

Corporate Presentation

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This presentation contains forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995. Words such as "may," "will," "should," "expect," "believe", "plan" 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 presentation. 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 presentation include, but are not limited to, statements about the design, progress, timing, scope and results of clinical trials, the preliminary nature of topline data, the anticipated regulatory development and future clinical trials involving Kezar's product candidates, the likelihood that 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 performance of audit and verification procedures on topline data, unexpected safety or efficacy data observed during clinical studies, changes in expected or existing competition, 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 presentation 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.

The Kezar Opportunity: First-in-Class, Small Molecule Therapies That Tackle Immune-Mediated Diseases and Cancer

Deep Expertise in Immunology and Oncology

Builds on 10+ years of R&D work in proteasome biology and protein secretion led by Kezar's scientific cofounders, Chris Kirk & Jack Taunton

Zetomipzomib (KZR-616): First-in-Class Immunoproteasome Inhibitor

A novel approach to harmonizing the immune system via immunomodulation. Potential to be a pipeline in a drug

KZR-261: First Clinical Candidate from Protein Secretion Platform

First-in-class inhibitor of Sec61 translocon. Impacts tumor proliferation, metastasis and immune invasion. Currently being evaluated in a Phase 1 study in solid tumors

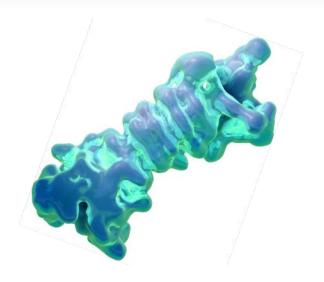
Strong Financial Position

\$307M cash, cash equivalents and marketable securities as of June 30, 2022; 68.3M common shares outstanding

Kezar's Novel, Complementary Programs Target Master Regulators of Cellular Function to Achieve Broad Therapeutic Activity

SELECTIVE IMMUNOPROTEASOME INHIBITION:

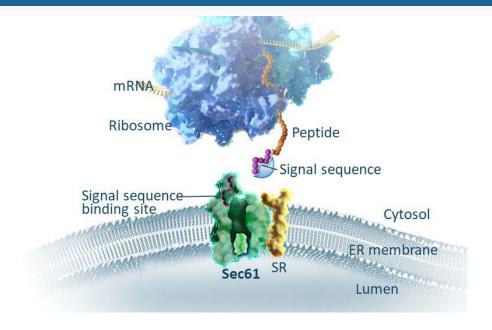
The Immunoproteasome



- Modulates multiple drivers of inflammation
- Restores normal immune responses, without evidence of immunosuppression

PROTEIN SECRETION INHIBITION:

The Sec61 Translocon

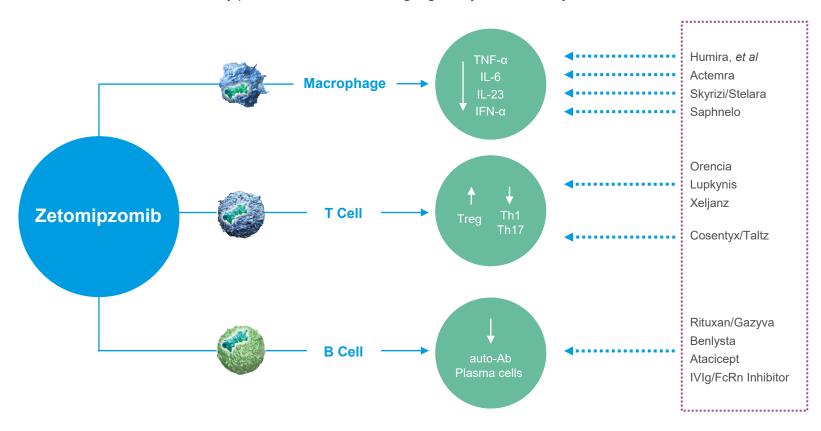


- Broad anti-tumor activity in preclinical models
- Applications in oncology, immuno-oncology, and autoimmunity
- Potential for small molecules to replace certain biologics

Zetomipzomib, a First-in-Class Inhibitor of the Immunoproteasome, Acts Across the Innate and Acquired Immune System

Immunoproteasome Inhibition with Zetomipzomib*

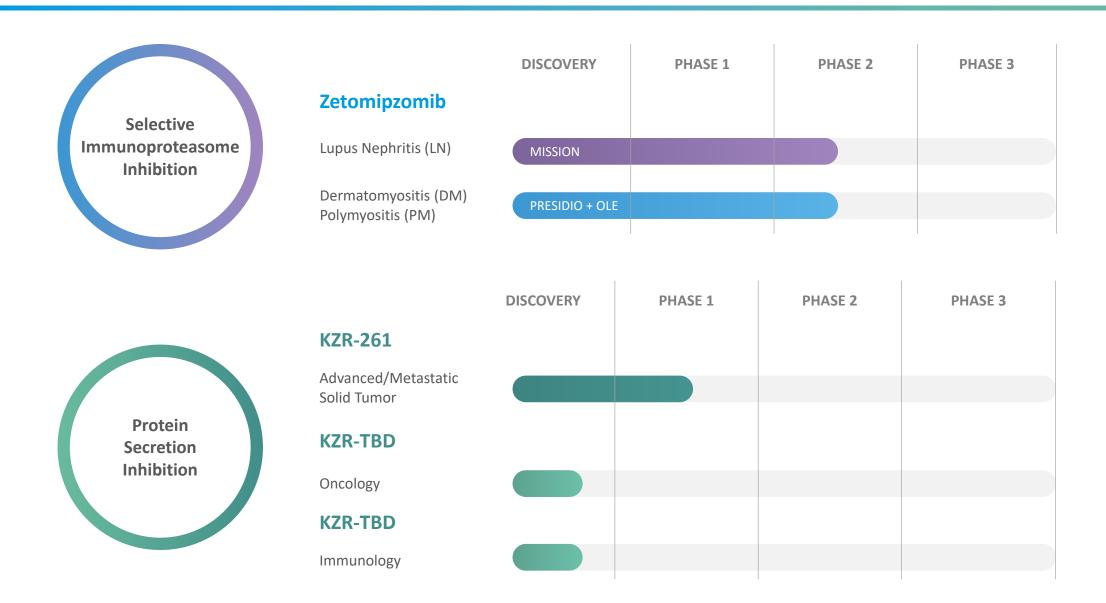
Decreases pro-inflammatory cytokine production, plasma cell activity and autoantibody production, while increasing regulatory T cells activity



Zetomipzomib Advantage

Inflammatory disorders are currently treated by targeting a single immune signal but zetomipzomib affects a broad spectrum of immune regulators

Our Programs Inhibit Multiple Drivers of Disease to Address a Diverse Pipeline of Indications







MISSION:

Topline Data MISSION Phase 2 Study Evaluating Zetomipzomib 60 mg SC QW for 24 Weeks in Lupus Nephritis



MISSION Phase 1b (Complete): Zetomipzomib Demonstrated Safety, Tolerability and Preliminary Efficacy in Patients with SLE with and without LN



- Treatment with zetomipzomib demonstrated improvements in multiple exploratory measures of disease activity across organ systems including SI FDAI-2K
- 2 of 2 subjects with active LN showed >50% reduction in UPCR and decreases in uCD163, a marker for inflammatory activity in LN
- Anti-dsDNA antibody titers improved over time in 8 of 8 patients with elevated levels at baseline
- Sustained or deepened improvement (tail-effect) in disease activity measures and key biomarkers in the post-treatment follow-up period observed
- The most common treatment-emergent adverse event was injection site erythema
- Favorable safety profile, with hematologic analysis showing no suppression of leukocytes through the duration of the study

MISSION Phase 2 Topline: Highlights Zetomipzomib Achieves Clinically Meaningful Overall Renal Response (ORR) in Refractory or Hard-to-Treat LN Patients without Standard Induction Therapy

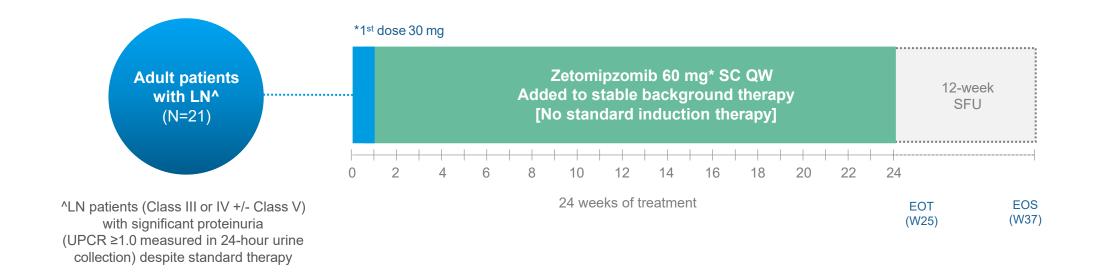


- 11/17 (64.7%) patients demonstrated an ORR of ≥50% UPCR reduction following 24 weeks of treatment (primary endpoint)
- 6/17 (35.2%) patients achieved a CRR with UPCR ≤0.5
- Treatment benefit was maintained or deepened following end-of-treatment period (measured at W29)
- Prednisone (or prednisone-equivalent) mean daily dose was decreased by 53% from baseline to end-of-treatment and continued to decrease following the end-of-treatment period
- Use of other standard immunosuppressive therapy was stable
- Mean eGFR remained stable
- Improvements observed in key SLE disease activity and disease biomarkers
- Favorable safety and tolerability profile observed, with no reports of opportunistic infections or immune cell depletion, supporting chronic administration

MISSION Phase 2: Study Design Open-Label Clinical Study to Evaluate the Efficacy and Safety of Zetomipzomib in Patients

with Active Proliferative Lupus Nephritis





Patients in MISSION Phase 2 did not receive standard induction therapy or protocol-mandated steroid taper

Lack of induction therapy is a significant difference compared to recently published trials in LN



MISSION Phase 2: Study Design Endpoints



Primary Endpoint: Overall Renal Response (ORR)

 Number of patients with ≥50% reduction in UPCR compared to baseline after 24 weeks of treatment with zetomipzomib

Key Secondary Endpoints

- Safety and tolerability of zetomipzomib
- The number of patients with a complete renal response (CRR) and partial renal response (PRR) after 24 weeks of treatment as defined by:

CRR:

- UPCR ≤0.5
- eGFR ≥60 mL/min/1.73m² or no worsening of eGFR from baseline of ≥25%
- Prednisone (or equivalent) ≤10 mg
- No use of prohibited medication

PRR:

- 50% reduction in UPCR and/or UPCR <1 (if baseline UPCR <3) and/or UPCR
 (if baseline UPCR >3)
- eGFR ≥60 mL/min/1.73m² or no worsening of eGFR from baseline of ≥25%
- No use of prohibited medication

Global SLE disease monitoring and key biomarkers

MISSION Phase 2 Topline: Safety Population Key Demographics and Baseline Characteristics



	Safety Population (N=21)
Age, mean (years), range	35.3 (19 - 69)
Female, n (%)	19 (90.5)
Race, n (%) White Black or African American Asian American Indian/Alaska Native Other Unknown Ethnicity, n (%) Hispanic or Latino	7 (33.3) 1 (4.8) 1 (4.8) 4 (19.0) 7 (33.3) 1 (4.8)
Hispanic or Latino Country, n (%) Australia Colombia Peru Russia Ukraine USA	3 (14.3) 4 (19.0) 6 (28.6) 4 (19.0) 1 (4.8) 3 (14.3)

	Safety Population (N=21)
SLE duration (years), median (min, max), N=20	8.7 (0.7, 26.4)
LN duration (years), median (min, max)	3.1 (-0.06*, 16.1)
LN class type, n	
Class III only	6
Class IV only	11
Class III + V	3
Class IV + V	1
24-hour UPCR (mg/mg)	
Mean (SD)	2.6 (2.6)
Median (Min, Max)	1.9 (0.93, 13.4)
eGFR (mL/min/1.73 m2)	
Mean (SD)	104.7 (32.8)
Median (Min, Max)	115.0 (36.5, 150)
Disease Activity Parameter, mean (SD)	
SLEDAI-2K	11.0 (4.7)
Corticosteroid (prednisone or equivalent) dose (mg), mean (min, max)	18.9 (5, 50)
Concomitant medications, n	
MMF/MPA	20
Prednisone (or equivalent)	21
Hydroxychloroquine	14
Azathioprine	2

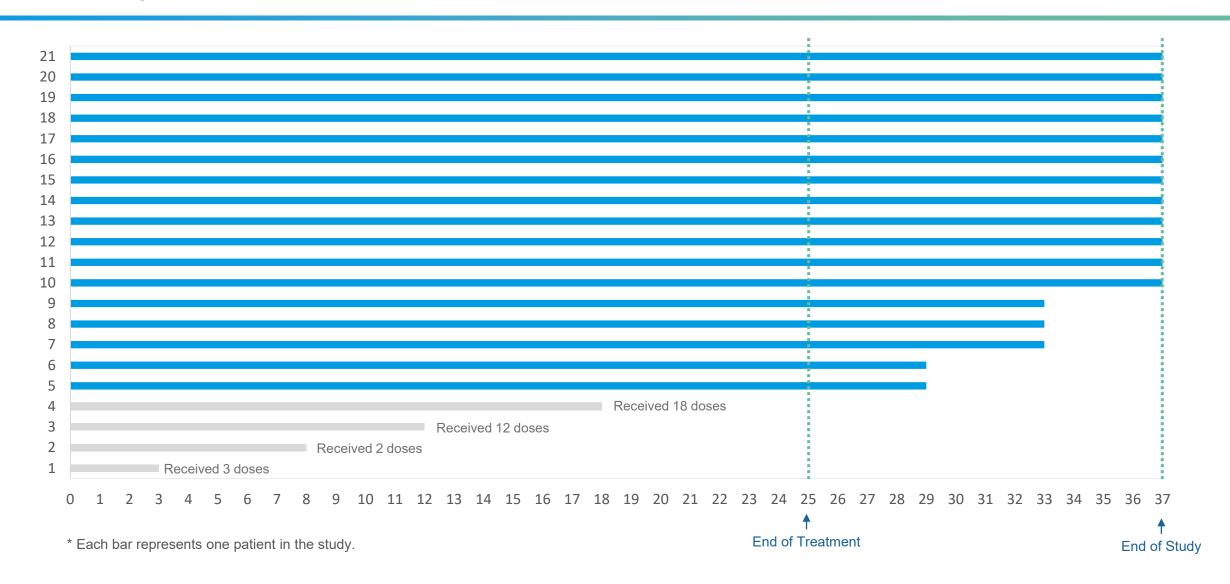
Abbreviation: SLE, systemic lupus erythematosus; LN, lupus nephritis; UPCR, urine protein to creatinine ratio; eGFR, estimated glomerular filtration rate; SLEDAI, Systemic Lupus Erythematosus Disease Activity index-2000; MMF, mycophenolate mofetil; MPA, mycophenolic acid.



^{*}LN biopsy confirming diagnosis was performed during screen period.

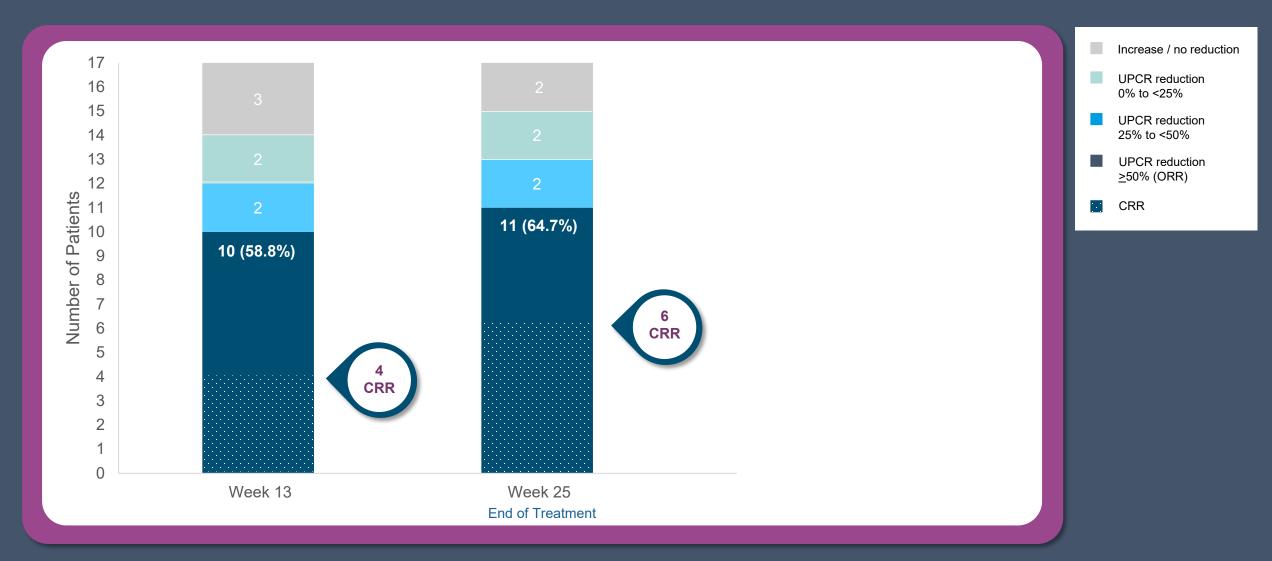
MISSION Phase 2 Topline: Patient Disposition*





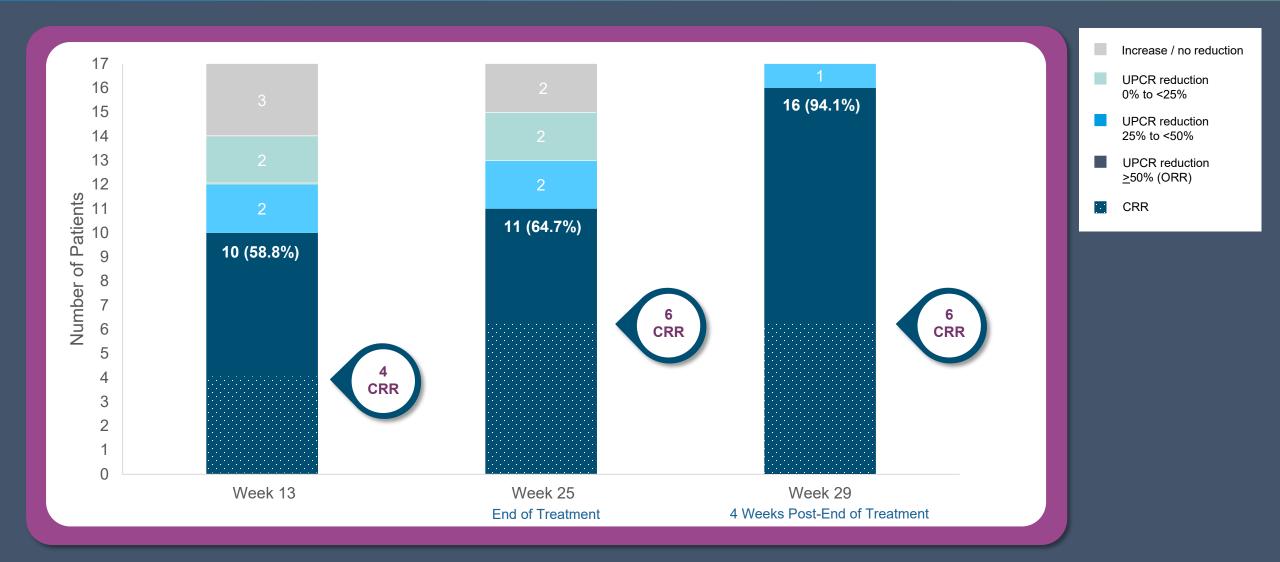
MISSION Phase 2 Topline: Evaluable Population Zetomipzomib Without Standard Induction Therapy Achieves ORR in 65% and CRR in 35% of Patients at Week 25





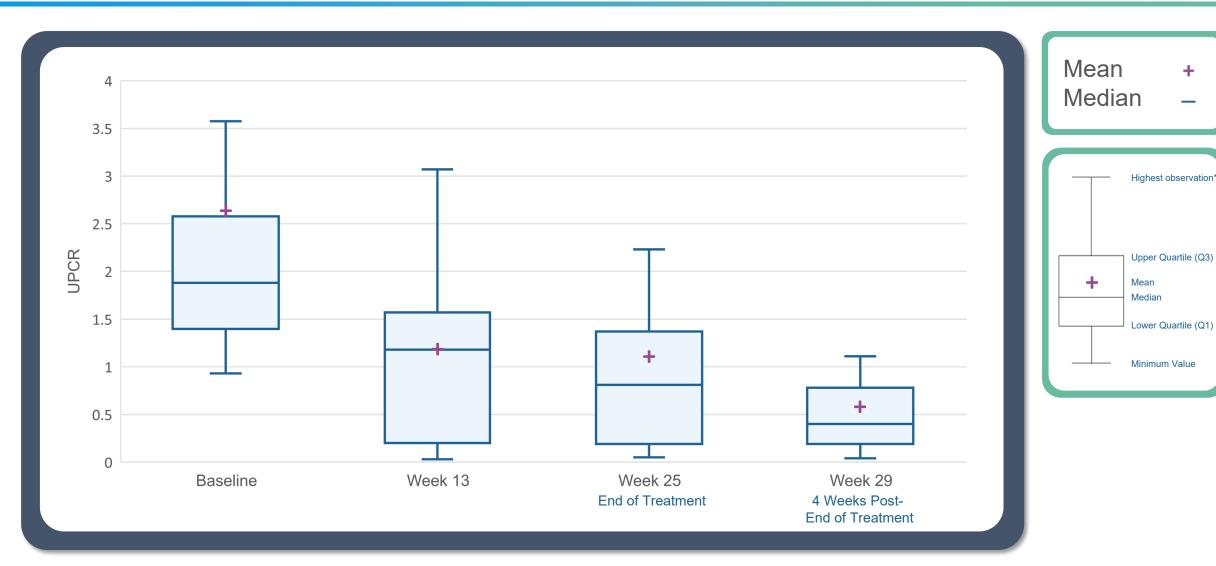
MISSION Phase 2 Topline: Evaluable Population CRRs Are Maintained and Additional ORRs are Observed at Week 29, Post-Treatment with Zetomipzomib





MISSION Phase 2 Topline: Evaluable Population Mean UPCR Decreases with 24 Weeks of Zetomipzomib Treatment



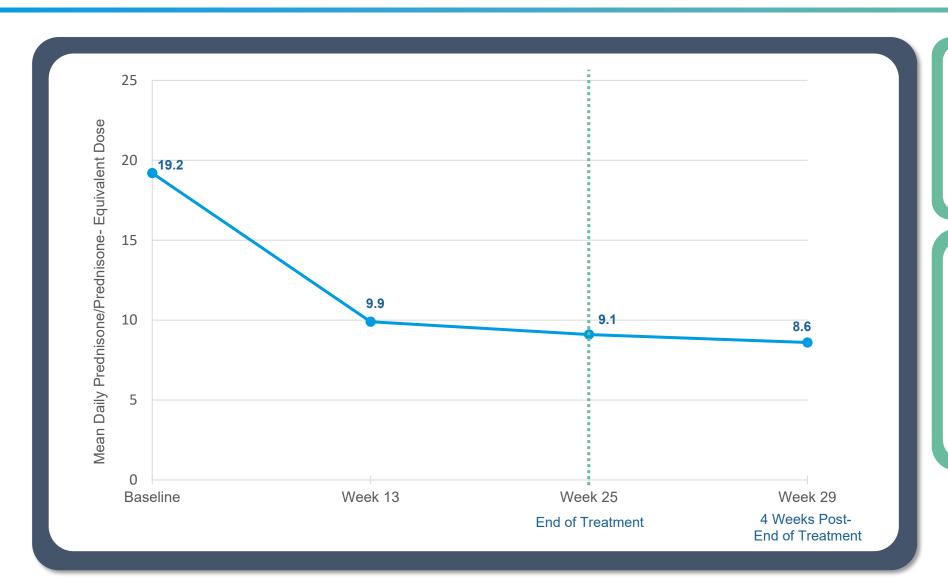


Evaluable population (n=17) are patients that did not withdraw before Week 25; Patients received 24 weeks of zetomipzomib; End-of-treatment assessments performed at Week 25. *Outliers are not shown on the plot but are incorporated in the mean/median values. Abbreviation: UPCR, urine protein to creatinine ratio. Topline data is subject to audit and verification procedures that could result in material changes in the final data.



MISSION Phase 2 Topline: With Zetomipzomib, Reductions in Proteinuria Achieved with 53% Less Corticosteroids







- No protocol-mandated steroid taper
- Other background immunosuppressive doses remained stable

MISSION Phase 2 Topline: Evaluable Population Mean eGFR Remains Stable





Evaluable population (n=17) are patients that did not withdraw before Week 25; Patients received 24 weeks of zetomipzomib; End-of-treatment assessments performed at Week 25. Stable eGFR defined as ≥60 mL/min/1.73m² or no worsening of eGFR from baseline of ≥25% **Abbreviations:** eGFR, estimated glomerular filtration rate.

MISSION Phase 2 Topline: Evaluable Population Reduction in Signs and Symptoms of SLE with Zetomipzomib Treatment



Tool	Baseline Mean (SD)	EOT (Week 25) Mean (SD)				
SLEDAI-2K	11.1 (4.7)	6.5 (3.1)				
Physician Global Assessment Score	57.2 (21.7)	23.9 (19.2)				
Patient Global Assessment Score	23.6 (21.1)	10.7 (12.2)				
HAQ-pain	20.8 (18.2)	12.1 (18.1)				
CLASI-A	3.7 (7.3)	1.9 (4.1)				
Tender Joint Count	1.3 (2.6)	0.1 (0.5)				
Swollen Joint Count	0.1 (0.5)	0.1 (0.2)				

Evaluable population (n=17) are patients that did not withdraw before Week 25.

Abbreviations: CLASI-A, Cutaneous Lupus Erythematosus Severity Index-Activity; EOT, end of treatment; HAQ, Health Assessment Questionnaire; SD, standard deviation; SLEDAI-2K, Systemic Lupus Erythematosus Disease Activity Index 2000.



MISSION Phase 2 Topline: Evaluable Population Improvement in Key Serologic Biomarkers Observed at Week 25 (EOT) in Patients with **Abnormal Levels at Baseline**



Biomarker	Patients with Abnormal Levels at Baseline	Patients with Improvement at W25 (EOT)	Patients with Normalization at Week 25 (EOT) 5			
Anti-dsDNA	12	10				
C3	5	4	2			
C4	4	2	2			

Cell counts remained stable in patients on study

Evaluable population (n=17) are patients that did not withdraw before Week 25. Reference ranges: dsDNA <20 IU/mL; C3 90 - 180 mg/dL; C4 10 - 40 mg/dL. Abbreviations: EOT, end of treatment.

MISSION Phase 2 Topline Data: Safety Population (N=21) Zetomipzomib Continues to Show Favorable Safety and Tolerability Profile



Treatment-Emergent Adverse Events

- TEAEs were generally mild to moderate (≤Grade 2) consistent with previous reports
- Most Common TEAEs occurring in >25% of patients:
 - Injection site reaction, pyrexia (fever), headache, or nausea with or without vomiting

Serious Adverse Events: 2 Patients

- Acute protracted migraine (related)
- 2. Worsening pulmonary arterial hypertension, AKI and UTI (unrelated)

Early Terminations: 4 Patients

- 1. Injection site infiltration (related)
- 2. Asthenia (related)
- 3. Reticulocytes increase (related)
- 4. Worsening pulmonary arterial hypertension (unrelated)

No opportunistic or Grade 3 infections reported

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ZETOMIPZOMIB:

Clinical Development Next Steps



Zetomipzomib, A First-in-Class Inhibitor of the Immunoproteasome: Key Attributes



- Modulates innate and acquired immune responses without signs of immunosuppression to date
- Once weekly subcutaneous administration leads to intermittent inhibition of the immunoproteasome with no accumulation observed with repeat dosing
 - Consistent exposure and clearance (T_{1/2} <5 hours)
- Not predicted to result in clinically significant drug-drug interactions
- No teratogenicity observed at any dose tested in animal models
- No monitoring required
- No immediate rebound of signs and symptoms of disease activity upon discontinuation of zetomipzomib in Mission Ph1b safety follow-up period

Zetomipzomib for the Treatment of Lupus Nephritis: Next Steps









Zetomipzomib is Well Positioned for Use as a **Chronic Therapy for the** Treatment of LN

- Potential to be used without induction therapy
- Potential to be steroidsparing
- Potential treatment for renal and extra-renal manifestations of SLE

Late Phase LN Trial

- Following the November 2021 interim data release, Kezar initiated preparations for registrational studies in LN
- Design will be informed by completion of the MISSION Phase 2 trial (EOS, July 2022) and discussions with regulatory authorities

Patient-Based **Education Initiatives**

In recognition of the significant unmet need in reaching patients with LN, Kezar will commence patient-based education initiatives

Additional Indication Exploration

- Given the extra-renal findings in the MISSION Phase 1b and Phase 2 studies, we are considering opportunities in SLF
- Further guidance will be available later this year



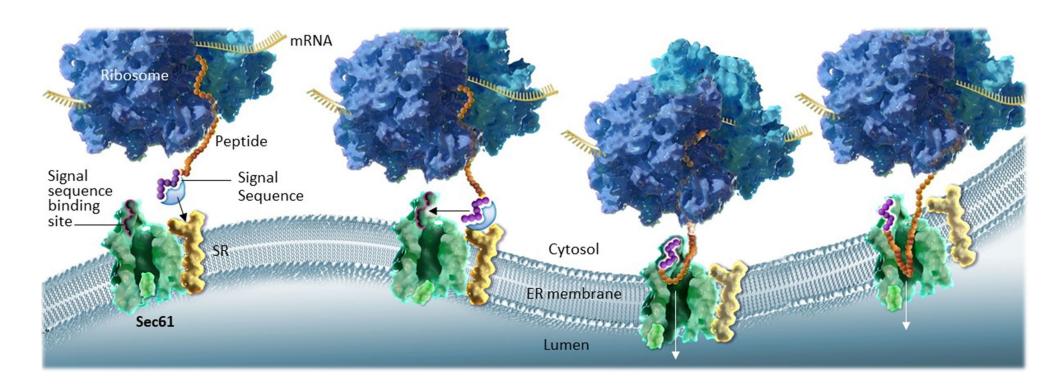
KZR-261:

Protein Secretion Inhibition

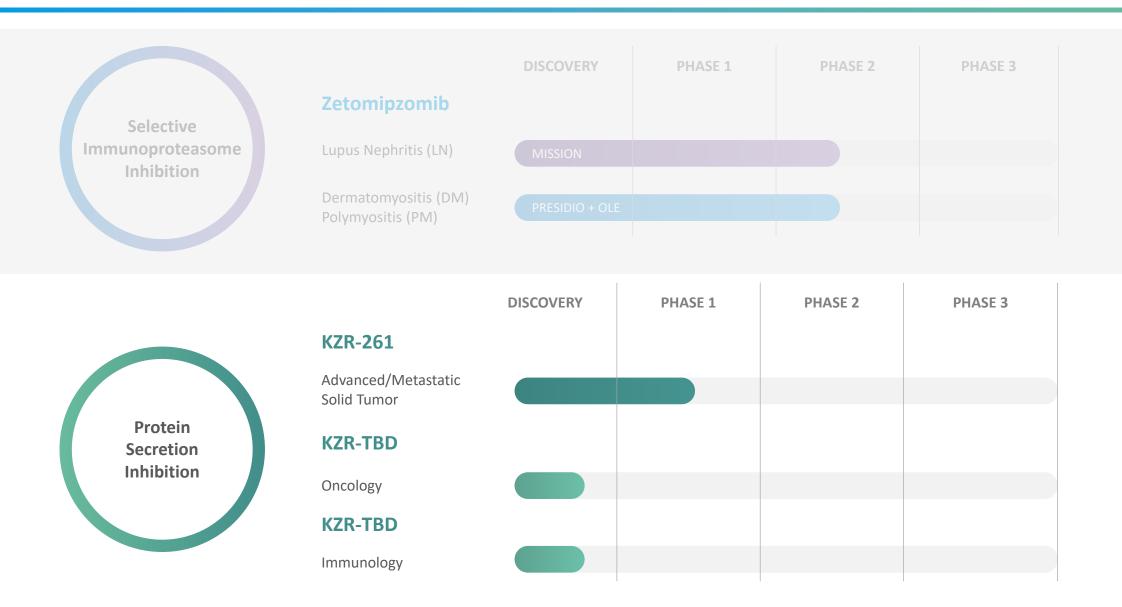


The Sec61 Translocation Channel (Translocon) is the Initiation of the Protein Secretion Pathway and a Novel Drug Target in Oncology

- Highly conserved process, functional in all cells
- Nearly all secreted and transmembrane proteins (5,000 7,000 proteins) utilize Sec61 to enter the ER
- Each protein expresses a unique signal sequence or transmembrane domain enabling selective inhibition of therapeutic targets
- Tumor cells utilize Sec61 for proliferation, metastasis and immune evasion

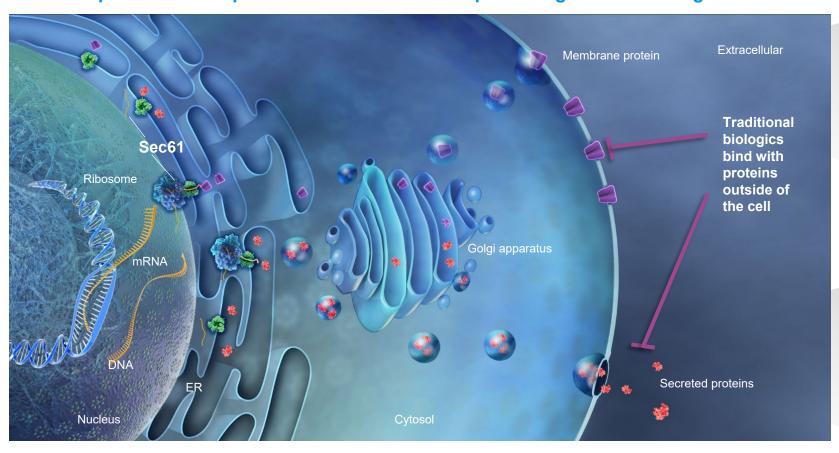


Our Programs Inhibit Multiple Drivers of Disease to Address a Diverse Pipeline of Indications



Targets of Most Biologics Utilize Sec61 for Secretion or Membrane Expression

A plethora of validated targets utilize the Sec61 translocon. Kezar's program holds the potential for superior small molecule therapeutics against these targets.



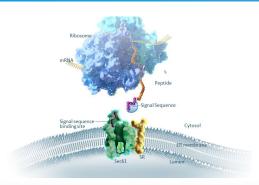
Membrane Proteins (partial list)

EGFR (ERBITUX) IL-6R (ACTEMRA) PD-1 (OPDIVO) PDL1 (TECENTRIQ) CTLA4 (YERVOY)

Secreted Proteins (partial list)

TNF-α (HUMIRA) IL-17 (COSENTYX) PCSK9 (REPATHA) IL-6 (SYLVANT) BAFF (BENLYSTA)

Kezar's Novel Platform for Drug Discovery Targets the Sec61 Translocon and the Protein Secretion Pathway



- Unique drug discovery engine developed with applications in multiple diseases
- Opportunity for orally bioavailable inhibitors of 1 or more high value targets with a single compound

Multi-Target Target Selective

Multi-Protein Secretion Inhibitors

- Inhibition of <u>multiple</u> secreted/membrane proteins
- Combination therapy in a single molecule
- Multiple oncology indications (tumor agnostic)

KZR-261: 1st clinical candidate

Subset Protein Secretion Inhibitors

- Inhibition of relevant <u>subset</u> secreted/membrane proteins
- Non-cytotoxic agents
- Indications: oncology, immuno-oncology, immunology

Single Protein Secretion Inhibitors

- Inhibition of a <u>single</u> secreted/membrane protein
- Non-cytotoxic agents
- Indications: Many...

KZR-261: Unique MOA Involving Direct Effects on Tumor Cells and the Microenvironment

Direct Effects on Tumor Cells

- Tumor cell death via proteotoxic stress
- Reduced growth factor & oncogenic RTK expression

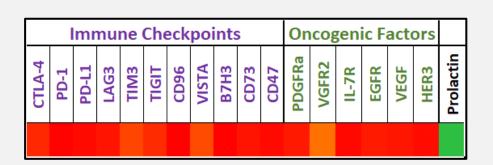


Tumor Microenvironment Modulation

- Reduced angiogenic factor expression (e.g., VEGF)
- Reduced immune checkpoint expression

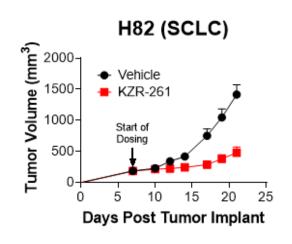
KZR-261 Blocks Expression of Immune Checkpoints and Oncogenic Factors

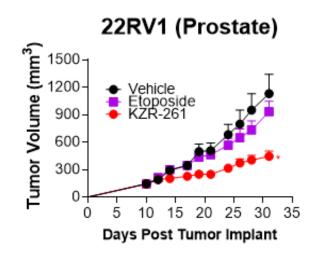
In vitro **Protein** Secretion **Assays**

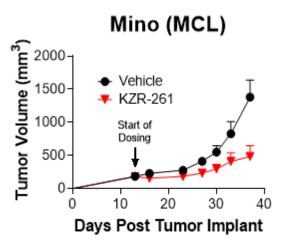




In vivo Tumor Xenografts

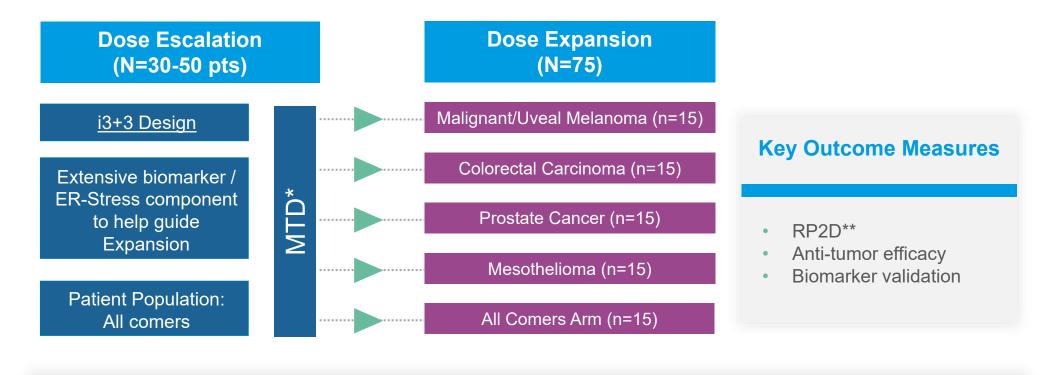






Lowe E et al. ASCO 2020

First-in-Human Dose Escalation and Expansion Study of KZR-261 Initiated



Goals for KZR-261-101

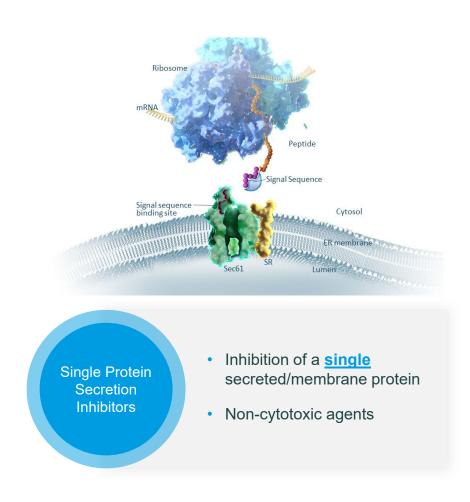
- Establish single agent activity
- Maximize opportunities for success for KZR-261
- Identify/confirm potential, predictive biomarkers

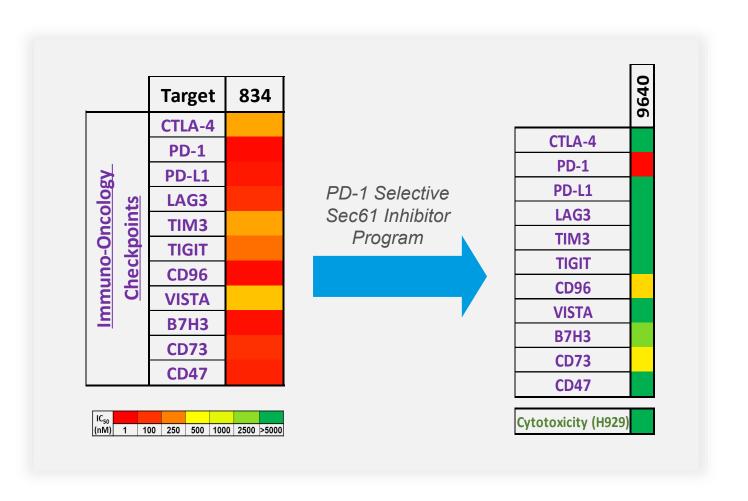
NCT05047536

^{*}Maximum Tolerated Dose

^{**}Recommended Phase 2 Dose

Kezar has Demonstrated Proof of Concept for Discovery of Highly Selective Protein Secretion Inhibitors





KZR-261 Represents Kezar's First of Multiple Opportunities to Bring Protein Secretion Inhibitors Into Clinical Development

Direct Effects on Tumor Cells

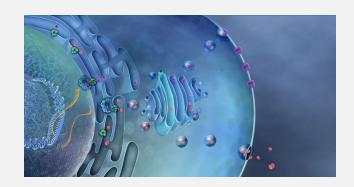
- Phase 1 study initiated in Q4'21
- Multiple oncology indications
- Potential to represent combination therapy in a single drug

IC ₅₀ (nl	IC ₅₀ (nM)							
1								
100								
250								
500								
750								
900								
>1000								

ĺ	Immune Checkpoints								Oncogenic Factors									
	CTLA-4	1-Qd	TT-Qd	F9V7	TIM3	LIBIL	96QD	VISTA	вунз	EZQ0	CD47	PDGFRa	VGFR2	IL-7R	EGFR	VEGF	HER3	Prolactin

Platform Potential of the Sec61 Translocon

- Gateway for multiple therapeutic targets across many indications
- High value drug target with ability to inhibit with hyper-selectivity as well as multi-valency
- World leading know-how in chemical targeting and target biology



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