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 8, 2024

Scholar Rock Holding Corporation
(Exact Name of Registrant as Specified in Charter)

Delaware	**001-38501**	**82-3750435**
(State or Other Jurisdiction of Incorporation)	(Commission File Number)	(I.R.S. Employer Identification Number)

301 Binney Street, 3rd Floor, Cambridge, MA 02142
(Address of Principal Executive Offices) (Zip Code)

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

(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))

Securities registered pursuant to Section 12(b) of the Act:

Title of each class	Trading Symbol(s)	Name of each exchange on which registered
Common Stock, par value $0.001 per share	SRRK	Nasdaq Global Select Market

Indicate by check mark whether the registrant is an emerging growth company as defined in Rule 405 of the Securities Act of 1933 (17 CFR §230.405) or Rule 12b-2 of the Securities Exchange Act of 1934 (17 CFR §240.12b-2). 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 2.02. Results of Operations and Financial Condition.

On August 8, 2024, Scholar Rock Holding Corporation (the "Company") issued a press release announcing its financial and operating results for the quarter ended June 30, 2024. A copy of the press release is being furnished as Exhibit 99.1 to this Report on Form 8-K.

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 8.01. Other Events.

A copy of the Company's current corporate slide presentation is being filed herewith as Exhibit 99.2 to this Report on Form 8-K and is incorporated herein by reference. The Company undertakes no obligation to update, supplement or amend the materials attached hereto as Exhibit 99.2.

Item 9.01. Financial Statements and Exhibits.

(d) Exhibits

Exhibit No.	Description
99.1	Press Release issued by the Company on August 8, 2024, furnished hereto.
99.2	Presentation Slide Deck.
104	Cover Page Interactive Data File (embedded within the Inline XBRL document).

SIGNATURE

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 8, 2024

By: /s/ Junlin Ho
Junlin Ho
General Counsel and Corporate Secretary

Scholar Rock Reports Second Quarter 2024 Financial Results and Highlights Business Progress

- *Remains on track to report topline data from pivotal Phase 3 SAPPHIRE trial in patients with Spinal Muscular Atrophy (SMA) in 4Q 2024*
- *New data from Phase 2 TOPAZ extension study in patients with nonambulatory SMA showed sustained clinical benefit over 48 months, a continued favorable safety profile with no new safety findings; patient retention rate of over 90%*
- *Phase 2 EMBRAZE proof-of-concept trial enrolling ahead of schedule, topline data expected in 2Q 2025*
- *Presented new SRK-439 preclinical data at American Diabetes Association's 84th Scientific Sessions (ADA) supporting the potential to contribute to a favorable body composition; increased lean mass and reduced fat mass regain following withdrawal from GLP-1 receptor agonist treatment*
- *Management to host update call today at 8:15 a.m. ET*

CAMBRIDGE, Mass.— (BUSINESS WIRE)— August 8, 2024—Scholar Rock (NASDAQ: SRRK), a late-stage biopharmaceutical company focused on advancing innovative treatments for spinal muscular atrophy (SMA), cardiometabolic disorders, and other serious diseases where protein growth factors play a fundamental role, today reported financial results and corporate updates for the second quarter ended June 30, 2024.

"Scholar Rock continues to execute across our portfolio of highly selective myostatin inhibition programs, further cementing our position as the global leader in harnessing the life-changing potential of TGF-beta superfamily biology," said Jay Backstrom, M.D., MPH, President & Chief Executive Officer of Scholar Rock. "With the only muscle-targeted program to demonstrate clinical proof-of-concept in SMA, our confidence in our lead program apitegromab continues to be supported by the clinical data generated over the past four years. At 48 months, over 90% of TOPAZ patients with nonambulatory SMA remained on apitegromab treatment on top of SMN therapy and we continued to observe sustained clinical benefit. We look forward to reporting topline data from the Phase 3 SAPPHIRE trial of apitegromab in SMA in the fourth quarter of this year."

Dr. Backstrom continued, "In addition, we are pleased with the progress of our cardiometabolic program. The enrollment of the Phase 2 proof-of-concept EMBRAZE study evaluating apitegromab in obesity has been advancing ahead of schedule and as a result, we are updating our guidance for topline data to the second quarter of 2025. We also presented new preclinical data supporting SRK-439's potential to help patients retain lean muscle mass at our investor event in May and at the ADA 84th Scientific Sessions in June. The hallmark of our approach in designing both apitegromab and SRK-439 is the exquisite selectivity for pro- and latent forms of myostatin. Our data in SMA suggest that this selectivity matters for patients, and we are excited to show how SRK-439 can become an integral component of the treatment and management of obesity helping to preserve lean muscle mass for sustainable and healthy weight loss management."

Company Highlights and Upcoming Milestones

SMA Program

Apitegromab is an investigational, fully human monoclonal antibody that inhibits myostatin activation by selectively binding the pro- and latent forms of myostatin in skeletal muscle and is being developed as a potential first muscle-targeted therapy for the treatment of SMA. Apitegromab is the only muscle-targeted therapy to show clinical proof-of-concept in SMA.

- **On track to report topline data from Phase 3 SAPPHIRE clinical trial in 4Q 2024**. If the trial is successful and apitegromab is approved, the Company expects to initiate a commercial product launch in 2025.
- **Reported that long-term apitegromab data continued to show substantial and sustained motor function improvements over 48 months[1].** The mean change in Hammersmith Functional Motor Scale (HFMSE) from baseline in nonambulatory patients (ages 2-21) on combination therapy (nusinersen and 20 mg/kg of apitegromab) was 5.3 points (95% CI: 1.5, 9.2; n=23), and for patients 2-12 was 6.4 points (95% CI: 1.8, 11.0, n=19). The mean change in RULM for the 2-21 age group was 3.6 points (95% CI: 2.0, 5.3; N=22) and for the 2-12 age group was 4.5 (95% CI: 2.7, 6.3; n=18). Of the 35 participants in the pooled nonambulatory population, 33 remained in the study over 4 years. The data analysis excluded the scores of 11 patients after undergoing scoliosis surgery, a known confounding factor for motor function assessment. Additional details will be discussed on the conference call this morning.

	12-Month Data	24-Month Data	36-Month Data	48-Month Data
Age 2-21 Years Mean Change from Baseline in HFMSE (95% Confidence Interval)	**3.6 points** (1.2, 6.0) n=32	**4.2 points** (1.9, 6.6) n=29	**4.0 points** (1.0, 6.9) n=28	**5.3 points** (1.5, 9.2) n=23
Age 2-12 Years Mean Change from Baseline in HFMSE (95% Confidence Interval)	**4.6 points** (1.8, 7.4) n=26	**5.2 points** (2.3, 8.0) n=23	**4.8 points** (1.3, 8.3) n=23	**6.4 points** (1.8, 11.0) n=19
Age 2-21 Years Mean Change from Baseline in RULM (95% Confidence Interval)	**1.3 points** (0.2, 2.3) n=31	**2.3 points** (1.2, 3.3) n=31	**2.4 points** (1.1, 3.7) n=27	**3.6 points** (2.0, 5.3) n=22
Age 2-12 Years Mean Change from Baseline in RULM (95% Confidence Interval)	**1.2 points** (0.1, 2.4) n=25	**2.2 points** (1.0, 3.5) n=25	**2.8 points** (1.4, 4.2) n=22	**4.5 points** (2.7, 6.3) n=18

1. For the 48-month evaluation, an observed case analysis was conducted using available data by analysis timepoint, censoring any HFMSE and RULM assessments after the patient received scoliosis surgery. The analysis population pooled the nonambulatory patients (Cohorts 2 and 3) and included patients receiving either low dose (2 mg/kg) or high dose (20 mg/kg) apitegromab (inclusive of patients in Cohort 3 who switched from 2 mg/kg to 20 mg/kg in Year 2). A total of 11 patients in the population had scoliosis surgery during the study and their data was excluded from any HFMSE or RULM assessments at 48 months. Visit windows were applied to utilize data from unscheduled or early termination visits if the patient was missing the HFMSE or RULM total score at the scheduled visit.

- **The ONYX open-label, multicenter extension study is ongoing.** The extension study is evaluating the long-term safety and efficacy of apitegromab in patients with Type 2 and Type 3 SMA who completed the TOPAZ or SAPPHIRE trials. More than 90 percent of patients on combination therapy in the TOPAZ study have completed 4 years of apitegromab treatment and enrolled into ONYX.

Cardiometabolic Program

SRK-439 is a novel, preclinical, investigational myostatin inhibitor that has high in vitro affinity for pro- and latent myostatin and maintains myostatin specificity (i.e., no GDF11 or Activin A binding), and is initially being developed for the treatment of obesity.

- **Presented preclinical data from the SRK-439 program.** In May, the Company announced new preclinical data comparing SRK-439 and an anti-activin receptor II (anti-ActRII) antibody that supported SRK-439's potential as best in class in preserving lean muscle mass in patients on GLP-1 receptor agonists (GLP-1 RAs). In June, the company presented new preclinical data at the American Diabetes Association 84th Scientific Sessions supporting the potential of SRK-439 to increase lean mass and contribute to a favorable body composition following withdrawal from GLP-1 RA treatment.
- **Initiated Phase 2 EMBRAZE proof-of-concept trial with apitegromab in combination with a GLP-1 receptor agonist (GLP-1 RA) in obesity in May.** The Phase 2 trial is a randomized, double-blind, placebo-controlled, multi-center study to evaluate the effect of apitegromab, a highly selective investigational myostatin inhibitor, to preserve muscle mass as an adjunctive therapy in overweight and obese adults who are taking a GLP-1 RA. Data are expected in the second quarter of 2025 and will be used to guide clinical development of SRK-439. The Company plans to file an IND for SRK-439 for the treatment of obesity in 2025.

Other Pipeline Updates

- **New SRK-181 data from the Phase 1 DRAGON proof-of-concept trial presented at the ASCO Annual Meeting in June.** SRK-181 is an investigational selective inhibitor of latent TGFβ-1 activation and developed with the aim of overcoming resistance to checkpoint therapy in patients with advanced cancer. Clinical data showed encouraging responses in heavily pretreated and anti-PD-(L)1 resistant patients across multiple tumor types. Enrollment of the DRAGON trial was completed in December 2023, and patients who remain on the study continue to be treated.
- **Published data on SRK-373 was featured on the cover of *Science Signaling* in July**. The article describes the selectivity of SRK-373 for LTBP-presented TGFβ-1, as well as efficacy data from two preclinical models that establish the feasibility of selectively targeting this particular form of TGFβ-1 for the treatment of fibrosis. SRK-373 is an investigational selective inhibitor of matrix associated TGFβ-1 in development for the treatment of fibrosis.

Second Quarter 2024 Financial Results

For the quarter ended June 30, 2024, net loss was $58.5 million or $0.60 per share compared to a net loss of $37.9 million or $0.47 per share for the quarter ended June 30, 2023.

- The Company did not record any revenue for the quarter ended June 30, 2024 or for the quarter ended June 30, 2023.
- Research and development expense was $42.4 million for the quarter ended June 30, 2024, compared to $26.9 million for the quarter ended June 30, 2023. The increase was primarily attributable to clinical trial and employee compensation costs.
- General and administrative expense was $17.1 million for the quarter ended June 30, 2024, compared to $12.2 million for the quarter ended June 30, 2023. The increase was due to employee-related costs.
- As of June 30, 2024, Scholar Rock had cash, cash equivalents, and marketable securities of approximately $190.5 million, which is expected to fund the Company's anticipated operating and capital expenditure requirements into the second half of 2025.

"Our year-to-date progress across our pipeline of industry-leading myostatin inhibition programs, combined with our highly experienced and disciplined team, provides us with a robust foundation for growth as we advance towards multiple milestones and our potential evolution into a commercial-stage company," said Ted Myles, Chief Operating Officer and Chief Financial Officer of Scholar Rock.

Conference Call Information

Management will provide an update on the Company and discuss second quarter 2024 results via conference call on Thursday, August 8 at 8:15 am ET. To access the live conference call, participants may register here. The live audio webcast of the call will be available under “Events and Presentations” in the Investor Relations section of the Scholar Rock website at http://investors.scholarrock.com. To participate via telephone, please register in advance here. Upon registration, all telephone participants will receive a confirmation email detailing how to join the conference call, including the dial-in number along with a unique passcode and registrant ID that can be used to access the call. An archived replay of the webcast will be available on the Company’s website for approximately 90 days.

About Apitegromab

Apitegromab is an investigational fully human monoclonal antibody inhibiting myostatin activation by selectively binding the pro- and latent forms of myostatin in the skeletal muscle. It is the first muscle-targeted treatment candidate to demonstrate clinical proof-of-concept in spinal muscular atrophy (SMA). Myostatin, a member of the TGFβ superfamily of growth factors, is expressed primarily by skeletal muscle cells, and the absence of its gene is associated with an increase in muscle mass and strength in multiple animal species, including humans. Scholar Rock believes that its highly selective targeting of pro- and latent forms of myostatin with apitegromab may lead to a clinically meaningful improvement in motor function in patients with SMA. The U.S. Food and Drug Administration (FDA) has granted Fast Track, Orphan Drug and Rare Pediatric Disease designations, and the European Medicines Agency (EMA) has granted Priority Medicines (PRIME) and Orphan Medicinal Product designations, to apitegromab for the treatment of SMA. The efficacy and safety of apitegromab have not been established and apitegromab has not been approved for any use by the FDA or any other regulatory agency.

About the Phase 3 SAPPHIRE Trial

SAPPHIRE is an ongoing randomized, double-blind, placebo-controlled, Phase 3 clinical trial evaluating the safety and efficacy of apitegromab in nonambulatory patients with Types 2 and 3 SMA who are receiving SMN-targeted therapy (either nusinersen or risdiplam). SAPPHIRE targeted enrolling approximately 156 patients aged 2-12 years old in the main efficacy population. These patients were randomized 1:1:1 to receive for 12 months either apitegromab 10 mg/kg, apitegromab 20 mg/kg, or placebo by intravenous (IV) infusion every 4 weeks. An exploratory population that targeted enrolling up to 48 patients aged 13-21 years old will also separately be evaluated. These patients were randomized 2:1 to receive either apitegromab 20 mg/kg or placebo. For more information about SAPPHIRE, visit www.clinicaltrials.gov. Apitegromab has not been approved for any use by the US FDA or any other health authority, and its safety and efficacy have not been established.

About EMBRAZE

EMBRAZE is a randomized, double-blind, placebo-controlled, Phase 2 proof-of-concept trial evaluating the efficacy, safety and pharmacokinetics of apitegromab in adults with a body mass index (BMI) of >27 (overweight) or a BMI of >30 (obese) and taking a GLP-1 RA (tirzepatide or semaglutide). The target enrollment of EMBRAZE is 100 subjects aged 18-65 who are overweight or obese without diabetes. As part of the study design, the treatment period is 24 weeks, and all subjects will receive a GLP-1 RA. In addition, all subjects will be randomized 1:1 to receive either apitegromab or placebo by intravenous (IV) infusion every four weeks during the 24-week treatment period. The primary endpoint is change from baseline at Week 24 in lean mass assessed by dual-energy X-ray absorptiometry. Secondary endpoints include additional weight loss measures, safety and tolerability, and pharmacokinetic outcomes. Exploratory endpoints at Weeks 24 and 32 include cardiometabolic parameters (e.g. HbA1c), body composition, and physical function.

About SRK-439

SRK-439 is a novel, preclinical, investigational myostatin inhibitor that has high in vitro affinity for pro- and latent myostatin and maintains myostatin specificity (i.e., no GDF11 or Activin-A binding), and is initially being developed for the treatment of cardiometabolic disorders, including obesity. Based on preclinical data, SRK-439 has the potential to

support healthier weight management by preserving lean mass during weight loss. The efficacy and safety of SRK-439 have not been established and SRK-439 has not been approved for any use by the FDA or any other regulatory agency.

About Scholar Rock

Scholar Rock is a biopharmaceutical company that discovers, develops, and delivers life-changing therapies for people with serious diseases that have high unmet need. As a global leader in the biology of the transforming growth factor beta (TGFβ) superfamily of cell proteins and named for the visual resemblance of a scholar rock to protein structures, the clinical-stage company is focused on advancing innovative treatments where protein growth factors are fundamental. Over the past decade, Scholar Rock has created a pipeline with the potential to advance the standard of care for neuromuscular disease, cardiometabolic disorders, cancer, and other conditions where growth factor-targeted drugs can play a transformational role.

Scholar Rock is the only company to show clinical proof-of-concept for a muscle-targeted treatment in spinal muscular atrophy (SMA). This commitment to unlocking fundamentally different therapeutic approaches is powered by broad application of a proprietary platform, which has developed novel monoclonal antibodies to modulate protein growth factors with extraordinary selectivity. By harnessing cutting-edge science in disease spaces that are historically under-addressed through traditional therapies, Scholar Rock works every day to create new possibilities for patients. Learn more about our approach at ScholarRock.com and follow @ScholarRock and on LinkedIn.

Availability of Other Information About Scholar Rock

Investors and others should note that we communicate with our investors and the public using our company website www.scholarrock.com, including, but not limited to, company disclosures, investor presentations and FAQs, Securities and Exchange Commission filings, press releases, public conference call transcripts and webcast transcripts, as well as on Twitter and LinkedIn. The information that we post on our website or on Twitter or LinkedIn could be deemed to be material information. As a result, we encourage investors, the media and others interested to review the information that we post there on a regular basis. The contents of our website or social media shall not be deemed incorporated by reference in any filing under the Securities Act of 1933, as amended.
Scholar Rock® is a registered trademark of Scholar Rock, Inc.

Forward-Looking Statements
This press release contains "forward-looking statements" within the meaning of the Private Securities Litigation Reform Act of 1995, including, but not limited to, statements regarding Scholar Rock's future expectations, plans and prospects, including without limitation, Scholar Rock's expectations regarding its growth, strategy, progress and timing of its clinical trials for apitegromab and SRK-181 and its preclinical programs, including SRK-439, and indication selection and development timing, including the therapeutic potential, clinical benefits and safety thereof, expectations regarding timing, success and data announcements of current ongoing preclinical and clinical trials, its cash runway, expectations regarding the achievement of important milestones, the ability of any product candidate to perform in humans in a manner consistent with earlier nonclinical, preclinical or clinical trial data, and the potential of its product candidates and proprietary platform. The use of words such as "may," "might," "could," "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. All such forward-looking statements are based on management's current expectations of future events and are subject to a number of risks and uncertainties that could cause actual results to differ materially and adversely from those set forth in or implied by such forward-looking statements. These risks and uncertainties include, without limitation, that preclinical and clinical data, including the results from the Phase 2 clinical trial of apitegromab, or Part A or Part B of the Phase 1 clinical trial of SRK-181, are not predictive of, may be inconsistent with, or more favorable than, data generated from future or ongoing clinical trials of the same product candidates, including, without limitation, the Phase 3 clinical trial of apitegromab in SMA or Part B of the Phase 1 clinical trial of SRK-181; Scholar Rock's ability to provide the financial support, resources and expertise necessary to identify and develop product candidates on the expected timeline; the data generated from Scholar Rock's nonclinical and preclinical studies and clinical trials; information provided or decisions made by regulatory authorities; competition from third parties that are 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, without limitation, 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, and our ability to continue as a going concern; as well as those risks more fully discussed in the section entitled "Risk Factors" in Scholar Rock's Quarterly Report on Form 10-Q for the quarter ended June 30, 2024, 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. All information in this press release is as of the date of the release, and Scholar Rock undertakes no duty to update this information unless required by law.

Scholar Rock Holding Corporation
Condensed Consolidated Statements of Operations
(unaudited)
(in thousands, except share and per share data)

	Three Months Ended June 30,		Six Months Ended June 30,	
	2024	2023	2024	2023
Operating expenses				
Research and development	$ 42,373	$ 26,867	$ 85,466	$ 56,602
General and administrative	17,125	12,215	32,451	22,989
Total operating expenses	59,498	39,082	117,917	79,591
Loss from operations	(59,498)	(39,082)	(117,917)	(79,591)
Other income (expense), net	990	1,157	2,556	2,287
Net loss	$ (58,508)	$ (37,925)	$ (115,361)	$ (77,304)
Net loss per share, basic and diluted	$ (0.60)	$ (0.47)	$ (1.20)	$ (0.97)
Weighted average common shares outstanding, basic and diluted	96,813,116	80,117,983	96,352,858	79,865,424

Scholar Rock Holding Corporation
Condensed Consolidated Balance Sheets
(unaudited)
(in thousands)

	June 30, 2024	December 31, 2023
Assets		
Cash, cash equivalents and marketable securities	$ 190,494	$ 279,938
Other current assets	8,643	8,256
Total current assets	199,137	288,194
Other assets	27,728	22,841
Total assets	$ 226,865	$ 311,035
Liabilities and Stockholders' Equity		
Current liabilities	$ 32,987	$ 32,741
Long-term liabilities	60,258	53,076
Total liabilities	93,245	85,817
Total stockholders' equity	133,620	225,218
Total liabilities and stockholders' equity	$ 226,865	$ 311,035

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Second Quarter 2024 Business Update

August 8, 2024

ScholarRock

Forward-Looking Statements

Various statements in this presentation concerning the future expectations, plans and prospects of Scholar Rock Holding Corporation and Scholar Rock, Inc. (collectiv "Scholar Rock"), including without limitation, Scholar Rock's expectations regarding its strategy, its product candidate selection and development timing, including timing for initiation of and reporting results from its preclinical studies and clinical trials for SRK-439, apitegromab, SRK-181 and other product candidates and indication selection development timing, its cash runway, the ability of any product candidate to perform in humans in a manner consistent with earlier nonclinical, preclinical or clinical trial data, the potential of its product candidates and proprietary platform. The use of words such as "may," "could," "might," "will," "should," "expect," "plan," "anticipate," "believ "estimate," "project," "intend," "future," "potential," or "continue," and other similar expressions are intended to identify such forward-looking statements for the purposes of safe harbor provisions under The Private Securities Litigation Reform Act of 1995. All such forward-looking statements are based on management's current expectations future events and are subject to a number of risks and uncertainties that could cause actual results to differ materially and adversely from those set forth in or implied by s forward-looking statements. These risks and uncertainties include, without limitation, that preclinical and clinical data, including the results from the Phase 2 trial of apitegrom or Part A or Part B of the Phase 1 trial of SRK-181, are not predictive of, may be inconsistent with, or more favorable than, data generated from future or ongoing clinical trial the same product candidate, including the Phase 3 clinical trial of apitegromab in SMA and Part B of the Phase 1 clinical trial of SRK-181, respectively, Scholar Rock's ability provide the financial support, resources and expertise necessary to identify and develop product candidates on the expected timeline, the data generated from Scholar Roc nonclinical and preclinical studies and clinical trials, information provided or decisions made by regulatory authorities, competition from third parties that are develop products for similar uses, Scholar Rock's ability to obtain, maintain and protect its intellectual property, the success of Scholar Rock's current and potential future collaboratio Scholar Rock's dependence on third parties for development and manufacture of product candidates including, without limitation, to supply any clinical trials, Scholar Roc ability to manage expenses and to obtain additional funding when needed to support its business activities and establish, and maintain strategic business alliances and business initiatives, and our ability to continue as a going concern as well as those risks more fully discussed in the section entitled "Risk Factors" in Scholar Rock's Form 10-K the year ended December 31, 2023, and Quarterly Report on Form 10-Q for the quarter ended June 30, 2024, as well as discussions of potential risks, uncertainties, and ot important factors in Scholar Rock's subsequent filings with the Securities and Exchange Commission. Any forward-looking statements represent Scholar Rock's views only a today and should not be relied upon as representing its views as of any subsequent date. All information in this press release is as of the date of the release, and Scholar R undertakes no duty to update this information unless required by law.

This presentation may also contain estimates and other statistical data made by independent parties and by us relating to market size and growth and other data about industry. This data involves a number of assumptions and limitations, and you are cautioned not to give undue weight to such estimates. In addition, projections, assumptio and estimates of our future performance and the future performance of the markets in which we compete are necessarily subject to a high degree of uncertainty and risk.

Apitegromab and SRK-181 are investigational drug candidates under evaluation. Apitegromab, SRK-181, SRK-373, SRK-256, and SRK-439 have not been approved for any use the FDA or any other regulatory agency and the safety and efficacy of apitegromab, SRK-181, SRK-373, SRK-256, and SRK-439 have not been established.

ScholarRoc

Introduction & Business Update

Jay Backstrom, M.D., MPH
President & Chief Executive Officer

ScholarRock

Today's Agenda

Topic	Speaker
▶ **Introduction & Business Update**	**Jay Backstrom**, President & Chief Executive Officer
▶ **Development Update**	**Jing Marantz**, Chief Medical Officer
▶ **Pipeline Update**	**Mo Qatanani**, Chief Scientific Officer
▶ **Upcoming Milestones**	**Jay Backstrom**
Q&A Session	

ScholarRock

INTRODUCTION

Advancing Towards Commercialization

1

Selectivity is the Key

The hallmark of our differentiated platform is unparalleled selectivity

2

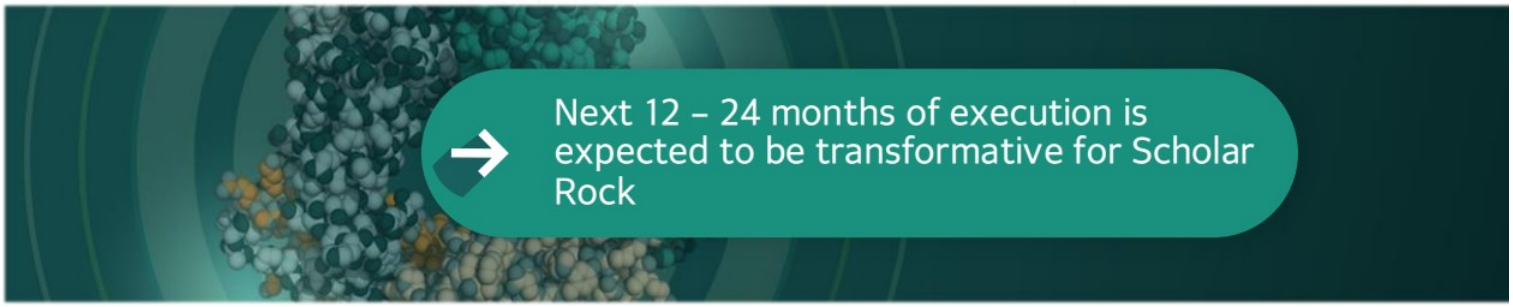
Productive Platform

Strong progress & momentum supports SRRK's scientific approach, capability to grow pipeline and ability to execute

3

Excellent Opportunity

SMA and obesity represent high value markets offering significant potential revenue opportunities



ScholarRock

Our Approach

Selectivity Drives Success

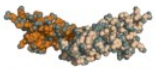
RIGHT TARGET → **Validated Biology**

RIGHT TIME → **Latent Form**

Deep structural insights to validated targets

Industry-leading antibody design and protein engineering to selectively target latent growth facto

Optimized for efficacy an mitigates off-target effe

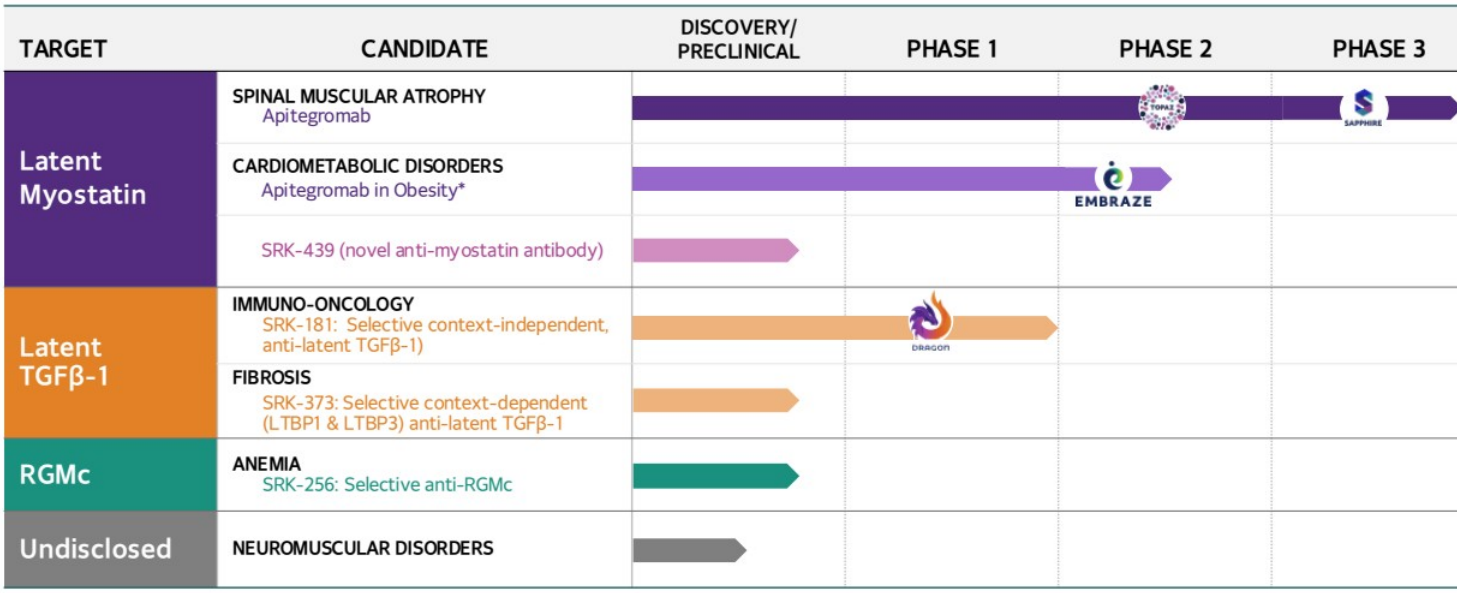


Traditional Target
"mature" active growth factor

Scholar Rock's Target
Latent Growth Factor Complex

ScholarRoc

Scientific Platform Yielding Growing Pipeline Across High Value Therapeutic Areas



*Utilized data from previously completed Ph 1 study in healthy volunteers and initiate a Ph 2 POC trial in 2024.
LTBP1=Latent transforming growth factor beta binding protein 1; LTBP3=Latent transforming growth factor beta binding protein 3; POC=Proof of concept; RGMc=Repulsive guidance molecule C; TGFβ-1=Transforming growth factor beta-1.

ScholarRock

High Value Growth Opportunities

Neuromuscular Disorders

Upcoming SAPPHIRE readout - Q4 2024

Initiate study in SMA patients < 2 years old: Planned for 2025

Exploring additional neuromuscular populations

Cardiometabolic Disorders

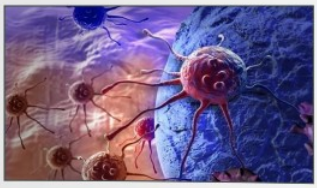
Ph 2 POC EMBRAZE study enrolling: Topline data expected in Q2 2025

Advancing SRK-439 to IND

Immuno-Oncology

Established PoC with SRK-181 in multiple advanced solid tumors

End of Ph 1 meeting planned



Fibrosis

Advance SRK-373, LTBP, to IND

Advancing nonclinical studies in renal and pulmonary fibrosis

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Strong Proprietary Platform

Cutting-Edge Research Recognized by Global Scientific Community

SRK-181	SRK-439	SRK-373
Promising objective response rates, safety, and novel biomarker data highlighted in oral presentation at ASCO	Compelling new preclinical data highlighted in oral presentation at ADA	Featured on the cover of **Science Signaling** and in *Focus* article: "More Velcro for the TGFB-1 Straightjack… *by Boris Hinz*

ASCO

Journal of Clinical Oncology

An American Society of Clinical Oncology Journal

American Diabetes Association®

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2024 & 2025 Anticipated Milestones

Milestones	2024	2025
SRK-181 data at ASCO • Oral presentation June 3 • Developmental Therapeutics-Immunotherapy	✓ ◆	
SRK-439 data at American Diabetes Association • Oral presentation June 23 • New Insights into Therapeutic Strategies for Obesity and Diabetes	✓ ◆	
SRK-439 IND submission		◆
EMBRAZE Ph 2 Trial (apitegromab in obesity) • Trial open for enrollment • Topline data expected in Q2 2025	✓ ▬▬▬	▬▬▬ ◆
SAPPHIRE Ph 3 Trial (apitegromab in SMA) • Topline readout expected in Q4 2024	◆	
Potential SMA launch in Q4 2025, if successful & approved		◆
Study in SMA Patients < 2 Years of Age • Study design endorsed by EMA's paediatric committee • Study initiation planned for 2025		▬▬▬

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Development Update

Jing Marantz, M.D., Ph.D.
Chief Medical Officer

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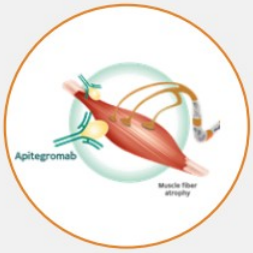
APITEGROMAB FOR SPINAL MUSCULAR ATROPHY

Why We Are Confident

1. Muscle Targeting

Selective muscle-targeting designed to improve motor function while minimizing off-target effects



2. Clinical Proof-Of-Concept

TOPAZ clinical proof-of-concept with substantial and durable effect across broad SMA patients ages 2-21

TOPAZ

3. SAPPHIRE Optimized for Success

Trial design informed by insights from TOPAZ

SAPPHIRE

4. TOPAZ Safety Profile

>90% patient retention,[1] well tolerated profile support durability of treatment

[1] Pooled nonambulatory patients

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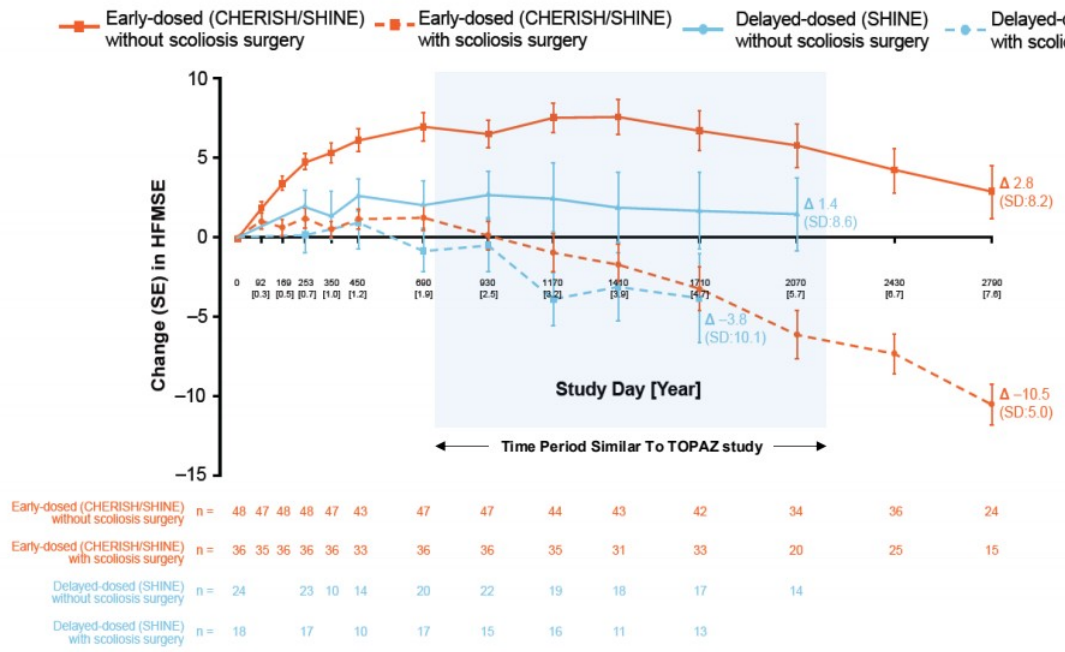
Motor function slowly declines despite following initial increase

CHERISH/SHINE study: Long-Term Results of Nusinersen-Treated Patients



CHERISH Study Baseline	Nusinersen	Contr
N	84	42
Age at Screening	4 years	3 year

- Decline in motor function is influenced by scoli surgery and contractures (Wolfe 2024; Dunawa 2020; Salazar 2018)
- TOPAZ participants had ~2 years of nusinersen treatment at baseline
- Blue-shaded area represents similar duration o nusinersen treatment

Finkel RS et al. "Final Safety and Efficacy Data From the SHINE Study in Participants With Infantile-Onset and Later-Onset SMA." Presented at Cure SMA Annual Conference, July 2024

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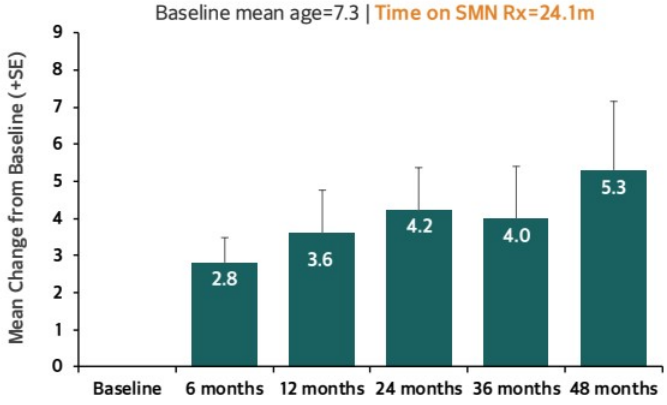
Apitegromab TOPAZ Clinical Trial: Motor Function Outcomes by HFMSE Over 48 Months

Improvements Were Substantial and Sustained

Pooled Nonambulatory Patients[1]

Age 2-21 Years
All Doses (N=35)

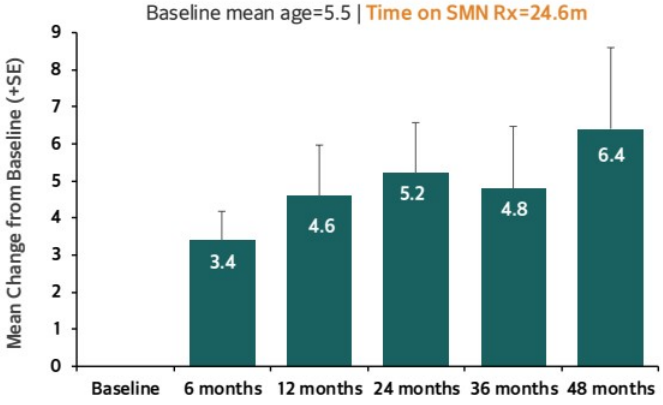
Baseline mean age=7.3 | Time on SMN Rx=24.1m



	Baseline	6 months	12 months	24 months	36 months	48 months
n=	35	29	32	29	28	23
95% CI=		(1.4, 4.1)	(1.2, 6.0)	(1.9, 6.6)	(1.0, 6.9)	(1.5, 9.2)

Age 2-12 Years
All Doses (N=29)

Baseline mean age=5.5 | Time on SMN Rx=24.6m



	Baseline	6 months	12 months	24 months	36 months	48 months
n=	29	23	26	23	23	19
95% CI=		(1.8, 5.0)	(1.8, 7.4)	(2.3, 8.0)	(1.3, 8.3)	(1.8, 11.0)

1. For the 48-month evaluation, an observed case analysis was conducted using available data by analysis timepoint, censoring any HFMSE assessments after the patient received scoliosis surgery. The analysis population pooled the nonambulatory patients (Cohorts 2 and 3) and included patients receiving either low dose (2 mg/kg) or high dose (20 mg/kg) apitegromab (inclusive of patients in Cohort 3 who switched from 2 mg/kg to 20 mg/kg in Year 2). A total of 11 patients in the population had scoliosis surgery during the study and their data was excluded from any HFMSE assessments at 48 months. Visit windows were applied to utilize data from unscheduled or early termination visits if the patient was missing the HFMSE total score at the scheduled visit. Error bars represent standard error (SE) and CI represents confidence interval. SMN Rx=SMN therapy. Apitegromab is an investigational drug candidate being evaluated for the treatment of spinal muscular atrophy. Apitegromab has not been approved for any use by the US FDA or any other health authority, and its safety and efficacy have not been established.

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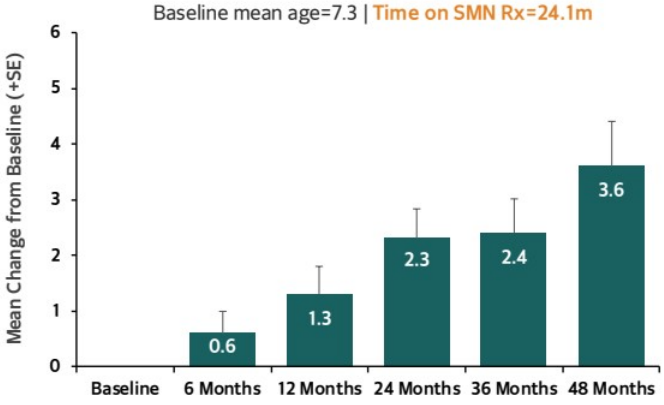
Apitegromab TOPAZ Clinical Trial: Motor Function Outcomes by RULM Over 48 Months

Improvements Were Substantial and Sustained

Pooled Nonambulatory Patients[1]

Age 2-21 Years
All Doses (N=35)

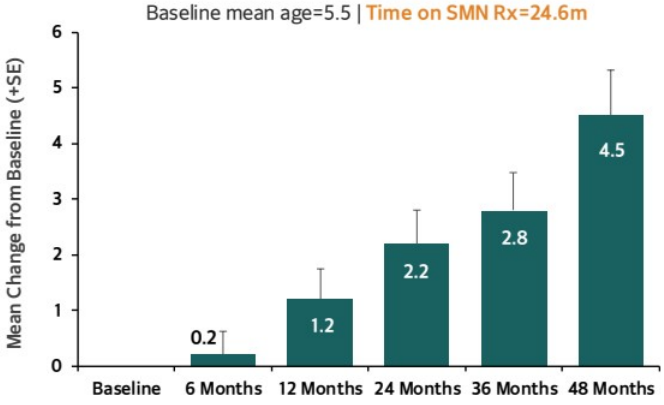
Baseline mean age=7.3 | Time on SMN Rx=24.1m



	Baseline	6 Months	12 Months	24 Months	36 Months	48 Months
n=	34	28	31	31	27	22
95% CI=		(-0.2, 1.4)	(0.2, 2.3)	(1.2, 3.3)	(1.1, 3.7)	(2.0, 5.3)

Age 2-12 Years
All Doses (N=29)

Baseline mean age=5.5 | Time on SMN Rx=24.6m



	Baseline	6 Months	12 Months	24 Months	36 Months	48 Months
n=	28	22	25	25	22	18
95% CI=		(-0.7, 1.1)	(0.1, 2.4)	(1.0, 3.5)	(1.4, 4.2)	(2.7, 6.3)

1. For the 48-month evaluation, an observed case analysis was conducted using available data by analysis timepoint, censoring any RULM assessments after the patient received scoliosis surgery. The analysis population pooled the nonambulatory patients (Cohorts 2 and 3) and included patients receiving either low dose (2 mg/kg) or high dose (20 mg/kg) apitegromab (inclusive of patients in Cohort 3 who switched from 2 mg/kg to 20 mg/kg in Year 2). A total of 11 patients in the population had scoliosis surgery during the study and their data was excluded from any RULM assessments at 48 months. Visit windows were applied to utilize data from unscheduled or early termination visits if the patient was missing the RULM total score at the scheduled visit. One patient did not have baseline RULM due to young age. Error bars represent standard error (SE) and CI represents confidence interval. SMN Rx=SMN therapy. Apitegromab is an investigational drug candidate being evaluated for the treatment of spinal muscular atrophy. Apitegromab has not been approved for any use by the US FDA or any other health authority, and its safety and efficacy have not been established.

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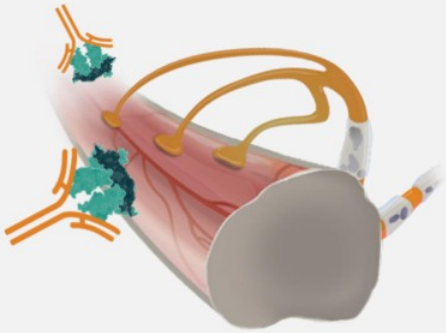
Summary of TOPAZ Data

Substantial and Sustained Improvement over 48 MONTHS[1]



TOPAZ

Data to date has shown substantial clinical benefit that is dose-dependent

Clinical benefit continued to improve or was sustained over 48 months

Consistency across functional scales and patient-reported outcomes

Well tolerated profil and low discontinuation rate supports durability of treatment

>90% of patients remain on therapy*

TOPAZ data suggest that apitegromab has the potential to transform care in SMA by directly addressing progressive muscle weakness

1- A total of 11 patients in the population had scoliosis surgery during the study and their data was excluded from any HFMSE and RULM assessments at 48 months.
PRO=Patient Reported Outcome
*Pooled non-ambulatory cohorts

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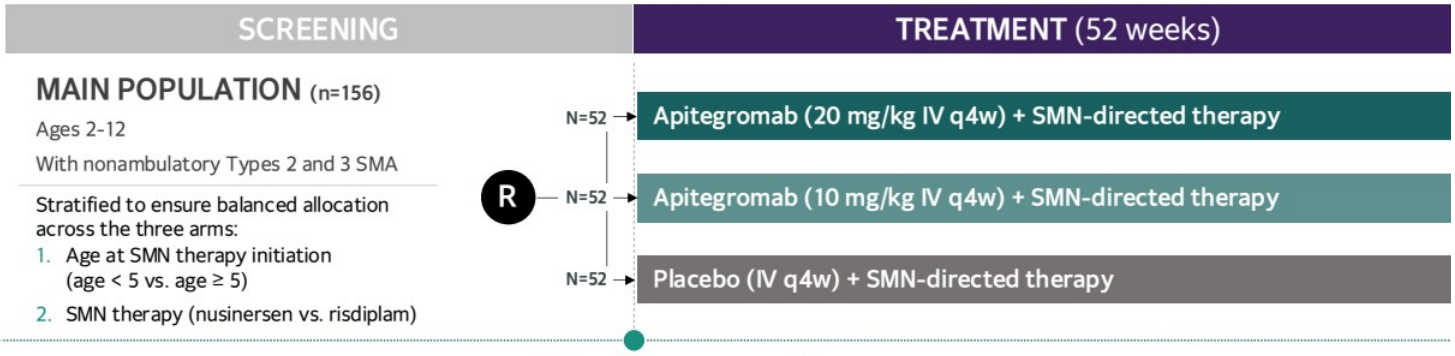
SAPPHIRE Trial Designed for Clinical Success

SAPPHIRE

Randomized, double-blind, placebo-controlled, parallel arm design (n=204)
Enrolling patients who are on SMN-directed therapy (nusinersen or risdiplam)
Completed enrollment in Q3 2023



ENDPOINTS

Primary Efficacy:
Mean HFMSE change from baseline at 12 months

Additional Efficacy Measures:
RULM, WHO, other outcome measures

Safety, PK/PD, ADA

Additional Data Opportunities

Exploratory population (age 13-21), in patients using SMN therapy
Focused upon safety & exploratory efficacy (n=48; 2:1 randomization between apitegromab 20 mg/kg vs placebo)

Separate open-label extension study (after patients complete 12-month treatment period)
Focused upon safety & exploratory long-term efficacy

ClinicalTrials.gov Identifier: NCT05156320
HFMSE=Hammersmith Functional Motor Scale Expanded; RULM=Revised Upper Limb Module; R=randomization; SMA=spinal muscular atrophy; SMN=survival motor neuron.

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Goals of the EMBRAZE Proof-of-Concept Study

EMBRAZE Study Aims to Demonstrate

- Effect of apitegromab to preserve lean mass in obese or overweight patients receiving a GLP-1 agonist
- Safety and tolerability data to provide initial support for long-term chronic use
- Explore the potential effect of apitegromab to improve metabolic profile and physical function

INSIGHTS GAINED FROM EMBRAZE STUDY

to inform SRK-439 development

Initiated trial in May 2024, ahead of target timeline	Strong enrollment momentum	Updating guidance for topline data to Q2 2025

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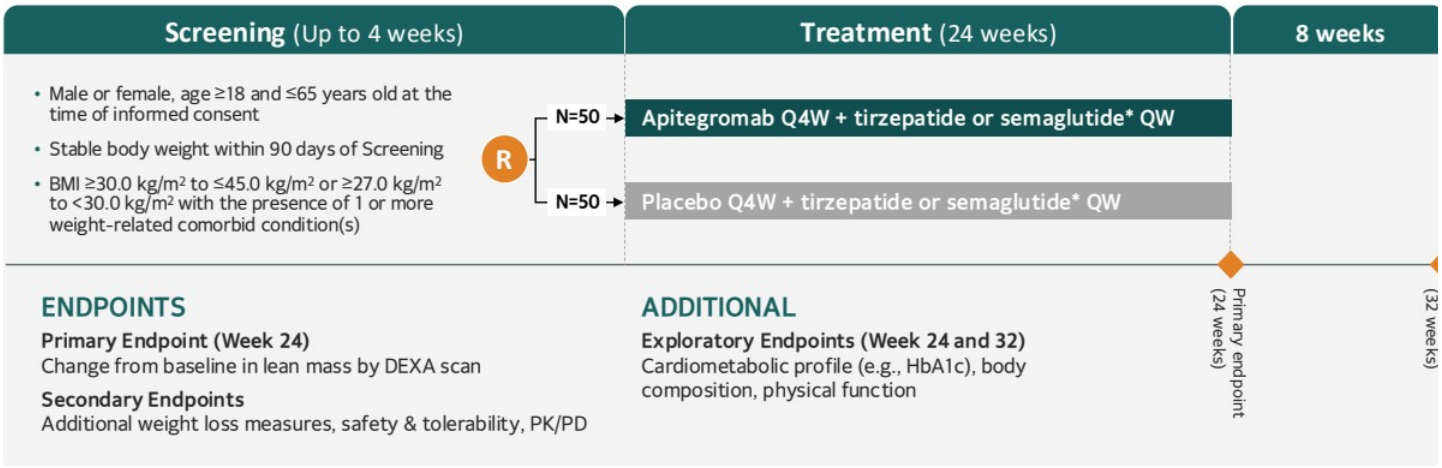
EMBRAZE TRIAL DESIGN

Enrolling Phase 2 Proof-of-Concept Study of Apitegromab in Obesity

EMBRAZE

Randomized, double-blind, placebo-controlled (n=100)
Enrolling patients who are overweight or obese
Enrollment ahead of schedule; topline data expected in Q2 2025



ENDPOINTS

Primary Endpoint (Week 24)
Change from baseline in lean mass by DEXA scan

Secondary Endpoints
Additional weight loss measures, safety & tolerability, PK/PD

ADDITIONAL

Exploratory Endpoints (Week 24 and 32)
Cardiometabolic profile (e.g., HbA1c), body composition, physical function

*Participating patient will be assigned to either tirzepatide or semaglutide depending on availability.
Apitegromab dose regimen will be 10 mg/kg Q4W, based on projected exposure in the obese population comparable to that of 20 mg/kg Q4W in SMA
Tirzepatide and semaglutide dose regimen will follow the United States Prescribing Information.

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Pipeline Update

Mo Qatanani, Ph.D.
Chief Scientific Officer

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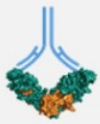
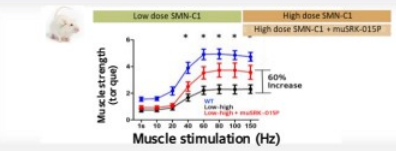
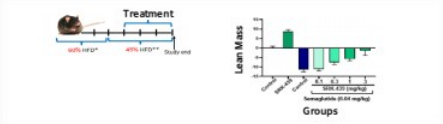
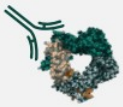
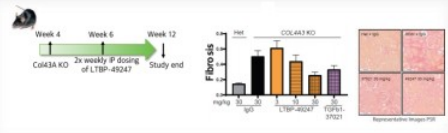
TRANSLATIONAL SUCCESS TO THE CLINIC

Platform and Expertise Drive Success in Clinic

Superb Engineering	Translational Models	Clinic
 Apitegromab Spinal Muscular Atrophy		TOPAZ SAPPHIRE ONY
 SRK-181 Immuno-oncology		DRAGON
 SRK-439 Cardiometabolic		IND planned for 2025
 SRK-373 Fibrosis		IND planned for 2026

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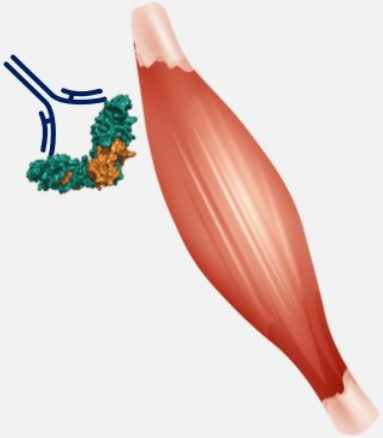
Strong Scientific Validation and Promising Preclinical Evidence

Give Us Confidence in SRK-439



Preclinical data to date show strong potential to support healthier weight loss in combination with GLP-1 RA:

- **Preservation of lean mass** during GLP-1 RA-induced weight loss and **improvement in metaboli parameters**
- **Increase in lean mass** and attenuation of fat mass regain following GLP-1 RA withdrawal
- **Greater potency** compared to an anti-ACTRII antibody

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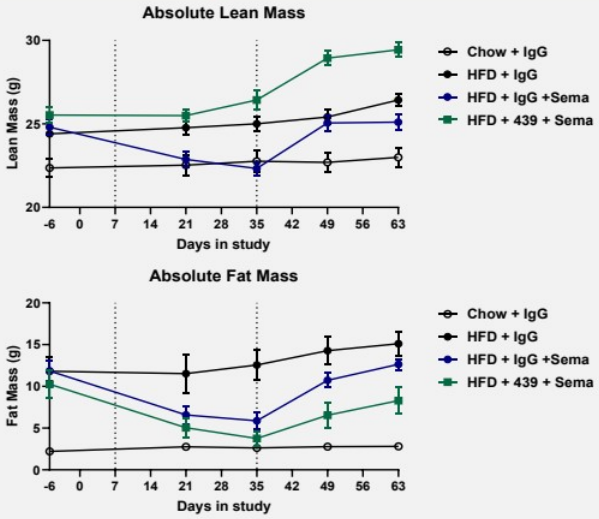
SRK-439 Increased Lean Mass and Attenuated Regain of Fat Mass After GLP-1 RA Withdrawal in Obesity Mouse Model

SRK-439 Increased Absolute Lean Mass and Attenuated Regain of Absolute Fat Mass



Key Observations

- Considerable lean mass loss seen with semaglutide treatment as expected
- Treatment with SRK-439 led to:
  - Preservation of lean mass during semaglutide treatment
  - Significant increase in lean mass upon semaglutide discontinuation
  - Attenuation of fat mass regain upon semaglutide discontinuation

American Diabetes Association.

Study conducted in Diet Induced Obesity (DIO) mouse model utilizing a murine chimera of SRK-439
GLP-1 RA: GLP1 receptor agonist. Day 7 is start of semaglutide and SRK-439 treatment. Day 35 is discontinuation of semaglutide treatment.

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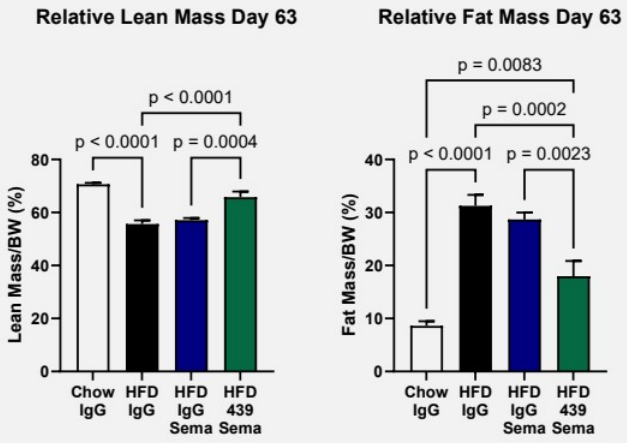
SRK-439 Improved Body Composition After GLP 1-RA Withdrawal

SRK-439 Improved Proportion of Lean and Fat Mass to Total Body Weight



Key Observations

- SRK-439 attenuates regain of fat mass after withdrawal of semaglutide compared to IgG control
- SRK-439 leads to higher lean mass proportion after withdrawal of semaglutide compared to IgG control

American Diabetes Association.

Study conducted in Diet Induced Obesity (DIO) mouse model utilizing a murine chimera of SRK-439
GLP-1 RA: GLP1 receptor agonist. HFD: High Fat Diet

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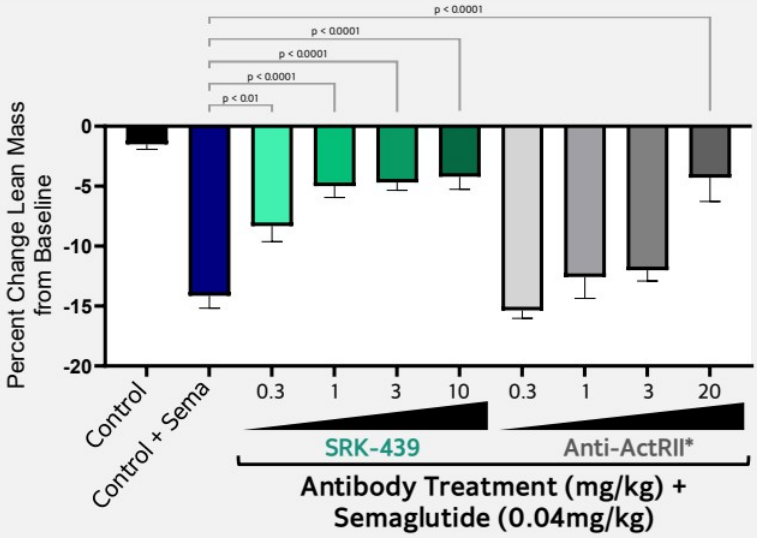
LOW EFFICACIOUS DOSE AND COMPETITIVE PROFILE

SRK-439 Is More Potent than Anti-ActRII Antibody at Maintaining Lean Mass During GLP-1 RA-Induced Weight Loss

Head-to-Head Comparison to Non-Selective Myostatin Inhibitor in DIO Mouse Model



Key Observations

- SRK-439 preserved semaglutide-driven lean mass loss dose-dependently and at lower do than anti-ActRII
- Highlights efficacy of SRK-439 and avoids potential liabilities of non-selective approach anti-ActRII antibodies
- Low target dose of SRK-439 supports subcutaneous and potentially best-in-class profile

*Murine chimera of Bimagrumab
Study conducted in Diet Induced Obesity (DIO) mouse model utilizing a murine chimera of SRK-439

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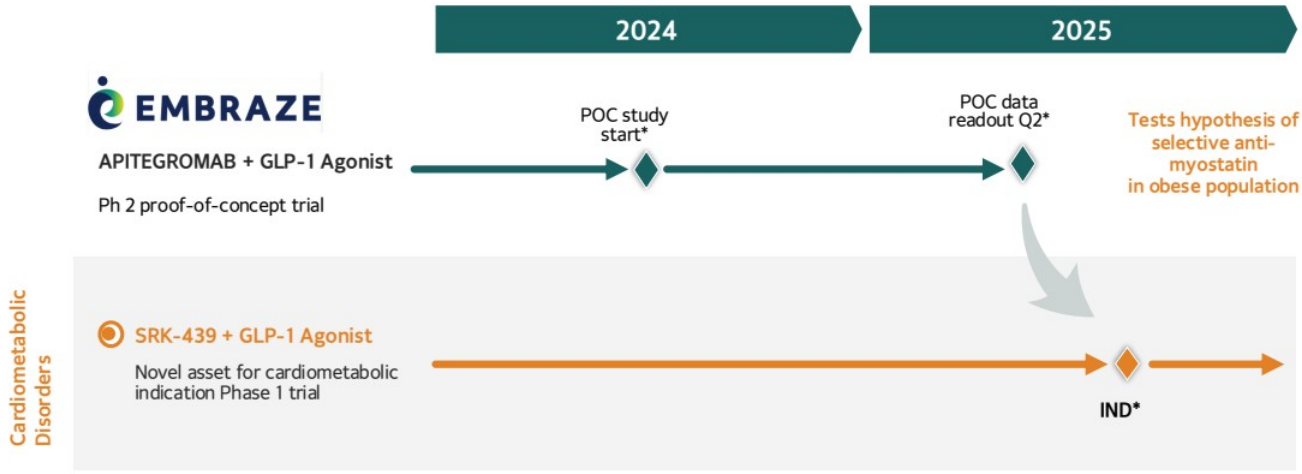
Leveraging Apitegromab to Inform Obesity Program



*Expected timelines
POC = Proof of Concept

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Upcoming Milestones

Jay Backstrom
Chief Executive Officer

ScholarRock

Looking Ahead: Upcoming Milestones

Q2 2024
ACCOMPLISHMENTS

- PRIME pre-submission meeting completed
- INITIATED EMBRAZE trial with apitegromab in obesity
- ADVANCED IND-enabling studies for SRK-439
- PRESENTED encouraging SRK-181 data at ASCO

2H 2024 focus

- SAPPHIRE Readout in Q4
- Prepare for commercialization
- Complete EMBRAZE enrollment
- Advance IND-enabling studies for SRK-439

IND=Investigational new drug; POC=Proof of concept;

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Q&A Session

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Company Speakers

Jay Backstrom, M.D., MPH
President & Chief
Executive Officer

Jing Marantz, M.D., Ph.D.
Chief Medical Officer

Mo Qatanani, Ph.D.
Chief Scientific Officer

Ted Myles, MBA
Chief Operating Officer
and Chief Financial Officer

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Thank you!

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