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Kezar Life Sciences Announces Completion of Enrollment of Its Phase 2 PRESIDIO Clinical Trial of KZR-616 in Polymyositis and Dermatomyositis

SOUTH SAN FRANCISCO, Calif.--(BUSINESS WIRE)-- Kezar Life Sciences, Inc. (Nasdaq: KZR), a clinical-stage biotechnology company discovering and developing breakthrough treatments for immune-mediated and oncologic disorders, today announced that PRESIDIO, its Phase 2 clinical trial to evaluate the safety and efficacy of KZR-616 for the treatment of polymyositis (PM) and dermatomyositis (DM) completed the target enrollment of 24 subjects.

PRESIDIO is a Phase 2 randomized, placebo-controlled, crossover design study of KZR-616 in patients with PM or DM. During the 32-week treatment period, patients receive either 45 mg of KZR-616 or placebo subcutaneously once weekly for 16 weeks followed by a crossover to the other treatment arm for an additional 16 weeks (the first two doses are at 30 mg). The primary endpoint of PRESIDIO is the mean change in the Total Improvement Score. Top line results from this trial are expected in the second quarter of 2022. Subjects who complete the 32 week study have the option to enroll into an open-label extension study and continue to receive 45mg weekly of KZR-616.

“Completing enrollment in PRESIDIO marks a significant milestone on our path to the full development of KZR-616 for the treatment of polymyositis and dermatomyositis. We are grateful to the patients, their families and caregivers, investigators and clinical research team who are participating in the trial. It goes without saying that their unwavering support of the trial during the course of the global pandemic was heroic,” said Noreen Roth Henig, MD, Chief Medical Officer of Kezar.

“Idiopathic inflammatory myopathies such as PM and DM are chronic and progressively debilitating inflammatory diseases that can have a significant impact on the quality of life of patients living with these rare diseases,” said Chrissy Thornton, Executive Director of the patient advocacy organization, The Myositis Association. “The completed enrollment of the Phase 2 PRESIDIO study is cause for celebration as it represents a significant step in bringing forward novel therapies for these diseases, where options to date have been limited.”

PM and DM are two of the five types of autoimmune myositis diseases. Both are chronic, debilitating, autoimmune diseases that are distinguished by inflammation of the muscles as well as the skin (in DM). An approximate 30,000-120,000 people in the United States are living with these severe and progressive inflammatory myopathies that are characterized by marked morbidity and associated mortality. While debilitating muscle weakness is the hallmark of these myopathies, including compromised muscles of respiration, other internal organ system dysfunctions can be equally disabling. The aim of treatment for these diseases

is to suppress inflammation, increase muscle strength and prevent long-term damage to muscles and extramuscular organs; however, treatment options are limited for DM, and there are currently no approved treatments for PM.

The U.S. Food and Drug Administration has granted Orphan Drug Designations for KZR-616 for the treatment of polymyositis and dermatomyositis.

More details about the PRESIDIO study [NCT04033926] and the PRESIDIO open-label extension study [NCT04628936] can be found on https://clinicaltrials.gov.

About KZR-616

KZR-616 is a novel, first-in-class, selective immunoproteasome inhibitor with broad therapeutic potential across multiple autoimmune diseases. Preclinical research demonstrates that selective immunoproteasome inhibition results in a broad anti-inflammatory response in animal models of several autoimmune diseases, while avoiding immunosuppression. Data generated from Phase 1a and 1b clinical trials provide evidence that KZR-616 exhibits a favorable safety and tolerability profile for development in severe, chronic autoimmune diseases. Phase 2 clinical trials are underway in severe autoimmune diseases.

About Kezar Life Sciences

Kezar Life Sciences is a clinical-stage biopharmaceutical company discovering and developing breakthrough treatments for immune-mediated and oncologic disorders. The company is pioneering first-in-class, small-molecule therapies that harness master regulators of cellular function to inhibit multiple drivers of disease via single, powerful targets. KZR-616, its lead development candidate, is a selective immunoproteasome inhibitor being evaluated in Phase 2 clinical trials in lupus nephritis, dermatomyositis and polymyositis. Additionally, KZR-261, is the first anti-cancer clinical candidate from the company's platform targeting the Sec61 translocon and the protein secretion pathway. An IND submission for KZR-261 in solid tumors was filed in August 2021, and Kezar plans to initiate an open-label dose-escalation Phase 1 clinical trial of KZR-261 to assess safety, tolerability and preliminary tumor activity in solid tumors. For more information, visit www.kezarlifesciences.com.

Cautionary Note on Forward-looking Statements

This press release contains forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995. Words such as "may," "will," "should," "expect," "believe" and similar expressions (as well as other words or expressions referencing future events, conditions or circumstances) are intended to identify forward-looking statements. These forward-looking statements are based on Kezar's expectations and assumptions as of the date of this press release. Each of these forward-looking statements involves risks and uncertainties that could cause Kezar's clinical development programs, future results or performance to differ materially from those expressed or implied by the forward-looking statements. Forward-looking statements contained in this press release include, but are not limited to, statements about the design, progress, timing, scope and results of clinical trials, the anticipated timing of disclosure of results of clinical trials and the anticipated regulatory development of Kezar's product candidates. Orphan Drug Designation does not provide any

assurance of regulatory approval or expedite regulatory review. Many factors may cause differences between current expectations and actual results, including the impacts of the COVID-19 pandemic on the company's business, clinical trials and financial position, unexpected safety or efficacy data observed during preclinical or clinical studies, clinical trial site activation or enrollment rates that are lower than expected, changes in expected or existing competition, changes in the regulatory environment, the uncertainties and timing of the regulatory approval process, and unexpected litigation or other disputes. Other factors that may cause actual results to differ from those expressed or implied in the forward-looking statements in this press release are discussed in Kezar's filings with the U.S. Securities and Exchange Commission, including the "Risk Factors" contained therein. Except as required by law, Kezar assumes no obligation to update any forward-looking statements contained herein to reflect any change in expectations, even as new information becomes available.

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