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October 2018

Forward Looking Statements

This presentation (together with any other statements or information that we may make in connection herewith) contains forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995 with respect to Kiniksa Pharmaceuticals, Ltd. (and its consolidated subsidiary, together, unless context otherwise requires, 'Kiniksa," "we" or our). In some cases, you can identify forward looking statements by terms such as "may," "will," "should," "expect," "plan," "anticipate," "could," "intend," "target," "project," "contemplate," "believe," "estimate," "predict," "potential" or "continue" or the negative of these terms or other similar expressions, although not all forward-looking statements contain these identifying words. All statements contained in this presentation that do not relate to matters of historical fact should be considered forward-looking statements, including without limitation statements regarding our strategy, plans and timing for initiation of new clinical trials, potential designs of our new clinical trials, proposed indications for the investigation of our product candidates, plans and timing to report clinical trial data, timing for the initiation of clinical trial sites, plans and timing for the submission of investigational new drug and other applications and submissions to regulatory authorities, and plans and timing to advance additional pipeline programs into clinical trials.

These statements involve known and unknown risks, uncertainties, and other important factors that may cause our actual results, performance or achievements to be materially different from those expressed or implied by the forward-looking statements, including without limitation the important factors discussed under the caption "Risk Factors" in our Quarterly Report on Form 10-Q filed with the Securities and Exchange Commission ("SEC") on August 7, 2018 and our other reports subsequently filed with the SEC. These forward-looking statements reflect various assumptions of Kiniksa's management that may or may not prove to be correct. No forward-looking statement is a guarantee of future results, performance, or achievements, and one should avoid placing undue reliance on such statements.

This presentation also contains estimates, projections, and other information regarding our industry, our business and the markets for certain of our product candidates, including data regarding the estimated size of those markets, and the incidence and prevalence of certain medical conditions. Unless otherwise expressly stated, we obtained this industry, business, market and other data from reports, research surveys, clinical trials, studies and similar data prepared by market research firms and other third parties, from industry, medical and general publications, and from government data and similar sources. Information that is based on estimates, forecasts, projections, market research, or similar methodologies is inherently subject to uncertainties and actual events or circumstances may differ materially from events and circumstances reflected in this information.

Except as otherwise indicated, this presentation speaks as of the date of this presentation. We undertake no obligation to update any forward-looking statements, whether as a result of new information, future events or otherwise.

KINIKSA

A company with a sequential pipeline, FIRMLY rooted in strong biologic rationale or validated mechanisms, potential for multiple indications, and designed to deliver near-, mid- and long-term value

The Kiniksa senior team has extensive experience in drug development and commercialization of valuable therapies

Sanj K. Patel
Chairman & Chief Executive Officer

Steve Mahoney
President & Chief Operating Officer

John Paolini
Chief Medical Officer

Christine Maurer
SVP Program Management

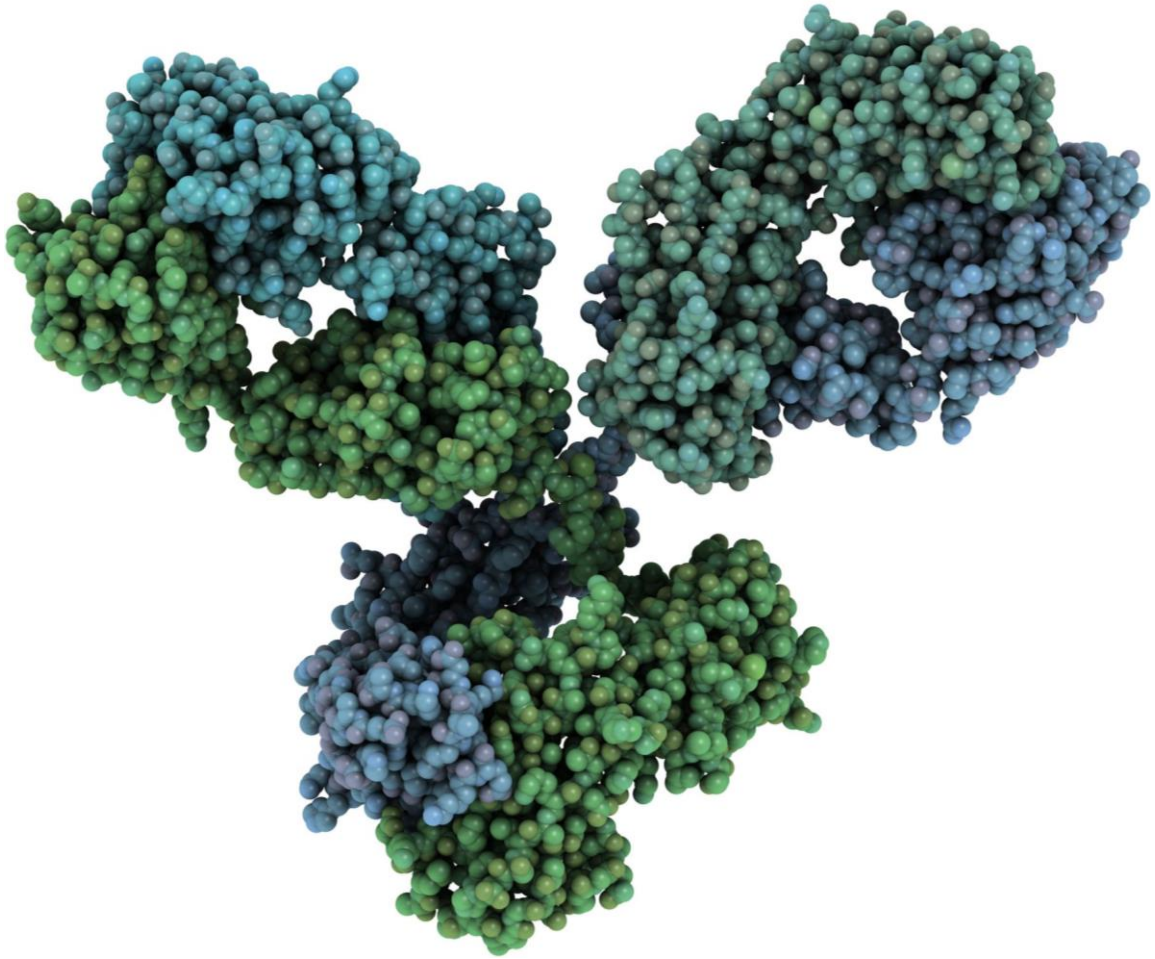
Carsten Boess
Corporate Affairs

Chris Heberlig
Chief Financial Officer

Eben Tessari
Chief Business Officer

Building a fully-integrated global biopharmaceutical company

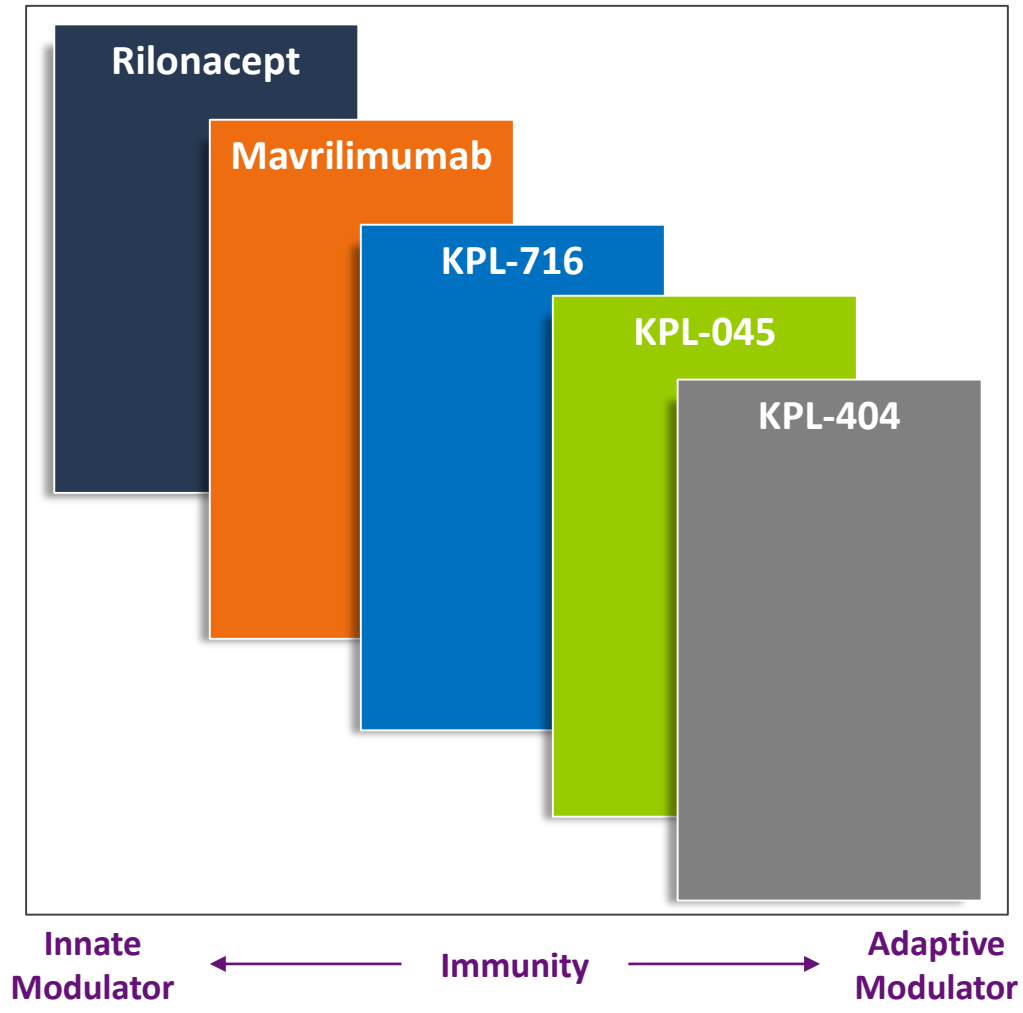
Discovering, acquiring, developing and commercializing life-changing therapies for debilitating diseases



- Focus on the Patients
- Rapid Product Development
- Expand Existing Portfolio Across Multiple Indications
- Build Core Capability in Autoimmune and Autoinflammatory Diseases
- Identify New Products

Strategic approach to building a portfolio

- **Deep literature and KOL reviews to map diseases with a specialty call point on two main axes:**
  - Clear Biology
  - Unmet need
  - Differentiation
- **Strict criteria / filter for:**
  - Market size / prevalence
  - Level of competition
- **Highly selective filter → 5 assets acquired out of >1k reviewed**



Identified pockets of unmet need ripe for innovation across a range of autoinflammatory & autoimmune diseases

A sequential pipeline across various stages of development

Designed to deliver near-, mid- and long-term value

Program & Target	Lead Indication	Phase (Preclin / 1 / 2 / 3)	Status and Anticipated Next Milestone	Rights
Rilonacept[1] IL-1α & IL-1β	Recurrent Pericarditis		• Expect to report additional data from ongoing open-label Phase 2 proof-of-concept trial in 2H 2018 • Plan to advance to a pivotal Phase 3 trial in 2H 2018	Worldwide (excluding MENA)
Mavrilimumab GM-CSFRα	Giant Cell Arteritis		• Plan to advance to a Phase 2 proof-of-concept trial in 2H 2018	Worldwide
KPL-716 OSMRβ	Prurigo Nodularis / Atopic Derm		• Plan advancement into multiple chronic pruritic diseases, including prurigo nodularis • Ongoing repeat single-dose Phase 1b trial in subjects with AtD	Worldwide
KPL-045[2] CD30L	Autoimmune		• IND filing planned for 2H 2019	Worldwide
KPL-404[2] CD40	Autoimmune		• IND filing planned for 2H 2019	Exclusive Option for Worldwide

1) Rilonacept (ARCALYST®) is approved and marketed for cryopyrin-associated periodic syndrome, in the United States by Regeneron. We will assume the rights to this indication upon receiving approval for rilonacept in the recurrent pericarditis indication; 2) We are planning IND-enabling studies for both KPL-045 and KPL-404 in T-cell-dependent, B-cell-mediated diseases, such as pemphigus/pemphigoid, myasthenia gravis, or graft versus host disease.

Rilonacept	Mavri	KPL-716	KPL-045	KPL-404
Approved for CAPs in US as ARCALYST Phase 2	Phase 2-ready	Phase 1a/1b	Pre-IND	Pre-IND

An opportunity in an inflammatory cardiovascular disease with established proof-of-concept and no currently-approved therapies

Lead Indication	• Recurrent Pericarditis
Patient Population[1]	• ~90k patients in the US experience an acute incident of pericarditis per year • ~3k refractory patients and ~9k who are not well-managed on existing therapies in the US • Incremental opportunity as a steroid-sparing agent and to treat Post Pericardiotomy and Dressler Syndromes
Mechanism of Action[2]	• IL-1α and IL-1β cytokine trap • Inhibition of IL-1 has been shown to be instrumental in the resolution of recurrent pericarditis
Competition[3]	• No currently-approved therapies • Differentiated from both existing marketed IL-1 agents
Clinical Development	• Expect to report additional data from ongoing open-label Phase 2 proof-of-concept trial in 2H 2018 • Plan to advance to a pivotal Phase 3 trial in 2H 2018

1) UptoDate, Trinity Partners, Mayo Clin Proc. 2010 ;85 (6): 572-593; New Diagnostic Criteria for Acute Pericarditis: A Cardiac MRI Perspective, 2015 American College of Cardiology; 2) Brucato et al. JAMA. 2016, 316 (18): 1906-1912; Arcalyst Prescribing Information; 3) Drugs@FDA: Arcalyst Prescribing Information, Ilaris Prescribing Information, Kineret Prescribing Information; Kaiser et al. Rheumatol Int (2012) 32:295–299; Theodoropoulou et al. Pediatric Rheumatology 2015, 13(Suppl 1):P155 ; Fleischmann et al, 2017 ACR/ARHP Abstract 1196; Kosloski et al, J of Clin Pharm 2016, 56 (12) 1582-1590; Cohen et al. Arthritis Research & Therapy 2011, 13:R125; Cardiel et al. Arthritis Research & Therapy 2010, 12:R192; Hong et al. Lancet Oncol 2014, 15: 656-666

The role of IL-1α signaling in the pathology of recurrent pericarditis and multiple diseases of sterile serosal inflammation

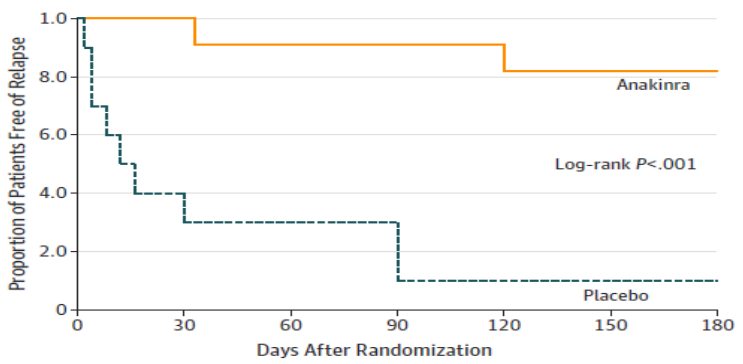
Proof-of-concept in RIP shown by anakinra (daily sc-administered IL-1α and IL-1β blocker) in AIRTRIP study

Anakinra in Patients with Cortico-Dependent Idiopathic Recurrent Pericarditis: A Randomised Double-Blind Placebo-Controlled Withdrawal Trial

AIR TRIP = AnakInRa - Treatment of Recurrent Idiopathic Pericarditis

Antonio Brucato, Massimo Imazio, Silvia Maestroni, Davide Faustino Cumetti, Anna Valenti, Renzo Marcolongo, George Lazaros, Mara Carraro, Fiorenzo Gaita, Gian Luca Erre, Martina Finetti, Marco Gattorno and Alberto Martini.

Patients With Recurrent Pericarditis Free of Relapse in the Double-Blind Withdrawal Phase, From Day 0 to Day 180 After Randomization (Intention-to-Treat Analysis)



IL-1β-only inhibition does not work in this patient population

Theodoropoulou et al. Pediatric Rheumatology 2015, 13(Suppl 1):P155
http://www.ped-rheum.com/content/13/S1/P155

PEDIATRIC RHEUMATOLOGY

POSTER PRESENTATION Open Access

A case of corticosteroid-dependent recurrent pericarditis with different response to two IL-1 blocking agents

K Theodoropoulou*, A von Scheven-Gête, S Bressieux-Degueldre, M Prsa, F Angelini, T Boulos, M Hofer

From 8th International Congress of Familial Mediterranean Fever and Systemic Autoinflammatory Diseases
Dresden, Germany. 30 September - 3 October 2015

Introduction
Recurrent pericarditis (RP) has a controversial pathogenesis that crosses infectious, auto-immune and auto-inflammatory pathways. It has been suggested that in some cases it might be an unrecognized auto-inflammatory disease. Recent studies have demonstrated that anakinra, an interleukin-1 receptor antagonist (IL-1RA), represents an effective treatment for the control of corticosteroid-dependent cases.

Objectives
Here we describe a case of cortico-dependent recurrent pericarditis with a different response to two IL-1 blocking agents, anakinra and canakinumab.

Materials and methods
Case report.

Results
A 11-year-old boy was admitted to our hospital with acute precordial pain, orthopnea, fever and increased levels of acute phase reactants. Acute pericarditis was confirmed by echocardiography and a treatment with prednisone was started with prompt clinical improvement. Pericarditis recurred twice during steroid tapering (at 1mg/kg/day and 0.5mg/kg/day respectively). After exclusion of infectious origin, therapy with anakinra (2mg/kg/day) was established (to avoid long term steroid side effects) followed by dramatic clinical response and normalisation of laboratory findings despite tapering and discontinuation of prednisone. Treatment with anakinra was discontinued after 5 months with recurrence of pericarditis one week later. Anakinra was resumed with an excellent response. Five months later, while being in complete remission, anakinra was replaced with canakinumab (2mg/kg/dose) due to patient's intolerance of daily injections. One week later, the patient experienced a new episode of pericarditis requiring corticotherapy. Two more relapses occured during steroid tapering, after 6 weeks and 2 months, in spite of the uptitration of canakinumab to 4mg/kg/dose. Anakinra was restarted with prompt clinical and biological remission and prednisone was discontinuated without recurrence of pericarditis. Four weeks later, anakinra was spaced out every 2 days and a treatment of colchicine was added. After further 12 weeks follow-up under anakinra and colchicine, the pericarditis is still in remission.

Conclusion
We describe a case of steroid-dependent RP with a dramatic therapeutic response to IL-1RA (anakinra) but without response to IL-1β monoclonal antibody (canakinumab). This unexpected observation could suggest that IL-1α might have a role in the pathogenesis of RP. A more precise usefulness of each IL-1 blocking agent requires confirmation in prospective controlled trials.

Consent to publish
Written informed consent for publication of their clinical details was obtained from the patient/parent/guardian/relative of the patient.

Published: 28 September 2015

doi:10.1186/1546-0096-13-S1-P155
Cite this article as: Theodoropoulou et al.: A case of corticosteroid-dependent recurrent pericarditis with different response to two IL-1 blocking agents. *Pediatric Rheumatology* 2015 13(Suppl 1):P155.

Department of Pediatrics, Lausanne University Hospital (CHUV), Lausanne, Switzerland

IL-1α signature across multiple diseases of sterile serosal inflammation

human reproduction ORIGINAL ARTICLE *Reproductive genetics*

Association between endometriosis and the interleukin 1A (*IL1A*) locus

J Res Med Sci. 2014 Dec; 19(12): 1145–1149. PMCID: PMC4333522

The diagnostic role of cervico-vaginal fluid interleukins-1α in endometriosis: A case-control study

Imbalance in Cytokines from Interleukin-1 Family – Role in Pathogenesis of Endometriosis

Justyna Sikora*, Aleksandra Mielczarek-Palacz, Zdzisława Kondera-Anasz

Department of Immunology and Serology, Medical University of Silesia, Katowice, Poland

Interleukin 1α and tissue-lytic matrix metalloproteinase-1 are elevated in ectopic endometrium of patients with endometriosis

Gut, 1990, **31**, 686–689

Role of interleukin 1 in inflammatory bowel disease – enhanced production during active disease

Epithelial Cell-Derived IL-1-alpha as a Novel Danger Signal in IBD Pathogenesis

Fiocchi, Claudio Stylianou, Eleni

Cleveland Clinic Lerner, Cleveland, OH, United States

Intestinal inflammation

ORIGINAL ARTICLE

Non-redundant properties of IL-1α and IL-1β during acute colon inflammation in mice

Marina Bersudsky,[1] Lotem Luski,[1] Daniel Fishman,[2] Rosalyn M White,[1] Nadya Ziv-Sokolovskaya,[3] Shahar Dotan,[1] Peleg Rider,[1] Irena Kaplanov,[1] Tegest Aychek,[4] Charles A Dinarello,[5] Ron N Apte,[1] Elena Voronov[1]

Sources: Brucato et al. JAMA. 2016, 316 (18): 1906-1912; Theodoropoulou et al. Pediatric Rheumatology 2015, 13 (Suppl 1): 155; Sapkota et al. Human Reproduction, 2015, 30 (1) 239-248 ; Mardanian et al. J Res Med Sci 2014, 19 (12), 1145-1149; Sikora et al. Am J of Reproductive Immunology, 2012, 68, 138-145; Hudelist et al. Human Reproduction 2005, 20 (6), 1695-1701; Ligumsky et al, Gut 1990, 31 (6) 686-689; Fiocchi et al. CTSC Project, Bersudsky et al. Gut 2014, 63, 598-609

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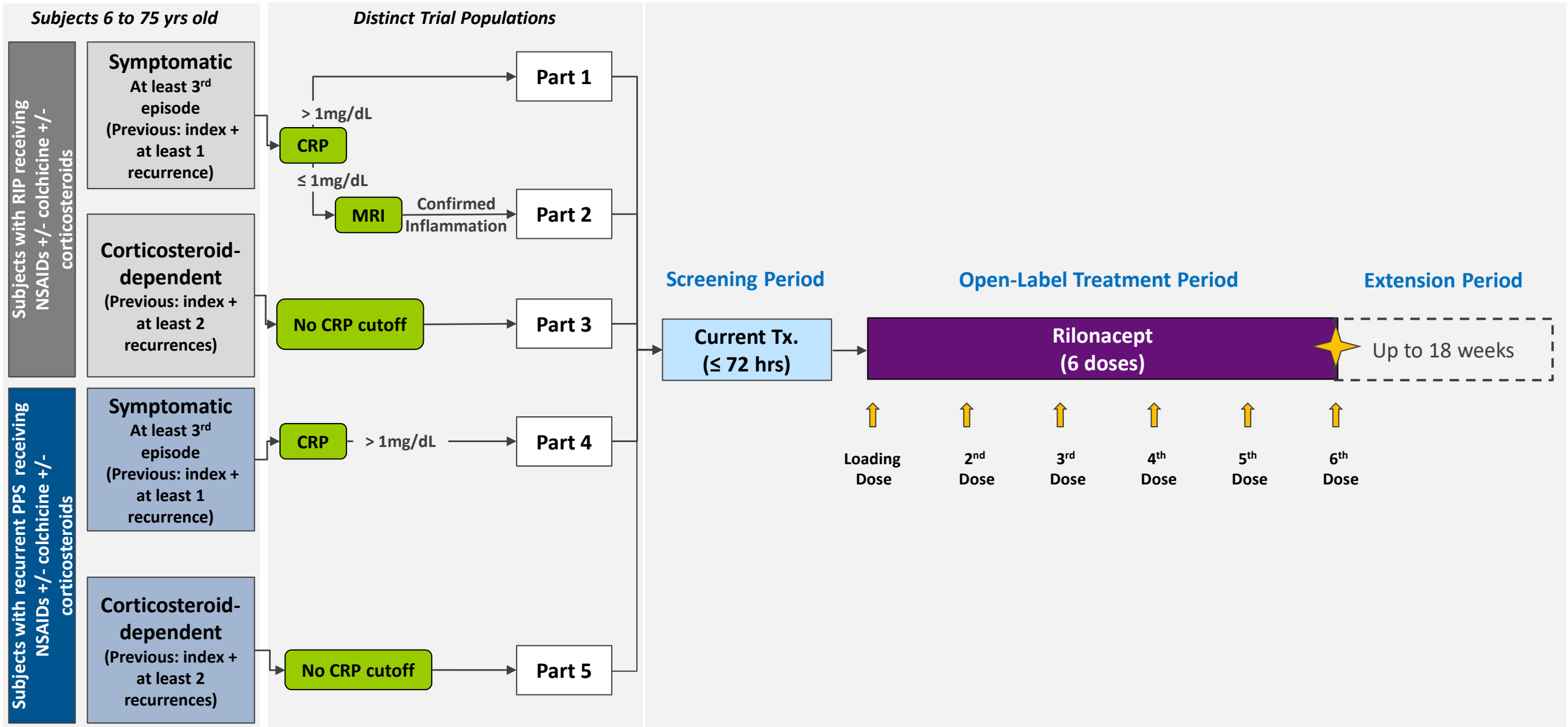
Open-label Phase 2 clinical trial of rilonacept in pericarditis populations



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Rilonacept	Mavrilimumab (KPL-301)	KPL-716	KPL-045	KPL-404
Approved for CAPs in US	Phase 2-ready			

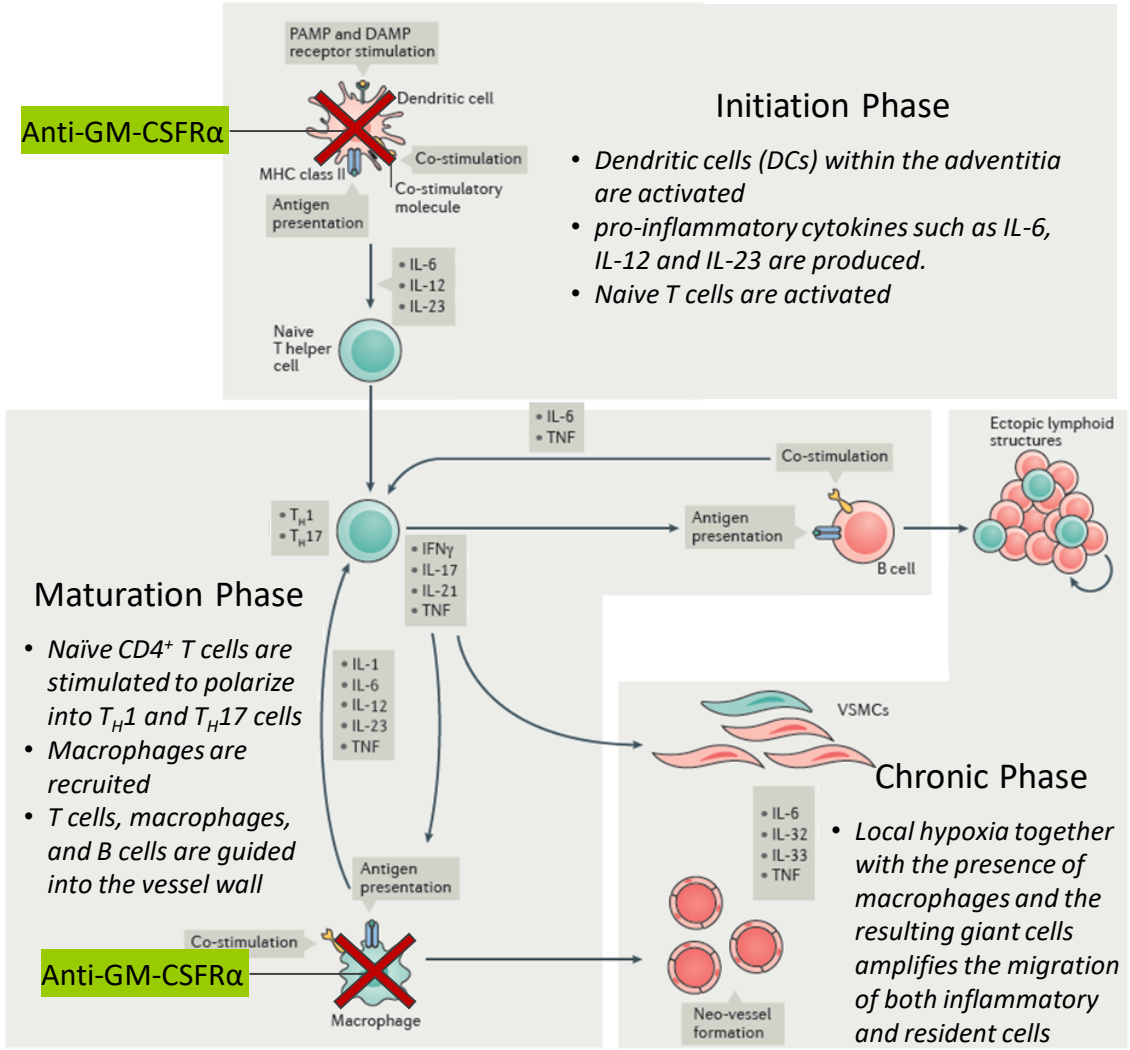
Phase 2 safety and efficacy data in over 550 rheumatoid arthritis subjects in the EU and mechanistic rationale for focusing on high unmet need vasculitides & inflammatory cardiomyopathies

Lead Indication	• Giant Cell Arteritis (GCA)
Patient Population[1]	• ~75k - 150k prevalent GCA patients in the US typically treated with steroid; similar prevalence in EU
Mechanism of Action[2]	• Monoclonal antibody inhibitor targeting GM-CSFRα • Blocks GM-CSF signaling, a key mediator of inflammation and autoimmunity
Competition[3]	• Potential first-in-class molecule • One FDA approved therapy for GCA but there is still a persistent unmet need
Clinical Development	• Plan to advance to a Phase 2 proof-of-concept trial in 2H 2018

1) Chandran et al., Scand J Rheumatol, 2015; Trinity Consulting – HCUP/Medicare Data, Quantitative Survey (n=102 rheumatologists); 2) Sources: Wicks, Roberts, Nature Review Immunology, 2015; Hamilton, Expert Review of Clinical Immunology, 11:4, 457-465; 3) Cortellis,;UpToDate; Correspondence, Trial of Tocilizumab in Giant-Cell Arteritis, NEJM, 2017

GM-CSF implicated in the pathology of giant cell formation by promoting activation, differentiation, survival and proliferation of key cell types

Pathology of GCA



- **GM-CSF is critical for the stimulation of hematopoietic progenitor cells in the bone marrow to generate differentiated myeloid-cell progeny (dendritic cells [DCs], neutrophils, eosinophils and monocyte/macrophages)**
- **GM-CSF enhances trafficking of these cell types through activated endothelium blood vessels, directly contributing to myeloid cell accumulation in blood vessels**
- **GM-CSF promotes activation, differentiation, survival and proliferation of trafficking monocytes and macrophages as well as resident tissue macrophages in inflamed tissues**
- **As shown in the diagram, DCs and macrophages are critical in all three stages of GCA pathology; therefore, blockade of GM-CSF with mavrilimumab has significant potential to attenuate both new-onset and relapsing disease**
  - In the chronic phase GM-CSF promotes giant cell formation, the hallmark pathologic finding of GCA, from activated macrophages
- **Further evidence is provided by data showing elevation of GM-CSF in the lesions of GCA patients**

Sources: Weyand, Ann Int Med 121:484, 1994; Watanabe Joint Bone Spine, in press; Wicks, Roberts, Nature Reviews Rheumatology, 2016; Dejaco et al., Nature Reviews Rheumatology, 2017

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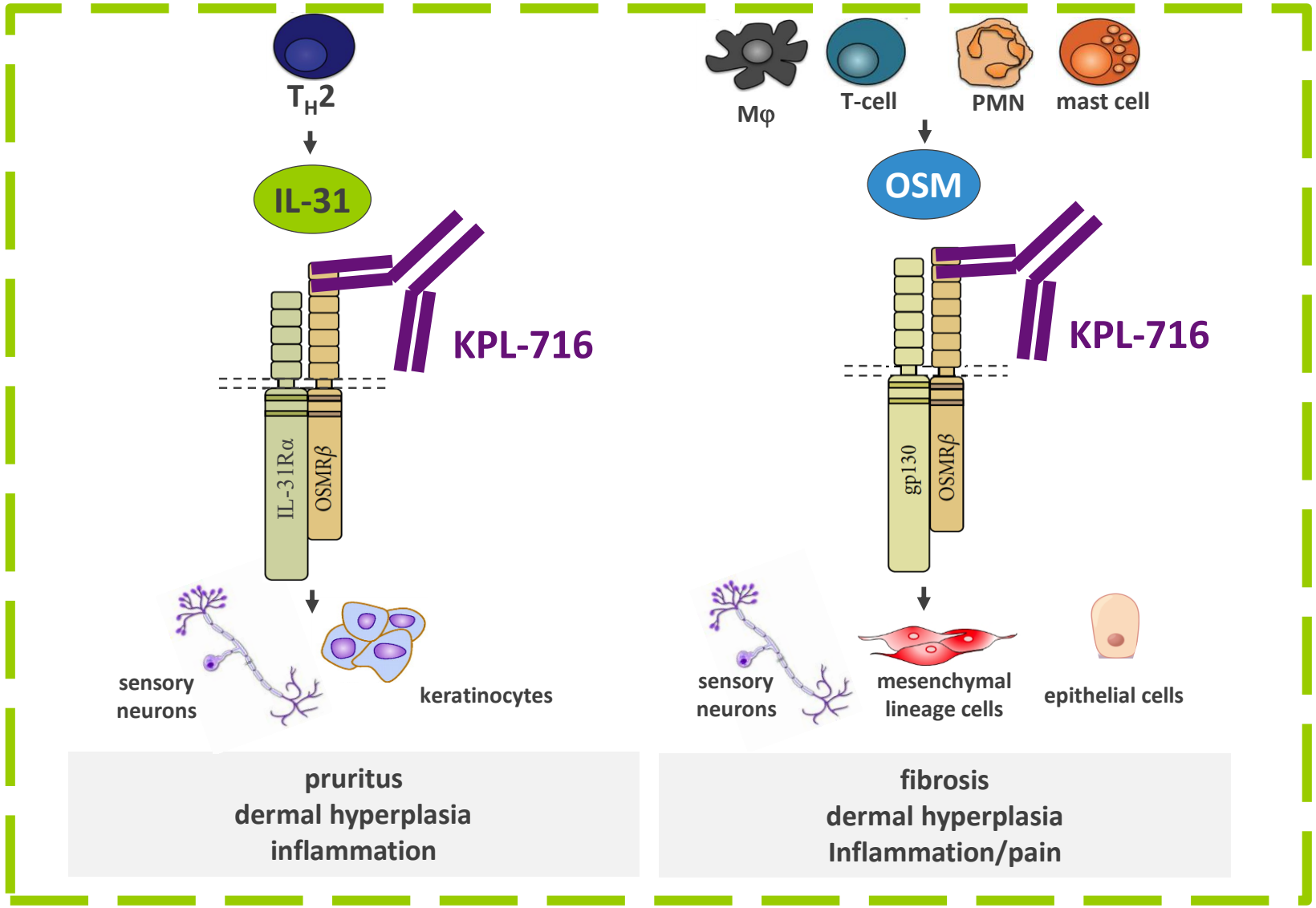
Rilonacept	Mavri	KPL-716	KPL-045	KPL-404
Approved for CAPs in US		Phase 1a/1b		

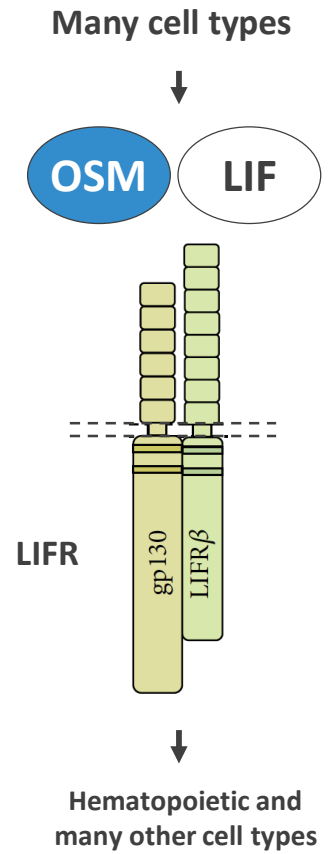
A differentiated molecule with potential to treat a variety of pruritic and fibrotic indications

Lead Indications	• Prurigo Nodularis (PN) • Atopic Dermatitis (AtD)
Patient Population[1]	• PN: Estimated ~300k patients in the US • AtD: Estimated ~1 M moderate-to-severe patients in the US; ~300k of which are eligible for systemic biologics
Mechanism of Action[2]	• Monoclonal antibody inhibitor of signaling through OSMRβ • OSMRβ is a key receptor subunit used by two inflammatory cytokines, IL-31 and Oncostatin M
Competition[3]	• Potential for differentiated efficacy and safety • Competitors block either IL-31 or OSM alone
Clinical Development	• Plan advancement into multiple chronic pruritic diseases, including prurigo nodularis • Ongoing repeat single-dose Phase 1b trial in subjects with AtD

1)Trinity Consulting - HCUP/Medicare Data 2012/2013; Quantitative Survey (n=100 dermatologists); Dantas, 2015, "Prevalence of dermatoses in dermatologic evaluation requests from patients admitted to a tertiary hospital for 10 years"; Mortz et al., Britis Journal of Dermatology, 2001; 2) Trinity Qualitative Interviews; 3) Simpson et al., N Engl J Med, 2016; Ruzicka et al., N Engl J Med, 2017; Reid et al., 2016 ACR Abstract # 1881; Cortellis

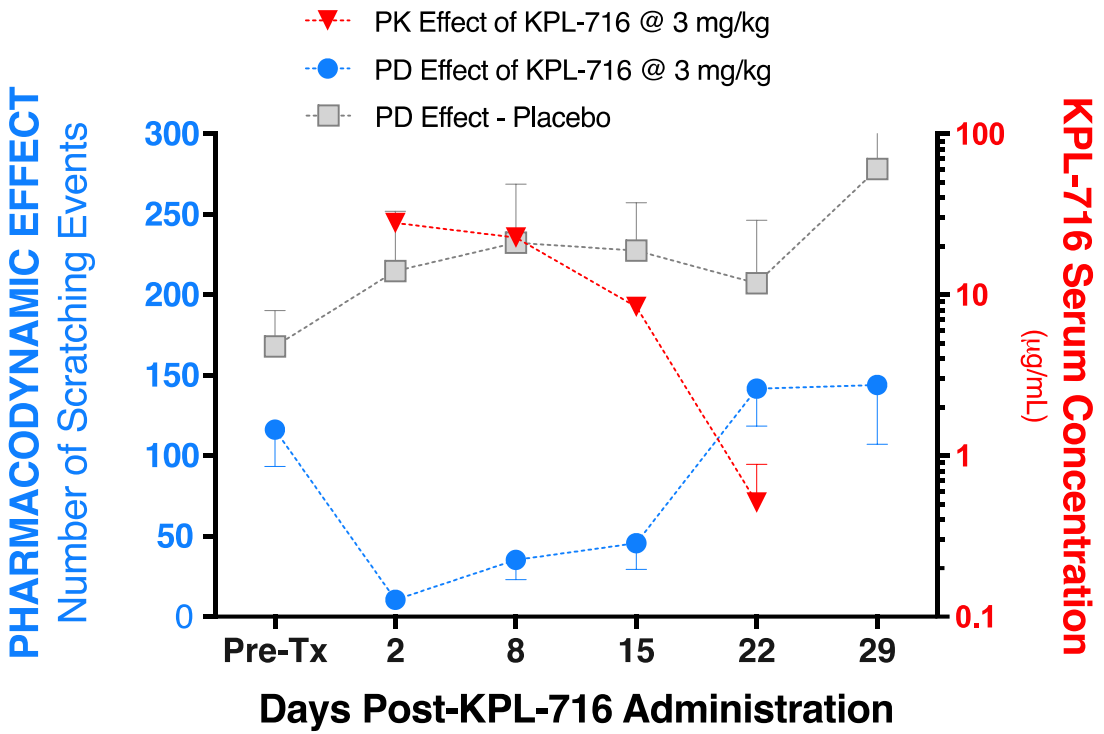
KPL-716 inhibits IL-31 & OSM signaling through OSMRβ but avoids inhibiting signaling critical to hematopoiesis through OSM/LIFR





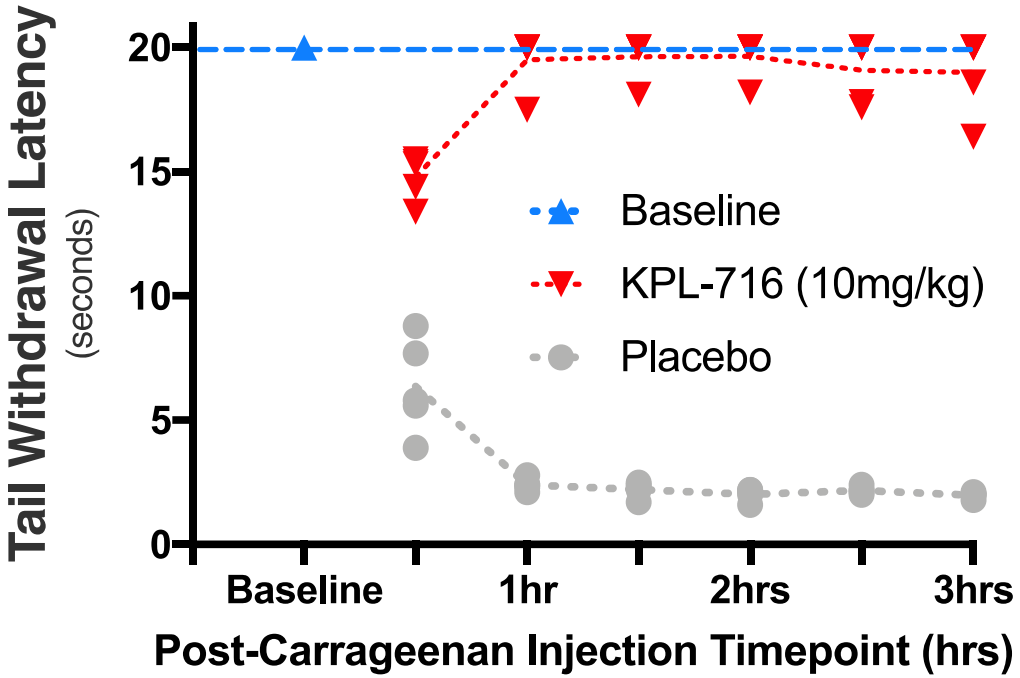
KPL-716 demonstrated potential efficacy in two validated primate models of pruritus and inflammation after a single dose

NHP Model of Pruritus[1]



A single dose of KPL-716 inhibited pruritic response driven by supraphysiologic levels of IL-31 for over 2 weeks at 3mg/kg

NHP Model of Inflammation[1]



Single dose of KPL-716 increased tail withdrawal latency → implicates OSMRβ in the inflammatory response

1) Unpublished data: not to be reproduced without Kiniksa's express permission

KPL-716 placebo-controlled, single-ascending-dose Phase 1a/1b study design

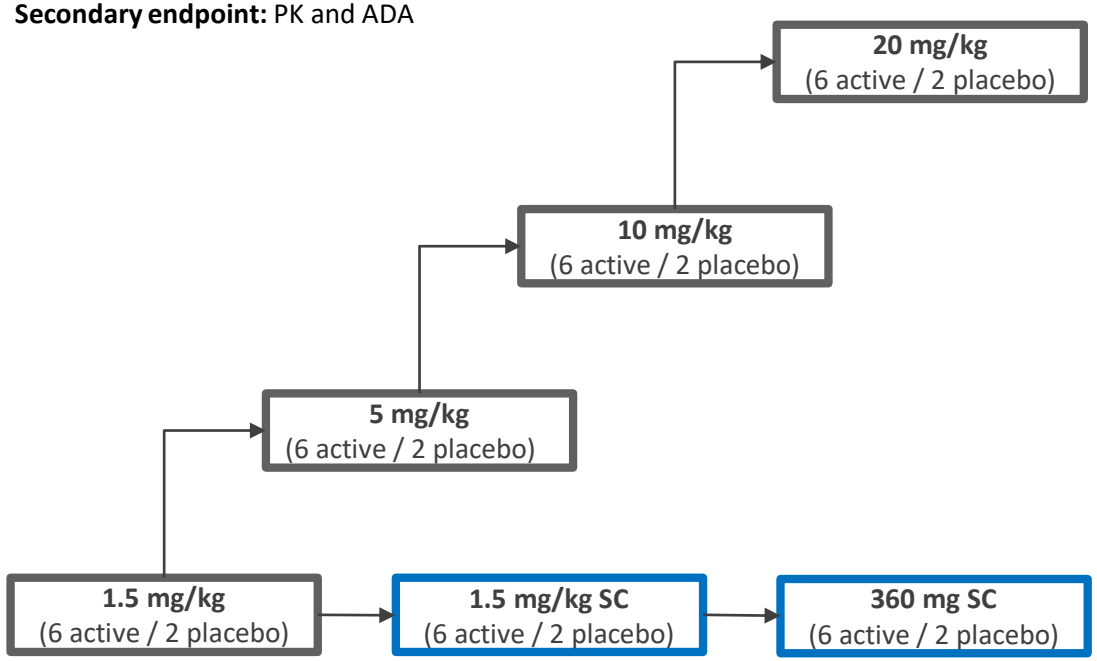
Phase 1a: Normal Healthy Volunteer (n=50)

Dose Groups

Single IV Dose

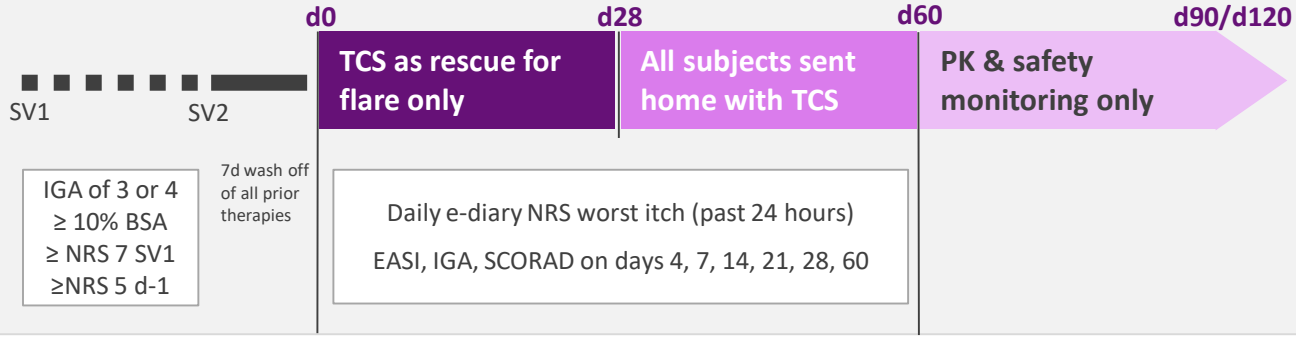
Single SC Dose

Primary endpoint: Safety and Tolerability
Secondary endpoint: PK and ADA



Phase 1b: Subjects with Atopic Dermatitis (n=32)

Patient Experience

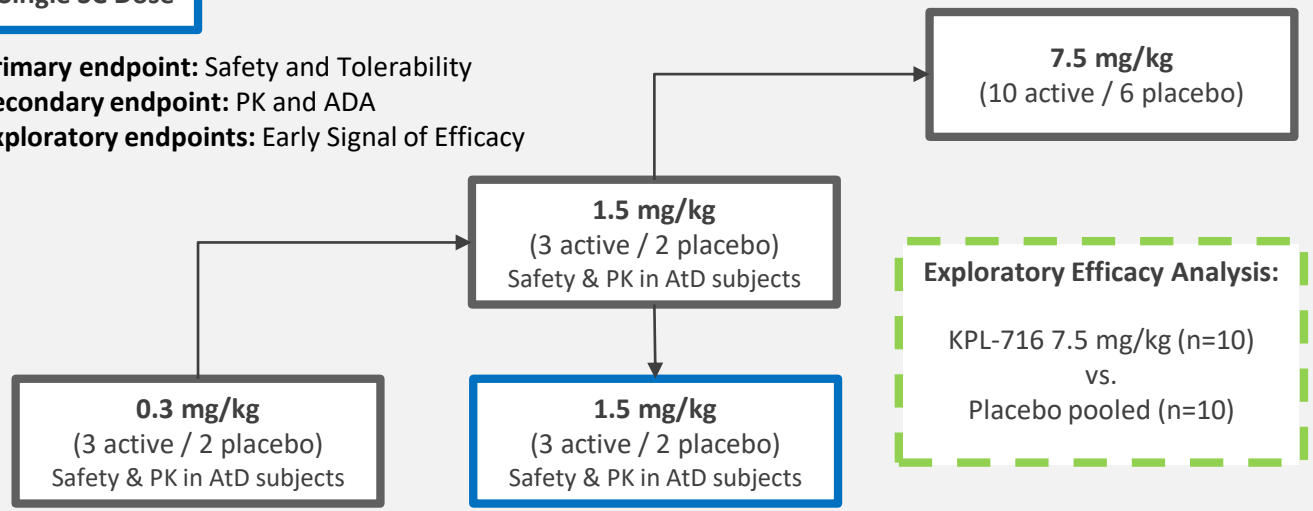


Dose Groups

Single IV Dose

Single SC Dose

Primary endpoint: Safety and Tolerability
Secondary endpoint: PK and ADA
Exploratory endpoints: Early Signal of Efficacy



Baseline parameters were balanced overall

KPL-716 recipients had more atopic dermatitis flares in the year prior to enrollment, suggesting more unstable disease at baseline compared with placebo

Baseline Demographics/Disease Characteristics: AD	KPL-716 7.5 mg/kg IV	Placebo Pooled IV
Age, mean (SD), years	29.7 (11.2)	41.7 (10.9)
Male, %	50	70
White, %	70	70
Elevated IgE, %	60	60
History of any allergic disease, %	40	60
#AD flares in past year, mean (SD)	28.1 (41.6)	3.7 (3.5)
Body surface area affected by AD, mean (SD)	24.2 (8.0)	34.1 (28.0)
Weekly average WI-NRS, mean (SD)	8.0 (1.3)	8.2 (0.7)
Total EASI, mean (SD)	19.9 (7.6)	25.3 (14.1)
Total SCORAD, mean (SD)	66.7 (10.7)	60.7 (13.7)
IGA=3, %	80	80
IGA=4, %	20	20

Baseline is defined as the last measurement prior to dosing, AD = atopic dermatitis, IV = intravenous, IGA = Investigator's Global Assessment (severity scale), WI-NRS = Worst Itch Numerical Rating Scale, EASI = Eczema Area and Severity Index, SCORAD = Scoring atopic dermatitis (severity scale)

KPL-716 was well-tolerated

- No Deaths
- No SAEs
- No Discontinuations due to AEs
- No Infusion Reactions
- No Injection Site Reactions
- No Thrombocytopenia
- No Peripheral Edema
- No Conjunctivitis
- Drug-Related Treatment Emergent Adverse Events (DR-TEAEs) infrequent and not related to dose

Normal Healthy Volunteers

AE	KPL-716 (IV)				Placebo (IV)	KPL-716 (SC)		Placebo (SC)
	1.5 mg/kg n=6	5 mg/kg n=6	10 mg/kg n=6	20 mg/kg n=6	Pooled n=8	1.5 mg/kg n=6	360 mg n=7	Pooled n=5
DR-TEAE	0	Mild headache (n=1)	0	0	0	Mild flushing (n=1)	Mild anemia (n=1)	0

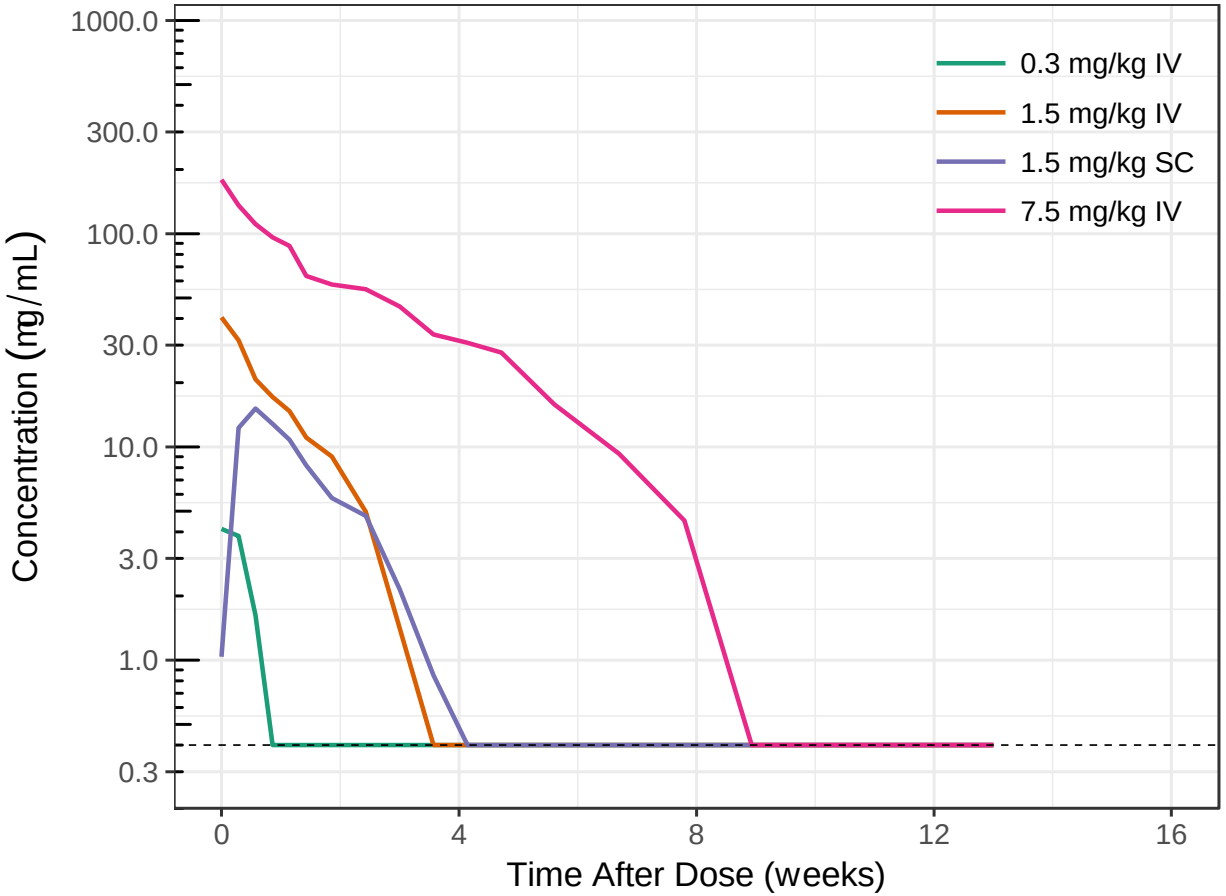
Subjects with Atopic Dermatitis

AE	KPL-716 (IV)			Placebo (IV)	KPL-716 (SC)	Placebo (SC)
	0.3 mg/kg n=3	1.5 mg/kg n=3	7.5 mg/kg n=10	Pooled n=10	1.5 mg/kg n=4	Pooled n=2
DR-TEAE†	0	Mild headache (n=1), Decreased appetite (n=1)	Moderate dizziness (n=1)	Mild somnolence (n=1)	Mild dizziness (n=1)	0
AD flare	1	0	2	3	0	0
Study day of AD flare	7	N/A	14, 20	1, 5, 45	N/A	N/A

†The only moderate DR-TEAE occurred after a protocol violation.

KPL-716 demonstrated dose-dependent elimination consistent with a target mediated drug disposition profile

7.5 mg/kg IV dose level was detectable through at least 8 weeks



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Exploratory efficacy endpoints and analysis plan

Efficacy Endpoints:

- Pruritus:
  - Weekly average of daily WI-NRS (worst itch in past 24 hours) collected by daily eDiary
  - Pruritus Visual Analog Scale, a component of SCORAD (average itch in past 3 days) collected at study visits
- Sleep loss VAS:
  - A component of SCORAD (average sleep loss in past 3 nights)
- Eczema Area Severity Index (EASI)

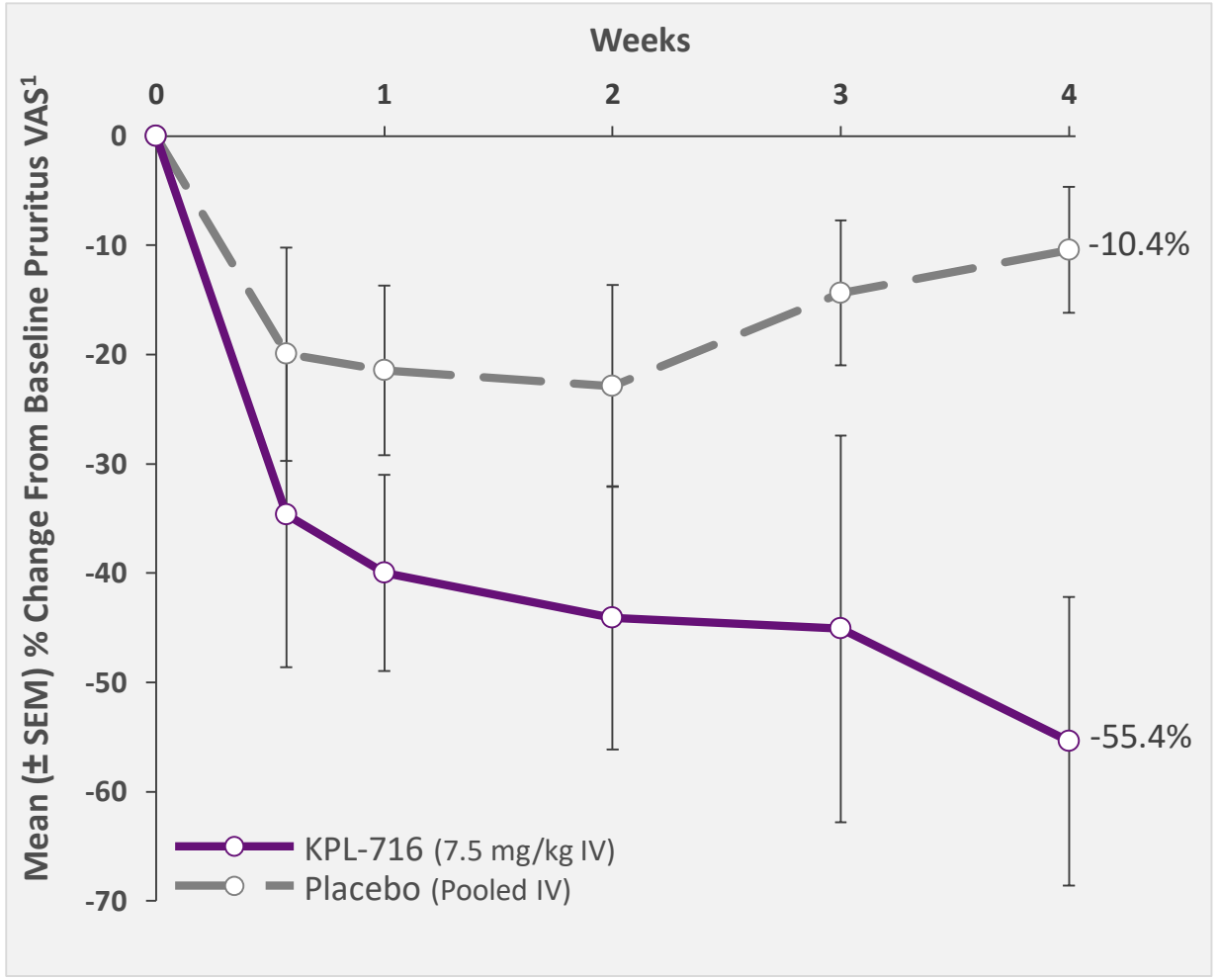
Efficacy Analysis:

- 10 KPL-716 subjects (7.5 mg/kg IV) versus 10 placebo subjects (pooled IV) from baseline to Day 28
- "Last Observation Carried Forward" approach used for data values after rescue medication administered. Subject was considered non-responder after rescue (responder analysis).
  - Two KPL-716: 2 AD flares (d15 and d21)
  - Three placebo: 2 AD flares (d3, d14), 1 anti-histamine use for upper respiratory infection (d26)
- Similar results obtained if data values after rescue medication administration were included or excluded

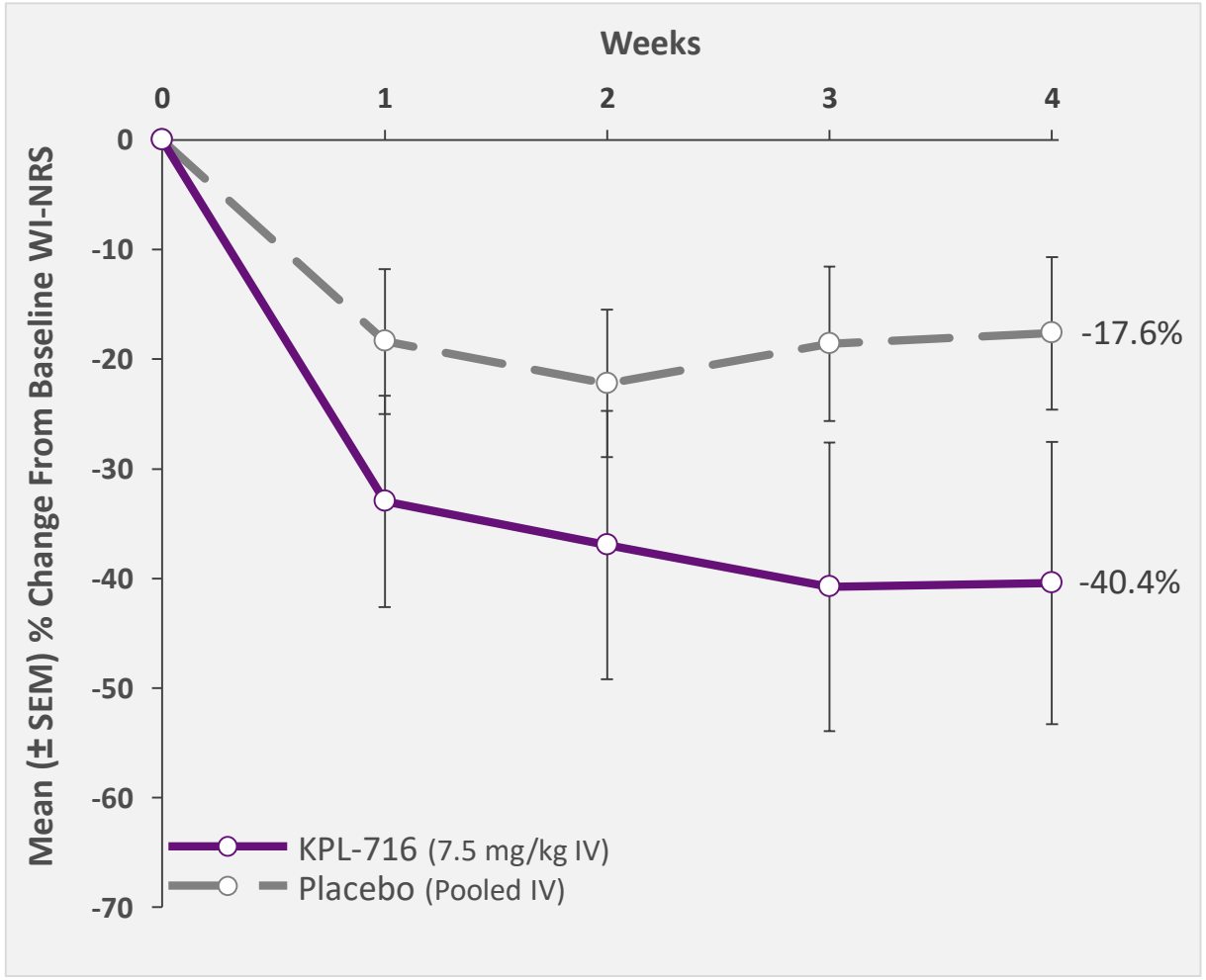
AD = atopic dermatitis, IV = intravenous, WI-NRS = Worst Itch Numerical Rating Scale, EASI = Eczema Area and Severity Index, SCORAD = Scoring atopic dermatitis (severity scale)

Single-dose of KPL-716 reduced pruritus versus placebo over the 28 day monotherapy period

Pruritus Visual Analog Scale (VAS)[1]



Weekly Average Worst Itch Numerical Rating Scale (WI-NRS)

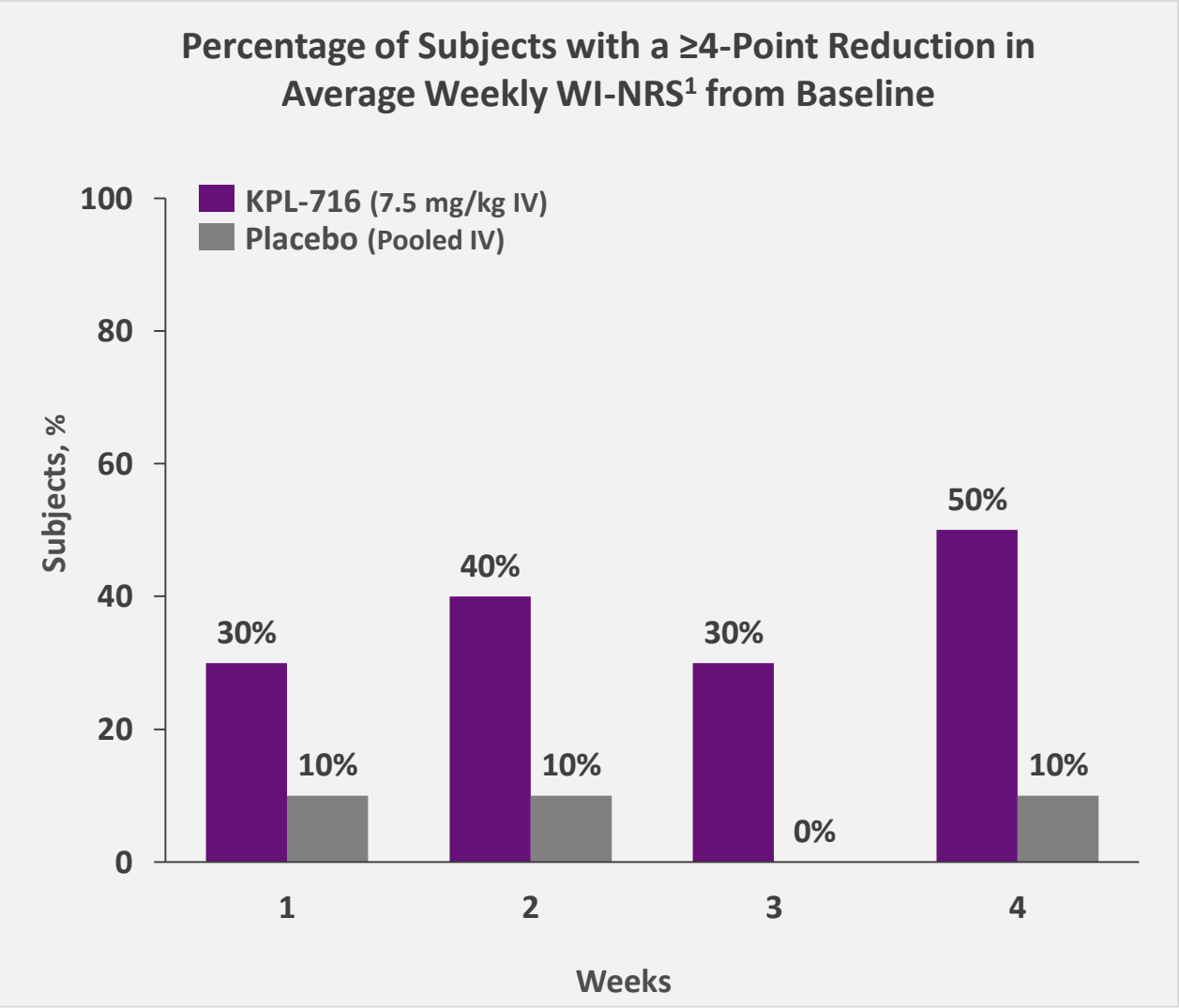


1) VAS = A component of SCORAD

SCORAD = Scoring atopic dermatitis (severity scale)

In subjects who received rescue medication, last observation was carried forward. Two KPL-716 recipients (d15, d21) and three placebo recipients (d3, d14, d26)

50% of KPL-716 recipients demonstrated a ≥ 4-point reduction in weekly average WI-NRS[1] compared to 10% of placebo at Day 28 in the absence of concomitant TCS[2]

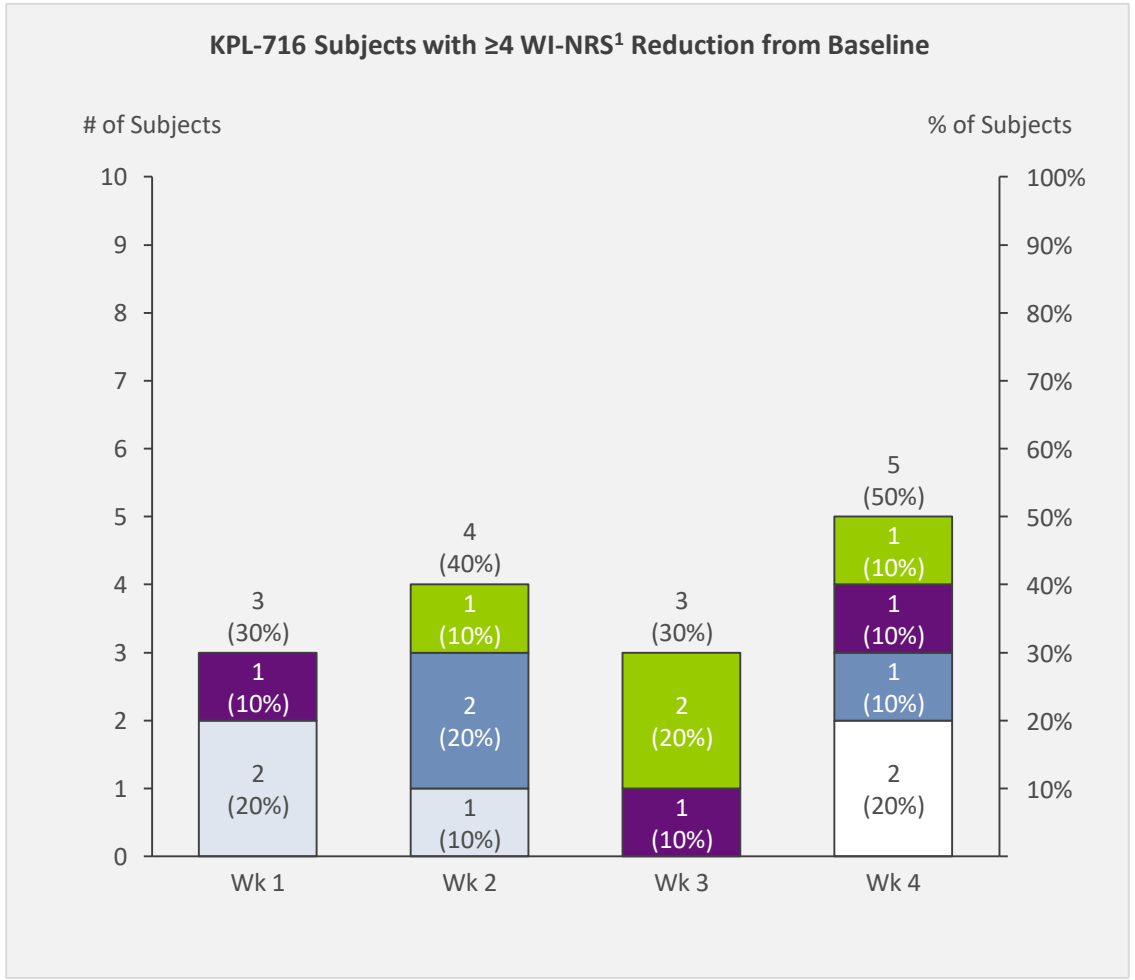


Subject was considered non-responder after rescue. Two KPL-716 recipients (d15, d 21) and three placebo recipients (d3, d14, d26).

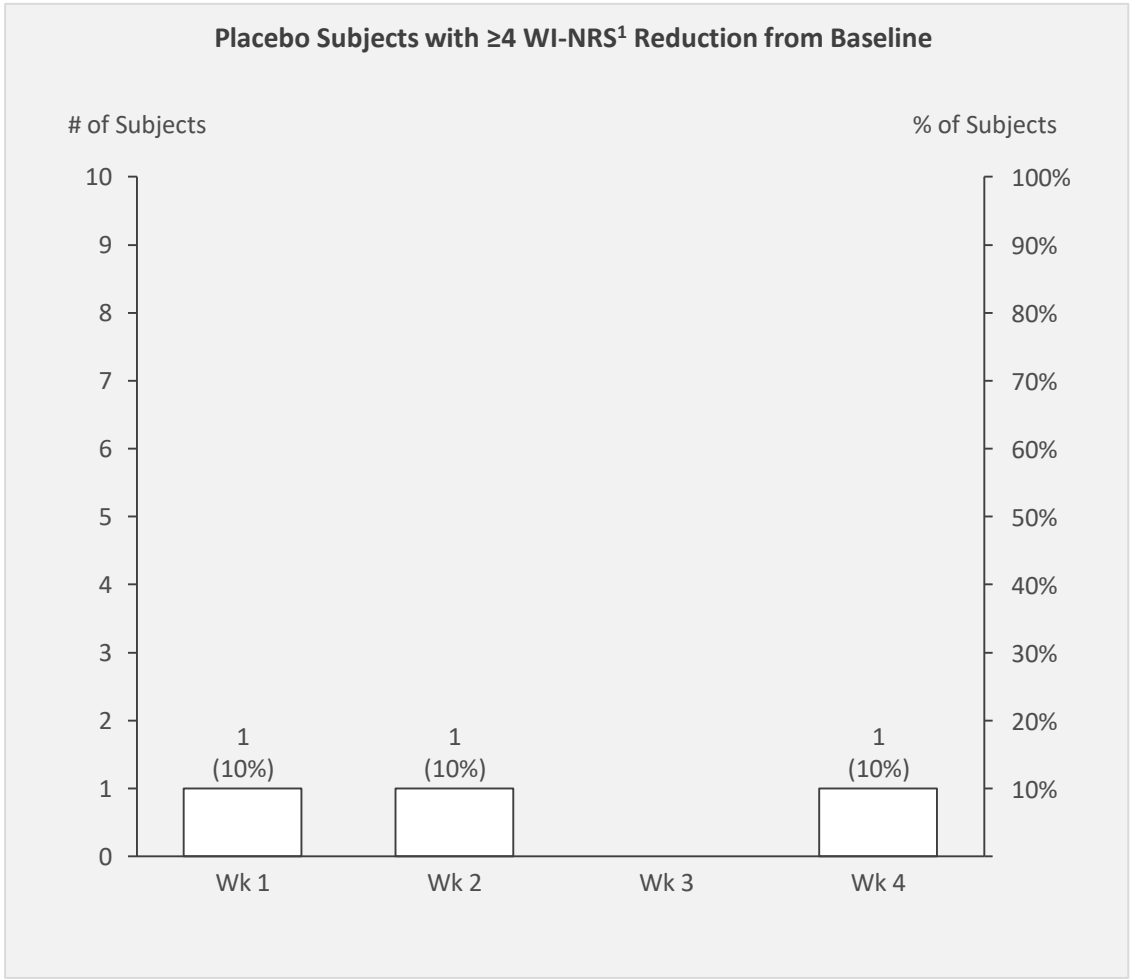
1) WI-NRS = Worst Itch Numerical Rating Scale; 2) TCS = topical corticosteroids

The maximum decrease in WI-NRS[1] at Day 28 in the absence of concomitant TCS[2] was ≥ 8-points in KPL-716 recipients compared to 4-points in placebo

KPL-716 (7.5mg/kg IV)



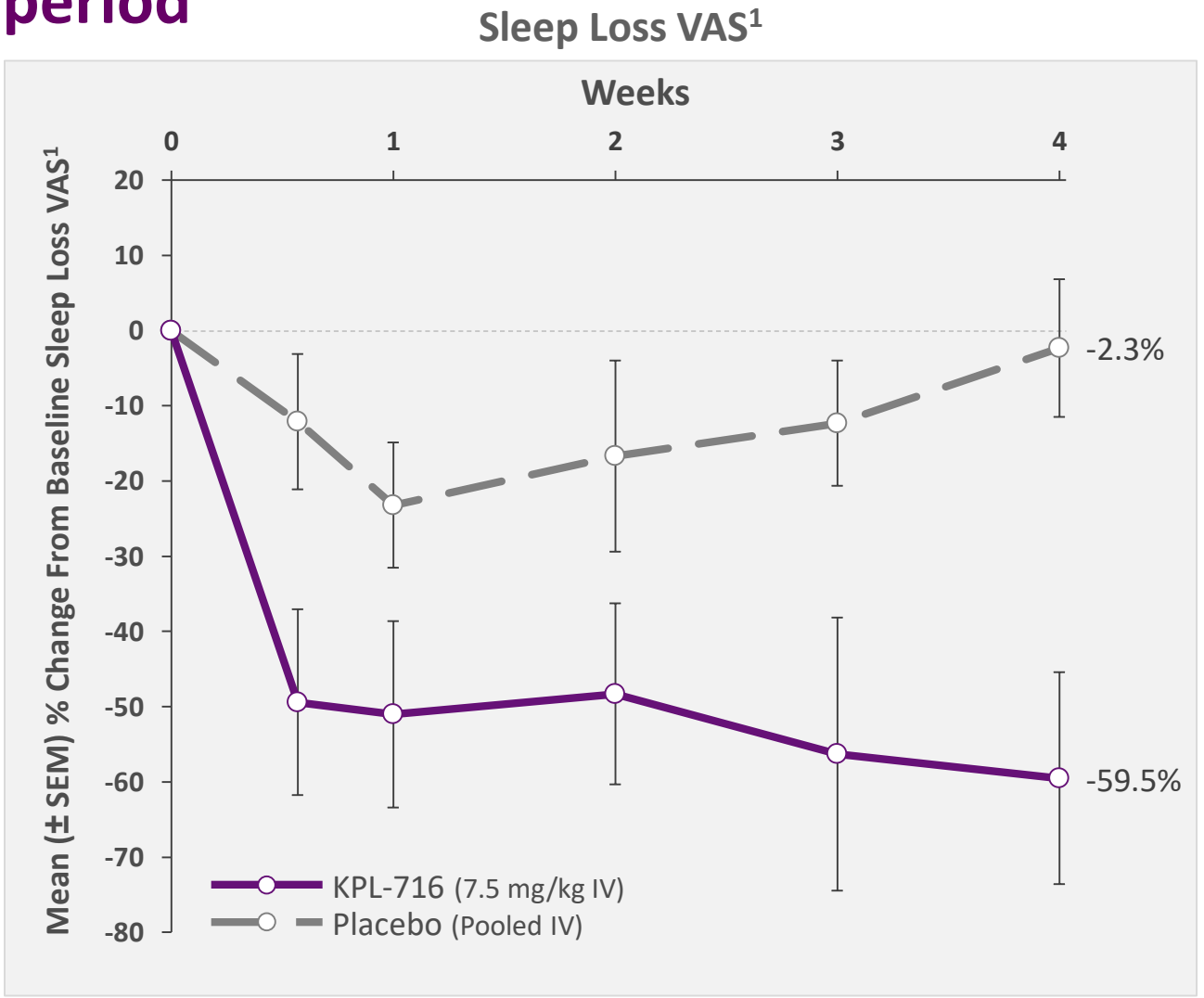
Placebo (Pooled IV)



Subject was considered non-responder after rescue. Two KPL-716 recipients (d15, d 21) and three placebo recipients (d3, d14, d26).

1) WI-NRS = Worst Itch Numerical Rating Scale; 2) TCS = topical corticosteroids

KPL-716 recipients reported reduced sleep loss VAS[1] versus placebo over the 28 day monotherapy period

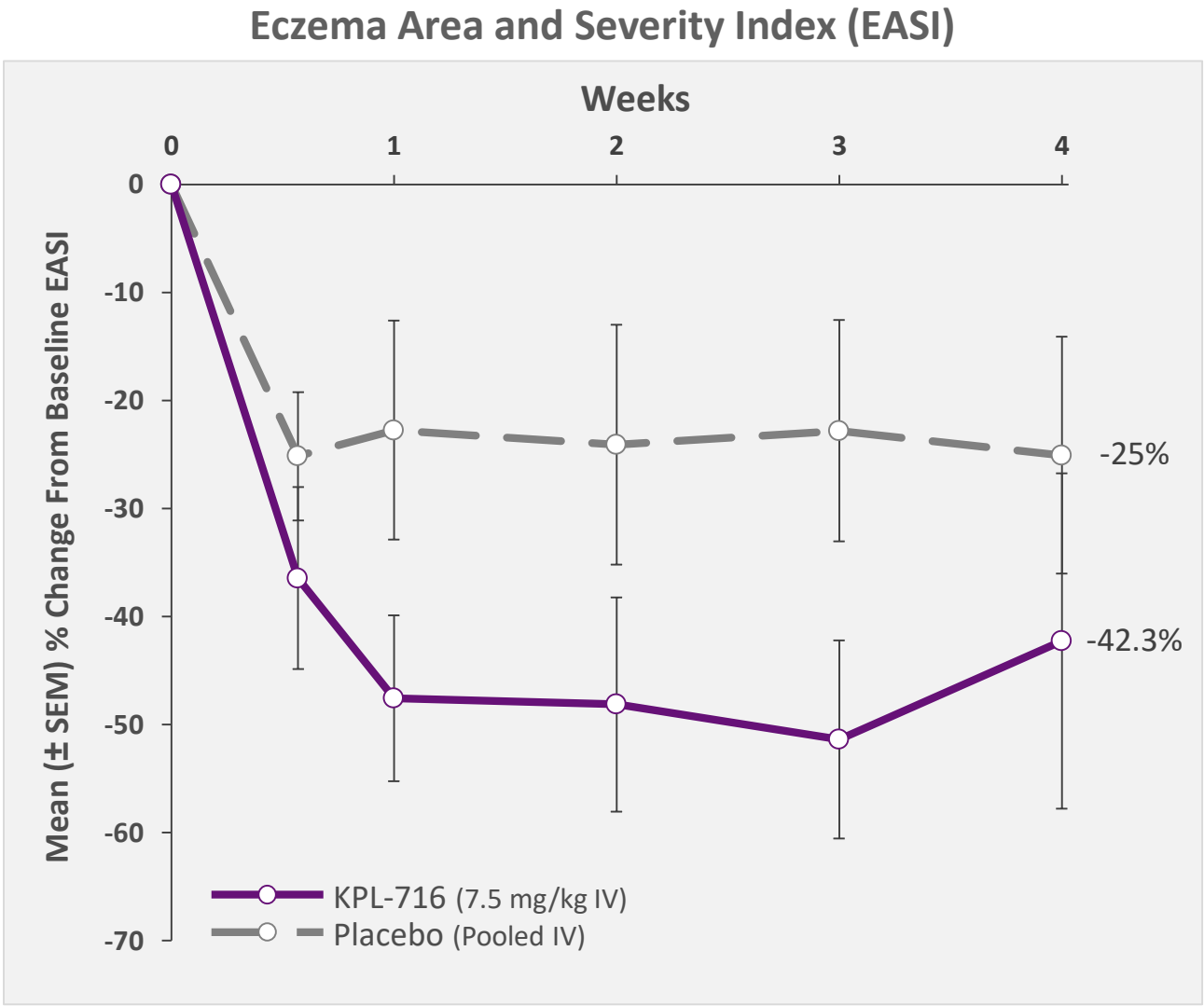


In subjects who received rescue medication, last observation was carried forward. Two KPL-716 recipients (d15, d 21) and three placebo recipients (d3, d14, d26)

1) VAS = Visual Analog Scale and a component of SCORAD.

KINIKSA

Single-dose of KPL-716 reduced atopic dermatitis disease severity versus placebo over the 28 day monotherapy period



In subjects who received rescue medication, last observation was carried forward. Two KPL-716 recipients (d15, d 21) and three placebo recipients (d3, d14, d26).

Rilonacept	Mavri	KPL-716	KPL-045	KPL-404
Approved for CAPs in US			Pre-IND	

Signal inhibitor of the CD30/CD30L interaction – a T-cell co-stimulatory receptor involved in activated T-memory function

- Biology important for activated T-memory cell function
- Comprehensive non-clinical package and POM established in models
- Favorable pharmacokinetic profile supports further development
- Preparing for IND enabling studies

Rilonacept Approved for CAPs in US	Mavri	KPL-716	KPL-045	KPL-404  Pre-IND

A central control node of T-cell-dependent, B-cell-mediated humoral adaptive immunity – designed to inhibit CD40/CD40L interaction

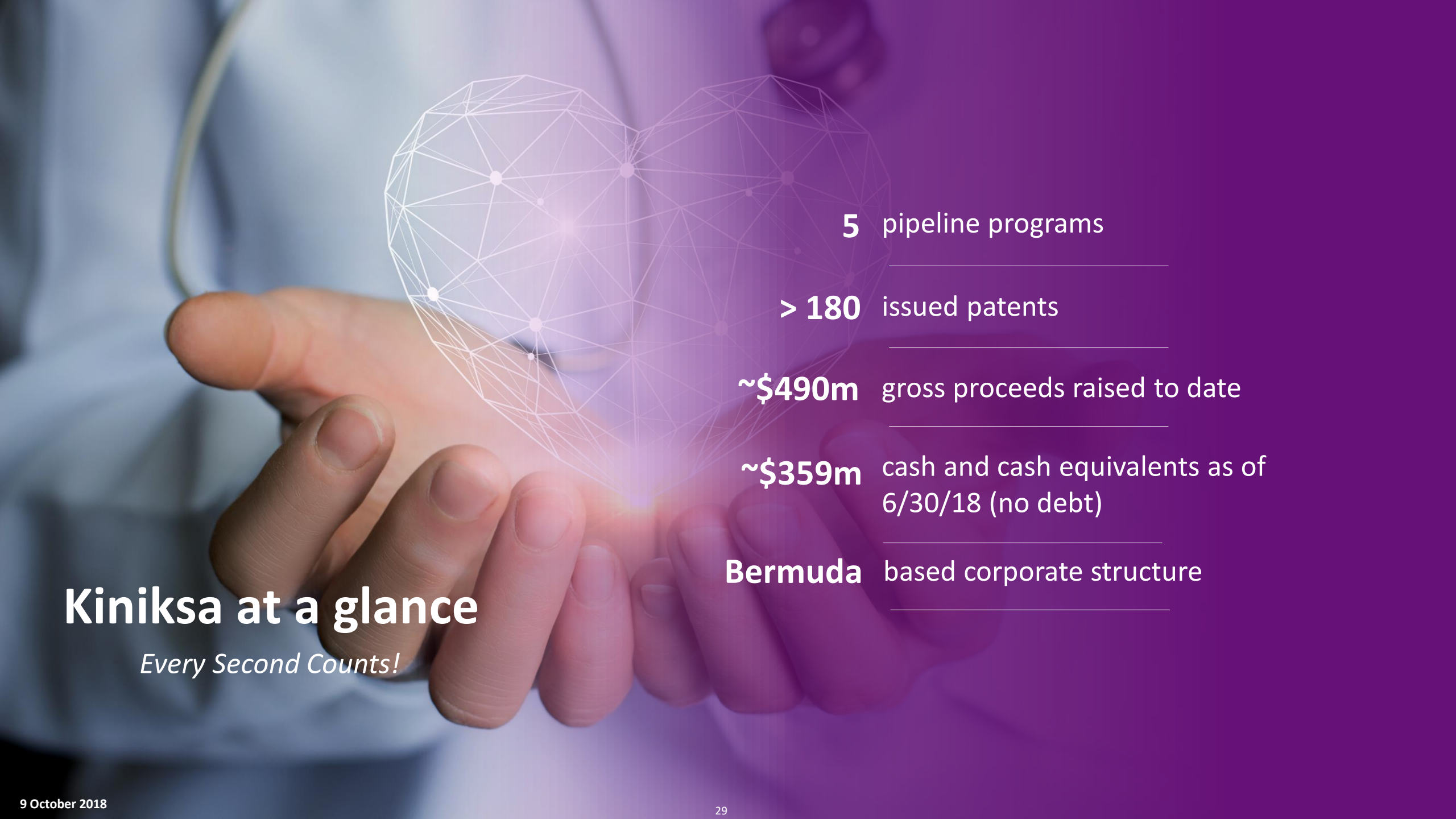
- External PoC of CD40 antagonism established by Novartis in autoimmune disease
- Favorable pharmacokinetic profile supports further development
- Preparing for IND enabling studies

A sequential pipeline across various stages of development

Designed to deliver near-, mid- and long-term value

Program & Target	Lead Indication	Phase (Preclin / 1 / 2 / 3)	Status and Anticipated Next Milestone	Rights
Rilonacept[1] IL-1α & IL-1β	Recurrent Pericarditis		• Expect to report additional data from ongoing open-label Phase 2 proof-of-concept trial in 2H 2018 • Plan to advance to a pivotal Phase 3 trial in 2H 2018	Worldwide (excluding MENA)
Mavrilimumab GM-CSFRα	Giant Cell Arteritis		• Plan to advance to a Phase 2 proof-of-concept trial in 2H 2018	Worldwide
KPL-716 OSMRβ	Prurigo Nodularis / Atopic Derm		• Plan advancement into multiple chronic pruritic diseases, including prurigo nodularis • Ongoing repeat single-dose Phase 1b trial in subjects with AtD	Worldwide
KPL-045[2] CD30L	Autoimmune		• IND filing planned for 2H 2019	Worldwide
KPL-404[2] CD40	Autoimmune		• IND filing planned for 2H 2019	Exclusive Option for Worldwide

1) Rilonacept (ARCALYST®) is approved and marketed for cryopyrin-associated periodic syndrome, in the United States by Regeneron. We will assume the rights to this indication upon receiving approval for rilonacept in the recurrent pericarditis indication; 2) We are planning IND-enabling studies for both KPL-045 and KPL-404 in T-cell-dependent, B-cell-mediated diseases, such as pemphigus/pemphigoid, myasthenia gravis, or graft versus host disease.



Kiniksa at a glance

Every Second Counts!

5 pipeline programs

> 180 issued patents

~$490m gross proceeds raised to date

~$359m cash and cash equivalents as of 6/30/18 (no debt)

Bermuda based corporate structure

Every Second Counts!™