



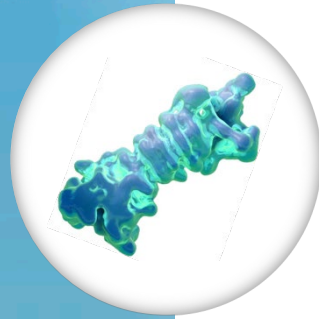
H.C. Wainwright 25th Annual Global Investment Conference

September 13th, 2023

Forward-Looking Statements and Topline Data Disclaimer

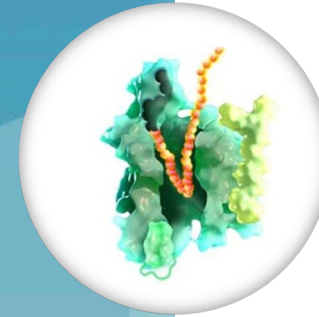
This presentation contains forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995. Words such as “may,” “will,” “should,” “expect,” “believe”, “plan” and similar expressions (as well as other words or expressions referencing future events, conditions or 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 initiation and timing of future clinical trials, the likelihood that data will support future development and therapeutic potential, the association of data with treatment outcomes, the anticipated therapeutic benefit and the likelihood of obtaining regulatory approval of Kezar’s product candidates. Many factors may cause differences between current expectations and actual results, including unexpected safety or efficacy data observed during preclinical and clinical studies, changes in expected or existing competition, lower than expected clinical trial enrollment rates, the uncertainty and timing of regulatory interactions and processes, financial audit and review procedures, 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.

Pursuing Paradigm Shifts in Immunology and Oncology



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

- Harmonizing the immune system via immunomodulation
- Potential “pipeline in a drug”
- Successfully completed MISSION Phase 2 study in lupus nephritis



KZR-261: First Candidate from Our Protein Secretion Platform

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



Strong Financial Position

- \$236.6M cash, cash equivalents and marketable securities as of June 30, 2023; 72.5M common shares outstanding

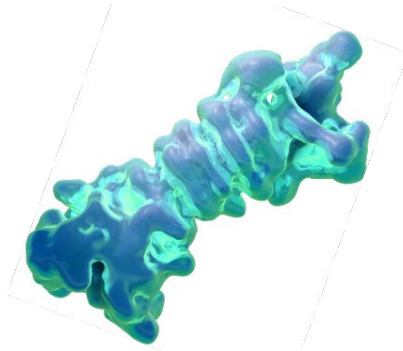
Targeting Master Regulators of Cellular Function to Treat a Range of Chronic Conditions

Kezar's Two Unique, Protein-Targeting Approaches

1

Selective Immunoproteasome Inhibition

Immunoproteasome



Targeting a range of autoimmune diseases through immune modulation versus direct immunosuppression

Clinical Programs

Lupus Nephritis

Autoimmune Hepatitis

2

Protein Secretion Inhibition

The Sec61 Translocon



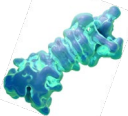
Halting the secretion of target proteins necessary for the proliferation of diseases spanning oncology and autoimmunity

Clinical Program

Solid Tumors

Building a First-In-Class Therapeutic Portfolio: “Pipeline in a Drug” Candidates with Multiple Shots on Goal, Supported by Novel Discovery Platform

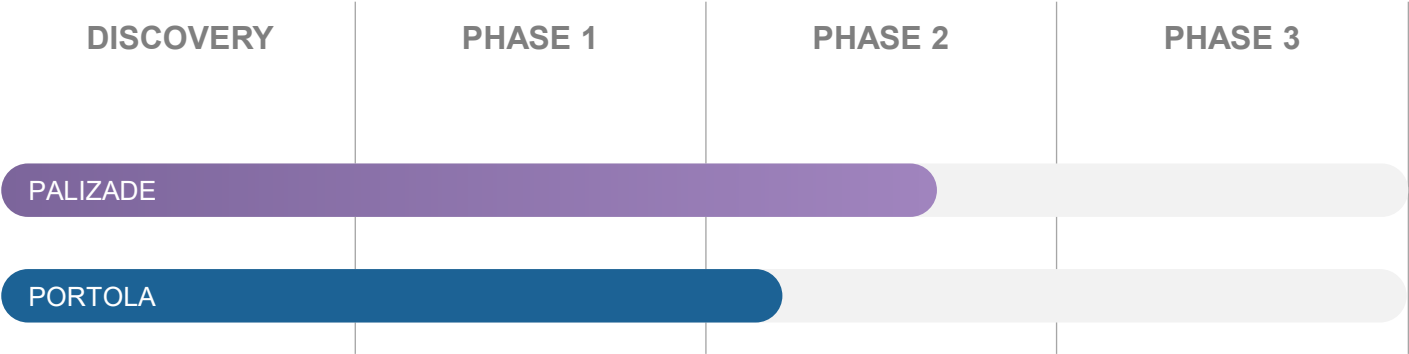
Selective Immunoproteasome Inhibition



Zetomipzomib

Lupus Nephritis (LN)

Autoimmune Hepatitis (AIH)



Protein Secretion Inhibition



KZR-261

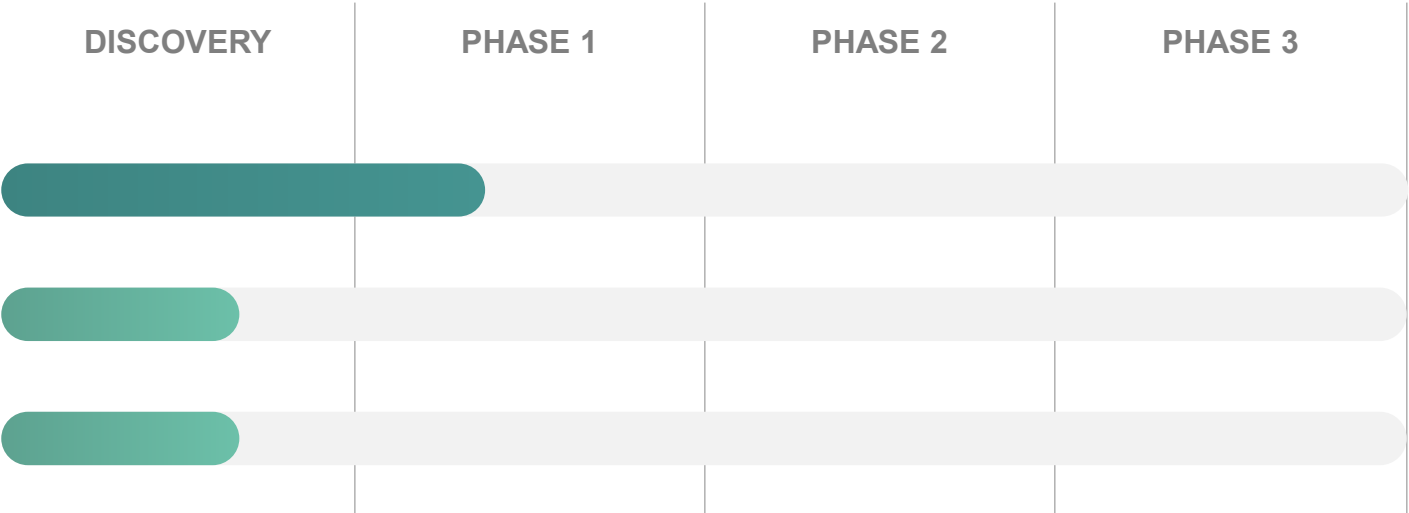
Advanced/Metastatic Solid Tumor

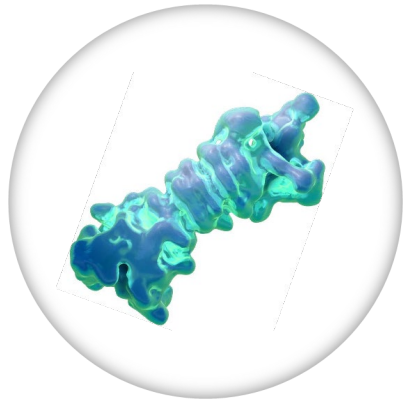
Oral Anti-PD-1

Single Target Oncology

KZR-TBD

Multi Target Oncology



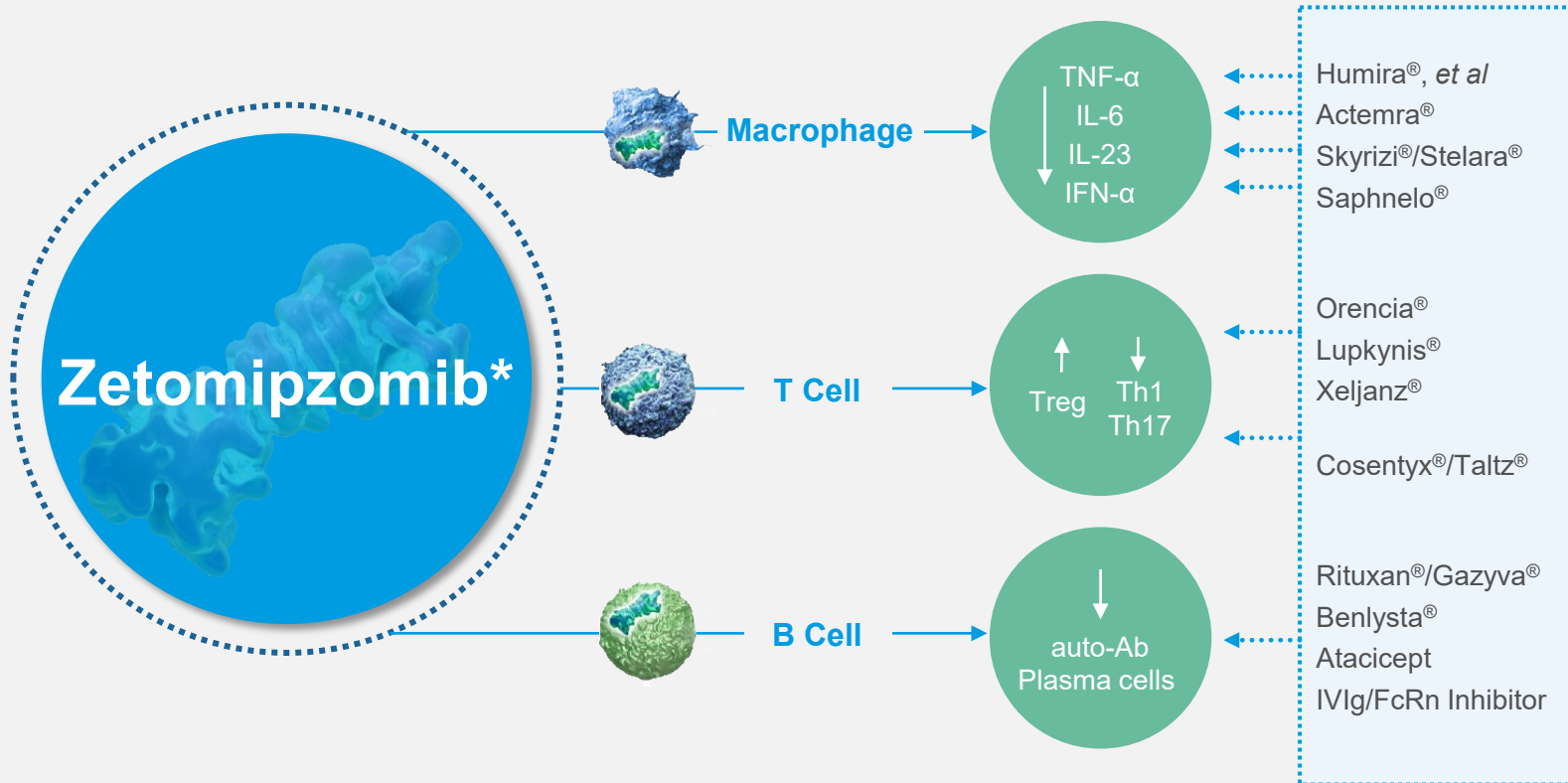


SELECTIVE
IMMUNOPROTEASOME
INHIBITION:

Zetomipzomib

Targeting a Range of Autoimmune
Diseases Through Immune Modulation
Versus Direct Immunosuppression

Zetomipzomib's Competitive Advantage: Immunomodulation Across the Entire Immune System



Zetomipzomib Advantage

- Targeted inhibition of immunoproteasome in immune cells and site of inflammation
- Inhibits multiple drivers of inflammation
- Normal immune response mechanisms remain intact

*Some preclinical studies were conducted with ONX 0914, a first-generation selective immunoproteasome inhibitor.

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

Zetomipzomib Modulates Innate and Acquired Immune Responses Without Evidence of Immunosuppression to Date



- Selective inhibition of the immunoproteasome down regulates inflammation without immunosuppression
 - Once-weekly SC administration
 - No accumulation observed with repeat dosing
 - Consistent exposure and clearance (T1/2 <5 hours)
- No immediate rebound of signs/symptoms of disease activity observed upon discontinuation
- No clinically significant opportunistic or serious infections observed
- No clinically significant immune cell depletion observed
- Not predicted to result in clinically significant drug-drug interactions (DDI)
- No off-target effects observed to date
- No teratogenicity observed in nonclinical studies
- No serum monitoring required



ZETOMIPZOMIB: MISSION

Phase 2 Study Evaluating
Zetomipzomib in Lupus Nephritis

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

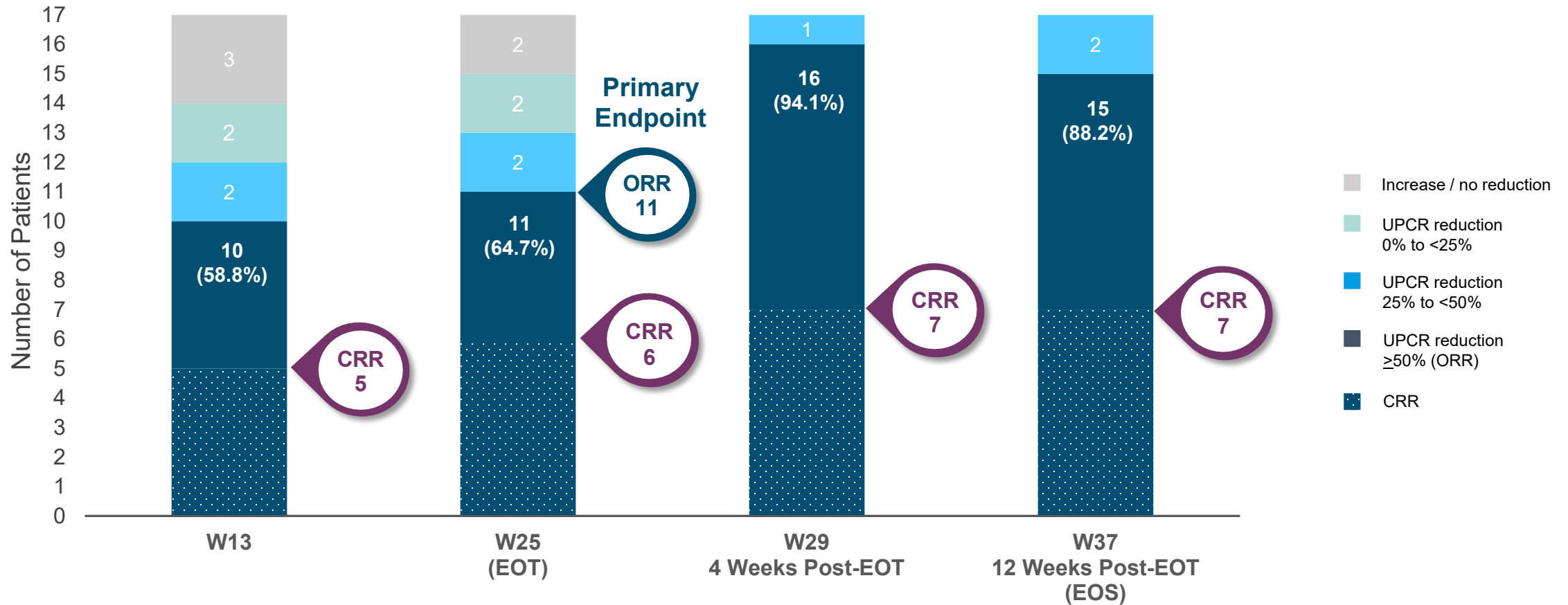


- Clinically meaningful ORR in 65% and CRR in 35% of patients at W25 (EOT)
- Renal response as early as W13 (ORR in 59% and CRR in 29%)
- Sustained renal response with additional ORRs/CRRs observed through W37
- Achievement of UPCR ≤ 0.5 in 65% of patients at W37 (EOS)
- Reduction of daily steroid dose to ≤ 10 mg/d in 82% of patients by W25 (EOT)
- Improvements in key SLE clinical disease activity scores and biomarkers
- Stable mean eGFR during the study
- Generally mild to moderate TEAEs (Grade 1/2)
- No evidence of immunosuppression (no serious/opportunistic infections or immune cell depletion)

Patients received 24 weeks of zetomipzomib; End-of-treatment assessments performed at Week 25.

Abbreviations: CRR, complete renal response; eGFR, estimated Glomerular Filtration Rate; EOS, end of study; EOT, end of treatment; LN, lupus nephritis; ORR, overall renal response; SLE, systemic lupus erythematosus; TEAEs, treatment emergent adverse events; UPCR, urine protein to creatinine ratio.

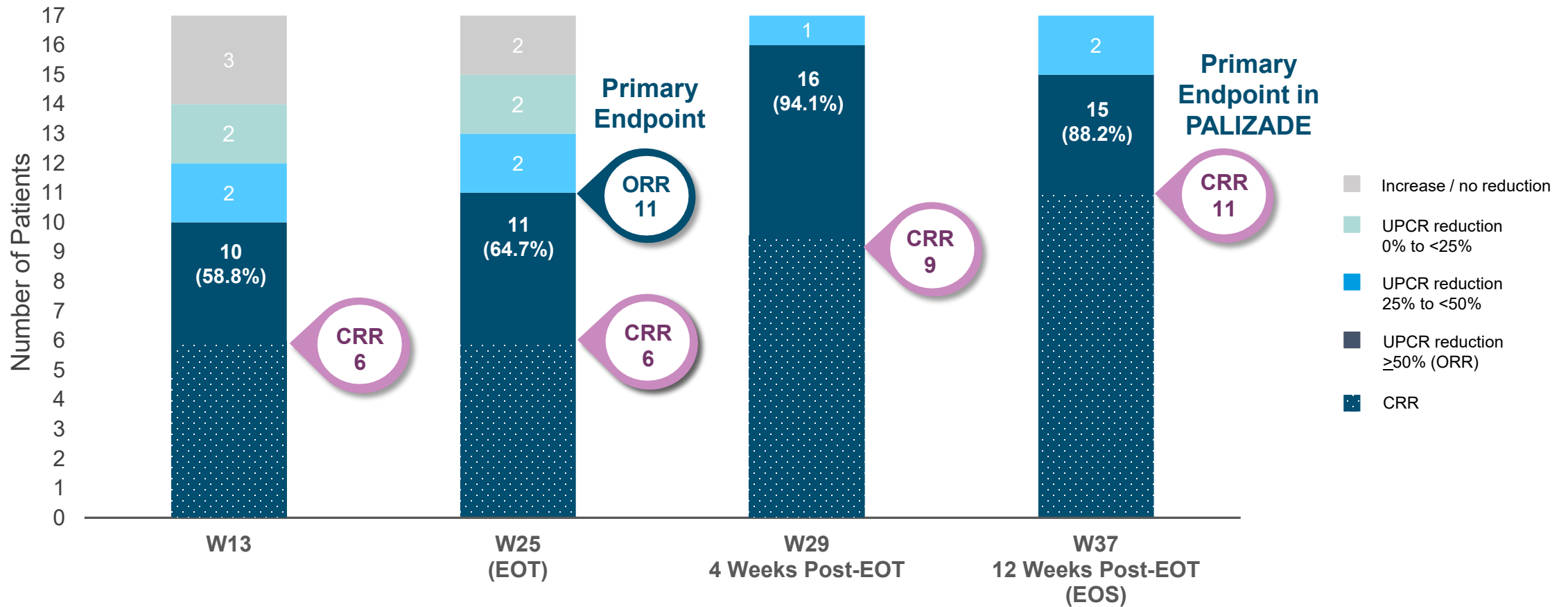
MISSION: Zetomipzomib Demonstrated Clinically Meaningful Renal Responses (Evaluable Population, n=17)



ORR: ≥50% reduction in UPCR compared to baseline; CRR: UPCR ≤0.5, eGFR ≥60 mL/min/1.73m² or no worsening of eGFR from baseline of ≥25%, prednisone (or equivalent) ≤10 mg and no use of prohibited medication; 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.

Abbreviations: CRR, complete renal response; eGFR, estimated Glomerular Filtration Rate; EOS, end of study, EOT, end of treatment; ORR, overall renal response; UPCR, urine protein to creatinine ratio.

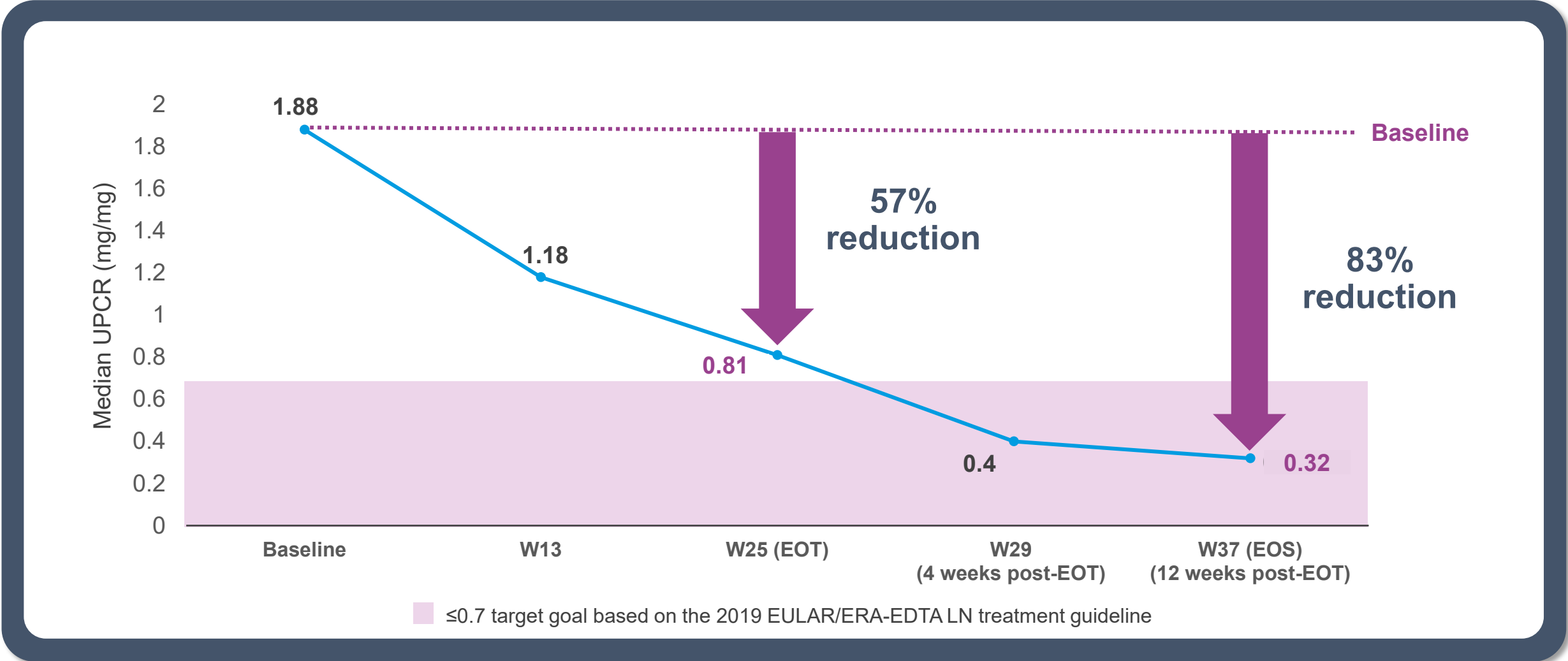
MISSION: Using PALIZADE CRR Criteria, Response Rates Further Improve



ORR: ≥50% reduction in UPCR compared to baseline; CRR: UPCR ≤0.5, eGFR ≥60 mL/min/1.73m² or no worsening of eGFR from baseline of ≥25%, prednisone (or equivalent) ≤10 mg and no use of prohibited medication; 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.

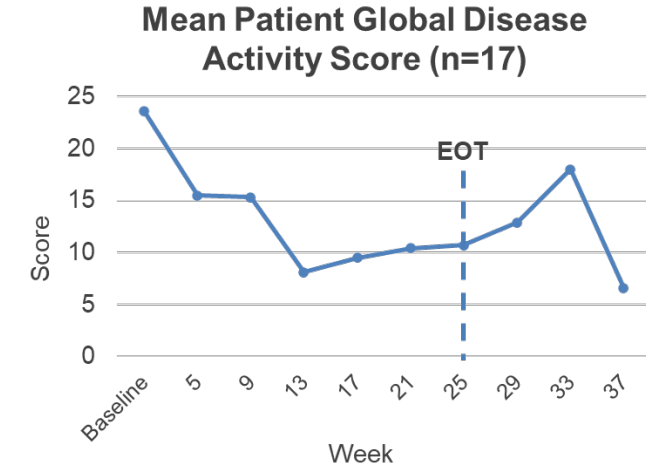
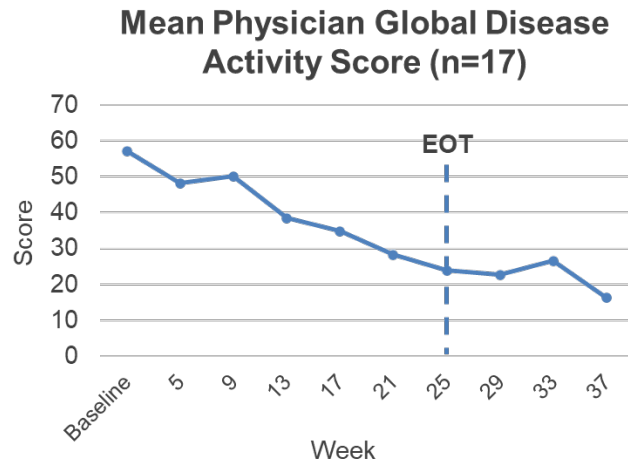
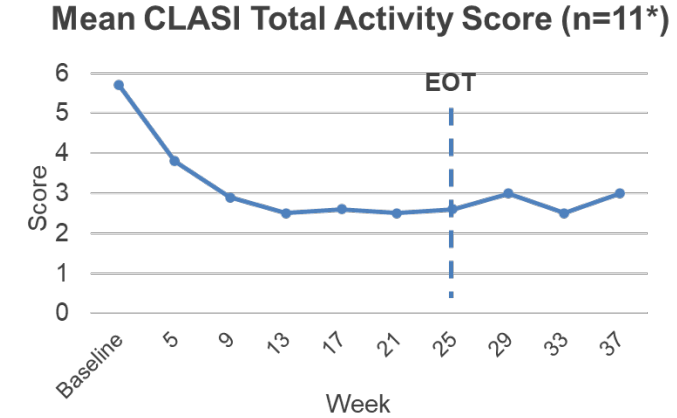
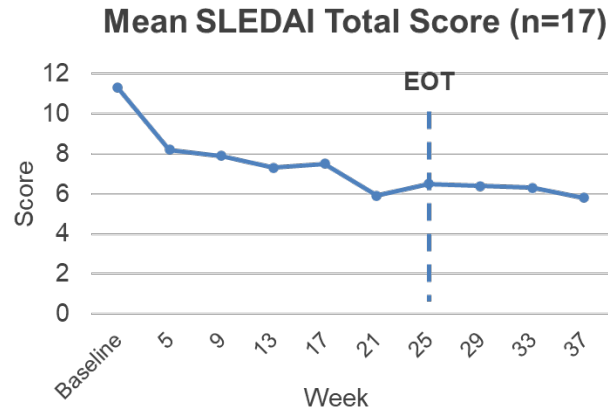
Abbreviations: CRR, complete renal response; eGFR, estimated Glomerular Filtration Rate; EOS, end of study, EOT, end of treatment; ORR, overall renal response; UPCR, urine protein to creatinine ratio.

MISSION: Continued Improvement in Median UPCR Observed With Zetomipzomib Treatment (Evaluable Population, n=17)



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.
Abbreviations: EOS, end of study; EOT, end of treatment; UPCR, urine protein to creatinine ratio.

MISSION: Zetomipzomib Improved Key SLE Clinical Disease Activity Scores (Evaluable Population, n=17)



*11 patients had active cutaneous SLE at baseline (CLASI-A >0). There were 5 patients with tender joint count >0 at baseline and 1 patient with swelling joint count >0 at baseline. 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

Abbreviations: CLASI, Cutaneous Lupus Erythematosus Severity Index-Activity; EOT, end of treatment; SLE, systemic lupus erythematosus; SLEDAI-2K, Systemic Lupus Erythematosus Disease Activity Index 2000.

MISSION: Zetomipzomib Demonstrated a Favorable Safety and Tolerability Profile (Safety Population, n=21)



Adverse Events	Zetomipzomib n (%)
Most Common TEAE: Injection-site Reaction	15 (71.4)
TEAE Leading to Study Drug Discontinuation[†]	4 (19.0)
Grade 3 TEAE	6 (28.6)
Serious TEAE[‡]	2 (9.5)
Grade ≥3 Infectious TEAE	0 (0)
Opportunistic Infections	0 (0)
Death	0 (0)

No Grade 4 TEAE was reported. [†]3 related TEAEs (injection site infiltration, asthenia, reticulocyte increase) and 1 unrelated serious TEAE (worsening pulmonary arterial hypertension [PAH] with acute kidney injury [AKI] and urinary tract infection [UTI]) led to study drug discontinuation. Patient subsequently had SAEs of AKI and UTI (unrelated) and has recovered. [‡]1 related serious TEAE of acute protracted migraine was reported. Study drug was temporarily interrupted, and patient has recovered and completed the study.

Abbreviation: TEAE, treatment emergent adverse event.

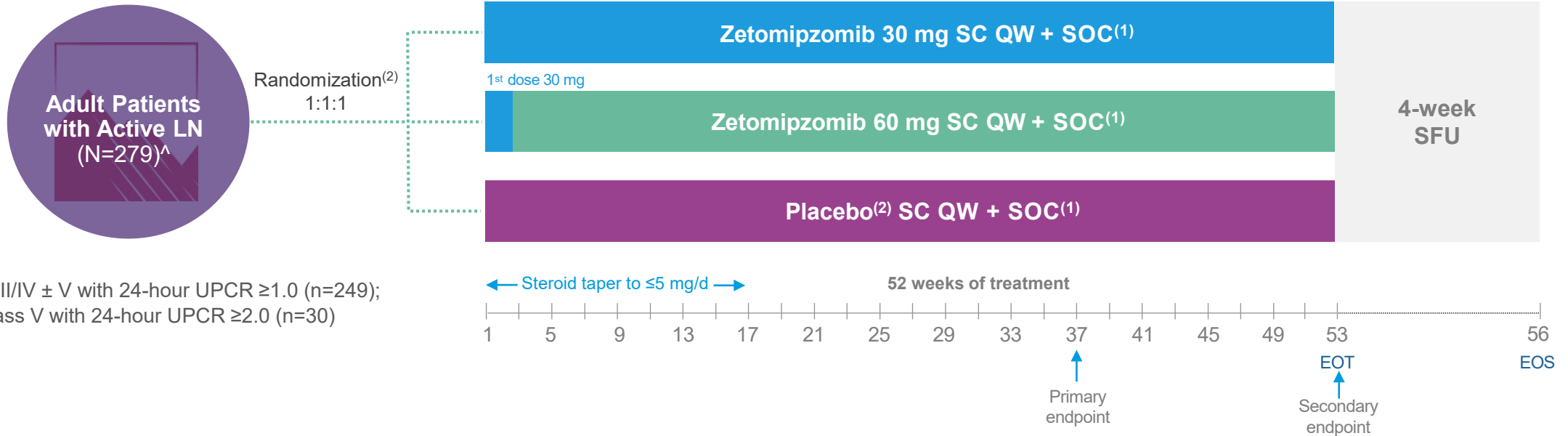


ZETOMIPZOMIB: PALIZADE

Phase 2b Study Evaluating
Zetomipzomib in Lupus Nephritis



PALIZADE: Phase 2b Placebo-Controlled Trial Evaluating the Efficacy and Safety of Zetomipzomib in Active Lupus Nephritis



[^]Class III/IV ± V with 24-hour UPCR ≥1.0 (n=249);
Class V with 24-hour UPCR ≥2.0 (n=30)

ENDPOINTS

Primary Efficacy Endpoint

- Proportion of patients achieving complete renal response (CRR⁽³⁾) at Week 37

Key Secondary Endpoints

- Proportion of patients achieving partial renal response (PRR⁽⁴⁾) at Week 37
- CRR at Weeks 25 and 53
- PRR at Weeks 25 and 53

Primary Safety Endpoint

- Severity of AEs

Other Endpoints

- SLE Disease Activity Measures (e.g. SLEDAI, 28-Joint Count, SLE Flare Index, PGA)

NCT05781750

1. MMF or equivalent (target dose 2 gm/d), oral corticosteroids (0.3-0.5 mg/kg/d, maximum 40 mg/d) and IV methylprednisolone (500 mg-1 gm, up to 3 gm, with opt-out for AE or lack of response).

2. Volume matched placebos with patients randomized 2:1 (zetomipzomib 30 mg: placebo) 2:1 (zetomipzomib 60 mg: placebo).

3. CRR: UPCR ≤0.5 and eGFR ≥60 mL/min/1.73 m² or no confirmed decrease of >20% from Baseline eGFR.

4. PRR: ≥50% reduction of UPCR from Baseline, and to <1.0 if the Baseline UPCR was <3.0 or to <3.0 if the Baseline value was ≥3.0.

Abbreviations: AE, adverse event; CRR, complete renal response; EOT, end of treatment; EOS, end of study; LN, lupus nephritis; PGA, Physician's Global Assessment; QW, once every week; SC, subcutaneous; SLE, Systemic Lupus Erythematosus; SLEDAI, Systemic Lupus Erythematosus Disease Activity Index; UPCR, urine protein to creatinine ratio; SFU, safety follow up.

Responder requirement: Should not have received >10 mg prednisone (or equivalent) for ≥3 consecutive days or for ≥7 days in total during the 8 weeks prior to a CRR assessment and no use of rescue or prohibited medication.



ZETOMIPZOMIB:
PORTOLA

Phase 2a Placebo-Controlled Study
Evaluating Zetomipzomib in AIH



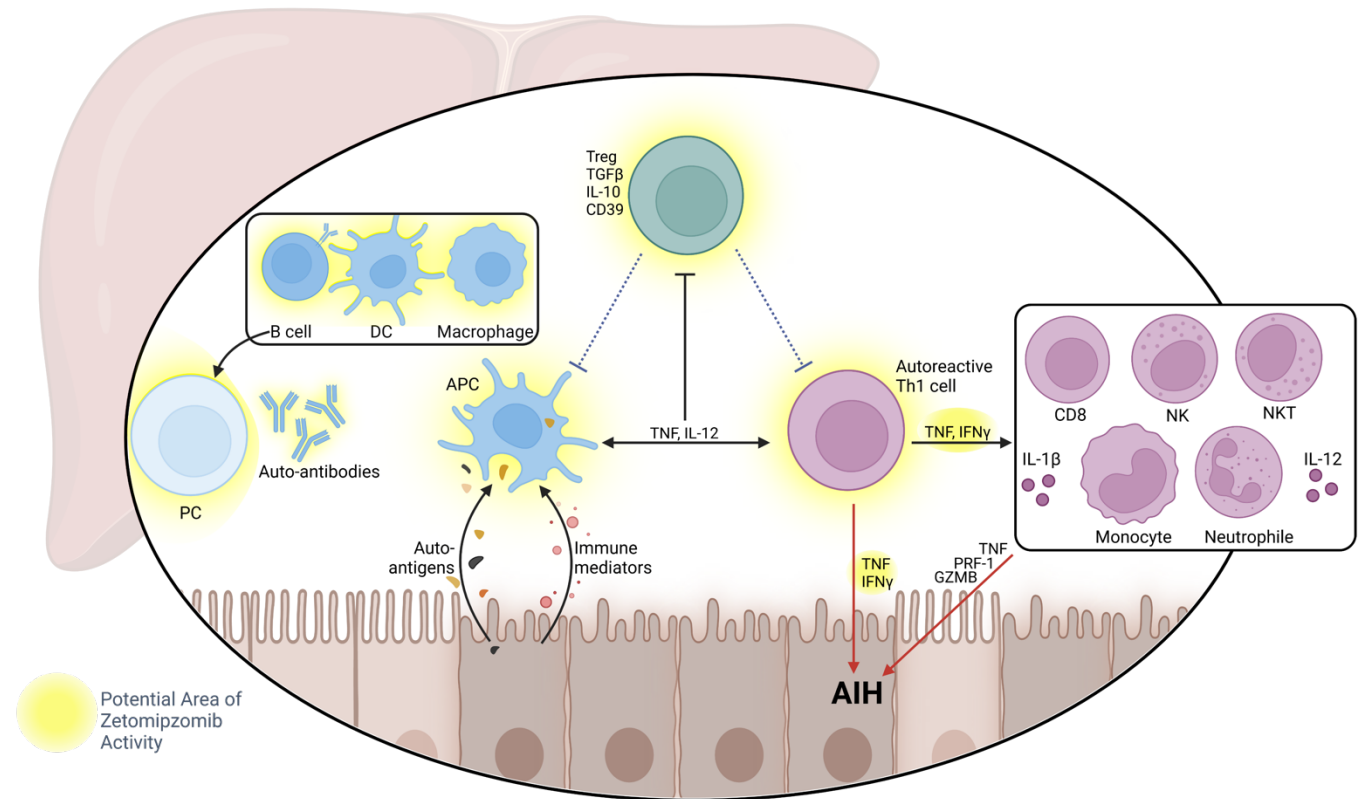
Newest Clinical Program for Zetomipzomib: Autoimmune Hepatitis (AIH) Significant Need For Treatments that Reduce Use of Chronic Immunosuppression

AIH: Complex Autoimmune Liver Disease with Increasing Prevalence

Significant Unmet Need Remains:

- Chronic, immunosuppressive steroids are the mainstay treatment¹
- 35% of patients on SOC do not go into remission²
- **Significant need for treatments that reduce the use of corticosteroids**

Zetomipzomib Targets Multiple Immune Effector Cells Involved in AIH

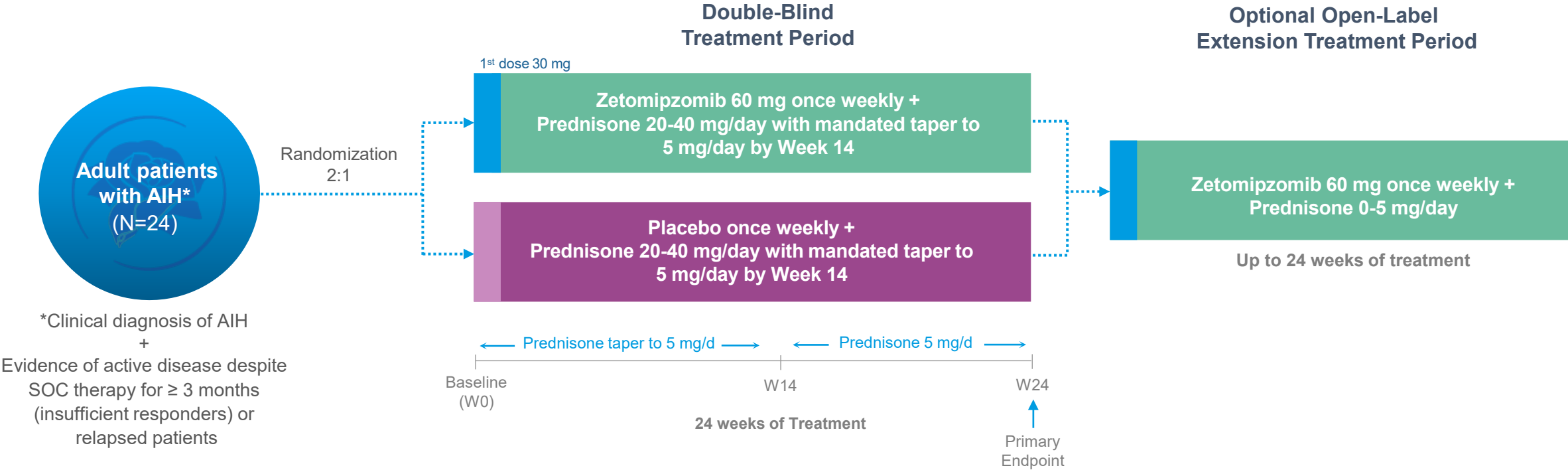


Adapted from Herkel et al., *Journal of Hepatology*. 2020,73(2):446-448.

¹Mack et al., *Hepatology*. 2020;72(2):671-722. ²Volk and Reau. *Clinical Liver Disease*. 2021;17(2):85-89.

Abbreviations: AIH, autoimmune hepatitis; SOC, standard of care.

PORTOLA: Phase 2a Placebo-Controlled Trial Evaluating the Safety and Efficacy of Zetomipzomib in Autoimmune Hepatitis



*Clinical diagnosis of AIH + Evidence of active disease despite SOC therapy for ≥ 3 months (insufficient responders) or relapsed patients

KEY ENDPOINTS

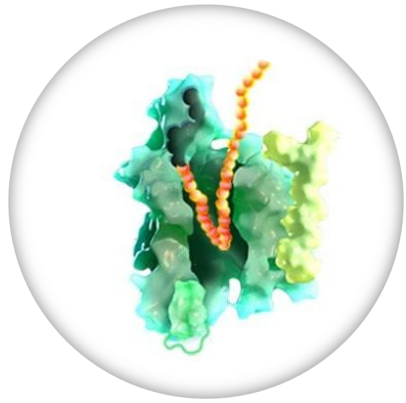
Primary Efficacy

- Proportion of patients who achieve complete remission (ALT/AST normalization) with successful corticosteroid taper by Week 24

Primary Safety

- Proportion of patients who experience adverse events and severe adverse events



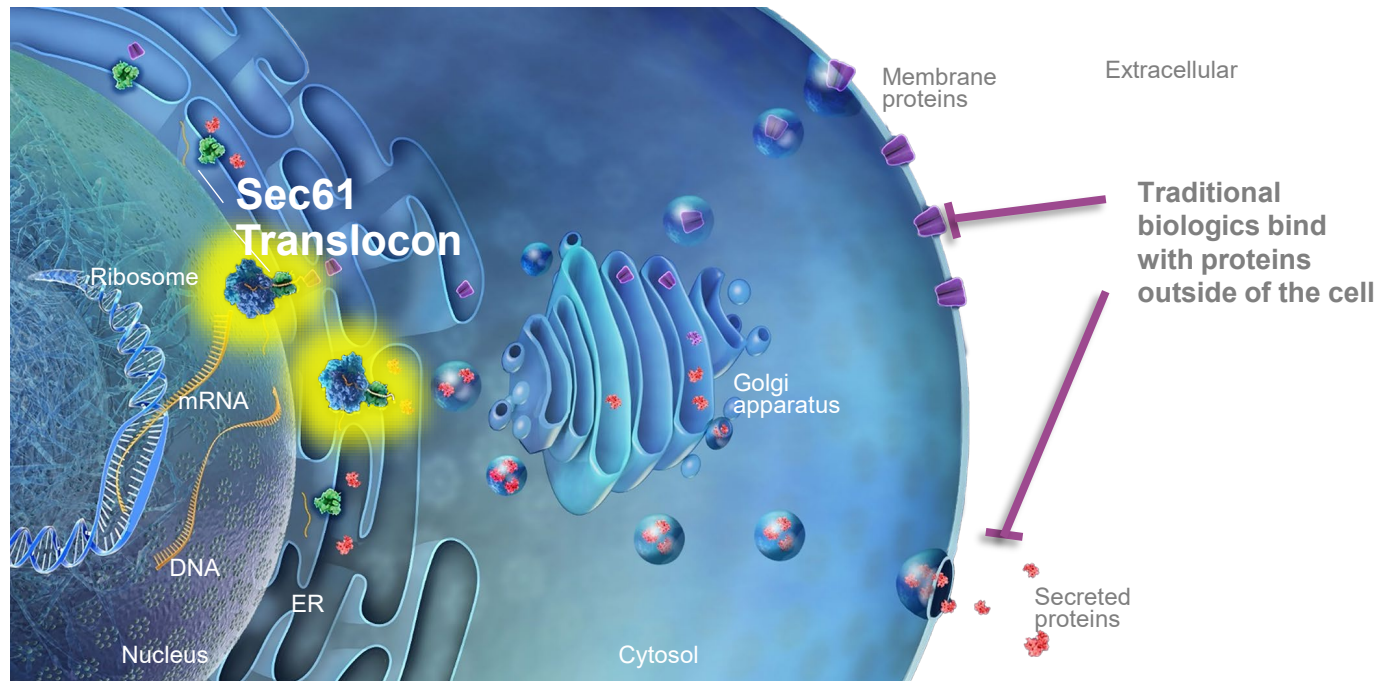


PROTEIN SECRETION
INHIBITION:
KZR-261

KZR-261: A First-in-Class Anti-Cancer
Agent Targeting the Sec61 Translocon

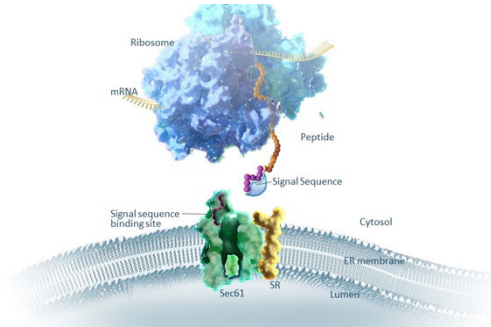
Protein Signals that Promote Tumor Proliferation, Metastasis, and Immune System Evasion Take Their First Steps Out of a Cell Via the Sec61 Translocation Channel (Translocon)

- The Sec61 Translocon is a highly conserved process, functional in all cells
- Nearly all secreted and transmembrane proteins (5,000 – 7,000 proteins) utilize Sec61 to enter the ER
- **Tumor cells rely on secreted and transmembrane proteins more than healthy cells. Thus, slowing traffic through the Sec61 Translocon will impact tumor cells to a greater degree.**
- **Currently available therapeutics target proteins that traffic through the Sec 61 translocon once they have been secreted or expressed. Our MOA inhibits expression of these proteins before they leave the cell.**



● - Target of Sec61 Inhibitors

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



- Unique drug discovery engine 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 **multiple** secreted/membrane proteins
- Combination therapy in a single molecule
- **Multiple potential oncology indications (tumor agnostic)**

KZR-261: 1st clinical candidate

Subset Protein Secretion Inhibitors

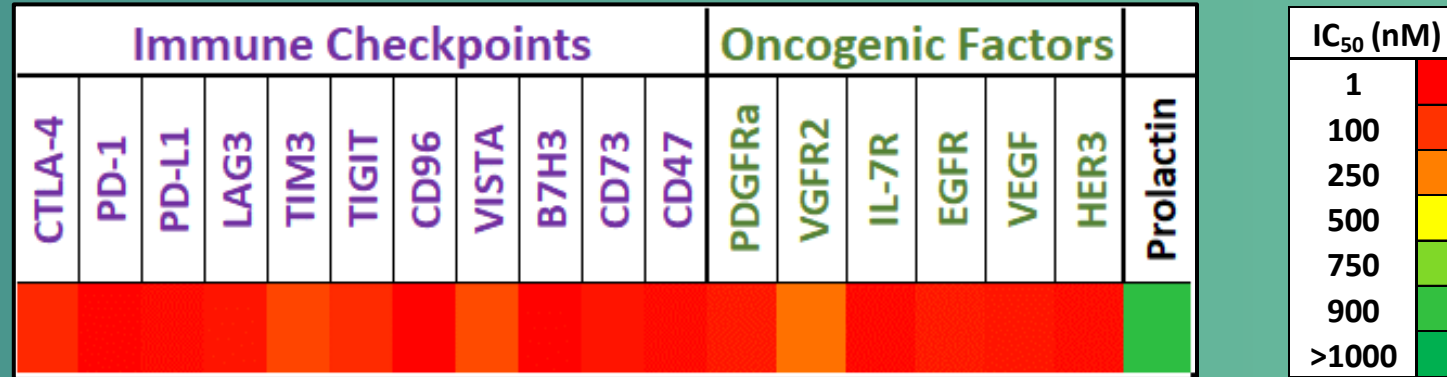
- Inhibition of relevant **subset** secreted/membrane proteins
- Non-cytotoxic agents
- **Indications: oncology, immuno-oncology, immunology**

Single Protein Secretion Inhibitors

- Inhibition of a **single** secreted/membrane protein
- **Preclinical oral PD1 inhibitor: KZR-540**
 - Data presented at SITC 2022
- Non-cytotoxic agents
- **Indications: Many...**

KZR-261: Combination Therapy in a Single Small Molecule

In vitro Protein Secretion Assays



Direct Effects on Tumor Cells

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



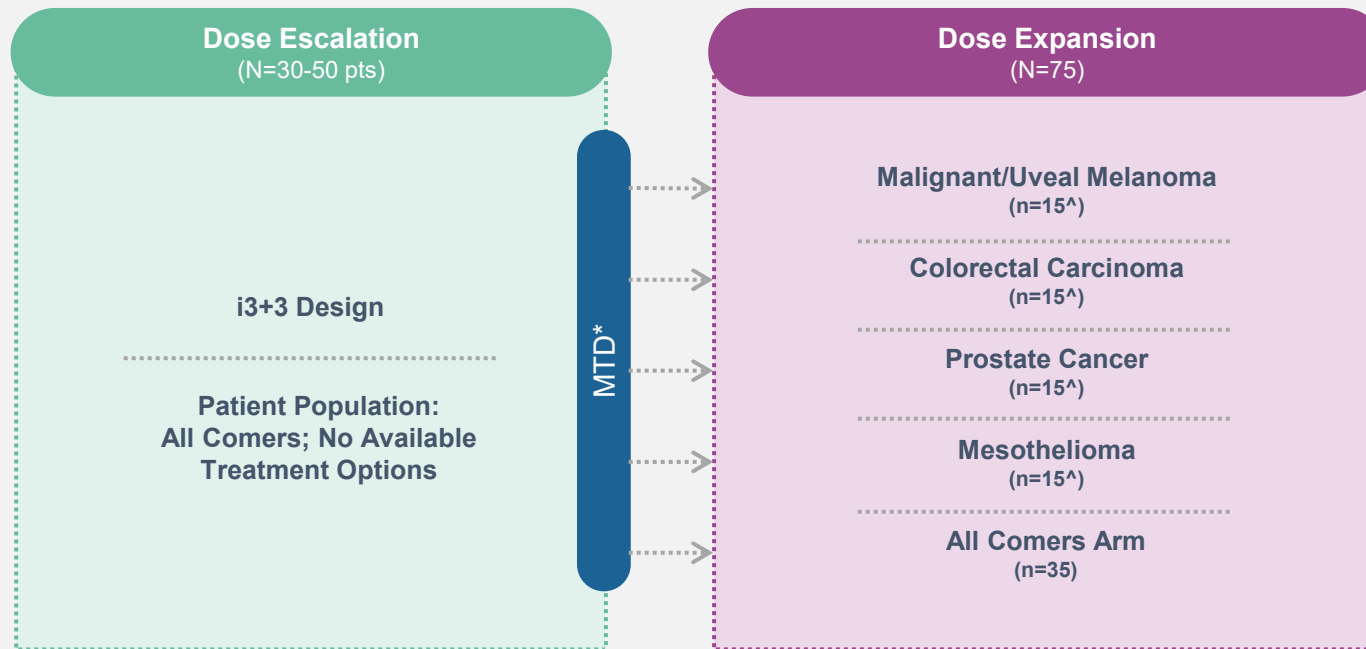
Tumor Microenvironment Modulation

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

Phase 1 Trial Ongoing

KZR-261: First-in-Human Study Ongoing

KZR-261-101 Phase 1 Trial Design



NCT05047536

*Maximum Tolerated Dose

^Fifteen subjects will be enrolled in each tumor specific cohort and may be increased to 35 subjects if sufficient efficacy is observed.



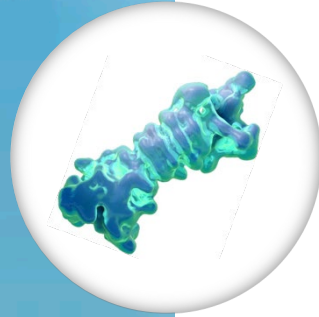
Key Outcome Measures

- Safety, tolerability & PK
- Recommended Phase 2 dose (RP2D)
- Anti-tumor efficacy
- Biomarker validation

Goals for KZR-261-101

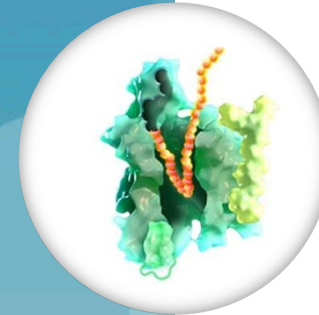
- Establish single agent activity
- Maximize opportunities for success for KZR-261
- Identify/confirm potential predictive biomarkers

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