Scholar Rock to Present New Data from SRK-181 Phase 1 DRAGON Trial at the American Society of Clinical Oncology (ASCO) Annual Meeting

May 28, 2024

- Updated safety and efficacy results on SRK-181 will be featured in an oral presentation on Monday, June 3rd, at 1:50 p.m CDT/2:50 p.m. EDT
- Company will host conference call to discuss the data on Tuesday, June 4th at 7:00 a.m. CDT/8:00 a.m. EDT

CAMBRIDGE, Mass.--(BUSINESS WIRE)--May 28, 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 announced it will present new data from its Phase 1 DRAGON proof-of-concept trial of SRK-181 in combination with pembrolizumab in patients with advanced solid tumors in an oral presentation during the American Society of Clinical Oncology (ASCO) Annual Meeting being held May 31 – June 4 in Chicago. Those data will be discussed further during a webcast on June 4th at 7 a.m. CDT/8 a.m. EDT that includes Jing Marantz, M.D., Ph.D., Chief Medical Officer at Scholar Rock, and Dr. Toni Choueiri, M.D., Director of the Lank Center for Genitourinary (GU) Oncology at Dana-Farber Cancer Institute (DFCI).

"We continued to observe encouraging responses and tolerability in heavily pretreated patients across multiple cancer types, providing further evidence of SRK-181's potential to overcome resistance to immune checkpoint inhibitors," said Jay Backstrom, M.D., M.P.H., President and Chief Executive Officer of Scholar Rock. "In addition, biomarker data indicated that treatment with SRK-181 is associated with an enhanced proinflammatory microenvironment and increased anti-tumor activity. Together, these new data provide further validation of the selective approach of our TGFβ platform. We look forward to sharing additional updated data at ASCO."

Data highlights are as follows:

- SRK-181 continued to be well tolerated in the dose expansion portion of the DRAGON trial (Part B).

- Encouraging responses were observed across multiple cancer types, including clear cell renal cell carcinoma (ccRCC), head and neck squamous cell carcinoma (HNSCC), melanoma (MEL), and urothelial carcinoma (UC).

- Biomarker data showed that the SRK-181 + pembrolizumab treatment combination created an enhanced proinflammatory microenvironment in anti-PD-(L)1 resistant patients.

- Patients with ccRCC whose tumors were infiltrated at baseline by CD8+ T cells and/or regulatory T cells showed positive correlations between infiltration of each cell type and response rate. This positive correlation between baseline infiltration status type and response rate suggests a potential patient selection strategy.

These results will be presented at the ASCO Annual Meeting in an oral presentation, details of which can be found below.

Title: Phase 1 study (DRAGON) of SRK-181 (linavonkibart), a latent TGFβ1 inhibitor, combined with pembrolizumab in anti-PD1 resistant patients with advanced solid tumors: Updated results of expansion part
Oral Session: Developmental Therapeutics—Immunotherapy
Presenter: Ulka N. Vaishampayan, MD, Division of Hematology/Oncology, University of Michigan
Location: Hall D2
Date/Time: June 3, 1:50 p.m. CDT

Conference Call Information
Scholar Rock will host a conference call on June 4 at 8 a.m. EDT that can be accessed by registering in advance at the Events and Presentations page of Scholar Rock's website. Members of Scholar Rock's executive management team will be joined by Dr. Toni Choueiri, M.D., Director of the Lank Center for Genitourinary (GU) Oncology at Dana-Farber Cancer Institute (DFCI).

The abstracts for these presentations are available on ASCO's website https://conferences.asco.org/am/abstracts. The presentations will be made available in the Publications & Posters section of Scholar Rock's website following the conference.

For conference information, visit https://conferences.asco.org/.

About SRK-181
SRK-181 is a selective inhibitor of TGFβ1 activation being developed to overcome primary resistance to checkpoint inhibitor therapy, such as anti-PD-(L)1 antibodies, in advanced cancer. TGFβ1 is the predominant TGFβ isoform expressed in many human tumor types. Based on analyses of various human tumors that are resistant to anti-PD-(L)1 therapy, data suggest that TGFβ1 is a key contributor to the immunosuppressive tumor microenvironment, excluding and preventing entry of cytotoxic T cells into the tumor, thereby inhibiting anti-tumor immunity. (2) SRK-181 specifically targets the latent TGFβ1 isoform in a context-independent manner, designed to enable complete inhibition of TGFβ1 in all compartments within the tumor microenvironment. Scholar Rock believes that SRK-181 has the potential to overcome this immune cell exclusion and induce tumor regression when administered in combination with anti-PD-(L)1 therapy while potentially avoiding toxicities associated with non-selective TGFβ inhibition. Enrollment of the DRAGON Phase 1 proof-of-concept clinical trial (NCT04291079) was completed in December 2023, and patients who remain on the study continue to be treated. The trial enrolled patients in multiple proof of concept cohorts conducted in parallel, including urothelial carcinoma (UC), cutaneous melanoma (MEL), non-small cell lung cancer (NSCLC), head and neck squamous cell carcinoma (HNSCC), and clear cell renal cell

carcinoma (ccRCC). SRK-181 is an investigational product candidate and its efficacy and safety have not been established. SRK-181 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.

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 SRK-181, 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, 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 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; 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; 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 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.

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Source: Scholar Rock