November 15, 2021

Kezar Life Sciences Announces Interim Results from the MISSION Phase 2 Trial in Patients with Lupus Nephritis

- *KZR-616 demonstrates clinically meaningful benefit in patients with lupus nephritis, with 4 out of 5 patients achieving either a partial or complete renal response at end of treatment*
- *KZR-616 maintained a favorable safety and tolerability profile over the six-month treatment period*
- *Company-hosted investor and analyst conference call and webcast with guest investigator to be held today at 4:30pm ET*

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 reported interim results from the Phase 2 portion of its MISSION clinical trial evaluating KZR-616, a first-in-class selective immunoproteasome inhibitor, in patients with active, proliferative lupus nephritis (LN).

“The MISSION Phase 2 interim results present a strong signal that KZR-616 is active and could be a meaningful therapy for patients with lupus nephritis, a long term and difficult to treat disease,” said Noreen R. Henig, M.D., Kezar’s Chief Medical Officer. “Reduction in proteinuria, as quickly as possible, is an important therapeutic goal for patients with lupus nephritis, and we observed meaningful reductions at 6 months as well as encouraging data at 3 months. KZR-616 continues to appear to be immunomodulatory rather than immunosuppressive, which we believe could offer advantages over current treatments available. Based on these interim findings, we look forward to reporting top-line data in the second quarter of 2022.”

Samir V. Parikh, M.D., Associate Professor of Medicine, Division of Nephrology, The Ohio State University Wexner Medical Center and an investigator in the MISSION study, added, “these interim results are important for patients living with lupus nephritis. One of the devastating consequences of the disease is kidney failure, so new immunomodulatory treatments that have the potential to protect kidney function would fulfill a substantial unmet need and could lead to better long-term outcomes.”

The MISSION Phase 2 clinical trial is an open-label study to demonstrate the responder rate of KZR-616 in patients with active lupus nephritis. During the 24 week treatment period, patients received 60 mg of KZR-616 subcutaneously once weekly (first dose of 30 mg) in addition to their background therapy. Patients in the MISSION Phase 2 trial do not receive KZR-616 as part of “induction” therapy, which represents a significant difference in comparison to other recently published trials in lupus nephritis. End of treatment assessments occurred at Week 25.

For the interim analysis, five patients had reached end of treatment, and ten patients had reached week 13 of treatment. The primary efficacy endpoint for the trial is the proportion of patients achieving a renal response measured by a 50% or greater reduction in urine protein to creatinine ratio (UPCR) at end of treatment. The secondary efficacy endpoint for the trial is the number of patients with a complete renal response (CRR) and partial renal response (PRR).

Key findings from the interim analysis of the MISSION Phase 2 are summarized below:

- Clinically meaningful renal response was observed at end of treatment.
    - 3 of 5 patients achieved a 50% or greater reduction in UPCR at week 25 compared to baseline, the primary efficacy endpoint of the clinical trial.
    - 4 of the 5 patients who completed treatment at week 25 with KZR-616 demonstrated clinically meaningful reduction in proteinuria to less than 0.8 UPCR:
        - 2 patients showed a CRR and had a reduction of absolute proteinuria values to equal to or less than 0.5 UPCR.
        - 2 patients showed a PRR and had a reduction of absolute proteinuria values to between 0.5 and 0.8 UPCR.

- Clinically meaningful reductions in UPCR were also observed in 5 of 10 patients at week 13 of KZR-616 and included improvements in key disease biomarkers.

- KZR-616 was well tolerated over the six-month treatment period.
    - No new safety signals were observed in the Phase 2 portion of the MISSION trial.
    - Adverse events were generally mild-to-moderate (Grade 1 or 2).
    - There were no study discontinuations due to drug related adverse events. There was one temporary interruption of study drug due to a Grade 3 serious adverse event, the occurrence of a migraine, and one discontinuation unrelated to the study drug.

Top-line data from the Phase 2 MISSION trial in patients with lupus nephritis are expected in the second quarter of 2022.

Data from the MISSION Phase 2 clinical trial are preliminary and will require confirmation in additional patients as well as longer follow-up to draw any clinical conclusion. Interim top-line and preliminary data from Kezar's clinical trials that it announces or publishes from time to time may change as more patient data become available and are subject to audit and verification procedures that could result in material changes in the final data.

Information on Today's Webcast

In connection with this announcement, Kezar will host a live video webcast at 4:30 p.m. ET today, November 15, 2021. The Investor and Analyst Day will highlight KZR-616, including these interim results, as well as a presentation from Dr. Parikh. Additionally, Kezar will present an overview of their protein secretion drug discovery platform and lead oncology candidate, KZR-261, including details around the Phase 1 trial design and the tumor selection process.

The live video webcast may be accessed through IR Calendar tab on the News & Events

page in the Investors section of Kezar's website at www.kezarlifesciences.com/. Alternatively, the conference call may be accessed through the following:

Live webcast: https://kvgo.com/corporate-services/kezar-investor-analyst-day-2021

For those unable to participate in the conference call or webcast, a replay will be available for 90 days on the Investors section of Kezar's website at www.kezarlifesciences.com/.

About Lupus Nephritis

Lupus nephritis (LN) is one of the most serious complications of systemic lupus erythematosus (SLE). LN is a disease comprising a spectrum of vascular, glomerular and tubulointerstitial lesions and develops in approximately 50% of SLE patients within 10 years of their initial diagnosis. LN is associated with considerable morbidity, including an increased risk of end-stage renal disease requiring dialysis or renal transplantation and an increased risk of death. There are limited approved therapies for the treatment of LN. Management typically consists of induction therapy to achieve remission and long-term maintenance therapy to prevent relapse.

About MISSION

MISSION (NCT03393013) is a Phase 1b/2 clinical trial evaluating KZR-616 in SLE patients with and without nephritis. The study consists of two parts. The Phase 1b portion is an open-label dose escalation study evaluating doses up to 75 mg of KZR-616 across 6 cohorts. The primary objective of the Phase 1b portion of MISSION is to assess safety and tolerability. Secondary objectives include evaluating pharmacokinetics (PK) and pharmacodynamics (PD) and selecting dose levels for Phase 2 trials. Several exploratory efficacy measures are also being assessed: Systemic Lupus Erythematosus Disease Activity Index 2000 (SLEDAI-2K), Cutaneous Lupus Erythematosus Severity Index-Activity (CLASI-A), Tender and Swollen Joint Counts (TJC/SJC), Physician Global Assessment (PhGA), Patient Global Assessment (PtGA) and Patient Assessment of Pain (PtP). The Phase 1b portion has been completed, and the Phase 2 portion evaluating KZR-616 in patients with LN has reached target enrollment.

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 trials are underway in multiple 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 asset, is a selective immunoproteasome inhibitor being evaluated in Phase 2 clinical trials in lupus nephritis, dermatomyositis and polymyositis. This asset also has the potential to address multiple chronic immune-mediated diseases. KZR-261, is the first anti-cancer clinical candidate from the company's platform targeting the Sec61 translocon and the protein secretion pathway. An open-label dose-escalation Phase 1 clinical trial of KZR-261 to assess safety, tolerability and preliminary tumor activity in solid tumors is underway. 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 potential use of Kezar's product candidates to treat patients, the design, progress, timing, scope and results of clinical trials, the anticipated timing of disclosure of top-line data from clinical trials and the likelihood that data, including interim and top-line data, will support future development and therapeutic potential, the association of data with treatment outcomes and the likelihood of obtaining regulatory approval of Kezar's product candidates. Many factors may cause differences between current expectations and actual results, including the availability of additional data, confirmation of data resulting from trial auditing and verification procedures, unexpected safety or efficacy data observed during clinical studies, clinical trial site activation or enrollment rates that are lower than expected, the impacts of the COVID-19 pandemic on the company's business and clinical trials, clinical trial audit and verification procedures that could result in material changes in the final data, 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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