Scholar Rock to Present Trials in Progress Poster for SRK-181 DRAGON Phase 1 Clinical Trial at the American Society of Clinical Oncology Annual Meeting

May 19, 2021

CAMBRIDGE, Mass.--(BUSINESS WIRE)--May 19, 2021-- Scholar Rock (NASDAQ: SRRK), a clinical-stage biopharmaceutical company focused on the treatment of serious diseases in which protein growth factors play a fundamental role, today announced a trials in progress poster presentation at the 2021 American Society of Clinical Oncology (ASCO) Annual Meeting, being held virtually from June 4-8, 2021.

The DRAGON Phase 1 proof-of-concept trial (NCT04291079) is evaluating SRK-181, a selective inhibitor of TGFβ1 activation, in patients with locally advanced or metastatic solid tumors that have shown primary resistance to checkpoint inhibitor therapies. Dose escalation is progressing in Part A of the trial and the Company expects to advance to the Part B dose expansion portion of the trial in mid-2021.

The trials in progress e-poster will provide an overview of the design of the DRAGON Phase 1 trial and highlight the biomarker analysis being implemented to support the trial. Details of the poster presentation are as follows:

- **Session:** Developmental Therapeutics--Molecularly Targeted Agents and Tumor Biology
- **Title:** “First-in-Human Phase 1 Trial (DRAGON) of SRK-181: A Potential First-in-Class Selective Latent TGFβ1 Inhibitor, Alone or in Combination with Anti-PD-(L)1 Treatment in Patients with Advanced Solid Tumors” (TPS3146)
- **Date and Time**: Friday, June 4, 11:00 AM - 2:00 PM EDT; Poster sessions available on demand in meeting library beginning 9:00 AM EDT on June 4, 2021.

About SRK-181

SRK-181 is a selective inhibitor of TGFβ1 activation and is an investigational product candidate being developed to overcome primary resistance to checkpoint inhibitor therapy, such as anti-PD-(L)1 antibodies. 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 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 [(1)]. Scholar Rock believes SRK-181, which specifically targets the latent TGFβ1 isoform, 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. The DRAGON Phase 1 proof-of-concept clinical trial (NCT04291079) in patients with locally advanced or metastatic solid tumors is ongoing. The efficacy and safety of SRK-181 have not been established. SRK-181 has not been approved for any use by the FDA nor any other regulatory agency.

(1) Martin et al., Sci. Transl. Med. 12: 25 March 2020

About Scholar Rock

Scholar Rock is a clinical-stage biopharmaceutical company focused on the discovery and development of innovative medicines for the treatment of serious diseases in which signaling by protein growth factors plays a fundamental role. Scholar Rock is creating a pipeline of novel product candidates with the potential to transform the lives of patients suffering from a wide range of serious diseases, including neuromuscular disorders, cancer, fibrosis and anemia. Scholar Rock’s approach to targeting the molecular mechanisms of growth factor activation enabled it to develop a proprietary platform for the discovery and development of monoclonal antibodies that locally and selectively target these signaling proteins at the cellular level. By developing product candidates that act in the disease microenvironment, the Company intends to avoid the historical challenges associated with inhibiting growth factors for therapeutic effect. Scholar Rock believes its focus on biologically validated growth factors may facilitate a more efficient development path. For more information, please visit https://scholarrock.com/ or follow Scholar Rock on Twitter (@ScholarRock) and LinkedIn (https://www.linkedin.com/company/scholar-rock/).

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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 expectations regarding its growth, strategy, progress and timing of its clinical trials and the ability of any product candidate to perform in humans in a manner consistent with earlier nonclinical, preclinical or clinical trial data. The use of words such as “may,” “might,” “will,” “should,” “expect,” “plan,” “anticipate,” “believe,” “estimate,” “project,” “intend,” “future,” “potential,” or “continue,” and other similar expressions are intended to identify such forward-looking statements. 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 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 and the impacts of public health pandemics such as COVID-19 on business operations and expectations, 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, 2021, 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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