



INVESTOR & ANALYST EVENT

JUNE 27, 2022

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Agenda

KEZAR SPEAKERS



John Fowler, MBA
Chief Executive Officer



Noreen R. Henig, MD
Chief Medical Officer

4:30 PM – 4:35 PM

Welcome, Introductions and Opening Remarks | *John Fowler, MBA*

4:35 PM – 5:05 PM

MISSION Phase 2 Clinical Update | *Noreen R. Henig, MD*

5:05 PM – 5:10 PM

Zetomipzomib Clinical Development Next Steps | *Noreen R. Henig, MD*

5:10 PM – 5:15 PM

Summary and Closing Remarks | *John Fowler, MBA*

5:15 PM – 5:30 PM

Question and Answer (sell side only)

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 co-founders, 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

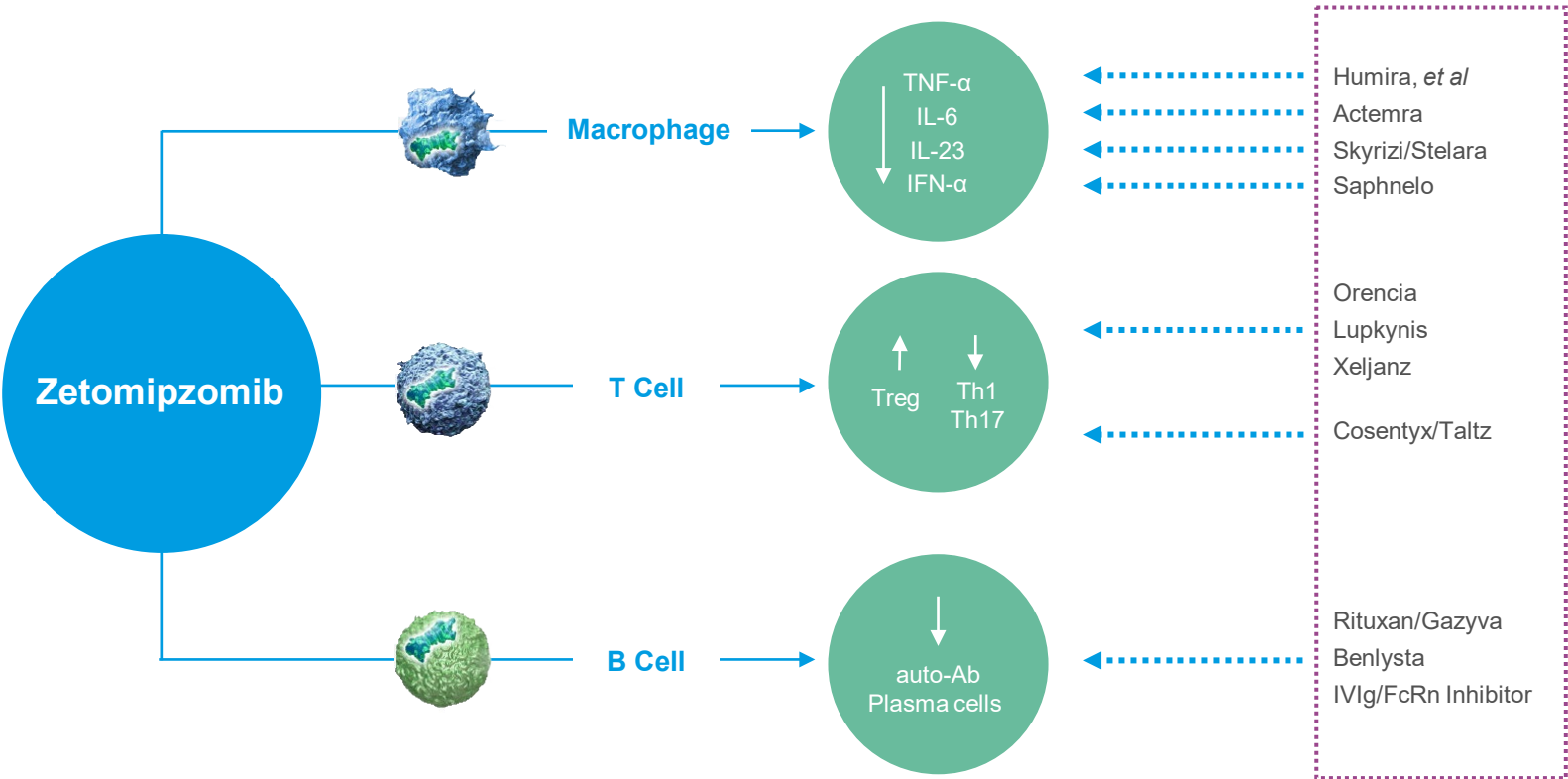
Strong Financial Position (as of 4/30/2022)

\$253M cash, cash equivalents and marketable securities; 60.4M common shares outstanding

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

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



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

A circular graphic with a purple background and a white dotted border. Inside the circle, the text "MISSION Phase 1b in SLE ± LN" is written in white. A faint background image of a building is visible behind the text.

MISSION Phase 1b in SLE ± LN

- Treatment with zetomipzomib demonstrated improvements in multiple exploratory measures of disease activity across organ systems including SLEDAI-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 $\geq 50\%$ 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

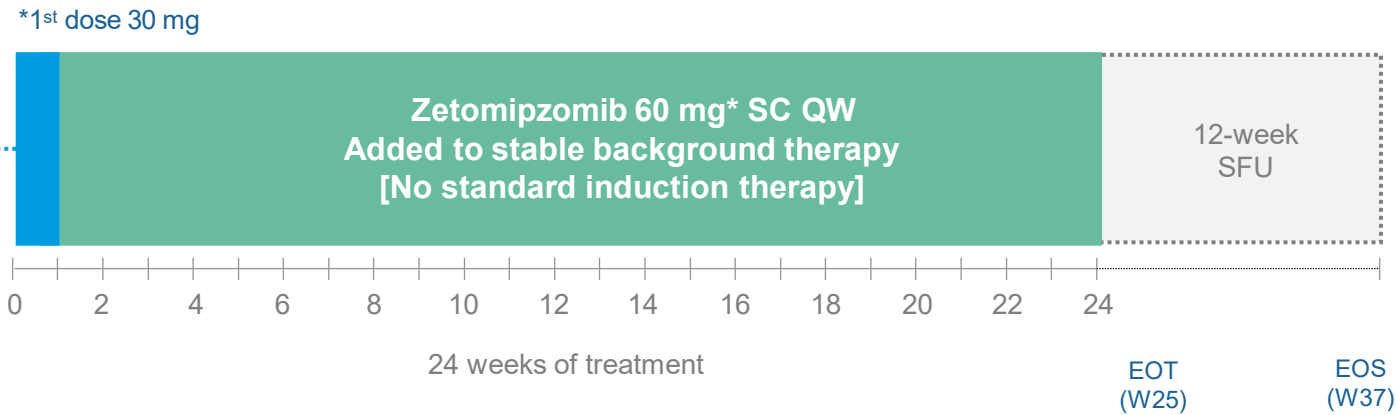
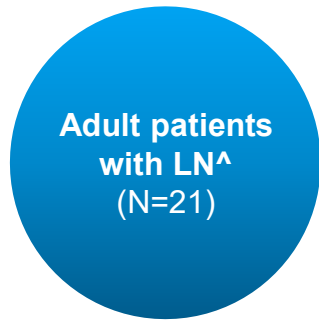
Patients received 24 weeks of zetomipzomib; End-of-treatment assessments performed at Week 25.

Abbreviations: LN, lupus nephritis; CRR, complete renal response; UPCR, urine protein to creatinine ratio, eGFR, estimated glomerular filtration rate; SLE, systemic lupus erythematosus.

Topline data is subject to audit and verification procedures that could result in material changes in the final data.

MISSION Phase 2: Study Design

Open-Label Clinical Study to Evaluate the Efficacy and Safety of Zetomipzomib in Patients with Active Proliferative Lupus Nephritis



^LN patients (Class III or IV +/- Class V) with significant proteinuria (UPCR ≥ 1.0 measured in 24-hour urine collection) despite standard therapy

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

<https://clinicaltrials.gov/ct2/show/NCT03393013>.

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

Abbreviations: UPCR, urine protein to creatinine ratio; LN, lupus nephritis; SC, subcutaneous; SFU, safety follow-up; QW, every week; W, week; EOT, end of treatment; EOS, end of study.



Primary Endpoint: Overall Renal Response (ORR)

- Number of patients with $\geq 50\%$ 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 $\geq 25\%$
- 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 < 3 (if baseline UPCR > 3)
- eGFR ≥ 60 mL/min/1.73m² or no worsening of eGFR from baseline of $\geq 25\%$
- No use of prohibited medication

- Global SLE disease monitoring and key biomarkers

MISSION Phase 2 Topline: Safety Population

Key Demographics and Baseline Characteristics



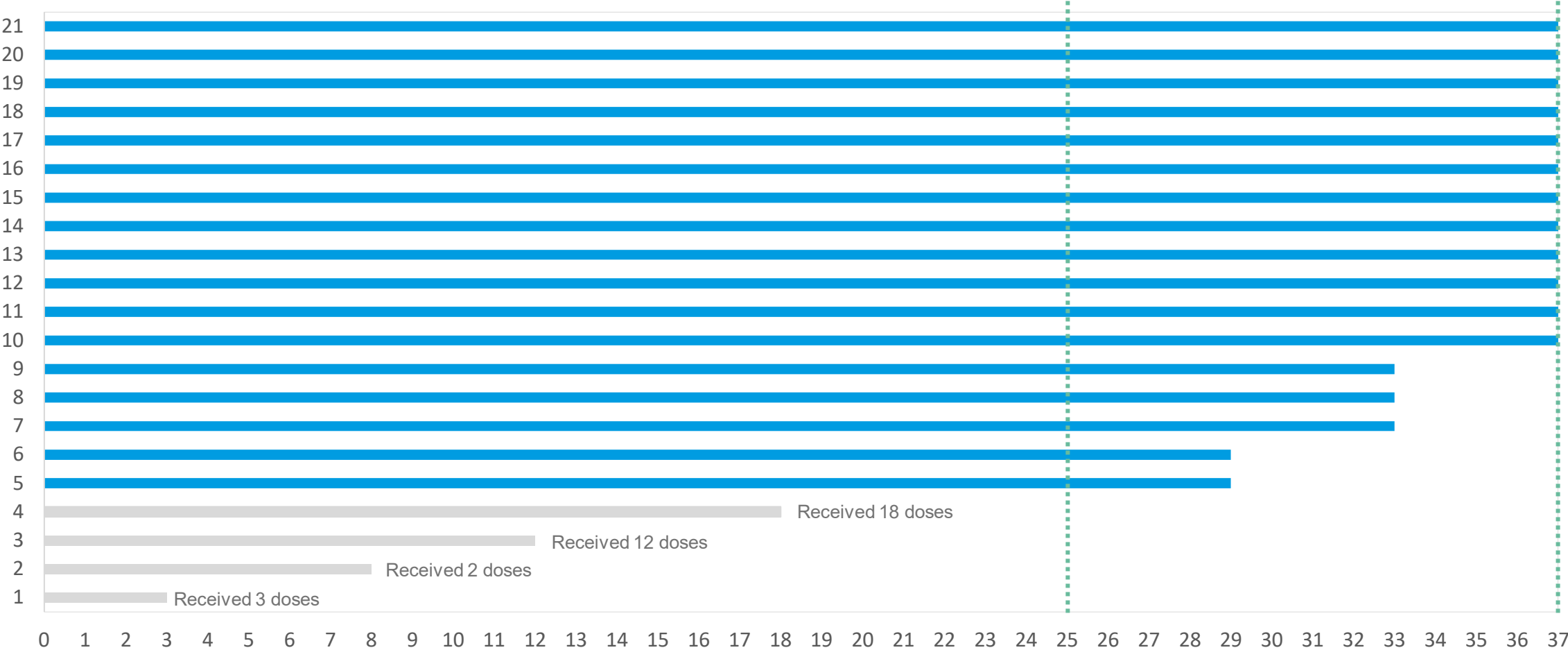
	Safety Population (N=21)		Safety Population (N=21)
Age, mean (years), range	35.3 (19 - 69)	SLE duration (years), median (min, max), N=20	8.7 (0.7, 26.4)
Female, n (%)	19 (90.5)	LN duration (years), median (min, max)	3.1 (-0.06*, 16.1)
Race, n (%)		LN class type, n	
White	7 (33.3)	Class III only	6
Black or African American	1 (4.8)	Class IV only	11
Asian	1 (4.8)	Class III + V	3
American Indian/Alaska Native	4 (19.0)	Class IV + V	1
Other	7 (33.3)	24-hour UPCR (mg/mg)	
Unknown	1 (4.8)	Mean (SD)	2.6 (2.6)
		Median (Min, Max)	1.9 (0.93, 13.4)
Ethnicity, n (%)		eGFR (mL/min/1.73 m2)	
Hispanic or Latino	11 (52.4)	Mean (SD)	104.7 (32.8)
		Median (Min, Max)	115.0 (36.5, 150)
Country, n (%)		Disease Activity Parameter, mean (SD)	
Australia	3 (14.3)	SLEDAI-2K	11.0 (4.7)
Colombia	4 (19.0)	Corticosteroid (prednisone or equivalent) dose (mg), mean (min, max)	18.9 (5, 50)
Peru	6 (28.6)	Concomitant medications, n	
Russia	4 (19.0)	MMF/MPA	20
Ukraine	1 (4.8)	Prednisone (or equivalent)	21
USA	3 (14.3)	Hydroxychloroquine	14
		Azathioprine	2

*LN biopsy confirming diagnosis was performed during screen period.

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.

Topline data is subject to audit and verification procedures that could result in material changes in the final data.

MISSION Phase 2 Topline: Patient Disposition*



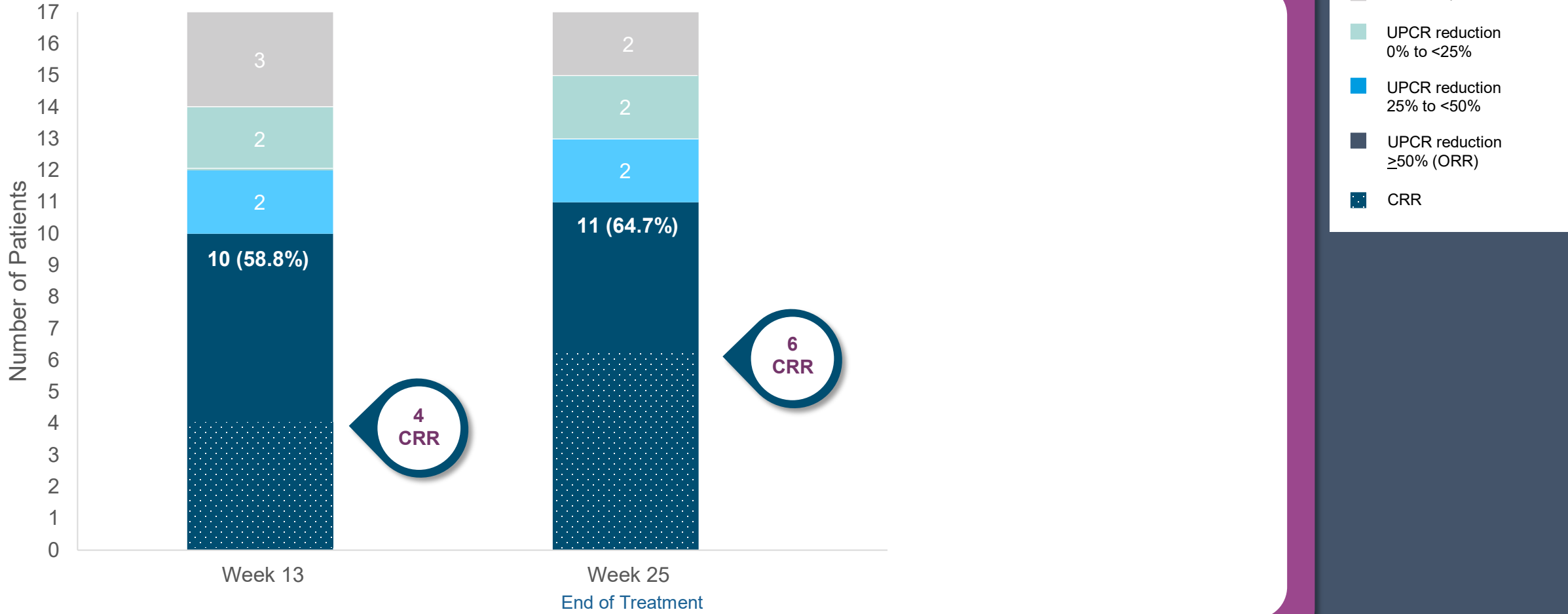
* Each bar represents one patient in the study.

↑
End of Treatment

↑
End of Study

MISSION Phase 2 Topline: Evaluable Population

Zetomipzomib Without Standard Induction Therapy Achieves ORR in 65% and CRR in 35% of Patients at Week 25



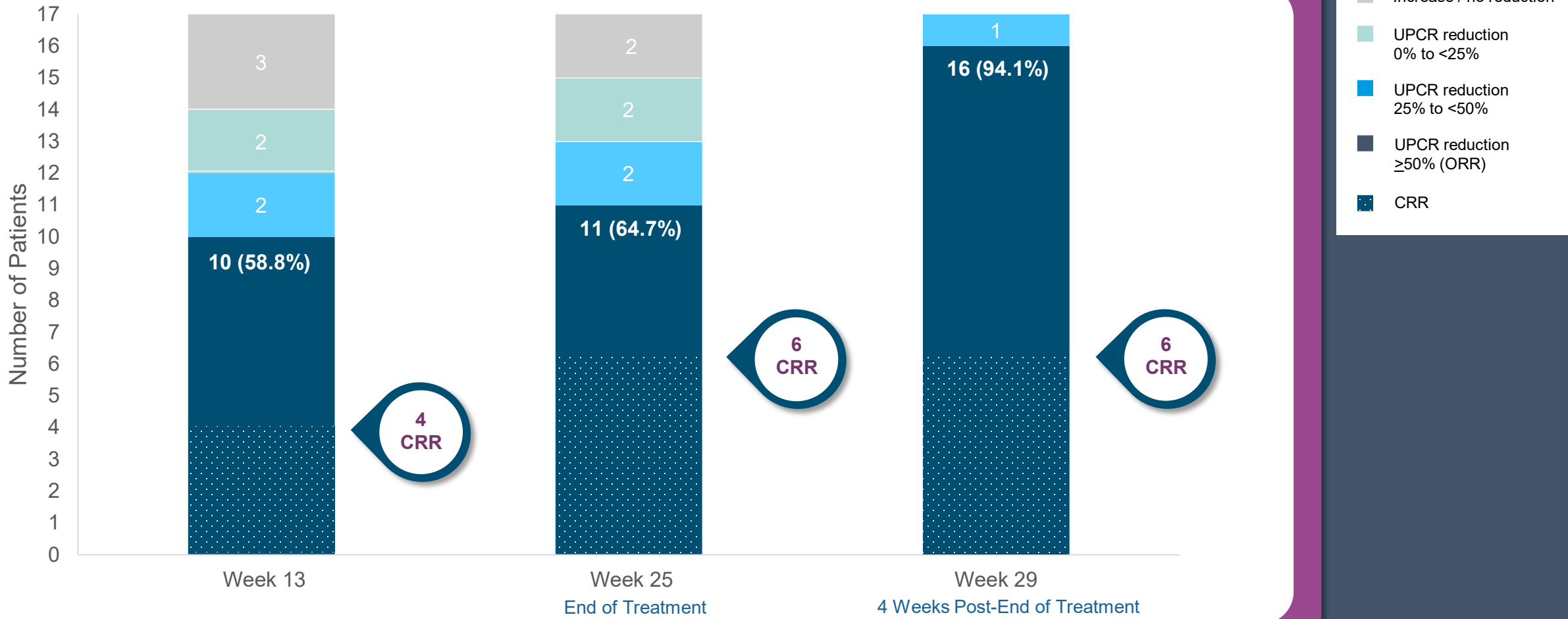
Evaluable population (n=17) are patients that did not withdraw before Week 25; Patients received 24 weeks of zetomipzomib; End-of-treatment assessments performed at Week 25.

Abbreviation: ORR, overall renal response; CRR, complete renal response; UPCR, urine protein to creatinine ratio.

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MISSION Phase 2 Topline: Evaluable Population

CRRs Are Maintained and Additional ORRs are Observed at Week 29, Post-Treatment with Zetomipzomib



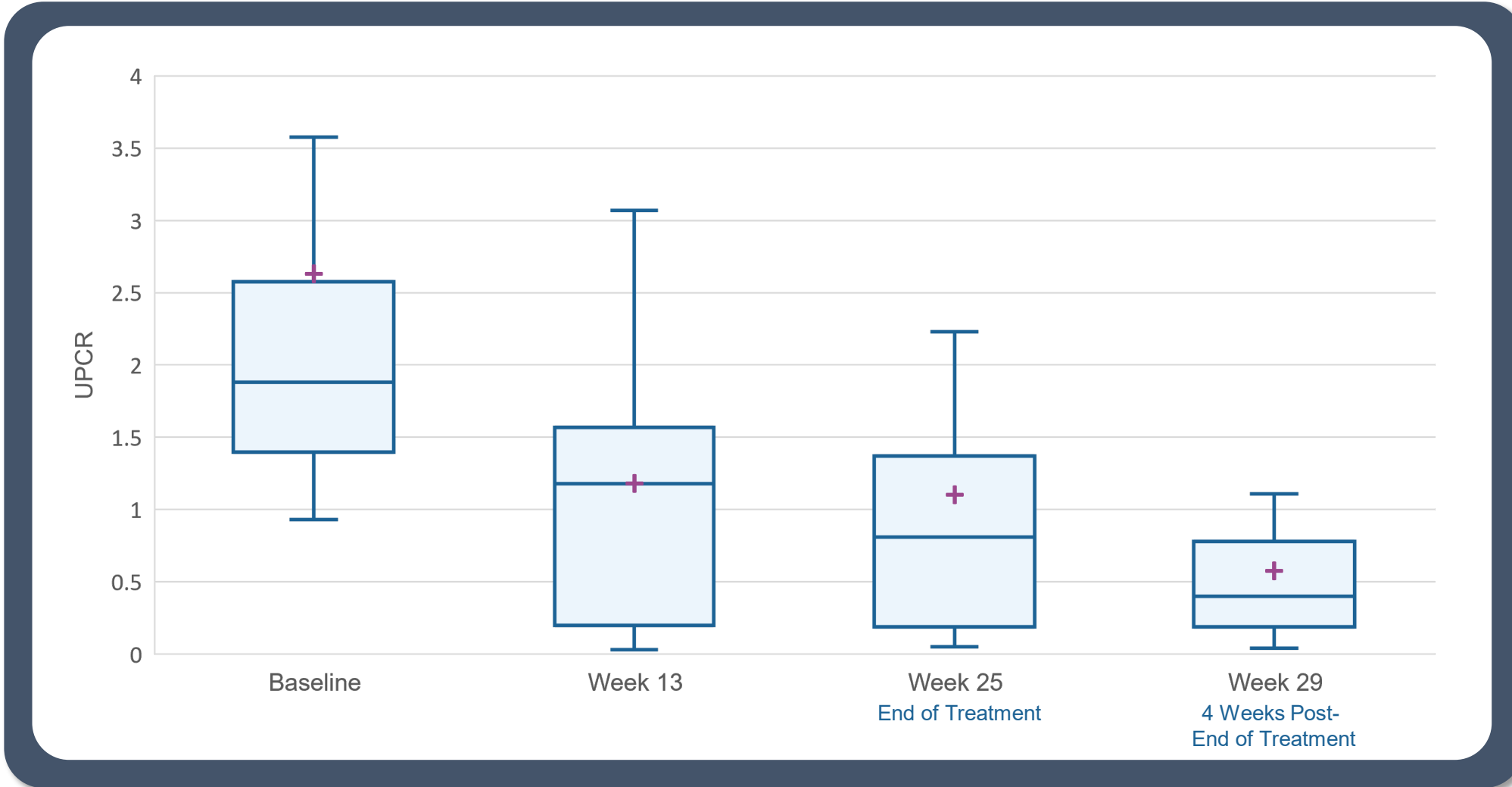
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MISSION Phase 2 Topline: Evaluable Population

Mean UPCR Decreases with 24 Weeks of Zetomipzomib Treatment



Mean +
Median —

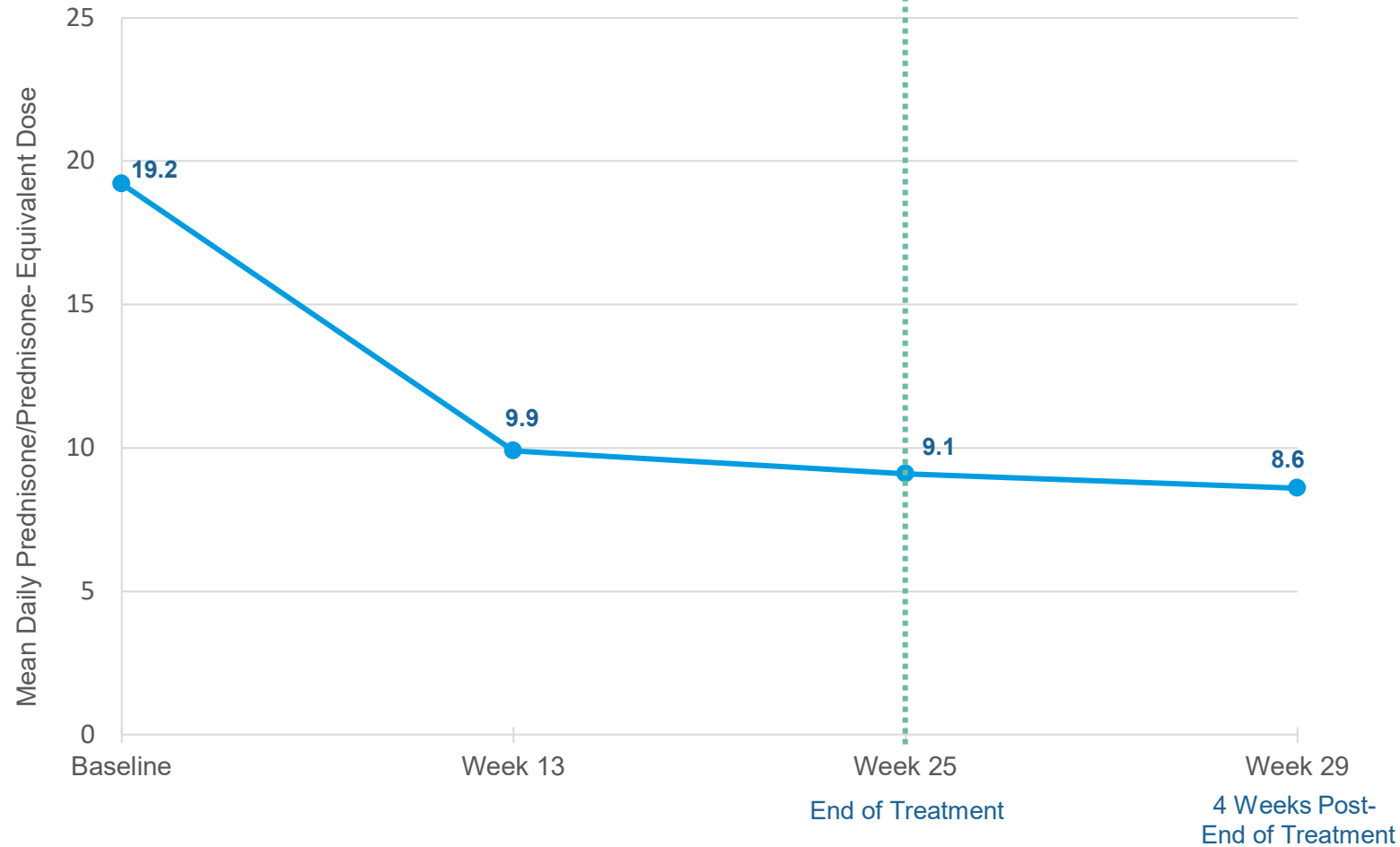
Highest observation*
Upper Quartile (Q3)
Mean +
Median
Lower Quartile (Q1)
Minimum Value


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.

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

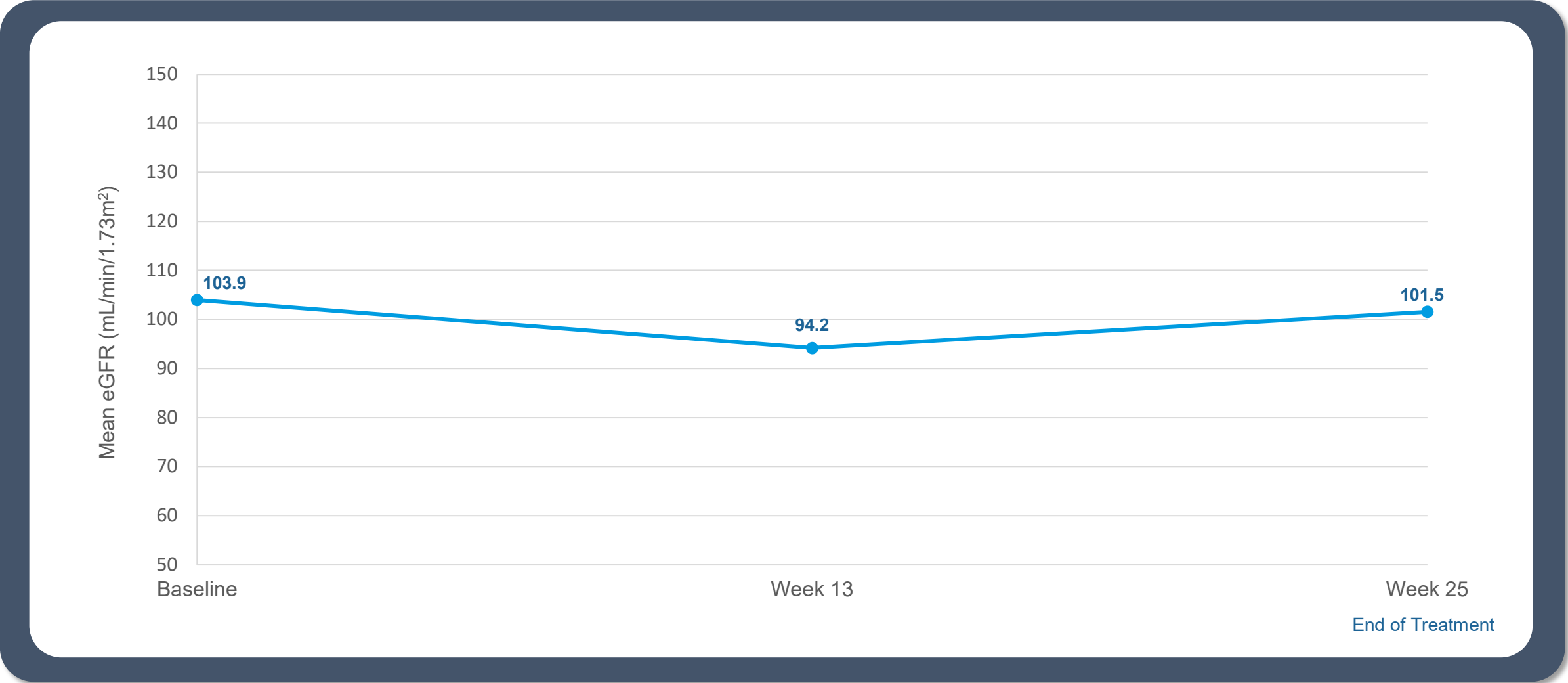


 **Mean Daily Steroid Dose**

- 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 $\geq 25\%$

Abbreviations: eGFR, estimated glomerular filtration rate.

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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.

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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)
Anti-dsDNA	12	10	5
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.

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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 (\leq 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

1. 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

MISSION Phase 2 Topline: Highlights

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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

1

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 steroid-sparing
- Potential treatment for renal and extra-renal manifestations of SLE

2

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

3

Patient-Based Education Initiatives

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

4

Additional Indication Exploration

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



Summary and Closing Remarks

SPEAKERS



John Fowler, MBA
Chief Executive Officer



Noreen R. Henig, MD
Chief Medical Officer



Christopher J. Kirk, PhD
Chief Scientific Officer



THANK YOU