stock Option shall be subject to and governed by all the terms and conditions of the Plan, including the powers of the Administrator set forth in Section 2(b) of the Plan. Capitalized terms in this Agreement shall have the meaning specified in the Plan, unless a different meaning is specified herein.

5. Transferability. This Agreement is personal to the Optionee, is non-assignable and is not transferable in any manner, by operation of law or otherwise, other than by will or the laws of descent and distribution. This Stock Option is exercisable, during the Optionee's lifetime, only by the Optionee, and thereafter, only by the Optionee's legal representative or legatee.

6. Tax Withholding. The Optionee shall, not later than the date as of which the exercise of this Stock Option becomes a taxable event for Federal income tax purposes, pay to the Company or make arrangements satisfactory to the Administrator for payment of any Federal, state, and local taxes required by law to be withheld on account of such taxable event. The Company shall have the authority to cause the required tax withholding obligation to be satisfied, in whole or in part, by withholding from shares of Stock to be issued to the Optionee a number of shares of Stock with an aggregate Fair Market Value that would satisfy the withholding amount due; provided, however, that the amount withheld does not exceed the maximum statutory tax rate or such lesser amount as is necessary to avoid adverse accounting treatment or as determined by the Administrator.

7. No Obligation to Continue Service Relationship. Neither the Company nor any Subsidiary is obligated by or as a result of the Plan or this Agreement to continue the Optionee's Service Relationship and neither the Plan nor this Agreement shall interfere in any way with the right of the Company or any Subsidiary to terminate the Service Relationship of the Optionee at any time.

8. Integration. This Agreement constitutes the entire agreement between the parties with respect to this Stock Option and supersedes all prior agreements and discussions between the parties concerning such subject matter.

9. Data Privacy Consent. In order to administer the Plan and this Agreement and to implement or structure future equity grants, the Company, its subsidiaries and affiliates and certain agents thereof (together, the "Relevant Companies") may process any and all personal or professional data, including but not limited to Social Security or other identification number, home address and telephone number, date of birth and other information that is necessary or

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desirable for the administration of the Plan and/or this Agreement (the "Relevant Information"). By entering into this Agreement, the Optionee (i) authorizes the Company to collect, process, register and transfer to the Relevant Companies all Relevant Information; (ii) waives any privacy rights the Optionee may have with respect to the Relevant Information; (iii) authorizes the Relevant Companies to store and transmit such information in electronic form; and (iv) authorizes the transfer of the Relevant Information to any jurisdiction in which the Relevant Companies consider appropriate. The Optionee shall have access to, and the right to change, the Relevant Information. Relevant Information will only be used in accordance with applicable law.

10. Notices. Notices hereunder shall be mailed or delivered to the Company at its principal place of business and shall be mailed or delivered to the Optionee at the address on file with the Company or, in either case, at such other address as one party may subsequently furnish to the other party in writing.

SCHOLAR ROCK HOLDING CORPORATION

By: _____
Title: _____

The foregoing Agreement is hereby accepted and the terms and conditions thereof hereby agreed to by the undersigned. Electronic acceptance of this Agreement pursuant to the Company's instructions to the Optionee (including through an online acceptance process) is acceptable.

Dated: _____
Optionee's Signature

Optionee's name and address:

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**NON-QUALIFIED STOCK OPTION AGREEMENT
FOR NON-EMPLOYEE CONSULTANTS
UNDER THE SCHOLAR ROCK HOLDING CORPORATION
2018 STOCK OPTION AND INCENTIVE PLAN**

Name of Optionee:

No. of Option Shares:

Option Exercise Price per Share: \$

[FMV on Grant Date]

Grant Date:

Expiration Date: [No more than 10 years]

Pursuant to the Scholar Rock Holding Corporation 2018 Stock Option and Incentive Plan as amended through the date hereof (the "Plan"), Scholar Rock Holding Corporation (the "Company") hereby grants to the Optionee named above, who is a Consultant of the Company, an option (the "Stock Option") to purchase on or prior to the Expiration Date specified above all or part of the number of shares of Common Stock, par value \$0.001 per share (the "Stock"), of the Company specified above at the Option Exercise Price per Share specified above subject to the terms and conditions set forth herein and in the Plan. This Stock Option is not intended to be an "incentive stock option" under Section 422 of the Internal Revenue Code of 1986, as amended.

1. Exercisability Schedule. No portion of this Stock Option may be exercised until such portion shall have become exercisable. Except as set forth below, and subject to the discretion of the Administrator (as defined in Section 2 of the Plan) to accelerate the exercisability schedule hereunder, this Stock Option shall be exercisable with respect to the following number of Option Shares on the dates indicated so long as the Optionee remains in service to the Company or a Subsidiary as a Consultant on such dates:

<u>Incremental Number of Option Shares Exercisable</u>	<u>Exercisability Date</u>
()%	
()%	
()%	
()%	
()%	

Once exercisable, this Stock Option shall continue to be exercisable at any time or times prior to the close of business on the Expiration Date, subject to the provisions hereof and of the Plan.

2. Manner of Exercise.

(a) The Optionee may exercise this Stock Option only in the following manner: from time to time on or prior to the Expiration Date of this Stock Option, the Optionee may give written notice to the Administrator of his or her election to purchase some or all of the Option Shares purchasable at the time of such notice. This notice shall specify the number of Option Shares to be purchased.

Payment of the purchase price for the Option Shares may be made by one or more of the following methods: (i) in cash, by certified or bank check or other instrument acceptable to the Administrator; (ii) through the delivery (or attestation to the ownership) of shares of Stock that have been purchased by the Optionee on the open market or that are beneficially owned by the Optionee and are not then subject to any restrictions under any Company plan and that otherwise satisfy any holding periods as may be required by the Administrator; (iii) by the Optionee delivering to the Company a properly executed exercise notice together with irrevocable instructions to a broker to promptly deliver to the Company cash or a check payable and acceptable to the Company to pay the option purchase price, provided that in the event the Optionee chooses to pay the option purchase price as so provided, the Optionee and the broker shall comply with such procedures and enter into such agreements of indemnity and other agreements as the Administrator shall prescribe as a condition of such payment procedure; (iv) by a "net exercise" arrangement pursuant to which the Company will reduce the number of shares of Stock issuable upon exercise by the largest whole number of shares with a Fair Market Value that does not exceed the aggregate exercise price; or (v) a combination of (i), (ii), (iii) and (iv) above. Payment instruments will be received subject to collection.

The transfer to the Optionee on the records of the Company or of the transfer agent of the Option Shares will be contingent upon (i) the Company's receipt from the Optionee of the full purchase price for the Option Shares, as set forth above, (ii) the fulfillment of any other requirements contained herein or in the Plan or in any other agreement or provision of laws, and (iii) the receipt by the Company of any agreement, statement or other evidence that the Company may require to satisfy itself that the issuance of Stock to be purchased pursuant to the exercise of Stock Options under the Plan and any subsequent resale of the shares of Stock will be in compliance with applicable laws and regulations. In the event the Optionee chooses to pay the purchase price by previously-owned shares of Stock through the attestation method, the number of shares of Stock transferred to the Optionee upon the exercise of the Stock Option shall be net of the Shares attested to.

(b) The shares of Stock purchased upon exercise of this Stock Option shall be transferred to the Optionee on the records of the Company or of the transfer agent upon compliance to the satisfaction of the Administrator with all requirements under applicable laws or regulations in connection with such transfer and with the requirements hereof and of the Plan. The determination of the Administrator as to such compliance shall be final and binding on the Optionee. The Optionee shall not be deemed to be the holder of, or to have any of the rights of a

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holder with respect to, any shares of Stock subject to this Stock Option unless and until this Stock Option shall have been exercised pursuant to the terms hereof, the Company or the transfer agent shall have transferred the shares to the Optionee, and the Optionee's name shall have been entered as the stockholder of record on the books of the Company. Thereupon, the Optionee shall have full voting, dividend and other ownership rights with respect to such shares of Stock.

(c) The minimum number of shares with respect to which this Stock Option may be exercised at any one time shall be 100 shares, unless the number of shares with respect to which this Stock Option is being exercised is the total number of shares subject to exercise under this Stock Option at the time.

(d) Notwithstanding any other provision hereof or of the Plan, no portion of this Stock Option shall be exercisable after the Expiration Date hereof.

3. **Termination of Service Relationship.** If the Optionee ceases to have a Service Relationship with the Company or a Subsidiary for any reason, any portion of this Stock Option outstanding on such date may be exercised, to the extent exercisable on the date the Optionee ceased to provide services, for a period of three months from the date the Optionee ceased to provide services or until the Expiration Date, if earlier. Any portion of this Stock Option that is not exercisable on the date the Optionee ceases to have a Service Relationship with the Company or a Subsidiary shall terminate immediately and be of no further force or effect.

4. **Incorporation of Plan.** Notwithstanding anything herein to the contrary, this Stock Option shall be subject to and governed by all the terms and conditions of the Plan, including the powers of the Administrator set forth in Section 2(b) of the Plan. Capitalized terms in this Agreement shall have the meaning specified in the Plan, unless a different meaning is specified herein.

5. **Transferability.** This Agreement is personal to the Optionee, is non-assignable and is not transferable in any manner, by operation of law or otherwise, other than by will or the laws of descent and distribution. This Stock Option is exercisable, during the Optionee's lifetime, only by the Optionee, and thereafter, only by the Optionee's legal representative or legatee.

6. **No Obligation to Continue Service Relationship.** Neither the Plan nor this Stock Option confers upon the Optionee any rights with respect to the continuance of Optionee's Service Relationship with the Company or a Subsidiary.

7. **Integration.** This Agreement constitutes the entire agreement between the parties with respect to this Stock Option and supersedes all prior agreements and discussions between the parties concerning such subject matter.

8. **Data Privacy Consent.** In order to administer the Plan and this Agreement and to implement or structure future equity grants, the Company, its subsidiaries and affiliates and certain agents thereof (together, the "Relevant Companies") may process any and all personal or professional data, including but not limited to Social Security or other identification number, home address and telephone number, date of birth and other information that is necessary or desirable for the administration of the Plan and/or this Agreement (the "Relevant Information").

By entering into this Agreement, the Optionee (i) authorizes the Company to collect, process, register and transfer to the Relevant Companies all Relevant Information; (ii) waives any privacy rights the Optionee may have with respect to the Relevant Information; (iii) authorizes the Relevant Companies to store and transmit such information in electronic form; and (iv) authorizes the transfer of the Relevant Information to any jurisdiction in which the Relevant Companies consider appropriate. The Optionee shall have access to, and the right to change, the Relevant Information. Relevant Information will only be used in accordance with applicable law.

9. **Notices.** Notices hereunder shall be mailed or delivered to the Company at its principal place of business and shall be mailed or delivered to the Optionee at the address on file with the Company or, in either case, at such other address as one party may subsequently furnish to the other party in writing.

SCHOLAR ROCK HOLDING CORPORATION

By: _____
Title:

The foregoing Agreement is hereby accepted and the terms and conditions thereof hereby agreed to by the undersigned. Electronic acceptance of this Agreement pursuant to the Company's instructions to the Optionee (including through an online acceptance process) is acceptable.

Dated: _____
Optionee's Signature

Optionee's name and address:

**RESTRICTED STOCK AWARD AGREEMENT
UNDER THE SCHOLAR ROCK HOLDING CORPORATION
2018 STOCK OPTION AND INCENTIVE PLAN**

Name of Grantee:

No. of Shares:

Grant Date:

Pursuant to the Scholar Rock Holding Corporation 2018 Stock Option and Incentive Plan as amended through the date hereof (the "Plan"), Scholar Rock Holding Corporation (the "Company") hereby grants a Restricted Stock Award (an "Award") to the Grantee named above. Upon acceptance of this Award, the Grantee shall receive the number of shares of Common Stock, par value \$0.001 per share (the "Stock") of the Company specified above, subject to the restrictions and conditions set forth herein and in the Plan. The Company acknowledges the receipt from the Grantee of consideration with respect to the par value of the Stock in the form of cash, past or future services rendered to the Company by the Grantee or such other form of consideration as is acceptable to the Administrator.

1. **Award.** The shares of Restricted Stock awarded hereunder shall be issued and held by the Company's transfer agent in book entry form, and the Grantee's name shall be entered as the stockholder of record on the books of the Company. Thereupon, the Grantee shall have all the rights of a stockholder with respect to such shares, including voting and dividend rights, subject, however, to the restrictions and conditions specified in Paragraph 2 below. The Grantee shall (i) sign and deliver to the Company a copy of this Award Agreement and (ii) deliver to the Company a stock power endorsed in blank.

2. **Restrictions and Conditions.**

(a) Any book entries for the shares of Restricted Stock granted herein shall bear an appropriate legend, as determined by the Administrator in its sole discretion, to the effect that such shares are subject to restrictions as set forth herein and in the Plan.

(b) Shares of Restricted Stock granted herein may not be sold, assigned, transferred, pledged or otherwise encumbered or disposed of by the Grantee prior to vesting.

(c) If the Grantee's Service Relationship with the Company and its Subsidiaries is voluntarily or involuntarily terminated for any reason (including death) prior to vesting of shares of Restricted Stock granted herein, all shares of Restricted Stock shall immediately and automatically be forfeited and returned to the Company.

3. **Vesting of Restricted Stock.** The restrictions and conditions in Paragraph 2 of this Agreement shall lapse on the Vesting Date or Dates specified in the following schedule so long as the Grantee continues to have a Service Relationship with the Company or a Subsidiary on such Dates. If a series of Vesting Dates is specified, then the restrictions and conditions in Paragraph 2 shall lapse only with respect to the number of shares of Restricted Stock specified as vested on such date.

Incremental Number of Shares Vested	Vesting Date
()%	
()%	
()%	
()%	

Subsequent to such Vesting Date or Dates, the shares of Stock on which all restrictions and conditions have lapsed shall no longer be deemed Restricted Stock. The Administrator may at any time accelerate the vesting schedule specified in this Paragraph 3.

4. **Dividends.** Dividends on shares of Restricted Stock shall be paid currently to the Grantee.

5. **Incorporation of Plan.** Notwithstanding anything herein to the contrary, this Award shall be subject to and governed by all the terms and conditions of the Plan, including the powers of the Administrator set forth in Section 2(b) of the Plan. Capitalized terms in this Agreement shall have the meaning specified in the Plan, unless a different meaning is specified herein.

6. **Transferability.** This Agreement is personal to the Grantee, is non-assignable and is not transferable in any manner, by operation of law or otherwise, other than by will or the laws of descent and distribution.

7. **Tax Withholding.** The Grantee shall, not later than the date as of which the receipt of this Award becomes a taxable event for Federal income tax purposes, pay to the Company or make arrangements satisfactory to the Administrator for payment of any Federal, state, and local taxes required by law to be withheld on account of such taxable event. Except in the case where an election is made pursuant to Paragraph 8 below, the Company shall have the authority to cause the required tax withholding obligation to be satisfied, in whole or in part, by withholding from shares of Stock to be issued or released by the transfer agent a number of shares of Stock with an aggregate Fair Market Value that would satisfy the withholding amount due; provided, however, that the amount withheld does not exceed the maximum statutory tax rate or such lesser amount as is necessary to avoid adverse accounting treatment or as determined by the Administrator.

8. **Election Under Section 83(b).** The Grantee and the Company hereby agree that the Grantee may, within 30 days following the Grant Date of this Award, file with the Internal Revenue Service and the Company an election under Section 83(b) of the Internal Revenue Code. In the event the Grantee makes such an election, he or she agrees to provide a copy of the election to the Company. The Grantee acknowledges that he or she is responsible for obtaining the advice of his or her tax advisors with regard to the Section 83(b) election and that he or she is relying solely on such advisors and not on any statements or representations of the Company or any of its agents with regard to such election.

9. **No Obligation to Continue Service Relationship.** Neither the Company nor any Subsidiary is obligated by or as a result of the Plan or this Agreement to continue the Grantee in the Service Relationship and neither the Plan nor this Agreement shall interfere in any way with the right of the Company or any Subsidiary to terminate the Service Relationship of the Grantee at any time.

10. **Integration.** This Agreement constitutes the entire agreement between the parties with respect to this Award and supersedes all prior agreements and discussions between the parties concerning such subject matter.

11. **Data Privacy Consent.** In order to administer the Plan and this Agreement and to implement or structure future equity grants, the Company, its subsidiaries and affiliates and certain agents thereof (together, the "Relevant Companies") may process any and all personal or professional data, including but not limited to Social Security or other identification number, home address and telephone number, date of birth and other information that is necessary or desirable for the administration of the Plan and/or this Agreement (the "Relevant Information"). By entering into this Agreement, the Grantee (i) authorizes the Company to collect, process, register and transfer to the Relevant Companies all Relevant Information; (ii) waives any privacy rights the Grantee may have with respect to the Relevant Information; (iii) authorizes the Relevant Companies to store and transmit such information in electronic form; and (iv) authorizes the transfer of the Relevant Information to any jurisdiction in which the Relevant Companies consider appropriate. The Grantee shall have access to, and the right to change, the Relevant Information. Relevant Information will only be used in accordance with applicable law.

12. **Notices.** Notices hereunder shall be mailed or delivered to the Company at its principal place of business and shall be mailed or delivered to the Grantee at the address on file with the Company or, in either case, at such other address as one party may subsequently furnish to the other party in writing.

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SCHOLAR ROCK HOLDING CORPORATION

By: _____

Name: _____

Title: _____

The foregoing Agreement is hereby accepted and the terms and conditions thereof hereby agreed to by the undersigned. Electronic acceptance of this Agreement pursuant to the Company's instructions to the Grantee (including through an online acceptance process) is acceptable.

Dated: _____

Grantee's Signature

Grantee's name and address:

**RESTRICTED STOCK UNIT AWARD AGREEMENT
FOR NON-EMPLOYEE DIRECTORS
UNDER THE SCHOLAR ROCK HOLDING CORPORATION
2018 STOCK OPTION AND INCENTIVE PLAN**

Name of Grantee: _____

No. of Restricted Stock Units: _____

Grant Date: _____

Pursuant to the Scholar Rock Holding Corporation 2018 Stock Option and Incentive Plan as amended through the date hereof (the "Plan"), Scholar Rock Holding Corporation (the "Company") hereby grants an award of the number of Restricted Stock Units listed above (an "Award") to the Grantee named above. Each Restricted Stock Unit shall relate to one share of Common Stock, par value \$0.001 per share (the "Stock") of the Company.

1. **Restrictions on Transfer of Award.** This Award may not be sold, transferred, pledged, assigned or otherwise encumbered or disposed of by the Grantee, and any shares of Stock issuable with respect to the Award may not be sold, transferred, pledged, assigned or otherwise encumbered or disposed of until (i) the Restricted Stock Units have vested as provided in Paragraph 2 of this Agreement and (ii) shares of Stock have been issued to the Grantee in accordance with the terms of the Plan and this Agreement.

2. **Vesting of Restricted Stock Units.** The restrictions and conditions of Paragraph 1 of this Agreement shall lapse on the Vesting Date or Dates specified in the following schedule so long as the Grantee remains in service as a member of the Board on such Dates. If a series of Vesting Dates is specified, then the restrictions and conditions in Paragraph 1 shall lapse only with respect to the number of Restricted Stock Units specified as vested on such date.

Incremental Number of Restricted Stock Units Vested	Vesting Date
()%	
()%	
()%	
()%	

The Administrator may at any time accelerate the vesting schedule specified in this Paragraph 2.

3. **Termination of Service.** If the Grantee's service with the Company and its Subsidiaries terminates for any reason (including death or disability) prior to the satisfaction of the vesting conditions set forth in Paragraph 2 above, any Restricted Stock Units that have not vested as of such date shall automatically and without notice terminate and be forfeited, and

neither the Grantee nor any of his or her successors, heirs, assigns, or personal representatives will thereafter have any further rights or interests in such unvested Restricted Stock Units.

4. **Issuance of Shares of Stock.** As soon as practicable following each Vesting Date (but in no event later than two and one-half months after the end of the year in which the Vesting Date occurs), the Company shall issue to the Grantee the number of shares of Stock equal to the aggregate number of Restricted Stock Units that have vested pursuant to Paragraph 2 of this Agreement on such date and the Grantee shall thereafter have all the rights of a stockholder of the Company with respect to such shares.

5. **Incorporation of Plan.** Notwithstanding anything herein to the contrary, this Agreement shall be subject to and governed by all the terms and conditions of the Plan, including the powers of the Administrator set forth in Section 2(b) of the Plan. Capitalized terms in this Agreement shall have the meaning specified in the Plan, unless a different meaning is specified herein.

6. **Section 409A of the Code.** This Agreement shall be interpreted in such a manner that all provisions relating to the settlement of the Award are exempt from the requirements of Section 409A of the Code as "short-term deferrals" as described in Section 409A of the Code.

7. **No Obligation to Continue as a Director.** Neither the Plan nor this Award confers upon the Grantee any rights with respect to continuance as a Director.

8. **Integration.** This Agreement constitutes the entire agreement between the parties with respect to this Award and supersedes all prior agreements and discussions between the parties concerning such subject matter.

9. **Data Privacy Consent.** In order to administer the Plan and this Agreement and to implement or structure future equity grants, the Company, its subsidiaries and affiliates and certain agents thereof (together, the "Relevant Companies") may process any and all personal or professional data, including but not limited to Social Security or other identification number, home address and telephone number, date of birth and other information that is necessary or desirable for the administration of the Plan and/or this Agreement (the "Relevant Information"). By entering into this Agreement, the Grantee (i) authorizes the Company to collect, process, register and transfer to the Relevant Companies all Relevant Information; (ii) waives any privacy rights the Grantee may have with respect to the Relevant Information; (iii) authorizes the Relevant Companies to store and transmit such information in electronic form; and (iv) authorizes the transfer of the Relevant Information to any jurisdiction in which the Relevant Companies consider appropriate. The Grantee shall have access to, and the right to change, the Relevant Information. Relevant Information will only be used in accordance with applicable law.

10. **Notices.** Notices hereunder shall be mailed or delivered to the Company at its principal place of business and shall be mailed or delivered to the Grantee at the address on file with the Company or, in either case, at such other address as one party may subsequently furnish to the other party in writing.

2

By: _____
Name: _____
Title: _____

The foregoing Agreement is hereby accepted and the terms and conditions thereof hereby agreed to by the undersigned. Electronic acceptance of this Agreement pursuant to the Company's instructions to the Grantee (including through an online acceptance process) is acceptable.

Dated: _____

Grantee's Signature

Grantee's name and address:

**RESTRICTED STOCK UNIT AWARD AGREEMENT
FOR COMPANY EMPLOYEES
UNDER THE SCHOLAR ROCK HOLDING CORPORATION
2018 STOCK OPTION AND INCENTIVE PLAN**

Name of Grantee:

No. of Restricted Stock Units:

Grant Date:

Pursuant to the Scholar Rock Holding Corporation 2018 Stock Option and Incentive Plan as amended through the date hereof (the "Plan"), Scholar Rock Holding Corporation (the "Company") hereby grants an award of the number of Restricted Stock Units listed above (an "Award") to the Grantee named above. Each Restricted Stock Unit shall relate to one share of Common Stock, par value \$0.001 per share (the "Stock") of the Company.

1. **Restrictions on Transfer of Award.** This Award may not be sold, transferred, pledged, assigned or otherwise encumbered or disposed of by the Grantee, and any shares of Stock issuable with respect to the Award may not be sold, transferred, pledged, assigned or otherwise encumbered or disposed of until (i) the Restricted Stock Units have vested as provided in Paragraph 2 of this Agreement and (ii) shares of Stock have been issued to the Grantee in accordance with the terms of the Plan and this Agreement.

2. **Vesting of Restricted Stock Units.** The restrictions and conditions of Paragraph 1 of this Agreement shall lapse on the Vesting Date or Dates specified in the following schedule so long as the Grantee continues to have a Service Relationship with the Company or a Subsidiary on such dates. If a series of Vesting Dates is specified, then the restrictions and conditions in Paragraph 1 shall lapse only with respect to the number of Restricted Stock Units specified as vested on such date.

Incremental Number of Restricted Stock Units Vested	Vesting Date
()%	
()%	
()%	
()%	

The Administrator may at any time accelerate the vesting schedule specified in this Paragraph 2.

3. **Termination of Service Relationship.** If the Grantee's Service Relationship with the Company and its Subsidiaries terminates for any reason (including death or disability) prior to the satisfaction of the vesting conditions set forth in Paragraph 2 above, any Restricted Stock Units that have not vested as of such date shall automatically and without notice terminate and be

forfeited, and neither the Grantee nor any of his or her successors, heirs, assigns, or personal representatives will thereafter have any further rights or interests in such unvested Restricted Stock Units.

4. **Issuance of Shares of Stock.** As soon as practicable following each Vesting Date (but in no event later than two and one-half months after the end of the year in which the Vesting Date occurs), the Company shall issue to the Grantee the number of shares of Stock equal to the aggregate number of Restricted Stock Units that have vested pursuant to Paragraph 2 of this Agreement on such date and the Grantee shall thereafter have all the rights of a stockholder of the Company with respect to such shares.

5. **Incorporation of Plan.** Notwithstanding anything herein to the contrary, this Agreement shall be subject to and governed by all the terms and conditions of the Plan, including the powers of the Administrator set forth in Section 2(b) of the Plan. Capitalized terms in this Agreement shall have the meaning specified in the Plan, unless a different meaning is specified herein.

6. **Tax Withholding.** The Grantee shall, not later than the date as of which the receipt of this Award becomes a taxable event for Federal income tax purposes, pay to the Company or make arrangements satisfactory to the Administrator for payment of any Federal, state, and local taxes required by law to be withheld on account of such taxable event. The Company shall have the authority to cause the required tax withholding obligation to be satisfied, in whole or in part, by withholding from shares of Stock to be issued to the Grantee a number of shares of Stock with an aggregate Fair Market Value that would satisfy the withholding amount due; provided, however, that the amount withheld does not exceed the maximum statutory tax rate or such lesser amount as is necessary to avoid adverse accounting treatment or as determined by the Administrator.

7. **Section 409A of the Code.** This Agreement shall be interpreted in such a manner that all provisions relating to the settlement of the Award are exempt from the requirements of Section 409A of the Code as "short-term deferrals" as described in Section 409A of the Code.

8. **No Obligation to Continue Service Relationship.** Neither the Company nor any Subsidiary is obligated by or as a result of the Plan or this Agreement to continue the Grantee in the Service Relationship and neither the Plan nor this Agreement shall interfere in any way with the right of the Company or any Subsidiary to terminate the Service Relationship of the Grantee at any time.

9. **Integration.** This Agreement constitutes the entire agreement between the parties with respect to this Award and supersedes all prior agreements and discussions between the parties concerning such subject matter.

10. **Data Privacy Consent.** In order to administer the Plan and this Agreement and to implement or structure future equity grants, the Company, its subsidiaries and affiliates and certain agents thereof (together, the "Relevant Companies") may process any and all personal or professional data, including but not limited to Social Security or other identification number, home address and telephone number, date of birth and other information that is necessary or desirable for the administration of the Plan and/or this Agreement (the "Relevant Information").

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By entering into this Agreement, the Grantee (i) authorizes the Company to collect, process, register and transfer to the Relevant Companies all Relevant Information; (ii) waives any privacy rights the Grantee may have with respect to the Relevant Information; (iii) authorizes the Relevant Companies to store and transmit such information in electronic form; and (iv) authorizes the transfer of the Relevant Information to any jurisdiction in which the Relevant Companies consider appropriate. The Grantee shall have access to, and the right to change, the Relevant Information. Relevant Information will only be used in accordance with applicable law.

11. **Notices.** Notices hereunder shall be mailed or delivered to the Company at its principal place of business and shall be mailed or delivered to the Grantee at the address on file with the Company or, in either case, at such other address as one party may subsequently furnish to the other party in writing.

3

SCHOLAR ROCK HOLDING CORPORATION

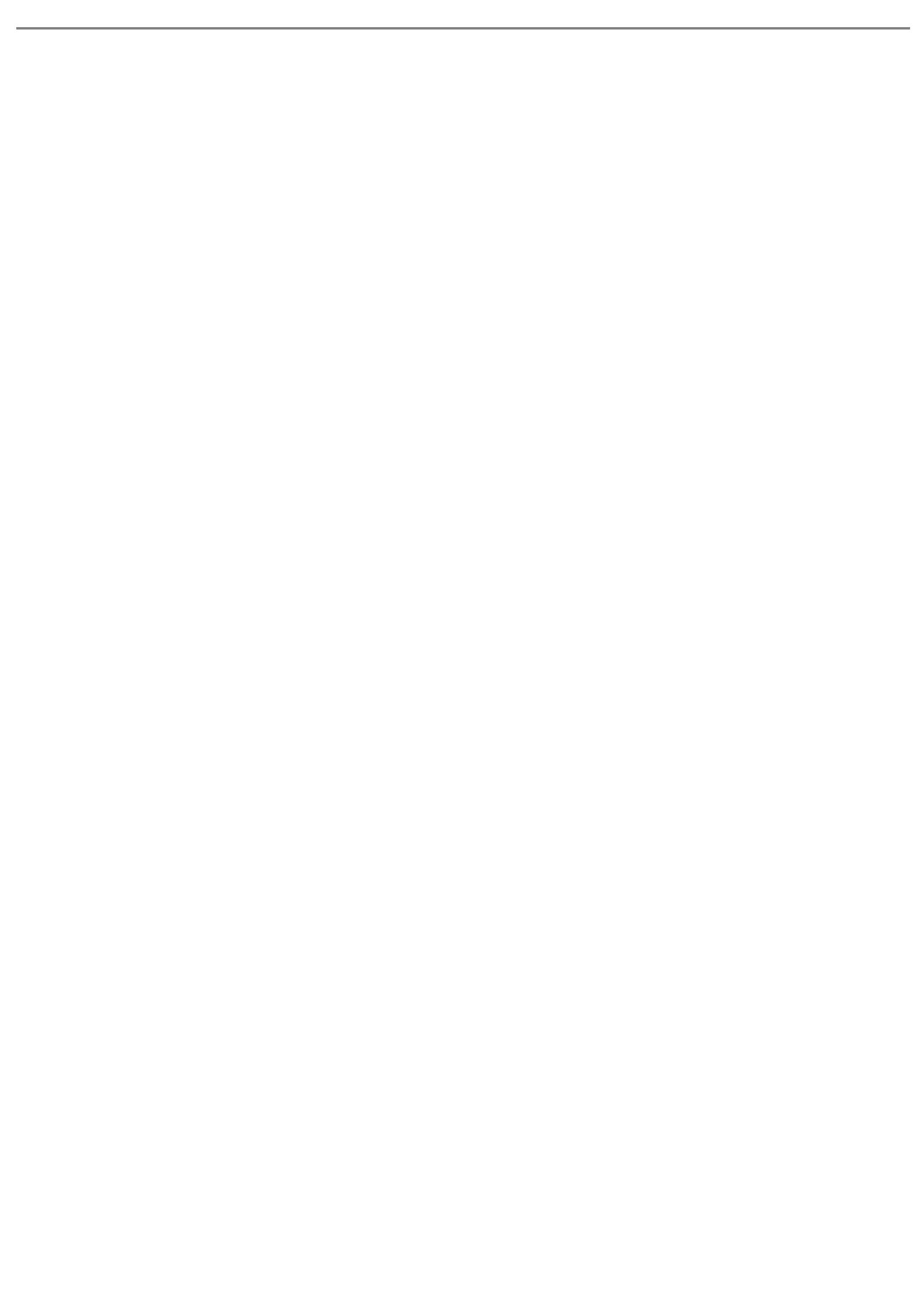
By: _____
Name: _____
Title: _____

The foregoing Agreement is hereby accepted and the terms and conditions thereof hereby agreed to by the undersigned. Electronic acceptance of this Agreement pursuant to the Company's instructions to the Grantee (including through an online acceptance process) is acceptable.

Dated: _____

Grantee's Signature

Grantee's name and address:



SCHOLAR ROCK HOLDING CORPORATION

2018 EMPLOYEE STOCK PURCHASE PLAN

The purpose of the Scholar Rock Holding Corporation 2018 Employee Stock Purchase Plan (the "Plan") is to provide eligible employees of Scholar Rock Holding Corporation (the "Company") and each Designated Subsidiary (as defined in Section 11) with opportunities to purchase shares of the Company's common stock, par value \$0.001 per share (the "Common Stock"). 235,743 shares of Common Stock have been approved and reserved for this purpose, plus on January 1, 2019, and each January 1 thereafter through January 1, 2028, the number of shares of Common Stock reserved and available for issuance under the Plan shall be cumulatively increased by the least of (i) 353,614 shares of Common Stock, (ii) 1% of the number of shares of Common Stock issued and outstanding on the immediately preceding December 31st, or (iii) such lesser number of shares of Common Stock as determined by the Administrator. The Plan is intended to constitute an "employee stock purchase plan" within the meaning of Section 423(b) of the Internal Revenue Code of 1986, as amended (the "Code"), and shall be interpreted in accordance with that intent.

1. Administration. The Plan will be administered by the person or persons (the "Administrator") appointed by the Company's Board of Directors (the "Board") for such purpose. The Administrator has authority at any time to: (i) adopt, alter and repeal such rules, guidelines and practices for the administration of the Plan and for its own acts and proceedings as it shall deem advisable; (ii) interpret the terms and provisions of the Plan; (iii) make all determinations it deems advisable for the administration of the Plan; (iv) decide all disputes arising in connection with the Plan; and (v) otherwise supervise the administration of the Plan. All interpretations and decisions of the Administrator shall be binding on all persons, including

the Company and the Participants. No member of the Board or individual exercising administrative authority with respect to the Plan shall be liable for any action or determination made in good faith with respect to the Plan or any option granted hereunder.

2. Offerings. The Company will make one or more offerings to eligible employees to purchase Common Stock under the Plan ("Offerings"). Unless otherwise determined by the Administrator, an Offering will begin on the first business day occurring on or after each December 1 and June 1 and will end on the last business day occurring on or before the following May 31 and November 30, respectively. The Administrator may, in its discretion, designate a different period for any Offering, provided that no Offering shall exceed one year in duration or overlap any other Offering.

3. Eligibility. All individuals classified as employees on the payroll records of the Company and each Designated Subsidiary are eligible to participate in any one or more of the Offerings under the Plan, provided that as of the first day of the applicable Offering (the "Offering Date") they are customarily employed by the Company or a Designated Subsidiary for more than twenty (20) hours a week and have completed at least three (3) months of employment. Notwithstanding any other provision herein, individuals who are not contemporaneously classified as employees of the Company or a Designated Subsidiary for purposes of the Company's or applicable Designated Subsidiary's payroll system are not considered to be eligible employees of the Company or any Designated Subsidiary and shall not be eligible to participate in the Plan. In the event any such individuals are reclassified as employees of the Company or a Designated Subsidiary for any purpose, including, without limitation, common law or statutory employees, by any action of any third party, including, without limitation, any government agency, or as a result of any private lawsuit, action or

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administrative proceeding, such individuals shall, notwithstanding such reclassification, remain ineligible for participation. Notwithstanding the foregoing, the exclusive means for individuals who are not contemporaneously classified as employees of the Company or a Designated Subsidiary on the Company's or Designated Subsidiary's payroll system to become eligible to participate in the Plan is through an amendment to the Plan, duly executed by the Company, which specifically renders such individuals eligible to participate herein.

4. Participation.

(a) Participants. An eligible employee who is not a Participant in any prior Offering may participate in a subsequent Offering by submitting an enrollment form to his or her appropriate payroll location at least fifteen (15) business days before the Offering Date (or by such other deadline as shall be established by the Administrator for the Offering).

(b) Enrollment. The enrollment form will (a) state a whole percentage to be deducted from an eligible employee's Compensation (as defined in Section 11) per pay period, (b) authorize the purchase of Common Stock in each Offering in accordance with the terms of the Plan and (c) specify the exact name or names in which shares of Common Stock purchased for such individual are to be issued pursuant to Section 10. An employee who does not enroll in accordance with these procedures will be deemed to have waived the right to participate. Unless a Participant files a new enrollment form or withdraws from the Plan, such Participant's deductions and purchases will continue at the same percentage of Compensation for future Offerings, provided he or she remains eligible.

(c) Notwithstanding the foregoing, participation in the Plan will neither be permitted nor be denied contrary to the requirements of the Code.

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5. Employee Contributions. Each eligible employee may authorize payroll deductions at a minimum of one percent (1%) up to a maximum of fifteen percent (15%) of such employee's Compensation for each pay period. The Company will maintain book accounts showing the amount of payroll deductions made by each Participant for each Offering. No interest will accrue or be paid on payroll deductions.

6. Deduction Changes. Except as may be determined by the Administrator in advance of an Offering, a Participant may not increase or decrease his or her payroll deduction during any Offering, but may increase or decrease his or her payroll deduction with respect to the next Offering (subject to the limitations of Section 5) by filing a new enrollment form at least fifteen (15) business days before the next Offering Date (or by such other deadline as shall be established by the Administrator for the Offering). The Administrator may, in advance of any Offering, establish rules permitting a Participant to increase, decrease or terminate his or her payroll deduction during an Offering.

7. Withdrawal. A Participant may withdraw from participation in the Plan by delivering a written notice of withdrawal to his or her appropriate payroll location. The Participant's withdrawal will be effective as of the next business day. Following a Participant's withdrawal, the Company will promptly refund such individual's entire account balance under the Plan to him or her (after payment for any Common Stock purchased before the effective date of withdrawal). Partial withdrawals are not permitted. Such an employee may not begin participation again during the remainder of the Offering, but may enroll in a subsequent Offering in accordance with Section 4.

8. Grant of Options. On each Offering Date, the Company will grant to each eligible employee who is then a Participant in the Plan an option ("Option") to purchase on the last day

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of such Offering (the "Exercise Date"), at the Option Price hereinafter provided for, the lowest of (a) a number of shares of Common Stock determined by dividing such Participant's accumulated payroll deductions on such Exercise Date by the lower of (i) eighty-five percent (85%) of the Fair Market Value of the Common Stock on the Offering Date, or (ii) eighty-five percent (85%) of the Fair Market Value of the Common Stock on the Exercise Date, (b) a number of shares determined by dividing \$25,000 by the Fair Market of the Common Stock on the Offering Date of such Offering, or (c) such other lesser maximum number of shares as shall have been established by the Administrator in advance of the Offering; provided, however, that such Option shall be subject to the limitations set forth below. Each Participant's Option shall be exercisable only to the extent of such Participant's accumulated payroll deductions on the Exercise Date. The purchase price for each share purchased under each Option (the "Option Price") will be eighty-five percent (85%) of the Fair Market Value of the Common Stock on the Offering Date or the Exercise Date, whichever is less.

Notwithstanding the foregoing, no Participant may be granted an option hereunder if such Participant, immediately after the option was granted, would be treated as owning stock possessing five percent (5%) or more of the total combined voting power or value of all classes of stock of the Company or any Parent or Subsidiary (as defined in Section 11). For purposes of the preceding sentence, the attribution rules of Section 424(d) of the Code shall apply in determining the stock ownership of a Participant, and all stock which the Participant has a contractual right to purchase shall be treated as stock owned by the Participant. In addition, no Participant may be granted an Option which permits his or her rights to purchase stock under the Plan, and any other employee stock purchase plan of the Company and its Parents and Subsidiaries, to accrue at a rate which exceeds \$25,000 of the fair market value of such stock

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(determined on the option grant date or dates) for each calendar year in which the Option is outstanding at any time. The purpose of the limitation in the preceding sentence is to comply with Section 423(b)(8) of the Code and shall be applied taking Options into account in the order in which they were granted.

9. Exercise of Option and Purchase of Shares. Each employee who continues to be a Participant in the Plan on the Exercise Date shall be deemed to have exercised his or her Option on such date and shall acquire from the Company such number of whole shares of Common Stock reserved for the purpose of the Plan as his or her accumulated payroll deductions on such date will purchase at the Option Price, subject to any other limitations contained in the Plan. Any amount remaining in a Participant's account at the end of an Offering solely by reason of the inability to purchase a fractional share will be carried forward to the next Offering; any other balance remaining in a Participant's account at the end of an Offering will be refunded to the Participant promptly.

10. Issuance of Certificates. Certificates representing shares of Common Stock purchased under the Plan may be issued only in the name of the employee, in the name of the employee and another person of legal age as joint tenants with rights of survivorship, or in the name of a broker authorized by the employee to be his, her or their, nominee for such purpose.

11. Definitions.

The term "Compensation" means the amount of base pay, prior to salary reduction pursuant to Sections 125, 132(f) or 401(k) of the Code, but excluding overtime, commissions, incentive or bonus awards, allowances and reimbursements for expenses such as relocation allowances or travel expenses, income or gains on the exercise of Company stock options, and similar items.

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The term "Designated Subsidiary" means any present or future Subsidiary (as defined below) that has been designated by the Board to participate in the Plan. The Board may so designate any Subsidiary, or revoke any such designation, at any time and from time to time, either before or after the Plan is approved by the stockholders.

The term "Fair Market Value of the Common Stock" on any given date means the fair market value of the Common Stock determined in good faith by the Administrator; provided, however, that if the Common Stock is admitted to quotation on the National Association of Securities Dealers Automated Quotation System ("Nasdaq"), Nasdaq Global Market or another national securities exchange, the determination shall be made by reference to the closing price on such date. If there is no closing price for such date, the determination shall be made by reference to the last date preceding such date for which there is a closing price.

The term "Initial Public Offering" means the first underwritten, firm commitment public offering pursuant to an effective registration statement under the Securities Act of 1933, as amended, covering the offer and sale by the Company of its Common Stock.

The term "Parent" means a "parent corporation" with respect to the Company, as defined in Section 424(e) of the Code.

The term "Participant" means an individual who is eligible as determined in Section 3 and who has complied with the provisions of Section 4.

The term "Subsidiary" means a "subsidiary corporation" with respect to the Company, as defined in Section 424(f) of the Code.

12. Rights on Termination of Employment. If a Participant's employment terminates for any reason before the Exercise Date for any Offering, no payroll deduction will be taken from any pay due and owing to the Participant and the balance in the Participant's account will

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be paid to such Participant or, in the case of such Participant's death, to his or her designated beneficiary as if such Participant had withdrawn from the Plan under Section 7. An employee will be deemed to have terminated employment, for this purpose, if the corporation that employs him or her, having been a Designated Subsidiary, ceases to be a Subsidiary, or if the employee is transferred to any corporation other than the Company or a Designated Subsidiary. An employee will not be deemed to have terminated employment for this purpose, if the employee is on an approved leave of absence for military service or sickness or for any other purpose approved by the Company, if the employee's right to reemployment is guaranteed either by a statute or by contract or under the policy pursuant to which the leave of absence was granted or if the Administrator otherwise provides in writing.

13. Special Rules. Notwithstanding anything herein to the contrary, the Administrator may adopt special rules applicable to the employees of a particular Designated Subsidiary, whenever the Administrator determines that such rules are necessary or appropriate for the implementation of the Plan in a jurisdiction where such Designated Subsidiary has employees; provided that such rules are consistent with the requirements of Section 423(b) of the Code. Any special rules established pursuant to this Section 13 shall, to the extent possible, result in the employees subject to such rules having substantially the same rights as other Participants in the Plan.

14. Options Not Stockholders. Neither the granting of an Option to a Participant nor the deductions from his or her pay shall constitute such Participant a holder of the shares of Common Stock covered by an Option under the Plan until such shares have been purchased by and issued to him or her.

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15. Rights Not Transferable. Rights under the Plan are not transferable by a Participant other than by will or the laws of descent and distribution, and are exercisable during the Participant's lifetime only by the Participant.

16. Application of Funds. All funds received or held by the Company under the Plan may be combined with other corporate funds and may be used for any corporate purpose.

17. Adjustment in Case of Changes Affecting Common Stock. In the event of a subdivision of outstanding shares of Common Stock, the payment of a dividend in Common Stock or any other change affecting the Common Stock, the number of shares approved for the Plan and the share limitation set forth in Section 8 shall be equitably or proportionately adjusted to give proper effect to such event.

18. Amendment of the Plan. The Board may at any time and from time to time amend the Plan in any respect, except that without the approval within twelve (12) months of such Board action by the stockholders, no amendment shall be made increasing the number of shares approved for the Plan or making any other change that would require stockholder approval in order for the Plan, as amended, to qualify as an "employee stock purchase plan" under Section 423(b) of the Code.

19. Insufficient Shares. If the total number of shares of Common Stock that would otherwise be purchased on any Exercise Date plus the number of shares purchased under previous Offerings under the Plan exceeds the maximum number of shares issuable under the Plan, the shares then available shall be apportioned among Participants in proportion to the amount of payroll deductions accumulated on behalf of each Participant that would otherwise be used to purchase Common Stock on such Exercise Date.

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20. Termination of the Plan. The Plan may be terminated at any time by the Board. Upon termination of the Plan, all amounts in the accounts of Participants shall be promptly refunded.

21. Governmental Regulations. The Company's obligation to sell and deliver Common Stock under the Plan is subject to obtaining all governmental approvals required in connection with the authorization, issuance, or sale of such stock.

22. Governing Law. The Plan and all Options and actions taken thereunder shall be governed by, and construed in accordance with, the laws of the State of Delaware, applied without regard to conflict of law principles.

23. Issuance of Shares. Shares may be issued upon exercise of an Option from authorized but unissued Common Stock, from shares held in the treasury of the Company, or from any other proper source.

24. Tax Withholding. Participation in the Plan is subject to any minimum required tax withholding on income of the Participant in connection with the Plan. Each Participant agrees, by entering the Plan, that the Company and its Subsidiaries shall have the right to deduct any such taxes from any payment of any kind otherwise due to the Participant, including shares issuable under the Plan.

25. Notification Upon Sale of Shares. Each Participant agrees, by entering the Plan, to give the Company prompt notice of any disposition of shares purchased under the Plan where such disposition occurs within two (2) years after the date of grant of the Option pursuant to which such shares were purchased or within one year after the date such shares were purchased.

26. Effective Date and Approval of Shareholders. The Plan shall take effect on the date on which the Company's registration statement on Form S-1 becomes effective following

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approval by the holders of a majority of the votes cast at a meeting of stockholders at which a quorum is present or by written consent of the stockholders.

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SCHOLAR ROCK HOLDING CORPORATION
FORM OF DIRECTOR INDEMNIFICATION AGREEMENT

This Indemnification Agreement ("Agreement") is made as of _____ by and between Scholar Rock Holding Corporation, a Delaware corporation (the "Company"), and [Director] ("Indemnitee").

RECITALS

WHEREAS, the Company desires to attract and retain the services of highly qualified individuals, such as Indemnitee, to serve the Company;

WHEREAS, in order to induce Indemnitee to provide or continue to provide services to the Company, the Company wishes to provide for the indemnification of, and advancement of expenses to, Indemnitee to the maximum extent permitted by law;

WHEREAS, the Amended and Restated Certificate of Incorporation (as amended and in effect from time to time, the "Charter") and the Amended and Restated Bylaws (as amended and in effect from time to time, the "Bylaws") of the Company require indemnification of the officers and directors of the Company, and Indemnitee may also be entitled to indemnification pursuant to the General Corporation Law of the State of Delaware (the "DGCL");

WHEREAS, the Charter, the Bylaws and the DGCL expressly provide that the indemnification provisions set forth therein are not exclusive, and thereby contemplate that contracts may be entered into between the Company and members of the board of directors, officers and other persons with respect to indemnification;

WHEREAS, the Board of Directors of the Company (the "Board") has determined that the increased difficulty in attracting and retaining highly qualified persons such as Indemnitee is detrimental to the best interests of the Company's stockholders;

WHEREAS, it is reasonable and prudent for the Company contractually to obligate itself to indemnify, and to advance expenses on behalf of, such persons to the fullest extent permitted by applicable law, regardless of any amendment or revocation of the Charter or the Bylaws, so that they will serve or continue to serve the Company free from undue concern that they will not be so indemnified;

WHEREAS, this Agreement is a supplement to and in furtherance of the indemnification provided in the Charter, the Bylaws and any resolutions adopted pursuant thereto, and shall not be deemed a substitute therefor, nor to diminish or abrogate any rights of Indemnitee thereunder; and

[WHEREAS, Indemnitee has certain rights to indemnification and/or insurance provided by [-] ("[-]") which Indemnitee and [-] intend to be secondary to the primary obligation of the Company to indemnify Indemnitee as provided in this Agreement, with the Company's

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acknowledgment and agreement to the foregoing being a material condition to Indemnitee's willingness to serve or continue to serve on the Board.]

NOW, THEREFORE, in consideration of the premises and the covenants contained herein, the Company and Indemnitee do hereby covenant and agree as follows:

Section 1. Services to the Company. Indemnitee agrees to [continue to] serve as a director of the Company. Indemnitee may at any time and for any reason resign from such position (subject to any other contractual obligation or any obligation imposed by law), in which event the Company shall have no obligation under this Agreement to continue Indemnitee in such position. This Agreement shall not be deemed an employment contract between the Company (or any of its subsidiaries or any Enterprise) and Indemnitee.

Section 2. Definitions.

As used in this Agreement:

(a) "Change in Control" shall mean (i) the sale of all or substantially all of the assets of the Company on a consolidated basis to an unrelated person or entity, (ii) a merger, reorganization or consolidation pursuant to which the holders of the Company's outstanding voting power and outstanding stock immediately prior to such transaction do not own a majority of the outstanding voting power and outstanding stock or other equity interests of the resulting or successor entity (or its ultimate parent, if applicable) immediately upon completion of such transaction, (iii) the sale of all of the Stock of the Company to an unrelated person, entity or group thereof acting in concert, or (iv) any other transaction in which the owners of the Company's outstanding voting power immediately prior to such transaction do not own at least a majority of the outstanding voting power of the Company or any successor entity immediately upon completion of the transaction other than as a result of the acquisition of securities directly from the Company.

(b) "Corporate Status" describes the status of a person as a current or former director of the Company or current or former director, manager, partner, officer, employee, agent or trustee of any other Enterprise which such person is or was serving at the request of the Company.

(c) "Enforcement Expenses" shall include all reasonable attorneys' fees, court costs, transcript costs, fees of experts, travel expenses, duplicating costs, printing and binding costs, telephone charges, postage, delivery service fees, and all other out-of-pocket disbursements or expenses incurred in connection with an action to enforce indemnification or advancement rights, or an appeal from such action. Expenses, however, shall not include fees, salaries, wages or benefits owed to Indemnitee.

(d) "Enterprise" shall mean any corporation (other than the Company), partnership, joint venture, trust, employee benefit plan, limited liability company, or other legal entity of which Indemnitee is or was serving at the request of the Company as a director, manager, partner, officer, employee, agent or trustee, including as a deemed fiduciary thereto.

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(e) "Expenses" shall include all reasonable attorneys' fees, court costs, transcript costs, fees of experts, travel expenses, duplicating costs, printing and binding costs, telephone charges, postage, delivery service fees, and all other out-of-pocket disbursements or expenses of the types customarily incurred in connection with prosecuting, defending, preparing to prosecute or defend, investigating, being or preparing to be a witness in, or otherwise participating in, a Proceeding or an appeal resulting from a Proceeding. Expenses, however, shall not include amounts paid in settlement by Indemnitee, the amount of judgments or fines against Indemnitee or fees, salaries, wages or benefits owed to Indemnitee.

(f) "Independent Counsel" means a law firm, or a partner (or, if applicable, member or shareholder) of such a law firm, that is experienced in matters of Delaware corporation law and neither presently is, nor in the past five (5) years has been, retained to represent: (i) the Company, any subsidiary of the Company, any Enterprise or Indemnitee in any matter material to any such party; or (ii) any other party to the Proceeding giving rise to a claim for indemnification hereunder. Notwithstanding the foregoing, the term "Independent Counsel" shall not include any person who, under the applicable standards of professional conduct then prevailing, would have a conflict of interest in representing either the Company or Indemnitee in an action to determine Indemnitee's rights under this Agreement. The Company agrees to pay the reasonable fees and expenses of the Independent Counsel referred to above and to fully indemnify such counsel against any and all expenses, claims, liabilities and damages arising out of or relating to this Agreement or its engagement pursuant hereto.

(g) The term "Proceeding" shall include any threatened, pending or completed action, suit, arbitration, alternate dispute resolution mechanism, investigation, inquiry, administrative hearing or any other actual, threatened or completed proceeding, whether brought in the right of the Company or otherwise and whether of a civil, criminal, administrative, regulatory or investigative nature, and whether formal or informal, in which Indemnitee was, is or will be involved as a party or otherwise by reason of the fact that Indemnitee is or was a director of the Company or is or was serving at the request of the Company as a director, manager, partner, officer, employee, agent or trustee of any Enterprise or by reason of any action taken by Indemnitee or of any action taken on his or her part while acting as a director of the Company or while serving at the request of the Company as a director, manager, partner, officer, employee, agent or trustee of any Enterprise, in each case whether or not serving in such capacity at the time any liability or expense is incurred for which indemnification, reimbursement or advancement of expenses can be provided under this Agreement; provided, however, that the term "Proceeding" shall not include any action, suit or arbitration, or part thereof, initiated by Indemnitee to enforce Indemnitee's rights under this Agreement as provided for in Section 12(a) of this Agreement.

Section 3. Indemnity in Third-Party Proceedings. The Company shall indemnify Indemnitee to the extent set forth in this Section 3 if Indemnitee is, or is threatened to be made, a party to or a participant in any Proceeding, other than a Proceeding by or in the right of the Company to procure a judgment in its favor. Pursuant to this Section 3, Indemnitee shall be indemnified against all Expenses, judgments, fines, penalties, excise taxes, and amounts paid in settlement actually and reasonably incurred by Indemnitee or on his or her behalf in connection

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with such Proceeding or any claim, issue or matter therein, if Indemnitee acted in good faith and in a manner he or she reasonably believed to be in or not opposed to the best interests of the Company and, in the case of a criminal proceeding, had no reasonable cause to believe that his or her conduct was unlawful.

Section 4. Indemnity in Proceedings by or in the Right of the Company. The Company shall indemnify Indemnitee to the extent set forth in this Section 4 if Indemnitee is, or is threatened to be made, a party to or a participant in any Proceeding by or in the right of the Company to procure a judgment in its favor. Pursuant to this Section 4, Indemnitee shall be indemnified against all Expenses actually and reasonably incurred by him or her in connection with such Proceeding or any claim, issue or matter therein, if Indemnitee acted in good faith and in a manner he or she reasonably believed to be in or not opposed to the best interests of the Company. No indemnification for Expenses shall be made under this Section 4 in respect of any claim, issue or matter as to which Indemnitee shall have been finally adjudged by a court to be liable to the Company, unless and only to the extent that the Delaware Court of Chancery (the "Delaware Court") shall determine upon application that, despite the adjudication of liability but in view of all the circumstances of the case, Indemnitee is fairly and reasonably entitled to indemnification for such expenses as the Delaware Court shall deem proper.

Section 5. Indemnification for Expenses of a Party Who is Wholly or Partly Successful. Notwithstanding any other provisions of this Agreement and except as provided in Section 7, to the extent that Indemnitee is a party to or a participant in any Proceeding and is successful in such Proceeding or in defense of any claim, issue or matter therein, the Company shall indemnify Indemnitee against all Expenses actually and reasonably incurred by him or her in connection therewith. If Indemnitee is not wholly successful in such Proceeding but is successful as to one or more but less than all claims, issues or matters in such Proceeding, the Company shall indemnify Indemnitee against all Expenses actually and reasonably incurred by Indemnitee or on his or her behalf in connection with each successfully resolved claim, issue or matter. For purposes of this Section and without limitation, the termination of any claim, issue or matter in such a Proceeding by dismissal, with or without prejudice, motion for summary judgment, settlement (with or without court approval), or upon a plea of nolo contendere or its equivalent, shall be deemed to be a successful result as to such claim, issue or matter.

Section 6. Reimbursement for Expenses of a Witness or in Response to a Subpoena. Notwithstanding any other provision of this Agreement, to the extent that Indemnitee, by reason of his or her Corporate Status, (i) is a witness in any Proceeding to which Indemnitee is not a party and is not threatened to be made a party or (ii) receives a subpoena with respect to any Proceeding to which Indemnitee is not a party and is not threatened to be made a party, the Company shall reimburse Indemnitee for all Expenses actually and reasonably incurred by him or her or on his or her behalf in connection therewith.

Section 7. Exclusions. Notwithstanding any provision in this Agreement to the contrary, the Company shall not be obligated under this Agreement:

- (a) to indemnify for amounts otherwise indemnifiable hereunder (or for which advancement is provided hereunder) if and to the extent that Indemnitee has otherwise actually

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received such amounts under any insurance policy, contract, agreement or otherwise; provided that the foregoing shall not affect the rights of Indemnitee or the Secondary Indemnitors as set forth in Section 13(c); provided further, however, that payment made to Indemnitee pursuant to an insurance policy purchased and maintained by Indemnitee at his or her own expense of any amounts otherwise indemnifiable or obligated to be made pursuant to this Agreement shall not reduce the Company's obligations to Indemnitee pursuant to this Agreement.

(b) to indemnify for an accounting of profits made from the purchase and sale (or sale and purchase) by Indemnitee of securities of the Company within the meaning of Section 16(b) of the Securities Exchange Act of 1934, as amended, or similar provisions of state statutory law or common law, or from the purchase or sale by Indemnitee of such securities in violation of Section 306 of the Sarbanes Oxley Act of 2002 ("SOX");

(c) to indemnify with respect to any Proceeding, or part thereof, brought by Indemnitee against the Company, any legal entity which it controls, any director or officer thereof or any third party, unless (i) the Board has consented to the initiation of such Proceeding or part thereof and (ii) the Company provides the indemnification, in its sole discretion, pursuant to the powers vested in the Company under applicable law; provided, however, that this Section 7(c) shall not apply to (A) counterclaims or affirmative defenses asserted by Indemnitee in an action brought against Indemnitee or (B) any action brought by Indemnitee for indemnification or advancement from the Company under this Agreement or under any directors' and officers' liability insurance policies maintained by the Company in the suit for which indemnification or advancement is being sought as described in Section 12; or

(d) to provide any indemnification or advancement of expenses that is prohibited by applicable law (as such law exists at the time payment would otherwise be required pursuant to this Agreement).

Section 8. Advancement of Expenses. Subject to Section 9(b), the Company shall advance, to the extent not prohibited by law, the Expenses incurred by Indemnitee in connection with any Proceeding, and such advancement shall be made as incurred, and such advancement shall be made within thirty (30) days after the receipt by the Company of a statement or statements requesting such advances (including any invoices received by Indemnitee, which such invoices may be redacted as necessary to avoid the waiver of any privilege accorded by applicable law) from time to time, whether prior to or after final disposition of any Proceeding. Advances shall be unsecured and insurance free. Advances shall be made without regard to Indemnitee's (i) ability to repay the expenses, (ii) ultimate entitlement to indemnification under the other provisions of this Agreement, and (iii) entitlement to and availability of insurance coverage, including advancement, payment or reimbursement of defense costs, expenses of covered loss under the provisions of any applicable insurance policy (including, without limitation, whether such advancement, payment or reimbursement is withheld, conditioned or delayed by the insurer(s)). Without limiting the generality or effect of the foregoing, within thirty days after any request for advancement by Indemnitee pursuant to the first sentence of this Section 8, the Company shall, in accordance with such request (but without duplication), (a) pay such Expenses on behalf of Indemnitee, (b) advance to Indemnitee funds in an amount sufficient to pay such Expenses, or (c) reimburse Indemnitee for such Expenses. The Company shall not

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seek from a court, or agree to, a "bar order" which would have the effect of prohibiting or limiting the Indemnitee's rights to receive advancement of expenses under this Agreement. Indemnitee shall qualify for advances (including any Enforcement Expenses as set forth in Section 12(e)) upon the execution and delivery to the Company of this Agreement which shall constitute an undertaking providing that Indemnitee undertakes to the fullest extent required by law to repay the advance if and to the extent that it is ultimately determined by a court of competent jurisdiction in a final judgment, not subject to appeal, that Indemnitee is not entitled to be indemnified by the Company. No other form of undertaking shall be required other than the execution of this Agreement. The right to advances under this paragraph shall in all events continue until final disposition of any Proceeding, including any appeal therein. Nothing in this Section 8 shall limit Indemnitee's right to advancement pursuant to Section 12(e) of this Agreement.

Section 9. Procedure for Notification and Defense of Claim.

(a) To obtain indemnification under this Agreement, Indemnitee shall submit to the Company a written request therefor specifying the basis for the claim, the amounts for which Indemnitee is seeking payment under this Agreement, and all documentation (which is not privileged or otherwise protected from disclosure and which is reasonably available to Indemnitee) related thereto as reasonably requested by the Company.

(b) In the event that the Company shall be obligated hereunder to provide indemnification or make any advancement of Expenses with respect to any Proceeding, the Company shall be entitled to assume the defense of such Proceeding, or any claim, issue or matter therein, with counsel approved by Indemnitee (which approval shall not be unreasonably withheld or delayed) upon the delivery to Indemnitee of written notice of the Company's election to do so. After delivery of such notice, approval of such counsel by Indemnitee and the retention of such counsel by the Company, the Company will not be liable to Indemnitee under this Agreement for any fees or expenses of separate counsel subsequently employed by or on behalf of Indemnitee with respect to the same Proceeding; provided that (i) Indemnitee shall have the right to employ separate counsel in any such Proceeding at Indemnitee's expense and (ii) if (A) the employment of separate counsel by Indemnitee has been previously authorized by the Company, (B) Indemnitee shall have reasonably concluded that there may be a conflict of interest between the Company and Indemnitee in the conduct of such defense, or (C) the Company shall not continue to retain such counsel to defend such Proceeding, then the fees and expenses actually and reasonably incurred by Indemnitee with respect to his or her separate counsel shall be Expenses hereunder. Indemnitee agrees that any such separate counsel retained by Indemnitee will be a member of any approved list of panel counsel under the Company's applicable directors' and officers' insurance policy, should the applicable policy provide for a panel of approved counsel and should such approved panel list comprise law firms with well-established reputations in the type of litigation at issue.

(c) In the event that the Company does not assume the defense in a Proceeding pursuant to paragraph (b) above, then the Company will be entitled to participate in the Proceeding at its own expense.

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(d) The Company shall not be liable to indemnify Indemnitee under this Agreement for any amounts paid in settlement of any Proceeding effected without its prior written consent (which consent shall not be unreasonably withheld or delayed). The Company shall not, without the prior written consent of Indemnitee (which consent shall not be unreasonably withheld or delayed), enter into any settlement which (i) includes an admission of fault of Indemnitee, any non-monetary remedy imposed on Indemnitee or any monetary damages for which Indemnitee is not wholly and actually indemnified hereunder or (ii) with respect to any Proceeding with respect to which Indemnitee may be or is made a party or may be otherwise entitled to seek indemnification hereunder, does not include the full release of Indemnitee from all liability in respect of such Proceeding.

Section 10. Procedure Upon Application for Indemnification.

(a) Upon written request by Indemnitee for indemnification pursuant to Section 9(a), a determination, if such determination is required by applicable law, with respect to Indemnitee's entitlement to indemnification hereunder shall be made in the specific case by one of the following methods: (x) if a Change in Control shall have occurred, by Independent Counsel in a written opinion to the Board; or (y) if a Change in Control shall not have occurred: (i) by a majority vote of the disinterested directors, even though less than a quorum; (ii) by a committee of disinterested directors designated by a majority vote of the disinterested directors, even though less than a quorum; or (iii) if there are no disinterested directors or if the disinterested directors so direct, by Independent Counsel in a written opinion to the Board. For purposes hereof, disinterested directors are those members of the Board who are not parties to the action, suit or proceeding in respect of which indemnification is sought. In the case that such determination is made by Independent Counsel, a copy of Independent Counsel's written opinion shall be delivered to Indemnitee and, if it is so determined that Indemnitee is entitled to indemnification, payment to Indemnitee shall be made within thirty (30) days after such determination. Indemnitee shall cooperate with the Independent Counsel or the Company, as applicable, in making such determination with respect to Indemnitee's entitlement to indemnification, including providing to such counsel or the Company, upon reasonable advance request, any documentation or information which is not privileged or otherwise protected from disclosure and which is reasonably available to Indemnitee and reasonably necessary to such determination. Any out-of-pocket costs or expenses (including reasonable attorneys' fees and disbursements) actually and reasonably incurred by Indemnitee in so cooperating with the Independent Counsel or the Company shall be borne by the Company (irrespective of the determination as to Indemnitee's entitlement to indemnification) and the Company hereby indemnifies and agrees to hold Indemnitee harmless therefrom.

(b) If the determination of entitlement to indemnification is to be made by Independent Counsel pursuant to Section 10(a), the Independent Counsel shall be selected by the Board if a Change in Control shall not have occurred or, if a Change in Control shall have occurred, by Indemnitee. Indemnitee or the Company, as the case may be, may, within ten (10) days after written notice of such selection, deliver to the Company or Indemnitee, as the case may be, a written objection to such selection; provided, however, that such objection may be asserted only on the ground that the Independent Counsel so selected does not meet the requirements of "Independent Counsel" as defined in Section 2 of this Agreement, and the objection shall set forth with particularity the factual basis of such assertion. Absent a proper

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and timely objection, the person so selected shall act as Independent Counsel. If such written objection is so made and substantiated, the Independent Counsel so selected may not serve as Independent Counsel unless and until such objection is withdrawn or the Delaware Court has determined that such objection is without merit. If, within twenty (20) days after the later of (i) submission by Indemnitee of a written request for indemnification pursuant to Section 9(a), and (ii) the final disposition of the Proceeding, including any appeal therein, no Independent Counsel shall have been selected without objection, either Indemnitee or the Company may petition the Delaware Court for resolution of any objection which shall have been made by Indemnitee or the Company to the selection of Independent Counsel and/or for the appointment as Independent Counsel of a person selected by the court or by such other person as the court shall designate. The person with respect to whom all objections are so resolved or the person so appointed shall act as Independent Counsel under Section 10(a) hereof. Upon the due commencement of any judicial proceeding or arbitration pursuant to Section 12(a) of this Agreement, Independent Counsel shall be discharged and relieved of any further responsibility in such capacity (subject to the applicable standards of professional conduct then prevailing).

Section 11. Presumptions and Effect of Certain Proceedings.

(a) To the extent permitted by applicable law, in making a determination with respect to entitlement to indemnification hereunder, it shall be presumed that Indemnitee is entitled to indemnification under this Agreement if Indemnitee has submitted a request for indemnification in accordance with Section 9(a) of this Agreement, and the Company shall have the burden of proof to overcome that presumption in connection with the making of any determination contrary to that presumption. Neither (i) the failure of the Company or of Independent Counsel to have made a determination prior to the commencement of any action pursuant to this Agreement that indemnification is proper in the circumstances because Indemnitee has met the applicable standard of conduct, nor (ii) an actual determination by the Company or by Independent Counsel that Indemnitee has not met such applicable standard of conduct, shall be a defense to the action or create a presumption that Indemnitee has not met the applicable standard of conduct.

(b) The termination of any Proceeding or of any claim, issue or matter therein, by judgment, order, settlement or conviction, or upon a plea of guilty, nolo contendere or its equivalent, shall not (except as otherwise expressly provided in this Agreement) of itself adversely affect the right of Indemnitee to indemnification or create a presumption that Indemnitee did not act in good faith and in a manner which he or she reasonably believed to be in or not opposed to the best interests of the Company or, with respect to any criminal Proceeding, that Indemnitee had reasonable cause to believe that his or her conduct was unlawful.

(c) The knowledge and/or actions, or failure to act, of any director, manager, partner, officer, employee, agent or trustee of the Company, any subsidiary of the Company, or any Enterprise shall not be imputed to Indemnitee for purposes of determining the right to indemnification under this Agreement.

Section 12. Remedies of Indemnitee.

(a) Subject to Section 12(f), in the event that (i) a determination is made pursuant to Section 10 of this Agreement that Indemnitee is not entitled to indemnification under this Agreement, (ii) advancement of Expenses is not timely made pursuant to Section 8 of this Agreement, (iii) no determination of entitlement to indemnification shall have been made pursuant to Section 10(a) of this Agreement within sixty (60) days after receipt by the Company of the request for indemnification for which a determination is to be made other than by Independent Counsel, (iv) payment of indemnification or reimbursement of expenses is not made pursuant to Section 5 or 6 or the last sentence of Section 10(a) of this Agreement within thirty (30) days after receipt by the Company of a written request therefor (including any invoices received by Indemnitee, which such invoices may be redacted as necessary to avoid the waiver of any privilege accorded by applicable law) or (v) payment of indemnification pursuant to Section 3 or 4 of this Agreement is not made within thirty (30) days after a determination has been made that Indemnitee is entitled to indemnification, Indemnitee shall be entitled to an adjudication by the Delaware Court of his or her entitlement to such indemnification or advancement. Alternatively, Indemnitee, at his or her option, may seek an award in arbitration to be conducted by a single arbitrator pursuant to the Commercial Arbitration Rules of the American Arbitration Association. Indemnitee shall commence such proceeding seeking an adjudication or an award in arbitration within 180 days following the date on which Indemnitee first has the right to commence such proceeding pursuant to this Section 12(a); provided, however, that the foregoing time limitation shall not apply in respect of a proceeding brought by Indemnitee to enforce his or her rights under Section 5 of this Agreement. The Company shall not oppose Indemnitee's right to seek any such adjudication or award in arbitration.

(b) In the event that a determination shall have been made pursuant to Section 10(a) of this Agreement that Indemnitee is not entitled to indemnification, any judicial proceeding or arbitration commenced pursuant to this Section 12 shall be conducted in all respects as a de novo trial, or arbitration, on the merits and Indemnitee shall not be prejudiced by reason of that adverse determination. In any judicial proceeding or arbitration commenced pursuant to this Section 12, the Company shall have the burden of proving Indemnitee is not entitled to indemnification or advancement, as the case may be.

(c) If a determination shall have been made pursuant to Section 10(a) of this Agreement that Indemnitee is entitled to indemnification, the Company shall be bound by such determination in any judicial proceeding or arbitration commenced pursuant to this Section 12, absent (i) a misstatement by Indemnitee of a material fact, or an omission of a material fact necessary to make Indemnitee's statement not materially misleading, in connection with the request for indemnification, or (ii) a prohibition of such indemnification under applicable law.

(d) The Company shall be precluded from asserting in any judicial proceeding or arbitration commenced pursuant to this Section 12 that the procedures and presumptions of this Agreement are not valid, binding and enforceable and shall stipulate in any such court or before any such arbitrator that the Company is bound by all the provisions of this Agreement.

(e) The Company shall indemnify Indemnitee to the fullest extent permitted by law against any and all Enforcement Expenses and, if requested by Indemnitee, shall (within thirty (30) days after receipt by the Company of a written request therefor) advance, to the extent

not prohibited by law, such Enforcement Expenses to Indemnitee, which are incurred by Indemnitee in connection with any action brought by Indemnitee for indemnification or advancement from the Company under this Agreement or under any directors' and officers' liability insurance policies maintained by the Company in the suit for which indemnification or advancement is being sought. Such written request for advancement shall include invoices received by Indemnitee in connection with such Enforcement Expenses but, in the case of invoices in connection with legal services, any references to legal work performed or to expenditures made that would cause Indemnitee to waive any privilege accorded by applicable law need not be included with the invoice. In the event of a change in control or the Company's becoming insolvent, the Company shall maintain in force any and all insurance policies then maintained by the Company in providing insurance — directors' and officers' liability, fiduciary, employment practices or otherwise — in respect of the individual directors and officers of the Company, for a fixed period of six years thereafter (a "Tail Policy"). Such coverage shall be non-cancellable and shall be placed and serviced for the duration of its term by the Company's incumbent insurance broker. Such broker shall place the Tail policy with the incumbent insurance carriers using the policies that were in place at the time of the change of control event (unless the incumbent carriers will not offer such policies, in which case the Tail Policy placed by the Company's insurance broker shall be substantially comparable in scope and amount as the expiring policies, and the insurance carriers for the Tail Policy shall have an AM Best rating that is the same or better than the AM Best ratings of the expiring policies).

(f) Notwithstanding anything in this Agreement to the contrary, no determination as to entitlement to indemnification under this Agreement shall be required to be made prior to the final disposition of the Proceeding, including any appeal therein.

Section 13. Non-exclusivity; Survival of Rights; Insurance; Primacy of Indemnification; Subrogation.

(a) The rights of indemnification and to receive advancement as provided by this Agreement shall not be deemed exclusive of any other rights to which Indemnitee may at any time be entitled under applicable law, the Charter, the Bylaws, any agreement, a vote of stockholders or a resolution of directors, or otherwise. No amendment, alteration or repeal of this Agreement or of any provision hereof shall limit or restrict any right of Indemnitee under this Agreement in respect of any action taken or omitted by such Indemnitee in his or her Corporate Status prior to such amendment, alteration or repeal. To the extent that a change in Delaware law, whether by statute or judicial decision, permits greater indemnification or advancement than would be afforded currently under the Charter, Bylaws and this Agreement, it is the intent of the parties hereto that Indemnitee shall enjoy by this Agreement the greater benefits so afforded by such change. No right or remedy herein conferred is intended to be exclusive of any other right or remedy, and every other right and remedy shall be cumulative and in addition to every other right and remedy given hereunder or now or hereafter existing at law or in equity or otherwise. The assertion or employment of any right or remedy hereunder, or otherwise, shall not prevent the concurrent assertion or employment of any other right or remedy.

(b) To the extent that the Company maintains an insurance policy or policies providing liability insurance for directors, managers, partners, officers, employees, agents or trustees of the Company or of any other Enterprise, Indemnitee shall be covered by such policy

or policies in accordance with its or their terms to the maximum extent of the coverage available for any such director, manager, partner, officer, employee, agent or trustee under such policy or policies. If, at the time of the receipt of a notice of a claim pursuant to the terms hereof, the Company has director and officer liability insurance in effect, the Company shall give prompt notice of the commencement of such proceeding to the insurers in accordance with the procedures set forth in the respective policies. The Company shall thereafter take all necessary or desirable action to cause such insurers to pay, on behalf of Indemnitee, all amounts payable as a result of such proceeding in accordance with the terms of such policies.

(c) [The Company hereby acknowledges that Indemnitee has certain rights to indemnification, advancement of expenses and/or insurance provided by [] and certain of its affiliates (collectively, the "Secondary Indemnitors"). The Company hereby agrees (i) that it is the indemnitor of first resort (i.e., its obligations to Indemnitee are primary and any obligation of the Secondary Indemnitors to advance expenses or to provide indemnification for the same expenses or liabilities incurred by Indemnitee are secondary), (ii) that it shall be required to advance the full amount of expenses incurred by Indemnitee and shall be liable for the full amount of all Expenses, judgments, penalties, fines and amounts paid in settlement to the extent legally permitted and as required by the terms of this Agreement and the Charter and/or Bylaws (or any other agreement between the Company and Indemnitee), without regard to any rights Indemnitee may have against the Secondary Indemnitors, and (iii) that it irrevocably waives, relinquishes and releases the Secondary Indemnitors from any and all claims against the Secondary Indemnitors for contribution, subrogation or any other recovery of any kind in respect thereof. The Company further agrees that no advancement or payment by the Secondary Indemnitors on behalf of Indemnitee with respect to any claim for which Indemnitee has sought indemnification from the Company shall affect the foregoing and the Secondary Indemnitors shall have a right of contribution and/or be subrogated to the extent of such advancement or payment to all of the rights of recovery of Indemnitee against the Company. The Company and Indemnitee agree that the Secondary Indemnitors are express third party beneficiaries of the terms of this Section 13(c).]

(d) [Except as provided in paragraph (c) above,] in the event of any payment under this Agreement, the Company shall be subrogated to the extent of such payment to all of the rights of recovery of Indemnitee [(other than against the Secondary Indemnitors)], who shall execute all papers required and take all action necessary to secure such rights, including execution of such documents as are necessary to enable the Company to bring suit to enforce such rights.

(e) [Except as provided in paragraph (c) above,] the Company's obligation to provide indemnification or advancement hereunder to Indemnitee who is or was serving at the request of the Company as a director, manager, partner, officer, employee, agent or trustee of any other Enterprise shall be reduced by any amount Indemnitee has actually received as indemnification or advancement from such other Enterprise.

Section 14. Duration of Agreement. This Agreement shall continue until and terminate upon the later of: (a) ten (10) years after the date that Indemnitee shall have ceased to serve as a director of the Company or (b) one (1) year after the final termination of any Proceeding, including any appeal, then pending in respect of which Indemnitee is granted rights

of indemnification or advancement hereunder and of any proceeding commenced by Indemnitee pursuant to Section 12 of this Agreement relating thereto. This Agreement shall be binding upon the Company and its successors and assigns and shall inure to the benefit of Indemnitee and his or her heirs, executors and administrators. The Company shall require and cause any successor (whether direct or indirect by purchase, merger, consolidation or otherwise) to all, substantially all or a substantial part, of the business and/or assets of the Company, by written agreement in form and substance satisfactory to Indemnitee, expressly to assume and agree to perform this Agreement to the fullest extent permitted by law.

Section 15. Severability. If any provision or provisions of this Agreement shall be held to be invalid, illegal or unenforceable for any reason whatsoever: (a) the validity, legality and enforceability of the remaining provisions of this Agreement (including, without limitation, each portion of any section of this Agreement containing any such provision held to be invalid, illegal or unenforceable, that is not itself invalid, illegal or unenforceable) shall not in any way be affected or impaired thereby and shall remain enforceable to the fullest extent permitted by law; (b) such provision or provisions shall be deemed reformed to the extent necessary to conform to applicable law and to give the maximum effect to the intent of the parties hereto; and (c) to the fullest extent possible, the provisions of this Agreement (including, without limitation, each portion of any section of this Agreement containing any such provision held to be invalid, illegal or unenforceable, that is not itself invalid, illegal or unenforceable) shall be construed so as to give effect to the intent manifested thereby.

Section 16. Enforcement.

(a) The Company expressly confirms and agrees that it has entered into this Agreement and assumed the obligations imposed on it hereby in order to induce Indemnitee to serve or continue to serve as a director of the Company, and the Company acknowledges that Indemnitee is relying upon this Agreement in serving as a director of the Company.

(b) This Agreement constitutes the entire agreement between the parties hereto with respect to the subject matter hereof and supersedes all prior agreements and understandings, oral, written and implied, between the parties hereto with respect to the subject matter hereof; provided, however, that this Agreement is a supplement to and in furtherance of the Charter, the Bylaws and applicable law, and shall not be deemed a substitute therefor, nor to diminish or abrogate any rights of Indemnitee thereunder.

Section 17. Modification and Waiver. No supplement, modification or amendment, or waiver of any provision, of this Agreement shall be binding unless executed in writing by the parties thereto. No waiver of any of the provisions of this Agreement shall be deemed or shall constitute a waiver of any other provisions of this Agreement nor shall any waiver constitute a continuing waiver. No supplement, modification or amendment of this Agreement or of

any provision hereof shall limit or restrict any right of Indemnitee under this Agreement in respect of any action taken or omitted by such Indemnitee prior to such supplement, modification or amendment.

Section 18. Notice by Indemnitee. Indemnitee agrees promptly to notify the Company in writing upon being served with any summons, citation, subpoena, complaint, indictment,

information or other document relating to any Proceeding or matter which may be subject to indemnification, reimbursement or advancement as provided hereunder. The failure of Indemnitee to so notify the Company shall not relieve the Company of any obligation which it may have to Indemnitee under this Agreement or otherwise.

Section 19. Notices. All notices, requests, demands and other communications under this Agreement shall be in writing and shall be deemed to have been duly given if (i) delivered by hand and received for by the party to whom said notice or other communication shall have been directed, (ii) mailed by certified or registered mail with postage prepaid, on the third business day after the date on which it is so mailed, (iii) mailed by reputable overnight courier and received for by the party to whom said notice or other communication shall have been directed or (iv) sent by facsimile transmission, with receipt of oral confirmation that such transmission has been received:

- (a) If to Indemnitee, at such address as Indemnitee shall provide to the Company.
- (b) If to the Company to:

Scholar Rock Holding Corporation.
620 Memorial Drive, 2nd Floor
Cambridge, Massachusetts 02139
Attention: [-]

or to any other address as may have been furnished to Indemnitee by the Company.

Section 20. Contribution. To the fullest extent permissible under applicable law, if the indemnification provided for in this Agreement is unavailable to Indemnitee for any reason whatsoever, the Company, in lieu of indemnifying Indemnitee, shall contribute to the amount incurred by Indemnitee, whether for judgments, fines, penalties, excise taxes, amounts paid or to be paid in settlement and/or for Expenses, in connection with any Proceeding in such proportion as is deemed fair and reasonable in light of all of the circumstances in order to reflect (i) the relative direct benefits received by the Company and Indemnitee in connection with the event(s) and/or transaction(s) giving rise to such Proceeding; and/or (ii) the relative fault of the Company (and its directors, officers, employees and agents) and Indemnitee in connection with such event(s) and/or transactions. The Company hereby agrees to fully indemnify and hold harmless Indemnitee from any claims for contribution which may be brought by Officers, Directors or employees of the Company (other than Indemnitee) who may be jointly liable with Indemnitee.

Section 21. Internal Revenue Code Section 409A. The Company intends for this Agreement to comply with the Indemnification exception under Section 1.409A-1(b)(10) of the regulations promulgated under the Internal Revenue Code of 1986, as amended (the "Code"), which provides that indemnification of, or the purchase of an insurance policy providing for payments of, all or part of the expenses incurred or damages paid or payable by Indemnitee with respect to a bona fide claim against Indemnitee or the Company do not provide for a deferral of compensation, subject to Section 409A of the Code, where such claim is based on actions or failures to act by Indemnitee in his or her capacity as a service provider of the Company. The parties intend that this Agreement be interpreted and construed with such intent.

Section 22. Applicable Law and Consent to Jurisdiction. This Agreement and the legal relations among the parties shall be governed by, and construed and enforced in accordance with, the laws of the State of Delaware, without regard to its conflict of laws rules. Except with respect to any arbitration commenced by Indemnitee pursuant to Section 12(a) of this Agreement, the Company and Indemnitee hereby irrevocably and unconditionally (i) agree that any action or proceeding arising out of or in connection with this Agreement shall be brought only in the Delaware Court, and not in any other state or federal court in the United States of America or any court in any other country, (ii) consent to submit to the exclusive jurisdiction of the Delaware Court for purposes of any action or proceeding arising out of or in connection with this Agreement, (iii) consent to service of process at the address set forth in Section 19 of this Agreement with the same legal force and validity as if served upon such party personally within the State of Delaware, (iv) waive any objection to the laying of venue of any such action or proceeding in the Delaware Court, and (v) waive, and agree not to plead or to make, any claim that any such action or proceeding brought in the Delaware Court has been brought in an improper or inconvenient forum.

Section 23. Determination of Good Faith/Safe Harbor. For purposes of any determination of good faith, Indemnitee [or "an independent director"] shall be presumed to have acted in good faith if Indemnitee's action is based on the records or books of account of the Enterprise, including financial statements, or on information supplied to Indemnitee by the officers of the Enterprise in the course of their duties, or on the advice of legal counsel for the Enterprise or the Board or counsel selected by any committee of the Board or on information or records given or reports made to the Enterprise by an independent certified public accountant or by an appraiser, investment banker, compensation consultant, or other expert selected with reasonable care by the Company or the Board or any committee of the Board. The provisions of this Section XXs shall not be deemed to be exclusive or to limit in any way the other circumstances in which the Indemnitee may be deemed to have met the applicable standard of conduct. Whether or not the foregoing provisions of this Section are satisfied, it shall in any event be presumed that Director has at all times acted in good faith and in a manner he or she reasonably believed to be in or not opposed to the best interests of the Company.

Section 24. Monetary Damages Insufficient/Specific Performance. The Company and Indemnitee agree that a monetary remedy for breach of this Agreement may be inadequate, impracticable and difficult of proof, and further agree that such breach may cause Indemnitee irreparable harm. Accordingly, the parties hereto agree that Indemnitee may enforce this Agreement by seeking injunctive relief and/or specific performance hereof, without any necessity of showing actual damage or irreparable harm (having agreed that actual and irreparable harm will result in not forcing the Company to specifically perform its obligations pursuant to this Agreement) and that by seeking injunctive relief and/or specific performance, Indemnitee shall not be precluded from seeking or obtaining any other relief to which he may be entitled. The Company and Indemnitee further agree that Indemnitee may be entitled to such specific performance and injunctive relief, including temporary restraining orders, preliminary injunctions and permanent injunctions, without the necessity of posting bonds or other undertaking in connection therewith. The Company acknowledges that in the absence of a waiver, a bond or undertaking may be required of Indemnitee by the Court, and the Company hereby waives any such requirement of a bond or undertaking.

Section 25. Headings. The headings of the paragraphs of this Agreement are inserted for convenience only and shall not be deemed to constitute part of this Agreement or to affect the construction thereof.

Section 26. Identical Counterparts. This Agreement may be executed in one or more counterparts, each of which shall for all purposes be deemed to be an original but all of which together shall constitute one and the same Agreement. Only one such counterpart signed by the party against whom enforceability is sought needs to be produced to evidence the existence of this Agreement.

[Remainder of Page Intentionally Left Blank]

IN WITNESS WHEREOF, the parties have caused this Agreement to be signed as of the day and year first above written.

SCHOLAR ROCK HOLDING CORPORATION

By:

Name: Nagesh K. Mahanthappa
Title: President and Chief Executive Officer

INDEMNITEE

Name:
Title:

SCHOLAR ROCK HOLDING CORPORATION

FORM OF OFFICER INDEMNIFICATION AGREEMENT

This Indemnification Agreement ("Agreement") is made as of _____ by and between Scholar Rock Holding Corporation, a Delaware corporation (the "Company"), and ("Indemnitee").(1)

RECITALS

WHEREAS, the Company desires to attract and retain the services of highly qualified individuals, such as Indemnitee, to serve the Company;

WHEREAS, in order to induce Indemnitee to provide or continue to provide services to the Company, the Company wishes to provide for the indemnification of, and advancement of expenses to, Indemnitee to the maximum extent permitted by law;

WHEREAS, the Amended and Restated Certificate of Incorporation (as amended and in effect from time to time, the "Charter") and the Amended and Restated Bylaws (as amended and in effect from time to time, the "Bylaws") of the Company require indemnification of the officers and directors of the Company, and Indemnitee may also be entitled to indemnification pursuant to the General Corporation Law of the State of Delaware (the "DGCL");

WHEREAS, the Charter, the Bylaws and the DGCL expressly provide that the indemnification provisions set forth therein are not exclusive, and thereby contemplate that contracts may be entered into between the Company and members of the board of directors, officers and other persons with respect to indemnification;

WHEREAS, the Board of Directors of the Company (the "Board") has determined that the increased difficulty in attracting and retaining highly qualified persons such as Indemnitee is detrimental to the best interests of the Company's stockholders;

WHEREAS, it is reasonable and prudent for the Company contractually to obligate itself to indemnify, and to advance expenses on behalf of, such persons to the fullest extent permitted by applicable law, regardless of any amendment or revocation of the Charter or the Bylaws, so that they will serve or continue to serve the Company free from undue concern that they will not be so indemnified; and

WHEREAS, this Agreement is a supplement to and in furtherance of the indemnification provided in the Charter, the Bylaws and any resolutions adopted pursuant thereto, and shall not be deemed a substitute therefor, nor to diminish or abrogate any rights of Indemnitee thereunder.

NOW, THEREFORE, in consideration of the premises and the covenants contained herein, the Company and Indemnitee do hereby covenant and agree as follows:

(1) To be entered into with all C-level officers, Section 16 officers and other officers and employees of the Company as specified by the Company.

Section 1. Services to the Company. Indemnitee agrees to [continue to] serve as [a director and] an officer of the Company. Indemnitee may at any time and for any reason resign from [any] such position (subject to any other contractual obligation or any obligation imposed by law), in which event the Company shall have no obligation under this Agreement to continue Indemnitee in such position. This Agreement shall not be deemed an employment contract between the Company (or any of its subsidiaries or any Enterprise) and Indemnitee.

Section 2. Definitions.

As used in this Agreement:

(a) "Change in Control" shall mean (i) the sale of all or substantially all of the assets of the Company on a consolidated basis to an unrelated person or entity, (ii) a merger, reorganization or consolidation pursuant to which the holders of the Company's outstanding voting power and outstanding stock immediately prior to such transaction do not own a majority of the outstanding voting power and outstanding stock or other equity interests of the resulting or successor entity (or its ultimate parent, if applicable) immediately upon completion of such transaction, (iii) the sale of all of the Stock of the Company to an unrelated person, entity or group thereof acting in concert, or (iv) any other transaction in which the owners of the Company's outstanding voting power immediately prior to such transaction do not own at least a majority of the outstanding voting power of the Company or any successor entity immediately upon completion of the transaction other than as a result of the acquisition of securities directly from the Company.

(b) "Corporate Status" describes the status of a person as a current or former [director or] officer of the Company or current or former director, manager, partner, officer, employee, agent or trustee of any other Enterprise which such person is or was serving at the request of the Company.

(c) "Enforcement Expenses" shall include all reasonable attorneys' fees, court costs, transcript costs, fees of experts, travel expenses, duplicating costs, printing and binding costs, telephone charges, postage, delivery service fees, and all other out-of-pocket disbursements or expenses incurred in connection with an action to enforce indemnification or advancement rights, or an appeal from such action. Expenses, however, shall not include fees, salaries, wages or benefits owed to Indemnitee.

(d) "Enterprise" shall mean any corporation (other than the Company), partnership, joint venture, trust, employee benefit plan, limited liability company, or other legal entity of which Indemnitee is or was serving at the request of the Company as a director, manager, partner, officer, employee, agent or trustee, including as a deemed fiduciary thereto.

(e) "Expenses" shall include all reasonable attorneys' fees, court costs, transcript costs, fees of experts, travel expenses, duplicating costs, printing and binding costs, telephone charges, postage, delivery service fees, and all other out-of-pocket disbursements or expenses of the types customarily incurred in connection with prosecuting, defending, preparing to prosecute or defend, investigating, being or preparing to be a witness in, or otherwise

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participating in, a Proceeding or an appeal resulting from a Proceeding. Expenses, however, shall not include amounts paid in settlement by Indemnitee, the amount of judgments or fines against Indemnitee or fees, salaries, wages or benefits owed to Indemnitee.

(f) "Independent Counsel" means a law firm, or a partner (or, if applicable, member or shareholder) of such a law firm, that is experienced in matters of Delaware corporation law and neither presently is, nor in the past five (5) years has been, retained to represent: (i) the Company, any subsidiary of the Company, any Enterprise or Indemnitee in any matter material to any such party; or (ii) any other party to the Proceeding giving rise to a claim for indemnification hereunder. Notwithstanding the foregoing, the term "Independent Counsel" shall not include any person who, under the applicable standards of professional conduct then prevailing, would have a conflict of interest in representing either the Company or Indemnitee in an action to determine Indemnitee's rights under this Agreement. The Company agrees to pay the reasonable fees and expenses of the Independent Counsel referred to above and to fully indemnify such counsel against any and all expenses, claims, liabilities and damages arising out of or relating to this Agreement or its engagement pursuant hereto.

(g) The term "Proceeding" shall include any threatened, pending or completed action, suit, arbitration, alternate dispute resolution mechanism, investigation, inquiry, administrative hearing or any other actual, threatened or completed proceeding, whether brought in the right of the Company or otherwise and whether of a civil, criminal, administrative, regulatory or investigative nature, and whether formal or informal, in which Indemnitee was, is or will be involved as a party or otherwise by reason of the fact that Indemnitee is or was [a director or] an officer of the Company or is or was serving at the request of the Company as a director, manager, partner, officer, employee, agent or trustee of any Enterprise or by reason of any action taken by Indemnitee or of any action taken on his or her part while acting as [a director or] an officer of the Company or while serving at the request of the Company as a director, manager, partner, officer, employee, agent or trustee of any Enterprise, in each case whether or not serving in such capacity at the time any liability or expense is incurred for which indemnification, reimbursement or advancement of expenses can be provided under this Agreement; provided, however, that the term "Proceeding" shall not include any action, suit or arbitration, or part thereof, initiated by Indemnitee to enforce Indemnitee's rights under this Agreement as provided for in Section 12(a) of this Agreement.

Section 3. Indemnity in Third-Party Proceedings. The Company shall indemnify Indemnitee to the extent set forth in this Section 3 if Indemnitee is, or is threatened to be made, a party to or a participant in any Proceeding, other than a Proceeding by or in the right of the Company to procure a judgment in its favor. Pursuant to this Section 3, Indemnitee shall be indemnified against all Expenses, judgments, fines, penalties, excise taxes, and amounts paid in settlement actually and reasonably incurred by Indemnitee or on his or her behalf in connection with such Proceeding or any claim, issue or matter therein, if Indemnitee acted in good faith and in a manner he or she reasonably believed to be in or not opposed to the best interests of the Company and, in the case of a criminal proceeding, had no reasonable cause to believe that his or her conduct was unlawful.

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Section 4. Indemnity in Proceedings by or in the Right of the Company. The Company shall indemnify Indemnitee to the extent set forth in this Section 4 if Indemnitee is, or is threatened to be made, a party to or a participant in any Proceeding by or in the right of the Company to procure a judgment in its favor. Pursuant to this Section 4, Indemnitee shall be indemnified against all Expenses actually and reasonably incurred by Indemnitee or on his or her behalf in connection with such Proceeding or any claim, issue or matter therein, if Indemnitee acted in good faith and in a manner he or she reasonably believed to be in or not opposed to the best interests of the Company. No indemnification for Expenses shall be made under this Section 4 in respect of any claim, issue or matter as to which Indemnitee shall have been finally adjudged by a court to be liable to the Company, unless and only to the extent that the Delaware Court of Chancery (the "Delaware Court") shall determine upon application that, despite the adjudication of liability but in view of all the circumstances of the case, Indemnitee is fairly and reasonably entitled to indemnification for such expenses as the Delaware Court shall deem proper.

Section 5. Indemnification for Expenses of a Party Who is Wholly or Partly Successful. Notwithstanding any other provisions of this Agreement and except as provided in Section 7, to the extent that Indemnitee is a party to or a participant in any Proceeding and is successful in such Proceeding or in defense of any claim, issue or matter therein, the Company shall indemnify Indemnitee against all Expenses actually and reasonably incurred by him or her in connection therewith. If Indemnitee is not wholly successful in such Proceeding but is successful as to one or more but less than all claims, issues or matters in such Proceeding, the Company shall indemnify Indemnitee against all Expenses actually and reasonably incurred by Indemnitee or on his or her behalf in connection with each successfully resolved claim, issue or matter. For purposes of this Section and without limitation, the termination of any claim, issue or matter in such a Proceeding by dismissal, with or without prejudice, motion for summary judgment, settlement (with or without court approval), or upon a plea of nolo contendere or its equivalent, shall be deemed to be a successful result as to such claim, issue or matter.

Section 6. Reimbursement for Expenses of a Witness or in Response to a Subpoena. Notwithstanding any other provision of this Agreement, to the extent that Indemnitee, by reason of his or her Corporate Status, (i) is a witness in any Proceeding to which Indemnitee is not a party and is not threatened to be made a party or (ii) receives a subpoena with respect to any Proceeding to which Indemnitee is not a party and is not threatened to be made a party, the Company shall reimburse Indemnitee for all Expenses actually and reasonably incurred by him or her or on his or her behalf in connection therewith.

Section 7. Exclusions. Notwithstanding any provision in this Agreement to the contrary, the Company shall not be obligated under this Agreement:

(a) to indemnify for amounts otherwise indemnifiable hereunder (or for which advancement is provided hereunder) if and to the extent that Indemnitee has otherwise actually received such amounts under any insurance policy, contract, agreement or otherwise; provided that the foregoing shall not affect the rights of Indemnitee or the Secondary Indemnitors as set forth in Section 13(c); provided further, however, that payment made to Indemnitee pursuant to an insurance policy purchased and maintained by Indemnitee at his or her own expense of any

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amounts otherwise indemnifiable or obligated to be made pursuant to this Agreement shall not reduce the Company's obligations to Indemnitee pursuant to this Agreement.

(b) to indemnify for an accounting of profits made from the purchase and sale (or sale and purchase) by Indemnitee of securities of the Company within the meaning of Section 16(b) of the Securities Exchange Act of 1934, as amended, or similar provisions of state statutory law or common law, or from the purchase or sale by Indemnitee of such securities in violation of Section 306 of the Sarbanes-Oxley Act of 2002 ("SOX");

(c) to indemnify for any reimbursement of, or payment to, the Company by Indemnitee of any bonus or other incentive-based or equity-based compensation or of any profits realized by Indemnitee from the sale of securities of the Company pursuant to Section 304 of SOX or any formal policy of the Company adopted by the Board (or a committee thereof), or any other remuneration paid to Indemnitee if it shall be determined by a final judgment or other final adjudication that such remuneration was in violation of law;

(d) to indemnify with respect to any Proceeding, or part thereof, brought by Indemnitee against the Company, any legal entity which it controls, any director or officer thereof or any third party, unless (i) the Board has consented to the initiation of such Proceeding or part thereof and (ii) the Company provides the indemnification, in its sole discretion, pursuant to the powers vested in the Company under applicable law; provided, however, that this

Section 7(d) shall not apply to (A) counterclaims or affirmative defenses asserted by Indemnitee in an action brought against Indemnitee or (B) any action brought by Indemnitee for indemnification or advancement from the Company under this Agreement or under any directors' and officers' liability insurance policies maintained by the Company in the suit for which indemnification or advancement is being sought as described in Section 12; or

- (e) to provide any indemnification or advancement of expenses that is prohibited by applicable law (as such law exists at the time payment would otherwise be required pursuant to this Agreement).

Section 8. Advancement of Expenses. Subject to Section 9(b), the Company shall advance, to the extent not prohibited by law, the Expenses incurred by Indemnitee in connection with any Proceeding, and such advancement shall be made as incurred, and such advancement shall be made within thirty (30) days after the receipt by the Company of a statement or statements requesting such advances (including any invoices received by Indemnitee, which such invoices may be redacted as necessary to avoid the waiver of any privilege accorded by applicable law) from time to time, whether prior to or after final disposition of any Proceeding. Advances shall be unsecured and interest free. Advances shall be made without regard to Indemnitee's (i) ability to repay the expenses, (ii) ultimate entitlement to indemnification under the other provisions of this Agreement, and (iii) entitlement to and availability of insurance coverage, including advancement, payment or reimbursement of defense costs, expenses of covered loss under the provisions of any applicable insurance policy (including, without limitation, whether such advancement, payment or reimbursement is withheld, conditioned or delayed by the insurer(s)). Without limiting the generality or effect of the foregoing, within thirty days after any request for advancement by Indemnitee pursuant to the first sentence of this Section 8, the Company shall, in accordance with such request (but without duplication), (a) pay

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such Expenses on behalf of Indemnitee, (b) advance to Indemnitee funds in an amount sufficient to pay such Expenses, or (c) reimburse Indemnitee for such Expenses. The Company shall not seek from a court, or agree to, a "bar order" which would have the effect of prohibiting or limiting the Indemnitee's rights to receive advancement of expenses under this Agreement. Indemnitee shall qualify for advances (including any Enforcement Expenses as set forth in Section 12(e)) upon the execution and delivery to the Company of this Agreement which shall constitute an undertaking providing that Indemnitee undertakes to the fullest extent required by law to repay the advance if and to the extent that it is ultimately determined by a court of competent jurisdiction in a final judgment, not subject to appeal, that Indemnitee is not entitled to be indemnified by the Company. No other form of undertaking shall be required other than the execution of this Agreement. The right to advances under this paragraph shall in all events continue until final disposition of any Proceeding, including any appeal therein. Nothing in this Section 8 shall limit Indemnitee's right to advancement pursuant to Section 12(e) of this Agreement.

Section 9. Procedure for Notification and Defense of Claim.

(a) To obtain indemnification under this Agreement, Indemnitee shall submit to the Company a written request therefor specifying the basis for the claim, the amounts for which Indemnitee is seeking payment under this Agreement, and all documentation (which is not privileged or otherwise protected from disclosure and which is reasonably available to Indemnitee) related thereto as reasonably requested by the Company.

(b) In the event that the Company shall be obligated hereunder to provide indemnification for or make any advancement of Expenses with respect to any Proceeding, the Company shall be entitled to assume the defense of such Proceeding, or any claim, issue or matter therein, with counsel approved by Indemnitee (which approval shall not be unreasonably withheld or delayed) upon the delivery to Indemnitee of written notice of the Company's election to do so. After delivery of such notice, approval of such counsel by Indemnitee and the retention of such counsel by the Company, the Company will not be liable to Indemnitee under this Agreement for any fees or expenses of separate counsel subsequently employed by or on behalf of Indemnitee with respect to the same Proceeding; provided that (i) Indemnitee shall have the right to employ separate counsel in any such Proceeding at Indemnitee's expense and (ii) if (A) the employment of separate counsel by Indemnitee has been previously authorized by the Company, (B) Indemnitee shall have reasonably concluded that there may be a conflict of interest between the Company and Indemnitee in the conduct of such defense, or (C) the Company shall not continue to retain such counsel to defend such Proceeding, then the fees and expenses actually and reasonably incurred by Indemnitee with respect to his or her separate counsel shall be Expenses hereunder. Indemnitee agrees that any such separate counsel retained by Indemnitee will be a member of any approved list of panel counsel under the Company's applicable directors' and officers' insurance policy, should the applicable policy provide for a panel of approved counsel and should such approved panel list comprise law firms with well-established reputations in the type of litigation at issue.

- (c) In the event that the Company does not assume the defense in a Proceeding pursuant to paragraph (b) above, then the Company will be entitled to participate in the Proceeding at its own expense.

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(d) The Company shall not be liable to indemnify Indemnitee under this Agreement for any amounts paid in settlement of any Proceeding effected without its prior written consent (which consent shall not be unreasonably withheld or delayed). The Company shall not, without the prior written consent of Indemnitee (which consent shall not be unreasonably withheld or delayed), enter into any settlement which (i) includes an admission of fault of Indemnitee, any non-monetary remedy imposed on Indemnitee or any monetary damages for which Indemnitee is not wholly and actually indemnified hereunder or (ii) with respect to any Proceeding with respect to which Indemnitee may be or is made a party or may be otherwise entitled to seek indemnification hereunder, does not include the full release of Indemnitee from all liability in respect of such Proceeding.

Section 10. Procedure Upon Application for Indemnification (2)

(a) Upon written request by Indemnitee for indemnification pursuant to Section 9(a), a determination, if such determination is required by applicable law, with respect to Indemnitee's entitlement to indemnification hereunder shall be made in the specific case by one of the following methods: [(x) if a Change in Control shall have occurred, by Independent Counsel in a written opinion to the Board; or (y) in any other case,] (i) by a majority vote of the disinterested directors, even though less than a quorum; (ii) by a committee of disinterested directors designated by a majority vote of the disinterested directors, even though less than a quorum; or (iii) if there are no disinterested directors or if the disinterested directors so direct, by Independent Counsel in a written opinion to the Board. For purposes hereof, disinterested directors are those members of the Board who are not parties to the action, suit or proceeding in respect of which indemnification is sought. In the case that such determination is made by Independent Counsel, a copy of Independent Counsel's written opinion shall be delivered to Indemnitee and, if it is so determined that Indemnitee is entitled to indemnification, payment to Indemnitee shall be made within thirty (30) days after such determination. Indemnitee shall cooperate with the Independent Counsel or the Company, as applicable, in making such determination with respect to Indemnitee's entitlement to indemnification, including providing to such counsel or the Company, upon reasonable advance request, any documentation or information which is not privileged or otherwise protected from disclosure and which is reasonably available to Indemnitee and reasonably necessary to such determination. Any out-of-pocket costs or expenses (including reasonable attorneys' fees and disbursements) actually and reasonably incurred by Indemnitee in so cooperating with the Independent Counsel or the Company shall be borne by the Company (irrespective of the determination as to Indemnitee's entitlement to indemnification) and the Company hereby indemnifies and agrees to hold Indemnitee harmless therefrom.

(b) If the determination of entitlement to indemnification is to be made by Independent Counsel pursuant to Section 10(a), the Independent Counsel shall be selected by the Board; provided that, if a Change in Control shall have occurred and indemnification is being requested by Indemnitee hereunder in his or her capacity as a director of the Company, the Independent Counsel shall be selected by Indemnitee. Indemnitee [or the Company, as the case may be,] may, within ten (10) days after written notice of such selection, deliver to the Company

(2) Bracketed portions for CEO Director version only

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[or Indemnitee, as the case may be,] a written objection to such selection; provided, however, that such objection may be asserted only on the ground that the Independent Counsel so selected does not meet the requirements of "Independent Counsel" as defined in Section 2 of this Agreement, and the objection shall set forth with particularity the factual basis of such assertion. Absent a proper and timely objection, the person so selected shall act as Independent Counsel. If such written objection is so made and substantiated, the Independent Counsel so selected may not serve as Independent Counsel unless and until such objection is withdrawn or the Delaware Court has determined that such objection is without merit. If, within twenty (20) days after the later of (i) submission by Indemnitee of a written request for indemnification pursuant to Section 9(a), and (ii) the final disposition of the Proceeding, including any appeal therein, no Independent Counsel shall have been selected without objection, either Indemnitee or the Company may petition the Delaware Court for resolution of any objection which shall have been made by Indemnitee or the Company to the selection of Independent Counsel and/or for the appointment as Independent Counsel of a person selected by the court or by such other person as the court shall designate. The person with respect to whom all objections are so resolved or the person so appointed shall act as Independent Counsel under Section 10(a) hereof. Upon the due commencement of any judicial proceeding or arbitration pursuant to Section 12(a) of this Agreement, Independent Counsel shall be discharged and relieved of any further responsibility in such capacity (subject to the applicable standards of professional conduct then prevailing).

Section 11. Presumptions and Effect of Certain Proceedings.

(a) To the extent permitted by applicable law, in making a determination with respect to entitlement to indemnification hereunder, it shall be presumed that Indemnitee is entitled to indemnification under this Agreement if Indemnitee has submitted a request for indemnification in accordance with Section 9(a) of this Agreement, and the Company shall have the burden of proof to overcome that presumption in connection with the making of any determination contrary to that presumption. Neither (i) the failure of the Company or of Independent Counsel to have made a determination prior to the commencement of any action pursuant to this Agreement that indemnification is proper in the circumstances because Indemnitee has met the applicable standard of conduct, nor (ii) an actual determination by the Company or by Independent Counsel that Indemnitee has not met such applicable standard of conduct, shall be a defense to the action or create a presumption that Indemnitee has not met the applicable standard of conduct.

(b) The termination of any Proceeding or of any claim, issue or matter therein, by judgment, order, settlement or conviction, or upon a plea of guilty, nolo contendere or its equivalent, shall not (except as otherwise expressly provided in this Agreement) of itself adversely affect the right of Indemnitee to indemnification or create a presumption that Indemnitee did not act in good faith and in a manner which he or she reasonably believed to be in or not opposed to the best interests of the Company or, with respect to any criminal Proceeding, that Indemnitee had reasonable cause to believe that his or her conduct was unlawful.

- (c) The knowledge and/or actions, or failure to act, of any director, manager, partner, officer, employee, agent or trustee of the Company, any subsidiary of the Company, or

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any Enterprise shall not be imputed to Indemnitee for purposes of determining the right to indemnification under this Agreement.

Section 12. Remedies of Indemnitee.

(a) Subject to Section 12(f), in the event that (i) a determination is made pursuant to Section 10 of this Agreement that Indemnitee is not entitled to indemnification under this Agreement, (ii) advancement of Expenses is not timely made pursuant to Section 8 of this Agreement, (iii) no determination of entitlement to indemnification shall have been made pursuant to Section 10(a) of this Agreement within sixty (60) days after receipt by the Company of the request for indemnification for which a determination is to be made other than by Independent Counsel, (iv) payment of indemnification or reimbursement of expenses is not made pursuant to Section 5 or 6 or the last sentence of Section 10(a) of this Agreement within thirty (30) days after receipt by the Company of a written request therefor (including any invoices received by Indemnitee, which such invoices may be redacted as necessary to avoid the waiver of any privilege accorded by applicable law) or (v) payment of indemnification pursuant to Section 3 or 4 of this Agreement is not made within thirty (30) days after a determination has been made that Indemnitee is entitled to indemnification, Indemnitee shall be entitled to an adjudication by the Delaware Court of his or her entitlement to such indemnification or advancement. Alternatively, Indemnitee, at his or her option, may seek an award in arbitration to be conducted by a single arbitrator pursuant to the Commercial Arbitration Rules of the American Arbitration Association. Indemnitee shall commence such proceeding seeking an adjudication or an award in arbitration within 180 days following the date on which Indemnitee first has the right to commence such proceeding pursuant to this Section 12(a); provided, however, that the foregoing time limitation shall not apply in respect of a proceeding brought by Indemnitee to enforce his or her rights under Section 5 of this Agreement. The Company shall not oppose Indemnitee's right to seek any such adjudication or award in arbitration.

(b) In the event that a determination shall have been made pursuant to Section 10(a) of this Agreement that Indemnitee is not entitled to indemnification, any judicial proceeding or arbitration commenced pursuant to this Section 12 shall be conducted in all respects as a de novo trial, or arbitration, on the merits and Indemnitee shall not be prejudiced by reason of that adverse determination. In any judicial proceeding or arbitration commenced pursuant to this Section 12, the Company shall have the burden of proving Indemnitee is not entitled to indemnification or advancement, as the case may be.

(c) If a determination shall have been made pursuant to Section 10(a) of this Agreement that Indemnitee is entitled to indemnification, the Company shall be bound by such determination in any judicial proceeding or arbitration commenced pursuant to this Section 12, absent (i) a misstatement by Indemnitee of a material fact, or an omission of a material fact necessary to make Indemnitee's statement not materially misleading, in connection with the request for indemnification, or (ii) a prohibition of such indemnification under applicable law.

(d) The Company shall be precluded from asserting in any judicial proceeding or arbitration commenced pursuant to this Section 12 that the procedures and presumptions of this Agreement are not valid, binding and enforceable and shall stipulate in any such court or before any such arbitrator that the Company is bound by all the provisions of this Agreement.

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(e) The Company shall indemnify Indemnitee to the fullest extent permitted by law against any and all Enforcement Expenses and, if requested by Indemnitee, shall (within thirty (30) days after receipt by the Company of a written request therefor) advance, to the extent not prohibited by law, such Enforcement Expenses to Indemnitee, which are incurred by Indemnitee in connection with any action brought by Indemnitee for indemnification or advancement from the Company under this Agreement or under any directors' and officers' liability insurance policies maintained by the Company in the suit for which indemnification or advancement is being sought. Such written request for advancement shall include invoices received by Indemnitee in connection with such Enforcement Expenses but, in the case of invoices in connection with legal services, any references to legal work performed or to expenditures made that would cause Indemnitee to waive any privilege accorded by applicable law need not be included with the invoice. In the event of a change in control or the Company's becoming insolvent, the Company shall maintain in force any and all insurance policies then maintained by the Company in providing insurance—directors' and officers' liability, fiduciary, employment practices or otherwise—in respect of the individual directors and officers of the Company, for a fixed period of six years thereafter (a "Tail Policy"). Such coverage shall be non-cancellable and shall be placed and serviced for the duration of its term by the Company's incumbent insurance broker. Such broker shall place the Tail policy with the incumbent insurance carriers using the policies that were in place at the time of the change of control event (unless the incumbent carriers will not offer such policies, in which case the Tail Policy placed by the Company's insurance broker shall be substantially comparable in scope and amount as the expiring policies, and the insurance carriers for the Tail Policy shall have an AM Best rating that is the same or better than the AM Best ratings of the expiring policies).

(f) Notwithstanding anything in this Agreement to the contrary, no determination as to entitlement to indemnification under this Agreement shall be required to be made prior to the final disposition of the Proceeding, including any appeal therein.

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Section 13. Non-exclusivity; Survival of Rights; Insurance; Subrogation.

(a) The rights of indemnification and to receive advancement as provided by this Agreement shall not be deemed exclusive of any other rights to which Indemnitee may at any time be entitled under applicable law, the Charter, the Bylaws, any agreement, a vote of stockholders or a resolution of directors, or otherwise. No amendment, alteration or repeal of this Agreement or of any provision hereof shall limit or restrict any right of Indemnitee under this Agreement in respect of any action taken or omitted by such Indemnitee in his or her Corporate Status prior to such amendment, alteration or repeal. To the extent that a change in Delaware law, whether by statute or judicial decision, permits greater indemnification or advancement than would be afforded currently under the Charter, Bylaws and this Agreement, it is the intent of the parties hereto that Indemnitee shall enjoy by this Agreement the greater benefits so afforded by such change. No right or remedy herein conferred is intended to be exclusive of any other right or remedy, and every other right and remedy shall be cumulative and in addition to every other right and remedy given hereunder or now or hereafter existing at law or in equity or otherwise. The assertion or employment of any right or remedy hereunder, or otherwise, shall not prevent the concurrent assertion or employment of any other right or remedy.

(b) To the extent that the Company maintains an insurance policy or policies providing liability insurance for directors, managers, partners, officers, employees, agents or trustees of the Company or of any other Enterprise, Indemnitee shall be covered by such policy or policies in accordance with its or their terms to the maximum extent of the coverage available for any such director, manager, partner, officer, employee, agent or trustee under such policy or policies. If, at the time of the receipt of a notice of a claim pursuant to the terms hereof, the Company has director and officer liability insurance in effect, the Company shall give prompt notice of the commencement of such proceeding to the insurers in accordance with the procedures set forth in the respective policies. The Company shall thereafter take all necessary or desirable action to cause such insurers to pay, on behalf of Indemnitee, all amounts payable as a result of such proceeding in accordance with the terms of such policies.

(c) In the event of any payment under this Agreement, the Company shall be subrogated to the extent of such payment to all of the rights of recovery of Indemnitee, who shall execute all papers required and take all action necessary to secure such rights, including execution of such documents as are necessary to enable the Company to bring suit to enforce such rights.

(d) The Company's obligation to provide indemnification or advancement hereunder to Indemnitee who is or was serving at the request of the Company as a director, manager, partner, officer, employee, agent or trustee of any other Enterprise shall be reduced by any amount Indemnitee has actually received as indemnification or advancement from such other Enterprise.

Section 14. Duration of Agreement. This Agreement shall continue until and terminate upon the later of: (a) ten (10) years after the date that Indemnitee shall have ceased to serve as [both a director and] an officer of the Company or (b) one (1) year after the final termination of any Proceeding, including any appeal, then pending in respect of which Indemnitee is granted rights of indemnification or advancement hereunder and of any proceeding

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commenced by Indemnitee pursuant to Section 12 of this Agreement relating thereto. This Agreement shall be binding upon the Company and its successors and assigns and shall inure to the benefit of Indemnitee and his or her heirs, executors and administrators. The Company shall require and cause any successor (whether direct or indirect by purchase, merger, consolidation or otherwise) to all, substantially all or a substantial part, of the business and/or assets of the Company, by written agreement in form and substance satisfactory to Indemnitee, expressly to assume and agree to perform this Agreement to the fullest extent permitted by law.

Section 15. Severability. If any provision or provisions of this Agreement shall be held to be invalid, illegal or unenforceable for any reason whatsoever: (a) the validity, legality and enforceability of the remaining provisions of this Agreement (including, without limitation, each portion of any section of this Agreement containing any such provision held to be invalid, illegal or unenforceable, that is not itself invalid, illegal or unenforceable) shall not in any way be affected or impaired thereby and shall remain enforceable to the fullest extent permitted by law; (b) such provision or provisions shall be deemed reformed to the extent necessary to conform to applicable law and to give the maximum effect to the intent of the parties hereto; and (c) to the fullest extent possible, the provisions of this Agreement (including, without limitation, each portion of any section of this Agreement containing any such provision held to be invalid, illegal or unenforceable, that is not itself invalid, illegal or unenforceable) shall be construed so as to give effect to the intent manifested thereby.

Section 16. Enforcement.

(a) The Company expressly confirms and agrees that it has entered into this Agreement and assumed the obligations imposed on it hereby in order to induce Indemnitee to serve or continue to serve as [a director and] an officer of the Company, and the Company acknowledges that Indemnitee is relying upon this Agreement in serving as [a director and] an officer of the Company.

(b) This Agreement constitutes the entire agreement between the parties hereto with respect to the subject matter hereof and supersedes all prior agreements and understandings, oral, written and implied, between the parties hereto with respect to the subject matter hereof; provided, however, that this Agreement is a supplement to and in furtherance of the Charter, the Bylaws and applicable law, and shall not be deemed a substitute therefor, nor to diminish or abrogate any rights of Indemnitee thereunder.

Section 17. Modification and Waiver. No supplement, modification or amendment, or waiver of any provision, of this Agreement shall be binding unless executed in writing by the parties thereto. No waiver of any of the provisions of this Agreement shall be deemed or shall constitute a waiver of any other provisions of this Agreement nor shall any waiver constitute a continuing waiver. No supplement, modification or amendment of this Agreement or of any provision hereof shall limit or restrict any right of Indemnitee under this Agreement in respect of any action taken or omitted by such Indemnitee prior to such supplement, modification or amendment.

Section 18. Notice by Indemnitee. Indemnitee agrees promptly to notify the Company in writing upon being served with any summons, citation, subpoena, complaint, indictment,

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information or other document relating to any Proceeding or matter which may be subject to indemnification, reimbursement or advancement as provided hereunder. The failure of Indemnitee to so notify the Company shall not relieve the Company of any obligation which it may have to Indemnitee under this Agreement or otherwise.

Section 19. Notices. All notices, requests, demands and other communications under this Agreement shall be in writing and shall be deemed to have been duly given if (i) delivered by hand and receipted for by the party to whom said notice or other communication shall have been directed, (ii) mailed by certified or registered mail with postage prepaid, on the third business day after the date on which it is so mailed, (iii) mailed by reputable overnight courier and receipted for by the party to whom said notice or other communication shall have been directed or (iv) sent by facsimile transmission, with receipt of oral confirmation that such transmission has been received:

(a) If to Indemnitee, at such address as Indemnitee shall provide to the Company.

(b) If to the Company to:

Scholar Rock Holding Corporation
620 Memorial Drive, 2nd Floor
Cambridge, Massachusetts 02139
Attention: []

or to any other address as may have been furnished to Indemnitee by the Company.

Section 20. Contribution. To the fullest extent permissible under applicable law, if the indemnification provided for in this Agreement is unavailable to Indemnitee for any reason whatsoever, the Company, in lieu of indemnifying Indemnitee, shall contribute to the amount incurred by Indemnitee, whether for judgments, fines, penalties, excise taxes, amounts paid or to be paid in settlement and/or for Expenses, in connection with any Proceeding in such proportion as is deemed fair and reasonable in light of all of the circumstances in order to reflect (i) the relative direct benefits received by the Company and Indemnitee in connection with the event(s) and/or transaction(s) giving rise to such Proceeding; and/or (ii) the relative fault of the Company (and its directors, officers, employees and agents) and Indemnitee in connection with such event(s) and/or transactions. The Company hereby agrees to fully indemnify and hold harmless Indemnitee from any claims for contribution which may be brought by Officers, Directors or employees of the Company (other than Indemnitee) who may be jointly liable with Indemnitee.

Section 21. Internal Revenue Code Section 409A. The Company intends for this Agreement to comply with the Indemnification exception under Section 1.409A-1(b)(10) of the regulations promulgated under the Internal Revenue Code of 1986, as amended (the "Code"), which provides that indemnification of, or the purchase of an insurance policy providing for payments of, all or part of the expenses incurred or damages paid or payable by Indemnitee with respect to a bona fide claim against Indemnitee or the Company do not provide for a deferral of compensation, subject to Section 409A of the Code, where such claim is based on actions or failures to act by Indemnitee in his or her capacity as a service provider of the Company. The parties intend that this Agreement be interpreted and construed with such intent.

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Section 22. Applicable Law and Consent to Jurisdiction. This Agreement and the legal relations among the parties shall be governed by, and construed and enforced in accordance with, the laws of the State of Delaware, without regard to its conflict of laws rules. Except with respect to any arbitration commenced by Indemnitee pursuant to Section 12(a) of this Agreement, the Company and Indemnitee hereby irrevocably and unconditionally (i) agree that any action or proceeding arising out of or in connection with this Agreement shall be brought only in the Delaware Court, and not in any other state or federal court in the United States of America or any court in any other country, (ii) consent to submit to the exclusive jurisdiction of the Delaware Court for purposes of any action or proceeding arising out of or in connection with this Agreement, (iii) consent to service of process at the address set forth in Section 19 of this Agreement with the same legal force and validity as if served upon such party personally within the State of Delaware, (iv) waive any objection to the laying of venue of any such action or proceeding in the Delaware Court, and (v) waive, and agree not to plead or to make, any claim that any such action or proceeding brought in the Delaware Court has been brought in an improper or inconvenient forum.

Section 23. Determination of Good Faith/Safe Harbor. For purposes of any determination of good faith, Indemnitee [or "an independent director"] shall be presumed to have acted in good faith if Indemnitee's action is based on the records or books of account of the Enterprise, including financial statements, or on information supplied to Indemnitee by the officers of the Enterprise in the course of their duties, or on the advice of legal counsel for the Enterprise or the Board or counsel selected by any committee of the Board or on information or records given or reports made to the Enterprise by an independent certified public accountant or by an appraiser, investment banker, compensation consultant, or other expert selected with reasonable care by the Company or the Board or any committee of the Board. The provisions of this Section XXs shall not be deemed to be exclusive or to limit in any way the other circumstances in which the Indemnitee may be deemed to have met the applicable standard of conduct. Whether or not the foregoing provisions of this Section are satisfied, it shall in any event be presumed that Director has at all times acted in good faith and in a manner he or she reasonably believed to be in or not opposed to the best interests of the Company.

Section 24. Monetary Damages Insufficient/Specific Performance. The Company and Indemnitee agree that a monetary remedy for breach of this Agreement may be inadequate, impracticable and difficult of proof, and further agree that such breach may cause Indemnitee irreparable harm. Accordingly, the parties hereto agree that Indemnitee may enforce this Agreement by seeking injunctive relief and/or specific performance hereof, without any necessity of showing actual damage or irreparable harm (having agreed that actual and irreparable harm will result in not forcing the Company to specifically perform its obligations pursuant to this Agreement) and that by seeking injunctive relief and/or specific performance, Indemnitee shall not be precluded from seeking or obtaining any other relief to which he may be entitled. The Company and Indemnitee further agree that Indemnitee may be entitled to such specific performance and injunctive relief, including temporary restraining orders, preliminary injunctions and permanent injunctions, without the necessity of posting bonds or other undertaking in connection therewith. The Company acknowledges that in the absence of a waiver, a bond or undertaking may be required of Indemnitee by the Court, and the Company hereby waives any such requirement of a bond or undertaking.

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Section 25. Headings. The headings of the paragraphs of this Agreement are inserted for convenience only and shall not be deemed to constitute part of this Agreement or to affect the construction thereof.

Section 26. Identical Counterparts. This Agreement may be executed in one or more counterparts, each of which shall for all purposes be deemed to be an original but all of which together shall constitute one and the same Agreement. Only one such counterpart signed by the party against whom enforceability is sought needs to be produced to evidence the existence of this Agreement.

[Remainder of Page Intentionally Left Blank]

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IN WITNESS WHEREOF, the parties have caused this Agreement to be signed as of the day and year first above written.

SCHOLAR ROCK HOLDING CORPORATION

By: _____

Name: Nagesh K. Mahanthappa
Title: President and Chief Executive Officer

INDEMNITEE

Name:

SCHOLAR ROCK HOLDING CORPORATION

EMPLOYMENT AGREEMENT

This Employment Agreement ("Agreement") is made as of the 11th day of May, 2018, between Scholar Rock Holding Corporation, a Delaware corporation (the "Company"), and Nagesh K. Mahanthappa (the "Employee") and is effective as of the closing of the Company's first underwritten public offering of its equity securities pursuant to an effective registration statement under the Securities Act of 1933, as amended (the "Effective Date").

WHEREAS, the Company and the Employee are parties to an offer letter, dated October 10, 2012, (the "Prior Agreement"); and

WHEREAS, the parties intend to replace the Prior Agreement with this Agreement, effective as of the Effective Date.

NOW, THEREFORE, in consideration of the mutual covenants and agreements herein contained and other good and valuable consideration, the receipt and sufficiency of which is hereby acknowledged, the parties agree as follows:

1. Employment.

(a) Term. The term of this Agreement shall commence on the Effective Date and continue until terminated in accordance with the provisions hereof (the "Term"). The Employee's employment with the Company will continue to be "at will," meaning that the Employee's employment may be terminated by the Company or the Employee at any time and for any reason subject to the terms of this Agreement.

(b) Position and Duties. During the Term, the Employee shall serve as the President and Chief Executive Officer of the Company, and shall have such duties and authorities as may from time to time be prescribed by the Board of Directors of the Company (the "Board"). During the Term, the Employee shall serve as a member of the Board. The Employee shall devote his full working time and efforts to the business and affairs of the Company. Notwithstanding the foregoing, the Employee may serve on other boards of directors, with the approval of the Board, or engage in religious, charitable or other community activities as long as such services and activities do not materially interfere with the Executive's performance of his duties to the Company as provided in this Agreement.

2. Compensation and Related Matters.

(a) Base Salary. During the Term, the Employee's annual base salary shall be \$475,000. The Employee's base salary shall be reviewed annually by the Compensation Committee of the Board (the "Compensation Committee"). The base salary in effect at any given time is referred to herein as "Base Salary." The Base Salary shall be payable in a manner that is consistent with the Company's usual payroll practices.

(b) Incentive Compensation. During the Term, the Employee shall be eligible to receive cash incentive compensation as determined by the Board or the Compensation Committee from time to time. The Employee's initial target annual incentive compensation shall be fifty percent (50%) of his Base Salary (the "Target Annual Incentive Compensation"). Except as otherwise provided herein, to earn incentive compensation, the Employee must be employed by the Company on the day such incentive compensation is paid.

(c) Expenses. The Employee shall be entitled to receive prompt reimbursement for all reasonable expenses incurred by him during the Term in performing services hereunder, in accordance with the policies and procedures then in effect and established by the Company.

(d) Other Benefits. During the Term, the Employee shall be eligible to participate in or receive benefits under the Company's employee benefit plans in effect from time to time, subject to the terms of such plans.

(e) Vacations. During the Term, the Employee shall be entitled to paid vacation in accordance with the Company's policies and procedures. The Employee shall also be entitled to all paid holidays given by the Company in accordance with the policies and procedures then in effect and established by the Company.

3. Termination. During the Term, the Employee's employment hereunder may be terminated without any breach of this Agreement under the following circumstances:

(a) Death. The Employee's employment hereunder shall terminate upon his death.

(b) Termination by Company for Cause. The Company may terminate the Employee's employment hereunder for Cause. For purposes of this Agreement, "Cause" shall mean: (i) conduct by the Employee constituting a material act of misconduct in connection with the performance of his duties, including, without limitation, misappropriation of funds or property of the Company or any of its subsidiaries or affiliates other than the occasional, customary and de minimis use of Company property for personal purposes; (ii) the commission by the Employee of any felony or a misdemeanor involving moral turpitude, deceit, dishonesty or fraud, or any conduct by the Employee that would reasonably be expected to result in material injury or reputational harm to the Company or any of its subsidiaries or affiliates if he were retained in his position; (iii) continued non-performance by the Employee of his duties hereunder (other than by reason of the Employee's physical or mental illness, incapacity or disability) which has continued for more than 30 days following written notice of such non-performance from the Board; (iv) a breach by the Employee of any of the provisions contained in Section 7 of this Agreement; (v) a material violation by the Employee of the Company's written employment policies; or (vi) failure to cooperate with a bona fide internal investigation or an investigation by regulatory or law enforcement authorities, after being instructed by the Company to cooperate, or the willful destruction or failure to preserve documents or other materials known to be relevant to such investigation or the inducement of others to fail to cooperate or to produce documents or other materials in connection with such investigation.

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(c) Termination Without Cause. The Company may terminate the Employee's employment hereunder at any time without Cause. Any termination by the Company of the Employee's employment under this Agreement which does not constitute a termination for Cause under Section 3(b) and does not result from the death of the Employee under Section 3(a) shall be deemed a termination without Cause.

(d) Termination by the Employee. The Employee may terminate his employment hereunder at any time for any reason, including but not limited to Good Reason. For purposes of this Agreement, "Good Reason" shall mean that the Employee has complied with the "Good Reason Process" (hereinafter defined) following the occurrence of any of the following events: (i) a material diminution in the Employee's responsibilities, authority or duties; (ii) a material diminution in the Employee's Base Salary except for across-the-board salary reductions based on the Company's financial performance similarly affecting all or substantially all senior management employees of the Company; (iii) a material change in the geographic location at which the Employee provides services to the Company, except for required travel for the Company's business; or (iv) the material breach of this Agreement by the Company. "Good Reason Process" shall mean that (i) the Employee reasonably determines in good faith that a "Good Reason" condition has occurred; (ii) the Employee notifies the Company in writing of the first occurrence of the Good Reason condition within 60 days of the first occurrence of such condition; (iii) the Employee cooperates in good faith with the Company's efforts, for a period not less than 30 days following such notice (the "Cure Period"), to remedy the condition; (iv) notwithstanding such efforts, the Good Reason condition continues to exist; and (v) the Employee terminates his employment within 60 days after the end of the Cure Period. If the Company cures the Good Reason condition during the Cure Period, Good Reason shall be deemed not to have occurred.

(e) Notice of Termination. Except for termination as specified in Section 3(a), any termination of the Employee's employment by the Company or any such termination by the Employee shall be communicated by written Notice of Termination to the other party hereto. For purposes of this Agreement, a "Notice of Termination" shall mean a notice which shall indicate the specific termination provision in this Agreement relied upon.

(f) Date of Termination. "Date of Termination" shall mean: (i) if the Employee's employment is terminated by his death, the date of his death; (ii) if the Employee's employment is terminated by the Company under Section 3(c), the date on which a Notice of Termination is given; (iii) if the Employee's employment is terminated by the Employee under Section 3(d) without Good Reason, 30 days after the date on which a Notice of Termination is given, and (iv) if the Employee's employment is terminated by the Employee under Section 3(d) with Good Reason, the date on which a Notice of Termination is given after the end of the Cure Period. Notwithstanding the foregoing, in the event that the Employee gives a Notice of Termination to the Company, the Company may unilaterally accelerate the Date of Termination and such acceleration shall not result in a termination by the Company for purposes of this Agreement.

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4. Compensation Upon Termination.

(a) Termination Generally. If the Employee's employment with the Company is terminated for any reason, the Company shall pay or provide to the Employee (or to his authorized representative or estate) (i) any Base Salary earned through the Date of Termination, unpaid expense reimbursements (subject to, and in accordance with, Section 2(c) of this Agreement) and unused vacation that accrued through the Date of Termination on or before the time required by law but in no event more than 30 days after the Employee's Date of Termination; and (ii) any vested benefits the Employee may have under any employee benefit plan of the Company through the Date of Termination, which vested benefits shall be paid and/or provided in accordance with the terms of such employee benefit plans (collectively, the "Accrued Benefit").

(b) Termination by the Company Without Cause or by the Employee with Good Reason. During the Term, if the Employee's employment is terminated by the Company without Cause as provided in Section 3(c), or the Employee terminates his employment for Good Reason as provided in Section 3(d), then the Company shall pay the Employee his Accrued Benefit. In addition, subject to the Employee signing a separation agreement containing, among other provisions, a general release of claims in favor of the Company and related persons and entities, confidentiality, return of property and non-disparagement, in a form and manner satisfactory to the Company (the "Separation Agreement and Release") and the Separation Agreement and Release becoming irrevocable and fully effective, all within 60 days after the Date of Termination (or such shorter time period provided in the Separation Agreement and Release):

(i) the Company shall pay the Employee an amount equal to 12 months of the Employee's Base Salary (the "Severance Amount"). Notwithstanding the foregoing, if the Employee breaches any of the provisions contained in Section 7 of this Agreement, all payments of the Severance Amount shall immediately cease;

(ii) RESERVED

(iii) if the Employee was participating in the Company's group health plan immediately prior to the Date of Termination and elects COBRA health continuation, then the Company shall pay to the Employee a monthly cash payment for 12 months or the Employee's COBRA health continuation period, whichever ends earlier, in an amount equal to the monthly employer contribution that the Company would have made to provide health insurance to the Employee if the Employee had remained employed by the Company; and

(iv) the amounts payable under Section 4(b)(i) and (ii) shall be paid out in substantially equal installments in accordance with the Company's payroll practice commencing within 60 days after the Date of Termination; provided, however, that if the 60-day period begins in one calendar year and ends in a second calendar year, the Severance Amount shall begin to be paid in the second calendar year by the last day of such 60-day period; provided, further, that the initial payment shall include a catch-up payment to cover amounts retroactive to the day immediately following the Date of

Termination. Each payment pursuant to this Agreement is intended to constitute a separate payment for purposes of Treasury Regulation Section 1.409A-2(b)(2).

5. **Change in Control Payment.** The provisions of this Section 5 set forth certain terms of an agreement reached between the Employee and the Company regarding the Employee's rights and obligations upon the occurrence of a Change in Control of the Company. These provisions are intended to assure and encourage in advance the Employee's continued attention and dedication to his assigned duties and his objectivity during the pendency and after the occurrence of any such event. These provisions shall apply in lieu of, and expressly supersede, the provisions of Section 4(b) regarding severance pay and benefits upon a termination of employment, if such termination of employment occurs within 18 months after the occurrence of the first event constituting a Change in Control. These provisions shall terminate and be of no further force or effect beginning 18 months after the occurrence of a Change in Control.

(a) **Change in Control.** During the Term, if within 18 months after a Change in Control, the Employee's employment is terminated by the Company without Cause as provided in Section 3(c) or the Employee terminates his employment for Good Reason as provided in Section 3(d), then, subject to the signing of the Separation Agreement and Release by the Employee and the Separation Agreement and Release becoming irrevocable and fully effective, all within 60 days after the Date of Termination (or such shorter time period provided in the Separation Agreement and Release):

(i) the Company shall pay the Employee a lump sum in cash in an amount equal to 1.5 times the sum of (A) the Employee's current Base Salary (or the Employee's Base Salary in effect immediately prior to the Change in Control, if higher) plus (B) the Employee's Average Incentive Compensation (For purposes of this Agreement, "Average Incentive Compensation" shall mean the Target Annual Incentive Compensation the Employee would have been entitled to receive in the fiscal year of termination (or the Employee's Target Annual Incentive Compensation in the fiscal year immediately prior to the Change in Control, if higher). In no event shall "Average Incentive Compensation" include any sign-on bonus, retention bonus or any other special bonus.);

(ii) notwithstanding anything to the contrary in any applicable option agreement or stock-based award agreement, all time-based stock options and other time-based stock-based awards held by the Employee shall immediately accelerate and become fully exercisable or nonforfeitable as of the Date of Termination;

(iii) if the Employee was participating in the Company's group health plan immediately prior to the Date of Termination and elects COBRA health continuation, then the Company shall pay to the Employee a monthly cash payment for 18 months or the Employee's COBRA health continuation period, whichever ends earlier, in an amount equal to the monthly employer contribution that the Company would have made to provide health insurance to the Employee if the Employee had remained employed by the Company; and

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(iv) The amounts payable under Section 5(a)(i) and (iii) shall be paid or commence to be paid within 60 days after the Date of Termination; provided, however, that if the 60-day period begins in one calendar year and ends in a second calendar year, such payment shall be paid or commence to be paid in the second calendar year by the last day of such 60-day period.

(b) **Additional Limitation.**

(i) Anything in this Agreement to the contrary notwithstanding, in the event that the amount of any compensation, payment or distribution by the Company to or for the benefit of the Employee, whether paid or payable or distributed or distributable pursuant to the terms of this Agreement or otherwise, calculated in a manner consistent with Section 280G of the Internal Revenue Code of 1986, as amended (the "Code") and the applicable regulations thereunder (the "Aggregate Payments"), would be subject to the excise tax imposed by Section 4999 of the Code, then the Aggregate Payments shall be reduced (but not below zero) so that the sum of all of the Aggregate Payments shall be \$1.00 less than the amount at which the Employee becomes subject to the excise tax imposed by Section 4999 of the Code; provided that such reduction shall only occur if it would result in the Employee receiving a higher After Tax Amount (as defined below) than the Employee would receive if the Aggregate Payments were not subject to such reduction. In such event, the Aggregate Payments shall be reduced in the following order, in each case, in reverse chronological order beginning with the Aggregate Payments that are to be paid the furthest in time from consummation of the transaction that is subject to Section 280G of the Code: (1) cash payments not subject to Section 409A of the Code; (2) cash payments subject to Section 409A of the Code; (3) equity-based payments and acceleration; and (4) non-cash forms of benefits; provided that in the case of all the foregoing Aggregate Payments all amounts or payments that are not subject to calculation under Treas. Reg. §1.280G-1, Q&A-24(b) or (c) shall be reduced before any amounts that are subject to calculation under Treas. Reg. §1.280G-1, Q&A-24(b) or (c).

(ii) For purposes of this Section 5(b), the "After Tax Amount" means the amount of the Aggregate Payments less all federal, state, and local income, excise and employment taxes imposed on the Employee as a result of the Employee's receipt of the Aggregate Payments. For purposes of determining the After Tax Amount, the Employee shall be deemed to pay federal income taxes at the highest marginal rate of federal income taxation applicable to individuals for the calendar year in which the determination is to be made, and state and local income taxes at the highest marginal rates of individual taxation in each applicable state and locality, net of the maximum reduction in federal income taxes which could be obtained from deduction of such state and local taxes.

(iii) The determination as to whether a reduction in the Aggregate Payments shall be made pursuant to Section 5(b)(i) shall be made by a nationally recognized accounting firm selected by the Company (the "Accounting Firm"), which shall provide detailed supporting calculations both to the Company and the Employee within 15 business days of the Date of Termination, if applicable, or at such earlier time

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as is reasonably requested by the Company or the Employee. Any determination by the Accounting Firm shall be binding upon the Company and the Employee.

(c) **Definitions.** For purposes of this Section 5, the following terms shall have the following meanings:

"Change in Control" shall mean any of the following:

(i) any "person," as such term is used in Sections 13(d) and 14(d) of the Securities Exchange Act of 1934, as amended (the "Act") (other than the Company, any of its subsidiaries, or any trustee, fiduciary or other person or entity holding securities under any employee benefit plan or trust of the Company or any of its subsidiaries), together with all "affiliates" and "associates" (as such terms are defined in Rule 12b-2 under the Act) of such person, shall become the "beneficial owner" (as such term is defined in Rule 13d-3 under the Act), directly or indirectly, of securities of the Company representing 50 percent or more of the combined voting power of the Company's then outstanding securities having the right to vote in an election of the Board ("Voting Securities") (in such case other than as a result of an acquisition of securities directly from the Company); or

(ii) the date a majority of the members of the Board is replaced during any 12-month period by directors whose appointment or election is not endorsed by a majority of the members of the Board before the date of the appointment or election; or

(iii) the consummation of (A) any consolidation or merger of the Company where the stockholders of the Company, immediately prior to the consolidation or merger, would not, immediately after the consolidation or merger, beneficially own (as such term is defined in Rule 13d-3 under the Act), directly or indirectly, shares representing in the aggregate more than 50 percent of the voting shares of the Company issuing cash or securities in the consolidation or merger (or of its ultimate parent corporation, if any), or (B) any sale or other transfer (in one transaction or a series of transactions contemplated or arranged by any party as a single plan) of all or substantially all of the assets of the Company and its affiliates on a consolidated basis.

Notwithstanding the foregoing, a "Change in Control" shall not be deemed to have occurred for purposes of the foregoing clause (i) solely as the result of an acquisition of securities by the Company which, by reducing the number of shares of Voting Securities outstanding, increases the proportionate number of Voting Securities beneficially owned by any person to 50 percent or more of the combined voting power of all of the then outstanding Voting Securities; provided, however, that if any person referred to in this sentence shall thereafter become the beneficial owner of any additional shares of Voting Securities (other than pursuant to a stock split, stock dividend, or similar transaction or as a result of an acquisition of securities directly from the Company) and immediately thereafter beneficially owns 50 percent or more of the combined voting power of all of the then outstanding Voting Securities, then a "Change in Control" shall be deemed to have occurred for purposes of the foregoing clause (i).

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6. **Section 409A.**

(a) Anything in this Agreement to the contrary notwithstanding, if at the time of the Employee's separation from service within the meaning of Section 409A of the Code, the Company determines that the Employee is a "specified employee" within the meaning of Section 409A(a)(2)(B)(i) of the Code, then to the extent any payment or benefit that the Employee becomes entitled to under this Agreement on account of the Employee's separation from service would be considered deferred compensation otherwise subject to the 20 percent additional tax imposed pursuant to Section 409A(a) of the Code as a result of the application of Section 409A(a)(2)(B)(i) of the Code, such payment shall not be payable and such benefit shall not be provided until the date that is the earlier of (A) six months and one day after the Employee's separation from service, or (B) the Employee's death. If any such delayed cash payment is otherwise payable on an installment basis, the first payment shall include a catch-up payment covering amounts that would otherwise have been paid during the six-month period but for the application of this provision, and the balance of the installments shall be payable in accordance with their original schedule.

(b) All in-kind benefits provided and expenses eligible for reimbursement under this Agreement shall be provided by the Company or incurred by the Employee during the time periods set forth in this Agreement. All reimbursements shall be paid as soon as administratively practicable, but in no event shall any reimbursement be paid after the last day of the taxable year following the taxable year in which the expense was incurred. The amount of in-kind benefits provided or reimbursable expenses incurred in one taxable year shall not affect the in-kind benefits to be provided or the expenses eligible for reimbursement in any other taxable year (except for any lifetime or other aggregate limitation applicable to medical expenses). Such right to reimbursement or in-kind benefits is not subject to liquidation or exchange for another benefit.

(c) To the extent that any payment or benefit described in this Agreement constitutes "non-qualified deferred compensation" under Section 409A of the Code, and to the extent that such payment or benefit is payable upon the Employee's termination of employment, then such payments or benefits shall be payable only upon the Employee's "separation from service." The determination of whether and when a separation from service has occurred shall be made in accordance with the presumptions set forth in Treasury Regulation Section 1.409A-1(h).

(d) The parties intend that this Agreement will be administered in accordance with Section 409A of the Code. To the extent that any provision of this Agreement is ambiguous as to its compliance with Section 409A of the Code, the provision shall be read in such a manner so that all payments hereunder comply with Section 409A of the Code. Each payment pursuant to this Agreement is intended to constitute a separate payment for purposes of Treasury Regulation Section 1.409A-2(b)(2). The parties agree that this Agreement may be amended, as reasonably requested by either party, and as may be necessary to fully comply with Section 409A of the Code and all related rules and regulations in order to preserve the payments and benefits provided hereunder without additional cost to either party.

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(e) The Company makes no representation or warranty and shall have no liability to the Employee or any other person if any provisions of this Agreement are determined to constitute deferred compensation subject to Section 409A of the Code but do not satisfy an exemption from, or the conditions of, such Section.

7. **Confidential Information, Noncompetition and Cooperation.** The terms of the Employee Non-Competition, Non-Solicitation, Confidentiality and Assignment Agreement (the "Restrictive Covenant Agreement"), between the Company and the Employee, attached hereto as Exhibit A, shall continue to be in full force and effect and are incorporated by reference in this Agreement. The Employee hereby reaffirms the terms of the Restrictive Covenant Agreement as material terms of this Agreement.

(a) **Litigation and Regulatory Cooperation.** During and after the Employee's employment, the Employee shall cooperate fully with the Company in the defense or prosecution of any claims or actions now in existence or which may be brought in the future against or on behalf of the Company which relate to events or occurrences that transpired while the Employee was employed by the Company. The Employee's full cooperation in connection with such claims or actions shall include, but not be limited to, being available to meet with counsel to prepare for discovery or trial and to act as a witness on behalf of the Company at mutually convenient times. During and after the Employee's employment, the Employee also shall cooperate fully with the Company in connection with any investigation or review of any federal, state or local regulatory authority as any such investigation or review relates to events or occurrences that transpired while the Employee was employed by the Company. The Company shall reimburse the Employee for any reasonable out-of-pocket expenses incurred in connection with the Employee's performance of obligations pursuant to this Section 7(a).

(b) **Relief.** The Employee agrees that it would be difficult to measure any damages caused to the Company which might result from any breach by the Employee of the promises set forth in this Section 7, and that in any event money damages would be an inadequate remedy for any such breach. Accordingly, subject to Section 8 of this Agreement, the Employee agrees that if the Employee breaches, or proposes to breach, any portion of this Agreement, the Company shall be entitled, in addition to all other remedies that it may have, to an injunction or other appropriate equitable relief to restrain any such breach without showing or proving any actual damage to the Company. In addition, in the event the Employee breaches this Section 7 during a period when he is receiving severance payments pursuant to Section 4 or Section 5 hereof, the Company shall have the right to suspend or terminate such severance payments. Such suspension or termination shall not limit the Company's other options with respect to relief for such breach and shall not relieve the Employee of his duties under this Agreement.

(c) **Protected Disclosures and Other Protected Action.** Nothing contained in this Agreement limits the Employee's ability to communicate with any federal, state or local governmental agency or commission, including to provide documents or other information, without notice to the Company.

8. **Arbitration of Disputes.** Any controversy or claim arising out of or relating to this Agreement or the breach thereof or otherwise arising out of the Employee's employment or the

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termination of that employment (including, without limitation, any claims of unlawful employment discrimination whether based on age or otherwise) shall, to the fullest extent permitted by law, be settled by arbitration in any forum and form agreed upon by the parties or, in the absence of such an agreement, under the auspices of the American Arbitration Association ("AAA") in Boston, Massachusetts in accordance with the Employment Dispute Resolution Rules of the AAA, including, but not limited to, the rules and procedures applicable to the selection of arbitrators. In the event that any person or entity other than the Employee or the Company may be a party with regard to any such controversy or claim, such controversy or claim shall be submitted to arbitration subject to such other person or entity's agreement. Judgment upon the award rendered by the arbitrator may be entered in any court having jurisdiction thereof. This Section 8 shall be specifically enforceable. Notwithstanding the foregoing, this Section 8 shall not preclude either party from pursuing a court action for the sole purpose of obtaining a temporary restraining order or a preliminary injunction in circumstances in which such relief is appropriate; provided that any other relief shall be pursued through an arbitration proceeding pursuant to this Section 8.

9. **Consent to Jurisdiction.** To the extent that any court action is permitted consistent with or to enforce Section 8 of this Agreement, the parties hereby consent to the jurisdiction of the Superior Court of the Commonwealth of Massachusetts and the United States District Court for the District of Massachusetts. Accordingly, with respect to any such court action, the Employee (a) submits to the personal jurisdiction of such courts; (b) consents to service of process; and (c) waives any other requirement (whether imposed by statute, rule of court, or otherwise) with respect to personal jurisdiction or service of process.

10. **Integration.** This Agreement constitutes the entire agreement between the parties with respect to the subject matter hereof and supersedes all prior agreements between the parties concerning such subject matter, including the Prior Agreement.

11. **Withholding.** All payments made by the Company to the Employee under this Agreement shall be net of any tax or other amounts required to be withheld by the Company under applicable law.

12. **Successor to the Employee.** This Agreement shall inure to the benefit of and be enforceable by the Employee's personal representatives, executors, administrators, heirs, distributees, devisees and legatees. In the event of the Employee's death after his termination of employment but prior to the completion by the Company of all payments due to him under this Agreement, the Company shall continue such payments to the Employee's beneficiary designated in writing to the Company prior to his death (or to his estate, if the Employee fails to make such designation).

13. **Enforceability.** If any portion or provision of this Agreement (including, without limitation, any portion or provision of any section of this Agreement) shall to any extent be declared illegal or unenforceable by a court of competent jurisdiction, then the remainder of this Agreement, or the application of such portion or provision in circumstances other than those as to which it is so declared illegal or unenforceable, shall not be affected thereby, and each portion and provision of this Agreement shall be valid and enforceable to the fullest extent permitted by law.

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14. **Survival.** The provisions of this Agreement shall survive the termination of this Agreement and/or the termination of the Employee's employment to the extent necessary to effectuate the terms contained herein.

15. **Waiver.** No waiver of any provision hereof shall be effective unless made in writing and signed by the waiving party. The failure of any party to require the performance of any term or obligation of this Agreement, or the waiver by any party of any breach of this Agreement, shall not prevent any subsequent enforcement of such term or obligation or be deemed a waiver of any subsequent breach.

16. **Notices.** Any notices, requests, demands and other communications provided for by this Agreement shall be sufficient if in writing and delivered in person or sent by a nationally recognized overnight courier service or by registered or certified mail, postage prepaid, return receipt requested, to the Employee at the last address the Employee has filed in writing with the Company or, in the case of the Company, at its main offices, attention of the Board.

17. **Amendment.** This Agreement may be amended or modified only by a written instrument signed by the Employee and by a duly authorized representative of the Company.

18. **Governing Law.** This is a Massachusetts contract and shall be construed under and be governed in all respects by the laws of the Commonwealth of Massachusetts without giving effect to the conflict of laws principles thereof.

19. **Counterparts.** This Agreement may be executed in any number of counterparts, each of which when so executed and delivered shall be taken to be an original; but such counterparts shall together constitute one and the same document.

20. **Successor to Company.** The Company shall require any successor (whether direct or indirect, by purchase, merger, consolidation or otherwise) to all or substantially all of the business or assets of the Company expressly to assume and agree to perform this Agreement to the same extent that the Company would be required to perform it if no succession had taken place. Failure of the Company to obtain an assumption of this Agreement at or prior to the effectiveness of any succession shall be a material breach of this Agreement.

21. **Gender Neutral.** Wherever used herein, a pronoun in the masculine gender shall be considered as including the feminine gender unless the context clearly indicates otherwise.

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IN WITNESS WHEREOF, the parties have executed this Agreement effective on the date and year first above written.

SCHOLAR ROCK HOLDING CORPORATION

/s/ Rhonda M. Chicko
By: Rhonda M. Chicko
Its: Chief Financial Officer

EMPLOYEE

/s/ Nagesh K. Mahanthappa
Nagesh K. Mahanthappa, Ph.D.

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SCHOLAR ROCK HOLDING CORPORATION

EMPLOYMENT AGREEMENT

This Employment Agreement ("Agreement") is made as of the 13th day of May, 2018, between Scholar Rock Holding Corporation, a Delaware corporation (the "Company"), and Rhonda M. Chicko (the "Employee") and is effective as of the closing of the Company's first underwritten public offering of its equity securities pursuant to an effective registration statement under the Securities Act of 1933, as amended (the "Effective Date").

WHEREAS, the Company and the Employee are parties to an offer letter, dated March 9, 2018, (the "Prior Agreement");

WHEREAS, the parties intend to replace the Prior Agreement with this Agreement, effective as of the Effective Date; and

WHEREAS, the Employee received a stock option to purchase 575,000 shares of the common stock of the Company, as approved by the Company's Board of Directors on April 3, 2018 (and subject to adjustment based on the reverse stock split effectuated by the Company on May 11, 2018) (the "Initial Hire Stock Option").

NOW, THEREFORE, in consideration of the mutual covenants and agreements herein contained and other good and valuable consideration, the receipt and sufficiency of which is hereby acknowledged, the parties agree as follows:

1. Employment.

(a) Term. The term of this Agreement shall commence on the Effective Date and continue until terminated in accordance with the provisions hereof (the "Term"). The Employee's employment with the Company will continue to be "at will," meaning that the Employee's employment may be terminated by the Company or the Employee at any time and for any reason subject to the terms of this Agreement.

(b) Position and Duties. During the Term, the Employee shall serve as the Chief Financial Officer of the Company, and shall have such duties and authorities as may from time to time be prescribed by the Chief Executive Officer of the Company (the "CEO"). The Employee shall devote her full working time and efforts to the business and affairs of the Company, provided that she may consult for her prior employer on terms that are acceptable to the Company for up to ten hours per week not to exceed a date agreed to by the CEO. Notwithstanding the foregoing, the Employee may serve on other boards of directors, with the approval of the CEO, or engage in religious, charitable or other community activities as long as such services and activities do not materially interfere with the Executive's performance of her duties to the Company as provided in this Agreement.

2. Compensation and Related Matters.

(a) Base Salary. During the Term, the Employee's annual base salary shall be \$363,000. The Employee's base salary shall be reviewed annually by the Compensation Committee of the Board (the "Compensation Committee") or the CEO. The base salary in effect

at any given time is referred to herein as "Base Salary." The Base Salary shall be payable in a manner that is consistent with the Company's usual payroll practices.

(b) Incentive Compensation. During the Term, the Employee shall be eligible to receive cash incentive compensation as determined by the Board or the Compensation Committee from time to time. The Employee's initial target annual incentive compensation shall be thirty-five percent (35%) of her Base Salary (the "Target Annual Incentive Compensation"). Except as otherwise provided herein, to earn incentive compensation, the Employee must be employed by the Company on the day such incentive compensation is paid. Under the Prior Agreement, the Employee received a one-time payment of \$50,000 upon joining the Company. If the Employee leaves the Company within 18 months of April 3, 2018, the Employee will be required to repay the Company the full amount of this one-time payment. All payments will be subject to legally required tax withholdings.

(c) Expenses. The Employee shall be entitled to receive prompt reimbursement for all reasonable expenses incurred by her during the Term in performing services hereunder, in accordance with the policies and procedures then in effect and established by the Company.

(d) Other Benefits. During the Term, the Employee shall be eligible to participate in or receive benefits under the Company's employee benefit plans in effect from time to time, subject to the terms of such plans.

(e) Vacations. During the Term, the Employee shall be entitled to paid vacation in accordance with the Company's policies and procedures. The Employee shall also be entitled to all paid holidays given by the Company in accordance with the policies and procedures then in effect and established by the Company.

3. Termination. During the Term, the Employee's employment hereunder may be terminated without any breach of this Agreement under the following circumstances:

(a) Death. The Employee's employment hereunder shall terminate upon her death.

(b) Termination by Company for Cause. The Company may terminate the Employee's employment hereunder for Cause. For purposes of this Agreement, "Cause" shall mean: (i) conduct by the Employee constituting a material act of misconduct in connection with the performance of her duties, including, without limitation, misappropriation of funds or property of the Company or any of its subsidiaries or affiliates other than the occasional, customary and de minimis use of Company property for personal purposes; (ii) the commission by the Employee of any felony or a misdemeanor involving moral turpitude, deceit, dishonesty or fraud, or any conduct by the Employee that would reasonably be expected to result in material injury or reputational harm to the Company or any of its subsidiaries or affiliates if she were retained in her position; (iii) continued non-performance by the Employee of her duties hereunder (other than by reason of the Employee's physical or mental illness, incapacity or disability) which has continued for more than 30 days following written notice of such non-performance from the CEO; (iv) a breach by the Employee of any of the provisions contained in

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Section 7 of this Agreement; (v) a material violation by the Employee of the Company's written employment policies; or (vi) failure to cooperate with a bona fide internal investigation or an investigation by regulatory or law enforcement authorities, after being instructed by the Company to cooperate, or the willful destruction or failure to preserve documents or other materials known to be relevant to such investigation or the inducement of others to fail to cooperate or to produce documents or other materials in connection with such investigation.

(c) Termination Without Cause. The Company may terminate the Employee's employment hereunder at any time without Cause. Any termination by the Company of the Employee's employment under this Agreement which does not constitute a termination for Cause under Section 3(b) and does not result from the death of the Employee under Section 3(a) shall be deemed a termination without Cause.

(d) Termination by the Employee. The Employee may terminate her employment hereunder at any time for any reason, including but not limited to Good Reason. For purposes of this Agreement, "Good Reason" shall mean that the Employee has complied with the "Good Reason Process" (hereinafter defined) following the occurrence of any of the following events: (i) a material diminution in the Employee's responsibilities, authority or duties; (ii) a material diminution in the Employee's Base Salary except for across-the-board salary reductions based on the Company's financial performance similarly affecting all or substantially all senior management employees of the Company; (iii) a material change in the geographic location at which the Employee provides services to the Company, except for required travel for the Company's business; or (iv) the material breach of this Agreement by the Company. "Good Reason Process" shall mean that (i) the Employee reasonably determines in good faith that a "Good Reason" condition has occurred; (ii) the Employee notifies the Company in writing of the first occurrence of the Good Reason condition within 60 days of the first occurrence of such condition; (iii) the Employee cooperates in good faith with the Company's efforts, for a period not less than 30 days following such notice (the "Cure Period"), to remedy the condition; (iv) notwithstanding such efforts, the Good Reason condition continues to exist; and (v) the Employee terminates her employment within 60 days after the end of the Cure Period. If the Company cures the Good Reason condition during the Cure Period, Good Reason shall be deemed not to have occurred.

(e) Notice of Termination. Except for termination as specified in Section 3(a), any termination of the Employee's employment by the Company or any such termination by the Employee shall be communicated by written Notice of Termination to the other party hereto. For purposes of this Agreement, a "Notice of Termination" shall mean a notice which shall indicate the specific termination provision in this Agreement relied upon.

(f) Date of Termination. "Date of Termination" shall mean: (i) if the Employee's employment is terminated by her death, the date of her death; (ii) if the Employee's employment is terminated by the Company under Section 3(c), the date on which a Notice of Termination is given; (iii) if the Employee's employment is terminated by the Employee under Section 3(d) without Good Reason, 30 days after the date on which a Notice of Termination is given, and (iv) if the Employee's employment is terminated by the Employee under Section 3(d) with Good Reason, the date on which a Notice of Termination is given after the end of the Cure Period. Notwithstanding the foregoing, in the event that the Employee gives a Notice of

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Termination to the Company, the Company may unilaterally accelerate the Date of Termination and such acceleration shall not result in a termination by the Company for purposes of this Agreement.

4. Compensation Upon Termination.

(a) Termination Generally. If the Employee's employment with the Company is terminated for any reason, the Company shall pay or provide to the Employee (or to her authorized representative or estate) (i) any Base Salary earned through the Date of Termination, unpaid expense reimbursements (subject to, and in accordance with, Section 2(c) of this Agreement) and unused vacation that accrued through the Date of Termination on or before the time required by law but in no event more than 30 days after the Employee's Date of Termination; and (ii) any vested benefits the Employee may have under any employee benefit plan of the Company through the Date of Termination, which vested benefits shall be paid and/or provided in accordance with the terms of such employee benefit plans (collectively, the "Accrued Benefit").

(b) Termination by the Company Without Cause or by the Employee with Good Reason. During the Term, if the Employee's employment is terminated by the Company without Cause as provided in Section 3(c), or the Employee terminates her employment for Good Reason as provided in Section 3(d), then the Company shall pay the Employee her Accrued Benefit. In addition, subject to the Employee signing a separation agreement containing, among other provisions, a general release of claims in favor of the Company and related persons and entities, confidentiality, return of property and non-disparagement, in a form and manner satisfactory to the Company (the "Separation Agreement and Release") and the Separation Agreement and Release becoming irrevocable and fully effective, all within 60 days after the Date of Termination (or such shorter time period provided in the Separation Agreement and Release):

(i) the Company shall pay the Employee an amount equal to 9 months of the Employee's Base Salary (the "Severance Amount"). Notwithstanding the foregoing, if the Employee breaches any of the provisions contained in Section 7 of this Agreement, all payments of the Severance Amount shall immediately cease;

(ii) any then unvested portion of the Initial Hire Stock Option (as described in the recitals of this Agreement) issued to the Employee shall immediately accelerate and become fully exercisable or nonforfeitable as of the Date of Termination;

(iii) if the Employee was participating in the Company's group health plan immediately prior to the Date of Termination and elects COBRA health continuation, then the Company shall pay to the Employee a monthly cash payment for 9 months or the Employee's COBRA health continuation period, whichever ends earlier, in an amount equal to the monthly employer contribution that the Company would have made to provide health insurance to the Employee if the Employee had remained employed by the Company; and

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(iv) the amounts payable under Section 4(b)(i) and (iii) shall be paid out in substantially equal installments in accordance with the Company's payroll practice commencing within 60 days after the Date of Termination; provided, however, that if the 60-day period begins in one calendar year and ends in a second calendar year, the Severance Amount shall begin to be paid in the second calendar year by the last day of such 60-day period; provided, further, that the initial payment shall include a catch-up payment to cover amounts retroactive to the day immediately following the Date of Termination. Each payment pursuant to this Agreement is intended to constitute a separate payment for purposes of Treasury Regulation Section 1.409A-2(b)(2).

5. **Change in Control Payment.** The provisions of this Section 5 set forth certain terms of an agreement reached between the Employee and the Company regarding the Employee's rights and obligations upon the occurrence of a Change in Control of the Company. These provisions are intended to assure and encourage in advance the Employee's continued attention and dedication to her assigned duties and her objectivity during the pendency and after the occurrence of any such event. These provisions shall apply in lieu of, and expressly supersede, the provisions of Section 4(b) regarding severance pay and benefits upon a termination of employment, if such termination of employment occurs within 18 months after the occurrence of the first event constituting a Change in Control. These provisions shall terminate and be of no further force or effect beginning 18 months after the occurrence of a Change in Control.

(a) **Change in Control.** During the Term, if within 18 months after a Change in Control, the Employee's employment is terminated by the Company without Cause as provided in Section 3(c) or the Employee terminates her employment for Good Reason as provided in Section 3(d), then, subject to the signing of the Separation Agreement and Release by the Employee and the Separation Agreement and Release becoming irrevocable and fully effective, all within 60 days after the Date of Termination (or such shorter time period provided in the Separation Agreement and Release):

(i) the Company shall pay the Employee a lump sum in cash in an amount equal to 1 times the sum of (A) the Employee's current Base Salary (or the Employee's Base Salary in effect immediately prior to the Change in Control, if higher) plus (B) the Employee's Average Incentive Compensation (For purposes of this Agreement, "Average Incentive Compensation" shall mean the Target Annual Incentive Compensation the Employee would have been entitled to receive in the fiscal year of termination (or the Employee's Target Annual Incentive Compensation in the fiscal year immediately prior to the Change in Control, if higher). In no event shall "Average Incentive Compensation" include any sign-on bonus, retention bonus or any other special bonus.);

(ii) notwithstanding anything to the contrary in any applicable option agreement or stock-based award agreement, all time-based stock options and other time-based stock-based awards held by the Employee shall immediately accelerate and become fully exercisable or nonforfeitable as of the Date of Termination;

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(iii) if the Employee was participating in the Company's group health plan immediately prior to the Date of Termination and elects COBRA health continuation, then the Company shall pay to the Employee a monthly cash payment for 12 months or the Employee's COBRA health continuation period, whichever ends earlier, in an amount equal to the monthly employer contribution that the Company would have made to provide health insurance to the Employee if the Employee had remained employed by the Company; and

(iv) The amounts payable under Section 5(a)(i) and (iii) shall be paid or commence to be paid within 60 days after the Date of Termination; provided, however, that if the 60-day period begins in one calendar year and ends in a second calendar year, such payment shall be paid or commence to be paid in the second calendar year by the last day of such 60-day period.

(b) **Additional Limitation.**

(i) Anything in this Agreement to the contrary notwithstanding, in the event that the amount of any compensation, payment or distribution by the Company to or for the benefit of the Employee, whether paid or payable or distributed or distributable pursuant to the terms of this Agreement or otherwise, calculated in a manner consistent with Section 280G of the Internal Revenue Code of 1986, as amended (the "Code") and the applicable regulations thereunder (the "Aggregate Payments"), would be subject to the excise tax imposed by Section 4999 of the Code, then the Aggregate Payments shall be reduced (but not below zero) so that the sum of all of the Aggregate Payments shall be \$1.00 less than the amount at which the Employee becomes subject to the excise tax imposed by Section 4999 of the Code; provided that such reduction shall only occur if it would result in the Employee receiving a higher After Tax Amount (as defined below) than the Employee would receive if the Aggregate Payments were not subject to such reduction. In such event, the Aggregate Payments shall be reduced in the following order, in each case, in reverse chronological order beginning with the Aggregate Payments that are to be paid the furthest in time from consummation of the transaction that is subject to Section 280G of the Code: (1) cash payments not subject to Section 409A of the Code; (2) cash payments subject to Section 409A of the Code; (3) equity-based payments and acceleration; and (4) non-cash forms of benefits; provided that in the case of all the foregoing Aggregate Payments all amounts or payments that are not subject to calculation under Treas. Reg. §1.280G-1, Q&A-24(b) or (c) shall be reduced before any amounts that are subject to calculation under Treas. Reg. §1.280G-1, Q&A-24(b) or (c).

(ii) For purposes of this Section 5(b), the "After Tax Amount" means the amount of the Aggregate Payments less all federal, state, and local income, excise and employment taxes imposed on the Employee as a result of the Employee's receipt of the Aggregate Payments. For purposes of determining the After Tax Amount, the Employee shall be deemed to pay federal income taxes at the highest marginal rate of federal income taxation applicable to individuals for the calendar year in which the determination is to be made, and state and local income taxes at the highest marginal rates of individual

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taxation in each applicable state and locality, net of the maximum reduction in federal income taxes which could be obtained from deduction of such state and local taxes.

(iii) The determination as to whether a reduction in the Aggregate Payments shall be made pursuant to Section 5(b)(i) shall be made by a nationally recognized accounting firm selected by the Company (the "Accounting Firm"), which shall provide detailed supporting calculations both to the Company and the Employee within 15 business days of the Date of Termination, if applicable, or at such earlier time as is reasonably requested by the Company or the Employee. Any determination by the Accounting Firm shall be binding upon the Company and the Employee.

(c) **Definitions.** For purposes of this Section 5, the following terms shall have the following meanings:

"Change in Control" shall mean any of the following:

(i) any "person," as such term is used in Sections 13(d) and 14(d) of the Securities Exchange Act of 1934, as amended (the "Act") (other than the Company, any of its subsidiaries, or any trustee, fiduciary or other person or entity holding securities under any employee benefit plan or trust of the Company or any of its subsidiaries), together with all "affiliates" and "associates" (as such terms are defined in Rule 12b-2 under the Act) of such person, shall become the "beneficial owner" (as such term is defined in Rule 13d-3 under the Act), directly or indirectly, of securities of the Company representing 50 percent or more of the combined voting power of the Company's then outstanding securities having the right to vote in an election of the Board ("Voting Securities") (in such case other than as a result of an acquisition of securities directly from the Company); or

(ii) the date a majority of the members of the Board is replaced during any 12-month period by directors whose appointment or election is not endorsed by a majority of the members of the Board before the date of the appointment or election; or

(iii) the consummation of (A) any consolidation or merger of the Company where the stockholders of the Company, immediately prior to the consolidation or merger, would not, immediately after the consolidation or merger, beneficially own (as such term is defined in Rule 13d-3 under the Act), directly or indirectly, shares representing in the aggregate more than 50 percent of the voting shares of the Company issuing cash or securities in the consolidation or merger (or of its ultimate parent corporation, if any), or (B) any sale or other transfer (in one transaction or a series of transactions contemplated or arranged by any party as a single plan) of all or substantially all of the assets of the Company and its affiliates on a consolidated basis.

Notwithstanding the foregoing, a "Change in Control" shall not be deemed to have occurred for purposes of the foregoing clause (i) solely as the result of an acquisition of securities by the Company which, by reducing the number of shares of Voting Securities outstanding, increases the proportionate number of Voting Securities beneficially owned by any person to 50 percent or more of the combined voting power of all of the then outstanding Voting

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Securities; provided, however, that if any person referred to in this sentence shall thereafter become the beneficial owner of any additional shares of Voting Securities (other than pursuant to a stock split, stock dividend, or similar transaction or as a result of an acquisition of securities directly from the Company) and immediately thereafter beneficially owns 50 percent or more of the combined voting power of all of the then outstanding Voting Securities, then a "Change in Control" shall be deemed to have occurred for purposes of the foregoing clause (i).

6. **Section 409A.**

(a) Anything in this Agreement to the contrary notwithstanding, if at the time of the Employee's separation from service within the meaning of Section 409A of the Code, the Company determines that the Employee is a "specified employee" within the meaning of Section 409A(a)(2)(B)(i) of the Code, then to the extent any payment or benefit that the Employee becomes entitled to under this Agreement on account of the Employee's separation from service would be considered deferred compensation otherwise subject to the 20 percent additional tax imposed pursuant to Section 409A(a) of the Code as a result of the application of Section 409A(a)(2)(B)(i) of the Code, such payment shall not be payable and such benefit shall not be provided until the date that is the earlier of (A) six months and one day after the Employee's separation from service, or (B) the Employee's death. If any such delayed cash payment is otherwise payable on an installment basis, the first payment shall include a catch-up payment covering amounts that would otherwise have been paid during the six-month period but for the application of this provision, and the balance of the installments shall be payable in accordance with their original schedule.

(b) All in-kind benefits provided and expenses eligible for reimbursement under this Agreement shall be provided by the Company or incurred by the Employee during the time periods set forth in this Agreement. All reimbursements shall be paid as soon as administratively practicable, but in no event shall any reimbursement be paid after the last day of the taxable year following the taxable year in which the expense was incurred. The amount of in-kind benefits provided or reimbursable expenses incurred in one taxable year shall not affect the in-kind benefits to be provided or the expenses eligible for reimbursement in any other taxable year (except for any lifetime or other aggregate limitation applicable to medical expenses). Such right to reimbursement or in-kind benefits is not subject to liquidation or exchange for another benefit.

(c) To the extent that any payment or benefit described in this Agreement constitutes "non-qualified deferred compensation" under Section 409A of the Code, and to the extent that such payment or benefit is payable upon the Employee's termination of employment, then such payments or benefits shall be payable only upon the Employee's "separation from service." The determination of whether and when a separation from service has occurred shall be made in accordance with the presumptions set forth in Treasury Regulation Section 1.409A-1(h).

(d) The parties intend that this Agreement will be administered in accordance with Section 409A of the Code. To the extent that any provision of this Agreement is ambiguous as to its compliance with Section 409A of the Code, the provision shall be read in such a manner so that all payments hereunder comply with Section 409A of the Code. Each payment pursuant

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to this Agreement is intended to constitute a separate payment for purposes of Treasury Regulation Section 1.409A-2(b)(2). The parties agree that this Agreement may be amended, as reasonably requested by either party, and as may be necessary to fully comply with Section 409A of the Code and all related rules and regulations in order to preserve the payments and benefits provided hereunder without additional cost to either party.

(e) The Company makes no representation or warranty and shall have no liability to the Employee or any other person if any provisions of this Agreement are determined to constitute deferred compensation subject to Section 409A of the Code but do not satisfy an exemption from, or the conditions of, such Section.

7. **Confidential Information, Noncompetition and Cooperation.** The terms of the Employee Non-Competition, Non-Solicitation, Confidentiality and Assignment Agreement (the "Restrictive Covenant Agreement"), between the Company and the Employee, attached hereto as Exhibit A, shall continue to be in full force and effect and are incorporated by reference in this Agreement. The Employee hereby reaffirms the terms of the Restrictive Covenant Agreement as material terms of this Agreement.

(a) **Litigation and Regulatory Cooperation.** During and after the Employee's employment, the Employee shall cooperate fully with the Company in the defense or prosecution of any claims or actions now in existence or which may be brought in the future against or on behalf of the Company which relate to events or occurrences that transpired while the Employee was employed by the Company. The Employee's full cooperation in connection with such claims or actions shall include, but not be limited to, being available to meet with counsel to prepare for discovery or trial and to act as a witness on behalf of the Company at mutually convenient times. During and after the Employee's employment, the Employee also shall cooperate fully with the Company in connection with any investigation or review of any federal, state or local regulatory authority as any such investigation or review relates to events or occurrences that transpired while the Employee was employed by the Company. The Company shall reimburse the Employee for any reasonable out-of-pocket expenses incurred in connection with the Employee's performance of obligations pursuant to this Section 7(a).

(b) **Relief.** The Employee agrees that it would be difficult to measure any damages caused to the Company which might result from any breach by the Employee of the promises set forth in this Section 7, and that in any event money damages would be an inadequate remedy for any such breach. Accordingly, subject to Section 8 of this Agreement, the Employee agrees that if the Employee breaches, or proposes to breach, any portion of this Agreement, the Company shall be entitled, in addition to all other remedies that it may have, to an injunction or other appropriate equitable relief to restrain any such breach without showing or proving any actual damage to the Company. In addition, in the event the Employee breaches this Section 7 during a period when she is receiving severance payments pursuant to Section 4 or Section 5 hereof, the Company shall have the right to suspend or terminate such severance payments. Such suspension or termination shall not limit the Company's other options with respect to relief for such breach and shall not relieve the Employee of her duties under this Agreement.

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(c) **Protected Disclosures and Other Protected Action.** Nothing contained in this Agreement limits the Employee's ability to communicate with any federal, state or local governmental agency or commission, including to provide documents or other information, without notice to the Company.

8. **Arbitration of Disputes.** Any controversy or claim arising out of or relating to this Agreement or the breach thereof or otherwise arising out of the Employee's employment or the termination of that employment (including, without limitation, any claims of unlawful employment discrimination whether based on age or otherwise) shall, to the fullest extent permitted by law, be settled by arbitration in any forum and form agreed upon by the parties or, in the absence of such an agreement, under the auspices of the American Arbitration Association ("AAA") in Boston, Massachusetts in accordance with the Employment Dispute Resolution Rules of the AAA, including, but not limited to, the rules and procedures applicable to the selection of arbitrators. In the event that any person or entity other than the Employee or the Company may be a party with regard to any such controversy or claim, such controversy or claim shall be submitted to arbitration subject to such other person or entity's agreement. Judgment upon the award rendered by the arbitrator may be entered in any court having jurisdiction thereof. This Section 8 shall be specifically enforceable. Notwithstanding the foregoing, this Section 8 shall not preclude either party from pursuing a court action for the sole purpose of obtaining a temporary restraining order or a preliminary injunction in circumstances in which such relief is appropriate; provided that any other relief shall be pursued through an arbitration proceeding pursuant to this Section 8.

9. **Consent to Jurisdiction.** To the extent that any court action is permitted consistent with or to enforce Section 8 of this Agreement, the parties hereby consent to the jurisdiction of the Superior Court of the Commonwealth of Massachusetts and the United States District Court for the District of Massachusetts. Accordingly, with respect to any such court action, the Employee (a) submits to the personal jurisdiction of such courts; (b) consents to service of process; and (c) waives any other requirement (whether imposed by statute, rule of court, or otherwise) with respect to personal jurisdiction or service of process.

10. **Integration.** This Agreement constitutes the entire agreement between the parties with respect to the subject matter hereof and supersedes all prior agreements between the parties concerning such subject matter, including the Prior Agreement.

11. **Withholding.** All payments made by the Company to the Employee under this Agreement shall be net of any tax or other amounts required to be withheld by the Company under applicable law.

12. **Successor to the Employee.** This Agreement shall inure to the benefit of and be enforceable by the Employee's personal representatives, executors, administrators, heirs, distributees, devisees and legatees. In the event of the Employee's death after her termination of employment but prior to the completion by the Company of all payments due to her under this Agreement, the Company shall continue such payments to the Employee's beneficiary designated in writing to the Company prior to her death (or to her estate, if the Employee fails to make such designation).

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13. **Enforceability.** If any portion or provision of this Agreement (including, without limitation, any portion or provision of any section of this Agreement) shall to any extent be declared illegal or unenforceable by a court of competent jurisdiction, then the remainder of this Agreement, or the application of such portion or provision in circumstances other than those as to which it is so declared illegal or unenforceable, shall not be affected thereby, and each portion and provision of this Agreement shall be valid and enforceable to the fullest extent permitted by law.

14. **Survival.** The provisions of this Agreement shall survive the termination of this Agreement and/or the termination of the Employee's employment to the extent necessary to effectuate the terms contained herein.

15. **Waiver.** No waiver of any provision hereof shall be effective unless made in writing and signed by the waiving party. The failure of any party to require the performance of any term or obligation of this Agreement, or the waiver by any party of any breach of this Agreement, shall not prevent any subsequent enforcement of such term or obligation or be deemed a waiver of any subsequent breach.

16. **Notices.** Any notices, requests, demands and other communications provided for by this Agreement shall be sufficient if in writing and delivered in person or sent by a nationally recognized overnight courier service or by registered or certified mail, postage prepaid, return receipt requested, to the Employee at the last address the Employee has filed in writing with the Company or, in the case of the Company, at its main offices, attention of the Board.

17. **Amendment.** This Agreement may be amended or modified only by a written instrument signed by the Employee and by a duly authorized representative of the Company.

18. **Governing Law.** This is a Massachusetts contract and shall be construed under and be governed in all respects by the laws of the Commonwealth of Massachusetts without giving effect to the conflict of laws principles thereof.

19. **Counterparts.** This Agreement may be executed in any number of counterparts, each of which when so executed and delivered shall be taken to be an original; but such counterparts shall together constitute one and the same document.

20. **Successor to Company.** The Company shall require any successor (whether direct or indirect, by purchase, merger, consolidation or otherwise) to all or substantially all of the business or assets of the Company expressly to assume and agree to perform this Agreement to the same extent that the Company would be required to perform it if no succession had taken place. Failure of the Company to obtain an assumption of this Agreement at or prior to the effectiveness of any succession shall be a material breach of this Agreement.

21. **Gender Neutral.** Wherever used herein, a pronoun in the masculine gender shall be considered as including the feminine gender unless the context clearly indicates otherwise.

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IN WITNESS WHEREOF, the parties have executed this Agreement effective on the date and year first above written.

SCHOLAR ROCK HOLDING CORPORATION

/s/ Nagesh K. Mahanthappa
By: Nagesh K. Mahanthappa, Ph.D.
Its: President and Chief Executive Officer

EMPLOYEE

/s/ Rhonda M. Chicko
Rhonda M. Chicko

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SCHOLAR ROCK HOLDING CORPORATION

EMPLOYMENT AGREEMENT

This Employment Agreement ("Agreement") is made as of the 11th day of May, 2018, between Scholar Rock Holding Corporation, a Delaware corporation (the "Company"), and Yung H. Chyung (the "Employee") and is effective as of the closing of the Company's first underwritten public offering of its equity securities pursuant to an effective registration statement under the Securities Act of 1933, as amended (the "Effective Date").

WHEREAS, the Company and the Employee are parties to an offer letter, dated February 2, 2016, (the "Prior Agreement"); and

WHEREAS, the parties intend to replace the Prior Agreement with this Agreement, effective as of the Effective Date.

NOW, THEREFORE, in consideration of the mutual covenants and agreements herein contained and other good and valuable consideration, the receipt and sufficiency of which is hereby acknowledged, the parties agree as follows:

1. Employment.

(a) Term. The term of this Agreement shall commence on the Effective Date and continue until terminated in accordance with the provisions hereof (the "Term"). The Employee's employment with the Company will continue to be "at will," meaning that the Employee's employment may be terminated by the Company or the Employee at any time and for any reason subject to the terms of this Agreement.

(b) Position and Duties. During the Term, the Employee shall serve as the Chief Medical Officer of the Company, and shall have such duties and authorities as may from time to time be prescribed by the Chief Executive Officer of the Company (the "CEO"). The Employee shall devote his full working time and efforts to the business and affairs of the Company. Notwithstanding the foregoing, the Employee may serve on other boards of directors, with the approval of the CEO, or engage in religious, charitable or other community activities as long as such services and activities do not materially interfere with the Executive's performance of his duties to the Company as provided in this Agreement.

2. Compensation and Related Matters.

(a) Base Salary. During the Term, the Employee's annual base salary shall be \$400,000. The Employee's base salary shall be reviewed annually by the Compensation Committee of the Board (the "Compensation Committee") or the CEO. The base salary in effect at any given time is referred to herein as "Base Salary." The Base Salary shall be payable in a manner that is consistent with the Company's usual payroll practices.

(b) Incentive Compensation. During the Term, the Employee shall be eligible to receive cash incentive compensation as determined by the Board or the Compensation

Committee from time to time. The Employee's initial target annual incentive compensation shall be thirty-five percent (35%) of his Base Salary (the "Target Annual Incentive Compensation"). Except as otherwise provided herein, to earn incentive compensation, the Employee must be employed by the Company on the day such incentive compensation is paid.

(c) Expenses. The Employee shall be entitled to receive prompt reimbursement for all reasonable expenses incurred by him during the Term in performing services hereunder, in accordance with the policies and procedures then in effect and established by the Company.

(d) Other Benefits. During the Term, the Employee shall be eligible to participate in or receive benefits under the Company's employee benefit plans in effect from time to time, subject to the terms of such plans.

(e) Vacations. During the Term, the Employee shall be entitled to paid vacation in accordance with the Company's policies and procedures. The Employee shall also be entitled to all paid holidays given by the Company in accordance with the policies and procedures then in effect and established by the Company.

3. Termination. During the Term, the Employee's employment hereunder may be terminated without any breach of this Agreement under the following circumstances:

(a) Death. The Employee's employment hereunder shall terminate upon his death.

(b) Termination by Company for Cause. The Company may terminate the Employee's employment hereunder for Cause. For purposes of this Agreement, "Cause" shall mean: (i) conduct by the Employee constituting a material act of misconduct in connection with the performance of his duties, including, without limitation, misappropriation of funds or property of the Company or any of its subsidiaries or affiliates other than the occasional, customary and de minimis use of Company property for personal purposes; (ii) the commission by the Employee of any felony or a misdemeanor involving moral turpitude, deceit, dishonesty or fraud, or any conduct by the Employee that would reasonably be expected to result in material injury or reputational harm to the Company or any of its subsidiaries or affiliates if he were retained in his position; (iii) continued non-performance by the Employee of his duties hereunder (other than by reason of the Employee's physical or mental illness, incapacity or disability) which has continued for more than 30 days following written notice of such non-performance from the CEO; (iv) a breach by the Employee of any of the provisions contained in Section 7 of this Agreement; (v) a material violation by the Employee of the Company's written employment policies; or (vi) failure to cooperate with a bona fide internal investigation or an investigation by regulatory or law enforcement authorities, after being instructed by the Company to cooperate, or the willful destruction or failure to preserve documents or other materials known to be relevant to such investigation or the inducement of others to fail to cooperate or to produce documents or other materials in connection with such investigation.

(c) Termination Without Cause. The Company may terminate the Employee's employment hereunder at any time without Cause. Any termination by the

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Company of the Employee's employment under this Agreement which does not constitute a termination for Cause under Section 3(b) and does not result from the death of the Employee under Section 3(a) shall be deemed a termination without Cause.

(d) Termination by the Employee. The Employee may terminate his employment hereunder at any time for any reason, including but not limited to Good Reason. For purposes of this Agreement, "Good Reason" shall mean that the Employee has complied with the "Good Reason Process" (hereinafter defined) following the occurrence of any of the following events: (i) a material diminution in the Employee's responsibilities, authority or duties; (ii) a material diminution in the Employee's Base Salary except for across-the-board salary reductions based on the Company's financial performance similarly affecting all or substantially all senior management employees of the Company; (iii) a material change in the geographic location at which the Employee provides services to the Company, except for required travel for the Company's business; or (iv) the material breach of this Agreement by the Company. "Good Reason Process" shall mean that (i) the Employee reasonably determines in good faith that a "Good Reason" condition has occurred; (ii) the Employee notifies the Company in writing of the first occurrence of the Good Reason condition within 60 days of the first occurrence of such condition; (iii) the Employee cooperates in good faith with the Company's efforts, for a period not less than 30 days following such notice (the "Cure Period"), to remedy the condition; (iv) notwithstanding such efforts, the Good Reason condition continues to exist; and (v) the Employee terminates his employment within 60 days after the end of the Cure Period. If the Company cures the Good Reason condition during the Cure Period, Good Reason shall be deemed not to have occurred.

(e) Notice of Termination. Except for termination as specified in Section 3(a), any termination of the Employee's employment by the Company or any such termination by the Employee shall be communicated by written Notice of Termination to the other party hereto. For purposes of this Agreement, a "Notice of Termination" shall mean a notice which shall indicate the specific termination provision in this Agreement relied upon.

(f) Date of Termination. "Date of Termination" shall mean: (i) if the Employee's employment is terminated by his death, the date of his death; (ii) if the Employee's employment is terminated by the Company under Section 3(c), the date on which a Notice of Termination is given; (iii) if the Employee's employment is terminated by the Employee under Section 3(d) without Good Reason, 30 days after the date on which a Notice of Termination is given, and (iv) if the Employee's employment is terminated by the Employee under Section 3(d) with Good Reason, the date on which a Notice of Termination is given after the end of the Cure Period. Notwithstanding the foregoing, in the event that the Employee gives a Notice of Termination to the Company, the Company may unilaterally accelerate the Date of Termination and such acceleration shall not result in a termination by the Company for purposes of this Agreement.

4. Compensation Upon Termination.

(a) Termination Generally. If the Employee's employment with the Company is terminated for any reason, the Company shall pay or provide to the Employee (or to his authorized representative or estate) (i) any Base Salary earned through the Date of Termination,

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unpaid expense reimbursements (subject to, and in accordance with, Section 2(c) of this Agreement) and unused vacation that accrued through the Date of Termination on or before the time required by law but in no event more than 30 days after the Employee's Date of Termination; and (ii) any vested benefits the Employee may have under any employee benefit plan of the Company through the Date of Termination, which vested benefits shall be paid and/or provided in accordance with the terms of such employee benefit plans (collectively, the "Accrued Benefit").

(b) Termination by the Company Without Cause or by the Employee with Good Reason. During the Term, if the Employee's employment is terminated by the Company without Cause as provided in Section 3(c), or the Employee terminates his employment for Good Reason as provided in Section 3(d), then the Company shall pay the Employee his Accrued Benefit. In addition, subject to the Employee signing a separation agreement containing, among other provisions, a general release of claims in favor of the Company and related persons and entities, confidentiality, return of property and non-disparagement, in a form and manner satisfactory to the Company (the "Separation Agreement and Release") and the Separation Agreement and Release becoming irrevocable and fully effective, all within 60 days after the Date of Termination (or such shorter time period provided in the Separation Agreement and Release):

(i) the Company shall pay the Employee an amount equal to 9 months of the Employee's Base Salary (the "Severance Amount"). Notwithstanding the foregoing, if the Employee breaches any of the provisions contained in Section 7 of this Agreement, all payments of the Severance Amount shall immediately cease;

(ii) RESERVED

(iii) if the Employee was participating in the Company's group health plan immediately prior to the Date of Termination and elects COBRA health continuation, then the Company shall pay to the Employee a monthly cash payment for 9 months or the Employee's COBRA health continuation period, whichever ends earlier, in an amount equal to the monthly employer contribution that the Company would have made to provide health

insurance to the Employee if the Employee had remained employed by the Company; and

(iv) the amounts payable under Section 4(b)(i) and (iii) shall be paid out in substantially equal installments in accordance with the Company's payroll practice commencing within 60 days after the Date of Termination; provided, however, that if the 60-day period begins in one calendar year and ends in a second calendar year, the Severance Amount shall begin to be paid in the second calendar year by the last day of such 60-day period; provided, further, that the initial payment shall include a catch-up payment to cover amounts retroactive to the day immediately following the Date of Termination. Each payment pursuant to this Agreement is intended to constitute a separate payment for purposes of Treasury Regulation Section 1.409A-2(b)(2).

5. Change in Control Payment. The provisions of this Section 5 set forth certain terms of an agreement reached between the Employee and the Company regarding the

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Employee's rights and obligations upon the occurrence of a Change in Control of the Company. These provisions are intended to assure and encourage in advance the Employee's continued attention and dedication to his assigned duties and his objectivity during the pendency and after the occurrence of any such event. These provisions shall apply in lieu of, and expressly supersede, the provisions of Section 4(b) regarding severance pay and benefits upon a termination of employment, if such termination of employment occurs within 18 months after the occurrence of the first event constituting a Change in Control. These provisions shall terminate and be of no further force or effect beginning 18 months after the occurrence of a Change in Control.

(a) Change in Control. During the Term, if within 18 months after a Change in Control, the Employee's employment is terminated by the Company without Cause as provided in Section 3(c) or the Employee terminates his employment for Good Reason as provided in Section 3(d), then, subject to the signing of the Separation Agreement and Release by the Employee and the Separation Agreement and Release becoming irrevocable and fully effective, all within 60 days after the Date of Termination (or such shorter time period provided in the Separation Agreement and Release):

(i) the Company shall pay the Employee a lump sum in cash in an amount equal to 1 times the sum of (A) the Employee's current Base Salary (or the Employee's Base Salary in effect immediately prior to the Change in Control, if higher) plus (B) the Employee's Average Incentive Compensation (For purposes of this Agreement, "Average Incentive Compensation" shall mean the Target Annual Incentive Compensation the Employee would have been entitled to receive in the fiscal year of termination (or the Employee's Target Annual Incentive Compensation in the fiscal year immediately prior to the Change in Control, if higher). In no event shall "Average Incentive Compensation" include any sign-on bonus, retention bonus or any other special bonus.);

(ii) notwithstanding anything to the contrary in any applicable option agreement or stock-based award agreement, all time-based stock options and other time-based stock-based awards held by the Employee shall immediately accelerate and become fully exercisable or nonforfeitable as of the Date of Termination;

(iii) if the Employee was participating in the Company's group health plan immediately prior to the Date of Termination and elects COBRA health continuation, then the Company shall pay to the Employee a monthly cash payment for 12 months or the Employee's COBRA health continuation period, whichever ends earlier, in an amount equal to the monthly employer contribution that the Company would have made to provide health insurance to the Employee if the Employee had remained employed by the Company; and

(iv) The amounts payable under Section 5(a)(i) and (iii) shall be paid or commence to be paid within 60 days after the Date of Termination; provided, however, that if the 60-day period begins in one calendar year and ends in a second calendar year, such payment shall be paid or commence to be paid in the second calendar year by the last day of such 60-day period.

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(b) Additional Limitation.

(i) Anything in this Agreement to the contrary notwithstanding, in the event that the amount of any compensation, payment or distribution by the Company to or for the benefit of the Employee, whether paid or payable or distributed or distributable pursuant to the terms of this Agreement or otherwise, calculated in a manner consistent with Section 280G of the Internal Revenue Code of 1986, as amended (the "Code") and the applicable regulations thereunder (the "Aggregate Payments"), would be subject to the excise tax imposed by Section 4999 of the Code, then the Aggregate Payments shall be reduced (but not below zero) so that the sum of all of the Aggregate Payments shall be \$1.00 less than the amount at which the Employee becomes subject to the excise tax imposed by Section 4999 of the Code; provided that such reduction shall only occur if it would result in the Employee receiving a higher After Tax Amount (as defined below) than the Employee would receive if the Aggregate Payments were not subject to such reduction. In such event, the Aggregate Payments shall be reduced in the following order, in each case, in reverse chronological order beginning with the Aggregate Payments that are to be paid the furthest in time from consummation of the transaction that is subject to Section 280G of the Code:

(1) cash payments not subject to Section 409A of the Code; (2) cash payments subject to Section 409A of the Code; (3) equity-based payments and acceleration; and (4) non-cash forms of benefits; provided that in the case of all the foregoing Aggregate Payments all amounts or payments that are not subject to calculation under Treas. Reg. §1.280G-1, Q&A-24(b) or (c) shall be reduced before any amounts that are subject to calculation under Treas. Reg. §1.280G-1, Q&A-24(b) or (c).

(ii) For purposes of this Section 5(b), the "After Tax Amount" means the amount of the Aggregate Payments less all federal, state, and local income, excise and employment taxes imposed on the Employee as a result of the Employee's receipt of the Aggregate Payments. For purposes of determining the After Tax Amount, the Employee shall be deemed to pay federal income taxes at the highest marginal rate of federal income taxation applicable to individuals for the calendar year in which the determination is to be made, and state and local income taxes at the highest marginal rates of individual taxation in each applicable state and locality, net of the maximum reduction in federal income taxes which could be obtained from deduction of such state and local taxes.

(iii) The determination as to whether a reduction in the Aggregate Payments shall be made pursuant to Section 5(b)(i) shall be made by a nationally recognized accounting firm selected by the Company (the "Accounting Firm"), which shall provide detailed supporting calculations both to the Company and the Employee within 15 business days of the Date of Termination, if applicable, or at such earlier time as is reasonably requested by the Company or the Employee. Any determination by the Accounting Firm shall be binding upon the Company and the Employee.

(c) Definitions. For purposes of this Section 5, the following terms shall have the following meanings:

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"Change in Control" shall mean any of the following:

(i) any "person," as such term is used in Sections 13(d) and 14(d) of the Securities Exchange Act of 1934, as amended (the "Act") (other than the Company, any of its subsidiaries, or any trustee, fiduciary or other person or entity holding securities under any employee benefit plan or trust of the Company or any of its subsidiaries), together with all "affiliates" and "associates" (as such terms are defined in Rule 12b-2 under the Act) of such person, shall become the "beneficial owner" (as such term is defined in Rule 13d-3 under the Act), directly or indirectly, of securities of the Company representing 50 percent or more of the combined voting power of the Company's then outstanding securities having the right to vote in an election of the Board ("Voting Securities") (in such case other than as a result of an acquisition of securities directly from the Company); or

(ii) the date a majority of the members of the Board is replaced during any 12-month period by directors whose appointment or election is not endorsed by a majority of the members of the Board before the date of the appointment or election; or

(iii) the consummation of (A) any consolidation or merger of the Company where the stockholders of the Company, immediately prior to the consolidation or merger, would not, immediately after the consolidation or merger, beneficially own (as such term is defined in Rule 13d-3 under the Act), directly or indirectly, shares representing in the aggregate more than 50 percent of the voting shares of the Company issuing cash or securities in the consolidation or merger (or of its ultimate parent corporation, if any), or (B) any sale or other transfer (in one transaction or a series of transactions contemplated or arranged by any party as a single plan) of all or substantially all of the assets of the Company and its affiliates on a consolidated basis.

Notwithstanding the foregoing, a "Change in Control" shall not be deemed to have occurred for purposes of the foregoing clause (i) solely as the result of an acquisition of securities by the Company which, by reducing the number of shares of Voting Securities outstanding, increases the proportionate number of Voting Securities beneficially owned by any person to 50 percent or more of the combined voting power of all of the then outstanding Voting Securities; provided, however, that if any person referred to in this sentence shall thereafter become the beneficial owner of any additional shares of Voting Securities (other than pursuant to a stock split, stock dividend, or similar transaction or as a result of an acquisition of securities directly from the Company) and immediately thereafter beneficially owns 50 percent or more of the combined voting power of all of the then outstanding Voting Securities, then a "Change in Control" shall be deemed to have occurred for purposes of the foregoing clause (i).

6. Section 409A.

(a) Anything in this Agreement to the contrary notwithstanding, if at the time of the Employee's separation from service within the meaning of Section 409A of the Code, the Company determines that the Employee is a "specified employee" within the meaning of Section 409A(a)(2)(B)(i) of the Code, then to the extent any payment or benefit that the Employee becomes entitled to under this Agreement on account of the Employee's separation from service

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would be considered deferred compensation otherwise subject to the 20 percent additional tax imposed pursuant to Section 409A(a) of the Code as a result of the application of Section 409A(a)(2)(B)(i) of the Code, such payment shall not be payable and such benefit shall not be provided until the date that is the earlier of (A) six months and one day after the Employee's separation from service, or (B) the Employee's death. If any such delayed cash payment is otherwise payable on an installment basis, the first payment shall include a catch-up payment covering amounts that would otherwise have been paid during the six-month period but for the application of this provision, and the balance of the installments shall be payable in accordance with their original schedule.

(b) All in-kind benefits provided and expenses eligible for reimbursement under this Agreement shall be provided by the Company or incurred by the Employee during the time periods set forth in this Agreement. All reimbursements shall be paid as soon as administratively practicable, but in no event shall any reimbursement be paid after the last day of the taxable year following the taxable year in which the expense was incurred. The amount of in-kind benefits provided or reimbursable expenses incurred in one taxable year shall not affect the in-kind benefits to be provided or the expenses eligible for reimbursement in any other taxable year (except for any lifetime or other aggregate limitation applicable to medical expenses). Such right to reimbursement or in-kind benefits is not subject to liquidation or exchange for another benefit.

(c) To the extent that any payment or benefit described in this Agreement constitutes "non-qualified deferred compensation" under Section 409A of the Code, and to the extent that such payment or benefit is payable upon the Employee's termination of employment, then such payments or benefits shall be payable only upon the Employee's "separation from service." The determination of whether and when a separation from service has occurred shall be made in accordance with the presumptions set forth in Treasury Regulation Section 1.409A-1(h).

(d) The parties intend that this Agreement will be administered in accordance with Section 409A of the Code. To the extent that any provision of this Agreement is ambiguous as to its compliance with Section 409A of the Code, the provision shall be read in such a manner so that all payments hereunder comply with Section 409A of the Code. Each payment pursuant to this Agreement is intended to constitute a separate payment for purposes of Treasury Regulation Section 1.409A-2(b)(2). The parties agree that this Agreement may be amended, as reasonably requested by either party, and as may be necessary to fully comply with Section 409A of the Code and all related rules and regulations in order to preserve the payments and benefits provided hereunder without additional cost to either party.

(e) The Company makes no representation or warranty and shall have no liability to the Employee or any other person if any provisions of this Agreement are determined to constitute deferred compensation subject to Section 409A of the Code but do not satisfy an exemption from, or the conditions of, such Section.

7. Confidential Information, Noncompetition and Cooperation. The terms of the Employee Non-Competition, Non-Solicitation, Confidentiality and Assignment Agreement (the "Restrictive Covenant Agreement"), between the Company and the Employee, attached hereto as

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Exhibit A, shall continue to be in full force and effect and are incorporated by reference in this Agreement. The Employee hereby reaffirms the terms of the Restrictive Covenant Agreement as material terms of this Agreement.

(a) Litigation and Regulatory Cooperation. During and after the Employee's employment, the Employee shall cooperate fully with the Company in the defense or prosecution of any claims or actions now in existence or which may be brought in the future against or on behalf of the Company which relate to events or occurrences that transpired while the Employee was employed by the Company. The Employee's full cooperation in connection with such claims or actions shall include, but not be limited to, being available to meet with counsel to prepare for discovery or trial and to act as a witness on behalf of the Company at mutually convenient times. During and after the Employee's employment, the Employee also shall cooperate fully with the Company in connection with any investigation or review of any federal, state or local regulatory authority as any such investigation or review relates to events or occurrences that transpired while the Employee was employed by the Company. The Company shall reimburse the Employee for any reasonable out-of-pocket expenses incurred in connection with the Employee's performance of obligations pursuant to this Section 7(a).

(b) Relief. The Employee agrees that it would be difficult to measure any damages caused to the Company which might result from any breach by the Employee of the promises set forth in this Section 7, and that in any event money damages would be an inadequate remedy for any such breach. Accordingly, subject to Section 8 of this Agreement, the Employee agrees that if the Employee breaches, or proposes to breach, any portion of this Agreement, the Company shall be entitled, in addition to all other remedies that it may have, to an injunction or other appropriate equitable relief to restrain any such breach without showing or proving any actual damage to the Company. In addition, in the event the Employee breaches this Section 7 during a period when he is receiving severance payments pursuant to Section 4 or Section 5 hereof, the Company shall have the right to suspend or terminate such severance payments. Such suspension or termination shall not limit the Company's other options with respect to relief for such breach and shall not relieve the Employee of his duties under this Agreement.

(c) Protected Disclosures and Other Protected Action. Nothing contained in this Agreement limits the Employee's ability to communicate with any federal, state or local governmental agency or commission, including to provide documents or other information, without notice to the Company.

8. Arbitration of Disputes. Any controversy or claim arising out of or relating to this Agreement or the breach thereof or otherwise arising out of the Employee's employment or the termination of that employment (including, without limitation, any claims of unlawful employment discrimination whether based on age or otherwise) shall, to the fullest extent permitted by law, be settled by arbitration in any forum and form agreed upon by the parties or, in the absence of such an agreement, under the auspices of the American Arbitration Association ("AAA") in Boston, Massachusetts in accordance with the Employment Dispute Resolution Rules of the AAA, including, but not limited to, the rules and procedures applicable to the selection of arbitrators. In the event that any person or entity other than the Employee or the Company may be a party with regard to any such controversy or claim, such controversy or

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claim shall be submitted to arbitration subject to such other person or entity's agreement. Judgment upon the award rendered by the arbitrator may be entered in any court having jurisdiction thereof. This Section 8 shall be specifically enforceable. Notwithstanding the foregoing, this Section 8 shall not preclude either party from pursuing a court action for the sole purpose of obtaining a temporary restraining order or a preliminary injunction in circumstances in which such relief is appropriate; provided that any other relief shall be pursued through an arbitration proceeding pursuant to this Section 8.

9. Consent to Jurisdiction. To the extent that any court action is permitted consistent with or to enforce Section 8 of this Agreement, the parties hereby consent to the jurisdiction of the Superior Court of the Commonwealth of Massachusetts and the United States District Court for the District of Massachusetts. Accordingly, with respect to any such court action, the Employee (a) submits to the personal jurisdiction of such courts; (b) consents to service of process; and (c) waives any other requirement (whether imposed by statute, rule of court, or otherwise) with respect to personal jurisdiction or service of process.

10. Integration. This Agreement constitutes the entire agreement between the parties with respect to the subject matter hereof and supersedes all prior agreements between the parties concerning such subject matter, including the Prior Agreement.

11. Withholding. All payments made by the Company to the Employee under this Agreement shall be net of any tax or other amounts required to be withheld by the Company under applicable law.

12. Successor to the Employee. This Agreement shall inure to the benefit of and be enforceable by the Employee's personal representatives, executors, administrators, heirs, distributees, devisees and legatees. In the event of the Employee's death after his termination of employment but prior to the completion by the Company of all payments due to him under this Agreement, the Company shall continue such payments to the Employee's beneficiary designated in writing to the Company prior to his death (or to his estate, if the Employee fails to make such designation).

13. Enforceability. If any portion or provision of this Agreement (including, without limitation, any portion or provision of any section of this Agreement) shall to any extent be declared illegal or unenforceable by a court of competent jurisdiction, then the remainder of this Agreement, or the application of such portion or provision in circumstances other than those as to which it is so declared illegal or unenforceable, shall not be affected thereby, and each portion and provision of this Agreement shall be valid and enforceable to the fullest extent permitted by law.

14. Survival. The provisions of this Agreement shall survive the termination of this Agreement and/or the termination of the Employee's employment to the extent necessary to effectuate the terms contained herein.

15. Waiver. No waiver of any provision hereof shall be effective unless made in writing and signed by the waiving party. The failure of any party to require the performance of any term or obligation of this Agreement, or the waiver by any party of any breach of this

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Agreement, shall not prevent any subsequent enforcement of such term or obligation or be deemed a waiver of any subsequent breach.

16. Notices. Any notices, requests, demands and other communications provided for by this Agreement shall be sufficient if in writing and delivered in person or sent by a nationally recognized overnight courier service or by registered or certified mail, postage prepaid, return receipt requested, to the Employee at the last address the Employee has filed in writing with the Company or, in the case of the Company, at its main offices, attention of the Board.

17. Amendment. This Agreement may be amended or modified only by a written instrument signed by the Employee and by a duly authorized representative of the Company.

18. Governing Law. This is a Massachusetts contract and shall be construed under and be governed in all respects by the laws of the Commonwealth of Massachusetts without giving effect to the conflict of laws principles thereof.

19. Counterparts. This Agreement may be executed in any number of counterparts, each of which when so executed and delivered shall be taken to be an original; but such counterparts shall together constitute one and the same document.

20. Successor to Company. The Company shall require any successor (whether direct or indirect, by purchase, merger, consolidation or otherwise) to all or substantially all of the business or assets of the Company expressly to assume and agree to perform this Agreement to the same extent that the Company would be required to perform it if no succession had taken place. Failure of the Company to obtain an assumption of this Agreement at or prior to the effectiveness of any succession shall be a material breach of this Agreement.

21. Gender Neutral. Wherever used herein, a pronoun in the masculine gender shall be considered as including the feminine gender unless the context clearly indicates otherwise.

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IN WITNESS WHEREOF, the parties have executed this Agreement effective on the date and year first above written.

SCHOLAR ROCK HOLDING CORPORATION

/s/ Nagesh K. Mahanthappa

By: Nagesh K. Mahanthappa, Ph.D.

Its: President and Chief Executive Officer

EMPLOYEE

/s/ Yung H. Chyung

Yung H. Chyung

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SCHOLAR ROCK HOLDING CORPORATION

EMPLOYMENT AGREEMENT

This Employment Agreement ("Agreement") is made as of the 11th day of May, 2018, between Scholar Rock Holding Corporation, a Delaware corporation (the "Company"), and Elan Z. Ezickson (the "Employee") and is effective as of the closing of the Company's first underwritten public offering of its equity securities pursuant to an effective registration statement under the Securities Act of 1933, as amended (the "Effective Date").

WHEREAS, the Company and the Employee are parties to an offer letter, dated July 17, 2014, (the "Prior Agreement"); and

WHEREAS, the parties intend to replace the Prior Agreement with this Agreement, effective as of the Effective Date.

NOW, THEREFORE, in consideration of the mutual covenants and agreements herein contained and other good and valuable consideration, the receipt and sufficiency of which is hereby acknowledged, the parties agree as follows:

1. Employment.

(a) Term. The term of this Agreement shall commence on the Effective Date and continue until terminated in accordance with the provisions hereof (the "Term"). The Employee's employment with the Company will continue to be "at will," meaning that the Employee's employment may be terminated by the Company or the Employee at any time and for any reason subject to the terms of this Agreement.

(b) Position and Duties. During the Term, the Employee shall serve as the Chief Operating Officer and Head of Corporate Development of the Company, and shall have such duties and authorities as may from time to time be prescribed by the Chief Executive Officer of the Company (the "CEO"). The Employee shall devote his full working time and efforts to the business and affairs of the Company. Notwithstanding the foregoing, the Employee may serve on other boards of directors, with the approval of the CEO, or engage in religious, charitable or other community activities as long as such services and activities do not materially interfere with the Executive's performance of his duties to the Company as provided in this Agreement.

2. Compensation and Related Matters.

(a) Base Salary. During the Term, the Employee's annual base salary shall be \$380,000. The Employee's base salary shall be reviewed annually by the Compensation Committee of the Board (the "Compensation Committee") or the CEO. The base salary in effect at any given time is referred to herein as "Base Salary." The Base Salary shall be payable in a manner that is consistent with the Company's usual payroll practices.

(b) Incentive Compensation. During the Term, the Employee shall be eligible to receive cash incentive compensation as determined by the Board or the Compensation Committee from time to time. The Employee's initial target annual incentive compensation shall be thirty-five percent (35%) of his Base Salary (the "Target Annual Incentive Compensation"). Except as otherwise provided herein, to earn incentive compensation, the Employee must be employed by the Company on the day such incentive compensation is paid.

(c) Expenses. The Employee shall be entitled to receive prompt reimbursement for all reasonable expenses incurred by him during the Term in performing services hereunder, in accordance with the policies and procedures then in effect and established by the Company.

(d) Other Benefits. During the Term, the Employee shall be eligible to participate in or receive benefits under the Company's employee benefit plans in effect from time to time, subject to the terms of such plans.

(e) Vacations. During the Term, the Employee shall be entitled to paid vacation in accordance with the Company's policies and procedures. The Employee shall also be entitled to all paid holidays given by the Company in accordance with the policies and procedures then in effect and established by the Company.

3. Termination. During the Term, the Employee's employment hereunder may be terminated without any breach of this Agreement under the following circumstances:

(a) Death. The Employee's employment hereunder shall terminate upon his death.

(b) Termination by Company for Cause. The Company may terminate the Employee's employment hereunder for Cause. For purposes of this Agreement, "Cause" shall mean: (i) conduct by the Employee constituting a material act of misconduct in connection with the performance of his duties, including, without limitation, misappropriation of funds or property of the Company or any of its subsidiaries or affiliates other than the occasional, customary and de minimis use of Company property for personal purposes; (ii) the commission by the Employee of any felony or a misdemeanor involving moral turpitude, deceit, dishonesty or fraud, or any conduct by the Employee that would reasonably be expected to result in material injury or reputational harm to the Company or any of its subsidiaries or affiliates if he were retained in his position; (iii) continued non-performance by the Employee of his duties hereunder (other than by reason of the Employee's physical or mental illness, incapacity or disability) which has continued for more than 30 days following written notice of such non-performance from the CEO; (iv) a breach by the Employee of any of the provisions contained in Section 7 of this Agreement; (v) a material violation by the Employee of the Company's written employment policies; or (vi) failure to cooperate with a bona fide internal investigation or an investigation by regulatory or law enforcement authorities, after being instructed by the Company to cooperate, or the willful destruction or failure to preserve documents or other materials known to be relevant to such investigation or the inducement of others to fail to cooperate or to produce documents or other materials in connection with such investigation.

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(c) Termination Without Cause. The Company may terminate the Employee's employment hereunder at any time without Cause. Any termination by the Company of the Employee's employment under this Agreement which does not constitute a termination for Cause under Section 3(b) and does not result from the death of the Employee under Section 3(a) shall be deemed a termination without Cause.

(d) Termination by the Employee. The Employee may terminate his employment hereunder at any time for any reason, including but not limited to Good Reason. For purposes of this Agreement, "Good Reason" shall mean that the Employee has complied with the "Good Reason Process" (hereinafter defined) following the occurrence of any of the following events: (i) a material diminution in the Employee's responsibilities, authority or duties, provided that the hiring by the Company of any Company officers with customary responsibilities, authority or duties (including any CBO, CFO or CMO), will not constitute any such material diminution; (ii) a material diminution in the Employee's Base Salary except for across-the-board salary reductions based on the Company's financial performance similarly affecting all or substantially all senior management employees of the Company; (iii) a material change in the geographic location at which the Employee provides services to the Company, except for required travel for the Company's business; or (iv) the material breach of this Agreement by the Company. "Good Reason Process" shall mean that (i) the Employee reasonably determines in good faith that a "Good Reason" condition has occurred; (ii) the Employee notifies the Company in writing of the first occurrence of the Good Reason condition within 60 days of the first occurrence of such condition; (iii) the Employee cooperates in good faith with the Company's efforts, for a period not less than 30 days following such notice (the "Cure Period"), to remedy the condition; (iv) notwithstanding such efforts, the Good Reason condition continues to exist; and (v) the Employee terminates his employment within 60 days after the end of the Cure Period. If the Company cures the Good Reason condition during the Cure Period, Good Reason shall be deemed not to have occurred.

(e) Notice of Termination. Except for termination as specified in Section 3(a), any termination of the Employee's employment by the Company or any such termination by the Employee shall be communicated by written Notice of Termination to the other party hereto. For purposes of this Agreement, a "Notice of Termination" shall mean a notice which shall indicate the specific termination provision in this Agreement relied upon.

(f) Date of Termination. "Date of Termination" shall mean: (i) if the Employee's employment is terminated by his death, the date of his death; (ii) if the Employee's employment is terminated by the Company under Section 3(c), the date on which a Notice of Termination is given; (iii) if the Employee's employment is terminated by the Employee under Section 3(d) without Good Reason, 30 days after the date on which a Notice of Termination is given, and (iv) if the Employee's employment is terminated by the Employee under Section 3(d) with Good Reason, the date on which a Notice of Termination is given after the end of the Cure Period. Notwithstanding the foregoing, in the event that the Employee gives a Notice of Termination to the Company, the Company may unilaterally accelerate the Date of Termination and such acceleration shall not result in a termination by the Company for purposes of this Agreement.

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4. Compensation Upon Termination.

(a) Termination Generally. If the Employee's employment with the Company is terminated for any reason, the Company shall pay or provide to the Employee (or to his authorized representative or estate) (i) any Base Salary earned through the Date of Termination, unpaid expense reimbursements (subject to, and in accordance with, Section 2(c) of this Agreement) and unused vacation that accrued through the Date of Termination on or before the time required by law but in no event more than 30 days after the Employee's Date of Termination; and (ii) any vested benefits the Employee may have under any employee benefit plan of the Company through the Date of Termination, which vested benefits shall be paid and/or provided in accordance with the terms of such employee benefit plans (collectively, the "Accrued Benefit").

(b) Termination by the Company Without Cause or by the Employee with Good Reason. During the Term, if the Employee's employment is terminated by the Company without Cause as provided in Section 3(c), or the Employee terminates his employment for Good Reason as provided in Section 3(d), then the Company shall pay the Employee his Accrued Benefit. In addition, subject to the Employee signing a separation agreement containing, among other provisions, a general release of claims in favor of the Company and related persons and entities, confidentiality, return of property and non-disparagement, in a form and manner satisfactory to the Company (the "Separation Agreement and Release") and the Separation Agreement and Release becoming irrevocable and fully effective, all within 60 days after the Date of Termination (or such shorter time period provided in the Separation Agreement and Release):

(i) the Company shall pay the Employee an amount equal to 9 months of the Employee's Base Salary (the "Severance Amount"). Notwithstanding the foregoing, if the Employee breaches any of the provisions contained in Section 7 of this Agreement, all payments of the Severance Amount shall immediately cease;

(ii) RESERVED

(iii) if the Employee was participating in the Company's group health plan immediately prior to the Date of Termination and elects COBRA health continuation, then the Company shall pay to the Employee a monthly cash payment for 9 months or the Employee's COBRA health continuation period, whichever ends earlier, in an amount equal to the monthly employer contribution that the Company would have made to provide health insurance to the Employee if the Employee had remained employed by the Company; and

(iv) the amounts payable under Section 4(b)(i) and (iii) shall be paid out in substantially equal installments in accordance with the Company's payroll practice commencing within 60 days after the Date of Termination; provided, however, that if the 60-day period begins in one calendar year and ends in a second calendar year, the Severance Amount shall begin to be paid in the second calendar year by the last day of such 60-day

Termination. Each payment pursuant to this Agreement is intended to constitute a separate payment for purposes of Treasury Regulation Section 1.409A-2(b)(2).

5. **Change in Control Payment.** The provisions of this Section 5 set forth certain terms of an agreement reached between the Employee and the Company regarding the Employee's rights and obligations upon the occurrence of a Change in Control of the Company. These provisions are intended to assure and encourage in advance the Employee's continued attention and dedication to his assigned duties and his objectivity during the pendency and after the occurrence of any such event. These provisions shall apply in lieu of, and expressly supersede, the provisions of Section 4(b) regarding severance pay and benefits upon a termination of employment, if such termination of employment occurs within 18 months after the occurrence of the first event constituting a Change in Control. These provisions shall terminate and be of no further force or effect beginning 18 months after the occurrence of a Change in Control.

(a) **Change in Control.** During the Term, if within 18 months after a Change in Control, the Employee's employment is terminated by the Company without Cause as provided in Section 3(c) or the Employee terminates his employment for Good Reason as provided in Section 3(d), then, subject to the signing of the Separation Agreement and Release by the Employee and the Separation Agreement and Release becoming irrevocable and fully effective, all within 60 days after the Date of Termination (or such shorter time period provided in the Separation Agreement and Release):

(i) the Company shall pay the Employee a lump sum in cash in an amount equal to 1 times the sum of (A) the Employee's current Base Salary (or the Employee's Base Salary in effect immediately prior to the Change in Control, if higher) plus (B) the Employee's Average Incentive Compensation (For purposes of this Agreement, "Average Incentive Compensation" shall mean the Target Annual Incentive Compensation the Employee would have been entitled to receive in the fiscal year of termination (or the Employee's Target Annual Incentive Compensation in the fiscal year immediately prior to the Change in Control, if higher). In no event shall "Average Incentive Compensation" include any sign-on bonus, retention bonus or any other special bonus.);

(ii) notwithstanding anything to the contrary in any applicable option agreement or stock-based award agreement, all time-based stock options and other time-based stock-based awards held by the Employee shall immediately accelerate and become fully exercisable or nonforfeitable as of the Date of Termination;

(iii) if the Employee was participating in the Company's group health plan immediately prior to the Date of Termination and elects COBRA health continuation, then the Company shall pay to the Employee a monthly cash payment for 12 months or the Employee's COBRA health continuation period, whichever ends earlier, in an amount equal to the monthly employer contribution that the Company would have made to provide health insurance to the Employee if the Employee had remained employed by the Company; and

(iv) The amounts payable under Section 5(a)(i) and (iii) shall be paid or commence to be paid within 60 days after the Date of Termination; provided, however, that if the 60-day period begins in one calendar year and ends in a second calendar year, such payment shall be paid or commence to be paid in the second calendar year by the last day of such 60-day period.

(b) **Additional Limitation.**

(i) Anything in this Agreement to the contrary notwithstanding, in the event that the amount of any compensation, payment or distribution by the Company to or for the benefit of the Employee, whether paid or payable or distributed or distributable pursuant to the terms of this Agreement or otherwise, calculated in a manner consistent with Section 280G of the Internal Revenue Code of 1986, as amended (the "Code") and the applicable regulations thereunder (the "Aggregate Payments"), would be subject to the excise tax imposed by Section 4999 of the Code, then the Aggregate Payments shall be reduced (but not below zero) so that the sum of all of the Aggregate Payments shall be \$1.00 less than the amount at which the Employee becomes subject to the excise tax imposed by Section 4999 of the Code; provided that such reduction shall only occur if it would result in the Employee receiving a higher After Tax Amount (as defined below) than the Employee would receive if the Aggregate Payments were not subject to such reduction. In such event, the Aggregate Payments shall be reduced in the following order, in each case, in reverse chronological order beginning with the Aggregate Payments that are to be paid the furthest in time from consummation of the transaction that is subject to Section 280G of the Code: (1) cash payments not subject to Section 409A of the Code; (2) cash payments subject to Section 409A of the Code; (3) equity-based payments and acceleration; and (4) non-cash forms of benefits; provided that in the case of all the foregoing Aggregate Payments all amounts or payments that are not subject to calculation under Treas. Reg. §1.280G-1, Q&A-24(b) or (c) shall be reduced before any amounts that are subject to calculation under Treas. Reg. §1.280G-1, Q&A-24(b) or (c).

(ii) For purposes of this Section 5(b), the "After Tax Amount" means the amount of the Aggregate Payments less all federal, state, and local income, excise and employment taxes imposed on the Employee as a result of the Employee's receipt of the Aggregate Payments. For purposes of determining the After Tax Amount, the Employee shall be deemed to pay federal income taxes at the highest marginal rate of federal income taxation applicable to individuals for the calendar year in which the determination is to be made, and state and local income taxes at the highest marginal rates of individual taxation in each applicable state and locality, net of the maximum reduction in federal income taxes which could be obtained from deduction of such state and local taxes.

(iii) The determination as to whether a reduction in the Aggregate Payments shall be made pursuant to Section 5(b)(i) shall be made by a nationally recognized accounting firm selected by the Company (the "Accounting Firm"), which shall provide detailed supporting calculations both to the Company and the Employee within 15 business days of the Date of Termination, if applicable, or at such earlier time

as is reasonably requested by the Company or the Employee. Any determination by the Accounting Firm shall be binding upon the Company and the Employee.

(c) **Definitions.** For purposes of this Section 5, the following terms shall have the following meanings:

"Change in Control" shall mean any of the following:

(i) any "person," as such term is used in Sections 13(d) and 14(d) of the Securities Exchange Act of 1934, as amended (the "Act") (other than the Company, any of its subsidiaries, or any trustee, fiduciary or other person or entity holding securities under any employee benefit plan or trust of the Company or any of its subsidiaries), together with all "affiliates" and "associates" (as such terms are defined in Rule 12b-2 under the Act) of such person, shall become the "beneficial owner" (as such term is defined in Rule 13d-3 under the Act), directly or indirectly, of securities of the Company representing 50 percent or more of the combined voting power of the Company's then outstanding securities having the right to vote in an election of the Board ("Voting Securities") (in such case other than as a result of an acquisition of securities directly from the Company); or

(ii) the date a majority of the members of the Board is replaced during any 12-month period by directors whose appointment or election is not endorsed by a majority of the members of the Board before the date of the appointment or election; or

(iii) the consummation of (A) any consolidation or merger of the Company where the stockholders of the Company, immediately prior to the consolidation or merger, would not, immediately after the consolidation or merger, beneficially own (as such term is defined in Rule 13d-3 under the Act), directly or indirectly, shares representing in the aggregate more than 50 percent of the voting shares of the Company issuing cash or securities in the consolidation or merger (or of its ultimate parent corporation, if any), or (B) any sale or other transfer (in one transaction or a series of transactions contemplated or arranged by any party as a single plan) of all or substantially all of the assets of the Company and its affiliates on a consolidated basis.

Notwithstanding the foregoing, a "Change in Control" shall not be deemed to have occurred for purposes of the foregoing clause (i) solely as the result of an acquisition of securities by the Company which, by reducing the number of shares of Voting Securities outstanding, increases the proportionate number of Voting Securities beneficially owned by any person to 50 percent or more of the combined voting power of all of the then outstanding Voting Securities; provided, however, that if any person referred to in this sentence shall thereafter become the beneficial owner of any additional shares of Voting Securities (other than pursuant to a stock split, stock dividend, or similar transaction or as a result of an acquisition of securities directly from the Company) and immediately thereafter beneficially owns 50 percent or more of the combined voting power of all of the then outstanding Voting Securities, then a "Change in Control" shall be deemed to have occurred for purposes of the foregoing clause (i).

6. **Section 409A.**

(a) Anything in this Agreement to the contrary notwithstanding, if at the time of the Employee's separation from service within the meaning of Section 409A of the Code, the Company determines that the Employee is a "specified employee" within the meaning of Section 409A(a)(2)(B)(i) of the Code, then to the extent any payment or benefit that the Employee becomes entitled to under this Agreement on account of the Employee's separation from service would be considered deferred compensation otherwise subject to the 20 percent additional tax imposed pursuant to Section 409A(a) of the Code as a result of the application of Section 409A(a)(2)(B)(i) of the Code, such payment shall not be payable and such benefit shall not be provided until the date that is the earlier of (A) six months and one day after the Employee's separation from service, or (B) the Employee's death. If any such delayed cash payment is otherwise payable on an installment basis, the first payment shall include a catch-up payment covering amounts that would otherwise have been paid during the six-month period but for the application of this provision, and the balance of the installments shall be payable in accordance with their original schedule.

(b) All in-kind benefits provided and expenses eligible for reimbursement under this Agreement shall be provided by the Company or incurred by the Employee during the time periods set forth in this Agreement. All reimbursements shall be paid as soon as administratively practicable, but in no event shall any reimbursement be paid after the last day of the taxable year following the taxable year in which the expense was incurred. The amount of in-kind benefits provided or reimbursable expenses incurred in one taxable year shall not affect the in-kind benefits to be provided or the expenses eligible for reimbursement in any other taxable year (except for any lifetime or other aggregate limitation applicable to medical expenses). Such right to reimbursement or in-kind benefits is not subject to liquidation or exchange for another benefit.

(c) To the extent that any payment or benefit described in this Agreement constitutes "non-qualified deferred compensation" under Section 409A of the Code, and to the extent that such payment or benefit is payable upon the Employee's termination of employment, then such payments or benefits shall be payable only upon the Employee's "separation from service." The determination of whether and when a separation from service has occurred shall be made in accordance with the presumptions set forth in Treasury Regulation Section 1.409A-1(h).

(d) The parties intend that this Agreement will be administered in accordance with Section 409A of the Code. To the extent that any provision of this Agreement is ambiguous as to its compliance with Section 409A of the Code, the provision shall be read in such a manner so that all payments hereunder comply with Section 409A of the Code. Each payment pursuant to this Agreement is intended to constitute a separate payment for purposes of Treasury Regulation Section 1.409A-2(b)(2). The parties agree that this Agreement may be amended, as reasonably requested by either party, and as may be necessary to fully comply with Section 409A of the Code and all related rules and regulations in order to preserve the payments and benefits provided hereunder without additional cost to either party.

(e) The Company makes no representation or warranty and shall have no liability to the Employee or any other person if any provisions of this Agreement are determined to constitute deferred compensation subject to Section 409A of the Code but do not satisfy an exemption from, or the conditions of, such Section.

7. **Confidential Information, Noncompetition and Cooperation.** The terms of the Employee Non-Competition, Non-Solicitation, Confidentiality and Assignment Agreement (the "Restrictive Covenant Agreement"), between the Company and the Employee, attached hereto as Exhibit A, shall continue to be in full force and effect and are incorporated by reference in this Agreement. The Employee hereby reaffirms the terms of the Restrictive Covenant Agreement as material terms of this Agreement.

(a) **Litigation and Regulatory Cooperation.** During and after the Employee's employment, the Employee shall cooperate fully with the Company in the defense or prosecution of any claims or actions now in existence or which may be brought in the future against or on behalf of the Company which relate to events or occurrences that transpired while the Employee was employed by the Company. The Employee's full cooperation in connection with such claims or actions shall include, but not be limited to, being available to meet with counsel to prepare for discovery or trial and to act as a witness on behalf of the Company at mutually convenient times. During and after the Employee's employment, the Employee also shall cooperate fully with the Company in connection with any investigation or review of any federal, state or local regulatory authority as any such investigation or review relates to events or occurrences that transpired while the Employee was employed by the Company. The Company shall reimburse the Employee for any reasonable out-of-pocket expenses incurred in connection with the Employee's performance of obligations pursuant to this Section 7(a).

(b) **Relief.** The Employee agrees that it would be difficult to measure any damages caused to the Company which might result from any breach by the Employee of the promises set forth in this Section 7, and that in any event money damages would be an inadequate remedy for any such breach. Accordingly, subject to Section 8 of this Agreement, the Employee agrees that if the Employee breaches, or proposes to breach, any portion of this Agreement, the Company shall be entitled, in addition to all other remedies that it may have, to an injunction or other appropriate equitable relief to restrain any such breach without showing or proving any actual damage to the Company. In addition, in the event the Employee breaches this Section 7 during a period when he is receiving severance payments pursuant to Section 4 or Section 5 hereof, the Company shall have the right to suspend or terminate such severance payments. Such suspension or termination shall not limit the Company's other options with respect to relief for such breach and shall not relieve the Employee of his duties under this Agreement.

(c) **Protected Disclosures and Other Protected Action.** Nothing contained in this Agreement limits the Employee's ability to communicate with any federal, state or local governmental agency or commission, including to provide documents or other information, without notice to the Company.

8. **Arbitration of Disputes.** Any controversy or claim arising out of or relating to this Agreement or the breach thereof or otherwise arising out of the Employee's employment or the

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termination of that employment (including, without limitation, any claims of unlawful employment discrimination whether based on age or otherwise) shall, to the fullest extent permitted by law, be settled by arbitration in any forum and form agreed upon by the parties or, in the absence of such an agreement, under the auspices of the American Arbitration Association ("AAA") in Boston, Massachusetts in accordance with the Employment Dispute Resolution Rules of the AAA, including, but not limited to, the rules and procedures applicable to the selection of arbitrators. In the event that any person or entity other than the Employee or the Company may be a party with regard to any such controversy or claim, such controversy or claim shall be submitted to arbitration subject to such other person or entity's agreement. Judgment upon the award rendered by the arbitrator may be entered in any court having jurisdiction thereof. This Section 8 shall be specifically enforceable. Notwithstanding the foregoing, this Section 8 shall not preclude either party from pursuing a court action for the sole purpose of obtaining a temporary restraining order or a preliminary injunction in circumstances in which such relief is appropriate; provided that any other relief shall be pursued through an arbitration proceeding pursuant to this Section 8.

9. **Consent to Jurisdiction.** To the extent that any court action is permitted consistent with or to enforce Section 8 of this Agreement, the parties hereby consent to the jurisdiction of the Superior Court of the Commonwealth of Massachusetts and the United States District Court for the District of Massachusetts. Accordingly, with respect to any such court action, the Employee (a) submits to the personal jurisdiction of such courts; (b) consents to service of process; and (c) waives any other requirement (whether imposed by statute, rule of court, or otherwise) with respect to personal jurisdiction or service of process.

10. **Integration.** This Agreement constitutes the entire agreement between the parties with respect to the subject matter hereof and supersedes all prior agreements between the parties concerning such subject matter, including the Prior Agreement.

11. **Withholding.** All payments made by the Company to the Employee under this Agreement shall be net of any tax or other amounts required to be withheld by the Company under applicable law.

12. **Successor to the Employee.** This Agreement shall inure to the benefit of and be enforceable by the Employee's personal representatives, executors, administrators, heirs, distributees, devisees and legatees. In the event of the Employee's death after his termination of employment but prior to the completion by the Company of all payments due to him under this Agreement, the Company shall continue such payments to the Employee's beneficiary designated in writing to the Company prior to his death (or to his estate, if the Employee fails to make such designation).

13. **Enforceability.** If any portion or provision of this Agreement (including, without limitation, any portion or provision of any section of this Agreement) shall to any extent be declared illegal or unenforceable by a court of competent jurisdiction, then the remainder of this Agreement, or the application of such portion or provision in circumstances other than those as to which it is so declared illegal or unenforceable, shall not be affected thereby, and each portion and provision of this Agreement shall be valid and enforceable to the fullest extent permitted by law.

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14. **Survival.** The provisions of this Agreement shall survive the termination of this Agreement and/or the termination of the Employee's employment to the extent necessary to effectuate the terms contained herein.

15. **Waiver.** No waiver of any provision hereof shall be effective unless made in writing and signed by the waiving party. The failure of any party to require the performance of any term or obligation of this Agreement, or the waiver by any party of any breach of this Agreement, shall not prevent any subsequent enforcement of such term or obligation or be deemed a waiver of any subsequent breach.

16. **Notices.** Any notices, requests, demands and other communications provided for by this Agreement shall be sufficient if in writing and delivered in person or sent by a nationally recognized overnight courier service or by registered or certified mail, postage prepaid, return receipt requested, to the Employee at the last address the Employee has filed in writing with the Company or, in the case of the Company, at its main offices, attention of the Board.

17. **Amendment.** This Agreement may be amended or modified only by a written instrument signed by the Employee and by a duly authorized representative of the Company.

18. **Governing Law.** This is a Massachusetts contract and shall be construed under and be governed in all respects by the laws of the Commonwealth of Massachusetts without giving effect to the conflict of laws principles thereof.

19. **Counterparts.** This Agreement may be executed in any number of counterparts, each of which when so executed and delivered shall be taken to be an original; but such counterparts shall together constitute one and the same document.

20. **Successor to Company.** The Company shall require any successor (whether direct or indirect, by purchase, merger, consolidation or otherwise) to all or substantially all of the business or assets of the Company expressly to assume and agree to perform this Agreement to the same extent that the Company would be required to perform it if no succession had taken place. Failure of the Company to obtain an assumption of this Agreement at or prior to the effectiveness of any succession shall be a material breach of this Agreement.

21. **Gender Neutral.** Wherever used herein, a pronoun in the masculine gender shall be considered as including the feminine gender unless the context clearly indicates otherwise.

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IN WITNESS WHEREOF, the parties have executed this Agreement effective on the date and year first above written.

SCHOLAR ROCK HOLDING CORPORATION

/s/ Nagesh K. Mahanthappa

By: Nagesh K. Mahanthappa, Ph.D.

Its: President and Chief Executive Officer

EMPLOYEE

/s/ Elan Z. Ezickson

Elan Z. Ezickson

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Consent of Independent Registered Public Accounting Firm

We consent to the reference to our firm under the caption "Experts" and to the use of our report dated May 8, 2018, except for Note 22, as to which the date is May 14, 2018, in Amendment No. 2 to the Registration Statement (Form S-1 No. 333-224493) and related Prospectus of Scholar Rock Holding Corporation for the registration of 6,164,000 shares of its common stock.

/s/ Ernst & Young LLP

Boston, Massachusetts
May 14, 2018

QuickLinks

[Exhibit 23.1](#)

[Consent of Independent Registered Public Accounting Firm](#)