



Selective Targets. Broad Impact.

Uniquely powerful approaches to tackling the toughest diseases

Corporate Overview

www.kezarlifesciences.com

Forward-Looking Statements Disclaimer

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,” “plan,” “anticipate,” “target,” and similar expressions (as well as other words or expressions referencing future events or circumstances) are intended to identify forward-looking statements. Each of these forward-looking statements involves substantial risks and uncertainties that could cause actual results to differ significantly from those expressed or implied by such forward-looking statements. Forward-looking statements contained in this presentation include, but are not limited to, statements regarding the disruption of our business and clinical trials from the global outbreak of a novel strain of coronavirus (COVID-19), the potential use of our product candidates to treat patients, the association of data with treatment outcomes, the design, timing of initiation, progress, enrollment and scope of clinical trials for our product candidates, the expected timing of program updates and data disclosures, the timing of filing INDs and other regulatory documents, the timing and likelihood of seeking regulatory approval for our product candidates, and the patient prevalence, regulatory pathway and competitive landscape for our product candidates.

These forward-looking statements reflect Kezar's current beliefs and expectations. Many factors may cause differences between current expectations and actual results, including unexpected safety or efficacy data observed during preclinical or clinical studies, clinical site activation rates or clinical trial enrollment rates that are lower than expected, changes in expected or existing competition, changes in the regulatory environment, and unexpected litigation or other disputes. Other factors that may cause our actual results to differ from current expectations are discussed in Kezar's most recent Form 10-K or Form 10-Q filed with the U.S. Securities and Exchange Commission (SEC), under the caption “Risk Factors” and elsewhere in such reports. Except as required by law, we assume 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: Harnessing Master Regulators of Cellular Function to Tackle Immune-mediated Diseases and Cancer

2 distinct scientific programs with potential across multiple indications of high unmet need



Rich Platform & Growing Pipeline



KZR-616:
First-in-Class Immunoproteasome Inhibitor

A novel approach to harmonizing the immune system; Potential to be a pipeline in a drug

First in class agent with broad anti-tumor activity; Potential to inhibit multiple targets with a single small molecule



KZR-261:
First candidate from Protein Secretion Platform



Strong Financial Position
(as of 9/30/2020)



\$150M cash, cash equivalents, and marketable securities; 46.3M common shares outstanding

Our Programs Employ Uniquely Powerful Approaches to Address a Diverse Pipeline of Indications

KZR-616 and KZR-261 are first-in-class small molecules that harness master regulators of cellular function to inhibit multiple drivers of disease via single targets

COMPOUND	THERAPEUTIC INDICATION	DEVELOPMENT STAGE				
		DISCOVERY	PRECLINICAL	PHASE 1	PHASE 2	PHASE 3

Selective Immunoproteasome Inhibition

KZR-616	Lupus Nephritis (LN)	 MISSION					
	Dermatomyositis (DM) / Polymyositis (PM)	 PRESIDIO					

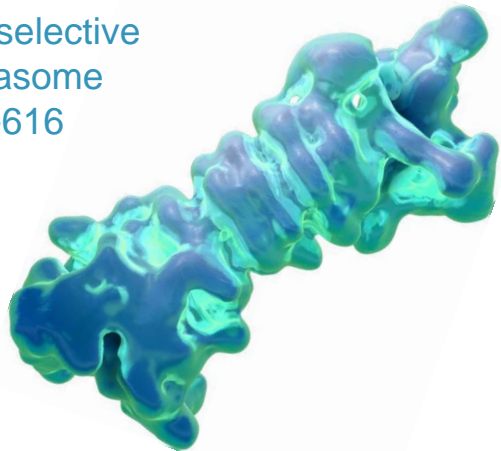
Protein Secretion Inhibition

KZR-261	Oncology	IND-enabling activities					
KZR-TBD	Oncology & Autoimmunity						

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

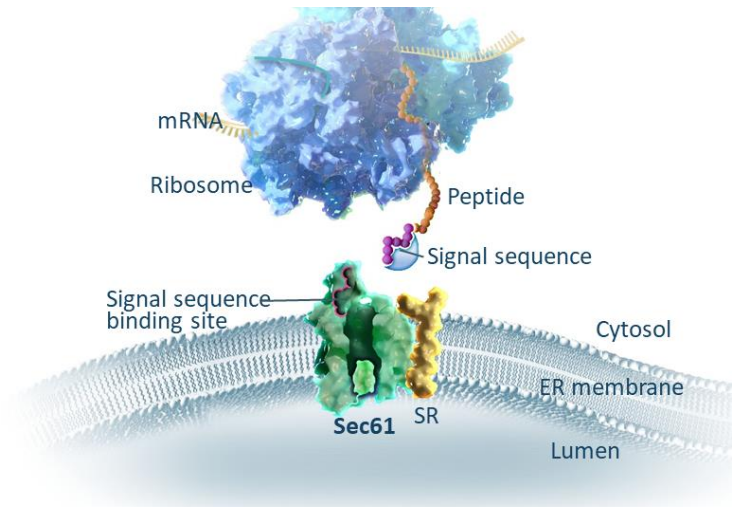
PROTEIN DEGRADATION: The Immunoproteasome

The target of selective immunoproteasome inhibitor KZR-616



- Modulates multiple drivers of inflammation
- Active in broad array of autoimmune disease models
- Restores normal immune responses in autoimmune disorders, while potentially avoiding immunosuppression

PROTEIN SECRETION: The Sec61 Translocon



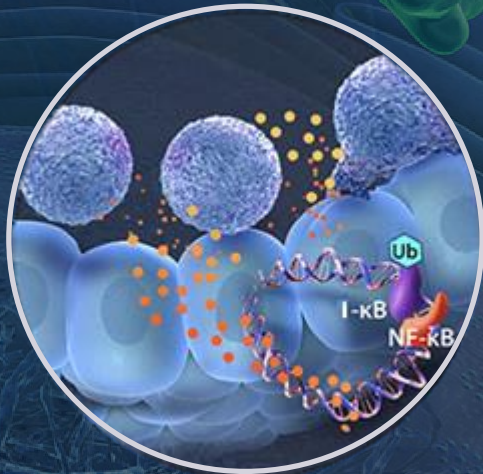
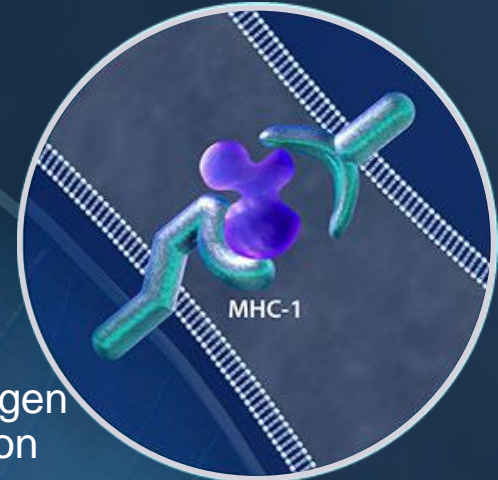
- Broad anti-tumor activity in preclinical models
- Applications in oncology, immuno-oncology, and autoimmunity
- Potential for small molecules to replace certain biologics

The Immunoproteasome Acts as a Master Regulator of Cellular Function in the Body's Immune System – its Dysfunction Underlies the Pathogenesis of Immune-mediated Diseases

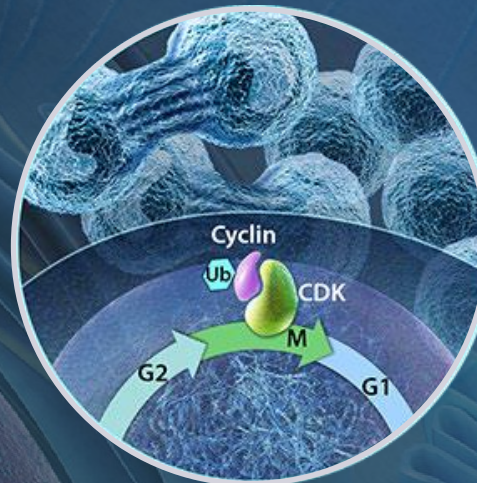
Immunoproteasome

Proteins to be degraded

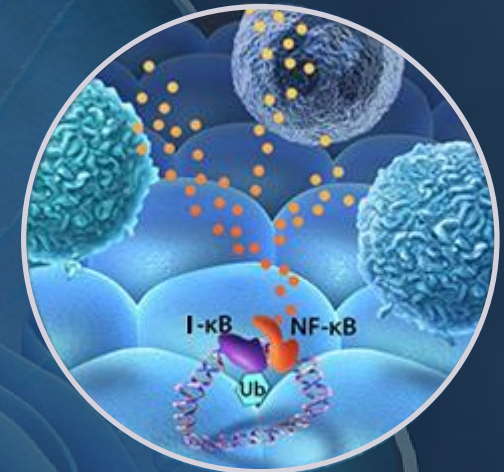
Class 1 Antigen Presentation



Lymphocyte migration & adhesion



Lymphocyte proliferation



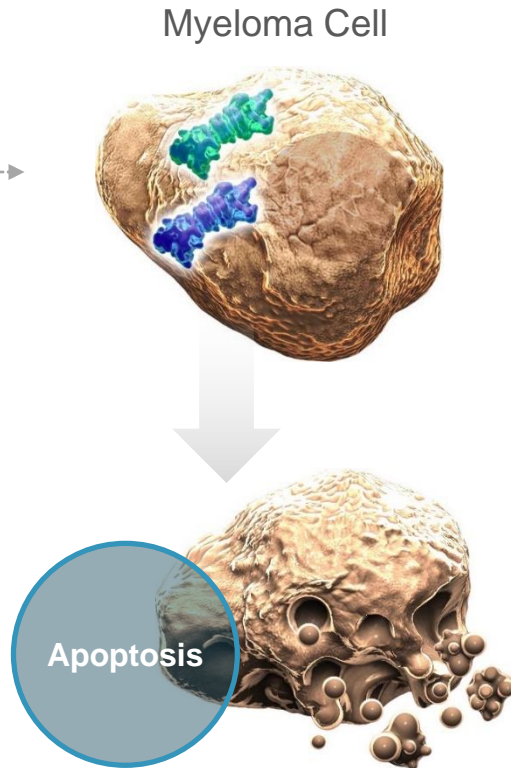
Cytokine expression

Selective Immunoproteasome Inhibition is not Cytotoxic and Results in Broad Immunomodulatory Activity

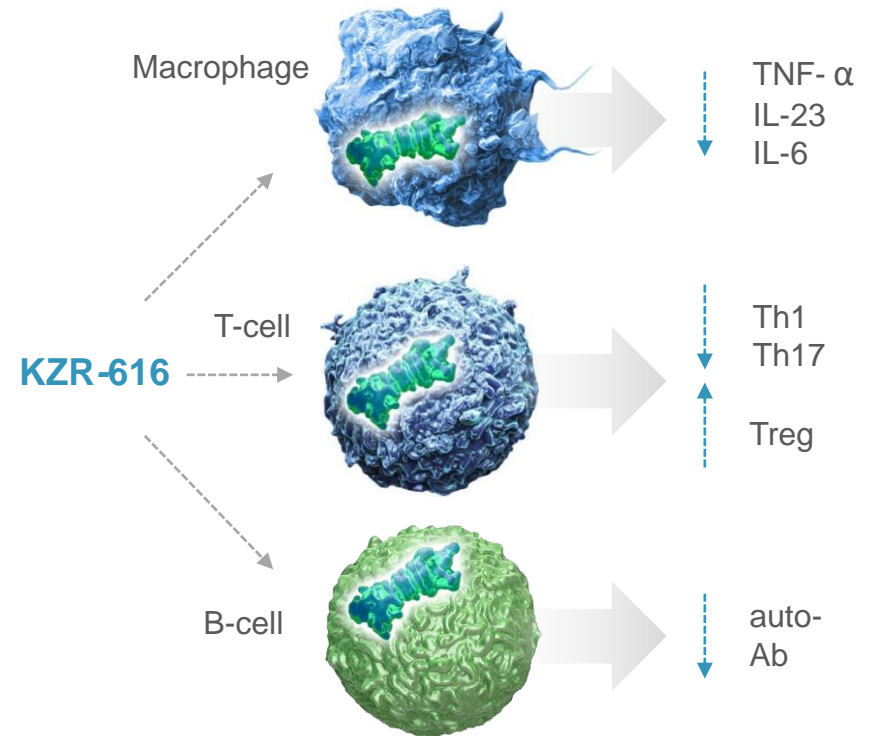
Dual-Targeting Proteasome Inhibitors

Dual proteasome inhibitors

- Dual inhibition required for cell death
- Death induced by protein buildup (UPR)



Selective Immunoproteasome Inhibitors

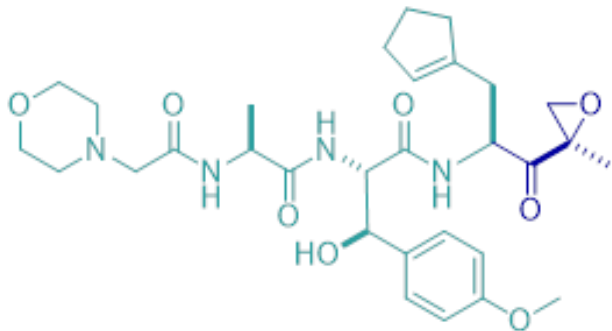


Parlati et al. Blood 2009

KZR-616, A First-in-Class Selective Immunoproteasome Inhibitor

Offers a Novel Approach for Harmonizing the Immune System for Immense Disease-Modifying Benefit

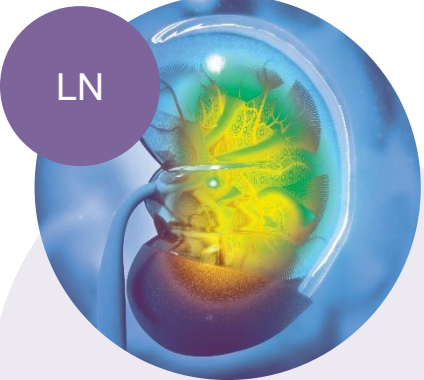
KZR-616



Strong Pharmaceutical Properties Exhibited in Studies to Date

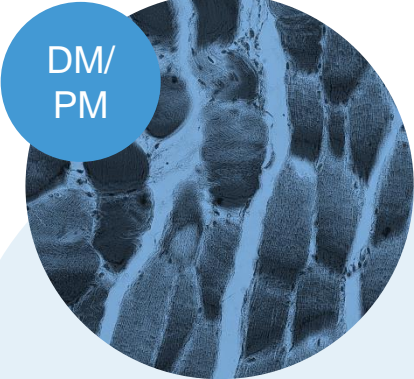
- Ideal clinical pharmacology parameters
- Low DDI risk
- 100% bioavailability with subcutaneous administration
- Amenable to patient self-administration
- Experienced CMC team prepared for commercial grade manufacturing with low COGS
- Extensive IP coverage and lifespan (2034+)

We are Leveraging KZR-616's Broad Immunomodulatory Potential to Initially Address Severe, Chronic Autoimmune Diseases



LN

Prevalence: ~150K US patients
Treatment: No approved treatments in US/EU



DM/PM

Prevalence: ~70K US patients
Treatment: Limited approved treatments in US/EU

CURRENT TREATMENT PARADIGMS ARE INADEQUATE

- Prolonged corticosteroid use results in significant complications
- Existing therapies are ineffective in many patients
- Targeted therapies (e.g., biologics) may not address needs of all patients with diseases characterized by defects in multiple arms of the immune system

Almani S et al. CJASN 2017
Oddis C et al. Nat Rev Rheumatol 2018
Barcellini W et al. Expert Rev Clin Immunol 2018

Lambert MP et al. Blood 2017
<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4386579/figure/Fig4>

Data From 130+ Healthy Volunteers and SLE Patients* Receiving KZR-616 Support Advancement into Ph2 Trials in Chronic Autoimmune Diseases



Safety & Tolerability

- Well-tolerated for 13 weeks of treatment
- Safety profile does not indicate need for patient monitoring



Efficacy & Biomarkers

- Improvement across all measured parameters of disease activity
- Rapid and sustained immunomodulatory gene expression changes
- Reduction in anti-dsDNA antibody titers in 7/7 patients with elevated levels at BL
- Rapid reduction in UPCR in 2/2 patients with active, proliferative LN

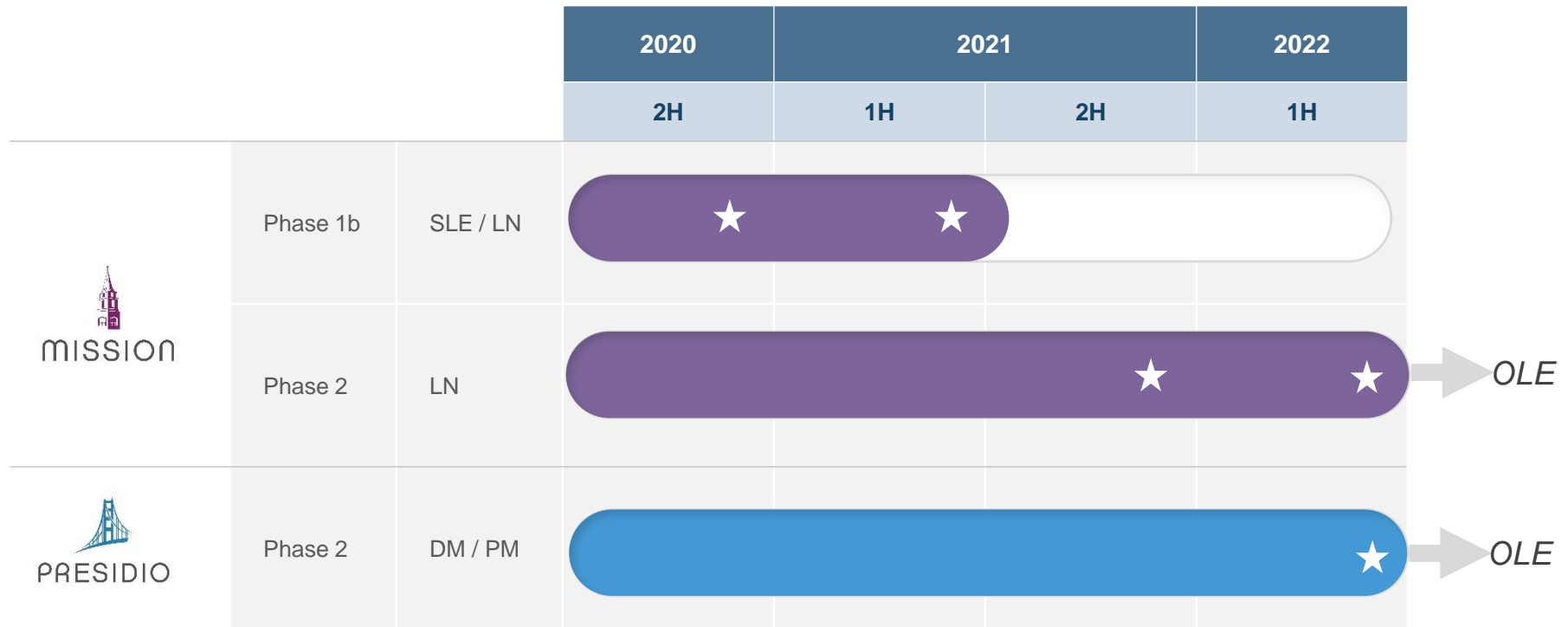


PK & PD

- Consistent PK and PD across subjects and with repeat dosing
- Target levels of immunoproteasome inhibition at doses $\geq 30\text{mg}$

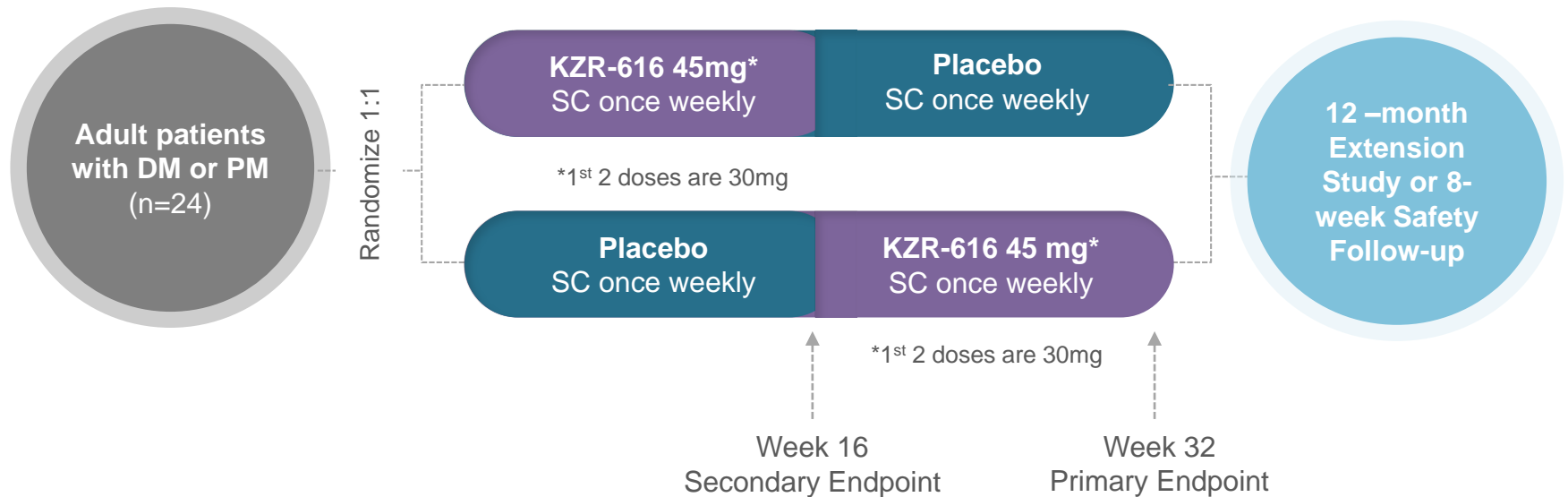
**data as of Sept 24, 2020*

KZR-616 is Now Being Evaluated Phase 2 Clinical Trials for the treatment of Chronic, Severe Autoimmune Diseases



★ Anticipated data updates

PRESIDIO Phase 2 Placebo-Controlled Cross-over Study for the Treatment of Dermatomyositis and Polymyositis Designed to Inform Late Stage Studies



ENDPOINTS

1°: Efficacy: Total Improvement Score (TIS)

2°: Safety and tolerability; Patient Reported Outcomes (PROs), PK

Exploratory: Biomarkers, PK/PD relationship

Study NCT04033926

KZR-616 Improved Muscle Function in a Mouse Model of Polymyositis and Dermatomyositis*

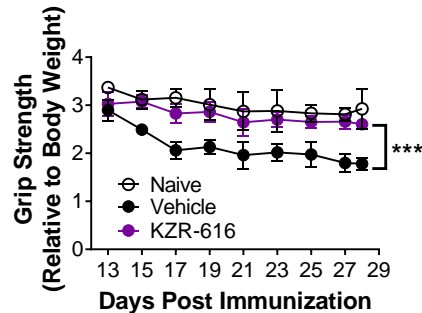
CIM MODEL

- Gold standard model for PM and DM (Sugihara 2007)
- Replicates multiple features of clinical disease



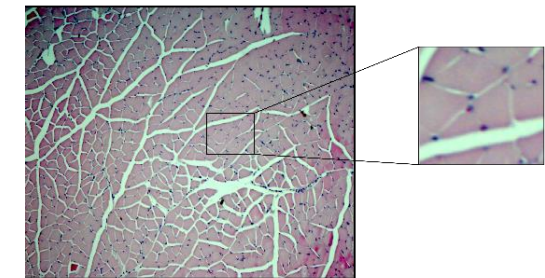
- KZR-616 treatment of diseased animals restored normal muscle function
- Significant reduction in tissue damage (histology and circulating enzyme levels)

Muscle Strength



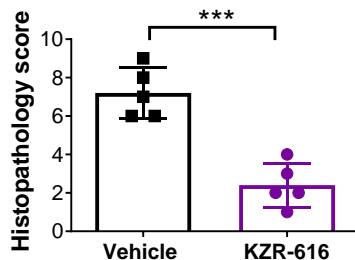
Triceps Histology

(H&E Staining)

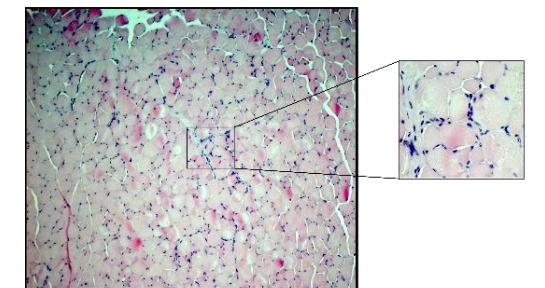
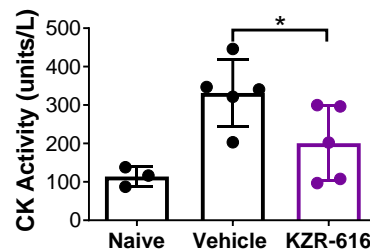


KZR-616

Muscle Histology



Muscle Enzymes



Vehicle

MISSION Phase 2 Trial to Demonstrate Responder Rate of KZR-616 60 mg Weekly and Inform Late Stage Studies in LN



KEY INCLUSION CRITERIA:

Biopsy-proven Class III or IV +/- Class V LN w/significant proteinuria (UPCR ≥ 1) despite standard therapy

Must be on stable therapy for at least 8 weeks prior to study entry

ENDPOINTS

1°: Efficacy
Number of patients with $\geq 50\%$ reduction in UPCR

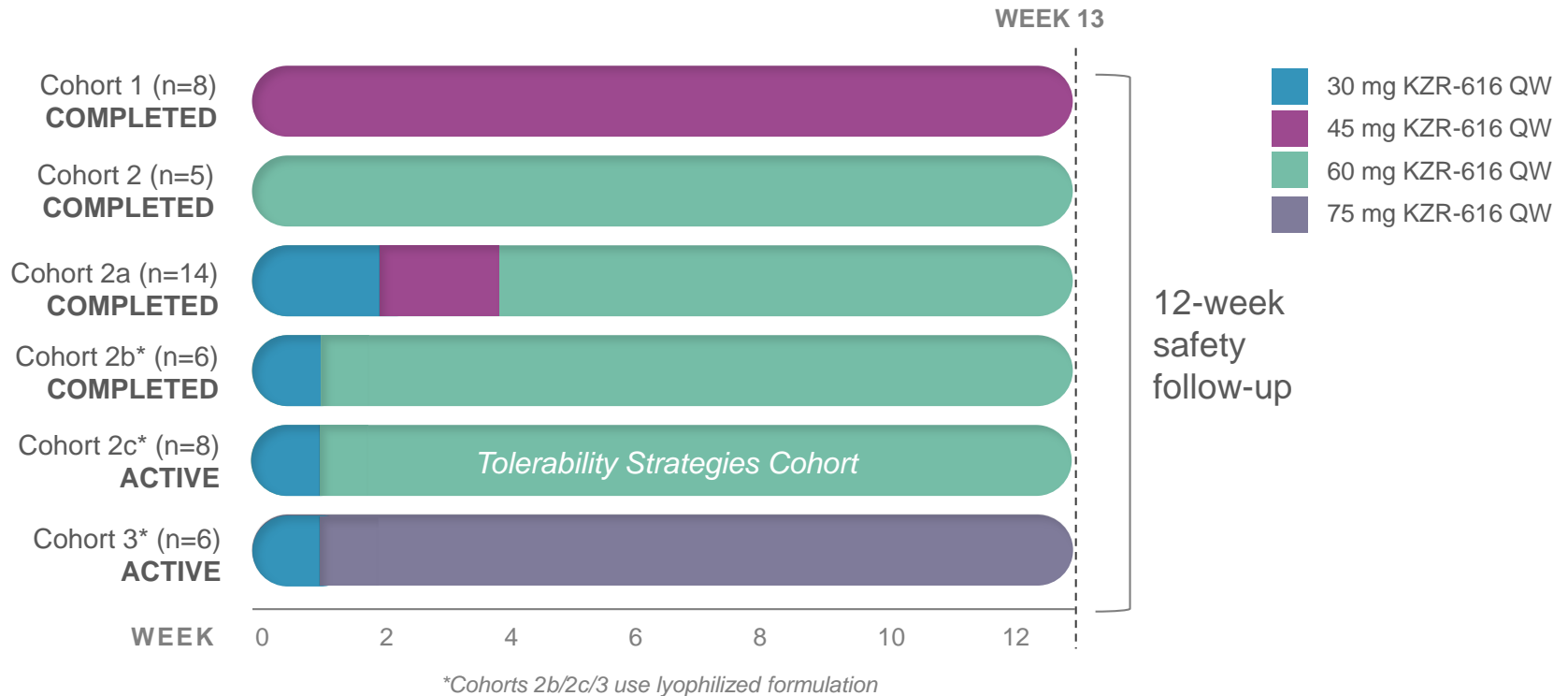
2°:
Safety and tolerability;
Additional renal response parameters (e.g. CRR);
Extra-renal SLE disease indices;
PROs

Exploratory:
Biomarkers

NCT03393013

MISSION Phase 1b is Designed to Evaluate Safety, Tolerability, and Early Efficacy Signals

Enrollment Complete



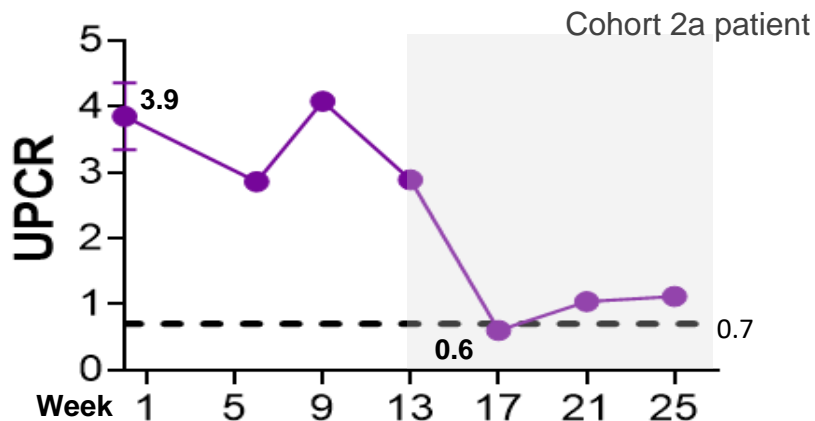
ENDPOINTS

1°: Safety

2°: Recommended Phase 2 doses, Plasma PK

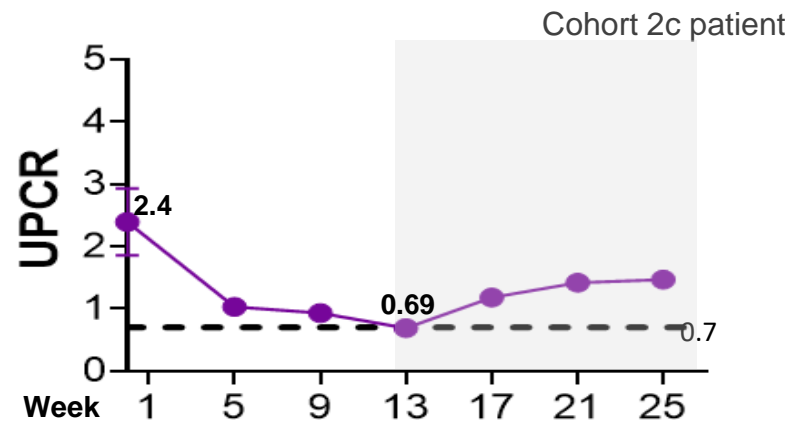
Exploratory:
Efficacy, PD, Biomarkers,
Pharmacogenomics

Results Observed in 2 of 2 LN Patients Treated in the MISSION Phase 1b Showed Rapid Improvement in Renal Function



	<u>SLEDAI-2K</u>	<u>anti-dsDNA(IU/ml)</u>
Baseline	17	137
Week 13	8	53
Week 17	8	73
Week 25	8	61

- Baseline stable treatment regimen of leflunomide, hydroxychloroquine, and prednisone (10mg/day); failed prior tacrolimus
- Nephrotic range
- >50% reduction in UPCR at week 17
- Reduced anti-dsDNA at week 13
- Drug holiday due to AE W2-4 & W11



	<u>SLEDAI-2K</u>	<u>anti-dsDNA(IU/ml)</u>
Baseline	14	123.5
Week 13	8	52
Week 17	8	40
Week 25	NA	53

- Baseline stable treatment regimen of MMF (2g), hydroxychloroquine, and prednisone (10mg/day) Nephrotic range
- >50% reduction in UPCR at week 5
- Improve symptom scores at week 5
- Reduced anti-dsDNA at week 5

KZR-616 Demonstrated Improvement on Exploratory Efficacy Measures of Disease Activity Across Organ Systems in SLE

Tool	Improvement	Mean Patient Score*		
		Baseline (n=33) ^a	EOT -W13 (n=32) ^{ab}	EOS -W25 (n=27) ^a
SLEDAI-2K	+	9.2	6.7	7.2
CLASI-A	+	5.8	2.9	3.4
Tender Joint Count	+	11.2	5.0	5.6
Swollen Joint Count	+	7.7	2.5	2.1
Physician Global Assessment Score	+	57.0	40.4	39.6
Patient Pain Assessment	+	59.3	37.8	44.7
Patient Global Assessment Score	+	59.3	44.2	44.6

^aNot all completing patients were evaluable because data had not been entered at the time of the data cut; ^b SLEDAI-2K at W13 is n=31

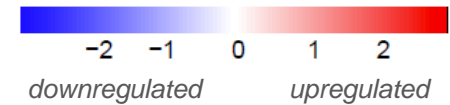
BL=Baseline; EOT=End of Treatment; EOS=End of Study; W13=week 13; W25=week 25; CLASI-A, Cutaneous Lupus Erythematosus Disease Area and Severity Index-Activity; SD, standard deviation; SLEDAI-2K, Systemic Lupus Erythematosus Disease Activity Index 2000.

**There are patients still active in cohorts 2c and 3 as of Sept 24, 2020*

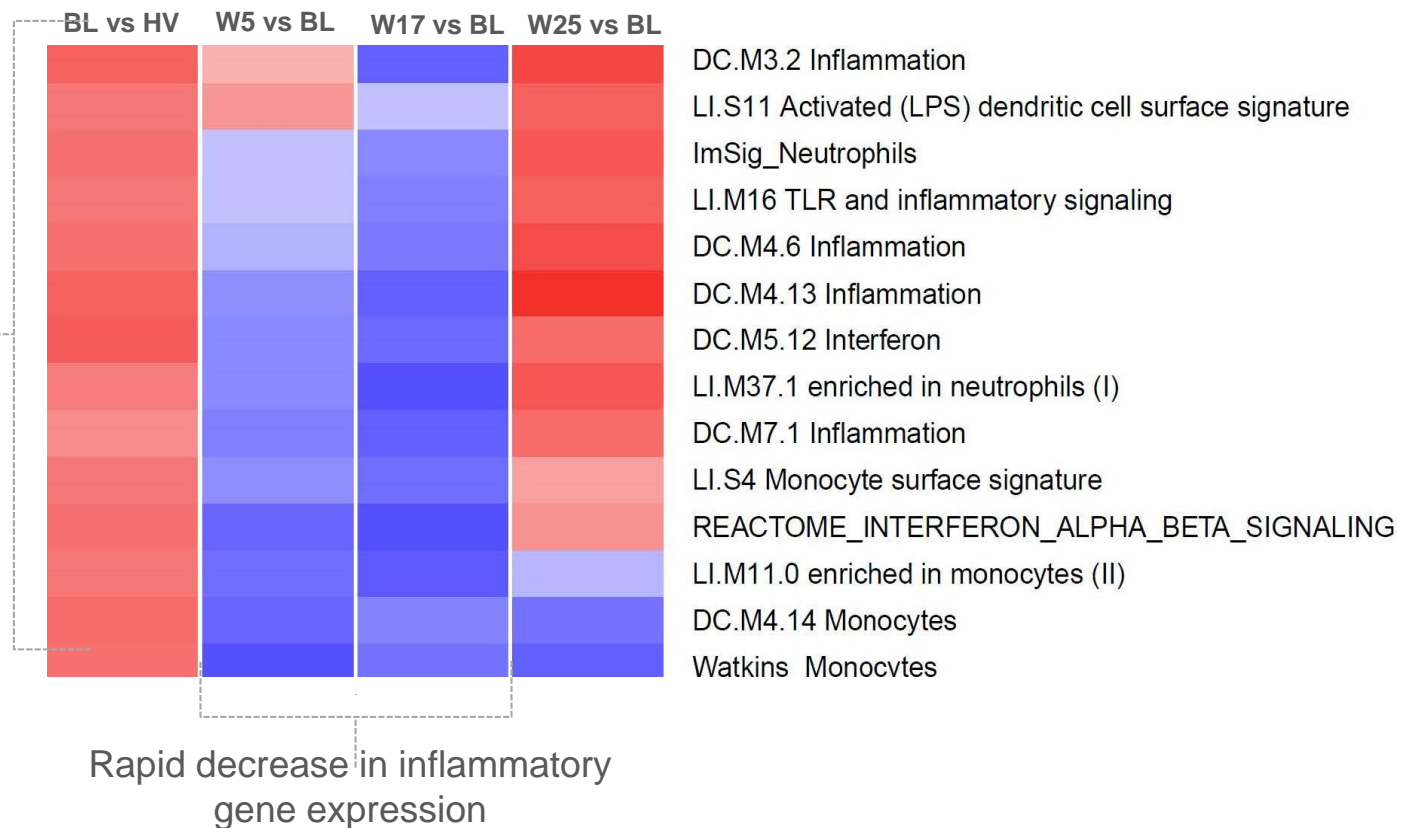
Whole Blood RNASeq Data from MISSION Phase 1b Patients Support Potential Broad Immunomodulatory Activity of KZR-616

GENE SIGNATURE COMPARISONS

4 wks post last dose 12 wks post last dose



Enriched inflammatory gene signatures in SLE pts (n=16)



All Patients with Elevated Anti-dsDNA Levels at Baseline (n=7) Experienced Reductions After Treatment with KZR-616

Patients Completing Study w/Elevated anti-dsDNA at BL*

Individual	Mean anti-dsDNA level, IU/mL (baseline)	% Change from baseline, week 13 (end of treatment)	% Change from baseline, week 25 (end of study)
Patient A	1015	-64.0	-82.0
Patient B ^a	87	-20.7	-33.3
Patient C	32	-6.3	-18.8
Patient D ^b	134	-60.4	-54.5
Patient E ^a	90	-76.7	-68.9
Patient F ^b	98	-46.9	-45.9
Patient G	29	-17.2	-24.1

^aHistory of nephritis. ^bActive nephritis.

Abbreviation: anti-dsDNA, anti-double-stranded DNA antibody

*Elevated levels of anti-dsDNA antibodies are highly specific markers of SLE disease activity (>20 IU/mL considered elevated)

KZR-616 Continues to Demonstrate a Favorable Safety and Tolerability Profile at Higher Doses

Measures, N% of patients	Cohort 2a ^a (n=14)	Lyophilized formulation		All patients* (Cohorts 1-3) (n=46)
		Cohort 2b (n=6)	Cohort 2c (n=8)	
At least 1 Treatment Emergent Adverse Event (TEAE)	12 (85.7)	4 (66.7)	7 (87.5)	38 (82.6)
Injection Site Erythema	5 (35.7)	2 (33.3)	6 (75.0)	20 (43.5)
Nausea	5 (35.7)	1 (16.7)	4 (50.0)	18 (39.1)
Vomiting	4 (28.6)	1 (16.7)	2 (25.0)	14 (30.4)
TEAEs ≥ Grade 3	3 (21.4)	0 (0.0)	0 (0.0)	4 (10.3)
Infectious TEAEs ≥ Grade 3	1 (7.1)	0 (0.0)	0 (0.0)	1 (2.2)
Infectious TEAEs; All Grades	5 (37.5)	2 (33.3)	2 (25.0)	10 (21.7)
Serious TEAEs	2 (14.3)	1 (16.7)	0 (0.0)	4 (8.7)
TEAEs leading to discontinuation	2 (14.3)	0 (0.0)	0 (0.0)	10 (21.7)

^aPatients received 4 doses to reach target dose. *All patients are inclusive of patients from Cohort 1. 47 patients were enrolled, but data for one patient were not entered at time of data cut.

Cohorts 2b, 2c and 3 utilize a lyophilized formulation of KZR-616, prophylactic oral electrolyte solution, non-sedating antihistamines, and antiemetics and/or dose escalation

****No patients in Cohort 3 had completed treatment as of Sept 24, 2020***

KZR-261, Our First Protein Secretion candidate, is Currently in IND-enabling Studies

COMPOUND	THERAPEUTIC INDICATION	DEVELOPMENT STAGE				
		DISCOVERY	PRECLINICAL	PHASE 1	PHASE 2	PHASE 3

Selective Immunoproteasome Inhibition

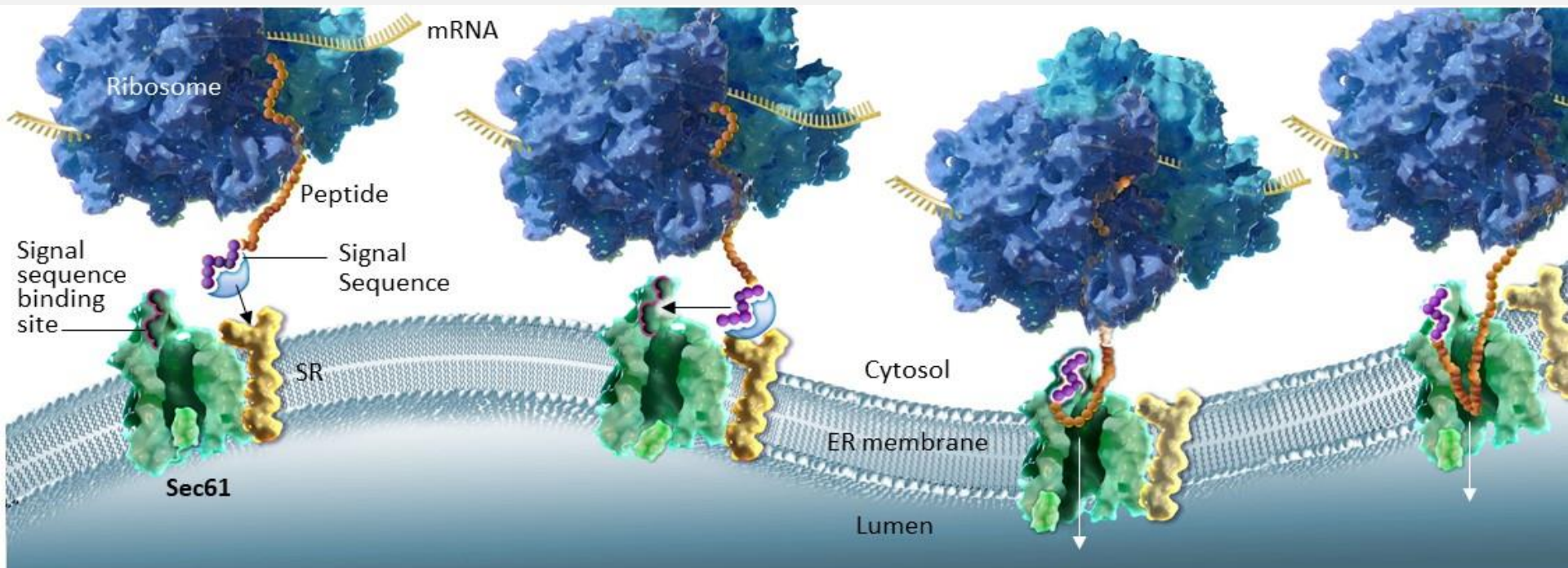
KZR-616	Lupus Nephritis (LN)	MISSION				
	Dermatomyositis (DM) / Polymyositis (PM)	PRESIDIO				

Protein Secretion Inhibition

KZR-261	Oncology	IND-enabling activities				
KZR-TBD	Oncology & Autoimmunity					

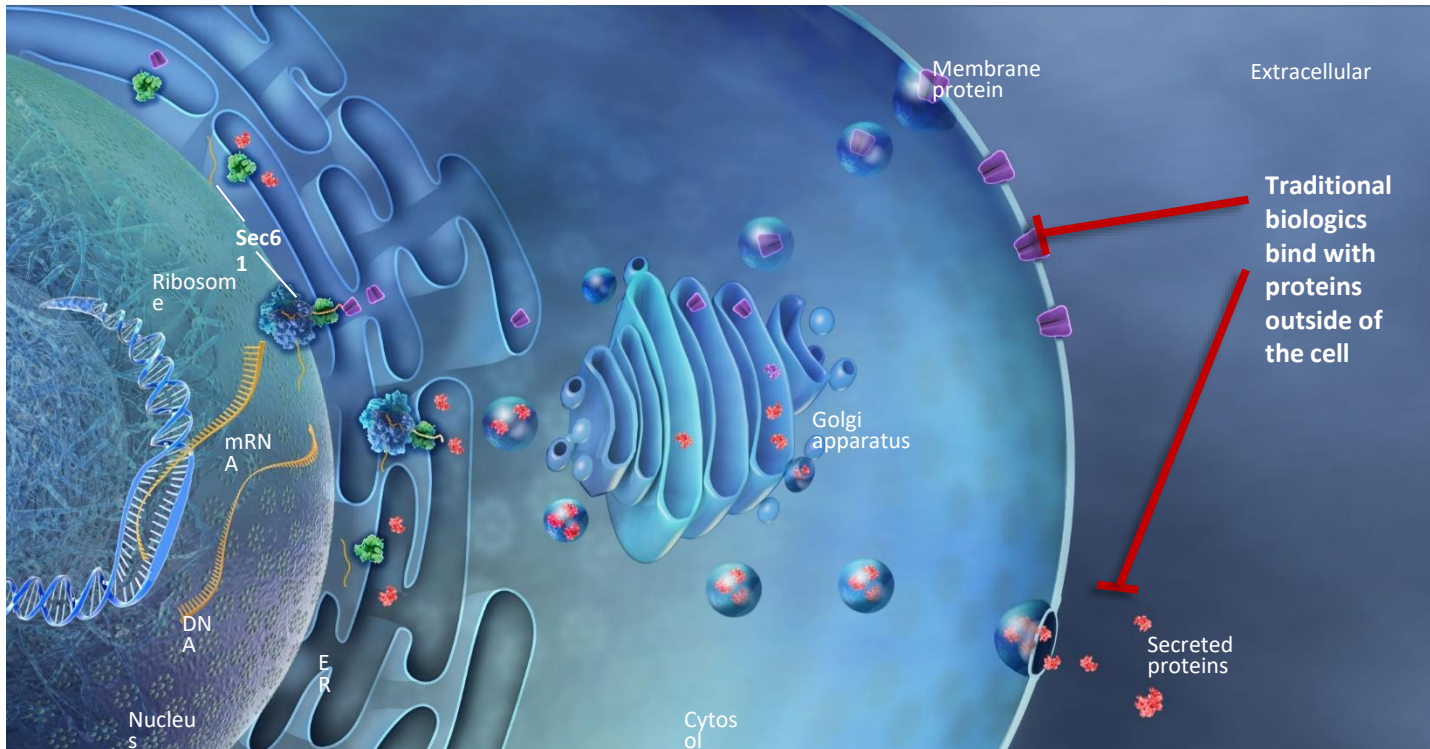
The Sec61 Translocon Represents Initiation of the Protein Secretion Pathway

- Highly conserved process
- Nearly all secreted and transmembrane proteins (5,000 – 7,000 proteins) utilize Sec61 to enter the endoplasmic reticulum (ER)
- Each protein expresses a unique signal sequence or transmembrane domain
- Kezar tool compounds block functional interaction of signal sequences and Sec61
- Blocking functional interaction of signal sequences inhibits the expression of a selected protein or proteins



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)

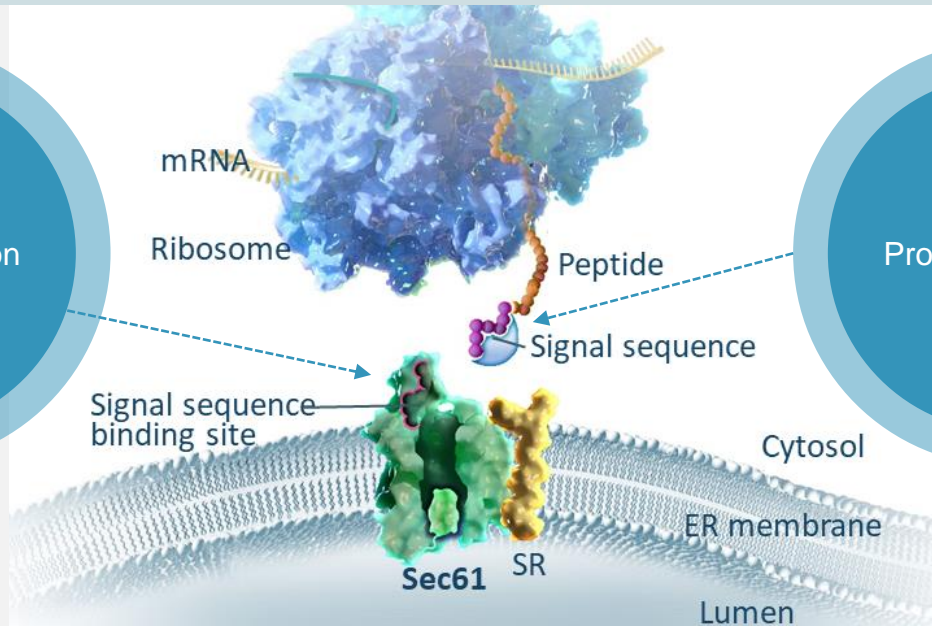
Secreted Proteins (partial list)

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

Kezar has Multiple Small Molecule Approaches to Modulating Protein Secretion at the Sec61 Translocon

Multiple Protein Secretion Inhibitors

- Potential for multiple clinical candidates
- Broad anti-tumor activity in vitro and in vivo
- KZR-261-first protein secretion clinical candidate; anticipated IND filing Q1 2021

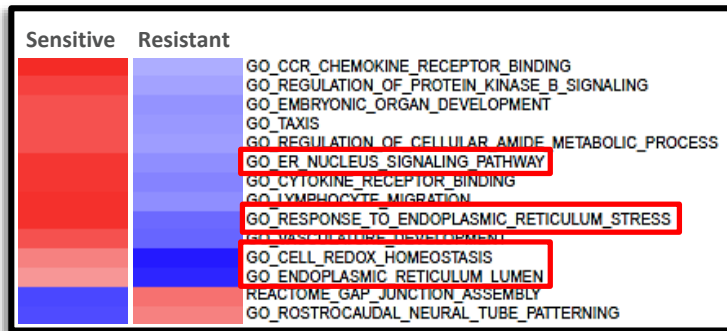


Specific Protein Secretion Inhibitors

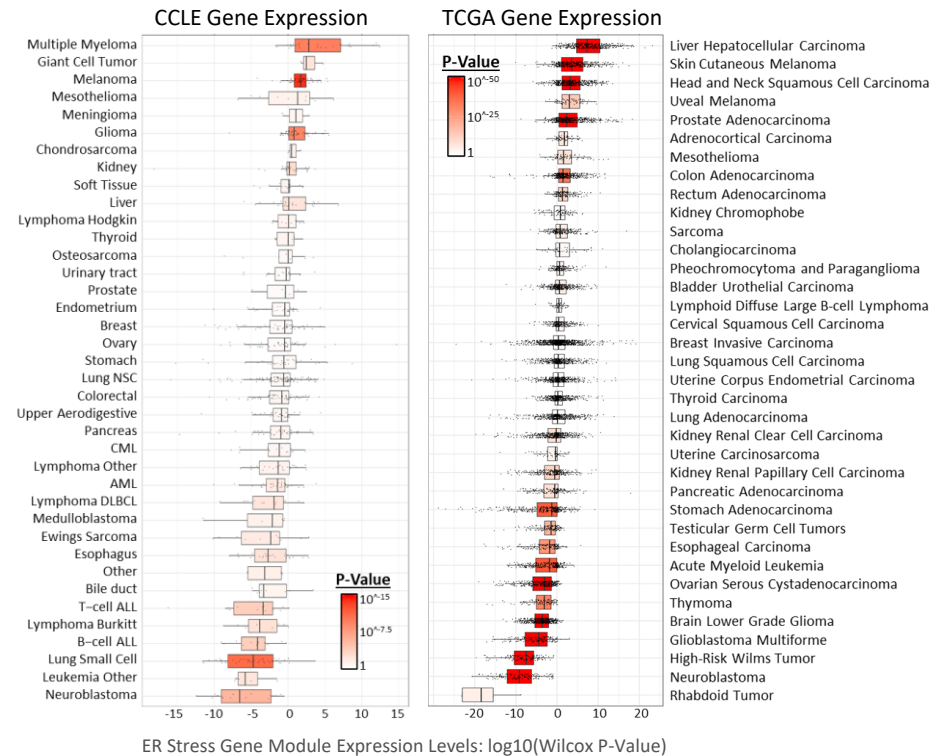
- Small molecule replacement of biologic therapies
- Target selective approach
- High value / validated targets
- Potential applications in oncology and autoimmunity

Potential to inhibit multiple therapeutically relevant targets with a single small molecule

Gene Expression Profiling Reveals Differential ER Stress Response: Potential Gene Signature Biomarker



- Sensitive and resistant myeloma cell lines were treated with a Sec61 inhibitor
- RNASeq analysis used to find gene modules that differed between the cell lines
- Sensitive cells: Increased expression of ER stress genes
- Enriched expression of ER stress genes in multiple solid tumor types



Positioned for Growth and Delivering Shareholder Value

Q4 2020

- Updated MISSION Phase 1b data (through Sept 2020)
- Initiate PRESIDIO Open-label Extension Study

1H 2021

- IND Submission for KZR-261
- Initiate Phase 1 basket study in solid tumors (KZR-261)
- Final Data from MISSION Phase 1b study (KZR-616 in SLE)
- Initiate MISSION Phase 2 Open-label extension study (KZR-616 in LN)

2H 2021

- Interim data from MISSION Phase 2 study (KZR-616 in LN)

1H 2022

- Top-line data from MISSION Phase 2 study (KZR-616 in LN)
- Top-line data from PRESIDIO Phase 2 study (KZR-616 in DM/PM)

The Kezar Opportunity: Harnessing Master Regulators of Cellular Function to Tackle Immune-mediated Diseases and Cancer

2 distinct scientific programs with potential across multiple indications of high unmet need



Rich Platform & Growing Pipeline



KZR-616:
First-in-Class
Immunoproteasome
Inhibitor

A novel approach to harmonizing the immune system; Potential to be a pipeline in a drug

First in class agent with broad anti-tumor activity; Potential to inhibit multiple targets with a single small molecule



KZR-261:
First candidate from
Protein Secretion
Platform



Strong Financial
Position
(as of 9/30/2020)

\$150M cash, cash equivalents, and marketable securities; 46.3M common shares outstanding



THANK YOU