Investor Day

May 22, 2024

ScholarRock

Welcome

Rushmie Nofsinger
Vice President
Investor Relations & Corporate Affairs

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. (collectively, “Scholar Rock”), 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 preclinical studies and clinical trials for SRK-439, apitegromab, SRK-181, and other product candidates and indication selection and 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, and the potential of its product candidates and proprietary platform. The use of words such as “may,” “could,” “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 for the purposes of the safe harbor provisions under The Private Securities Litigation Reform Act of 1995. 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 trial of apitegromab 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 trials of 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 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, the success of Scholar Rock’s current and potential future collaborations, Scholar Rock’s dependence on third parties for development and manufacture of product candidates including, without limitation, to supply any clinical trials, 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 Scholar Rock’s Form 10-K for the year ended December 31, 2023, and Quarterly Report on Form 10-Q for the quarter ended March 31, 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.

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 our 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, assumptions, 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, and SRK-439 have not been approved for any use by the FDA or any other regulatory agency and the safety and efficacy of apitegromab, SRK-181 and SRK-439 have not been established.

ScholarRock

Company Speakers

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

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

Tracey Sacco
Chief Commercial Officer

Mo Qatanani, Ph.D.
Chief Scientific Officer

Expert Speakers

Diana Castro, M.D.

Founder of Neurology & Neuromuscular Care Center and Neurology Rare Disease Center

Former Associate Professor of Pediatrics, Neurology and Neurotherapeutics, University of Texas Southwestern, Director of the Neuromuscular Program & Fellowship and Director of the Pediatric Muscular Dystrophy Association Clinic at Children's Health

Ania Jastreboff, M.D., Ph.D.

Associate Professor of Medicine (Endocrinology), Yale School of Medicine

Director, Yale Obesity Research Center (Y-Weight)

Co-Director, Yale Center for Weight Management

Member of Board of Directors for the American Board of Obesity Medicine

Today's Agenda

Topic	8:30 – 12:00	Speaker
▶ **Welcome**		**Rushmie Nofsinger**, VP of IR & Corporate Affairs
▶ **Vision & Strategic Overview**		**Jay Backstrom**, President & Chief Executive Officer
▶ **SMA: The Patient Journey**		**Diana Castro**, M.D., Neurology & Neuromuscular Care Center and Neurology Rare Disease Center
▶ **Apitegromab: Development Program**		**Jing Marantz**, Chief Medical Officer
▶ **Apitegromab: Commercial Readiness**		**Tracey Sacco**, Chief Commercial Officer
	Q&A	
	10-minute Break	
▶ **Obesity: Muscle Matters**		**Ania Jastreboff**, M.D., Ph.D., Yale School of Medicine
▶ **SRK-439: Differentiated Approach**		**Mo Qatanani**, Chief Scientific Officer
▶ **Cardiometabolic Development Program**		**Jing Marantz**, Chief Medical Officer
	Q&A	
▶ **Closing Remarks**		**Jay Backstrom**, President & Chief Executive Officer

Building a Fully Integrated Biopharma Company

Jay Backstrom
President & Chief Executive Officer

ScholarRock

We are a global leader in harnessing the life-changing potential of TGFβ biology

OUR MISSION

To discover, develop, and deliver life-changing therapies by harnessing cutting-edge science to create new possibilities for people living with serious diseases

TGFβ=Transforming growth factor-beta.

Advancing Our Journey Towards Commercialization

1

Selectivity is the Key

The hallmark of our differentiated platform is unparalleled selectivity

2

Large Unmet Needs

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

3

Positioned for Success

Experienced team executing on strategy and goals

Next 12 – 24 months of execution is expected to be transformative for our company

AGENDA

Evolving into a Fully Integrated Biopharma Company

Leveraging our proprietary platform

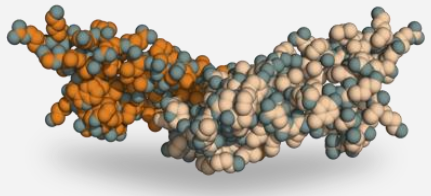
Building a muscle-targeted franchise

The road ahead – value drivers

SELECTIVITY DRIVES SUCCESS

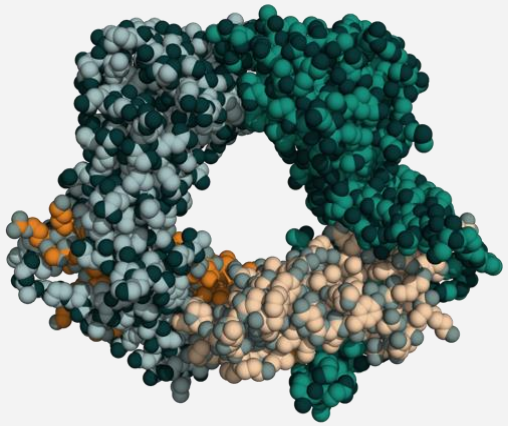
Scholar Rock Has Succeeded Where Others Have Failed

Traditional Target
"Mature" Active Growth Factor



Has been challenging to target because of high homology across super-family

Scholar Rock's Target
Latent Growth Factor

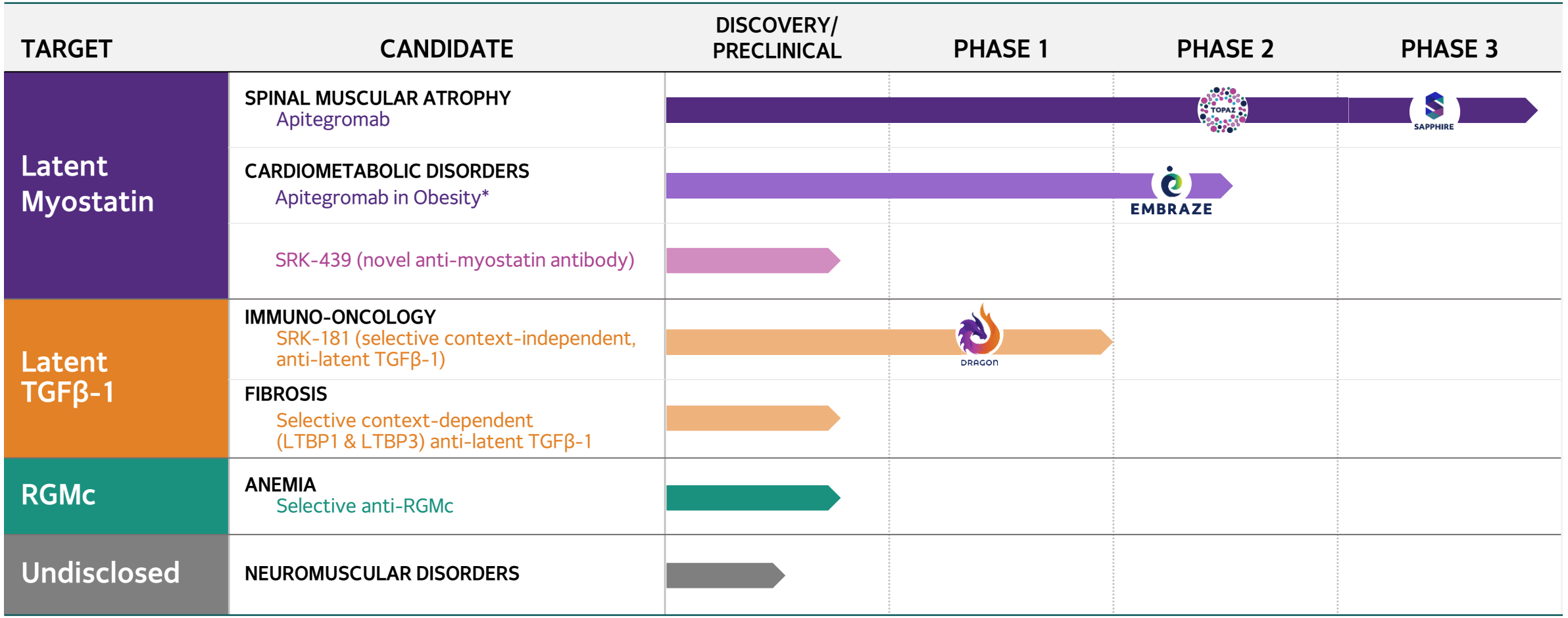


Targeting the 'cage' before growth factor is released allows for exquisite selectivity

RIGHT TARGET → Validated Biology

RIGHT TIME → Latent Form

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.

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High Value Growth Opportunities

Neuromuscular Disorders

Ph 3 SAPPHIRE in SMA

Apitegromab additional neuromuscular populations

Cardiometabolic Disorders

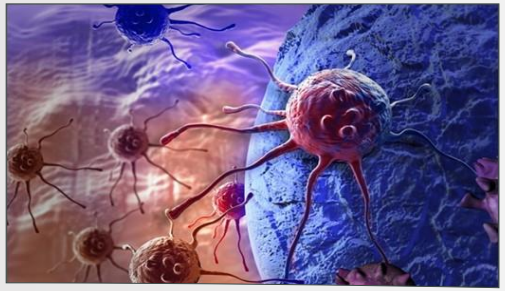
EMBRAZE PoC in obesity

Advance SRK-439 to IND

Immuno-Oncology

Established PoC in ccRCC

End of Ph 1 meeting



Fibrosis

Advance LTBP to IND

Nonclinical studies in renal and pulmonary fibrosis

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Strong Proprietary Platform

AGENDA

Evolving into a Fully Integrated Biopharma Company

Leveraging our proprietary platform

Building a muscle-targeted franchise

The road ahead – value drivers

Distinct & High Value Opportunities for Myostatin Inhibition

Selective Myostatin Inhibition

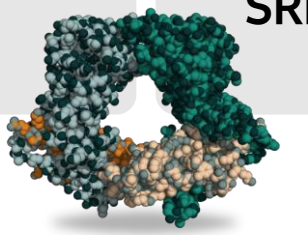
	Therapeutic Area	Current Market Size
Apitegromab		**$4.5B**[1] Spinal Muscular Atrophy SRRK Commercialization
SRK-439		**~$20B**[2] Projected to reach $100B by 2030[2] Obesity Ideal for Partnership

[1] Revenue as of Biogen 4Q23 financial update, Roche 4Q23 financial update, and Novartis 4Q23 financial update
[2] Morgan Stanley Research, "Obesity Medication, Ripple Effects." April 14, 2024

Leveraging Our Building Blocks, Transformative 18 Months Ahead

Powerful Building Blocks

Novel Scientific Platform	Experienced Team	Established Markets with High Unmet Need	Global Rights Across the Portfolio
Robust Pipeline across 5 therapeutic areas **3** clinical programs **Multiple** preclinical programs	**Deep rare disease**, R&D, FDA/EMA approval experience **~150+** Employees **~74%** R&D	**Apitegromab** in SMA **SRK-439** in Obesity **SRK-181** in Immuno-oncology	**29** patent families pending **Exclusivity** through 2036 to 2043 for key assets



* Subject to regulatory approval

Leveraging Our Building Blocks, Transformative 18 Months Ahead

Value Drivers

Significant Inflection Points in Next Year

Phase 3 SAPPHIRE Trial

Proof of Concept
in Obesity

Preparing to Launch SMA in US and EU*

Phased approach
to building key capabilities

Well established presence within SMA Community

Powerful Building Blocks

Novel Scientific Platform

Robust Pipeline
across 5 therapeutic areas

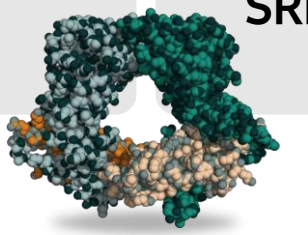
3 clinical programs

Multiple preclinical programs

Experienced Team

Deep rare disease, R&D, FDA/EMA approval experience

~150+ Employees
~74% R&D



Established Markets with High Unmet Need

Apitegromab in SMA

SRK-439 in Obesity

SRK-181 in Immuno-oncology

Global Rights Across the Portfolio

29 patent families pending

Exclusivity through 2036 to 2043 for key assets

* Subject to regulatory approval

AGENDA

Evolving into a Fully Integrated Biopharma Company

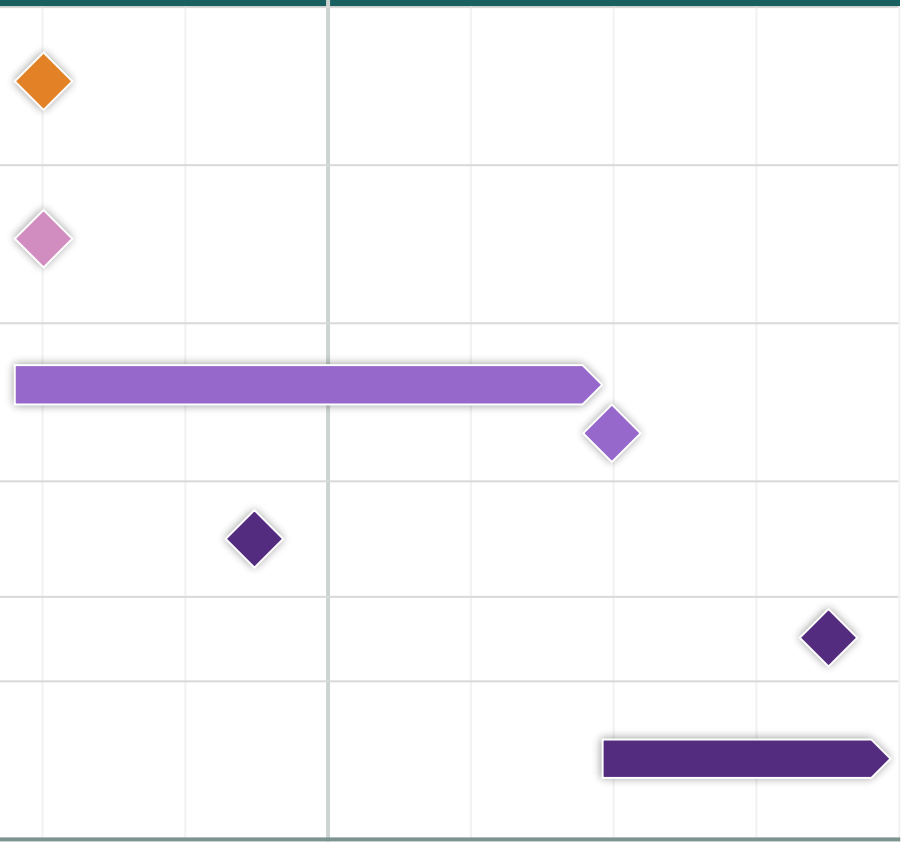
Leveraging our proprietary platform

Building a muscle-targeted franchise

The road ahead – value drivers

Near-Term 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		
EMBRAZE Ph 2a Trial (apitegromab in obesity) • Trial open for enrollment • Topline data expected mid-2025		
SAPPHIRE Ph 3 Trial (apitegromab in SMA) • Topline readout 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		



Strategic Roadmap: Our Vision for 2030

Scholar Rock Today

- Building Commercial Preparedness in US & EU
- 3 Clinical Programs
- Multiple Research Programs

Scholar Rock 2030

- **Multi-Billion Dollar Global Neuromuscular Franchise**
  Established in key major markets
- **5 Clinical Programs**
  Including additional neuromuscular assets
- **Multiple Research Programs**
  Across TGFβ superfamily of growth factors
- **New Targets**
  Leveraging our world-leading antibody engineering capabilities to pursue targets beyond TGFβ superfamily

From a World-Class Scientific Platform to a World-Class Biopharma Company

PLATFORM	Unparalleled selectivity to optimize efficacy and safety
PIPELINE	Broad opportunities to improve patient outcomes in areas of high unmet need
PEOPLE	Seasoned research, development and commercial teams

Industry-leading anti-myostatin portfolio

SMA & Obesity represent significant revenue opportunity

Apitegromab: Development Program

Jing Marantz
Chief Medical Officer

ScholarRock

Upcoming Catalyst: Topline Data Expected in Q4 2024

Where We Were
Phase 2

Where We Are
Phase 3

Future

TOPAZ

12-Month
COMPLETE

48-Month EXTENSION
COMPLETE

SAPPHIRE

12-Month
IN PROCESS

ONYX

Long-Term
EXTENSION

SAPPHIRE

- Q4 2024 data[1]
- 2025 launch[2]

[1] Anticipated
[2] Subject to regulatory approval

AGENDA

Apitegromab for SMA Development Program

Pivotal SAPPHIRE trial: Why we are confident

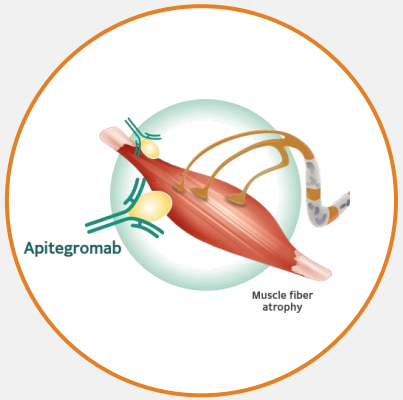
Evidence supporting apitegromab's potential

Where we plan to go

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 supports durability of treatment

[1] Based on TOPAZ patients receiving combination therapy after 4 years of treatment. Data cutoff date: April 2024

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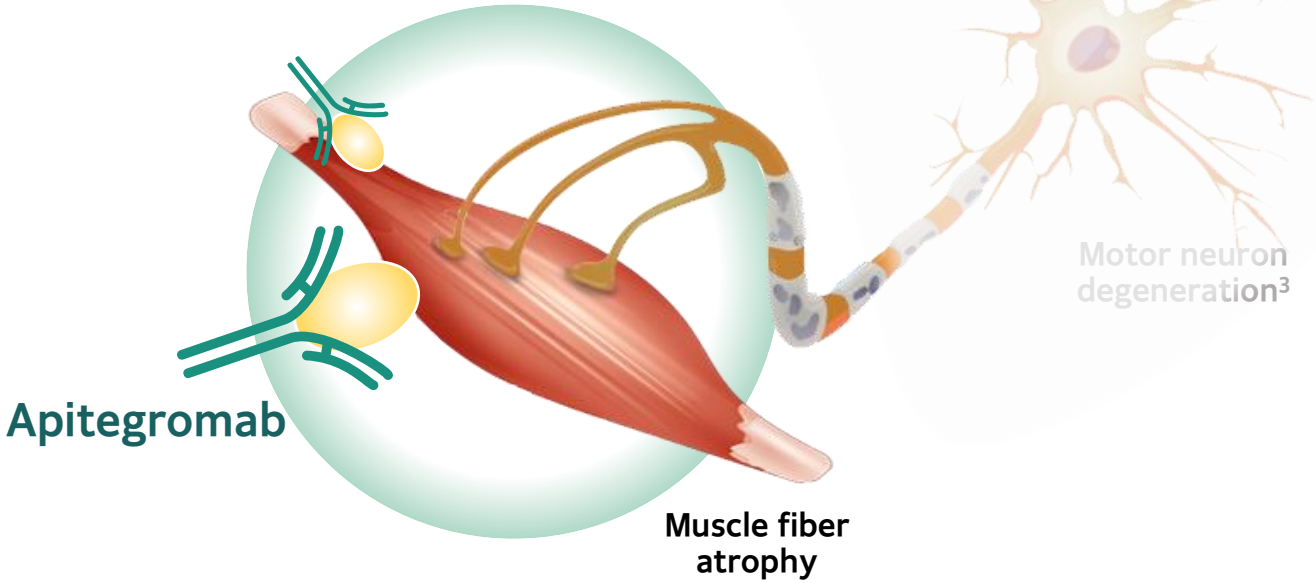
Selectively Targets Muscle to Address Unmet Needs

Apitegromab is a selective MUSCLE-TARGETED APPROACH designed to improve motor function*[1,2]



Evrysdi risdiplam

SPINRAZA (nusinersen) injection 12 mg/5 mL

zolgensma (onasemnogene abeparvovec-xioi) suspension for intravenous infusion

Motor neuron degeneration[3]


Myostatin


Apitegromab

- Myostatin is a negative modulator of muscle growth
- Evidence indicates upstream targeting of structurally differentiated pro- and latent myostatin minimizes undesirable off-target effects

* Based on Animal Model Data; 1. Long KK, et al. Hum Mol Genet. 2019;28(7):1077-1088; 2. Pirruccello-Straub M, et al. Sci Reports. 2018;8(1):2292. doi:10.1038/s41598-018-20524-9 3. Figure adapted from: SMA Foundation Overview. http://www.smafoundation.org/wp-content/uploads/2012/03/SMA-Overview.pdf.; Accessed April 18, 2021. For illustrative purposes only

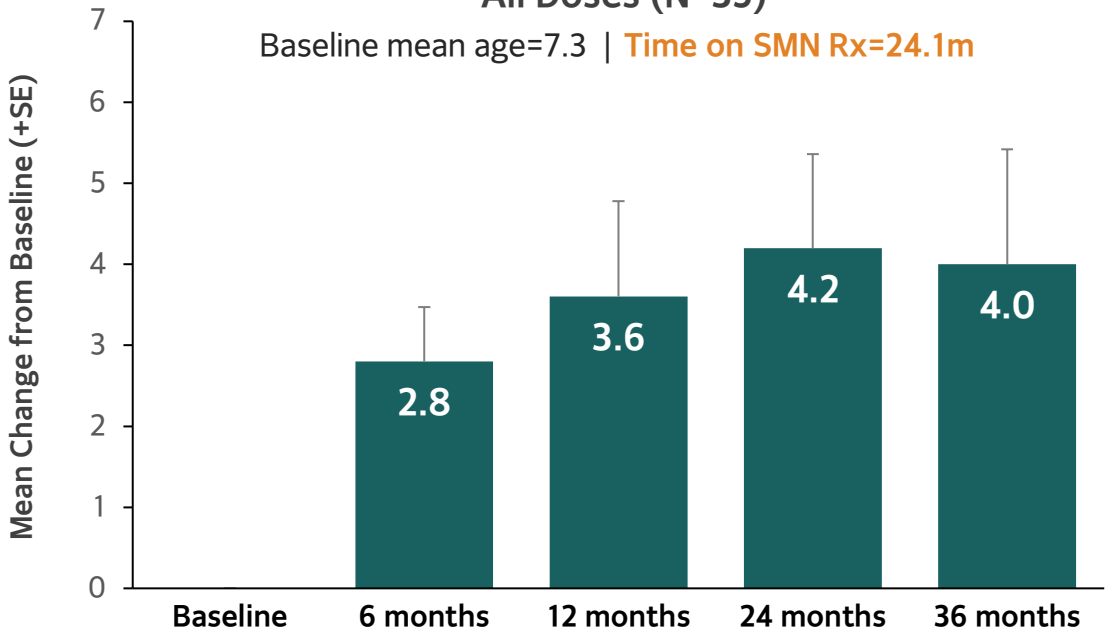
Motor Function Improvements Were Substantial & Sustained Over 36 Months

Pooled Nonambulatory Patients

HFMSE

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

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

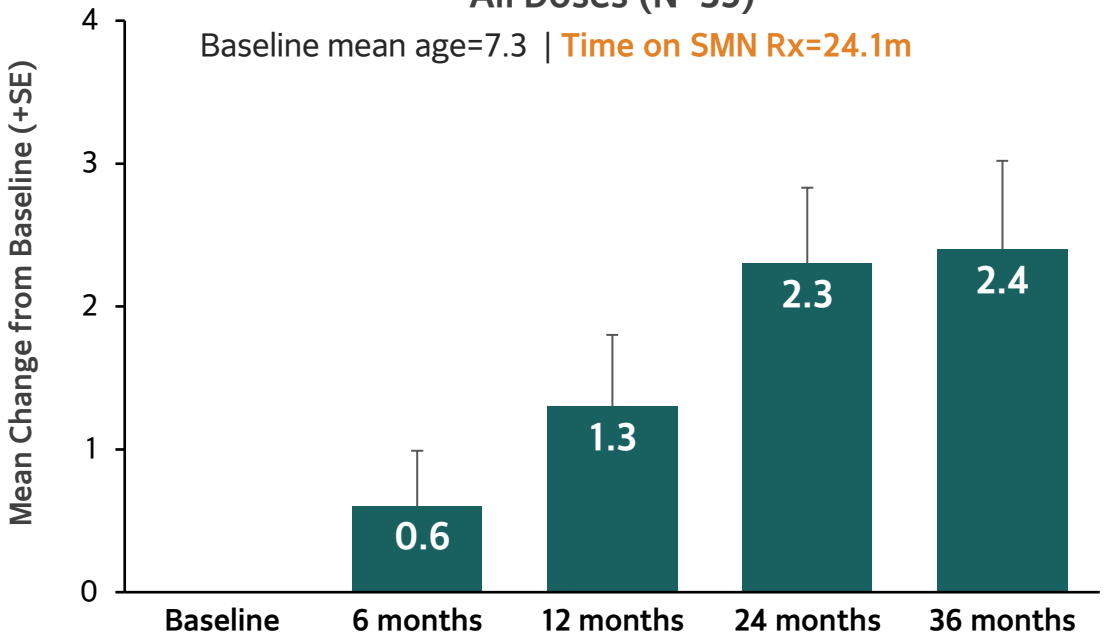


n=	35	29	32	29	28
95% CI=		(1.4, 4.1)	(1.2, 6.0)	(1.9, 6.6)	(1.0, 6.9)

RULM

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

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



n=	35	28	31	31	27
95% CI=		(-0.2, 1.4)	(0.2, 2.3)	(1.2, 3.3)	(1.1, 3.7)

For the 36-month evaluation, an observed case analysis was conducted, which pooled all the nonambulatory patients (Cohorts 2 and 3) and was based upon the available data for given timepoints. This analysis population 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). This analysis excludes data post scoliosis surgery from seven patients. One patient did not conduct HFMSE at time of database lock for 24 months, however, this patient had an unscheduled HFMSE score one month prior to their scheduled visit. In the most recent analysis, this result was included in the 24-month analysis. Error bars represent SE. CI represents confidence interval. SMN Rx=SMN therapy. In the age 2-21 group, 18/28 patients achieved ≥ 1-pt gains, and 11/28 patients ≥ 3-pt gains at 36 months. Data cutoff date as of March 13, 2023. 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.**

SAPPHIRE Phase 3 Design is Optimized by Insights from TOPAZ

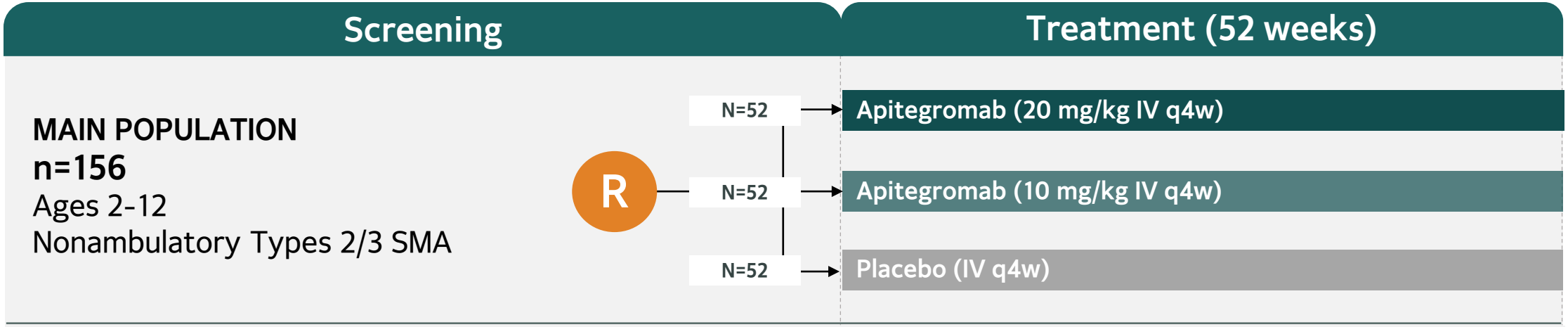
	TOPAZ Learnings	SAPPHIRE Trial Design
Study Population	• Substantial HFMSE gains in nonambulatory* patients • Type 2/3 SMA cohorts	• Nonambulatory* Type 2/3 SMA patients • Primary efficacy endpoint: HFMSE
Age	• Exploratory age 2-12 analysis in nonambulatory Type 2/3 showed transformative potential	• Age 2-12 main efficacy population • Age 13-21 exploratory population
Duration	• HFMSE gains substantial by 12 months of treatment	• 12-month treatment duration
Dose	• Clear dose response with greater effect observed with 20 mg/kg over 2 mg/kg	• 20 mg/kg apitegromab dose • 10 mg/kg apitegromab dose

HFMSE=Hammersmith Functional Motor Scale Expanded.
*Nonambulatory is defined as unable to independently ambulate without aids or orthotics over 10 steps at time of walk test during screening.

SAPPHIRE Trial Design – Calibrated and Targeted

SAPPHIRE

Randomized, double-blind, placebo-controlled (enrolled n=188)
Enrolled patients receiving SMN-targeted therapy (nusinersen or risdiplam)
Completed enrollment in 3Q 2023



Exploratory Population

N = 32, Ages 13-21
2:1 apitegromab 20 mg/kg vs placebo

Stratification (across main and exploratory)

Age at SMN therapy initiation (age < 5 vs age ≥ 5)
SMN-targeted therapy (nusinersen vs. risdiplam)

Endpoints

Primary Efficacy:
Mean HFMSE change from baseline at 12 months

Additional Efficacy Measures:
RULM, WHO, other outcome measures

Safety, PK/PD, ADA

Nonambulatory is defined as unable to independently ambulate without aids or orthotics over 10 steps at time of walk test during screening

SAPPHIRE – Clear Goals

SAPPHIRE

Study Aims to Demonstrate

- **Improved motor function** (HFMSE[1] and RULM) across broad SMA patients ages 2-21 and regardless of type of SMN-targeted therapy
- **Improved caregiver-reported outcomes** (PROMIS-Fatigue and PEDI-CAT) consistent with motor function improvement
- **Safety and tolerability profile** that supports long-term use

[1] Primary efficacy endpoint of the SAPPHIRE trial

Well Tolerated Safety Profile & Low Discontinuation Rate

RIGHT TARGET → Myostatin

RIGHT TIME → Latent Form

- **>90%** of patients on combination therapy remain on study after 4 years of treatment[1]
- **Consistent** treatment-emergent adverse events (TEAEs) with previous reports with no new findings after >200 patient years of exposure in SMA[2]
  - Most frequently reported TEAEs included headache, pyrexia, COVID-19, nasopharyngitis, & upper respiratory tract infection
  - TEAEs were mostly mild to moderate and generally consistent with the underlying patient population and nusinersen therapy
- **No** treatment-related serious AEs or hypersensitivity reactions
- **No** report of positive anti-apitegromab antibodies (ADA)

[1] Excludes patients on monotherapy
[2] Data cutoff date: April 2024

AGENDA

Apitegromab for SMA Development Program

Pivotal SAPPHIRE trial: Why we are confident

Evidence supporting apitegromab's potential

Where we plan to go

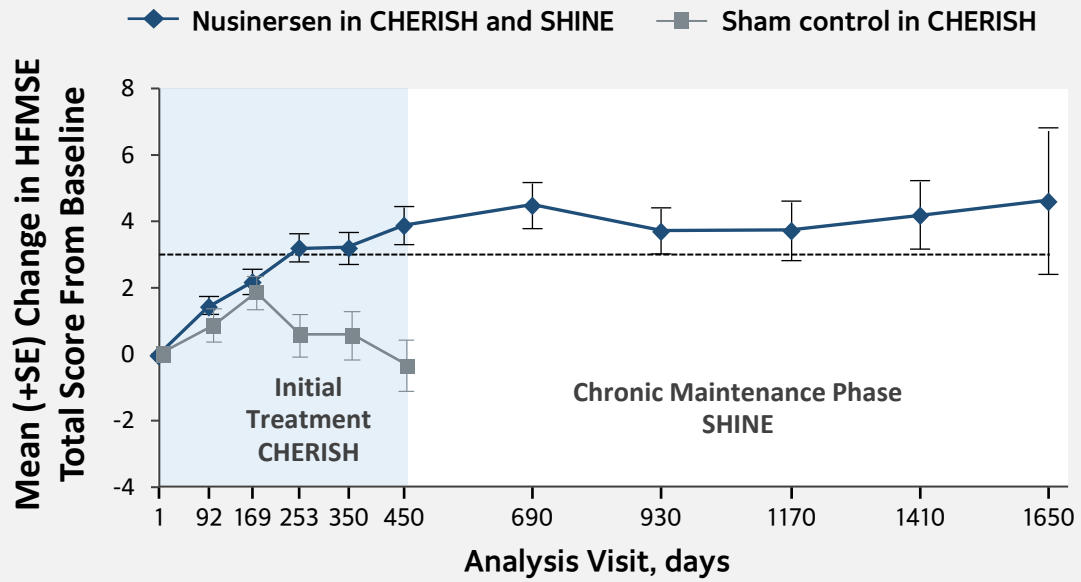
Evidence from TOPAZ to Support Apitegromab's Potential in SMA

1. SMN therapies generally plateau after initial gains
2. Substantial HFMSE gains on top of SMN therapy
3. Motor function improvement showed dose response and no clear correlation with nusinersen exposure
4. Caregiver-reported outcomes demonstrated improved fatigue and daily activities
5. High patient retention on study

Unmet Need to Address Muscle Atrophy

Change in HFMSE Over Four Years with Nusinersen[1]

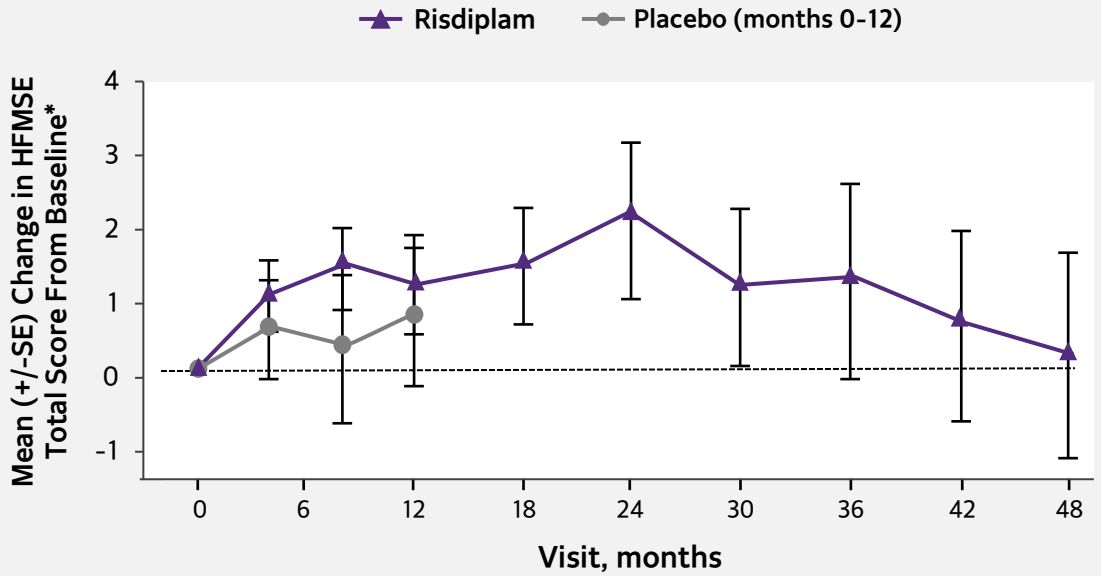
Overall population age 2-12



Nusinersen n=	84	82	84	84	83	76	83	83	79	61	20
Placebo n=	42	41	41	42	42	39					

Change in HFMSE* Over Four Years with Risdiplam[2]

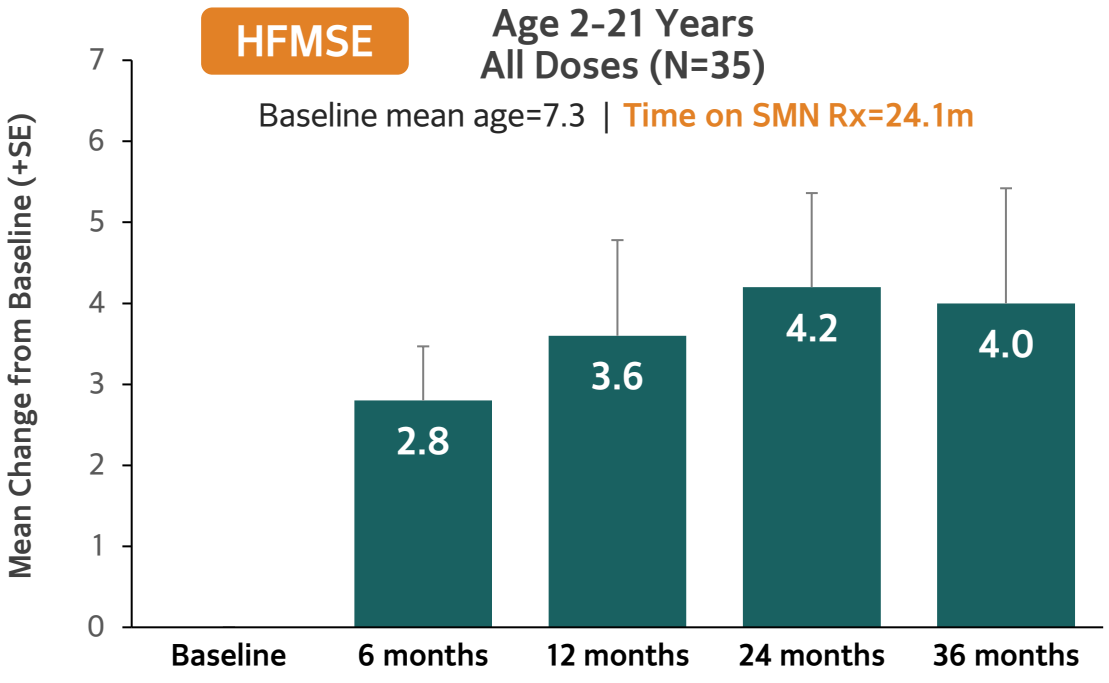
Overall population age 2-25



Risdiplam n=	120	120	119	117	109	106	89	99	101	97	
Placebo n=	60	60	58	58							

[1] Mercuri E, et al. Presented at: World Muscle Society Congress 2020, P. 257
[2] Oskoui M, et al. Presented at: 2021 Muscular Dystrophy Association Clinical & Scientific Conference; March 15-18, 2021. Poster 80.
HFMSE, Hammersmith Functional Motor Scale–Expanded; SE, standard error.
*MFM was primary efficacy endpoint of SUNFISH. HFMSE was a secondary endpoint. This third-party information is provided for background only and is not intended to convey or imply a comparison to the TOPAZ clinical trial results.

Motor Function Gains Were Substantial & Sustained Over 36 Months



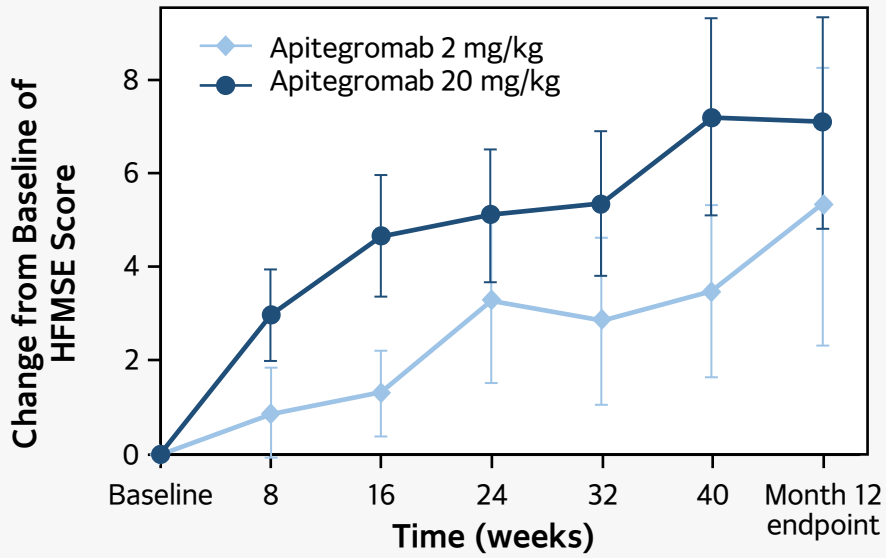
n=	35	29	32	29	28
95% CI=		(1.4, 4.1)	(1.2, 6.0)	(1.9, 6.6)	(1.0, 6.9)

- Patients enrolled in the TOPAZ study had received nusinersen for a mean of ~2 years, well into the steady maintenance phase
- HFMSE gain stands above outcomes observed well into the plateau phase of nusinersen treatment

For the 36-month evaluation, an observed case analysis was conducted, which pooled all the nonambulatory patients (Cohorts 2 and 3) and was based upon the available data for given timepoints. This analysis population 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). This analysis excludes data post scoliosis surgery from seven patients. One patient did not conduct HFMSE at time of database lock for 24 months, however, this patient had an unscheduled HFMSE score one month prior to their scheduled visit. In the most recent analysis, this result was included in the 24-month analysis. Error bars represent SE. CI represents confidence interval. SMN Rx=SMN therapy. In the age 2-21 group, 18/28 patients achieved ≥ 1-pt gains, and 11/28 patients ≥ 3-pt gains at 36 months. Data cutoff date as of March 13, 2023. 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.**

Motor Function Improvement Mainly Attributable to Apitegromab

Clear Benefit Attributable to Apitegromab

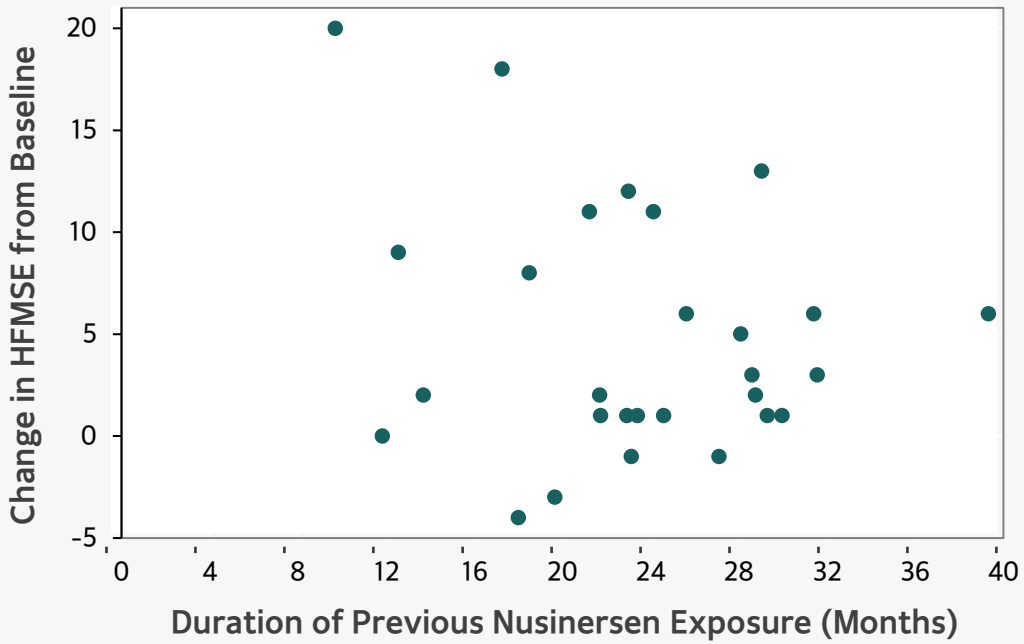


Apitegromab sample size at each visit

2 mg/kg	10	10	10	7	8	8	9
20 mg/kg	10	10	10	8	8	8	8

- Dose response observed in HFMSE in nonambulatory ≥Age 2 group randomized to 2 mg/kg and 20 mg/kg in a double-blind fashion
- Both arms showed early benefit with a greater latency of the low dose arm, supporting that the effect is mainly attributable to apitegromab

Lack of Correlation Suggests Improvement Attributable to Apitegromab

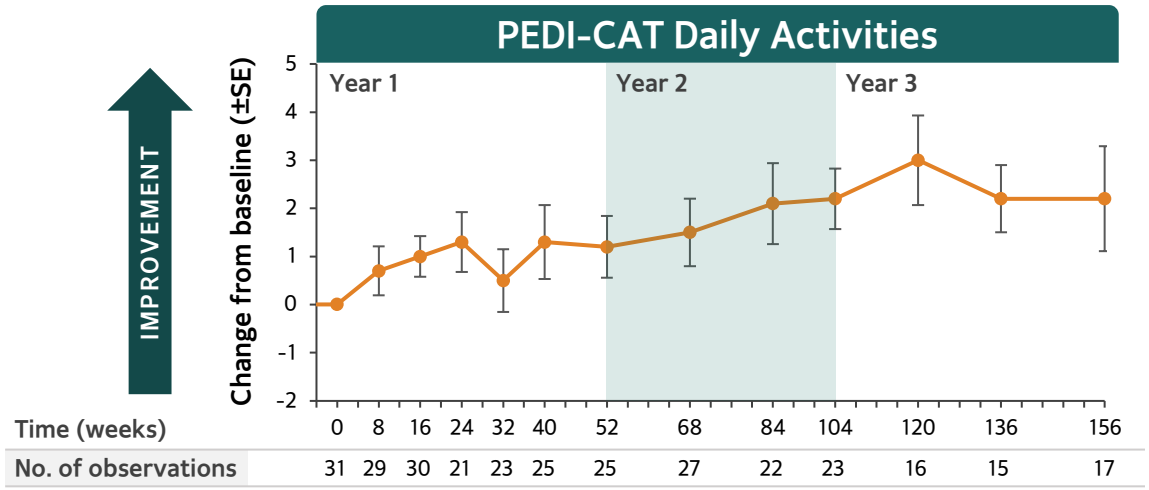


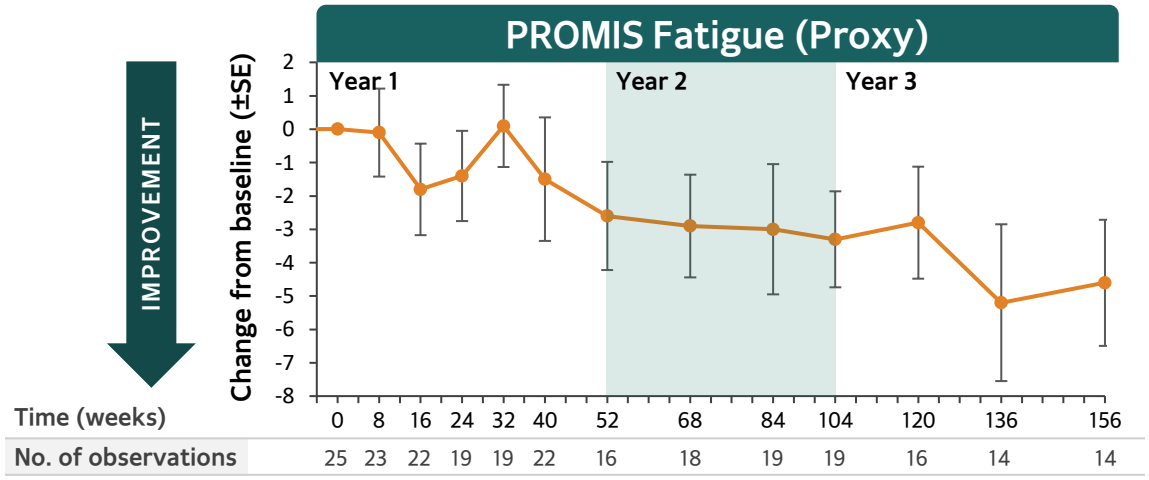
- Patients enrolled were already in the chronic maintenance phase of nusinersen (mean prior exposure ~2 years) where motor function generally plateaus
- Lack of clear correlation between 12-month HFMSE & duration of prior nusinersen exposure in patients aged 2 – 21 suggests motor function improvement mainly attributable to apitegromab

HFMSE, Hammersmith Functional Motor Scale Expanded.
Dose response graph: Crawford TO, et al. Neurology. 2024; 102 (5). Scatter plot of prior nusinersen treatment duration vs change in HFMSE from baseline, a post-hoc analysis in nonambulatory. Types 2 and 3 participants in TOPAZ. Patients skipped 3 or more doses due to COVID-site restrictions excluded;.
Apitegromab is an investigational product candidate under development.

ScholarRock

Caregivers Report Improved Self-Sufficiency and Fatigue





N = 35; Baseline mean age=7.3 | **Time on SMN Rx=24.1m**

Motor improvements:
"…since the trial she's been able to go from lying down to sitting on her own, to getting into the sitting position."

Independence:
"She has more recently gained the ability to crawl. She can get into the crawling position on her own and move across the room, and stand on her own…"

Fine Motor Skills:
"She can take lids off markers on her own. She is able to use crayons…She can brush her own teeth and dress Barbie."

Autonomy:
"She is able to access a lot of her world way more than she was able to [before the trial]."

[1] Pokrzywinski R et al. Presented at AAN, 2024.
[2] Data on file, Scholar Rock, Inc.

HFMSE=Hammersmith Functional Motor Scale Expanded; OC=observed case; PEDI-CAT=Pediatric Evaluation of Disability Inventory Computer Adaptive Test; PROMIS=Patient Reported Outcome Measurement Information System; RULM=Revised upper limb module; SE=standard error of the mean. SMN Rx=SMN therapy. Data on File. Scholar Rock, Inc. Cambridge, MA. Data cutoff date as of March 13, 2023. The updated PEDI-CAT analysis included additional records (2 at 12 months and 1 at 24 months) that were not available at the time of previous analysis. 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.**

#5 HIGH PATIENT RETENTION ON STUDY

>90 Percent of Patients on Combination Therapy Remain on Study

TOPAZ Safety Profile

Primary Treatment (12 Months)

- **58** enrolled
- **57** completed primary treatment period and enrolled in the extension study
- **1** withdrew consent

Extension Phase

- **57** enrolled
- **8** discontinued: **2** due to COVID-19 concerns; **1** due to work schedule, **5** on monotherapy due to perceived lack of benefit
- **>90%** of patients on combination therapy remain on study with 4 years of treatment[1]

~94% (33/35) of nonambulatory* patients remain on study with 4 years of treatment

[1]Excludes patients on monotherapy. Data cutoff date: April 2024
*Nonambulatory is defined as unable to independently ambulate without aids or orthotics over 10 steps at time of walk test during screening.

AGENDA

Apitegromab for SMA Development Program

Pivotal SAPPHIRE trial: Why we are confident

Evidence supporting apitegromab's potential

Where we plan to go

Where We Plan to Go: Expanding to Benefit More People Living with SMA

2024

SAPPHIRE

Q4 Data Readout

2025

- **BLA /MAA Filing**
- **Regulatory Approval***
- **Study in SMA Patients < 2 Years of Age**
  - Study design endorsed by EMA's paediatric committee
  - Study initiation planned for 2025

*Pending approval

IN SUMMARY

Advancing a Novel Muscle-Targeted Therapy for SMA

- High confidence based on proof-of-concept data in SMA
- Q4 pivotal readout with potential 2025 regulatory approval & commercialization
- Studies planned to support additional indications for apitegromab

Apitegromab: Commercial Readiness

Tracey Sacco
Chief Commercial Officer

ScholarRock

AGENDA

Apitegromab: Commercial Readiness

SMA market insights

Commercialization planning

Our Purpose: Create Possibilities for Those Living with SMA

> Muscle is everything. I want to live knowing that I have the strength **to take care of myself** if left alone.
>
> - Lyza

Listening to the Customer Voice to Stay Focused on Our Purpose

Building Deep Insights

- **>15** market research and insights projects with US, EU, and UK participants
  - → **250+** caregivers of or people living with SMA
  - → **340+** HCPs, including both neurologists and physical therapists
  - → **60+** payer insights
- Ongoing discussions with SMA patient advocacy organizations and SMA treaters

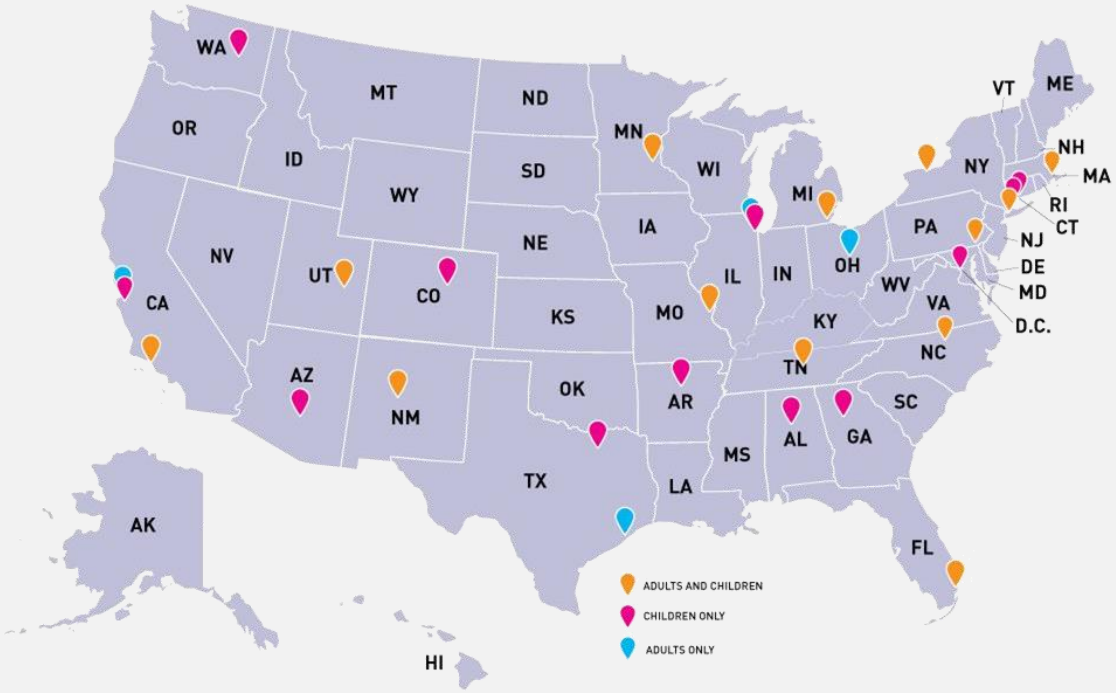
Source: Scholar Rock Internal Research 2022-2024
HCP=Healthcare Professional

SMA Has Evolved – Today Patients are Diagnosed, Treated, and Surviving Longer

SMA Has Evolved

- Newborn screening leading to earlier treatment
- Broad and expanding global access to treatment
- Concentrated care in US and EU
- Engaged and organized global patient advocacy

Cure SMA Care Center Network



Sources: https://www.curesma.org/sma-care-center-network accessed April 22, 2024; https://odysma.sma-europe.eu accessed April 22, 2024; Internal Scholar Rock market research

Patient Demand, HCP Feedback, and Payer Proactivity Suggest: More is Still Needed

Patient Demand

% of patients who identified ***improvement in muscle strength*** as what they seek most from a new treatment in SMA

97%

Physician Feedback

% of SMA patients for whom HCPs ***would prescribe a muscle targeted therapy***

Payer Proactivity

% of US commercial lives whose plan already covers ***combination SMA therapy****

1/3

Sources: Cure SMA. Education on adult patient expectations according to copy number and disease status at time of report. September 2022. Internal Scholar Rock market research; Managed Markets Insight & Technology, LLC
*As measured by covered lives; coverage for SMA approved therapy following demonstrated decline post-treatment with SMA approved gene therapy

Progressive Muscle Weakness Remains a Core Unmet Need

> Personal hygiene, using the toilet and the shower on my own would be huge. My four-year-old can do it on her own. It's degrading.
>
> – **US Patient**

> Muscle atrophy and loss of strength is a key issue in these patients. Increasing a patients' HFMSE score is really important. Its measurable and meaningful.
>
> – **Pediatric Neurologist (UK)**

> Patients treated with [nusinersen and risdiplam] are receiving only modest benefit. We need to restore motor function that enables practical improvement in daily activities / independence.
>
> – **National MCO**

Source: Internal Scholar Rock market research
HFMSE = Hammersmith Functional Motor Scale Extended; MCO = Managed Care Organization

SMA is the Right Opportunity for Scholar Rock's First Launch

1.

Clear Unmet Need

- Patient, HCP, and payer communities recognize remaining needs in SMA

2.

Concentrated and Proactive Care

- Patients already diagnosed and treated proactively
- High concentration of care

3.

Engaged Patient Community

- Proactive advocacy, patient, and caregiver community
- Aligned on need for muscle targeted therapy

4.

Payer Receptivity

- Established value of improving motor function

AGENDA

Apitegromab: Commercial Readiness

SMA market insights

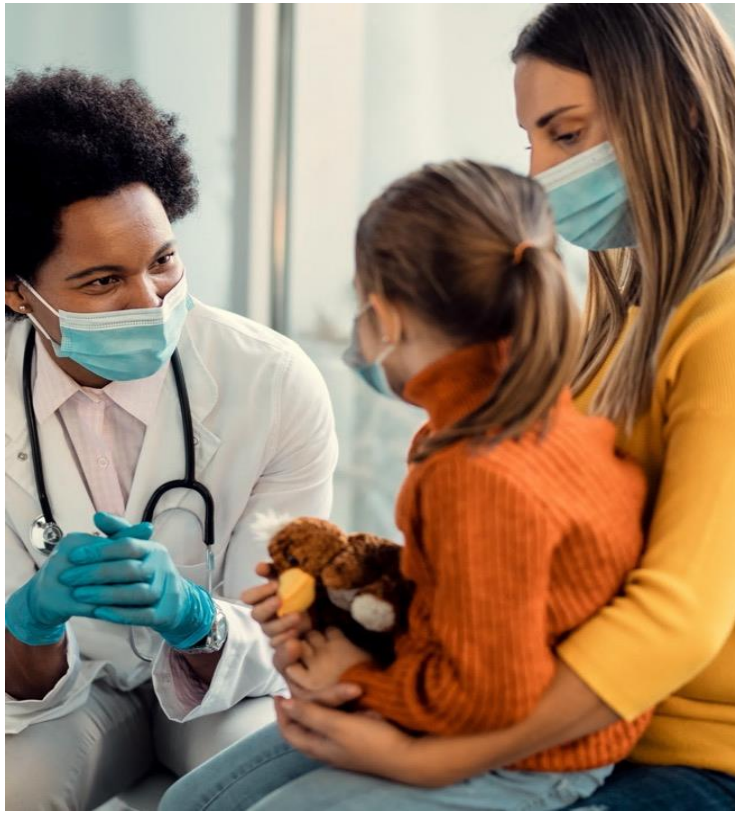
Commercialization planning

Why We Are Confident About Potential Commercial Success of Apitegromab

Gold standard efficacy measure in SMA
- HFMSE is a SMA-specific, validated functional scale
- Commonly used in practice by both HCPs and payers

Long-term treatment experience in SMA patients
- SMA patients treated >4 years on apitegromab[1]
- High retention rate in TOPAZ

Fits into current SMA practice
- Used with nusinersen or risdiplam
- Monthly dosing



[1] Ph 2 TOPAZ trial (apitegromab in SMA), data as of April 2024
HFMSE = Hammersmith Functional Motor Scale Extended

SMA Represents a Significant Opportunity for Apitegromab

~$4.5B[1]

global revenue for three SMN targeted therapies

SPINRAZA® (nusinersen) injection 12 mg/5 mL

Evrysdi™ risdiplam 60 mg powder for oral solution

zolgensma® (onasemnogene abeparvovec-xioi) suspension for intravenous infusion

Apitegromab global revenue potential

>$1.0B[2]

[1] Revenue as of Biogen 4Q23 financial update, Roche 4Q23 financial update, and Novartis 4Q23 financial update
[2] Scholar Rock internal estimates as of May 2024
SMA=Spinal muscular atrophy; SMN=Survival motor neuron.

ScholarRock

Commercialization Approach: Three Key Elements

1. Engagement

- Partnering with SMA communities

2. Patient Experience

- Ensure excellent patient experience

3. Execution

- Build team to deliver US launch & global expansion

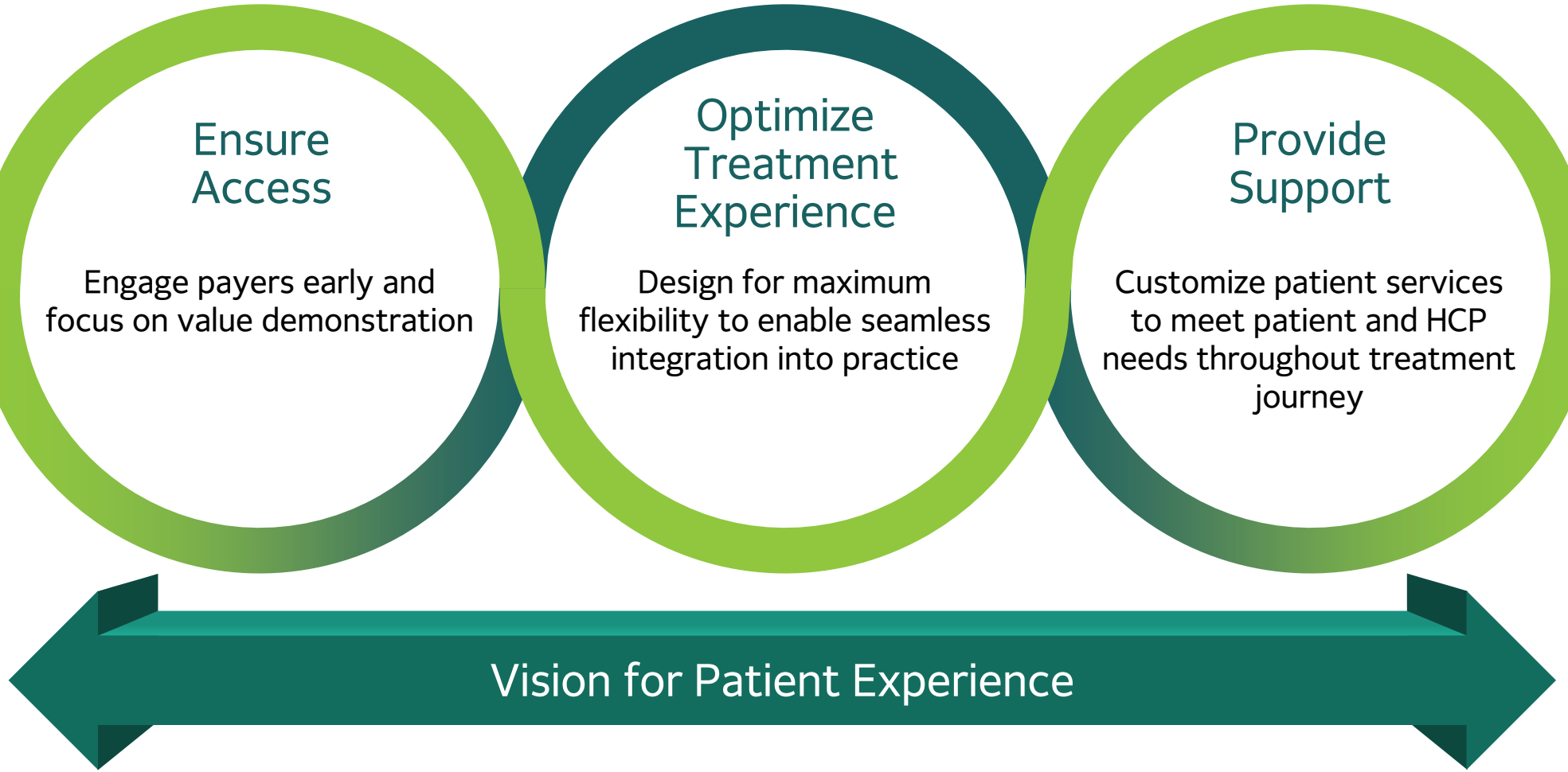
Commercialization Plan

Partnering With the SMA Community

- MSL team on the ground
- Continued engagement with US and EU patient advocacy
- Amplify patient voice with muscle-focused education

MSL= Medical Science Liaison

Secure Access and Customize Treatment to Meet Patient Needs



Building to Achieve Commercial Success

Building the Foundation

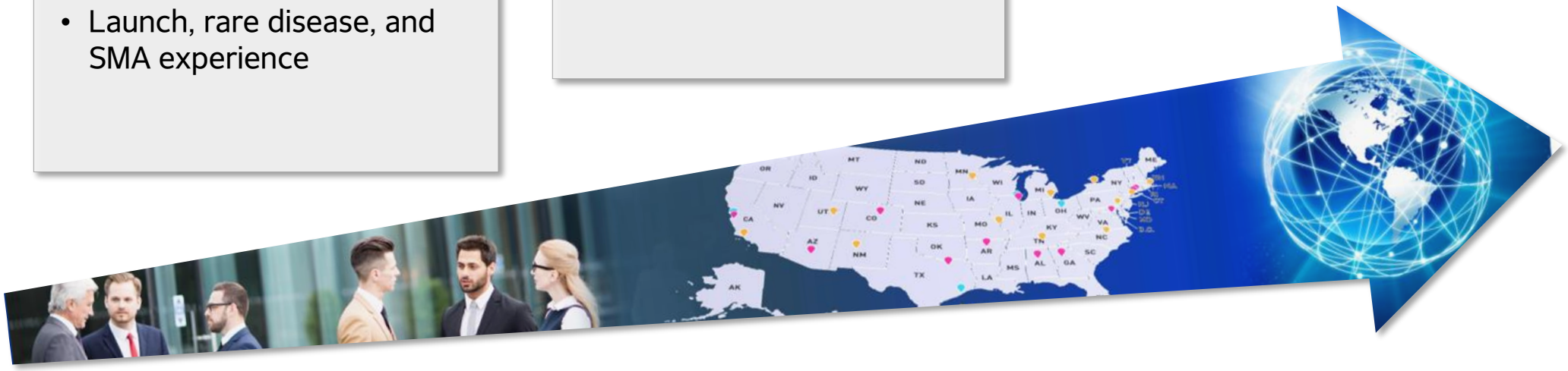
- Focused and experienced team
- Launch, rare disease, and SMA experience

U.S. Launch*

- Expand commercial capabilities
- Efficient US customer facing footprint (30-50 FTEs)

Geographic Expansion

- Commercialize in selected European countries*
- Remaining EU and ROW expansion through distributorships & partnerships



2024 2025 2026+

* Subject to regulatory approval
ROW= Rest of world

IN SUMMARY

Path to Achieving Commercial Success in SMA

The right market

→ Clear unmet need and favorable market dynamics

The right medicine

→ Competitive and attractive potential profile

The right plan

→ Engagement, patient-focus & execution

[1] Scholar Rock internal estimates as of May 2024

SRK-439: Selective Anti-Myostatin Designed for Optimal Profile in Obesity

Mo Qatanani
Chief Scientific Officer

AGENDA

A Differentiated Approach

Best-in-class platform for selectivity

Different approaches to muscle preservation

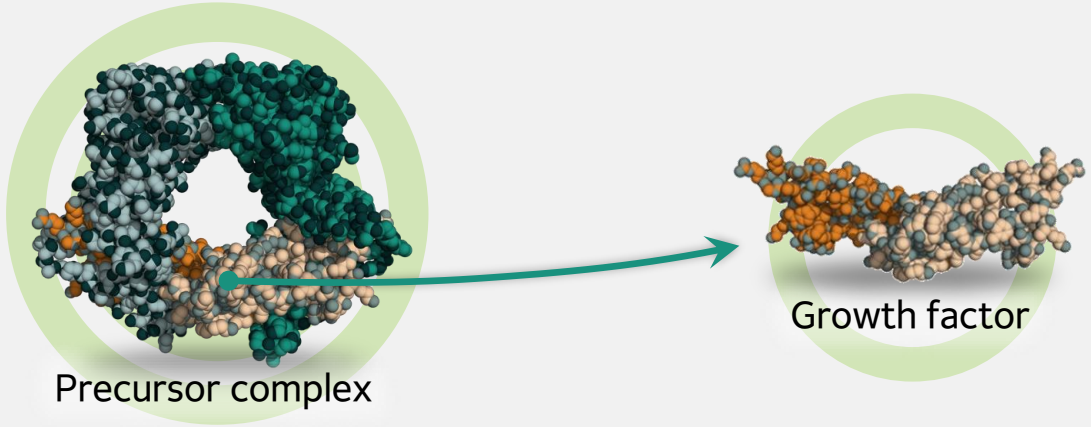
SRK-439: novel asset, differentiated profile

Differentiated Approach to Targeting Growth Factors

Advantages of Scholar Rock's Approach

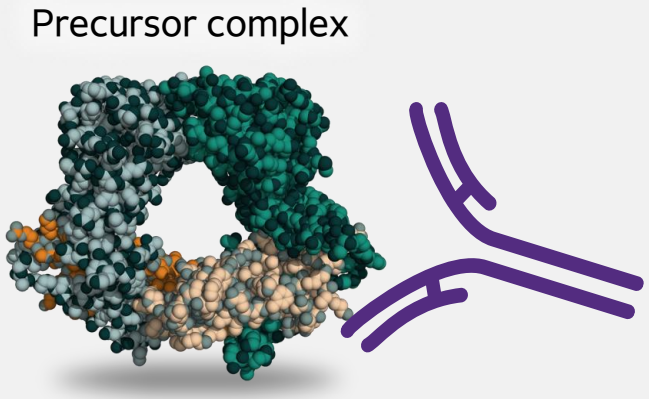
Scholar Rock's Target
Latent growth factor

Traditional Target
"Mature" active growth factor



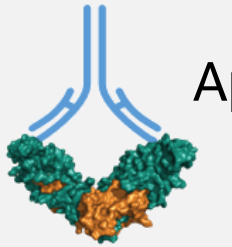
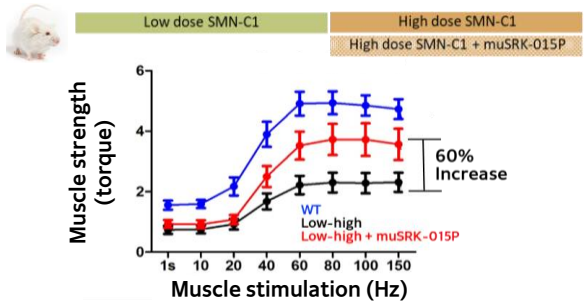
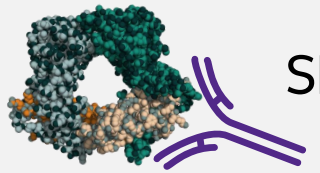
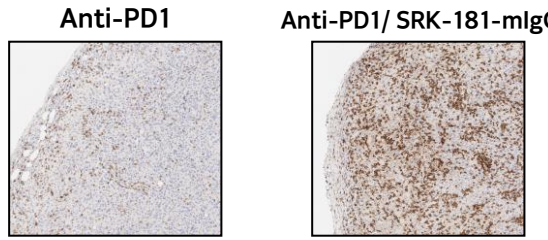
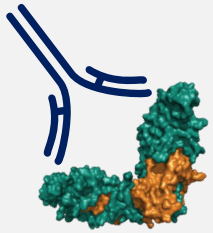
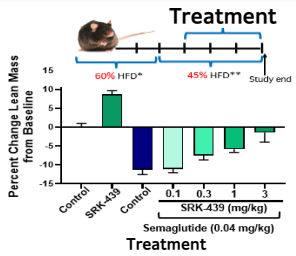
- Deep structural insights to validated targets
- Industry-leading antibody design and protein engineering to selectively target latent growth factors
- Optimized for efficacy and avoids off-target effects

Transformational Medicines



Highly selective monoclonal antibodies

Platform and Expertise Drive Success in Clinic

Superb Engineering	Translational Models	Clinic
 Apitegromab		TOPAZ SAPPHIRE ONYX
 SRK-181		DRAGON
 SRK-439		FDA IND in 2025

AGENDA

A Differentiated Approach

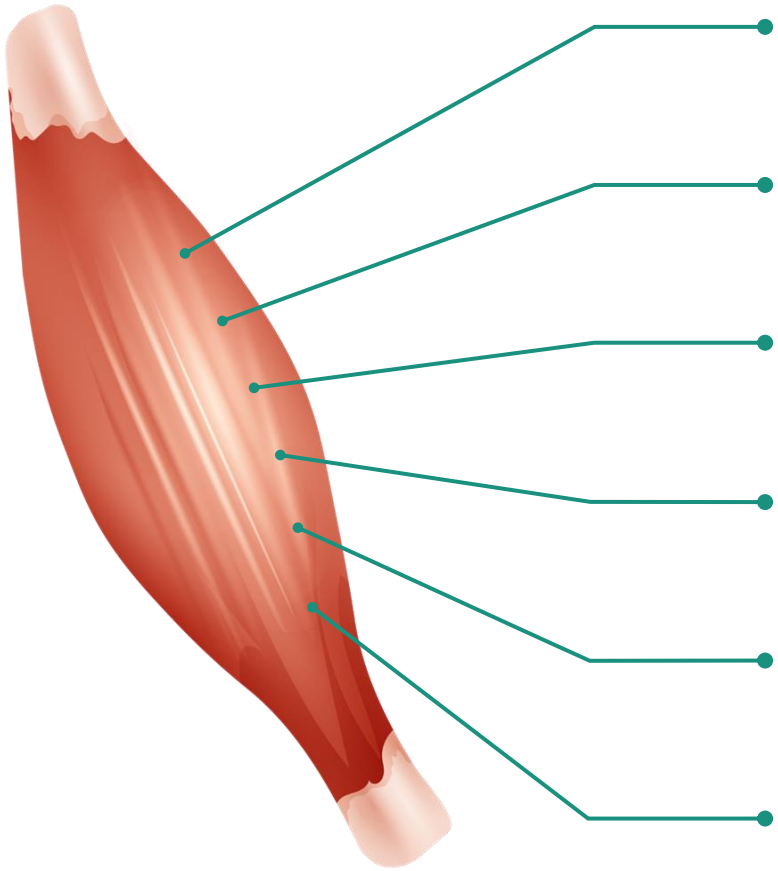
Best-in-class platform for selectivity

Different approaches to muscle preservation

SRK-439: novel asset, differentiated profile

KEY METABOLIC ORGAN

Muscle is Critical for Overall Health



- Increase basal metabolic rate (BMR)[1]
- Enhance glucose homeostasis[2] and better insulin sensitivity and lower risk of prediabetes[3]
- Signal to other systems as an endocrine organ impacting overall health[4]
- Reduce visceral fat[5]
- Increase caloric expenditure post-exercise[6]
- Increase bone density, strength, function, and longevity and decreased risk of injury and disability[7-9]

1. Aristizabal JC et al. Eur J Clin Nutr 2015; 2. Lindegaard B et al. J Clin Endocrinol Metab 2008; 3. Srikanthan P, Karlamangla AS J Clin Endocrinol Metab 2011; 4. Severinsen et al. Endocr Rev. 2020; 5. Wewege MA, et al. Sport Med 2022; 6. Zurlo F. et al. J Clin Invest 1990; 7. Fukushima Y et al. Diabetes Metab J. 2016; 8. Roh E, Choi KM. Front. Endocrinol. 2020; 9. Volpi E, et al Curr Opin Clin Nutr Metab Care. 2004

Myostatin is the Right Target for Muscle Growth

Advantages

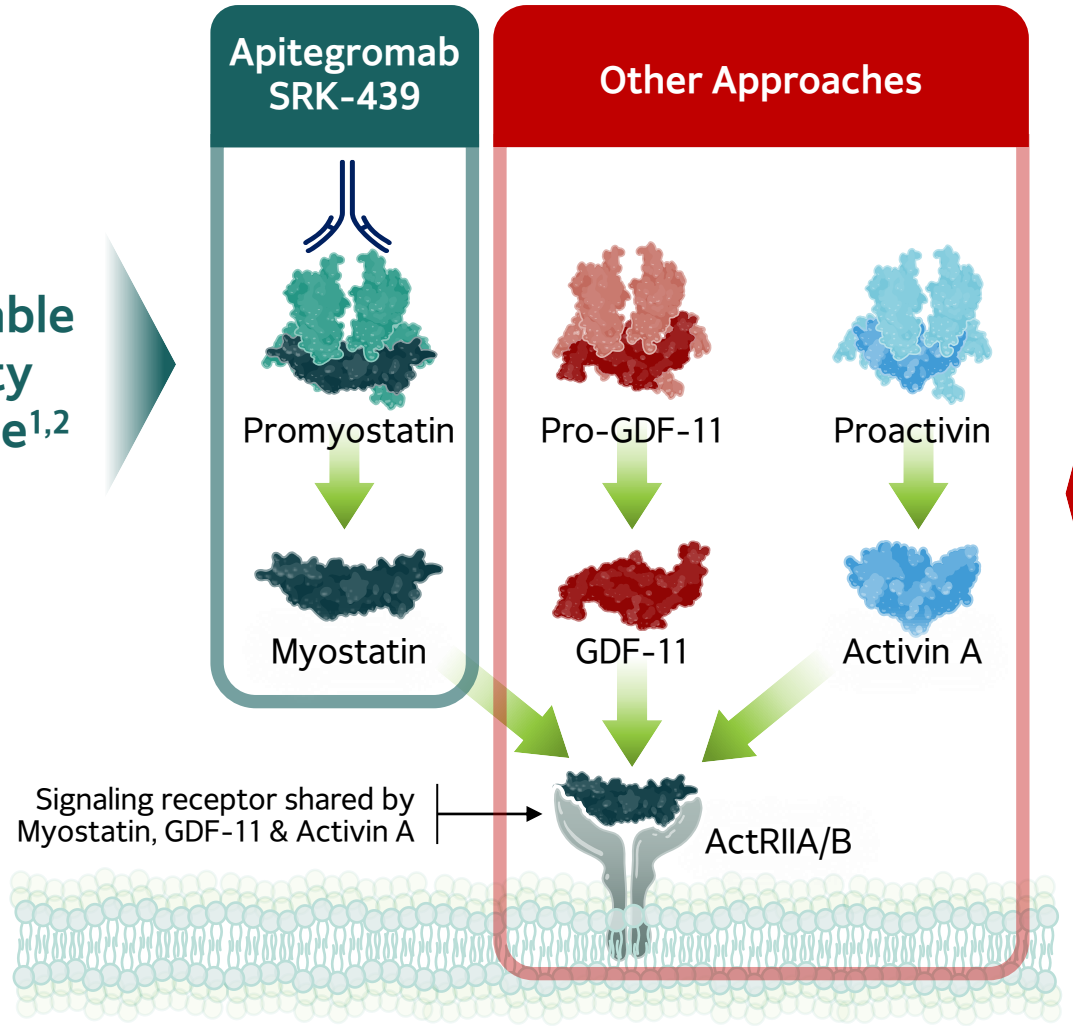
- Myostatin is specific to muscle[1]
- Validated genetically with no evident safety liabilities [1-4]
- Inhibition leads to muscle growth in adults[5]
- Selective targeting leads to improved motor function as seen in TOPAZ with favorable tolerability profile observed in >200 patient years of exposure in SMA [6]

Pictures depict increase in muscle mass in myostatin null animals and humans

1. McPherron, A.C., et al. Nature 1997; 2. Schuelke, M., et al. NEJM 2004; 3. Kambadur, R., et al Genome Res. 1997; 4. Mosher, D.S., et al. PLoS 2007; 5. Abati E, et al. Cell Mol Life Sci. 2022; 6. Ph 2 TOPAZ trial (apitegromab in SMA), data as of April 2024

Multiple Risks Associated with Non-Selective Targeting

Favorable safety profile[1,2]



Health Risks Observed with Non-Selective Inhibition of ActRII Pathway:

- GI problems, e.g., diarrhea, pancreatitis [3-6]
- Nose bleeds (epistaxis), low platelet count, telangiectasias[7-10]
- Reduction in reproductive hormones in males and females[3, 7, 11, 12]
- Acne, rash, skin abscesses[5, 13, 14]
- Madarosis (loss of eyebrows or eyelashes)[14]

1. Barrett et al., Adv Therapy 2021; 2. Crawford et al., Neurology 2024; 3. Garito T et al Clin Endocrinol 2018; 4. Amato AA et al Neurology 2021; 5. Heymsfield SB et al. JAMA 2021; 6. Vanhoutte F et al. J Clin Pharmacol 2020; 7. Attie KM et al Muscle Nerve 2013; 8. Attie KM et al Am J Hematol 2014; 9. Campbell C et al. Muscle Nerve 2017; 10. Hoeper MM et al NEJM 2023; 11. Ruckle J et al, JBMR 2009; 12. Sherman ML et al J Clin Pharm 2013; 13. Muntoni F et al. Neurol Ther. 2024. 14. Rocco MD et al Nat Med 2023;

AGENDA

A Differentiated Approach

Best-in-class platform for selectivity

Different approaches to muscle preservation

SRK-439: novel asset, differentiated profile

Why We Are Confident in SRK-439

Scholar Rock's Unique Approach

- A new anti-myostatin specifically suited for obesity

Exquisite Selectivity

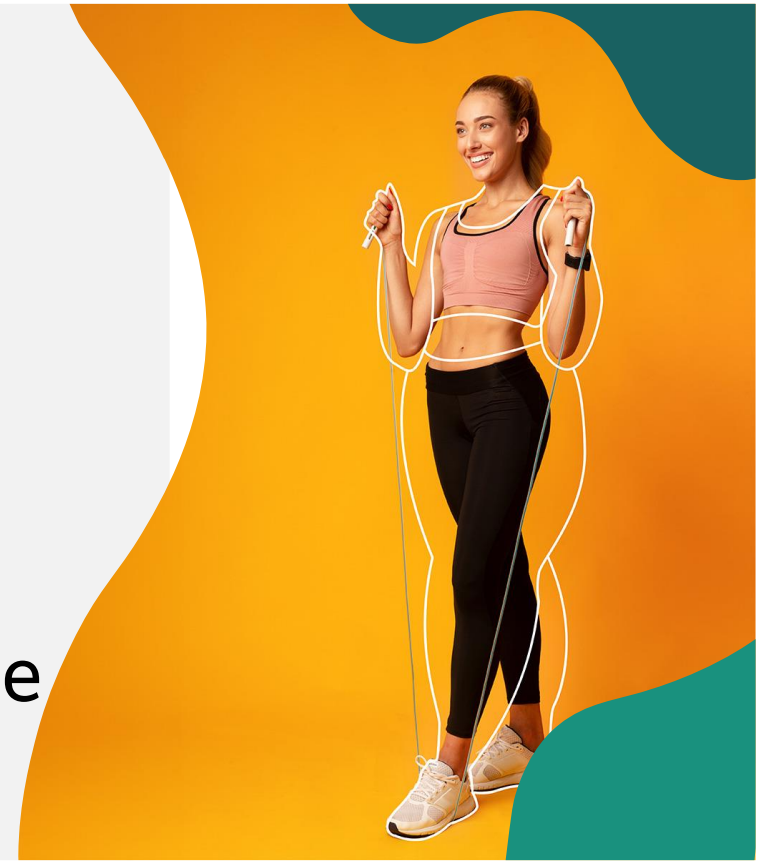
- Targets pro and latent forms of myostatin designed to minimize undesirable off-target effects

Strong Scientific Validation

- Preclinical data demonstrated favorable muscle mass preservation and metabolic effects

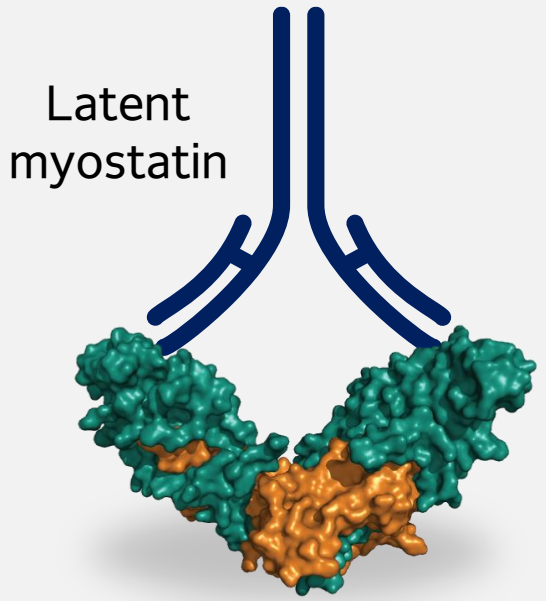
SRK-439: Differentiated Profile

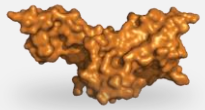
1. Exquisite selectivity for myostatin
2. Potential for healthier weight loss in combination with GLP-1 RA
3. Low efficacious dose and competitive profile



SRK-439: Exquisite Selectivity for Myostatin

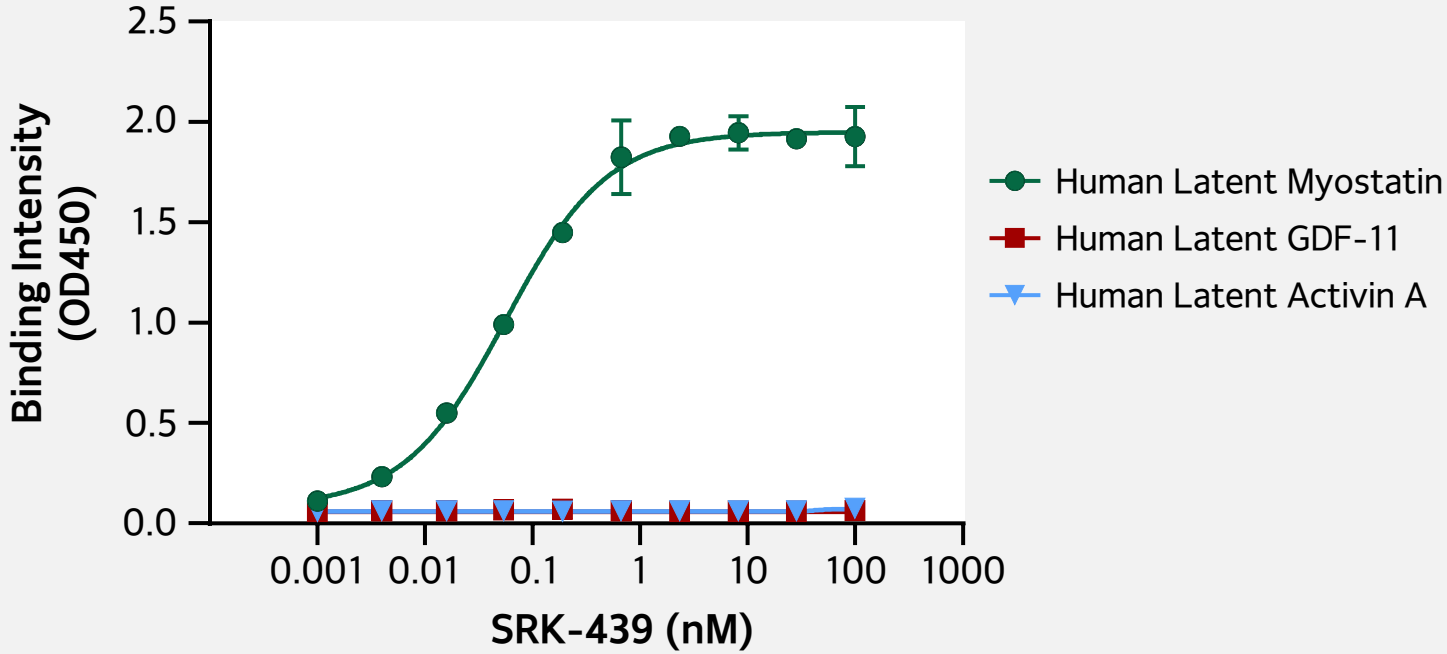
Scholar Rock's Target: Latent Myostatin





"mature" myostatin

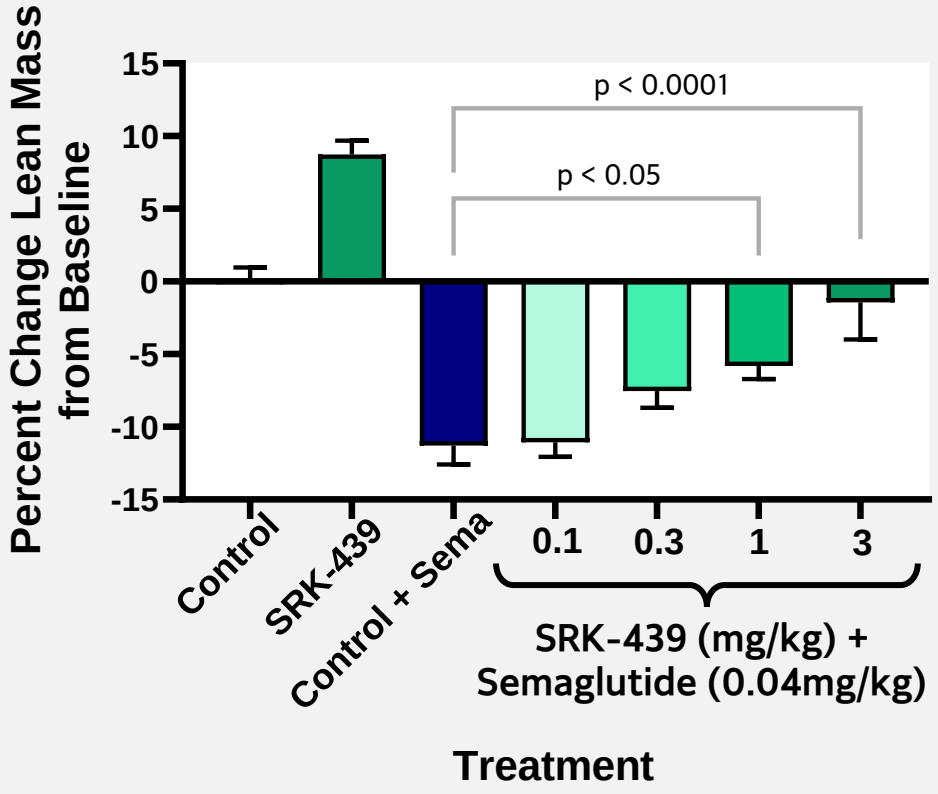
SRK-439 Selectively Binds Latent Myostatin



- Increasing SRK-439 concentrations lead to increased signal of binding to myostatin with no signal observed for GDF-11 or Activin A

SRK-439: Potential for Healthier Weight Loss Management in Combination with GLP-1 RA

SRK-439 Maintained Lean Mass in Semaglutide Treated Animals



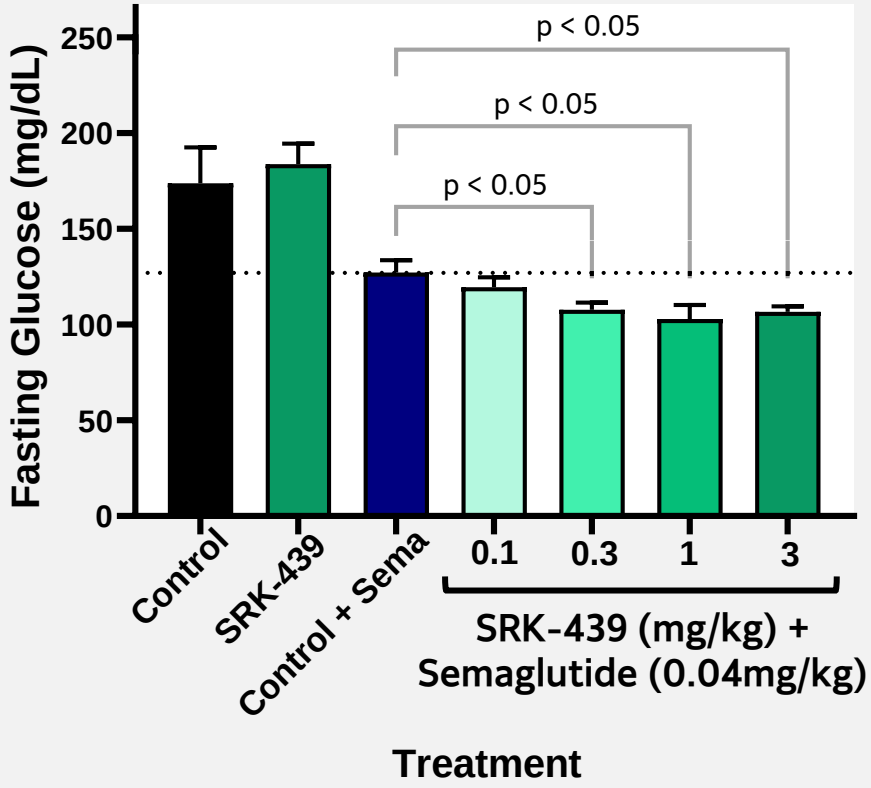
Key Observations

- Considerable lean mass loss with semaglutide treatment
- Combination with SRK-439 led to dose-dependent lean mass preservation
  - Effects seen with doses as low as 0.3 mg/kg
- Dose dependent enhancement of fat mass loss also observed, improving overall body composition

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

SRK-439: Further Improvement of Metabolic Health

SRK-439 Further Improved Fasting Glucose in Semaglutide Treated Animals

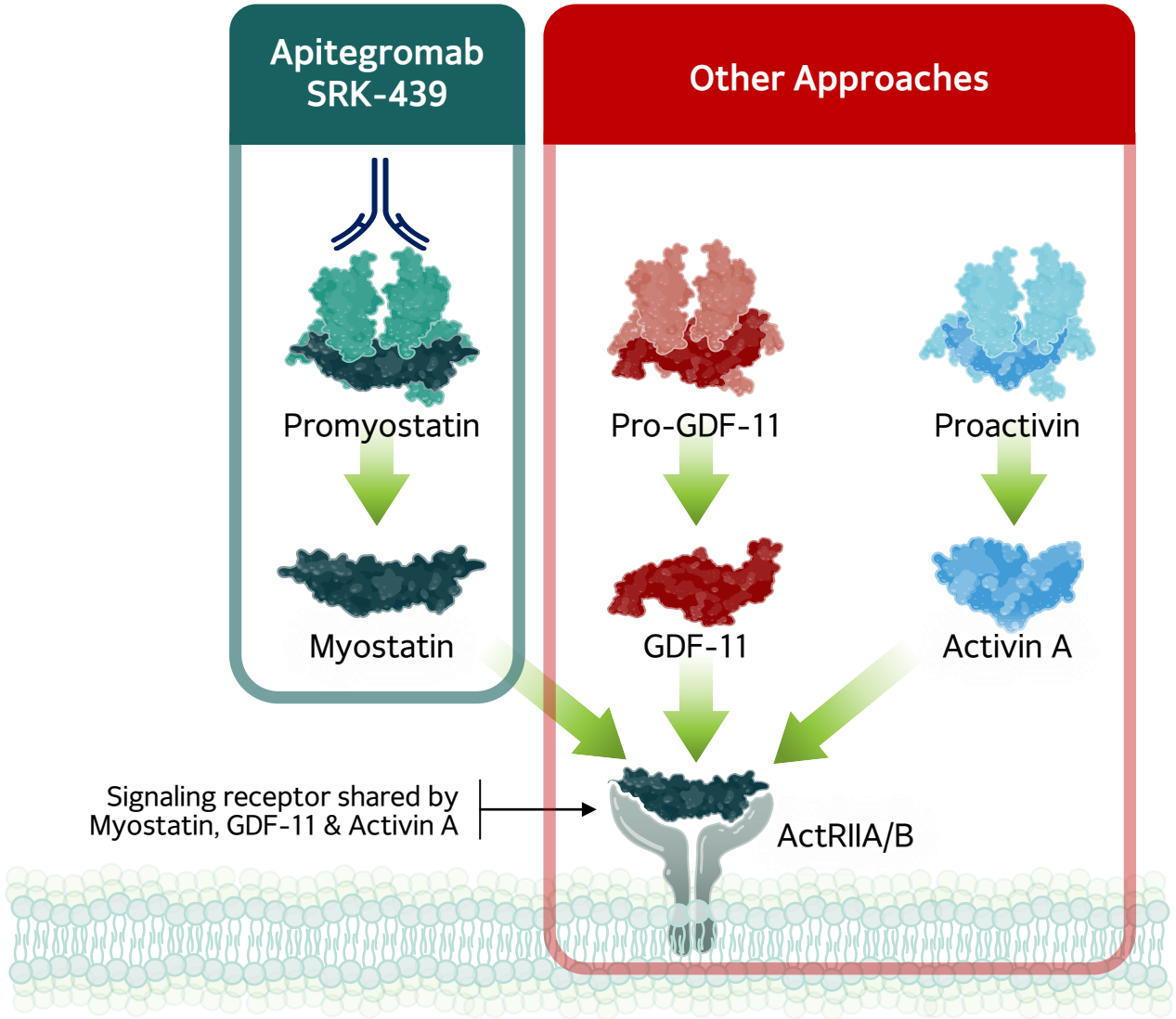


Key Observations

- Semaglutide reduced fasting glucose levels as expected
- Combination with SRK-439 led to further improvement in fasting glucose levels by ~20% in a dose-dependent manner
- Highlights the role of muscle preservation in improving long term metabolic profile

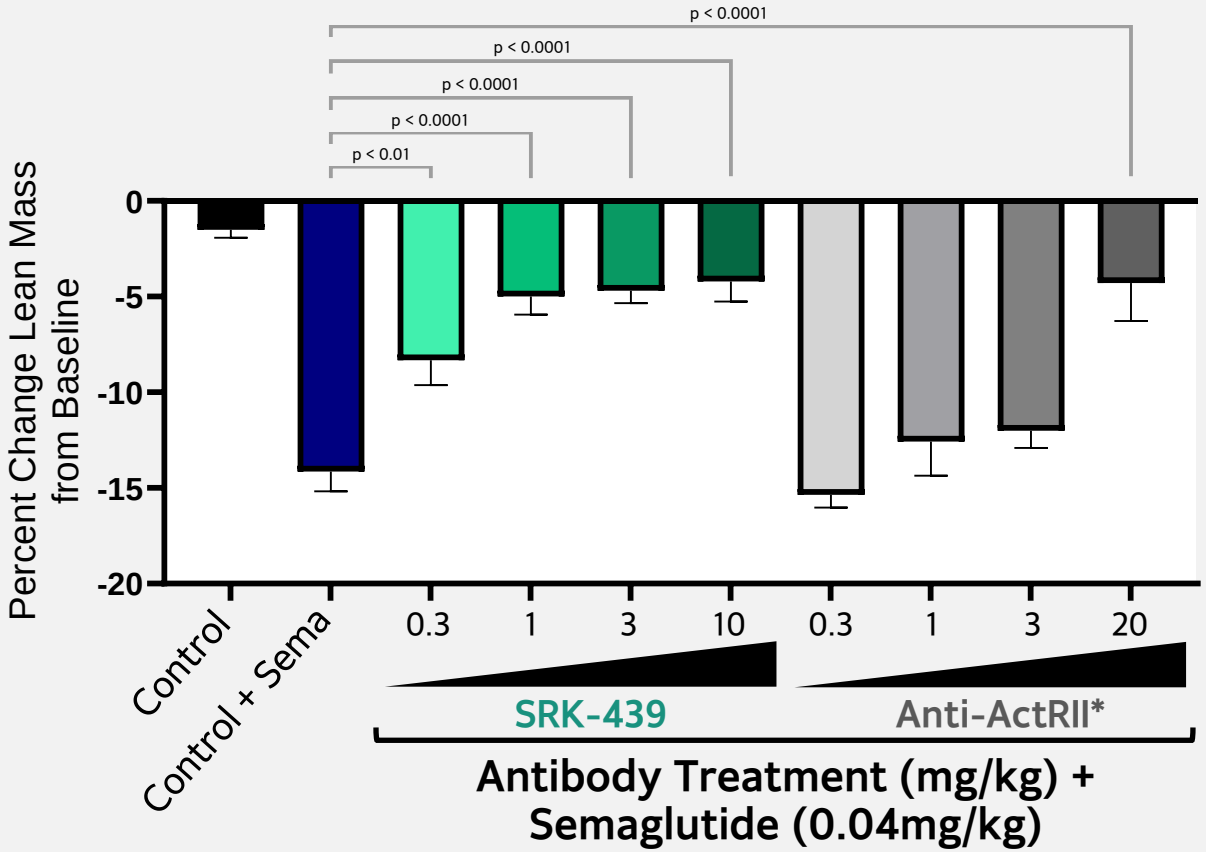
Study conducted in Diet Induced Obesity (DIO) mouse model utilizing a murine chimera of SRK-439

Are We Limiting Efficacy with Selective Targeting of Myostatin?



SRK-439: More Potent than Anti-ActRII Antibody at Maintaining Lean Mass

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



Key Observations

- SRK-439 preserved GLP-1 driven lean mass loss dose-dependently and at lower doses than anti-ActRII
- Highlights efficacy of SRK-439 and avoids potential liabilities of non-selective approach
- Low target dose of SRK-439 supports subcutaneous profile

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

SRK-439: Best in Class Potential

	SRK-439	ActRII Ab	Ligand Trap	Adnectin
Selectivity for myostatin	✓	✗	✗	✗
Action limited to muscle	✓	✗	✗	✗
Combination GLP-1 RA data in obesity preclinical models[1-3]	✓	✓	✓	✓
Low efficacious dose in preclinical obesity models[1-3]	✓	✗	✗	✗
Lower risk of potential undesirable effects in clinic[4]	✓	✗	✗	✗

GLP-1 RA: GLP1 receptor agonist

1. Nunn E, et al., Mol Metab 2024; 2. Schang G., et al, J. Endoc Soc 2023; 3. Ackerman, P, et al. Obesity Week 2023 Poster 211;
4. See also references on slide titled, “Multiple Risks Associated with Non-Selective Targeting” in this presentation.

IN SUMMARY

SRK-439: The Right Molecule for Healthy Weight Loss

The right target

→ Highly selective approach

The right tissue

→ Targets muscle

The right safety profile

→ Efficacy without potential liabilities of non-selective approaches

The right product profile

→ Designed for subcutaneous low frequency dosing with robust subcutaneous exposure and long half life

Cardiometabolic Development Program

Aiming for Healthier Weight Loss

Jing Marantz
Chief Medical Officer

ScholarRock

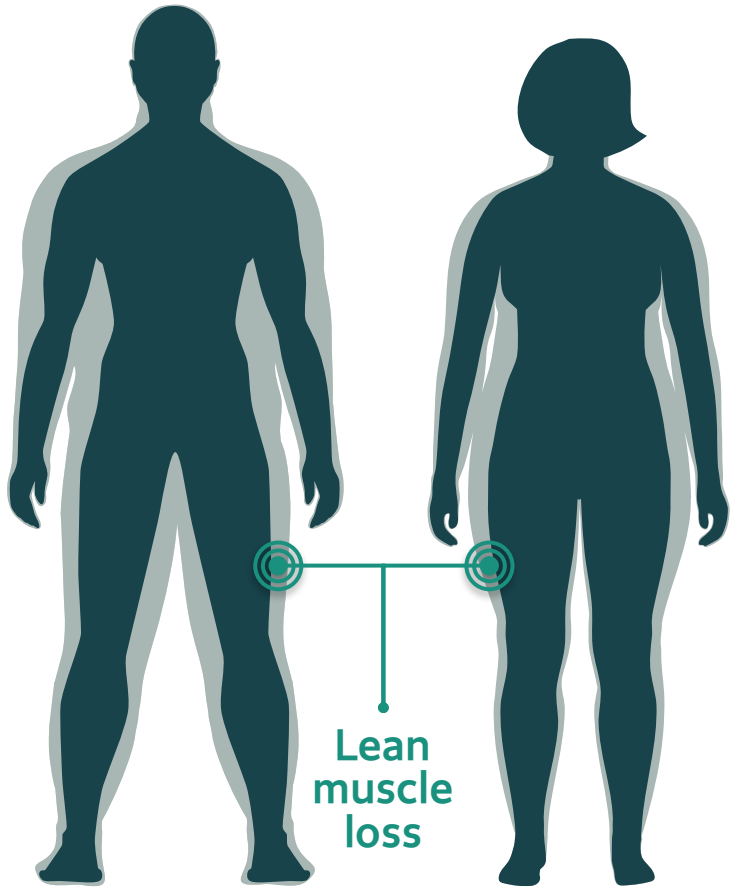
AGENDA

Cardiometabolic Development Program

Overview of muscle-targeted opportunity

SRK-439 Development Pathway

Significant Proportion of Weight Loss Due to Loss of Lean Muscle Mass



Current weight loss strategies challenged by:

- Tolerability
- Lack of durability
- **Significant muscle loss**[1-3]

GLP-1 Agonist=Glucagon-like peptide-1 receptor agonist

1. Muller TD, et al Anti-obesity drug discovery: advances and challenges. Nature Reviews Drug Discovery 2022; 21, 201–223;
2. Wilding JPH, Batterham RL, Calanna S, et al. Once-Weekly Semaglutide in Adults with Overweight or Obesity. N Engl J Med. 2021;384(11):989-1002;
3. Jastreboff AM, et al Tirzepatide Once Weekly for the Treatment of Obesity. NEJM 2022; 387 (3): 205-216;

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Lean Muscle Is Essential to Healthy Metabolic Function

- **Increased** basal metabolic rate[1]
- **Enhanced** glucose homeostasis[2]
- **Better** insulin sensitivity and lower risk of prediabetes[3]
- **Reduced** visceral fat[4]
- **Increased** caloric expenditure post-exercise[5]
- **Increased** bone density, strength, function, longevity, and decreased risk of injury and disability[6-8]

GLP-1 Agonist=Glucagon-like peptide-1 receptor agonist

1. Aristizabal JC, Freidenreich DJ, Volk BM, et al. Effect of resistance training on resting metabolic rate and its estimation by a dual-energy X-ray absorptiometry metabolic map. Eur J Clin Nutr.2015; 69, 831–836. https://doi.org/10.1038/ejcn.2014.216; 2. Lindegaard B, Hansen T, Hvid T, et al. The effect of strength and endurance training on insulin sensitivity and fat distribution in human immunodeficiency virus-infected patients with lipodystrophy. J Clin Endocrinol Metab. 2008; 93:3860–9; 3. Srikanthan P, Karlamangla AS. Relative muscle mass is inversely associated with insulin resistance and prediabetes. Findings from the third National Health and Nutrition Examination Survey. J Clin Endocrinol Metab. 2011; 96:2898–903. doi: 10.1210/jc.2011-0435; 4. Wewege MA, Desai I, Honey C, et al. The effect of resistance training in healthy adults on Body fat percentage, fat mass and visceral fat: A systematic review and meta-analysis. Sports Med. 2022(Feb);52(2):287-300. doi: 10.1007/s40279-021-01562-2; 5. Zurlo, F., Larson, K., Bogardus, C., et al. Skeletal muscle metabolism is a major determinant of resting energy expenditure. J Clin Invest. 1990;86(5), 1423-1427; 6. Fukushima Y, Kurose S, Shinno H, et al. Importance of lean muscle maintenance to improve insulin resistance by body weight reduction in female patients with obesity. Diabetes Metab J. 2016;40: 147-153; 7. Roh E, Choi KM. Health consequences of sarcopenic obesity: a narrative review. Front. Endocrinol. 2020;11: 332; 8. Volpi E, Nazemi R, Fujita S. Muscle tissue changes with aging. Curr Opin Clin Nutr Metab Care. 2004;7(4): 405-410.

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Our Solution Delivers Attractive Clinical Risk/Benefit Profile

Key Points

- **Inhibition of myostatin**, a negative regulator of muscle, is known to promote muscle growth and function
- Apitegromab, a selective myostatin inhibitor, has been shown in a Phase 2 Proof-of-Concept study to **improve motor function**
- Preserving muscle, an endocrine organ with important role in energy metabolism, has the potential to **improve durability of weight loss**
- Selective targeting **minimizes off-target effects**, potentially supporting long-term use for healthy weight management

Potential Benefits

- **Preserving muscle**
- **Improving function**
- **Improving durability**
- **Long-term safety**

AGENDA

Cardiometabolic Development Program

Overview of muscle-targeted opportunity

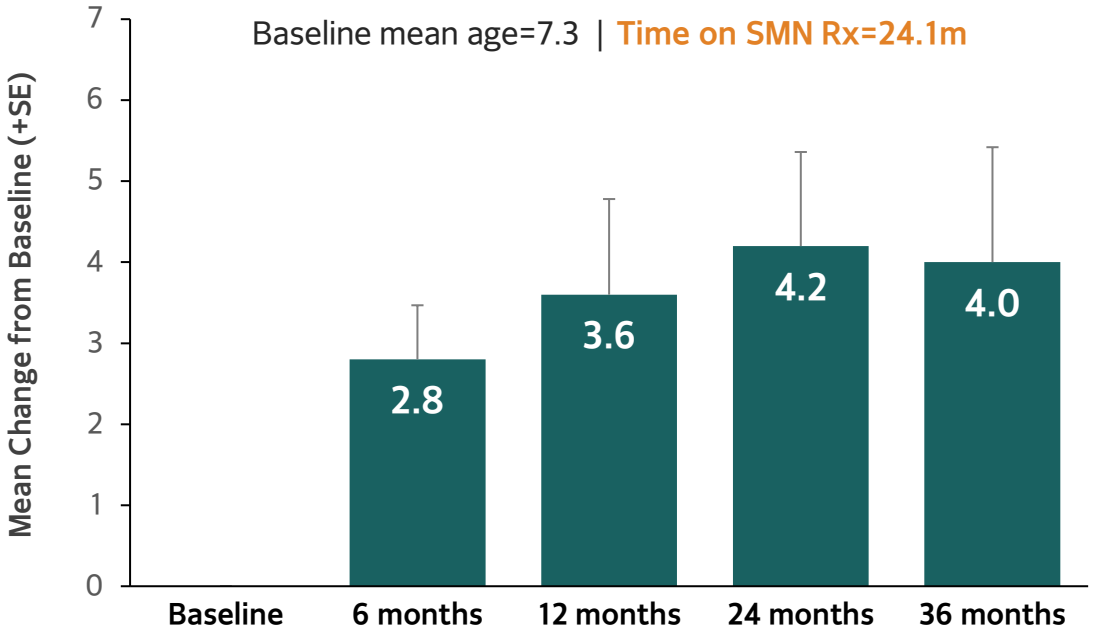
SRK-439 Development Pathway

Apitegromab in SMA Improved Motor Function from Baseline

HFMSE Gain

Age 2-21 Years
Pooled Nonambulatory Patients (N=35)

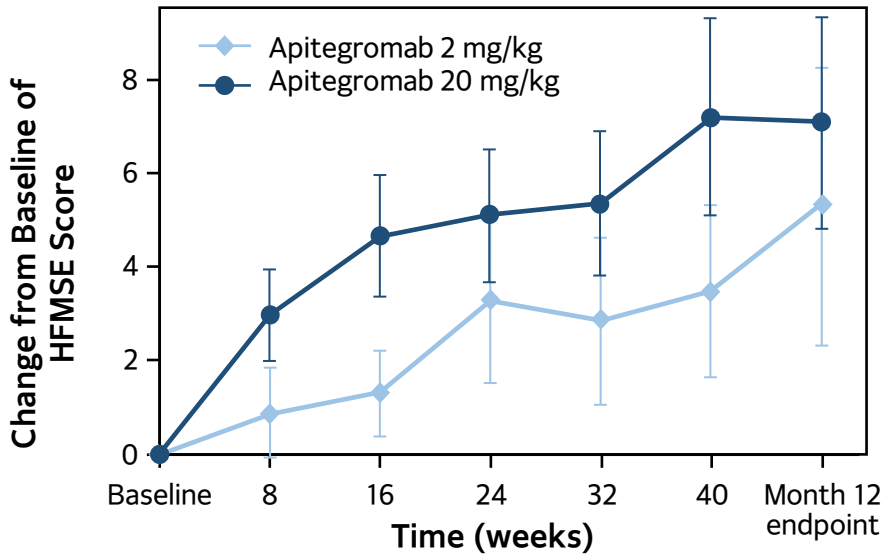
Baseline mean age=7.3 | Time on SMN Rx=24.1m



n=	35	29	32	29	28
95% CI=		(1.4, 4.1)	(1.2, 6.0)	(1.9, 6.6)	(1.0, 6.9)

Dose Response in HFMSE

TOPAZ nonambulatory Cohort 3 patients randomized to 2 mg/kg and 20 mg/kg in a double-blind fashion

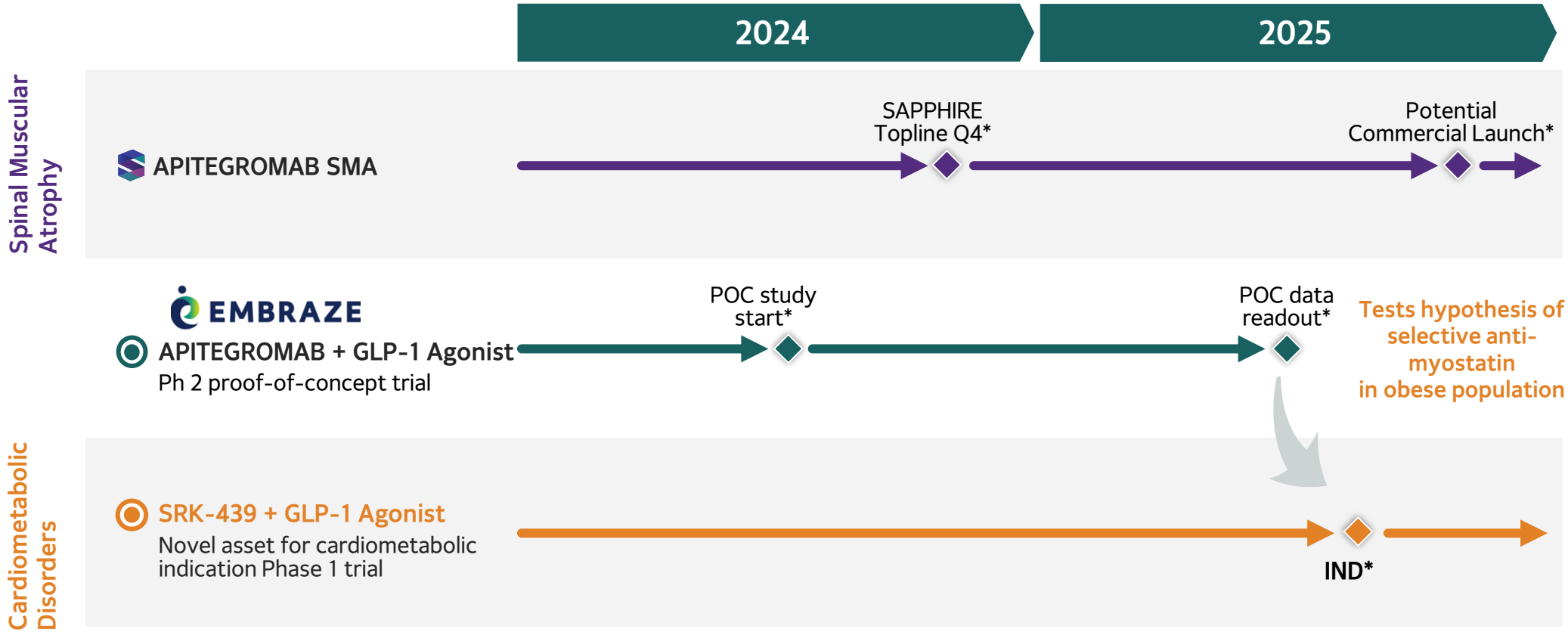


Apitegromab sample size at each visit

2 mg/kg	10	10	10	7	8	8	9
20 mg/kg	10	10	10	8	8	8	8

HFMSE, Hammersmith Functional Motor Scale Expanded.
Dose response graph: Crawford TO, et al. Neurology. 2024; 102:e209151
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.

Leveraging Apitegromab POC Study to Inform SRK-439 Development



Expected timelines
POC = Proof of Concept

EMBRAZE Proof-of-Concept Study

EMBRAZE Study Aims to Demonstrate

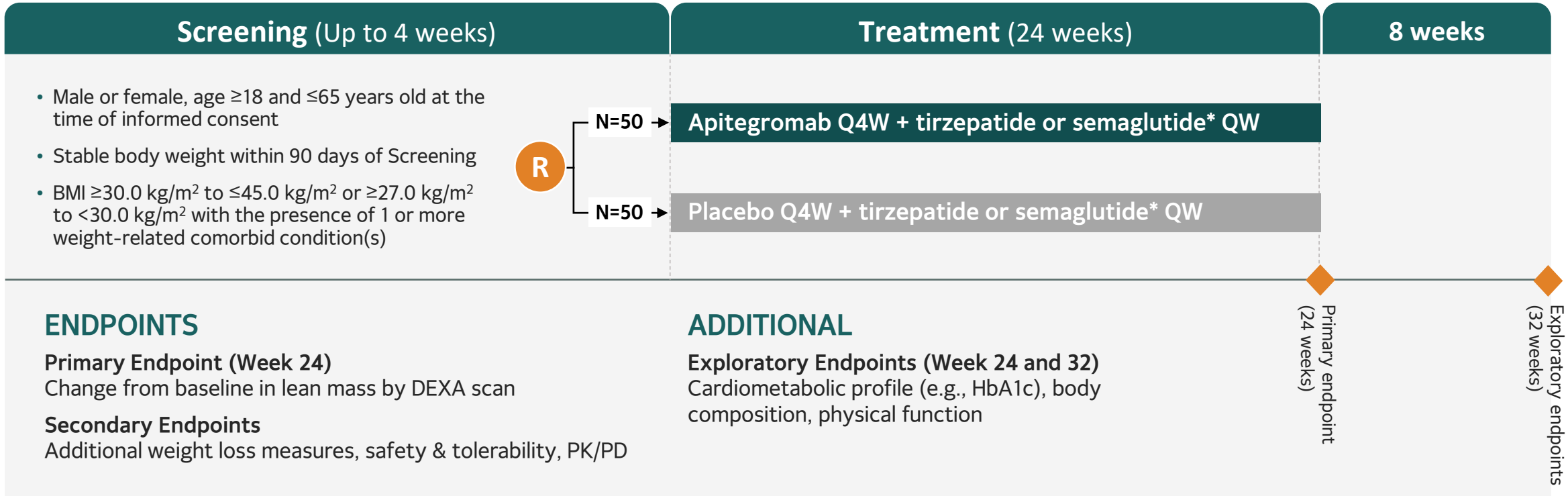
- Effect of apitegromab to preserve lean mass in obese or overweight patients receiving a GLP-1 agonist
- Safety and tolerability data to provide initial support for long-term chronic use
- Explore the potential effect of apitegromab to improve metabolic profile and physical function

Insights gained from EMBRAZE study to inform SRK-439 development

Launching 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 expected to start in 2Q 2024



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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Regulatory Pathway

FDA Guidelines

A clinical outcome assessment is a measure that describes or reflects how a patient feels, functions, or survives.*

- FDA guidance supports combination strategy
- Need to demonstrate the added clinical benefit of the combination

Added Clinical Benefit

Incremental Weight Loss

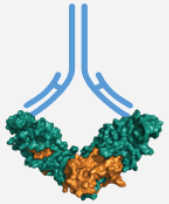
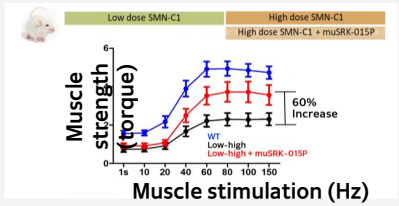
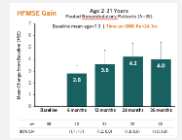
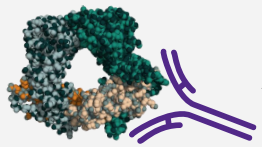
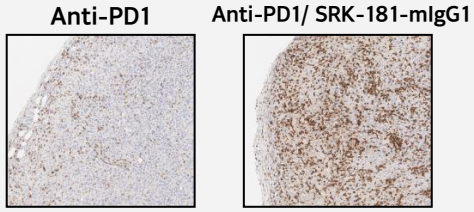
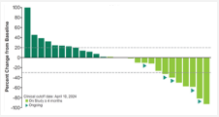
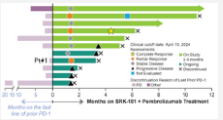
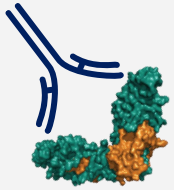
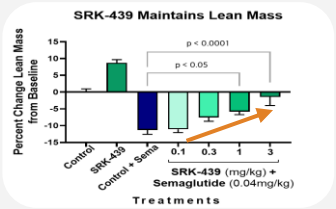
- Current weight management drugs approved based on total body weight loss
- Incremental weight loss as primary endpoint – preservation of lean mass may lead to additional weight loss incremental to that mediated by GLP-1 agonist

Incremental Clinical Benefit

- Increased muscle mass has the potential to improve metabolic profile (e.g., HbA1c)
- Preserving lean body mass is expected to improve physical function

*Clinical Outcome Assessment: Frequently Asked Questions: https://www.fda.gov/about-fda/clinical-outcome-assessment-coa-frequently-asked-questions#Consideration1

Platform and Expertise Drive Success in Clinic

Differentiated Engineering	Translational Models	Clinic
 Apitegromab		TOPAZ  SAPPHIRE ONYX
 SRK-181		DRAGON  
 SRK-439		FDA IND in 2025

Cardiometabolic Program Has Key Ingredients For Success

Leverages Apitegromab Study

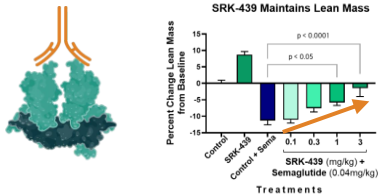
First and only anti-myostatin to show clinical proof of concept, apitegromab's study in obesity informs SRK-439 development

EMBRAZE

SRK-439 Designed for Obesity

Specificity minimizes off-target effects; effect in translational models on preservation of lean mass, enhanced fat loss, & improved metabolic profile

SRK-439



Regulatory Path Forward

FDA guidance supports combination strategy; SRK-439 development has the potential to improve function & incremental weight loss

FDA

IND in 2025

Closing Remarks

Jay Backstrom
President & Chief Executive Officer

IN SUMMARY

Advancing Our Journey Towards Commercialization

1

Selectivity is the Key

The hallmark of our differentiated platform is unparalleled selectivity

2

Large Unmet Needs

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

3

Positioned for Success

Experienced team executing on strategy and goals

Next 12 – 24 months of execution is expected to be transformative for our company