UNITED STATES
SECURITIES AND EXCHANGE COMMISSION

Washington, D.C. 20549

FORM 8-K

CURRENT REPORT
Pursuant to Section 13 or 15(d)
of the Securities Exchange Act of 1934

Date of Report (Date of earliest event reported): **August 13, 2018**

SCHOLAR ROCK HOLDING CORPORATION

(Exact name of registrant as specified in its charter)

Delaware	**001-38501**	**82-3750435**
(State or other jurisdiction of incorporation)	(Commission File Number)	(I.R.S. Employer Identification No.)

620 Memorial Drive, 2nd Floor
Cambridge, MA 02139
(Address of principal executive offices, including zip code)

(857) 259-3860
(Registrant's telephone number, including area code)

Not Applicable
(Former Name or Former Address, if Changed Since Last Report)

Check the appropriate box below if the Form 8-K filing is intended to simultaneously satisfy the filing obligation of the registrant under any of the following provisions:

☐ Written communications pursuant to Rule 425 under the Securities Act (17 CFR 230.425)

☐ Soliciting material pursuant to Rule 14a-12 under the Exchange Act (17 CFR 240.14a-12)

☐ Pre-commencement communications pursuant to Rule 14d-2(b) under the Exchange Act (17 CFR 240.14d-2(b))

☐ Pre-commencement communications pursuant to Rule 13e-4(c) under the Exchange Act (17 CFR 240.13e-4(c))

Indicate by check mark whether the registrant is an emerging growth company as defined in Rule 405 of the Securities Act of 1933 (§ 230.405 of this chapter) or Rule 12b-2 of the Securities Exchange Act of 1934 (§ 240.12b-2 of this chapter).

Emerging growth company ☒

If an emerging growth company, indicate by check mark if the registrant has elected not to use the extended transition period for complying with any new or revised financial accounting standards provided pursuant to Section 13(a) of the Exchange Act. ☒

Item 7.01. Regulation FD Disclosure.

The management of Scholar Rock Holding Corporation will participate in the Wedbush PacGrow Healthcare Conference in New York, NY on Tuesday, August 14, 2018 with a presentation at 1:55 p.m. ET.

A copy of the presentation slide deck that will be presented at the Wedbush PacGrow Healthcare Conference is being furnished as Exhibit 99.1 to this Report on Form 8-K. A live webcast of the Wedbush PacGrow presentation may be accessed by visiting the Investors & Media section of the Scholar Rock website at http://investors.scholarrock.com.

The information in this Report on Form 8-K and Exhibit 99.1 attached hereto is intended to be furnished and shall not be deemed "filed" for purposes of Section 18 of the Securities Exchange Act of 1934, as amended (the "Exchange Act"), or otherwise subject to the liabilities of that section, nor shall it be deemed incorporated by reference in any filing under the Securities Act of 1933, as amended, or the Exchange Act, except as expressly set forth by specific reference in such filing.

Item 9.01. Financial Statements and Exhibits.

(d) Exhibits

99.1 Presentation Slide Deck, furnished hereto.

SIGNATURES

Pursuant to the requirements of the Securities Exchange Act of 1934, the registrant has duly caused this report to be signed on its behalf by the undersigned hereunto duly authorized.

Scholar Rock Holding Corporation

Date: August 13, 2018

By: /s/ Junlin Ho
Junlin Ho
VP, Head of Corporate Legal

SCHOLAR ROCK

From New Insights to New Medicines

August 2018

Disclaimers

Various statements in this presentation concerning Scholar Rock's future expectations, plans and prospects, including without limitation, Scholar Rock's expectations regarding its strategy, its product candidate selection and development timing, including timing for the initiation of and reporting results from its clinical trials for its product candidates, its product candidate selection and development timing, its disease indication selection timing, its management team capabilities, and the ability of SRK-015 to affect the treatment of patients suffering from SMA either as a monotherapy or in conjunction with the current standard of care, constitute forward-looking statements for the purposes of the safe harbor provisions under The Private Securities Litigation Reform Act of 1995. The use of words such as "may," "might," "will," "should," "expect," "plan," "anticipate," "believe," "estimate," "project," "intend," "future," "potential," or "continue," and other similar expressions are intended to identify such forward looking statements. Actual results may differ materially from those indicated by these forward-looking statements as a result of various important factors, including, without limitation, Scholar Rock's ability to provide the financial support and resources necessary to identify and develop product candidates on the expected timeline, competition from others developing products for similar uses, Scholar Rock's ability to obtain, maintain and protect its intellectual property, Scholar Rock's dependence on third parties for development and manufacture of product candidates including to supply any clinical trials, and Scholar Rock's ability to manage expenses and to obtain additional funding when needed to support its business activities and establish and maintain strategic business alliances and new business initiatives as well as those risks more fully discussed in the section entitled "Risk Factors" in the Quarterly Report on Form 10-Q for the quarter ended June 30, 2018, which is on file with the Securities and Exchange Commission, as well as discussions of potential risks, uncertainties, and other important factors in Scholar Rock's subsequent filings with the Securities and Exchange Commission. Any forward-looking statements represent Scholar Rock's views only as of today and should not be relied upon as representing its views as of any subsequent date. Scholar Rock explicitly disclaims any obligation to update any forward-looking statements unless required by law.

Scholar Rock's Mission

Leverage our newly elucidated understanding of growth factor activation to develop highly selective therapies for underserved patients

SCHOLAR ROCK

Compelling Strategy

- Utilize our proprietary platform to unlock the therapeutic potential of targeting growth factor signaling in the disease microenvironment
- Focus on biologically validated growth factors
- Selectively seek strategic collaborations to maximize value of platform and pipeline
- Deliver novel therapies to underserved patients suffering from serious diseases

Robust Pipeline

- Advance pipeline focused on neuromuscular disorders, cancer, fibrosis and anemia
- Initiated Phase 1 clinical trial for lead product candidate, SRK-015, in May 2018
- Plan to initiate Phase 2 proof-of-concept trial for SRK-015 in patients with later-onset spinal muscular atrophy in 1Q19
- Identify next indication for SRK-015 in 1H19
- Nominate TGFβ1 product candidate and lead indication by the end of 1H19
    - Targeting indication in oncology, immuno-oncology or fibrosis
- Explore additional indications for existing and emerging product candidates

Experienced Management, Board and Founders

- Expertise in drug discovery, development, and commercialization
- Highly respected scientific founders – Dr. Timothy Springer and Dr. Leonard Zon

Highly Experienced Leadership Team

NAGESH MAHANTHAPPA, PHD
CEO & President

ELAN EZICKSON
COO & Head, Corporate Development

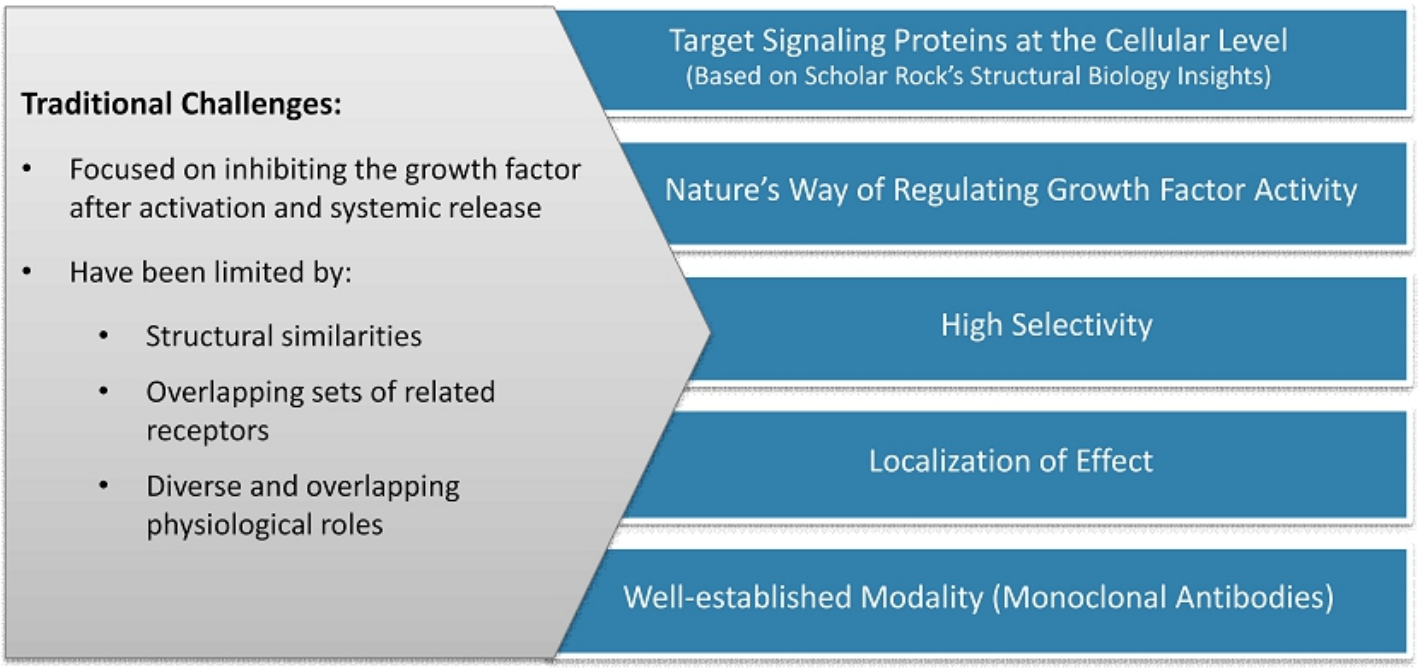
YUNG CHYUNG, MD
Chief Medical Officer

ALAN BUCKLER, PHD
Chief Scientific Officer

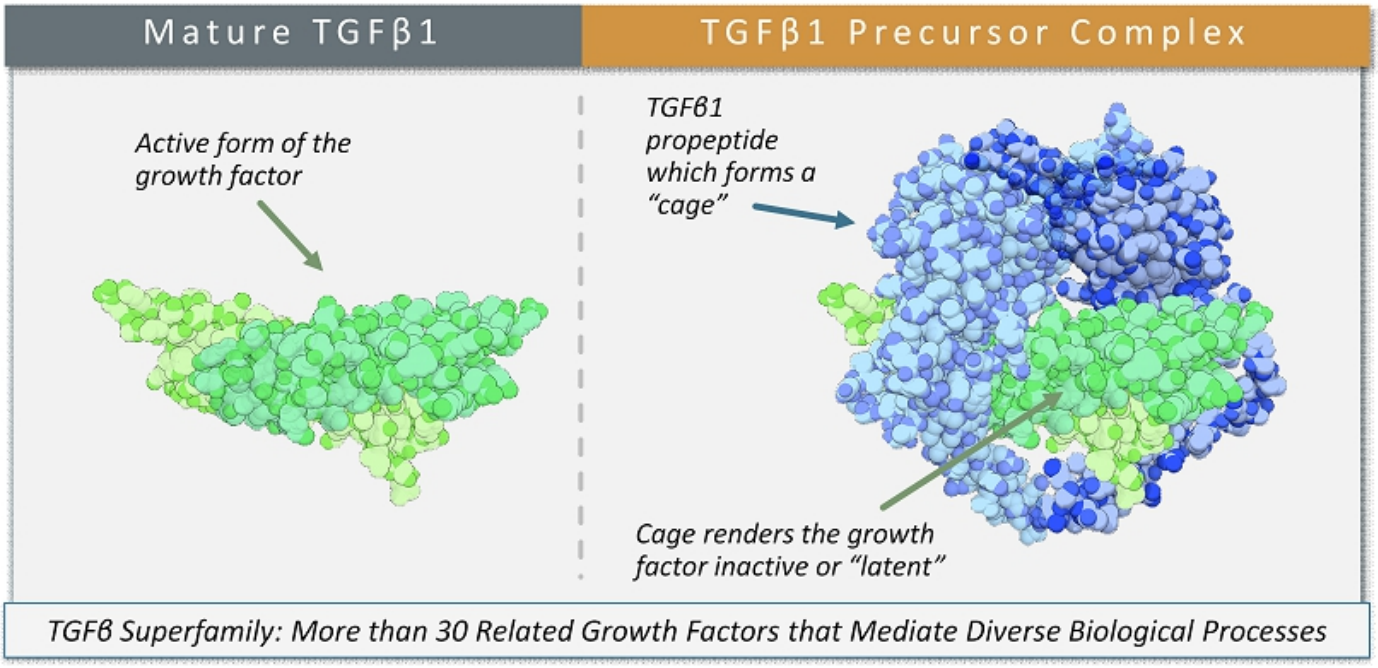
RHONDA CHICKO, CPA
Chief Financial Officer

AVILA · Alnylam · AVEO · Biogen · Dyax · genzyme · Biogen · NOVARTIS · HARVARD MEDICAL SCHOOL · editas MEDICINE · Ironwood

Scholar Rock's Solution to Traditional Challenges



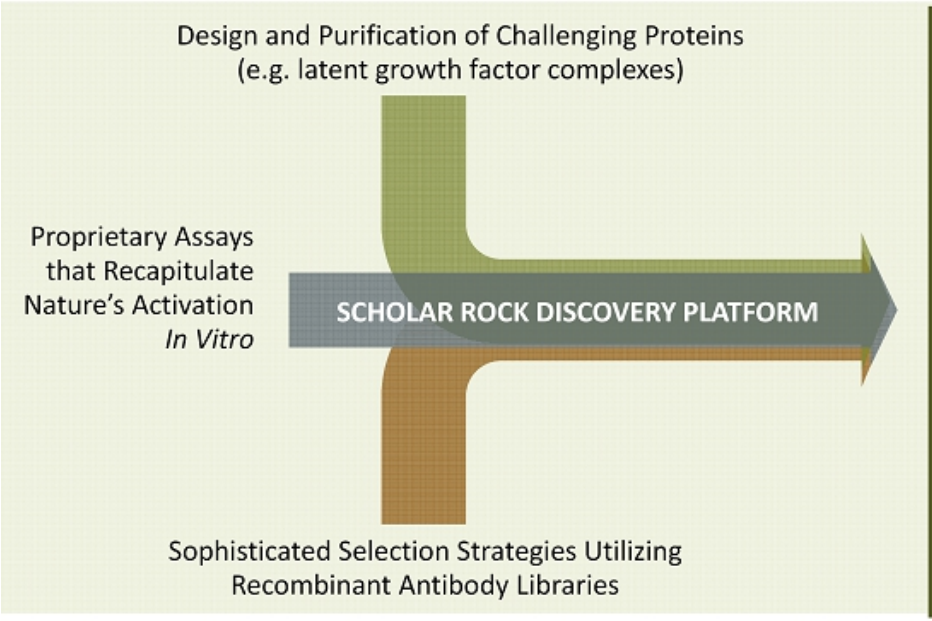
Nature's Activation Machinery



TGFβ Superfamily: More than 30 Related Growth Factors that Mediate Diverse Biological Processes

 Shi, M., et al. Nature 474, 7351, 343-9 (2011)

Proprietary Platform to Target Growth Factor Activation



Broad IP Portfolio Covering Compositions and Methods

Exemplified by

- **US Patent 9,758,576**
- **Issued in September 2017 (expiry in 2034)**
- **Covers monoclonal antibodies that inhibit the activation of the myostatin precursor**

Robust Pipeline Portfolio

PROGRAM		STAGE OF DEVELOPMENT					STATUS	
Target	Indication	Late-Stage Discovery	Preclinical	Phase 1	Phase 2	Phase 3	Worldwide Rights	Next Anticipated Milestone
SRK-015 Program								
Latent Myostatin	Spinal Muscular Atrophy						Scholar Rock	Phase 1 Trial Ongoing
Latent Myostatin	Additional Myostatin-Related Disorders						Scholar Rock	1H:2019 – Identify Next Indication
TGFβ1 Program								
Context-Independent								
Latent TGFβ1	Oncology/Immuno-oncology; Fibrosis						Scholar Rock	1H:2019 – Nominate Product Candidate
Context-Dependent								
Latent TGFβ1 / GARP	Oncology/Immuno-oncology						Janssen Biotech, Inc.	
Latent TGFβ1 / GARP & LRRC33	Oncology/Immuno-oncology						Scholar Rock	
Latent TGFβ1 / LRRC33	Oncology/Immuno-oncology						Scholar Rock	
Latent TGFβ1 / LTBP1 & LTBP3	Fibrosis						Scholar Rock	
BMP6 Program								
BMP6 Signaling Pathway	Anemia						Scholar Rock	

SRK-015: Inhibitor of Myostatin Activation

Potential First Muscle-Directed Therapy for SMA

SCHOLAR ROCK

Spinal Muscular Atrophy (SMA): Severe and Debilitating Disorder

Overview

Rare neuromuscular disorder

A child with SMA is born roughly every day in the U.S.

Caused by mutation or deletion in SMN1 gene leading to loss of motor neurons

Symptoms

Debilitating muscle atrophy and loss of motor function

Difficulty sitting, walking, raising arm, breathing, swallowing etc; shortened life expectancy

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Symptoms range from very severe to milder depending on SMA type

Treatment Options

First SMA therapy: nusinersen
(SMN upregulator, FDA approved 2016)

Spinal stabilization surgery, tracheostomy/ventilator, feeding tube placement

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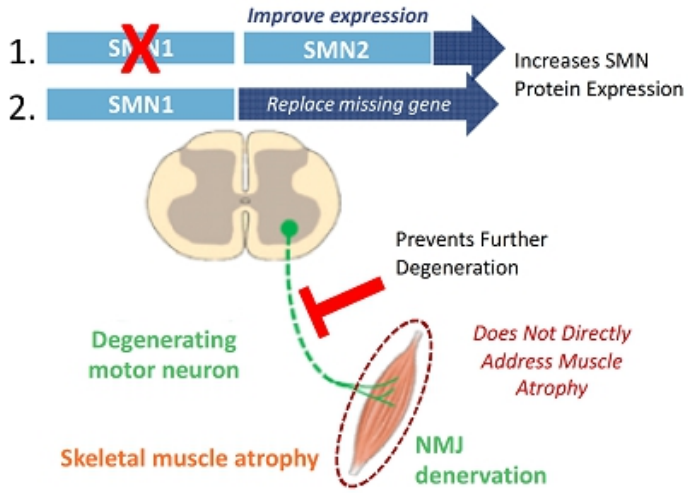
Significant unmet need in muscle function

SMN Upregulators Do Not Directly Address Muscle Atrophy

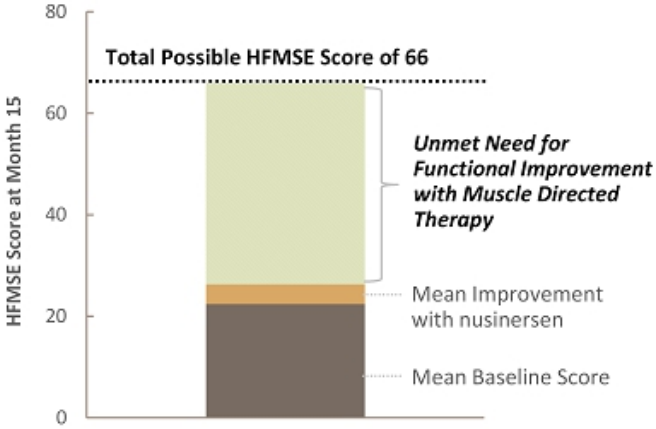
Significant Unmet Need Remains Despite Current Therapeutic Strategies



Adapted from images that were courtesy of the SMA Foundation

Muscle Function in SMA (Human)

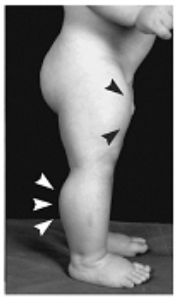
Hammersmith Functional Motor Scale Expanded (HFMSE)



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

SRK-015: Muscle-Directed Approach to Treating SMA

Vertebrates lacking the myostatin gene are healthy and display increased muscle mass and strength



- Myostatin is a genetically-validated, negative regulator of muscle mass expressed in skeletal muscle tissue
- There has been high interest from pharma as a potential drug target
- Clinical trial results with traditional systemic inhibitors of mature myostatin (or its receptor) have been mixed

Differentiated approach with SRK-015:

Inhibition of myostatin activation to potentially improve muscle function

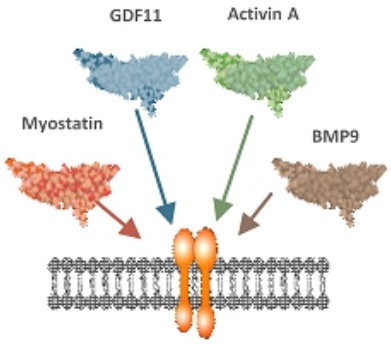
SRK-015: Aligning Therapeutic Approach with Myostatin Biology

Optimal Setting for Myostatin Inhibition		Key Characteristics of Spinal Muscular Atrophy (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

Traditional Approaches Can Raise Significant Safety Concerns

Traditional Approaches Lack Selectivity

- Most inhibitors of active myostatin also inhibit GDF11 and may inhibit other growth factors as well
- Antibodies to ActRIIb and ligand trap approaches inhibit signaling of multiple ligands



Potential Consequences

Developmental:

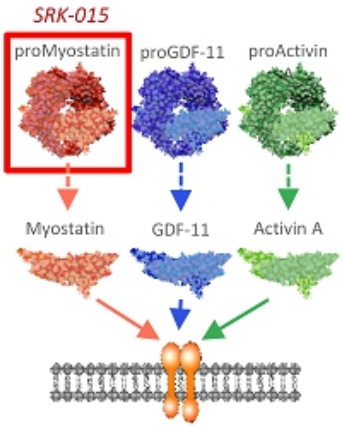
- Loss of GDF11 signaling in embryonic mice resulted in:
  - Kidney agenesis
  - Skeletal transformations

Clinical:

- Non-specific ligand trap approach resulted in nosebleeds, gum bleeds, and small dilated blood vessels within the skin attributed to inhibition of BMP9 signaling
- Blockade of ActRIIB suppressed FSH levels in postmenopausal and premenopausal women attributed to inhibition of activin signaling

SRK-015: High Selectivity by Targeting Precursor Form of Myostatin

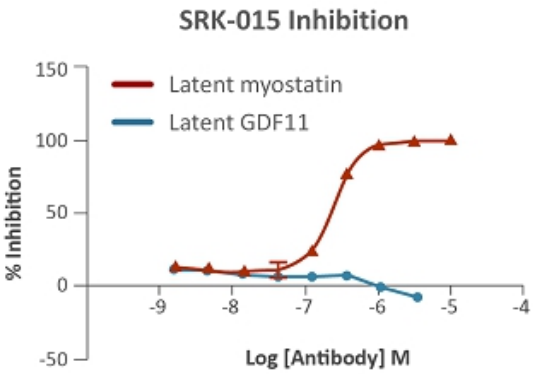
Scholar Rock Approach



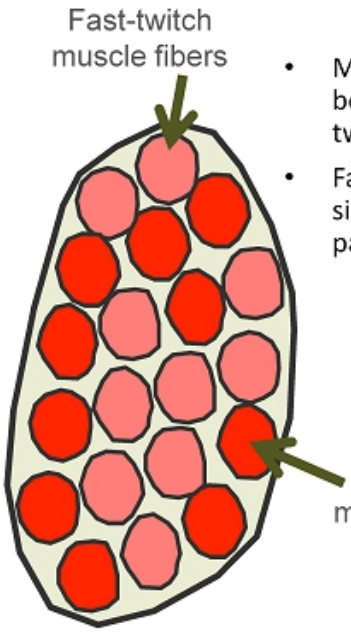
Exquisite Selectivity for Myostatin

	SRK-015 binding
ProMyostatin	++
Latent Myostatin	++
Myostatin	-
ProGDF11	-
GDF11	-
ProActivin A	-
Activin A	-
BMP9	-
BMP10	-
TGFβ1	-

Specific Inhibition of Myostatin Activation



Fast-Twitch Muscle Fibers May Be Preferentially Benefited by SRK-015

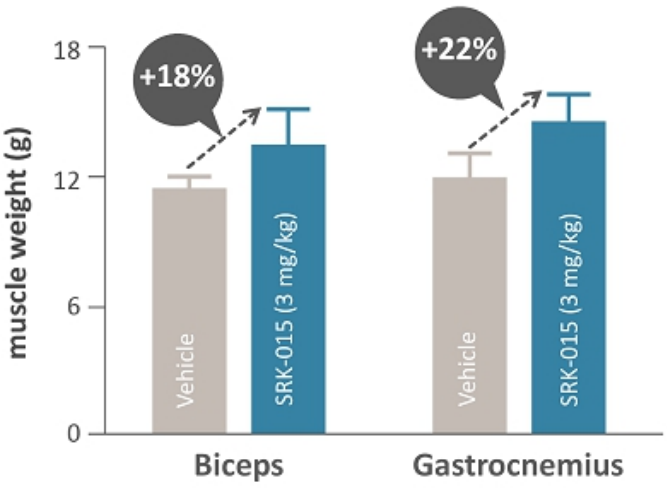


Muscle cross section

- Muscle is comprised of both slow-twitch and fast-twitch fibers
- Fast-twitch fibers significantly atrophy in patients with SMA

Healthy Non-Human Primate (Cyno)

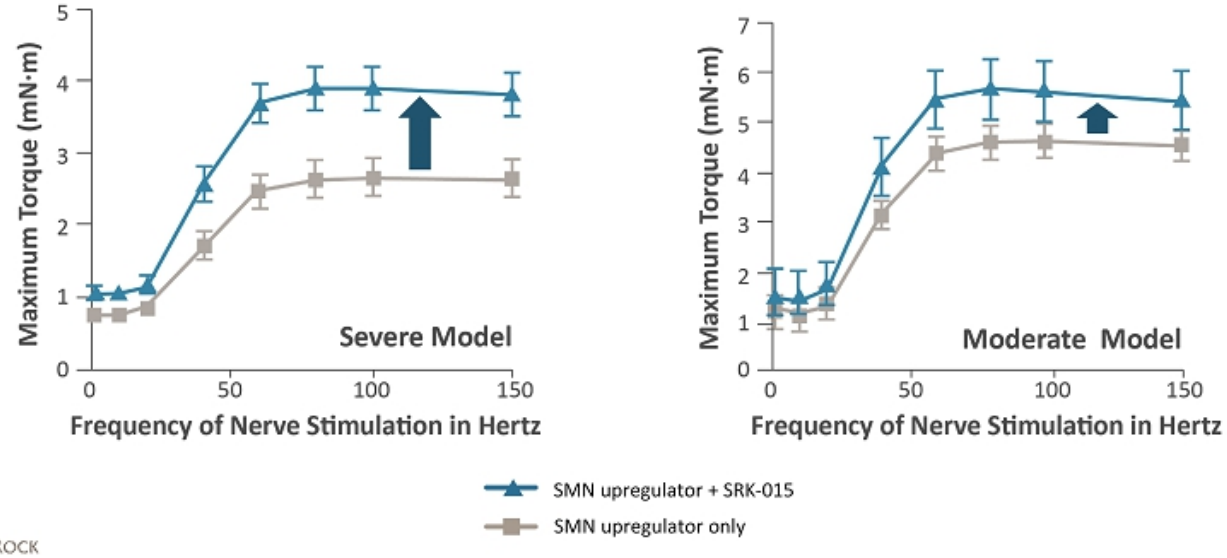
Pronounced Effect on Fast-Twitch Muscle



SRK-015 Demonstrates Potential Benefits Across SMA Severities

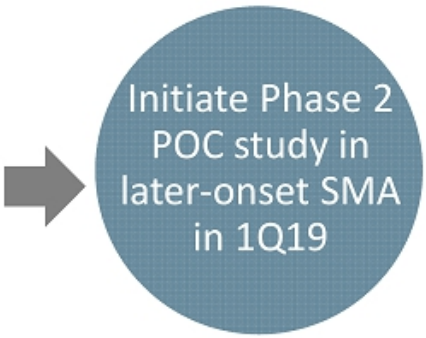
Genetic Model of SMA ("Δ7 Mouse") Demonstrate Improved *In Vivo* Muscle Force Generation

Strength of Plantarflexor Muscle Group



SRK-015 Phase 1 Trial Design

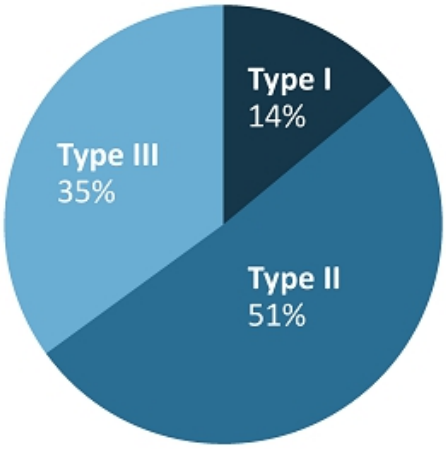
Design	• Double-blind, placebo-controlled • 3:1 randomization • Single- and multiple-ascending dose
Subjects	• Adult healthy volunteers (ages 18-55) • Anticipate enrolling ~60 subjects total
Key Objectives	• Safety and tolerability of SRK-015 IV • PK/PD • Immunogenicity
Status	• Enrollment and dosing ongoing



SRK-015 Opportunity in Spinal Muscular Atrophy

Overall Prevalence of 30,000-35,000 in U.S. and Europe

RELATIVE PREVALENCE AMONG PATIENTS LIVING WITH SMA



Type I:

- Infant-onset; often fatal

Type II and non-ambulatory type III:

- Later-onset but still early childhood
- Severe deficits in motor function

Potential to use SRK-015 in conjunction with current standard of care

Ambulatory type III:

- Limited mobility and substantial morbidity

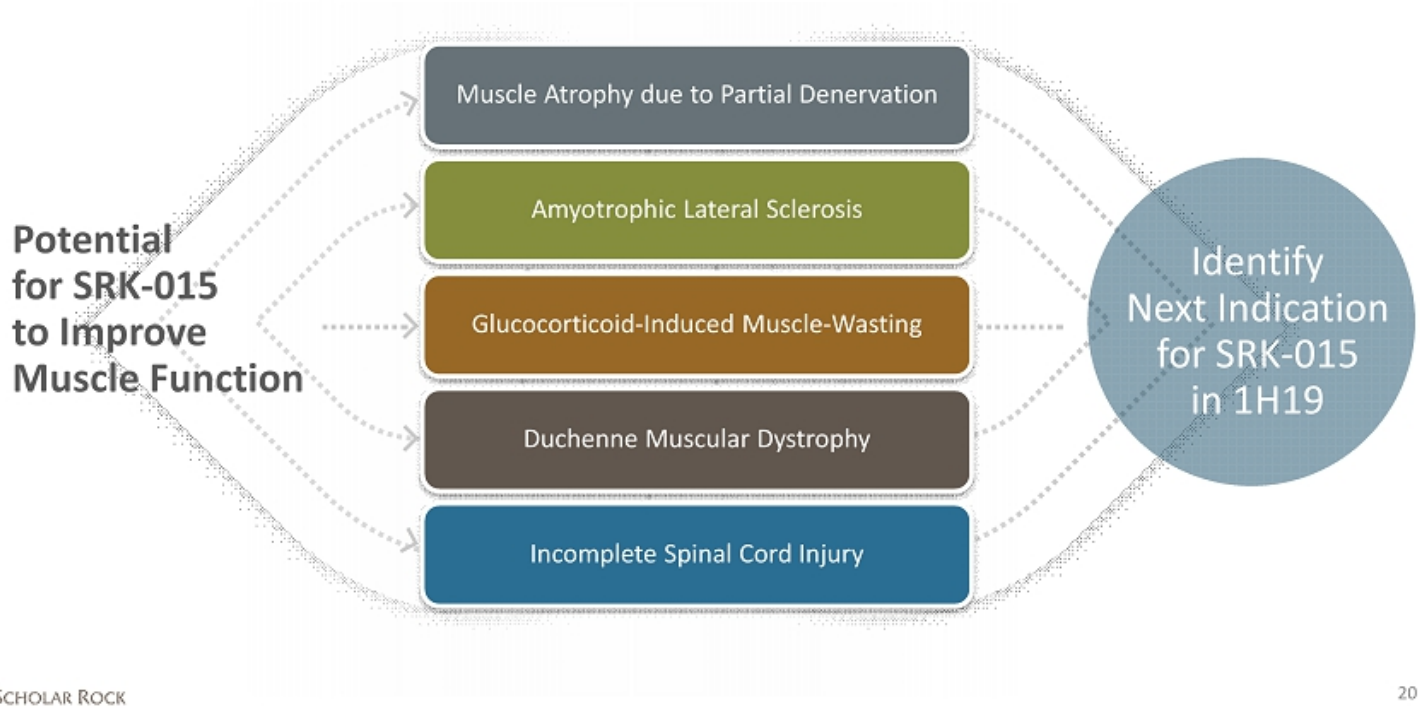
Potential to use SRK-015 as monotherapy

Type IV:

- Population not well-characterized

 Lally et al, Orphanet Journal of Rare Diseases, 2017

Examples of Potential Opportunities for SRK-015 in Additional Muscle-Wasting Disorders



Recent Achievements and Upcoming Milestones

- ✓ Orphan Drug Designation granted by FDA
- ✓ Completed IND-enabling toxicology studies
- ✓ IND in SMA submitted to FDA in March and cleared in April 2018
- ✓ Initiated Phase 1 clinical trial in healthy volunteers in May 2018
  - ✓ Advanced to multiple-ascending dose portion of trial
- ❑ Initiate Phase 2 proof-of-concept study in patients with later-onset SMA in 1Q19
  - ❑ Top-line results from Phase 2 proof-of-concept study expected in 2H19
- ❑ Identify next indication for SRK-015 in 1H19

TGFβ1: Significant Opportunities in Oncology/Immuno-oncology and Fibrosis

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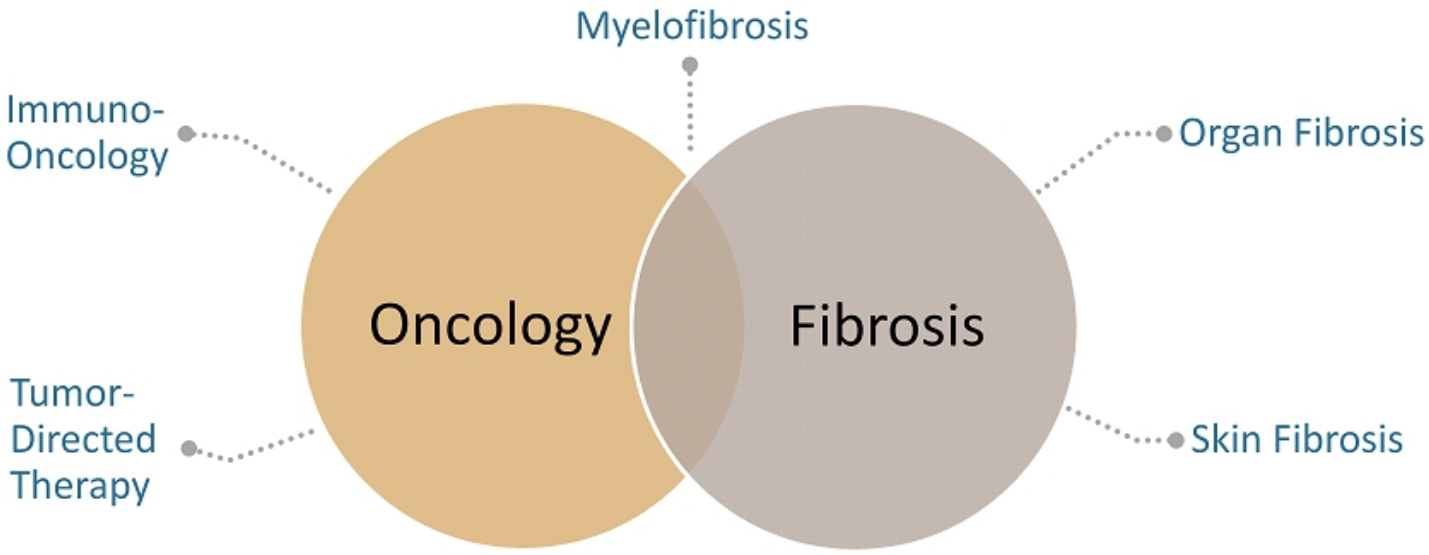
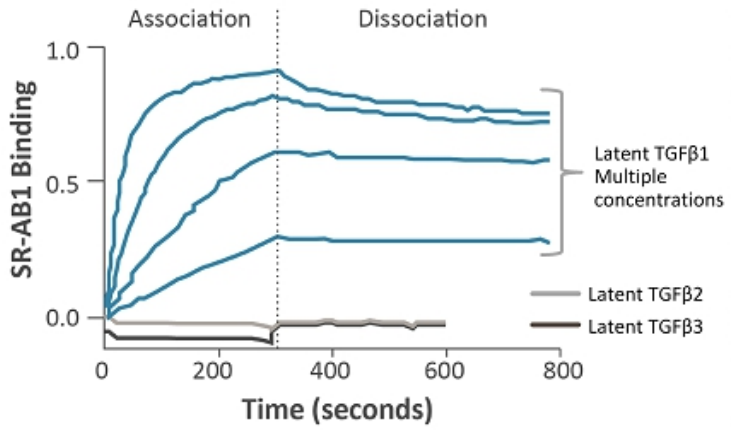
TGFβ1 Plays Central Role in Multiple Diseases with Unmet Need

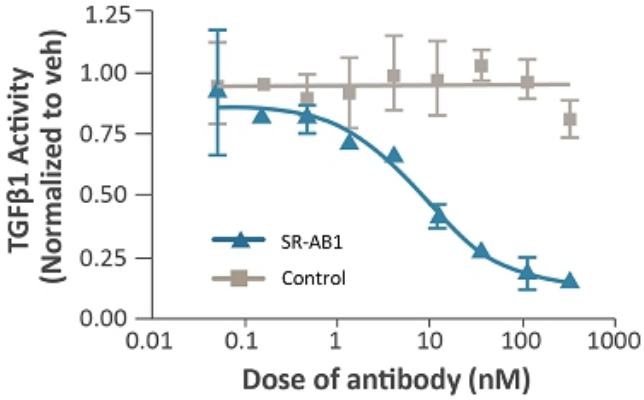


Scholar Rock TGFβ1 Activation Inhibitor Demonstrates Selectivity and In Vitro Inhibitory Activity

SR-AB1 Binds to Latent TGFβ1 with High Affinity

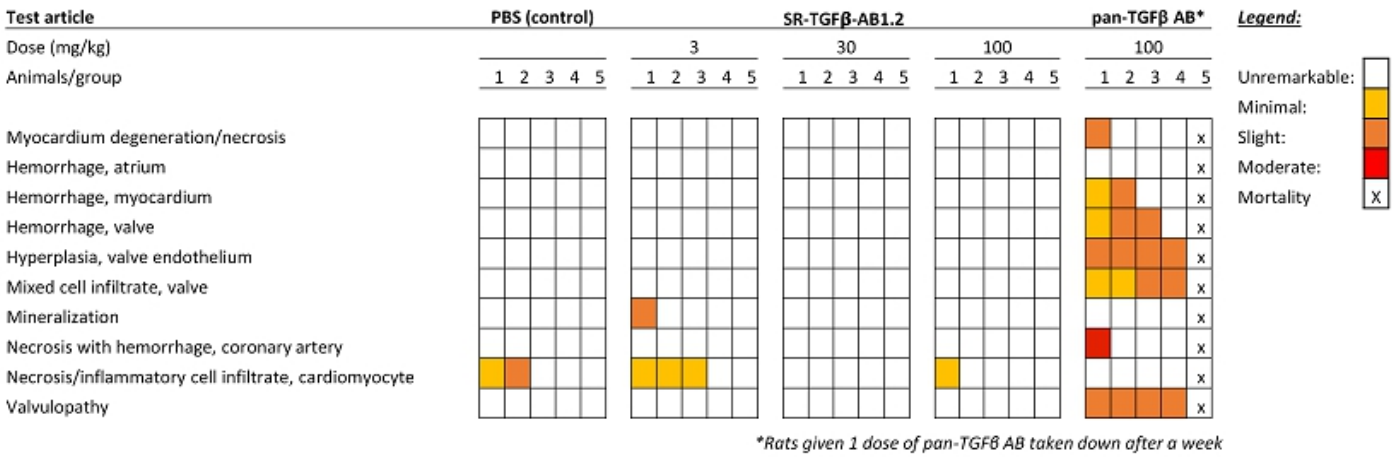


SR-AB1 Shows Dose-Dependent Inhibition of TGFβ1 Activity



Scholar Rock Selective Inhibitor Avoids Cardiac Valve Toxicity Induced by Pan-TGFβ Inhibitors in Four-Week Safety Study

Heart Example, microscopic findings

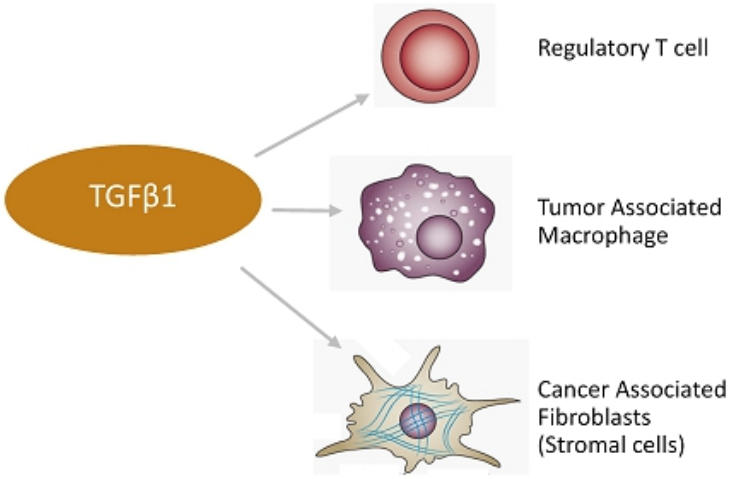


**Rats given 1 dose of pan-TGFβ AB taken down after a week*

No drug related toxicities identified with Scholar Rock selective inhibitor up to 100 mg/kg QW (highest dose tested)

Inhibition of TGFβ1: Multipronged Approach for Immuno-Oncology

TGFβ1 is a key driver of immune system evasion by cancer cells



Nature (on-line), Feb. 14, 2018
doi:10.1038/nature25501

TGFβ attenuates tumour response to PD-L1 blockade by contributing to exclusion of T cells

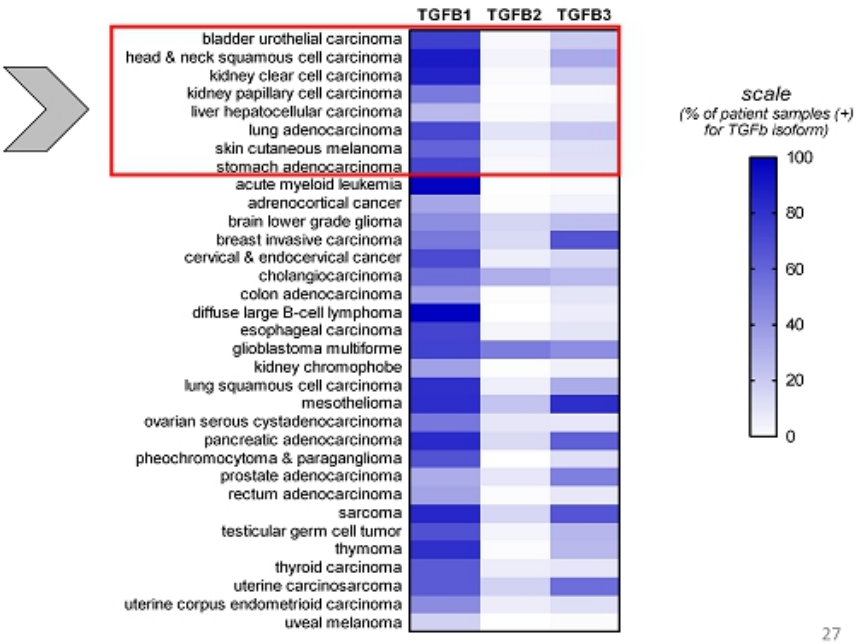
Sanjeev Mariathasan[1]*, Shannon J. Turley[1]*, Dorothee Nickles[1]*, Alessandra Castiglioni[1], Kobe Yuen[1], Yulei Wang[1], Edward E. Kadel III[2], Hartmut Koeppen[1], Jillian L. Astarita[3], Rafael Cubas[2], Suchit Jhunjhunwala[2], Romain Banchereau[1], Yagai Yang[1], Yinghui Guan[1], Cecile Chalouni[1], James Ziai[1], Yasin Şenbabaoğlu[1], Stephen Santoro[1], Daniel Sheinson[1], Jeffrey Hung[1], Jennifer M. Giltnane[1], Andrew A. Pierce[1], Kathryn Mesh[1], Steve Lianoglou[1], Johannes Riegler[1], Richard A. D. Carano[1], Pontus Eriksson[2], Mattias Höglund[2], Loan Somarriba[3], Daniel L. Halligan[3], Michiel S. van der Heijden[4], Yohann Loriot[5], Jonathan E. Rosenberg[6], Lawrence Fong[7], Ira Mellman[3], Daniel S. Chen[1], Marjorie Green[1], Christina Derleth[1], Gregg D. Fine[1], Priti S. Hegde[1], Richard Bourgon[1] & Thomas Powles[8]

- Pathway analysis points to TGFβ1 as major determinant of resistance to anti-PD-L1 (atezolizumab)
- TGFβ1 creates 'immune excluded' tumor microenvironment
- Anti-TGFβ antibody enhances anti-PDL1 treatment response in syngeneic EMT-6 tumor model

TGFβ1 is the Predominant Isoform in Most Human Tumors

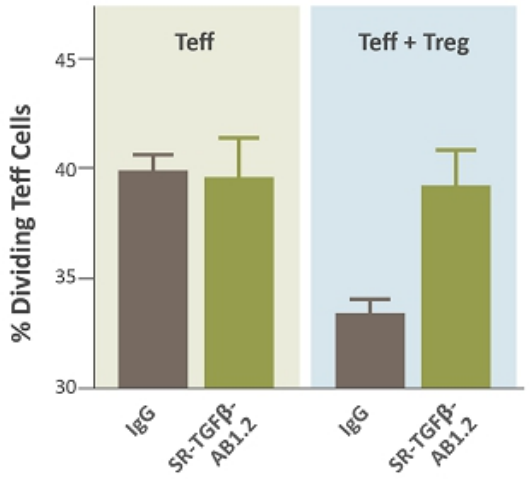
The Cancer Genome Atlas RNAseq analysis: >10,000 samples spanning 33 tumor types

- TGFβ1 prevalent in human cancers for which checkpoint therapies are approved
- Expression data for most tumor types suggest that TGFβ signaling mainly driven by TGFβ1



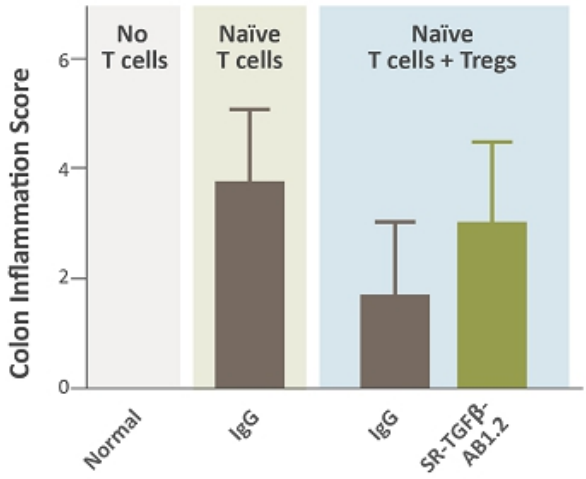
Immuno-Oncology: Inhibition of Latent TGFβ1 Activation Suppresses Treg Activity

SR-AB1.2 suppresses Human Treg Activity Ex Vivo



Human Treg Suppression Assay

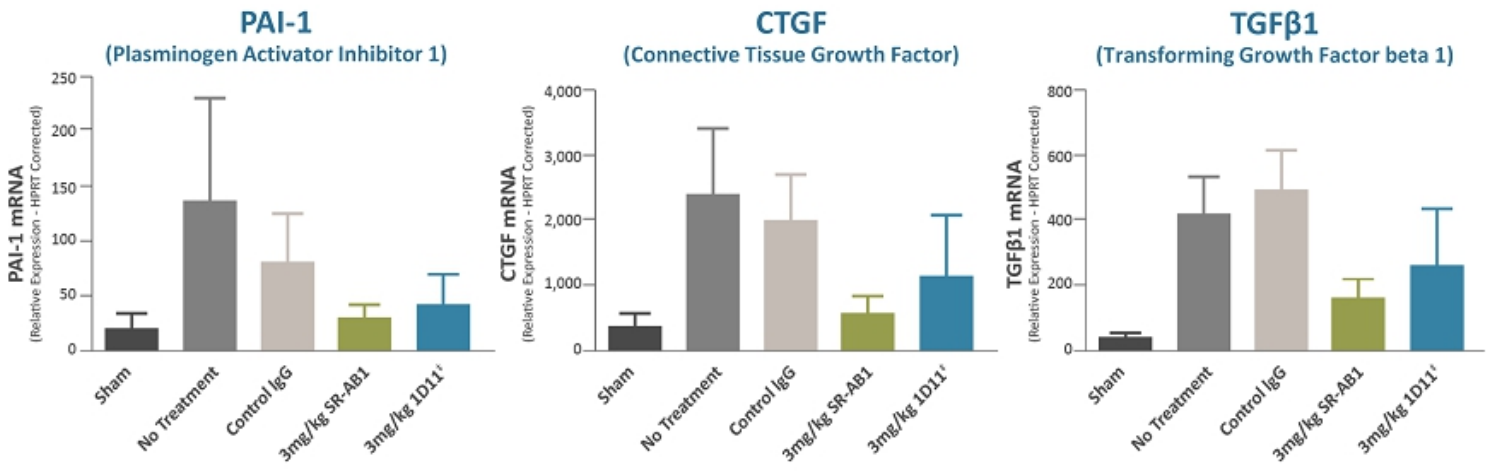
SR-AB1.2 suppresses mouse Treg Activity In Vivo



T cell Transfer Colitis Model

TGFβ is a Central Driver of Fibrotic Disease

Acute Kidney Fibrosis Model (mouse UUO): Gene Expression

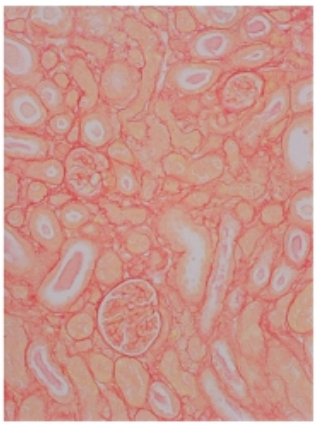


‡1D11 is an inhibitor of all three isoforms of mature TGFß

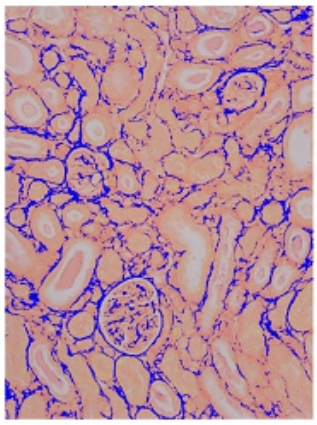
Inhibition of Latent TGFβ1 Activation Prevents Fibrosis

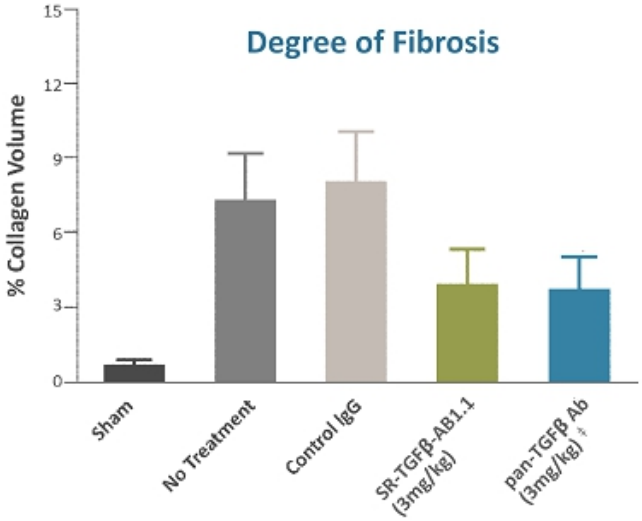
Acute Kidney Fibrosis Model (mouse UUO): Histomorphometry

Picrosirius red stain



Blue segmentation mask

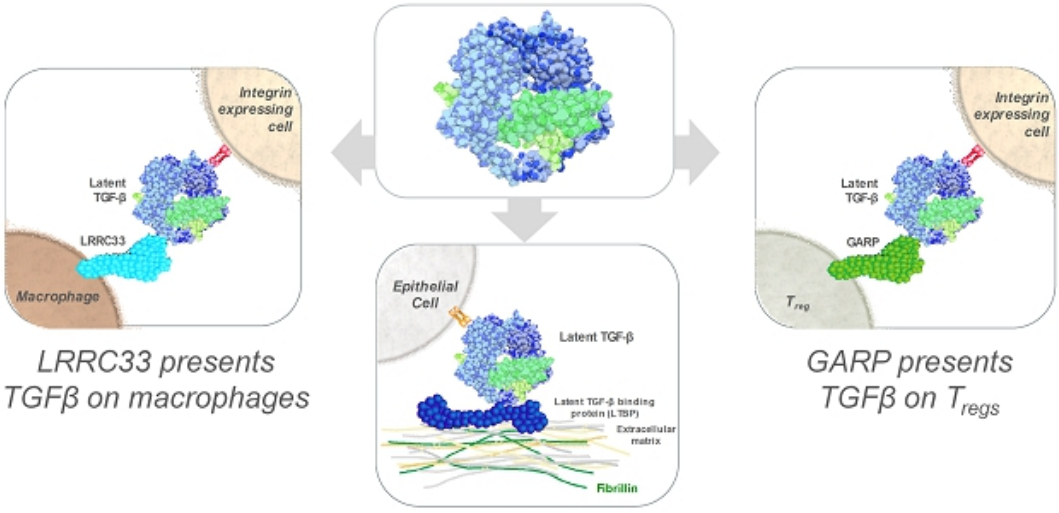




†1D11 is an inhibitor of all three isoforms of mature TGFβ

Targeting Latent TGFβs Creates Multiple "Handles" For Selectivity

Context-Dependent Inhibition of TGFβ1



LRRC33 presents TGFβ on macrophages

LTBP1 & 3 present TGFβ in connective tissue

GARP presents TGFβ on T_{regs}

Recent Achievements and Upcoming Milestones

- ✓ Observed inhibition of TGFβ1 activation in vitro and immunomodulatory and anti-fibrotic activity in multiple in vivo disease models
- ✓ Completed 28-day pilot toxicology study of our leading antibody
  - ✓ Have not observed any drug-related toxicity
- ❑ Actively evaluating our selective inhibitors of the activation of TGFβ1 in multiple disease models
- ❑ Nominate product candidate and lead indication by the end of 1H19
- ❑ Continue to advance active discovery programs for context-dependent inhibition of TGFβ1

Recent Achievements

- ✓ Initiated Phase 1 clinical trial of SRK-015 in May 2018
  - ✓ Advanced to multiple-ascending dose portion of trial
- ✓ IND for SRK-015 in SMA was submitted to FDA in March and cleared in April 2018
- ✓ FDA granted Orphan Drug Designation for SRK-015 for the treatment of SMA
- ✓ Issuance of U.S. Patent 9,758,576 covering myostatin activation inhibitors; exclusivity May 2034
- ✓ Raised $86M in gross proceeds from IPO in May 2018

Upcoming Milestones

- ❑ Initiate Phase 2 proof-of-concept study of SRK-015 in patients with later-onset SMA in 1Q19
- ❑ Identify next indication for SRK-015 in 1H19
- ❑ Nominate product candidate and lead indication in TGFβ1 program by the end of 1H19
- ❑ Top-line results from Phase 2 proof-of-concept study of SRK-015 in patients with later-onset SMA in 2H19

Building for Success