June 3, 2020

Kezar Provides Data Update from MISSION Study of KZR-616

- ***Two of two patients with Lupus Nephritis saw greater than 50% reduction in proteinuria***
- ***KZR-616 demonstrates improvement on exploratory efficacy measures across seven measures of disease activity***
- ***Step-up dosing drives positive safety and tolerability profile***

SOUTH SAN FRANCISCO, Calif., June 03, 2020 (GLOBE NEWSWIRE) -- Kezar Life Sciences, Inc. (Nasdaq: KZR), a clinical-stage biotechnology company discovering and developing novel small molecule therapeutics to treat unmet needs in autoimmune disease and cancer, today announced updated results from the Phase 1b (Ph1b) portion of MISSION, which is evaluating the safety and tolerability of KZR-616 in patients with systemic lupus erythematosus (SLE) with and without nephritis.

Overall, improvements were seen across seven measures of disease activity, and two of two patients with lupus nephritis (LN) experienced a greater than 50% reduction in proteinuria, a biomarker of disease severity. A positive safety and tolerability profile was observed with step-up dosing of KZR-616. The Phase 1b dataset builds on the extensive safety and tolerability testing performed in 100 healthy subjects from two Phase 1a studies.

“KZR-616 is demonstrating a favorable safety and tolerability profile at doses consistent with potent and selective inhibition of immunoproteasome activity. Most strikingly, there are early and strong signs of modification of SLE disease activity, including a reduction in symptoms and biomarkers. Given the encouraging exploratory efficacy data, we are evaluating opportunities to quickly bring KZR-616 to patients with a wide array of severe autoimmune diseases,” said Noreen Henig, MD, Kezar’s Chief Medical Officer.

As of the May 4, 2020 data cutoff, the Ph1b portion of MISSION enrolled 39 SLE patients across five dose cohorts evaluating 45 mg and step-up dosing to 60 mg weekly for 13 weeks. Patients are followed to week 25 and kept on stable background treatment. Below are the results for step-up dosing Cohorts 2a, 2b, and 2c, which enrolled 26 patients. At this time point, a total of 16 patients from these cohorts completed 13 weeks of treatment (10 from Cohort 2a and 6 from Cohort 2b) and are included in the exploratory efficacy measures reported below.

- Two SLE patients with biopsy-proven lupus nephritis were included in the Phase 1b portion. Both patients showed a greater than 50% reduction in proteinuria as measured by urine protein to creatine ratio (UPCR), as well as reductions in SLEDAI and reductions in anti-dsDNA (double-stranded DNA) antibody levels.

Patient 1:

	UPCR	SLEDAI-2K	Anti-dsDNA antibodies (IU/ml)
Baseline	3.85	17	134
Week 13	2.89	8	53
Week 17	0.6	8	73
Week 25	1.0	8	61

Patient 2 (patient has not yet completed study):

	UPCR	SLEDAI-2K	Anti-dsDNA antibodies (IU/ml)
Baseline	2.39	14	123.5
Week 5	1.03	10	82
Week 13	0.69	NA	52

- Among patients completing treatment in Cohorts 2a and 2b, all seven measures of disease activity improved (decrease in score) in the majority of patients from Baseline to Week 13. Improvement in disease activity persisted following the end-of-treatment.

Mean Score for Cohorts 2a and 2b Patients completing Study (n=16)			
	Baseline	**End of Treatment (Week 13)**	**End of Study (Week 25)**
Systemic Lupus Erythematosus Disease Activity Index 2000 (SLEDAI-2K)	9.4	6.8	6.7
Cutaneous Lupus Erythematosus Severity Index-Activity (CLASI-A)	5.7	3.1	2.9
Tender Joint Count (TJC)	10.8	4.6	5.6
Swollen Joint Count (SJC)	7.1	2.6	2.6
Physician Global Assessment (PhGA)	55.7	40.8	35.6
Patient Global Assessment (PtGA)	60.5	40.6	41.5
Patient Assessment of Pain (PtP)	60.3	49.4	41.6

Note: No patients in Cohort 2c have reached 13 weeks of treatment as of May 4, 2020.

- Step-up dosing of KZR-616 improved overall tolerability. Most treatment emergent adverse events (TEAEs) occurred early and diminished with later doses. To date, no patients have discontinued treatment in Cohorts 2b and 2c, which utilize a lyophilized formulation of KZR-616.

- The most common treatment emergent adverse events were transient injection site reactions.

N (%) of patients	Cohort 2a (n=14)	Cohort 2b (n=6)	Cohort 2c* (n=6)	All Patients (Cohorts 1-2c) (n=39)
TEAEs	12 (85.7)	4 (66.7)	5 (83.3)	34 (87.2)
Injection Site Reactions	9 (64.3)	2 (33.3)	4 (66.7)	25 (64.1)
Nausea	5 (35.7)	1 (16.7)	2 (33.3)	14 (35.9)
Vomiting	4 (28.6)	1 (16.7)	1 (16.7)	12 (30.8)
TEAEs ≥ Grade 3	2 (14.3)	1 (16.7)	0 (0.0)	4 (10.3)
Infectious TEAEs ≥ Grade 3	1 (7.1)	1 (16.7)	0 (0.0)	2 (5.1)
Infectious TEAEs; All Grades	5 (37.5)	2 (33.3)	0 (0.0)	8 (20.5)
Serious TEAEs	2 (14.3)	1 (16.7)	0 (0.0)	4 (10.3)
Any Discontinuation	4 (28.6)	0 (0.0)	0 (0.0)	10 (25.6)

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 which is evaluating doses of 45 mg, 60 mg and 75 mg of KZR-616 across 6 cohorts. The primary objective of the Ph1b portion of MISSION is to assess safety and tolerability. Secondary objectives include evaluating pharmacokinetics (PK) and pharmacodynamics (PD) and selecting dose levels for the 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), and Physician Global Assessment (PhGA)/Patient Global Assessment (PtGA)/Patient Assessment of Pain (PtP).

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 collected to date from two Phase 1a studies in healthy volunteers and the Phase 1b clinical trial in patients with systemic lupus erythematosus establish the positive safety and tolerability profile of KZR-616. Additionally, results from the Phase 1b study provide early evidence that KZR-616 demonstrates improvement across measures of disease activity. Phase 2 trials are underway for the treatment of lupus nephritis (MISSION), dermatomyositis and polymyositis (PRESIDIO), and autoimmune hemolytic anemia and immune thrombocytopenia (MARINA).

About Kezar Life Sciences

Based in South San Francisco, Kezar Life Sciences is a clinical-stage biotechnology company committed to revolutionizing treatments for patients with autoimmune diseases and cancer. Kezar is translating its innovative research on the immunoproteasome and protein secretion pathways to advance novel therapeutic approaches. KZR-616, a first-in-class selective immunoproteasome inhibitor, is being evaluated in severe autoimmune diseases, including systemic lupus erythematosus (SLE), lupus nephritis (LN), dermatomyositis (DM), polymyositis (PM), autoimmune hemolytic anemia (AIHA) and immune thrombocytopenia (ITP). Additionally, Kezar has nominated KZR-261 as its first clinical candidate for the

treatment of cancer from its protein secretion program and is undergoing IND-enabling studies for the program. 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 likelihood that data will support future development, the association of data with treatment outcomes, the design, progress, timing, scope and results of clinical trials, the anticipated timing of disclosure of results of clinical trials 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 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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