Second Quarter 2024 Business Update

August 8, 2024

ScholarRock

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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, 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 Form 10-K for 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 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, SRK-373, SRK-256, 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, SRK-373, SRK-256, and SRK-439 have not been established.

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Introduction & Business Update

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

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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	

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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

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Our Approach

Selectivity Drives Success

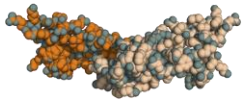
RIGHT TARGET → **Validated Biology**

RIGHT TIME → **Latent Form**

Deep structural insights to validated targets

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

Optimized for efficacy and mitigates off-target effects



Traditional Target
"mature" active growth factor

Scholar Rock's Target
Latent Growth Factor Complex

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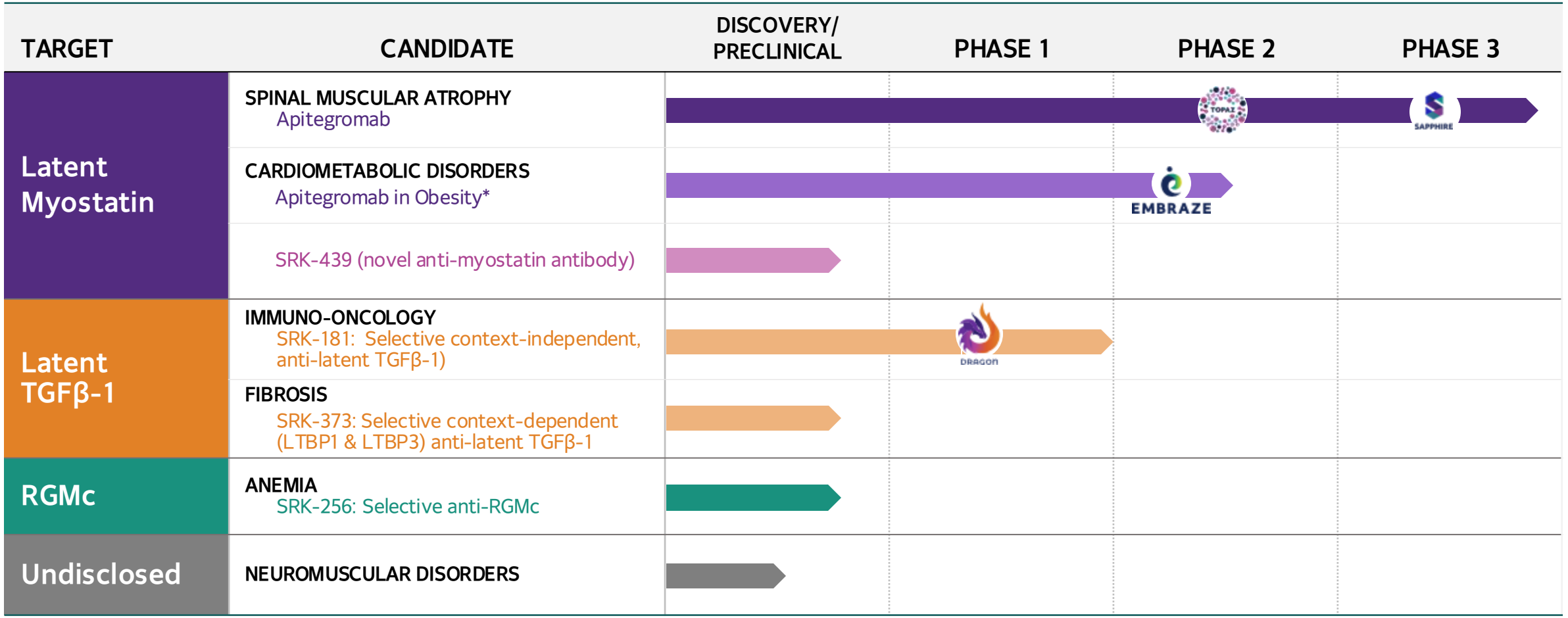
Scientific Platform Yielding Growing Pipeline Across High Value Therapeutic Areas



*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 protein3; 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

Upcoming SAPPHIRE readout - Q4 2024

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

Exploring additional neuromuscular populations

Cardiometabolic Disorders

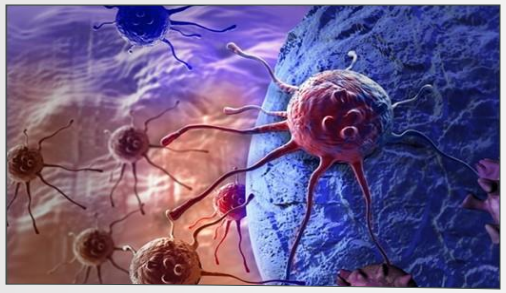
Ph 2 POC EMBRAZE study enrolling: Topline data expected in Q2 2025

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

Promising objective response rates, safety, and novel biomarker data highlighted in oral presentation at ASCO

ASCO

Journal of Clinical Oncology
An American Society of Clinical Oncology Journal

SRK-439

Compelling new preclinical data highlighted in oral presentation at ADA

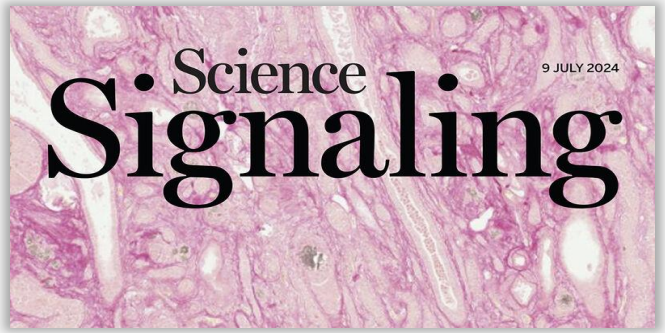
American Diabetes Association®

SRK-373

Featured on the cover of **Science Signaling** and in *Focus* article:

"More Velcro for the TGFB-1 Straightjacket"

by Boris Hinz



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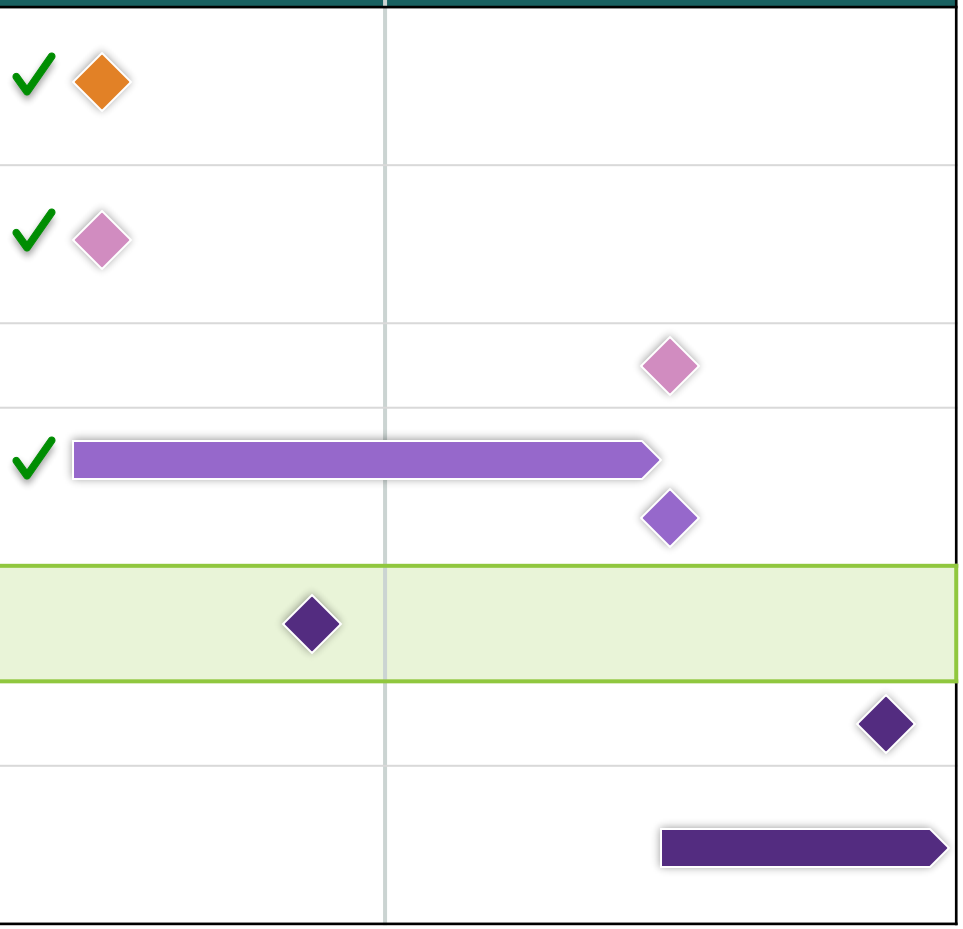
2024 & 2025 Anticipated Milestones

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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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] Pooled nonambulatory patients

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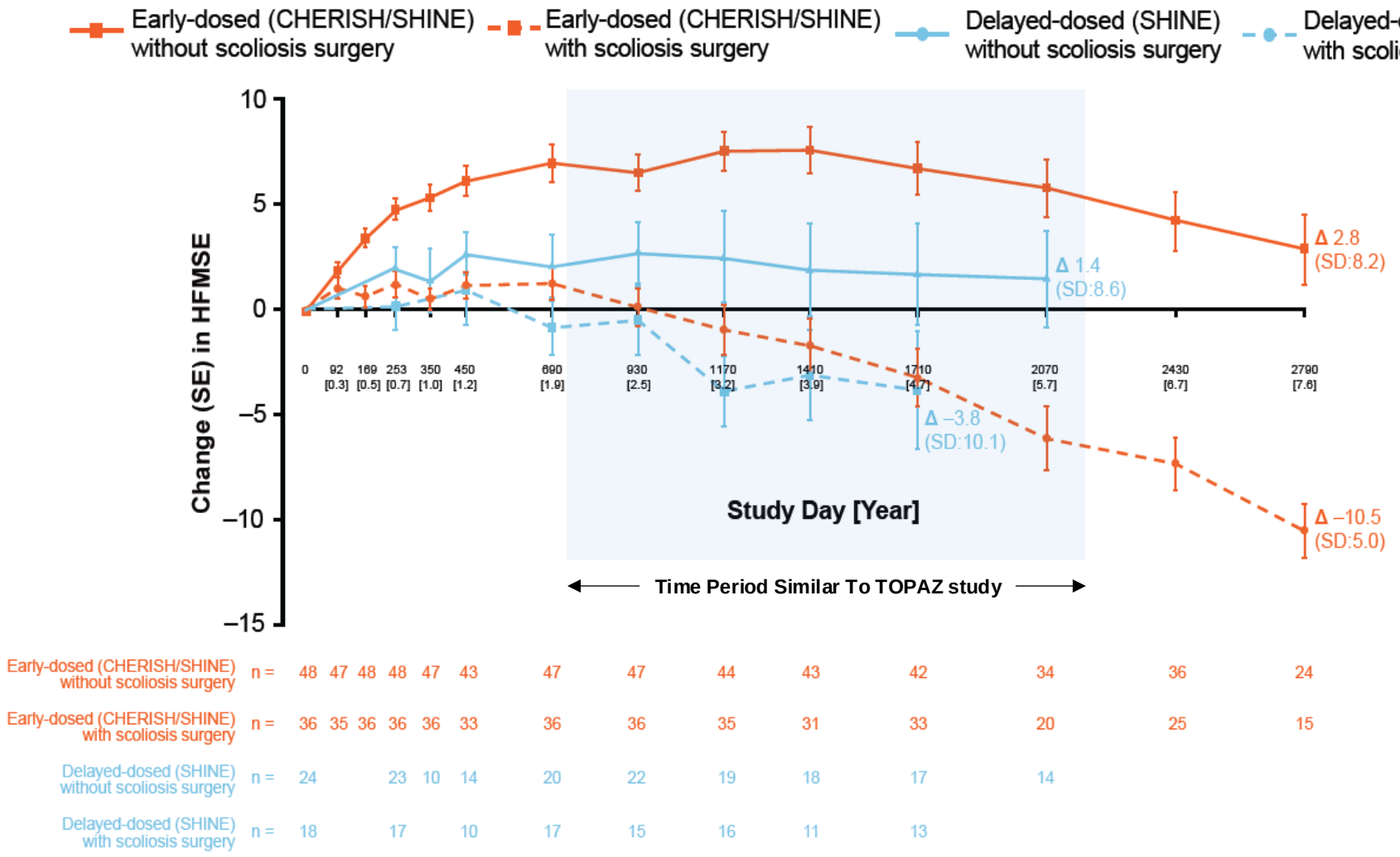
Motor function slowly declines despite following initial increase

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



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

- Decline in motor function is influenced by scoliosis surgery and contractures (Wolfe 2024; Dunaway 2020; Salazar 2018)
- TOPAZ participants had ~2 years of nusinersen treatment at baseline
- Blue-shaded area represents similar duration of 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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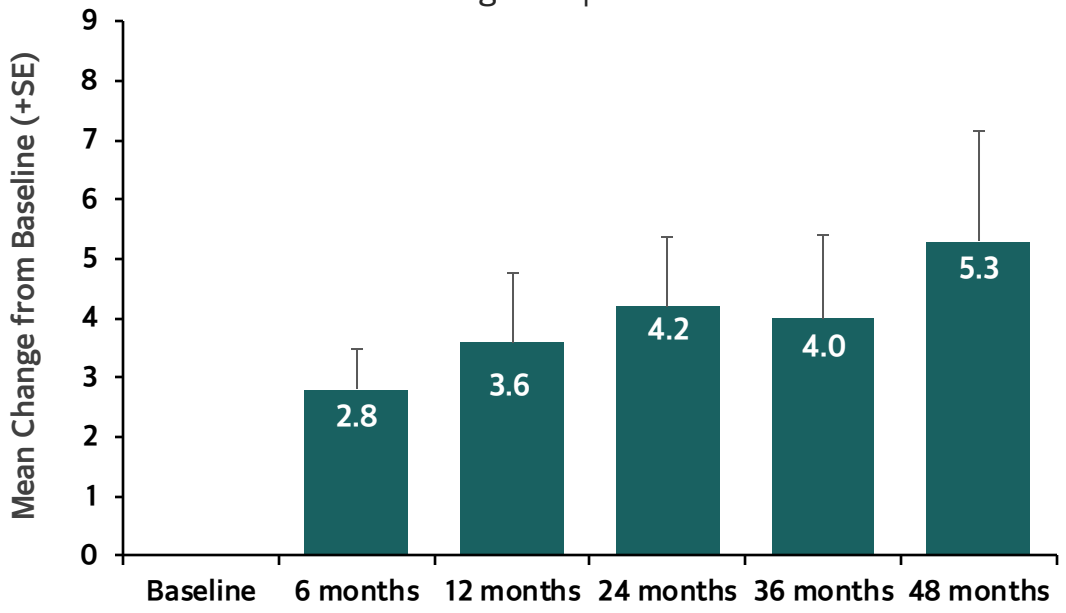
Apitegromab TOPAZ Clinical Trial: Motor Function Outcomes by HFMSE Over 48 Months

Improvements Were Substantial and Sustained

Pooled Nonambulatory Patients[1]

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

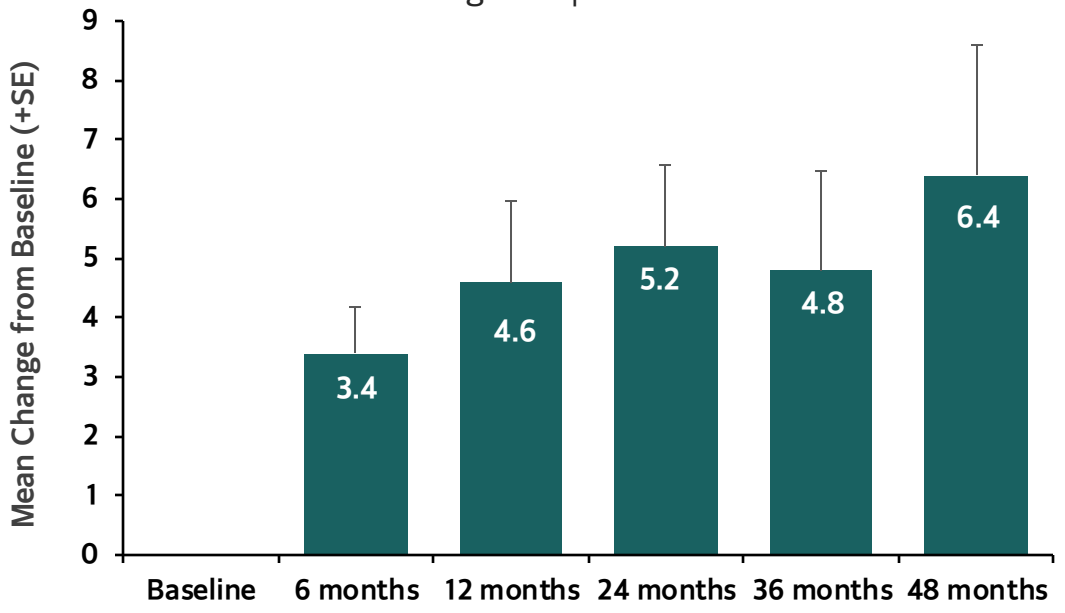
Baseline mean age=7.3 | **Time on SMN Rx=24.1m**



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**



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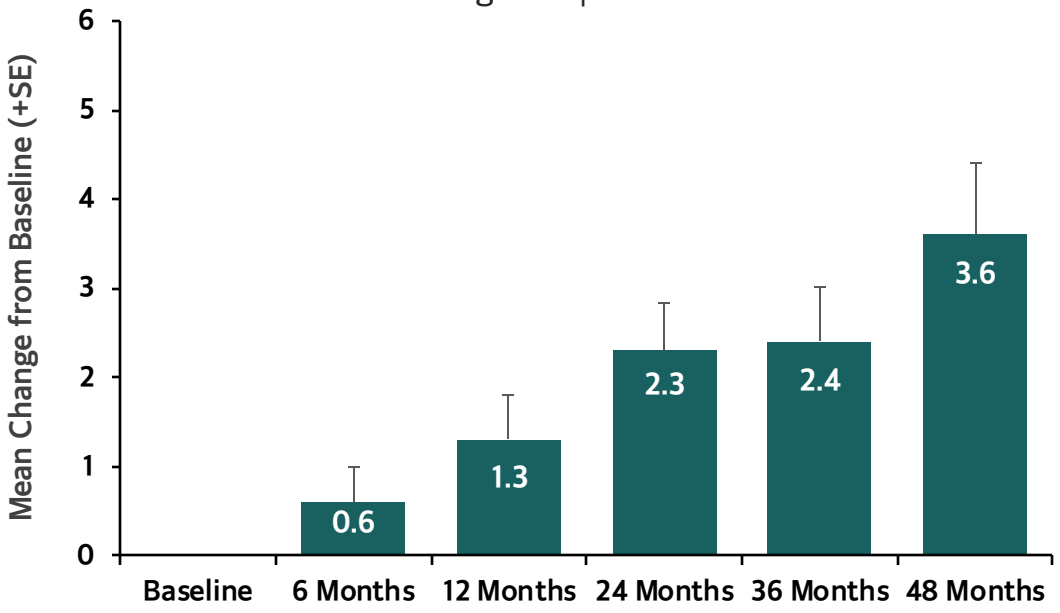
Apitegromab TOPAZ Clinical Trial: Motor Function Outcomes by RULM Over 48 Months

Improvements Were Substantial and Sustained

Pooled Nonambulatory Patients[1]

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

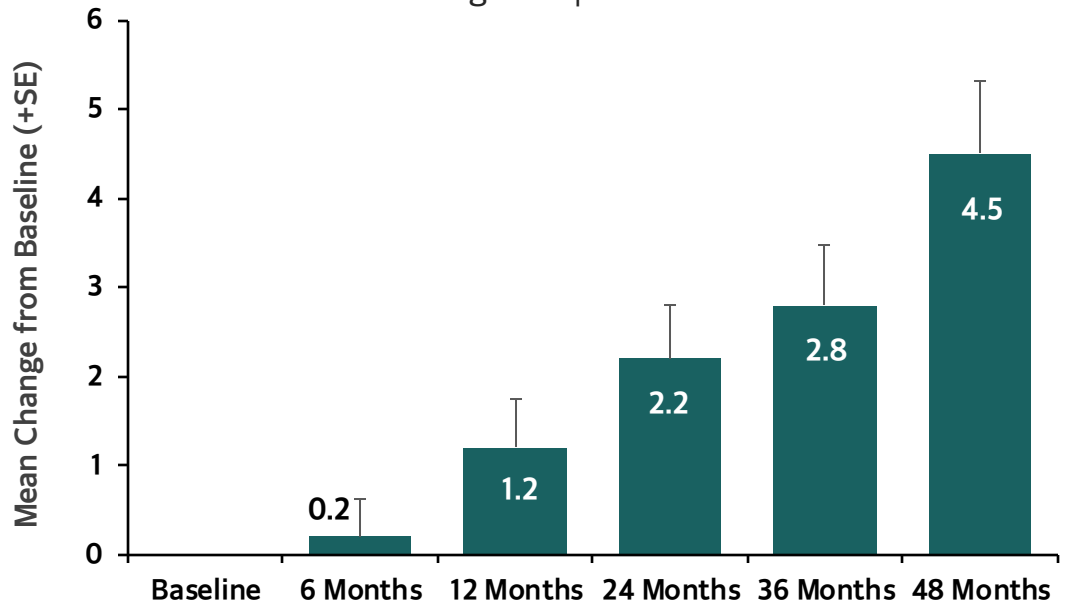
Baseline mean age=7.3 | Time on SMN Rx=24.1m



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



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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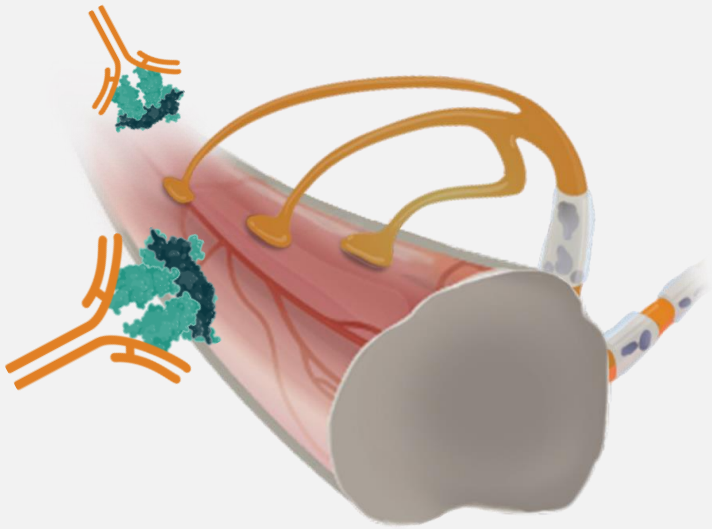
Summary of TOPAZ Data

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 profile 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

SCREENING

MAIN POPULATION (n=156)

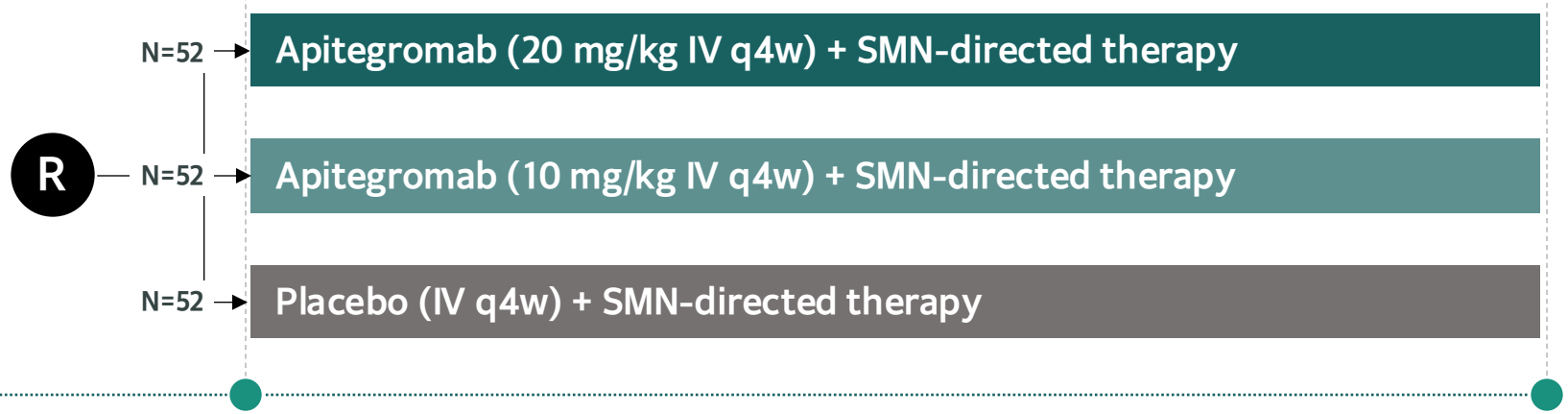
Ages 2-12

With nonambulatory Types 2 and 3 SMA

Stratified to ensure balanced allocation across the three arms:

1. Age at SMN therapy initiation (age < 5 vs. age ≥ 5)
2. SMN therapy (nusinersen vs. risdiplam)

TREATMENT (52 weeks)



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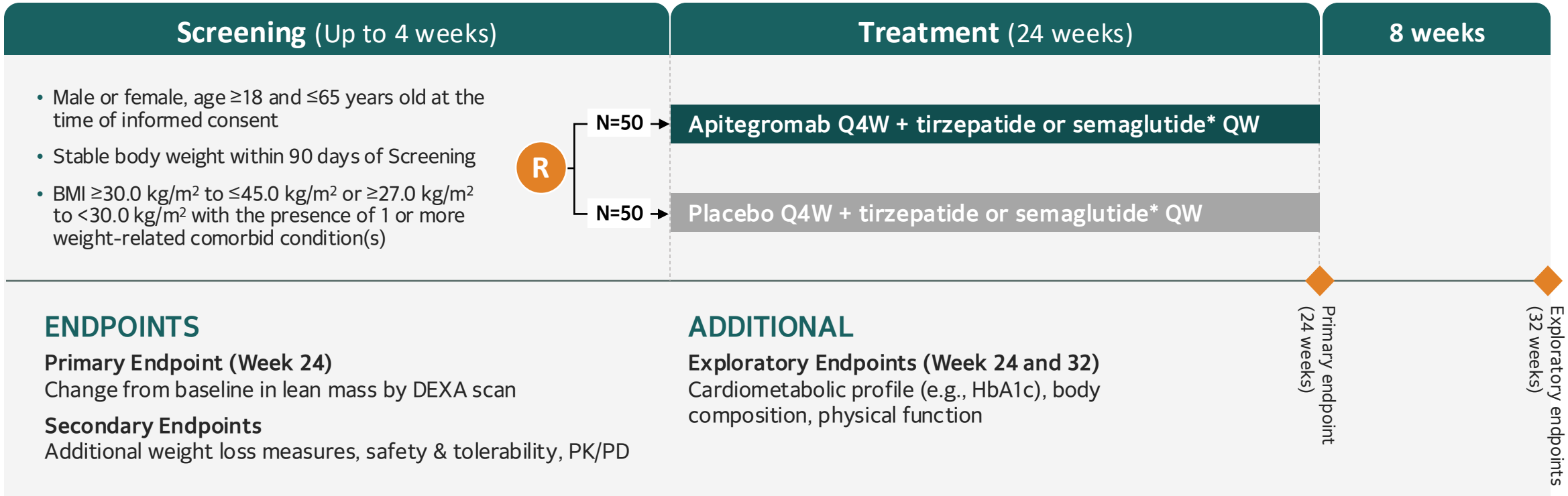
Enrolling Phase 2 Proof-of-Concept Study of Apitegromab in Obesity

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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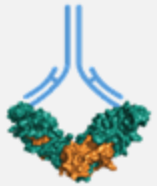
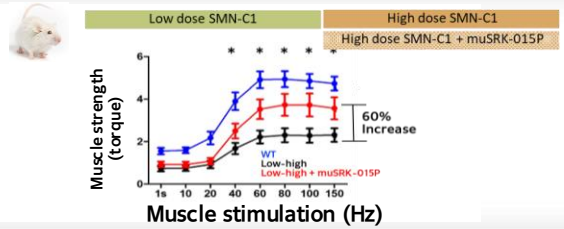
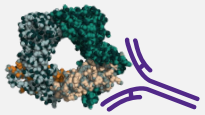
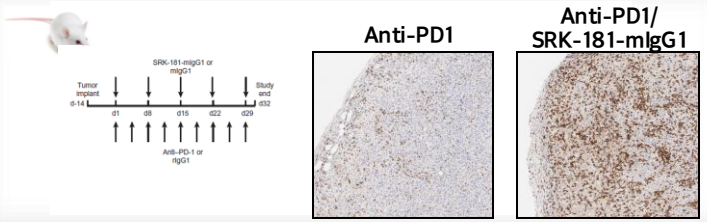
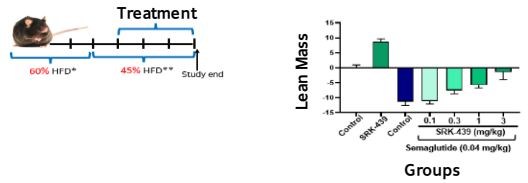
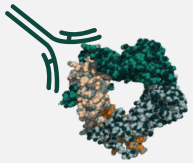
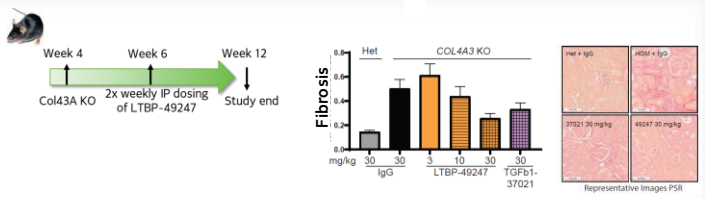
Platform and Expertise Drive Success in Clinic

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

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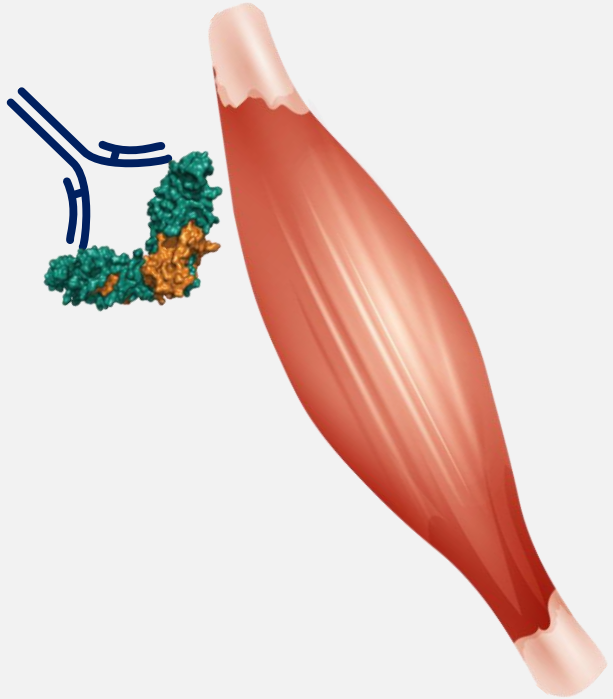
Strong Scientific Validation and Promising Preclinical Evidence

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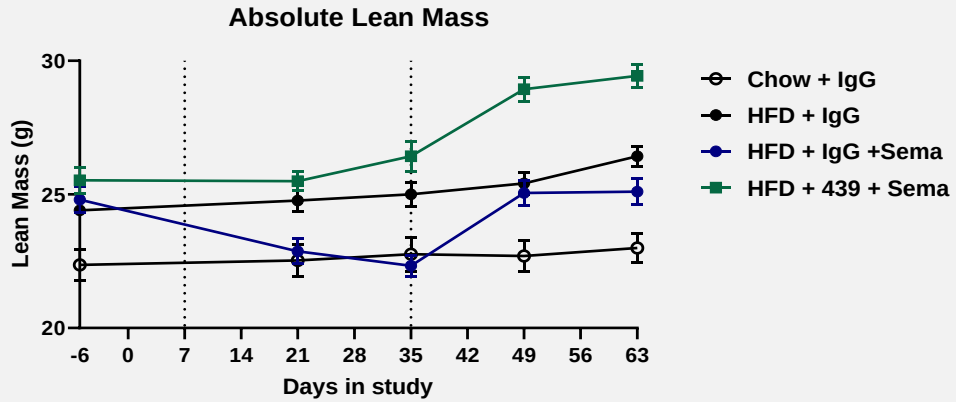
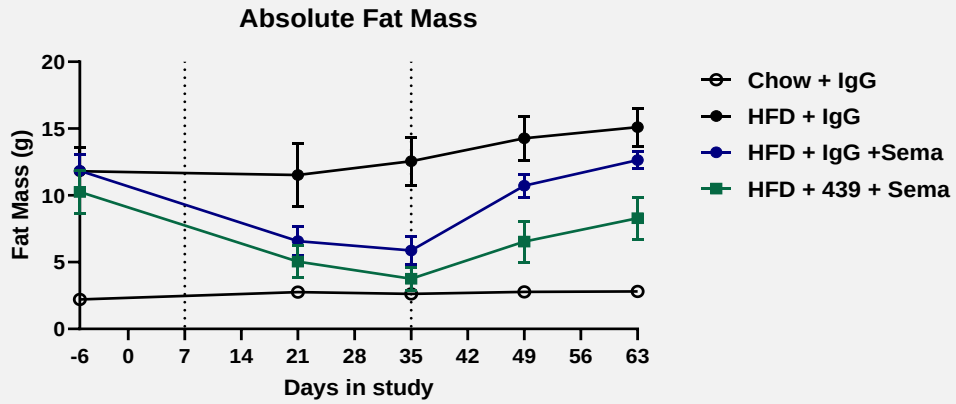
SRK-439 Increased Lean Mass and Attenuated Regain of Fat Mass After GLP-1 RA Withdrawal in Obesity Mouse Model

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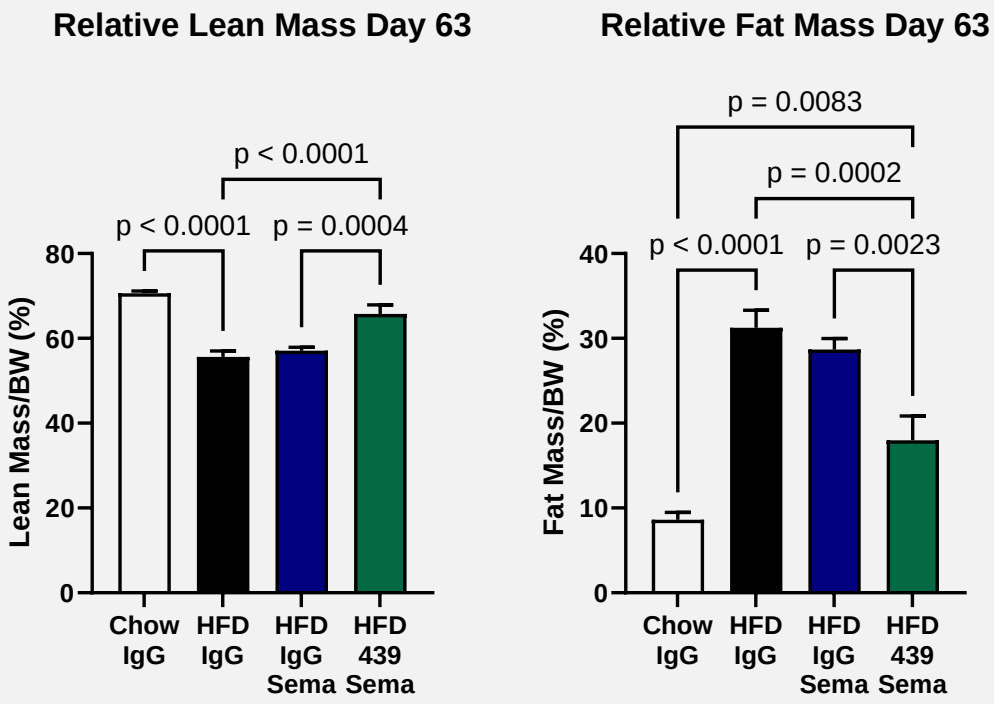
SRK-439 Improved Body Composition After GLP 1-RA Withdrawal

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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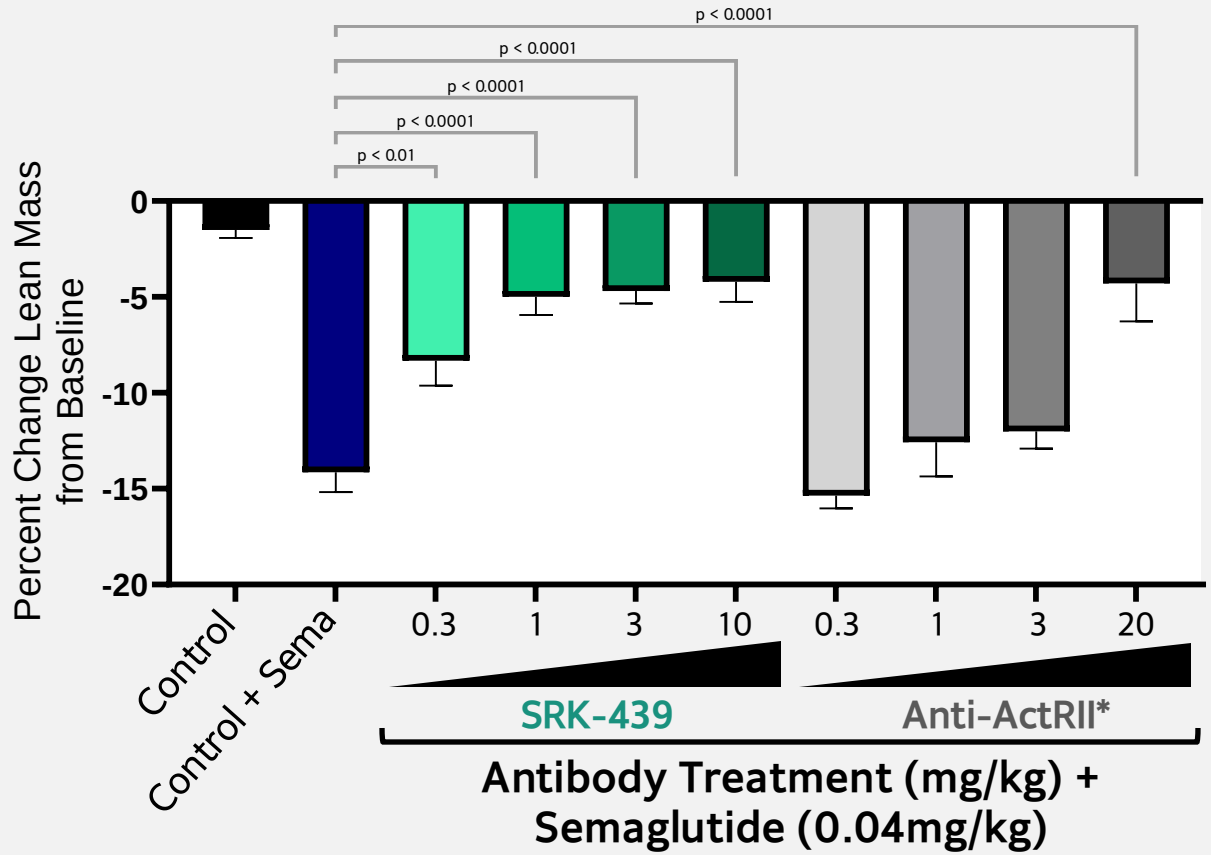
LOW EFFICACIOUS DOSE AND COMPETITIVE PROFILE

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 doses than anti-ActRII
- Highlights efficacy of SRK-439 and avoids potential liabilities of non-selective approach of 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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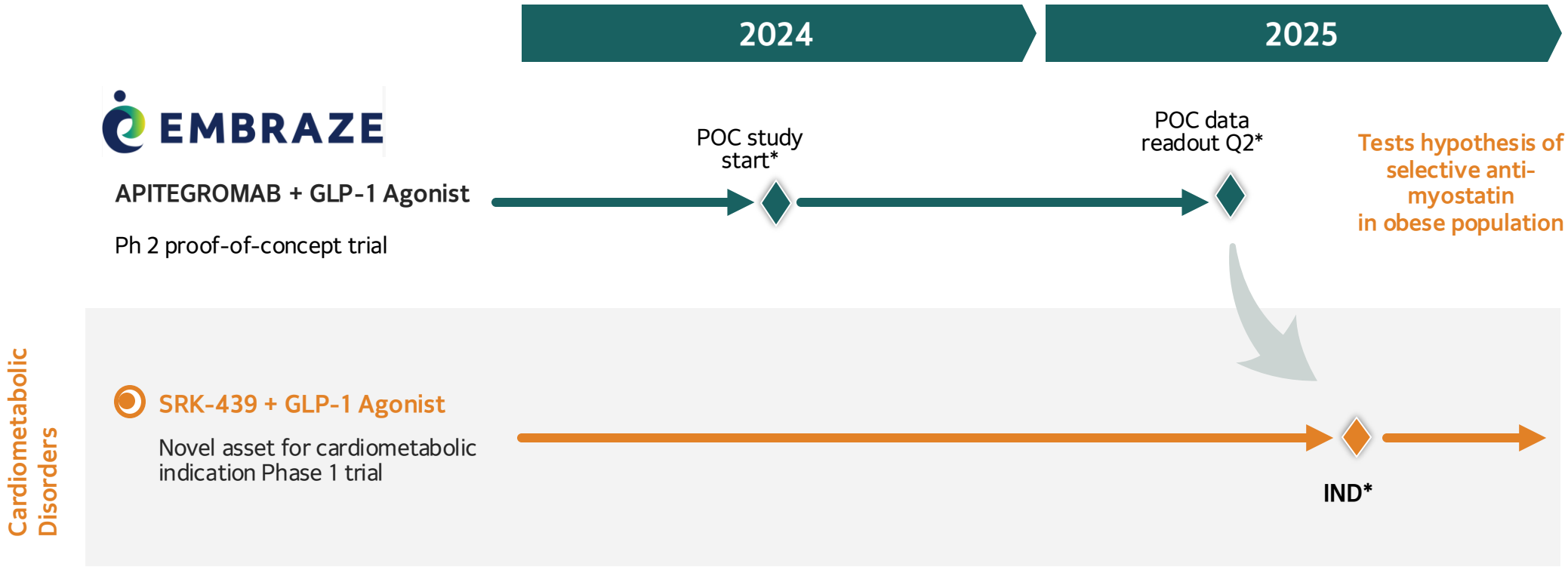
Leveraging Apitegromab to Inform Obesity Program



*Expected timelines
POC = Proof of Concept

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Upcoming Milestones

Jay Backstrom
Chief Executive Officer

ScholarRock

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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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