August 12, 2021

Kezar Life Sciences Reports Second Quarter Financial Results and Provides Business Updates

- *KZR-616 MISSION Phase 1b study results support continued development for multiple immune mediated diseases*
- *Investigational new drug (IND) application filed for first-in-class protein secretion inhibitor, KZR-261*
- *Expertise in autoimmune drug development added with appointment of Micki Klearman, MD, to Board of Directors and creation of Clinical Advisory Committee*

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 its second quarter 2021 financial results and corporate highlights.

“We were pleased this quarter to share the final data update from our completed Phase 1b portion of the MISSION study, building on the positive safety, tolerability and early efficacy profile of KZR-616. I commend the entire team at Kezar on another quarter of outstanding execution across both of our programs and look forward to providing meaningful updates from both before the end of the year,” said John Fowler, Kezar’s Co-founder and Chief Executive Officer. “We are also pleased to announce that Kezar recently submitted an IND application to the FDA for KZR-261, our first-in-class protein secretion inhibitor. Like the platform potential we see for KZR-616 in autoimmunity, we believe KZR-261 could have broad potential across the oncology landscape. We look forward to sharing more about our planned Phase 1 trial design and the potential of our protein secretion drug discovery platform in the coming months.”

Clinical Highlights & Updates

KZR-616: Selective Immunoproteasome Inhibitor

MISSION – Phase 2 clinical trial in patients with lupus nephritis (LN) (NCT03393013)

At the European Congress of Rheumatology (EULAR 2021) in June, Kezar presented final clinical data from the completed Phase 1b dose escalation portion of the MISSION study in 47 patients with systemic lupus erythematous, including two patients with active proliferative LN. KZR-616 demonstrated improvement across all exploratory efficacy measures and was well-tolerated up to 75 mg subcutaneously once weekly for 13 weeks. In the two patients with LN, improvements in renal function correlated with reductions in a key biomarker of kidney inflammation, uCD163.

- The safety and tolerability profile observed with KZR-616 was consistent with previously reported data and supports treatment for chronic use. No new safety or

tolerability signals were observed from previously reported data.
- These data support the development of KZR-616 in multiple immune-mediated diseases.
    - Doses being investigated in the Phase 2 clinical trials are 60 mg (MISSION Phase 2 in LN) and 45 mg (PRESIDIO Phase 2 in dermatomyositis and polymyositis) with weekly subcutaneous dosing.
- The amended Phase 2 open-label portion of the MISSION trial in patients with active, proliferative LN opened for enrollment in August 2020 and is actively recruiting. 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 six months.
    - Kezar reiterates prior guidance and expects interim data to be reported in Q4 2021, and topline data are expected in the first half of 2022.

PRESIDIO – Phase 2 clinical trial in patients with dermatomyositis (DM) and polymyositis (PM) (NCT04033926)

- The PRESIDIO Phase 2, placebo controlled cross-over trial of KZR-616 in DM and PM is actively enrolling. Additionally, a 12-month open-label extension study is open to patients completing the 32-week placebo-controlled trial (NCT04628936).
    - Topline data are expected in the first half of 2022.

KZR-261: Protein Secretion Inhibitor

- KZR-261 is a first-in-class protein secretion inhibitor that targets the Sec61 translocon and has demonstrated broad anti-tumor activity in preclinical models of both solid and hematologic malignancies.
    - Kezar submitted an investigational new drug (IND) application for KZR-261 to the U.S. Food and Drug Administration (FDA) in August 2021. The company plans to initiate a Phase 1 clinical trial of KZR-261, which will assess safety, tolerability and preliminary tumor activity of KZR-261 in solid tumors.
    - At the American Association of Cancer Research (AACR) 2021 Virtual Annual Meeting in April, Kezar presented preclinical data on its novel small molecule inhibitors of the Sec61 translocon. These data support the therapeutic potential of inhibiting Sec61 and the protein secretion pathway as a way to generate novel therapies to treat multiple tumor indications.

Corporate Updates

- Kezar formed a Clinical Advisory Committee in June, comprised of world-renowned thought leaders in immunology, rheumatology, neurology and nephrology. Appointments to the committee include: Rohit Aggarwal, MD, MS, Professor of Medicine, University of Pittsburgh; Prof. Olivier Benveniste, MD, PhD, Professor of Internal Medicine & Immunology, Sorbonne Université, Pitie Salpetriere Hospital; Mazen Dimachkie, MD, Professor of Neurology, University of Kansas Medical Center; Ingrid Lundberg, MD, PhD, Professor of Medicine, Karolinska Institute; Samir V. Parikh, MD, Assistant Professor of Medicine, Ohio State University Wexner Medical Center; and Onno Teng, MD, PhD, Nephrology Clinical Scientist, Leiden University Medical Center.

- Micki Klearman, MD, rheumatologist and internist, was appointed to Kezar's Board of Directors, bringing over two decades of biopharmaceutical experience with her significant contributions to the fields of immunology and rheumatology.

Second Quarter 2021 Financial Results

- **Cash, cash equivalents and marketable securities** totaled $129 million as of June 30, 2021, compared to $140 million as of December 31, 2020. The decrease in cash, cash equivalents and marketable securities was primarily attributable to cash used by the company in operations to advance its clinical-stage programs and preclinical research and development, offset by the net proceeds from the issuance of common stock in February 2021, under the "at-the-market" Sales Agreement with Cowen and Company, LLC.
- **Research and development expenses** for the second quarter of 2021 increased by $2.2 million to $9.3 million compared to $7.1 million in the second quarter of 2020. This increase was primarily related to advancing the KZR-616 clinical program in multiple indications and the protein secretion preclinical program.
- **General and administrative expenses** for the second quarter of 2021 increased by $1.0 million to $3.7 million compared to $2.7 million in the second quarter of 2020. The increase was primarily due to an increase in stock-based compensation and personnel and recruiting expenses as a result of an increase in headcount and salaries and an increase in the cost of directors' and officers' liability insurance.
- **Net loss** for the second quarter of 2021 was $13.0 million, or $0.25 per basic and diluted common share, compared to a net loss of $9.5 million, or $0.22 per basic and diluted common share, for the second quarter of 2020.
- **Total shares of common stock outstanding** were 48.1 million shares as of June 30, 2021. Additionally, there were outstanding pre-funded warrants to purchase 3.8 million shares of common stock at an exercise price of $0.001 per share and outstanding options to purchase 7.3 million shares of common stock at a weighted-average exercise price of $5.86 per share as of June 30, 2021.

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 severe autoimmune diseases.

About KZR-261

KZR-261, a novel, first-in-class protein secretion inhibitor, is the first clinical candidate to be nominated from Kezar's research and discovery efforts targeting protein secretion pathway. KZR-261 is a broad-spectrum anti-tumor agent that acts through direct interaction and inhibition of Sec61 activity. The compound was discovered by Kezar through a robust medicinal chemistry campaign in which several scaffolds were progressed through the company's proprietary platform evaluating Sec61 modulation. As a result, Kezar has established a broad library of protein secretion inhibitors. KZR-261 has demonstrated

several encouraging properties that lead to its potential to be an anti-cancer agent for the treatment of solid and hematologic malignancies. An IND submission in solid tumors was filed in August 2021 and a Phase 1 trial is expected to commence once the IND goes into effect.

About Lupus Nephritis

Lupus nephritis (LN) is one of the most serious complications of systemic lupus erythematosus. 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 Dermatomyositis and Polymyositis

Dermatomyositis (DM) and Polymyositis (PM) are two of the five types of autoimmune myositis diseases. Both are chronic, debilitating, inflammatory autoimmune myopathies that are distinguished by inflammation of the muscles as well as the skin (in DM). Approximately 30,000 to 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.

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, the first anti-cancer clinical candidate from the company's platform targeting the Sec61 translocon and the protein secretion pathway, is expected to be evaluated in Phase 1 clinical trials in solid tumors, pending effectiveness of its IND application. 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 regulatory development of Kezar's product candidates, the anticipated timing of disclosure of results, including interim and topline data, of clinical trials, the likelihood 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 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.

KEZAR LIFE SCIENCES, INC.
Selected Balance Sheets Data
(In thousands)

	June 30, 2021	December 31, 2020
	(unaudited)	
Cash, cash equivalents and marketable securities	$ 128,975	$ 140,447
Total assets	139,672	151,842
Total current liabilities	5,944	6,442
Total stockholders' equity	129,886	140,978

Summary of Operations Data
(Unaudited in thousands except share and per share data)

	Three Months Ended June 30,		Six Months Ended June 30,	
	2021	2020	2021	2020
	(unaudited)		(unaudited)	
Operating expenses:				
Research and development	$9,341	$7,148	$18,627	$14,605
General and administrative	3,668	2,705	7,430	5,726
Total operating expenses	13,009	9,853	26,057	20,331
Loss from operations	(13,009)	(9,853)	(26,057)	(20,331)
Interest income	47	353	101	819
Net loss	$(12,962)	$(9,500)	$(25,956)	$(19,512)
Net loss per common share, basic and diluted	$(0.25)	$(0.22)	$(0.50)	$(0.51)
Weighted-average shares used to compute net loss per common share, basic and diluted	51,904,701	42,936,991	51,483,709	37,902,294

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