



Every Second Counts!™

Corporate Presentation
March 2020

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 subsidiaries, collectively, unless context otherwise requires, “Kiniksa,” “we,” “us” or “our”). In some cases, you can identify forward looking statements by terms such as “may,” “will,” “should,” “expect,” “plan,” “anticipate,” “could,” “intend,” “goal,” “design,” “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; potential acquisitions and collaborations; potential value drivers; potential market opportunities and competitive position; clinical trials and other studies; timing and potential impact of clinical data; regulatory and other submissions, applications and approvals; commercial strategy and pre-commercial activities; expected cash, cash equivalents and short-term investments for FY 2019; expected funding of our operating plan; and capital allocation.

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 potential delays or difficulties with our clinical trials; potential inability to demonstrate safety or efficacy or otherwise producing negative, inconclusive or uncompetitive results; potential for changes in final data from preliminary or interim data; potential inability to replicate in later clinical trials positive results from earlier trials and studies; our reliance on third parties for manufacturing and conducting clinical trials, research and other studies; potential changes in our strategy, operating plan and funding requirements; substantial new or existing competition; and our ability to attract and retain qualified personnel. These and the important factors discussed under the caption “Risk Factors” in our Quarterly Report on Form 10-Q filed with the Securities and Exchange Commission (the “SEC”) on November 5, 2019 and other filings 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. 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.

This presentation also contains estimates, projections, and/or 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.

Building a Fully-Integrated Global Biopharmaceutical Company



Every Second Counts!™

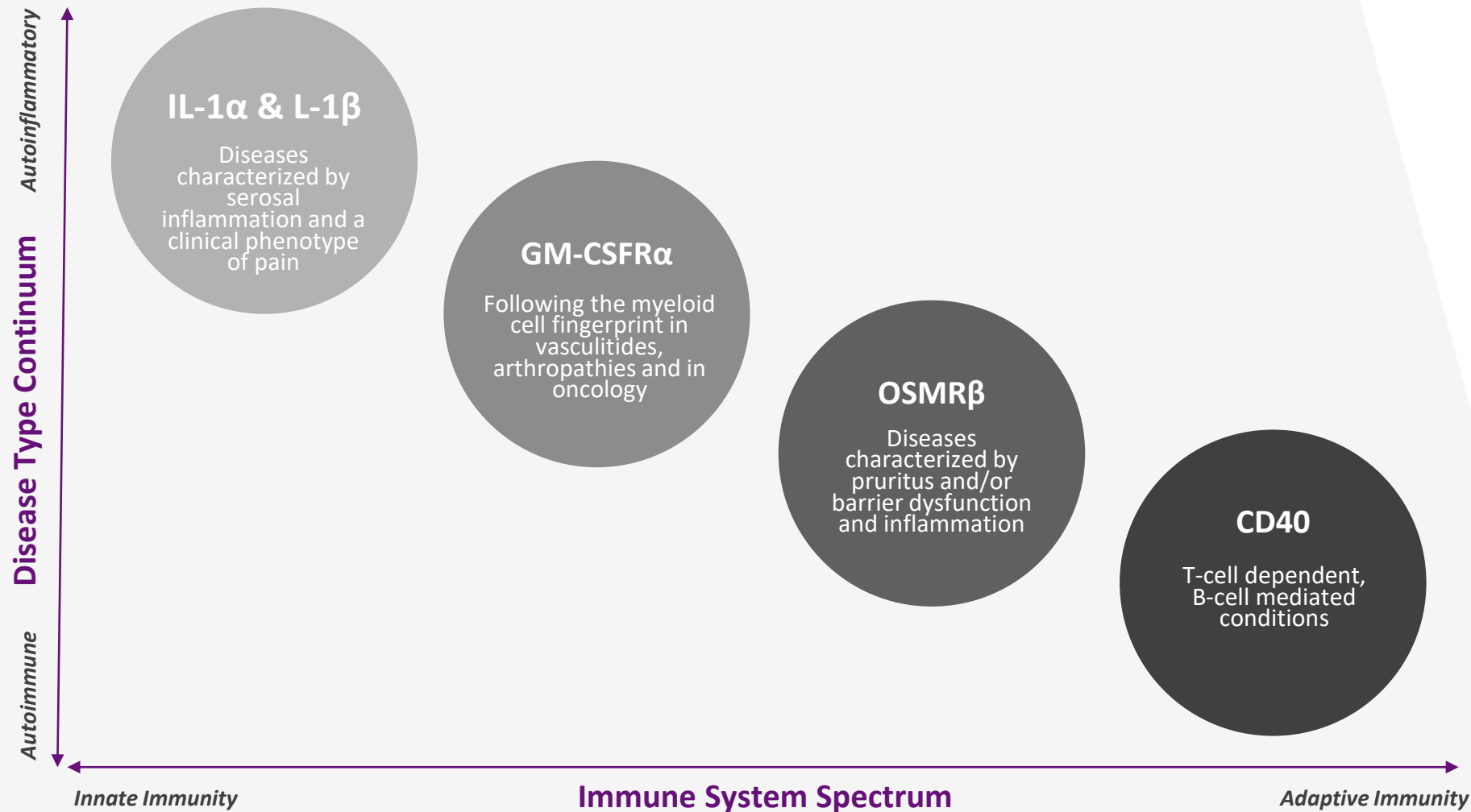
Focused on modulating different parts of the innate and adaptive immune system

Product candidates based on validated mechanisms and/or strong biologic rationale

Target underserved conditions and offer potential differentiation

Allocate capital across portfolio relative to opportunity

Development Strategy Focused on Modulating Central Nodes of the Immune System



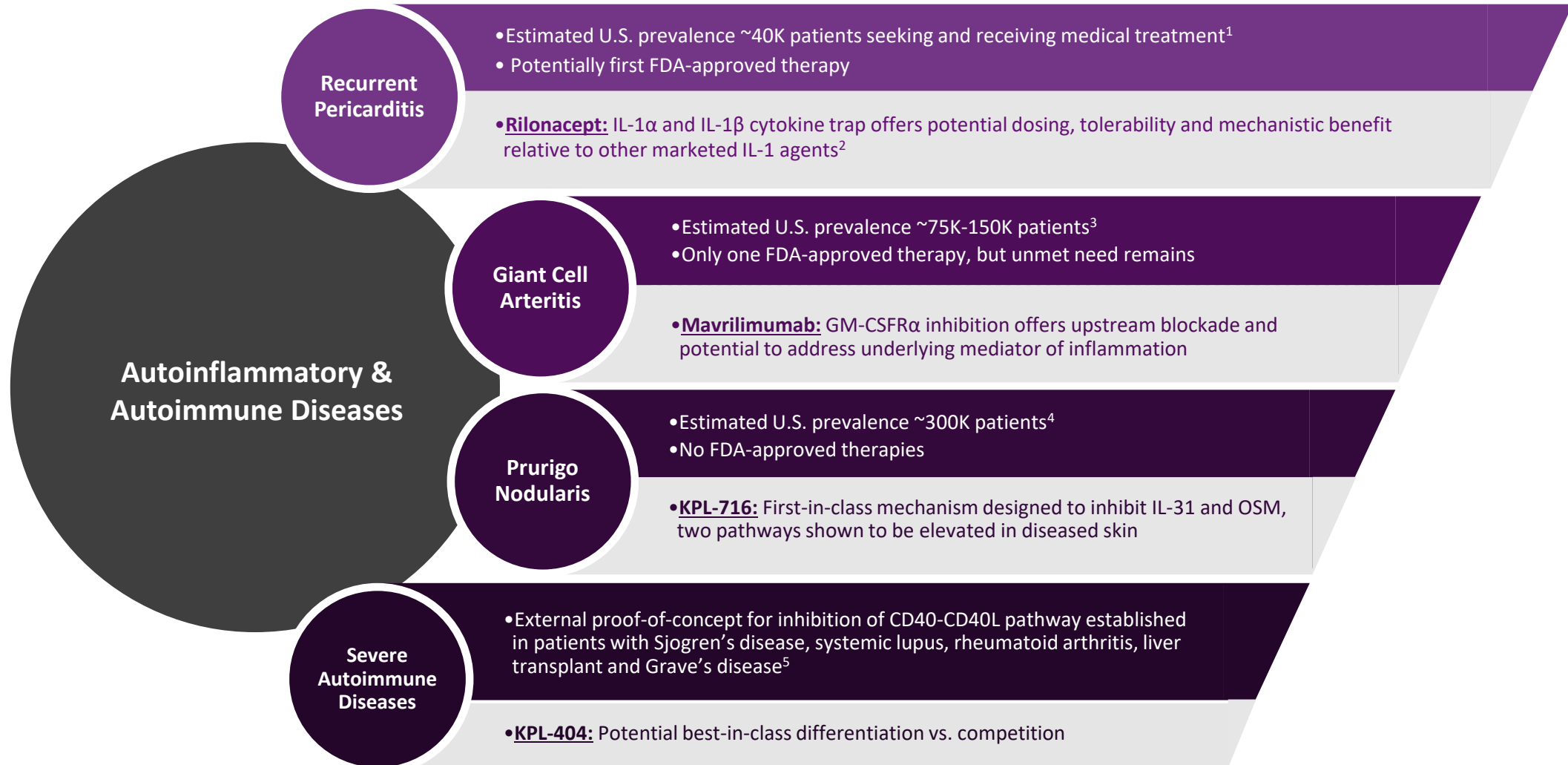
Clinical-Stage Pipeline of Immune-Modulating Product Candidates

Indication	Program & Target	Preclinical	Phase 1	Phase 2	Phase 3	Status
Recurrent Pericarditis	Rilonacept¹ IL-1 α & IL-1 β					Pivotal Phase 3 (RHAPSODY)
Giant Cell Arteritis	Mavrilimumab GM-CSFR α					Phase 2
Relapsed or Refractory Large B-Cell Lymphoma ²	Mavrilimumab GM-CSFR α					Phase 2 (Initiation Expected 2H 2020)
Prurigo Nodularis	KPL-716 OSMR β					Phase 2
Diseases Characterized by Chronic Pruritus ³	KPL-716 OSMR β					Phase 2
Severe Autoimmune Diseases	KPL-404 CD40					Phase 1

Clinical-Stage Assets Based on Validated Mechanisms and/or Strong Biologic Rationale

Mechanism of Action	Rationale	Initial Indication
Rilonacept IL-1 α and IL-1 β cytokine trap	IL-1 α and IL-1 β cytokines shown to play key role in inflammatory diseases ¹	Phase 2 data in recurrent pericarditis showed resolution of pericarditis episodes, reduction in recurrences while on treatment, and tapering/discontinuation of corticosteroids ⁶
Mavrimumab monoclonal antibody inhibitor targeting GM-CSFR α	GM-CSF is a key growth factor and cytokine in autoinflammation and autoimmunity ²	GM-CSF and GM-CSFR α are highly expressed in biopsies of giant cell arteritis patients vs. normal healthy controls ⁷
KPL-716 monoclonal antibody inhibitor targeting OSMR β	IL-31 and oncostatin M (OSM) are key cytokines implicated in prurigo nodularis ³	IL-31, OSM and OSMR β mRNA are upregulated in lesional vs. non-lesional skin biopsies of prurigo nodularis subjects ³
KPL-404 monoclonal antibody inhibitor of CD40 / CD40L interaction	CD40-CD40L interaction is an attractive mechanism for targeting T-cell dependent, B-cell-mediated autoimmune diseases ^{4,5}	External proof-of-concept for inhibition of pathway has been established in a broad range of autoimmune diseases ⁸

Clinical-Stage Assets Target Underserved Diseases and Offer Potential Differentiation



Multiple Clinical Data Readouts Expected in 2020

KPL-716 – Phase 2a (monoclonal antibody inhibitor targeting OSMR β)	Prurigo Nodularis	By the End of April 2020
KPL-716 – Phase 2 (monoclonal antibody inhibitor targeting OSMR β)	Diseases Characterized by Chronic Pruritus¹	1H 2020
Rilonacept – Pivotal Phase 3 (IL-1 α and IL-1 β cytokine trap) 	Recurrent Pericarditis	2H 2020
Mavrilimumab – Phase 2 (monoclonal antibody inhibitor targeting GM-CSFR α)	Giant Cell Arteritis	2H 2020
KPL-404 – Phase 1 (monoclonal antibody inhibitor of CD40-CD40L interaction)	First-in-Human w/ Antigen Challenge TDAR	2H 2020



Rilonacept

Mavrilimumab

KPL-716

KPL-404

First Indication¹	Recurrent Pericarditis: Painful and debilitating autoinflammatory cardiovascular disease
Mechanism of Action²	IL-1α and IL-1β cytokine trap
Scientific Rationale²	IL-1α and IL-1β are cytokines shown to play key role in inflammatory diseases
Prevalence³	~40k prevalent in U.S.; addressable opportunity of ~14k in U.S.
Competition⁴	No FDA-approved therapies for recurrent pericarditis
Status	Breakthrough Therapy designation granted; enrollment target achieved in pivotal Phase 3 clinical trial
Economics	Regulatory milestones; 50/50 profit split upon commercialization
Rights	BLA transfers to Kiniksa after receipt of positive Phase 3 clinical data; upon approval Kiniksa has the rights to recurrent pericarditis worldwide (excluding MENA)



Recurrent Pericarditis Patients Currently Have Limited Treatment Options

Patients with pericarditis are deemed recurrent after symptom-free period of 4-6 weeks

Recurrent Pericarditis

1st Line

NSAID +/- Colchicine

2nd Line

Systemic Corticosteroids

Steroid-Sparing
Opportunity

3rd Line

IVIG, Azathioprine, Methotrexate, or Anakinra (off-label)

Refractory Patients

4th Line

Pericardiectomy

Key Areas of Unmet Need in Patients with Recurrent Pericarditis

Recurrent pericarditis episodes: painful, debilitating and disruptive to quality of life

**Resolution of
Episodes**

*~50% Have Symptoms
that Persist for >4 wks*

**Prevention of
Future Episodes¹**

*50% Annual
Recurrence Rate*

**Steroid-Sparing
Disease Control**

*Unable to Wean
off Steroids*

Quality of Life

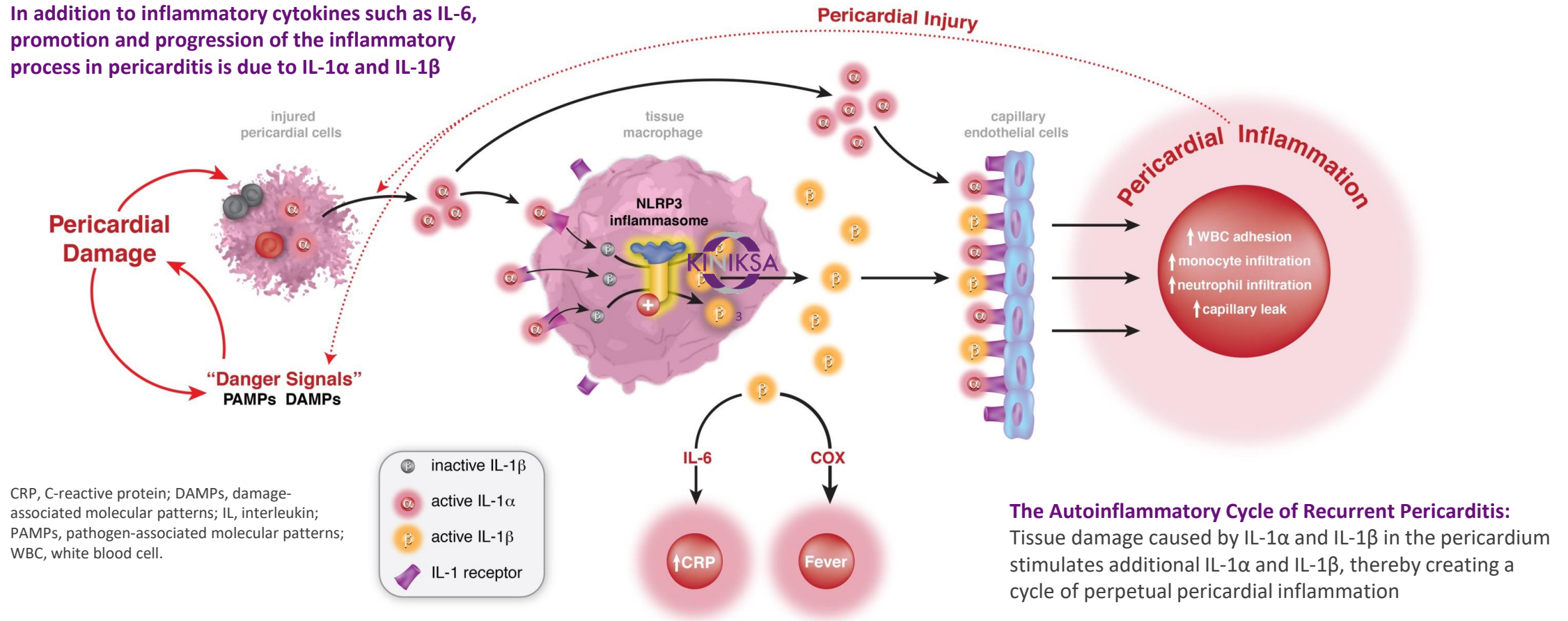
*Increased Rates of
Anxiety and Depression*

“ The worst thing about pericarditis is its unpredictability and its chronicity. It’s a permanent condition, so it has the potential to impact everything...work, exercise, family plans, travel. ”

- Patient quote, 2019

Role of IL-1 α and IL-1 β in the Autoinflammatory Cycle of Recurrent Pericarditis

In addition to inflammatory cytokines such as IL-6, promotion and progression of the inflammatory process in pericarditis is due to IL-1 α and IL-1 β



Brucato A, et al. Int Emerg Med 2018 <https://doi.org/10.1007/s11739-018-1907-x>
Dinarello CA, et al. Nat Rev Drug Discov 2012;11:633-652

Clinical Development Plan for Rilonacept in Recurrent Pericarditis

Designed to generate data on clinically meaningful outcomes

Phase 2

- Open-label, 5-part clinical trial with rilonacept in range of recurrent pericarditis populations
- Provided first evidence that rilonacept treatment improved clinically meaningful outcomes in study¹
- Rilonacept was well-tolerated in study, with safety profile consistent with FDA-approved label for CAPS²

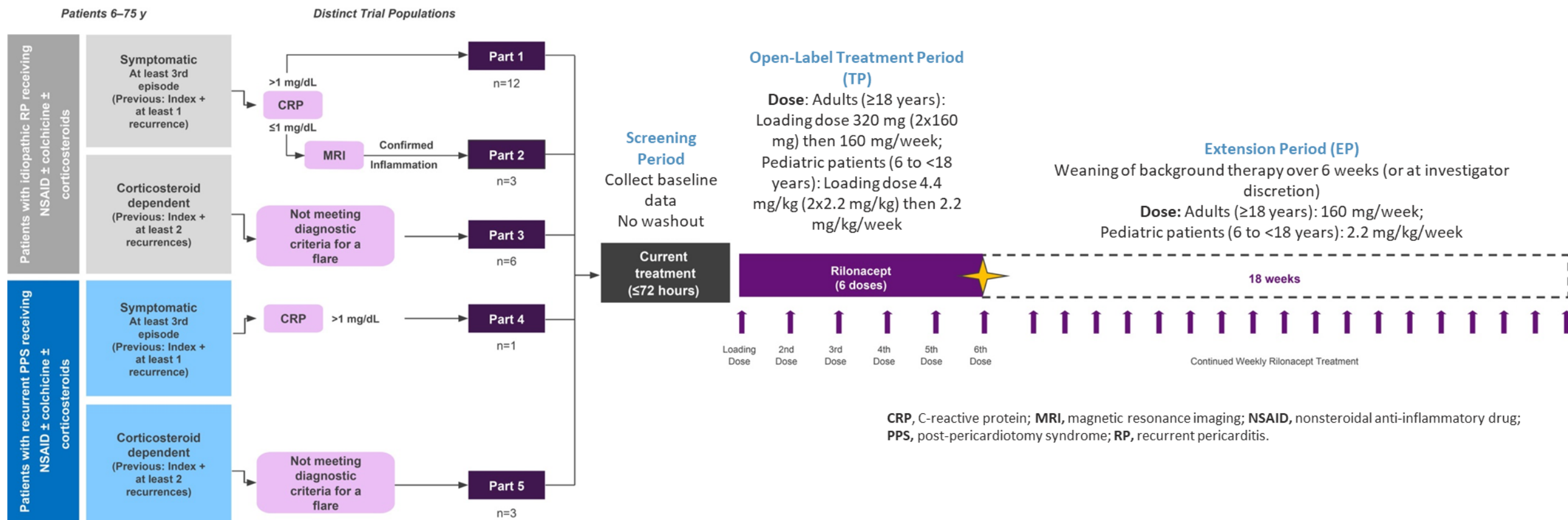
Completed

Phase 3 (RHAPSODY)

- **Enrollment completed**
- Pivotal clinical trial of rilonacept for treatment of recurrent pericarditis
- 24-week, double-blind, placebo-controlled, randomized-withdrawal (RW) study with open-label extension
- Primary efficacy endpoint is time-to-first-adjudicated pericarditis-recurrence in the RW period
- Continuing to enroll patients for a limited period to facilitate the accrual of primary efficacy endpoint events

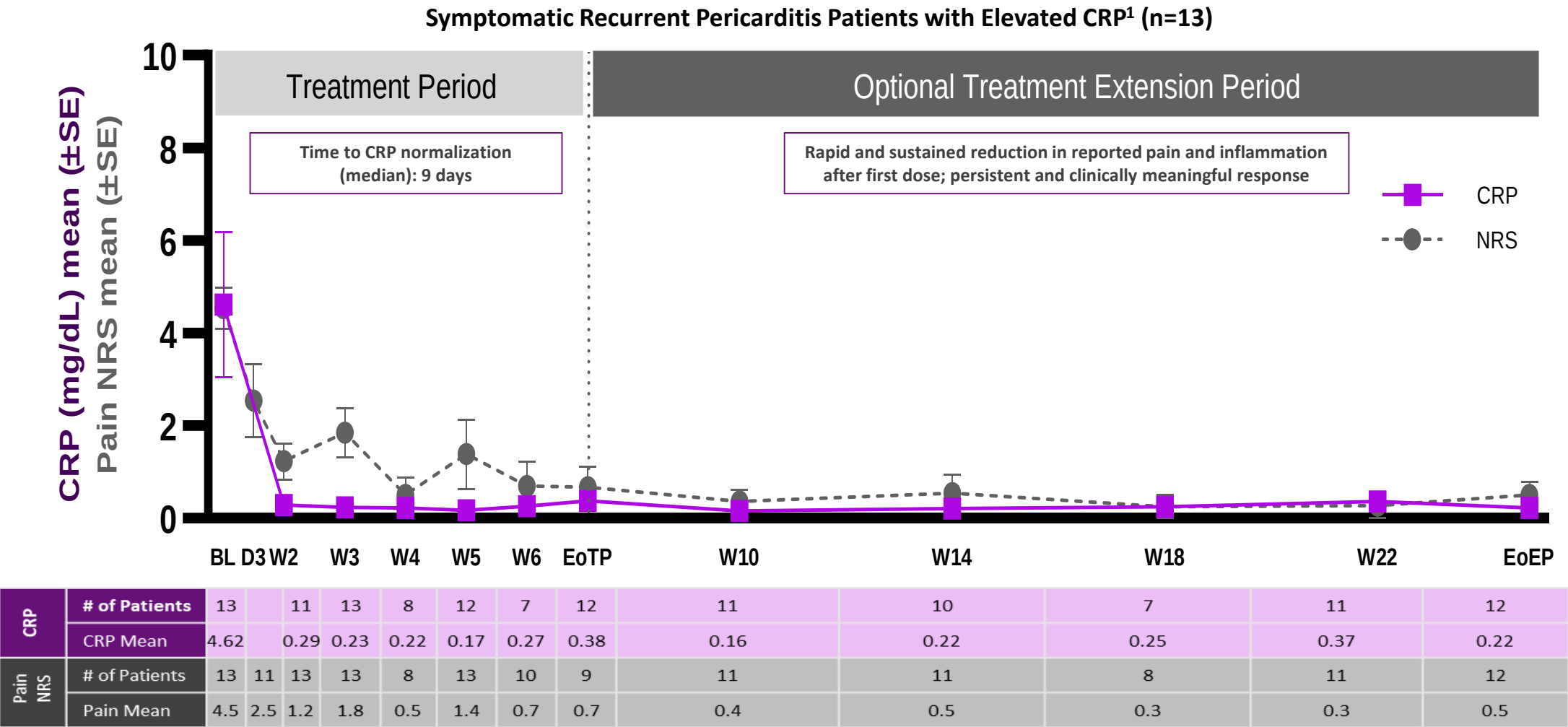
Top-line data expected 2H 2020

Open-Label Phase 2 Clinical Trial of Rilonacept in Pericarditis Populations



Phase 2 Rilonacept Data

Resolution of pericarditis episodes in symptomatic patients (parts 1 and 4)



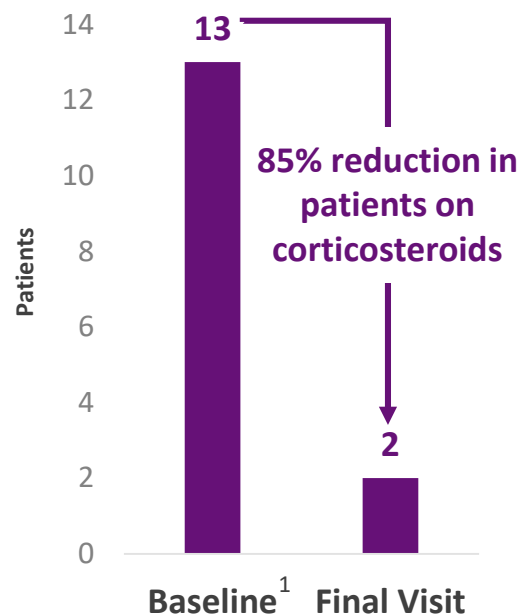
15 1) Patients with elevated CRP and symptomatic disease (Parts 1 and 4) are most representative of real-world recurrent pericarditis. Inclusion and exclusion criteria for the ongoing Phase 3 study RHAPSODY align with this patient population (clinicaltrials.gov/NCT03737110). EoTP = end of treatment period; EoEP = end of extension period; CRP = C-Reactive Protein; NRS = Numeric Rating Scale



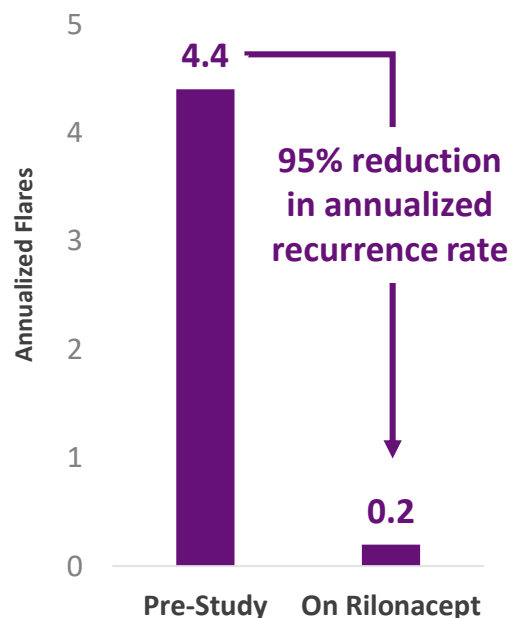
Phase 2 Rilonacept Data

Discontinuation of corticosteroids, decrease in incidence of pericarditis episodes while on treatment and improvement in quality of life scores

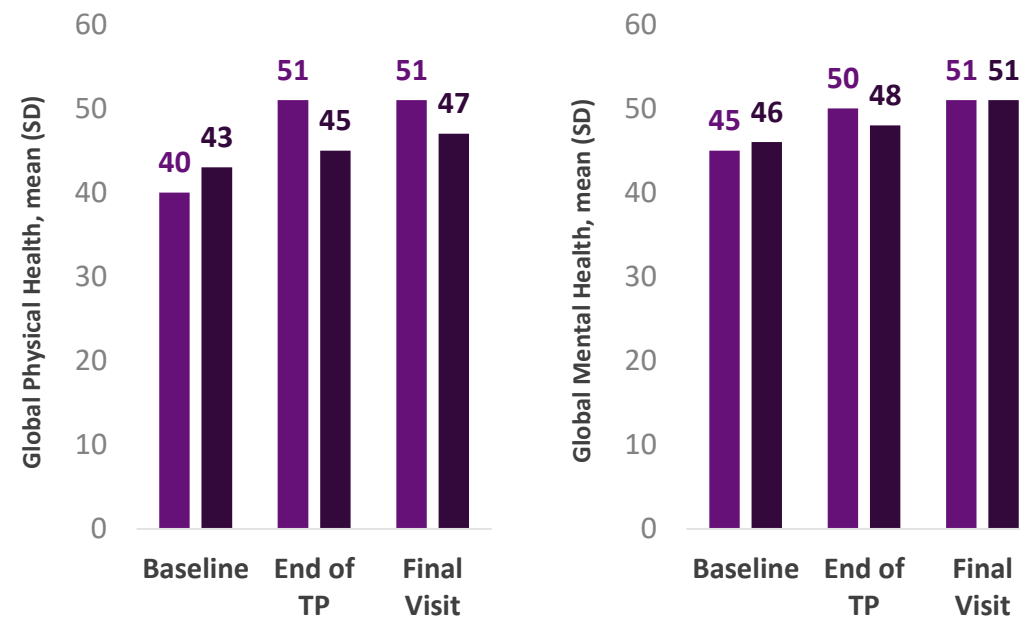
Discontinuation of Corticosteroids Without Pericarditis Recurrence



Decrease in Annualized Incidence of Pericarditis Episodes While on Treatment

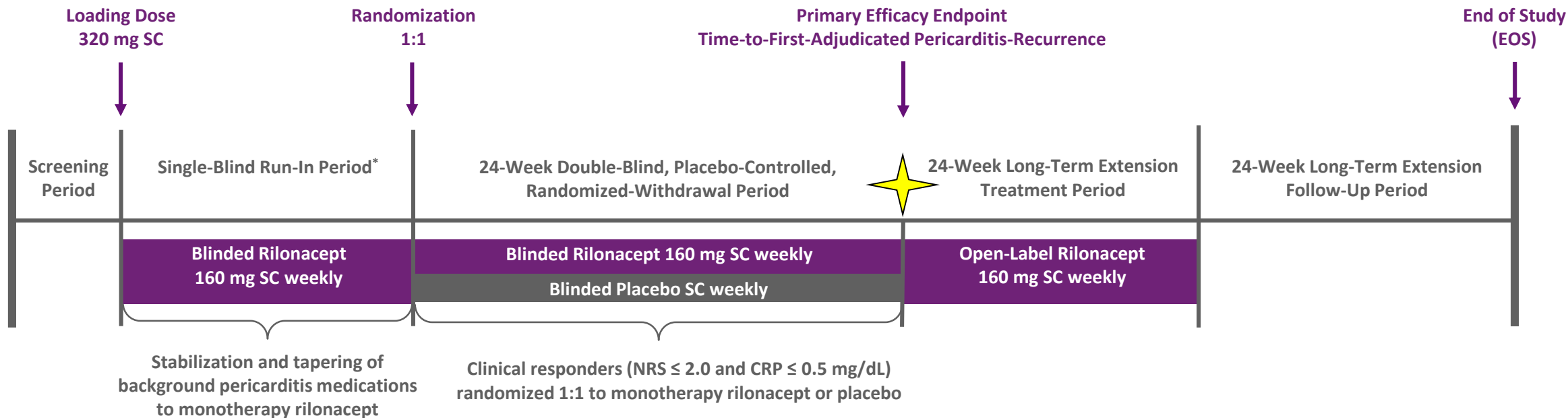


Improved Quality of Life Scores²



Idiopathic or PPS: ■ Active³ (n=16) ■ CS-dependent⁴ (n=9)

Pivotal Phase 3 Clinical Trial of Rilonacept for Recurrent Pericarditis



Inclusion Criteria:

- All etiologies except infection and malignancy
- Present at screening with at least a third pericarditis episode, defined as at least 1 day with NRS pain of ≥ 4 and CRP value ≥ 1 mg/dL within the 7-day period prior to first study drug administration
- Concomitant NSAIDs and/or colchicine and/or oral corticosteroid treatment in any combination

Primary Outcome Measure (24 weeks):

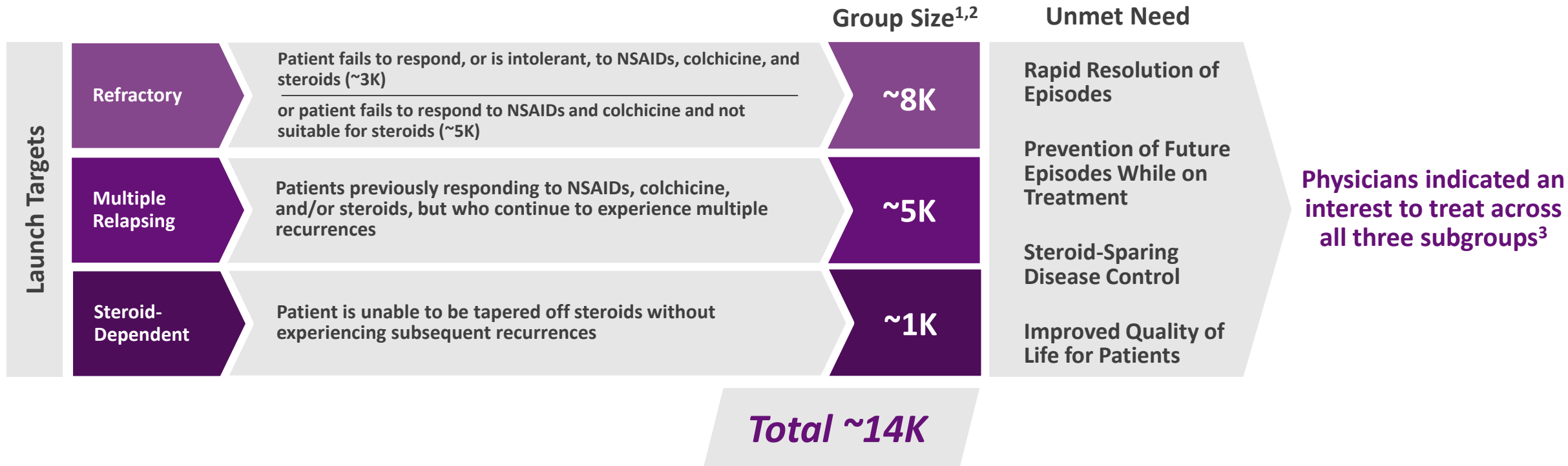
- Time-to-first-adjudicated pericarditis-recurrence in the RW period

Secondary Outcome Measures (24-weeks):

- Proportion of subjects who maintained Clinical Response
- Percentage of days with no or minimal pain
- Proportion of subjects with absent or minimal pericarditis symptoms
- Proportion of subjects with adverse events

Recurrent Pericarditis U.S. Prevalence Estimated to be ~40K Patients

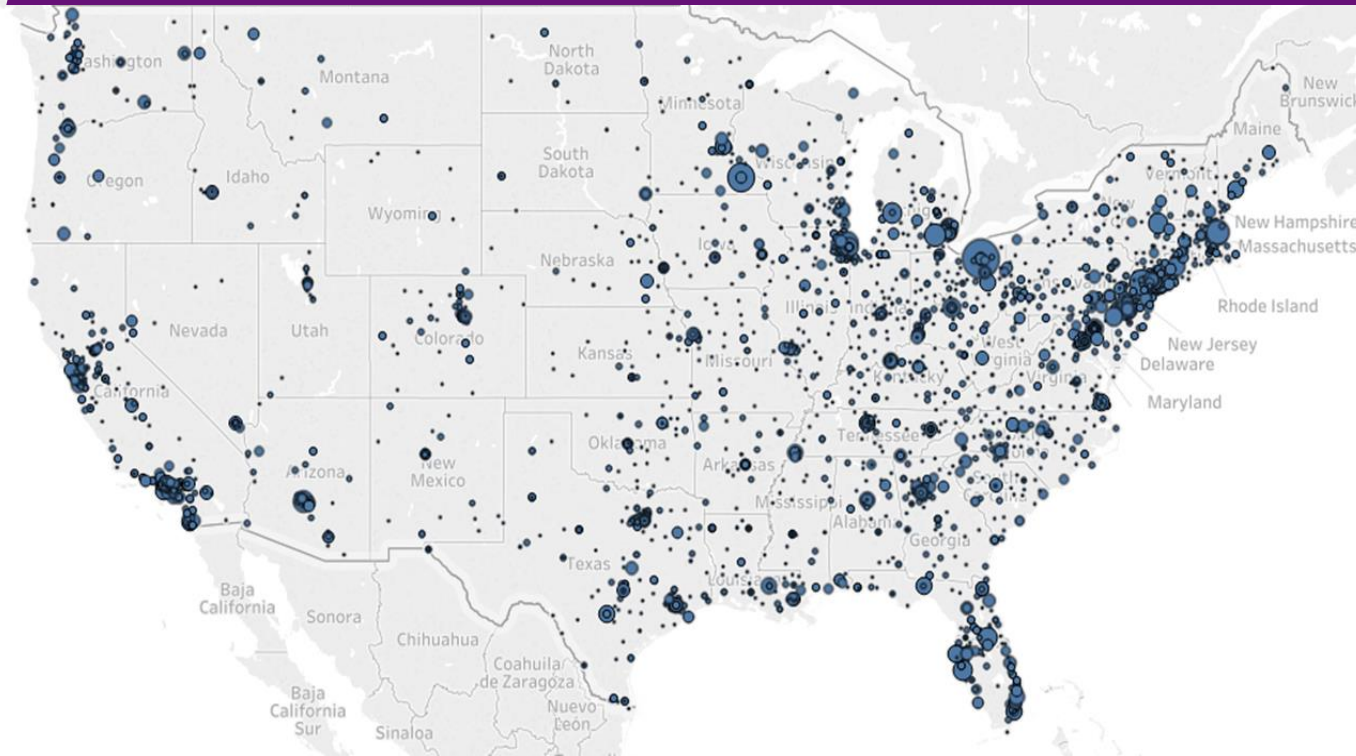
Addressable U.S. opportunity for rilonacept estimated to be ~14K patients



Commercial Strategy

Planned launch focused on high-volume specialists

Recurrent Pericarditis Patient Volume by Account



Commercialization Plan Linked to Opportunity

- Specialty cardiology sales force of ~30 reps to call on high volume specialists
- Supported by current MSL team
- Efficient digital marketing to educate lower volume specialists
- Robust patient services capabilities to maintain appropriate patients on therapy
- Duration of therapy expected to be at least 6-12 months
- Pricing in-line with high unmet need in rare disease

Mavrilimumab – Phase 2

Rilonacept

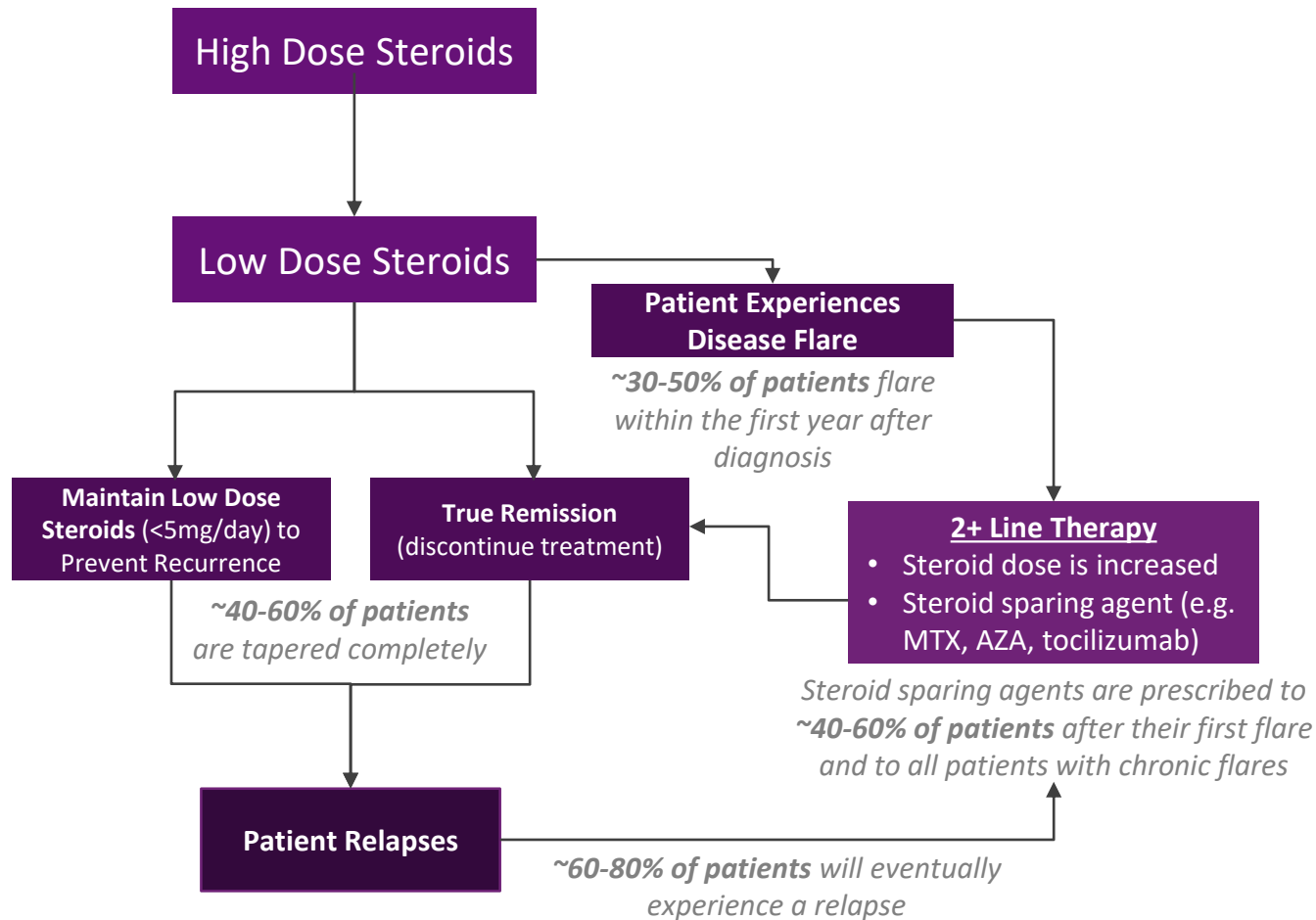
Mavrilimumab

KPL-716

KPL-404

First Indication	Giant Cell Arteritis: Chronic inflammatory disease of medium-to-large arteries
Mechanism of Action¹	Monoclonal antibody inhibitor targeting GM-CSFR α
Scientific Rationale^{2,3}	Reported data implicate the GM-CSF is key growth factor and cytokine in GCA
Prevalence⁴	~75k - 150k prevalent in U.S.; similar prevalence in other major markets
Competition⁵	Only one FDA-approved therapy for GCA and unmet needs remain
Status	Enrollment target achieved in global Phase 2 clinical trial; collaboration with Kite Gilead in R/R LBCL ⁶
Economics	Clinical, regulatory and sales milestones; tiered royalty on annual net sales
Rights	Worldwide

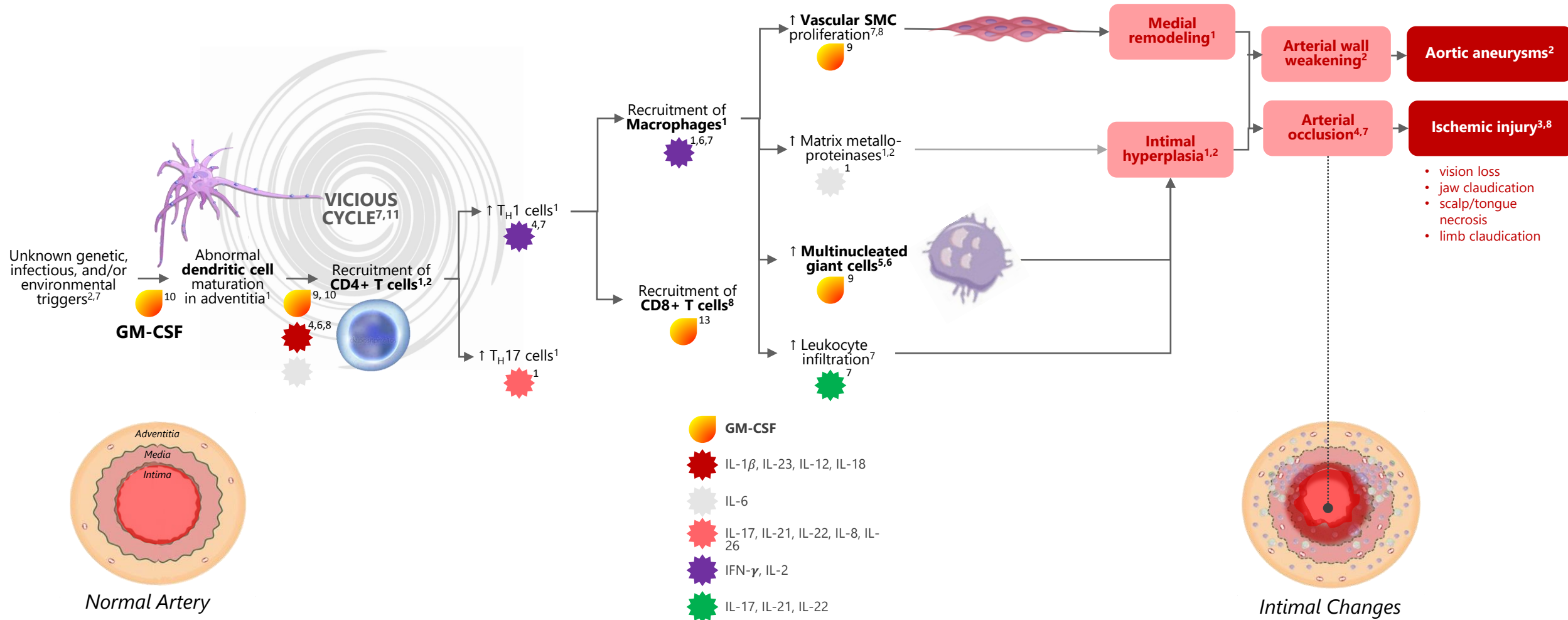
Current Treatment Paradigm for GCA Involves High-Dose Steroids Upon Clinical Suspicion



Treatment Approach:

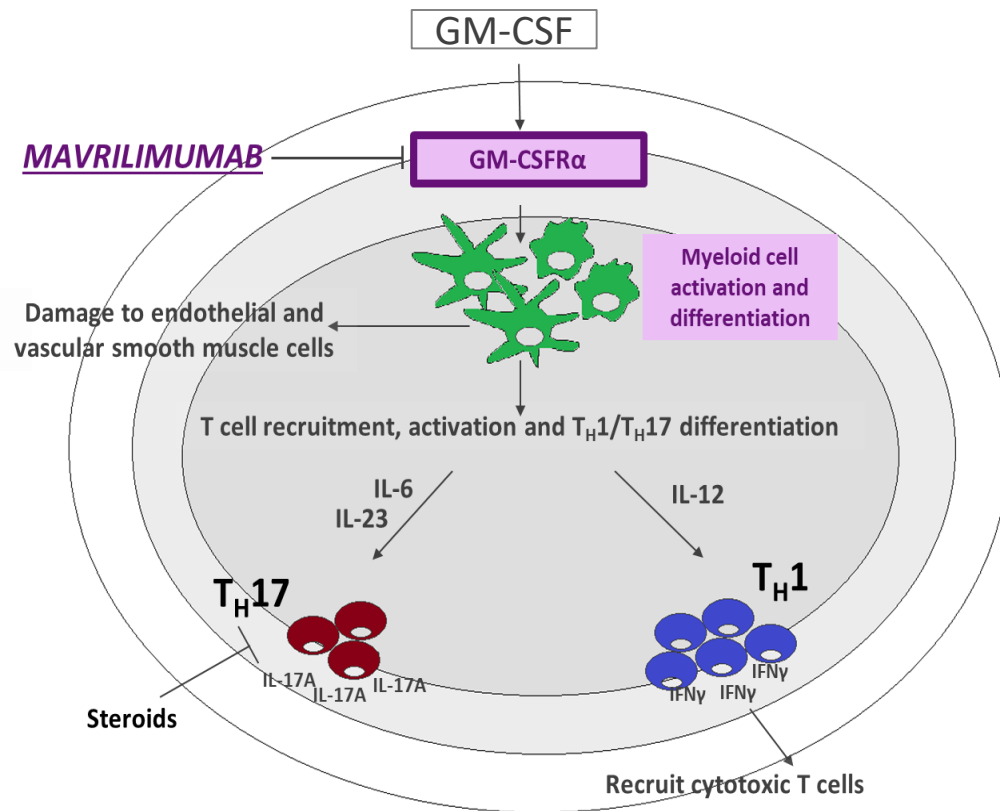
- All patients receive **high-dose steroids**, which are effective at preventing disease related complications; however, **they may lead to life altering side-effects** like osteoporosis and diabetes
- A few treaters initiate **steroid sparing agents** early in the treatment paradigm, relying on them more for the chronic treatment of GCA
- Others treat GCA in more of a stepwise fashion, adding new agents on top of steroids only following disease flares/relapse

Central Role of GM-CSF in Pathophysiology of Giant Cell Arteritis

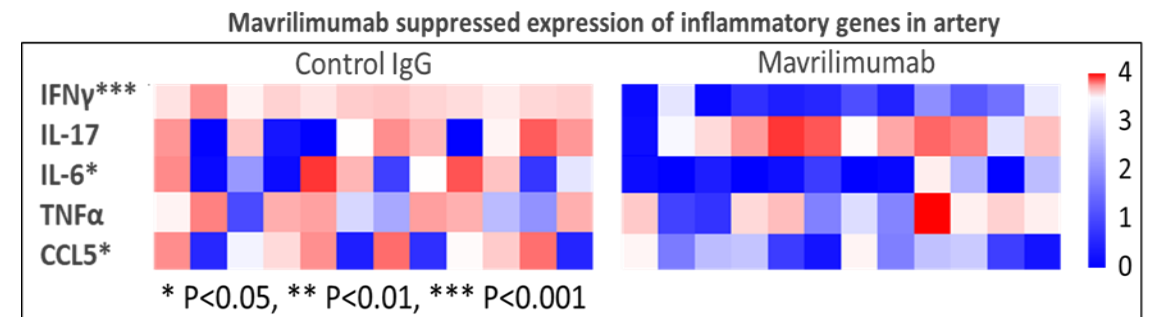
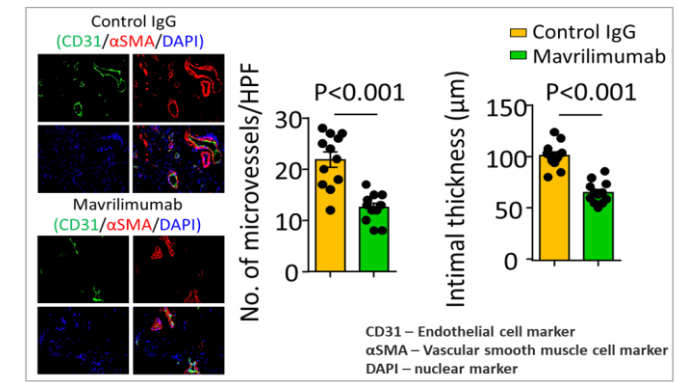
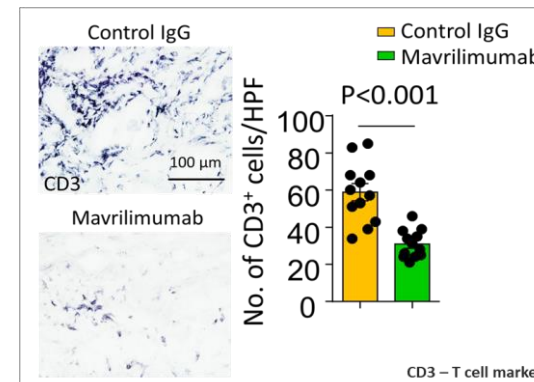


Preclinical Data Support the Mechanistic Rationale of Targeting GM-CSF in GCA

GM-CSF and its receptor, GM-CSFR α , shown to be elevated in GCA biopsies compared to control¹



Mavrilimumab reduced arterial inflammation compared to control in an *in vivo* model of vasculitis²



Clinical Development Plan for Mavrilimumab

Phase 2 Giant Cell Arteritis

- **Enrollment completed**
- 26-week, double-blind, randomized, placebo-controlled clinical trial of mavrilimumab with a corticosteroid taper in subjects with new-onset or refractory GCA
- Primary efficacy endpoint involves measuring GCA flares during 26-week treatment period
- Continuing to enroll patients for a limited period to facilitate the accrual of primary efficacy endpoint events

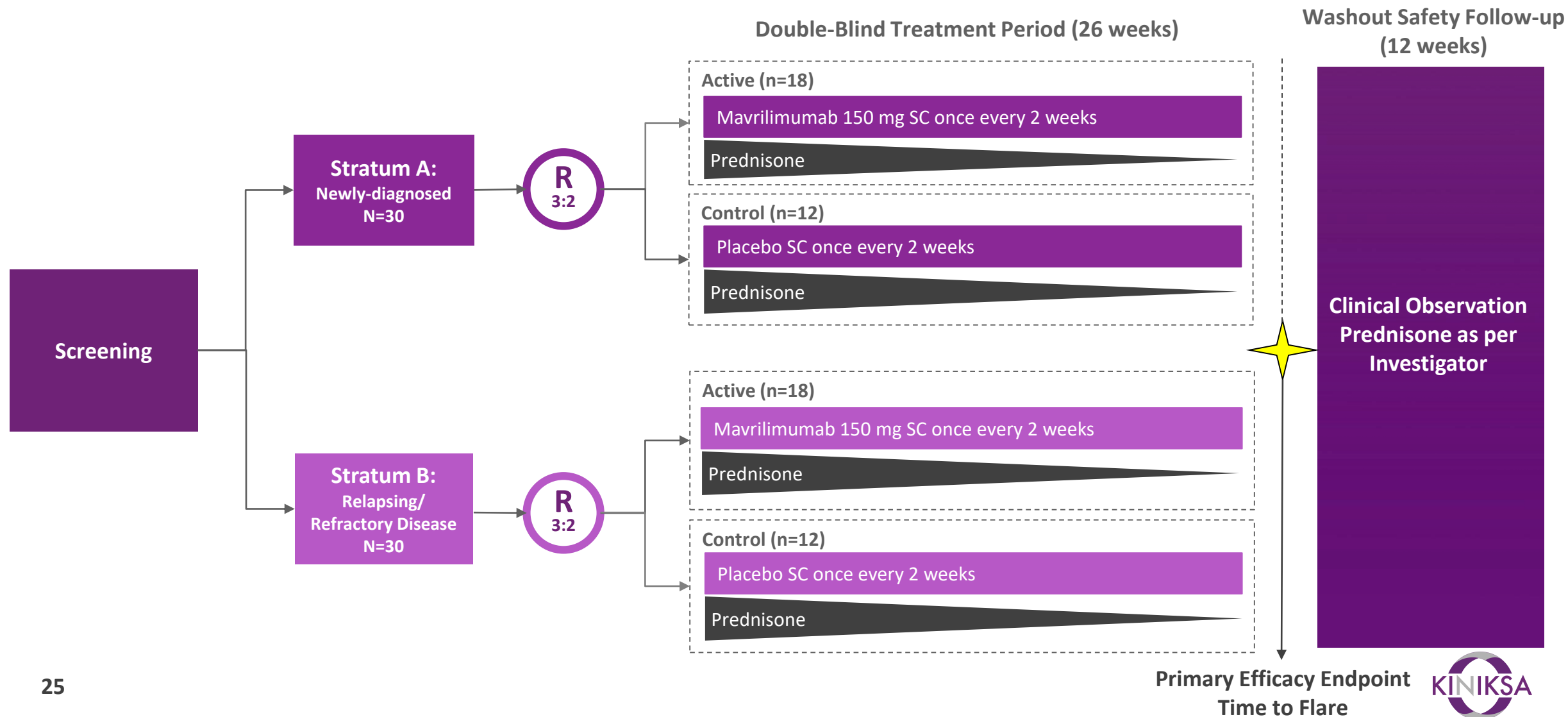
Top-line data expected 2H 2020

Phase 2 Relapsed/Refractory Large B-Cell Lymphoma

- **Clinical collaboration with Kite, a Gilead Company**
- Study of mavrilimumab with Yescarta® (axicabtagene ciloleucel) in patients with relapsed or refractory large B-cell lymphoma
- Preclinical evidence shows the potential for granulocyte macrophage colony stimulating factor (GM-CSF) to disrupt chimeric antigen receptor T (CAR T) cell mediated inflammation without disrupting anti-tumor efficacy¹

Initiation expected 2H 2020

Phase 2 Clinical Trial of Mavrilimumab in GCA



KPL-716 – Phase 2

Rilonacept

Mavrilimumab

KPL-716

KPL-404

First Indication	Prurigo Nodularis: Chronic inflammatory skin disease with pruritic lesions
Mechanism of Action¹	Monoclonal antibody inhibitor targeting OSMR β
Scientific Rationale²	OSMR β is a key receptor subunit shared by IL-31 and OSM; cytokines implicated in prurigo nodularis
Prevalence³	~300k prevalent in U.S.
Competition⁴	No FDA-approved therapies for prurigo nodularis
Status	Enrolling Phase 2a clinical trial in prurigo nodularis and exploratory Phase 2 study in diseases characterized by chronic pruritus
Economics	Clinical, regulatory and sales milestones; tiered royalty on annual net sales
Rights	Worldwide

Prurigo Nodularis is Typically Treated by Dermatologists Through a Combination of Medications and Behavioral Therapies; Treatment is Usually Unsuccessful

Diagnosis of Prurigo Nodularis By Dermatologists

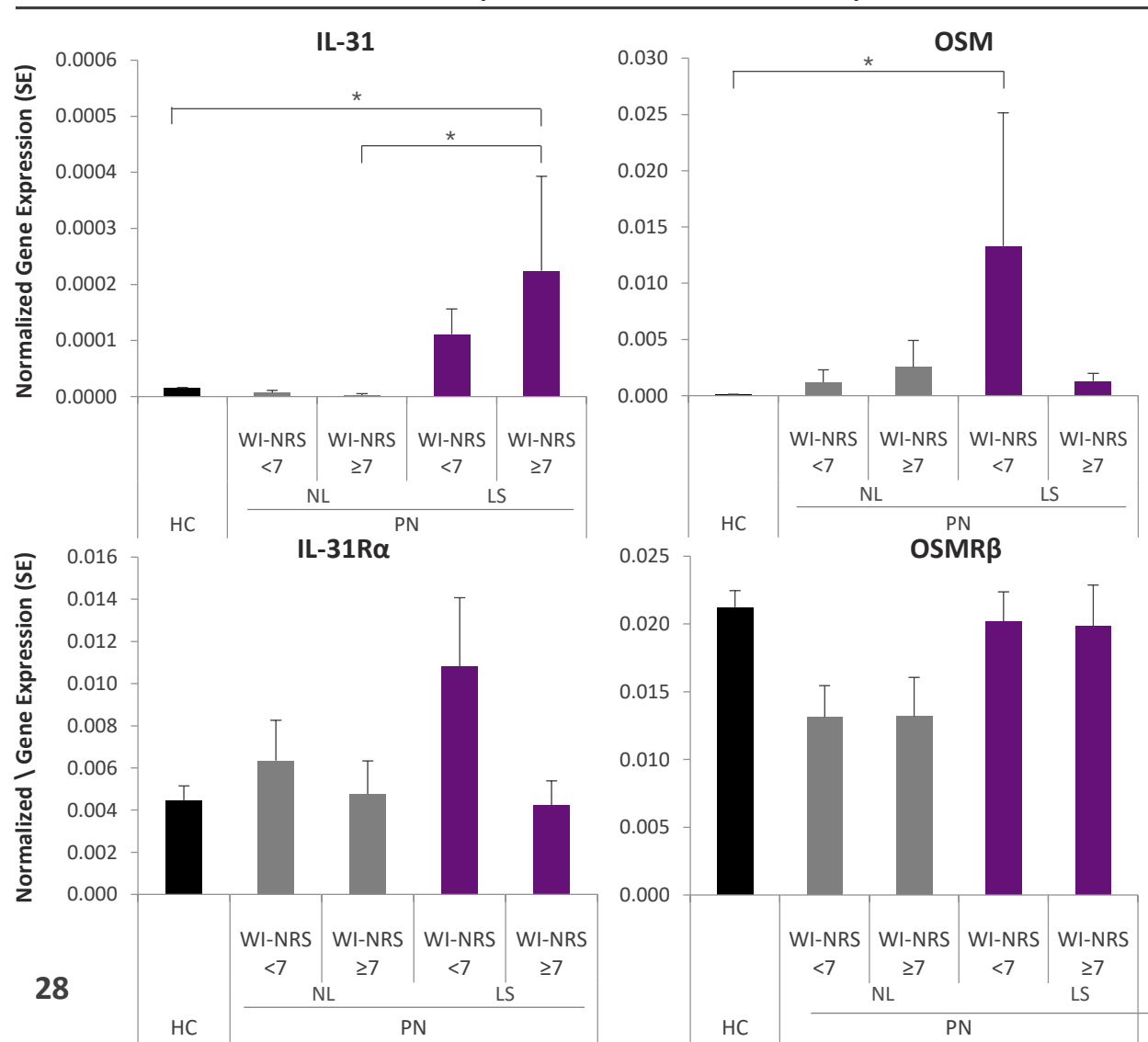
1 st Line	~100%	Emollients + Antipruritic Creams + Topical Corticosteroids + Antihistamines	
2 nd Line	~60-70%	Low-Dose Oral Corticosteroids, Intralesional Steroids, Occlusive Steroid Wrap	KPL-716 may initially slot after steroids
3 rd Line	~25-30%	UV Phototherapy	
4 th Line	~20-30%	Other Systemic Therapy (e.g. MTX, Cyclosporine, Doxepin, Thalidomide)	

Note: none of the above therapies are approved specifically for prurigo nodularis

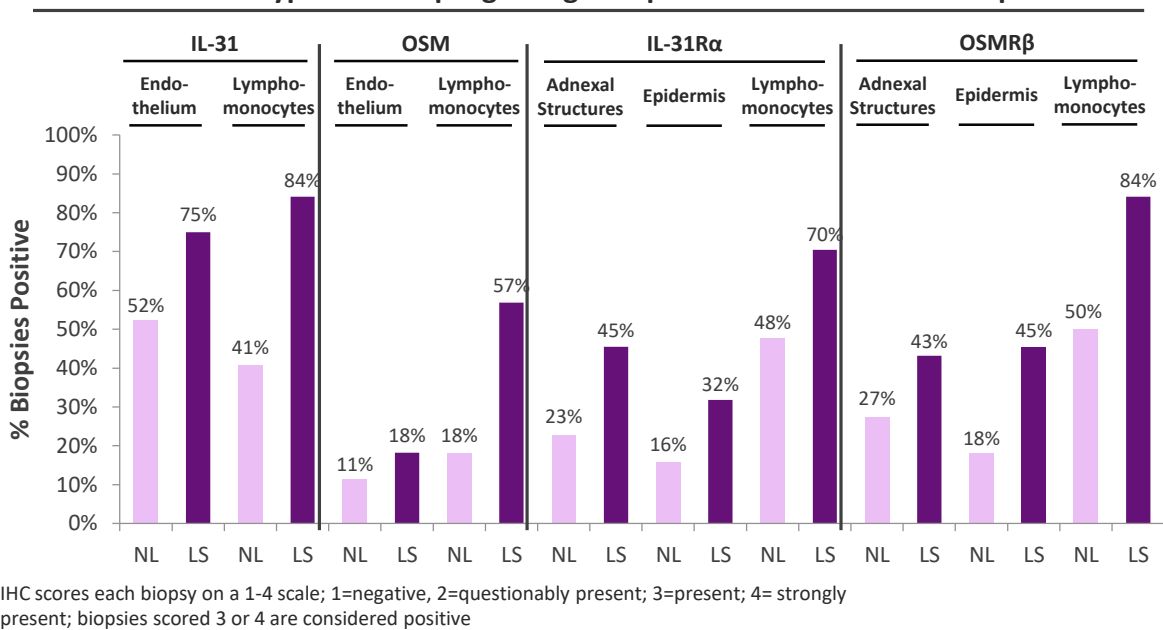


All Components of the Type II OSMR β Signaling Complex Show Upregulation in Lesional Skin of PN Patients; IL-31 is More Highly Expressed in Those Reporting Severe Pruritus

Levels of Gene Expression in PN and HC Skin Biopsies



Presence of Type II OSMR β Signaling Complex Protein in PN Skin Biopsies*



IHC scores each biopsy on a 1-4 scale; 1=negative, 2=questionably present; 3=present; 4=strongly present; biopsies scored 3 or 4 are considered positive

- OSM, OSMR β , IL-31, and IL-31R α mRNA expression was higher in lesional (LS) PN biopsies compared with non-lesional (NL) biopsies; all components except for OSMR β , which is known to be constitutively expressed, showed elevation compared to healthy controls (HC)
 - LS samples from PN patients with WI-NRS $\geq$ 7 expressed higher levels of IL-31 mRNA compared with HC samples ($p<0.05$) and NL samples
- Protein, analyzed through immunohistochemistry (IHC), for each of the Type II OSMR β signaling proteins shows upregulation in LS vs NL biopsies of PN patients' skin

These data suggest a role for the OSMR β axis (IL-31, OSM, IL-31R α , OSMR β) in the pathogenesis of PN given its prevalent expression in PN lesional skin

HC, healthy volunteers; IL-31R α , interleukin 31 receptor α ; LS, lesional; NL, non-lesional; SE, standard error; WI-NRS, Worst Itch Numeric Rating Scale. WI-NRS ranges from 0 ("no itch") to 10 ("worst imaginable itch"). * $P<0.05$

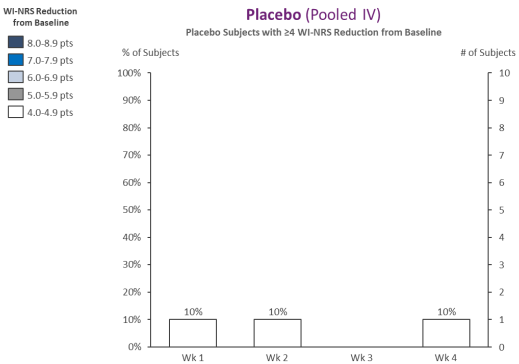
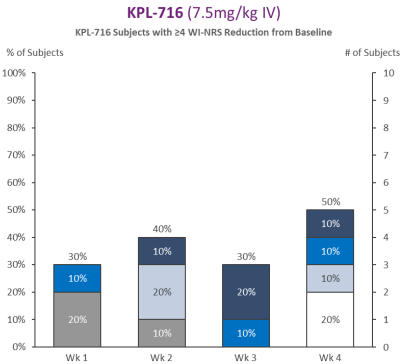
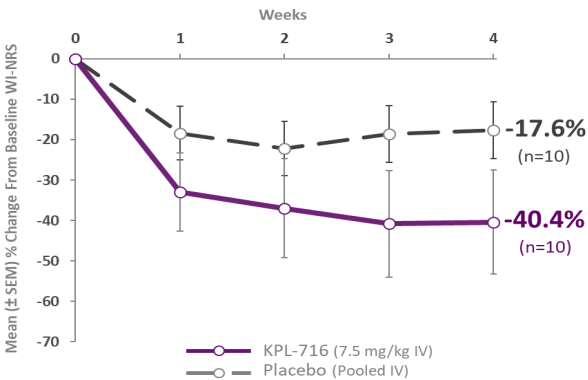
*Key tissue compartments for each component included; data for additional tissue compartments available



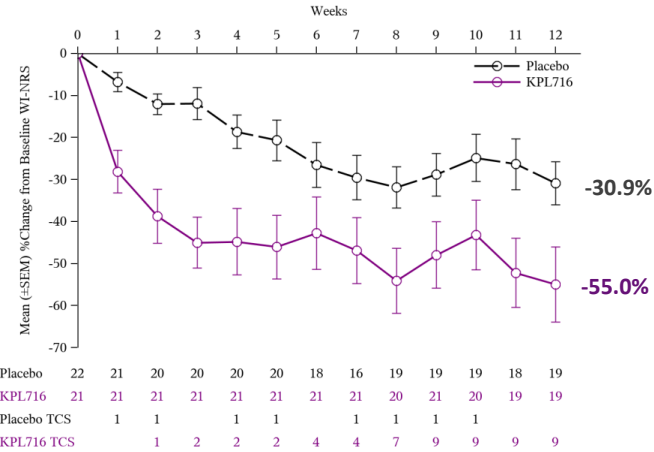
KPL-716 Phase 1b Data Showed Rapid and Sustained Reduction in Pruritus Versus Placebo

Single-Dose Phase 1b¹

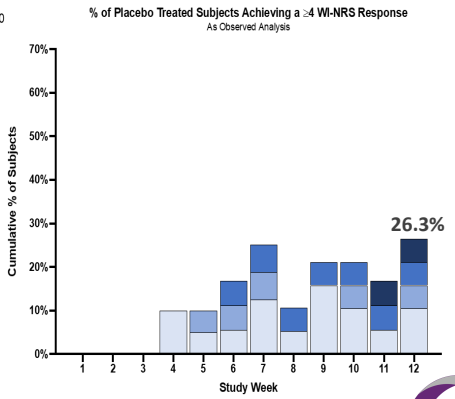
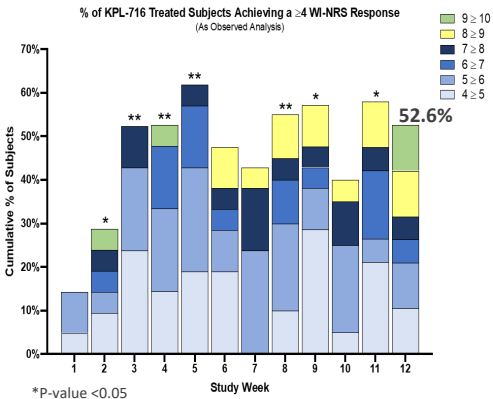
Weekly Average Worst Itch Numerical Rating Scale (WI-NRS)



Repeated-Single-Dose Phase 1b²



Placebo	22	21	20	20	20	20	18	16	19	19	19	18	19
KPL716	21	21	21	21	21	21	21	21	20	21	20	19	19
Placebo TCS	1	1	1	1	1	1	1	1	1	1	1	1	1
KPL716 TCS			1	2	2	2	4	4	7	9	9	9	9



*P-value <0.05
** P-value<0.01

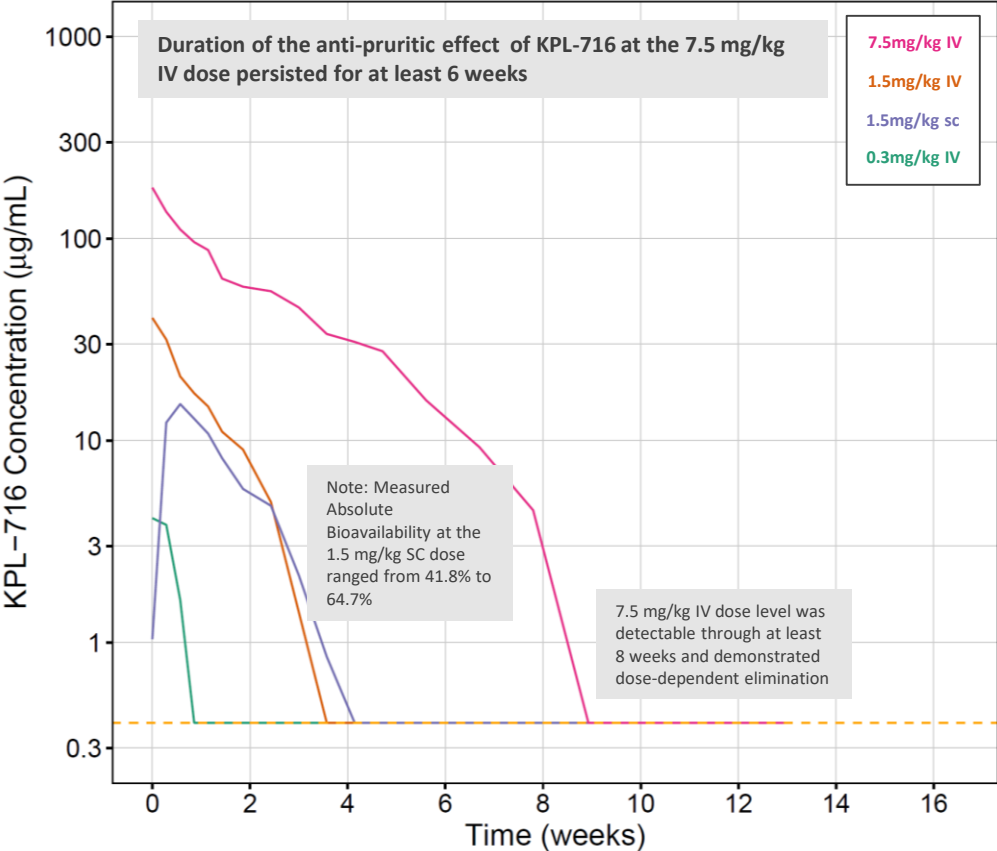
1) Oral presentation at the 27th European Academy of Dermatology and Venereology (EADV) Congress: First-In-Human Study of KPL-716, Anti-Oncostatin M Receptor Beta Monoclonal Antibody, in Healthy Volunteers and Subjects with Atopic Dermatitis

2) Interim data results 8/12/19 – available through Investors & Media section of Kiniksa's website at www.kiniksa.com

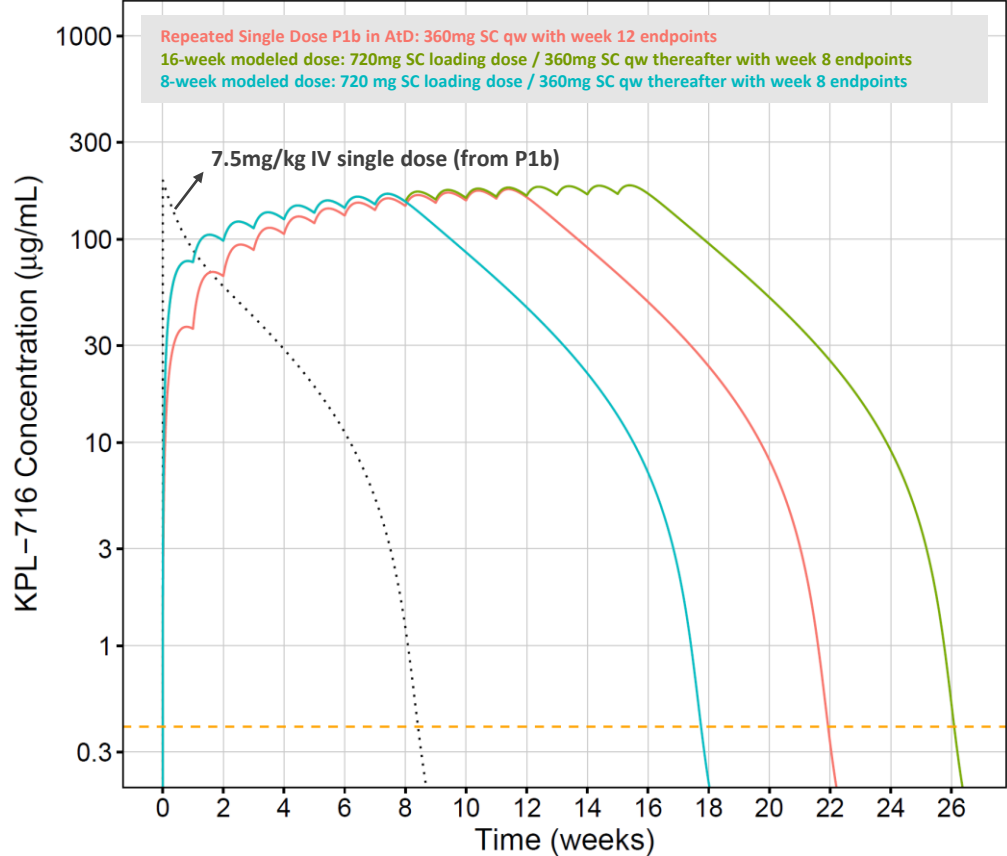


PK/PD Model: Weekly SC Dosing Provides Sufficient/High Exposures for POC Studies and Alternate Dosing Regimens in Future Dose-Finding Studies (e.g., q2w and/or qm)

Measured KPL-716 PK From P1b Single Dose



Phase 1b data used to build predictive PK/dosing model for multiple-dose studies (RSD, PN, Chronic Pruritic Diseases)



Note: Model based upon Absolute Bioavailability of 65% at the 360 mg SC dose



Clinical Development Plan for KPL-716

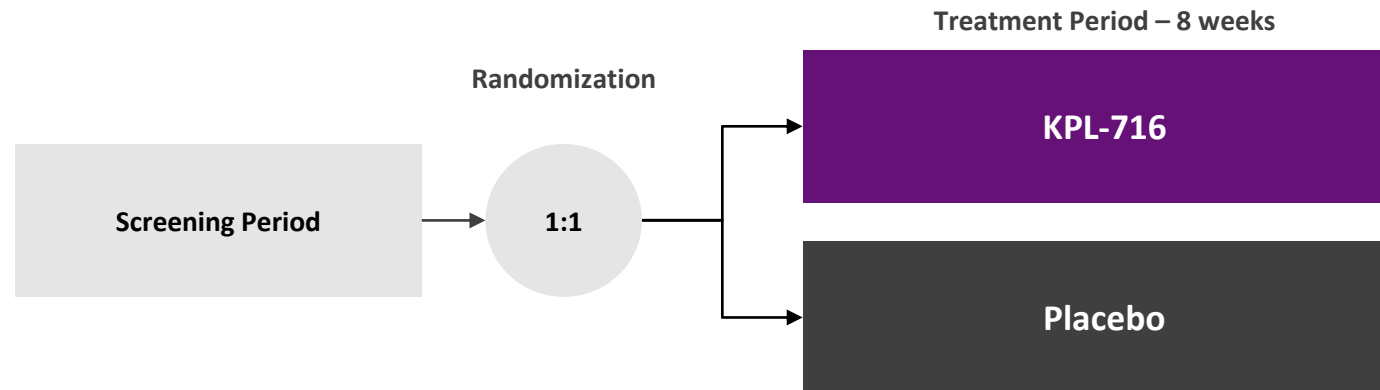
Phase 2a Prurigo Nodularis	Phase 2 Multiple Chronic Pruritic Diseases
• Evaluating KPL-716 in an 8-week, double-blind, randomized, placebo-controlled clinical trial in subjects with prurigo nodularis • Primary efficacy endpoint is percent change from baseline in weekly average Worst-Itch Numeric Rating Scale (WI-NRS) at 8 weeks	• Evaluating KPL-716 in an 8-week, double-blind, randomized, placebo-controlled clinical trial in subjects with chronic idiopathic urticaria, chronic idiopathic pruritus, lichen planus, lichen simplex chronicus and plaque psoriasis • Primary efficacy endpoint is percent change from baseline in weekly average WI-NRS at 8 weeks
Data expected by the end of April 2020	Interim data from cohorts expected 1H 2020

KPL-716 Phase 2a Trial in Prurigo Nodularis

Phase 2a Proof-of-Concept

Objective: Assess pruritus reduction

Dose: 720 mg SC loading dose --> 360 mg single SC QW thereafter



Primary Endpoint

% change from baseline in weekly average Worst Itch-Numeric Rating Scale (WI-NRS)

Key Secondary Endpoints

Proportion of subjects achieving at least a 4-point reduction from baseline in weekly average WI-NRS % change from baseline in pruritus Visual Analog Scale (VAS)

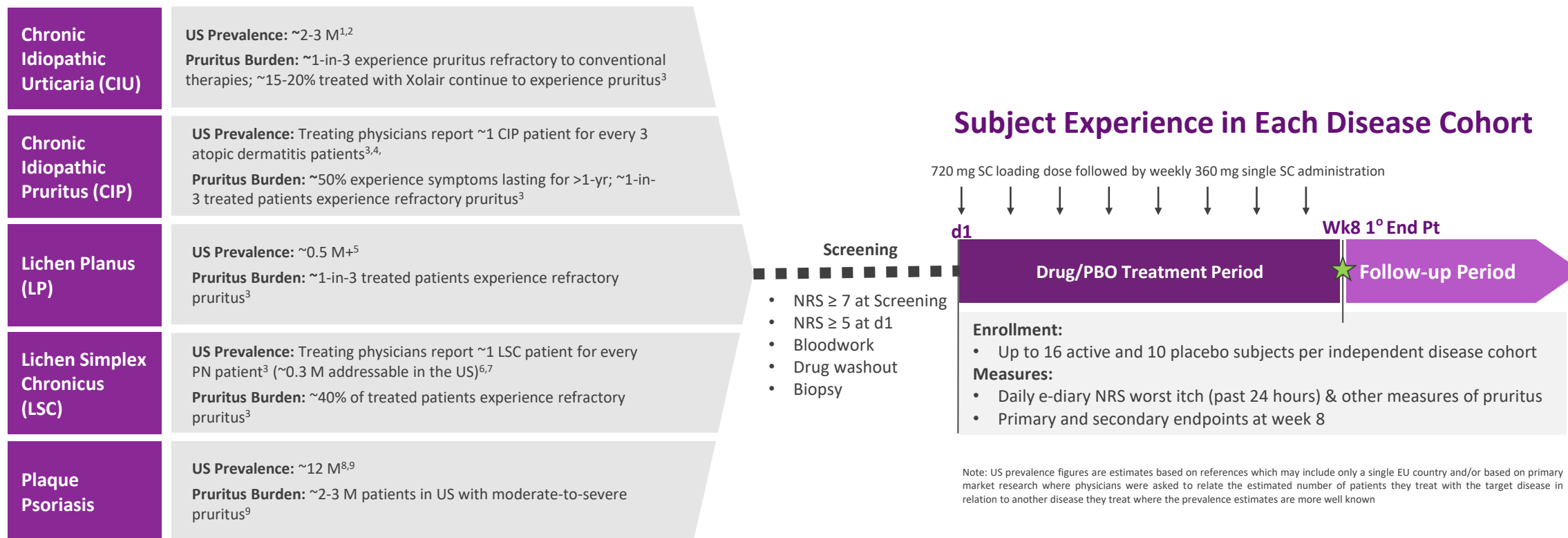
Other Secondary Endpoints

Exploratory tools will be used to measure disease modification

KPL-716 Exploratory Phase 2 Study in Diseases Characterized by Chronic Pruritus

Pilot Study Rationale

- 1 Investigate presence of IL-31 & OSM signature in multiple diseases characterized by chronic pruritus
- 2 In diseases where IL-31 is present (based on post-hoc biopsy analysis) → link inhibition of IL-31 with KPL-716 to clinical response
- 3 Diseases where IL-31 is NOT present (based on post-hoc biopsy analysis) → Investigate whether blocking OSMRβ has any effect



KPL-404 – Phase 1

Rilonacept

Mavrilimumab

KPL-716

KPL-404

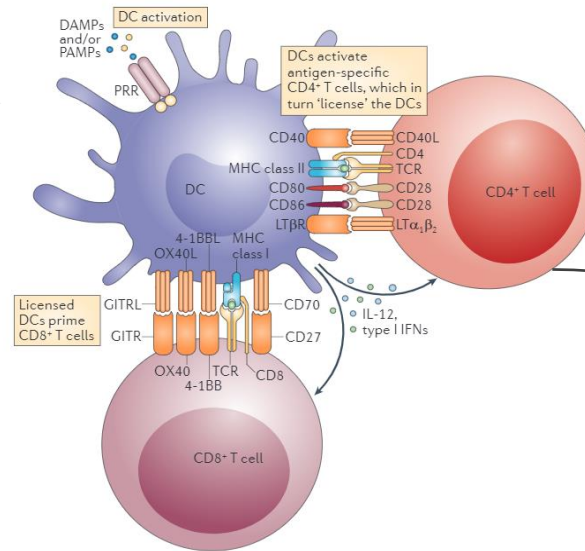
Autoimmune Diseases¹	External proof-of-concept previously established in broad range of autoimmune diseases: Sjogren's disease, systemic lupus, rheumatoid arthritis, solid organ transplant and Graves' disease ¹
Mechanism of Action²	Monoclonal antibody inhibitor of CD40-CD40L interaction
Scientific Rationale^{3,4}	Attractive target for blocking T-cell dependent, B-cell-mediated autoimmunity
Status	Enrolling first-in-human study with antigen challenge TDAR ⁵
Economics	Wholly-owned; insignificant clinical and regulatory milestones and royalty on annual net sales
Rights	Worldwide

KPL-404: Potential Best-in-Class Molecule for a Broad Range of Autoimmune Diseases

Mechanism	Humanized mAb inhibitor of CD40-CD40L interaction ¹	Designed to inhibit CD40-CD40L, a T-cell co-stimulatory pathway critical for B-cell maturation and immunoglobulin class switching
Rationale	External POC for CD40-CD40L inhibition established in a range of autoimmune diseases ^{2,3}	Published Positive Class-Related Clinical Data: Sjogren's syndrome, systemic lupus erythematosus, solid organ transplant, rheumatoid arthritis, Graves' disease Ongoing Class-Related Studies: type 1 diabetes, ulcerative colitis, lupus nephritis, hidradenitis suppurativa, kidney transplant and focal segmental glomerulosclerosis
Preclinical Data	Robust preclinical package supports development potential	Favorable pharmacokinetic and pharmacodynamic profiles, including engagement of CD40 target and block of antigen-specific primary and secondary antibody responses in a T-cell dependent antibody response cynomolgus monkey model
Competition	Potential differentiation	- KPL-404 at 10mg/kg achieved/maintained ~100% receptor occupancy in 7/7 non-human primates (NHP) through 4 weeks - KPL-404 10mg/kg suppressed T-cell dependent antibody responses (TDAR) in NHP model to tetanus toxoid (TT) and keyhole limpet hemocyanin (KLH) for >4 weeks
Status	Enrolling first-in-human study	- Enrolling a single-ascending-dose Phase 1 study in healthy volunteers which will provide safety data and pharmacokinetics as well as receptor occupancy and TDAR - Top-line data are expected in the second half of 2020

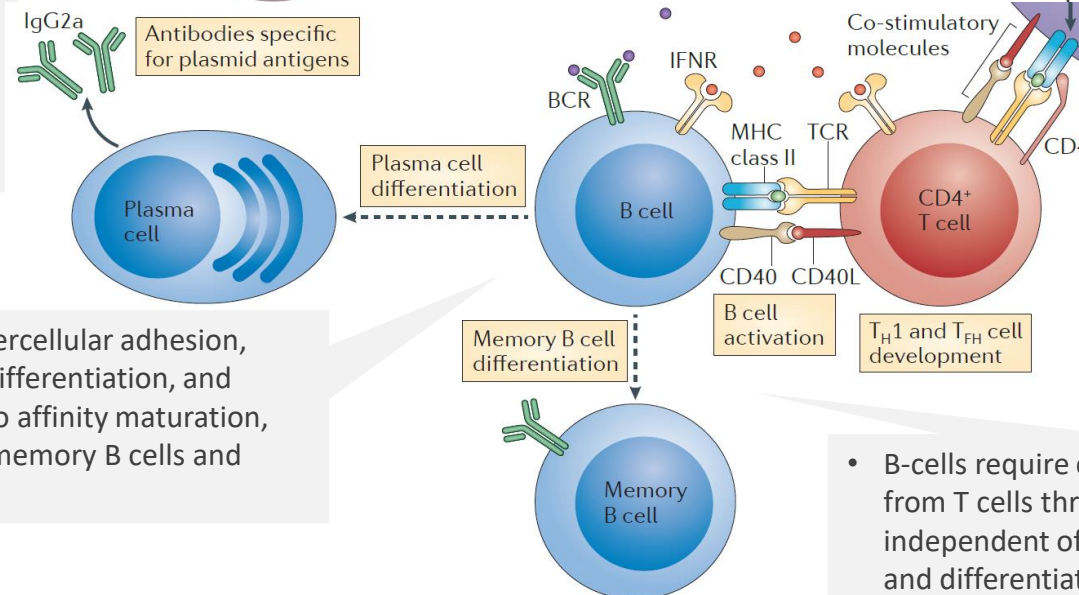
CD40/CD40L is an Essential Immune Pathway for T-Cell Priming and T-Cell Dependent B-Cell Responses

- CD40 is expressed on the surface of dendritic cells, B-cells, antigen-presenting cells and non-immune cell types
- Its ligand, CD40L (CD154), is expressed by activated T-cells, platelets, and other cell types



- CD40 ligation on DCs induces cell maturation by promoting antigen presentation and enhancing their costimulatory activity
- Mature DCs stimulate activated T-cells to increase IL-2 production that facilitates T-helper cells (Th) and cytolytic T-Lymphocyte (CTL) expansion
- CD40-stimulated DCs also secrete cytokines favoring Th1 cell differentiation and promoting Th cell migration to sites of inflammation
- CD40 ligation also provides a pro-inflammatory signal within the mononuclear phagocyte system

- Humoral immunity is dependent on a thriving B cell population and activation by Th cells; blockade of CD40/CD40L interaction has been shown to completely ablate primary and secondary TDAR response



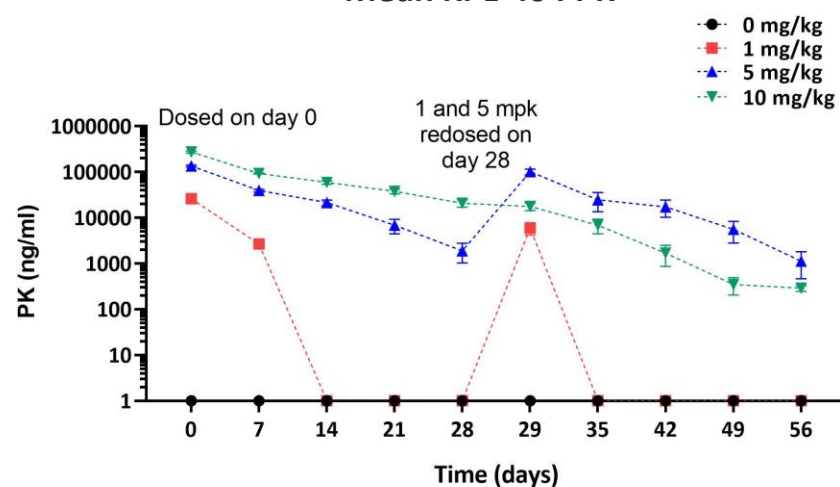
- CD40 engagement triggers B-cell intercellular adhesion, sustained proliferation, expansion, differentiation, and antibody isotype switching leading to affinity maturation, which is essential for generation of memory B cells and long-lived plasma cells

- B-cells require contact-dependent stimulus from T cells through CD40/CD40L interaction independent of cytokines to trigger growth and differentiation

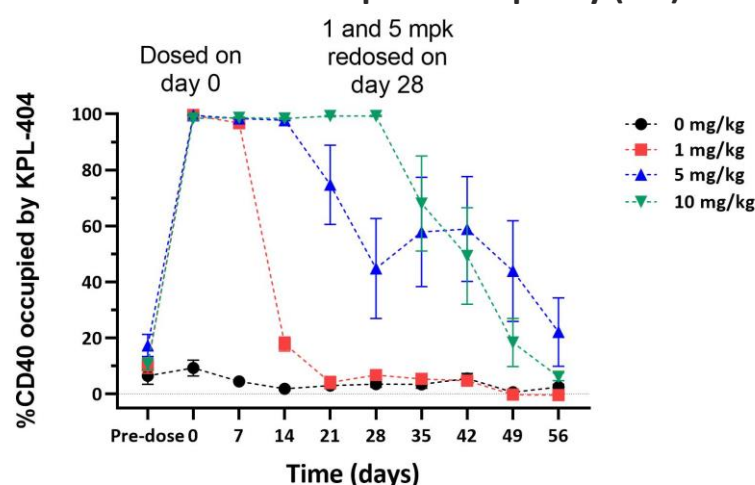
Sources: Elgueta et al., Immunol Rev, 2009; Peters et al., Semin Immunol, 2009; Kambayashi et al., Nature Reviews: Immunology, 14, 2014; Desmet et al., Nature Reviews: Immunology, 12, 2012

KPL-404 Shows Strong Results in a Non-Human Primate Model of TDAR

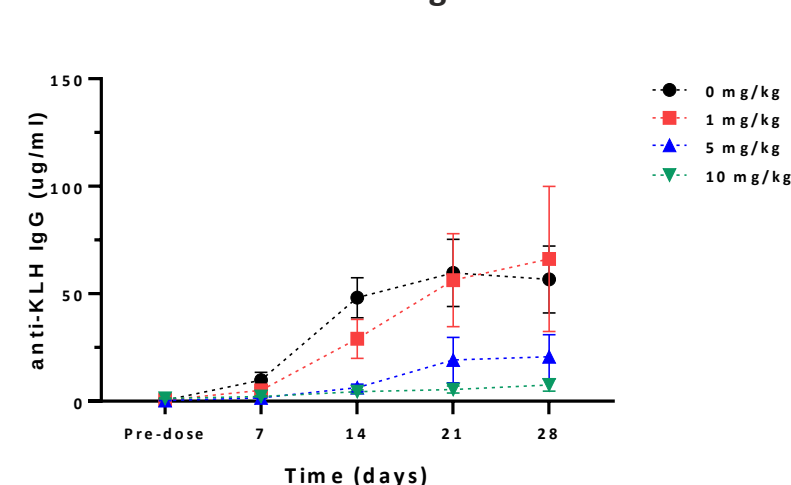
Mean KPL-404 PK



Mean KPL-404 Receptor Occupancy (RO)



Mean KLH IgG



Demonstrated linear pharmacokinetic profile with low variability between non-human primate subjects (n=7)

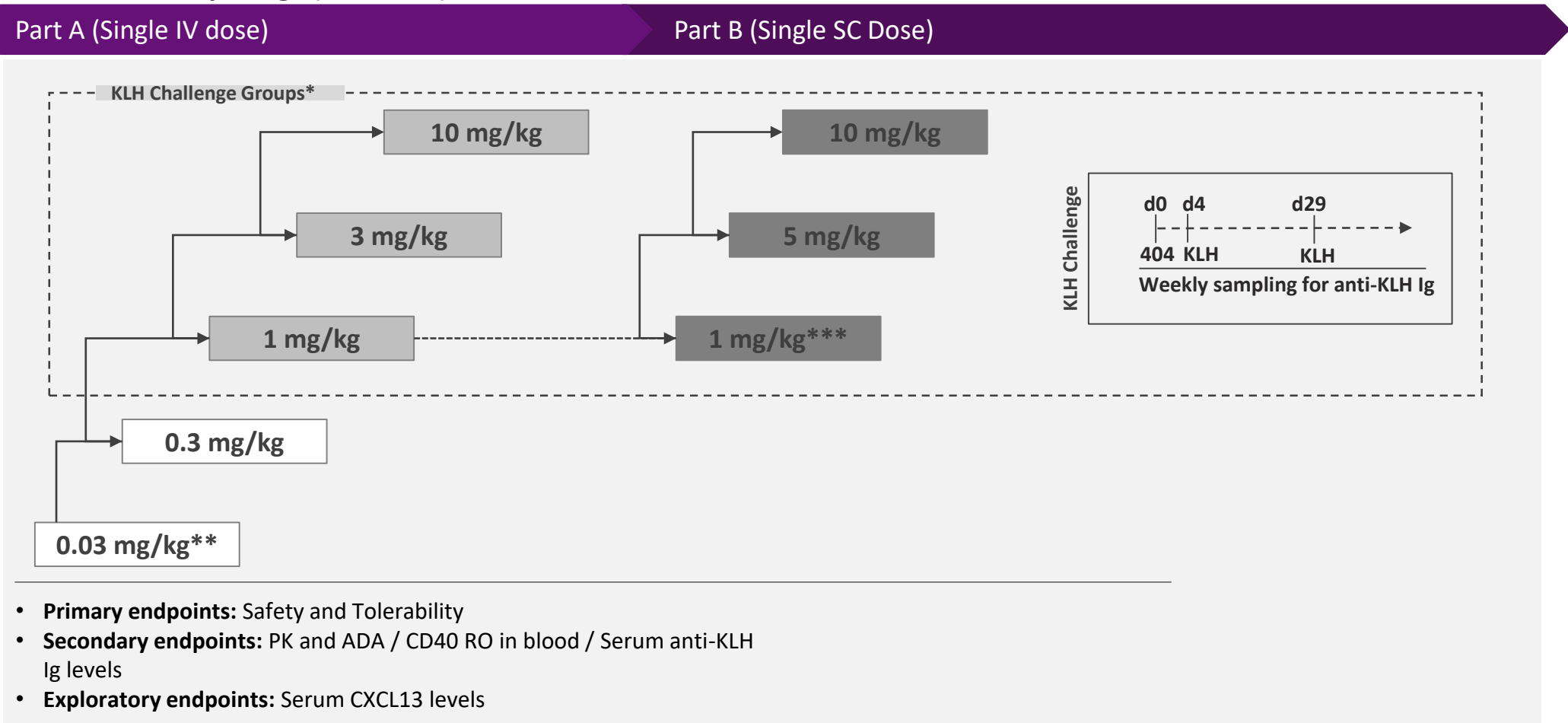
KPL-404 achieved 100% receptor occupancy for 2 weeks in all animals at 5mg/kg and 4 weeks in all animals at 10mg/kg

Complete suppression of primary T-cell dependent antigen response correlated with 100% receptor occupancy

KPL-404 Single-Ascending-Dose Phase 1 Study

First-in-human study to provide safety data and pharmacokinetics as well as receptor occupancy and TDAR

Phase 1 SAD Study Design (n=60 NHV)



Notes: Unless otherwise noted dose groups included 6 active/2 placebo subjects; *1° KLH challenge for all SAD dose groups except 0.03 and 0.3 mg/kg, 2° KLH re-challenge only in 1, 3, and 10 mg/kg IV; ** Cohort included 2 active and 2 placebo subjects; *** The 1 mg/kg SC dose arm will enroll after review of the 1 mg/kg IV SMC

SAD = single-ascending-dose; TDAR = T-cell dependent antibody response; KLH = keyhole limpet hemocyanin; RO = receptor occupancy; ADA = anti-drug antibodies



Multiple Clinical Data Readouts Expected in 2020

KPL-716 – Phase 2a (monoclonal antibody inhibitor targeting OSMR β)	Prurigo Nodularis	By the End of April 2020
KPL-716 – Phase 2 (monoclonal antibody inhibitor targeting OSMR β)	Diseases Characterized by Chronic Pruritus¹	1H 2020
Rilonacept – Pivotal Phase 3 (IL-1 α and IL-1 β cytokine trap)	Recurrent Pericarditis	2H 2020
Mavrilimumab – Phase 2 (monoclonal antibody inhibitor targeting GM-CSFR α)	Giant Cell Arteritis	2H 2020
KPL-404 – Phase 1 (monoclonal antibody inhibitor of CD40-CD40L interaction)	First-in-Human w/ Antigen Challenge TDAR	2H 2020





Immune-Modulating Product Candidates

Validated Mechanisms or Strong Biologic Rationale

Debilitating Diseases with Unmet Medical Need

~\$233M YE 2019 Cash Reserves Extend into 2H 2021¹

Multiple Clinical Data Readouts Expected in 2020



Every Second Counts!™

Appendix



Every Second Counts!™

Appendix – Rilonacept

Recurrent Pericarditis is a Debilitating Disease with No FDA-Approved Therapies

Pericarditis is chest pain caused by pericardial inflammation

Acute Pericarditis is diagnosed in patients with two of the following:

- (1) Retrosternal, pleuritic chest pain (85-90% of cases), (2) Abnormal ECG (ST elevation or PR depression); (4) Pericardial effusion^{1,2}

Often Idiopathic Etiology:

- Absent a clear sign of infection, it is assumed that most cases are post-viral, but are termed “idiopathic”

Recurrent Pericarditis:

- Diagnosed if there is recurrence after initial episode of acute pericarditis, with a symptom-free interval of > 4-6 weeks → First recurrence is followed by more recurrences between 20% - 30% of the time^{1,2}

Involvement of IL-1 in Idiopathic Recurrent Pericarditis:

- IL-1 has been implicated by several case reports and the AIRTRIP Study in idiopathic pericarditis

Recurrent pericarditis causes significant impairment of quality of life

Recurrent Disease Creates Burden on QoL:

- Although pericarditis is rarely life-threatening, patients may have significant impairment on quality of life due to chest pain:
 - Interference with sleep, as chest pain worsens while reclining
 - Lower productivity at work or school
 - Some patients may be on disability or close to it
 - Standard of care treatments have significant AEs

Complications Are Rare but Severe:

- Complications of pericarditis are rare (i.e., effusion, tamponade, constrictive pericarditis), but, when they occur, they can be life threatening and often require invasive therapy

Phase 2 Rilonacept Data

Baseline demographics and clinical characteristics

Baseline Demographics

General Characteristics	All Patients (n=25)
Unique patients, n	25
Mean age (range), yrs	42.8 (26-62)
Sex (male/female)	10/15
Race (white/African American)	22/3
Mean pericarditis episodes at enrollment ¹ (range)	4.3 (3-10)
Mean disease duration (range), yrs	2.2 (0.2-7.9)

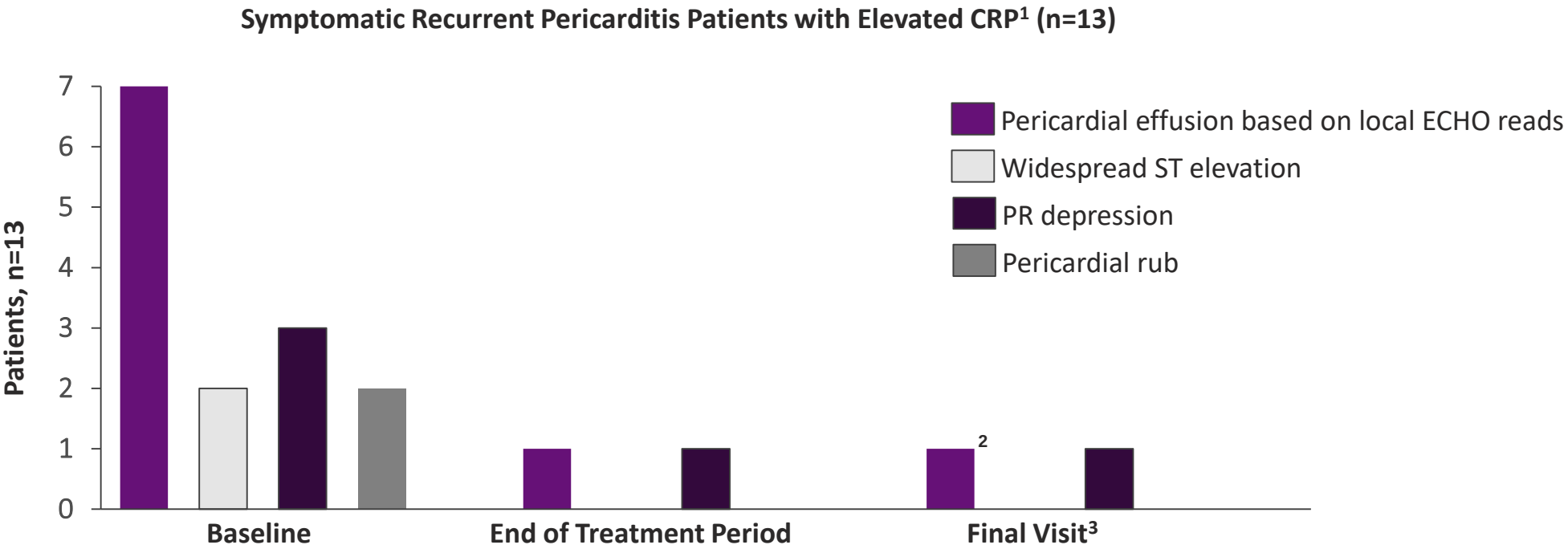
1)Includes index, recurrent, and qualifying (if applicable) episodes

Clinical Characteristics

Disease Status: CRP requirement (mg/dL): N:	Idiopathic RP			PPS	
	Active ^a	Active ^b	CS-dep ^c	Active ^d	CS-dep ^e
	>1	≤1	N/A	>1	N/A
Mean NRS ^f (SD)	4.6 (1.7)	4.7 (3.1)	1.2 (0.8)	4.0 (N/A)	2.0 (2.7)
Mean CRP (SD), mg/dL	4.9 (5.8)	0.5 (0.4)	0.2 (0.1)	1.1 (N/A)	0.1 (0.1)

Phase 2 Rilonacept Data

Pericardial signs resolved or improved in all patients (parts 1 and 4)

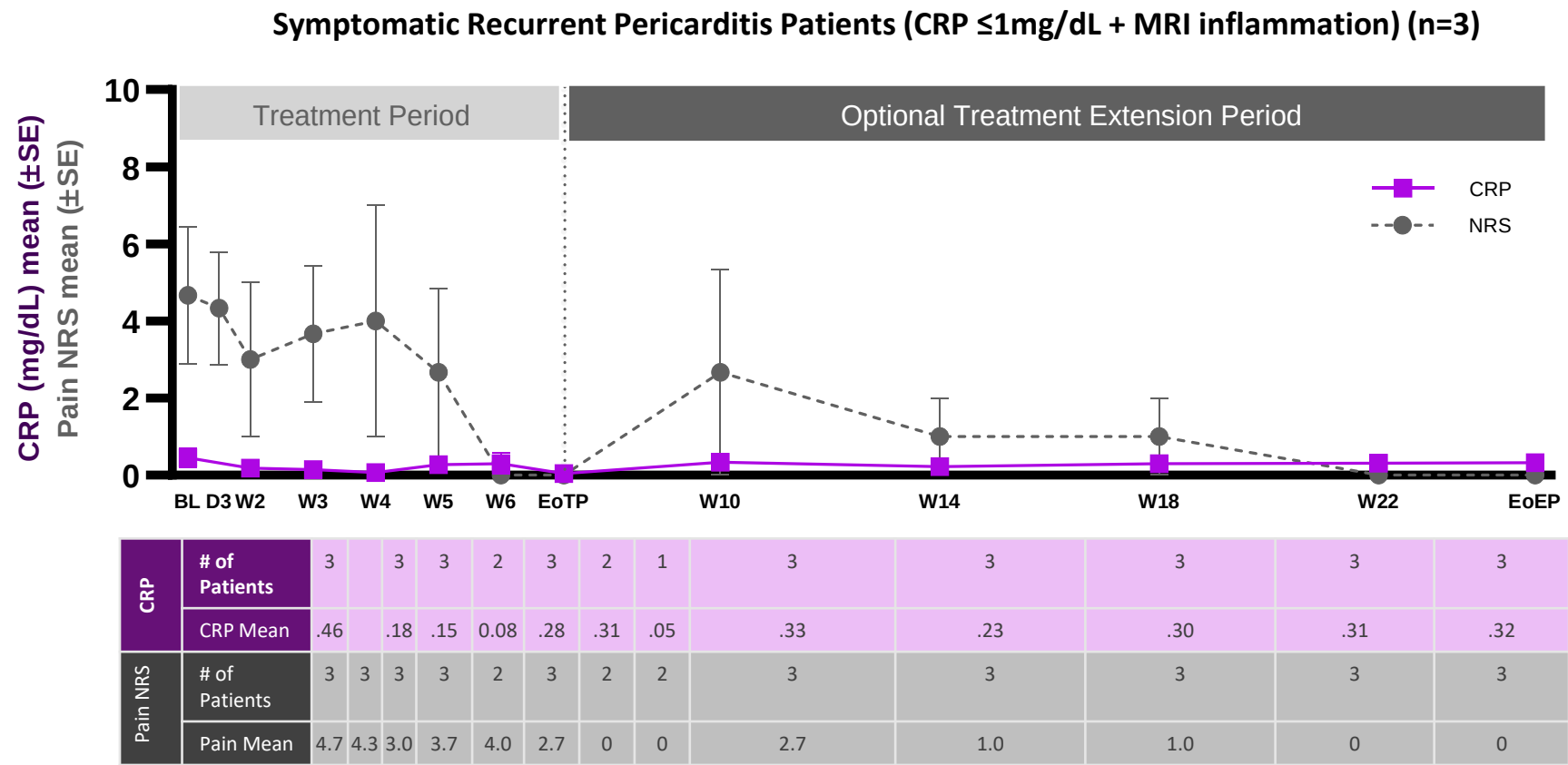


1) Patients with elevated CRP and symptomatic disease (Parts 1 and 4) are most representative of real-world recurrent pericarditis. Inclusion and exclusion criteria for the ongoing Phase 3 study RHAPSODY align with this patient population (clinicaltrials.gov/NCT03737110); 2) patient with effusion at baseline, no effusion at EoT Visit and trivial effusion (not pathological) at Final Visit; 3) n=12; one patient discontinued study drug in TP due to SAE; no effusion at baseline or EoT Visit; CRP = C-Reactive Protein



Phase 2 Rilonacept Data

Reduction in both reported pain and inflammation in symptomatic patients without elevated CRP and with MRI inflammation (Part 2)



Phase 2 Rilonacept Data

Corticosteroid tapering in corticosteroid-dependent patients (Parts 3 and 5)

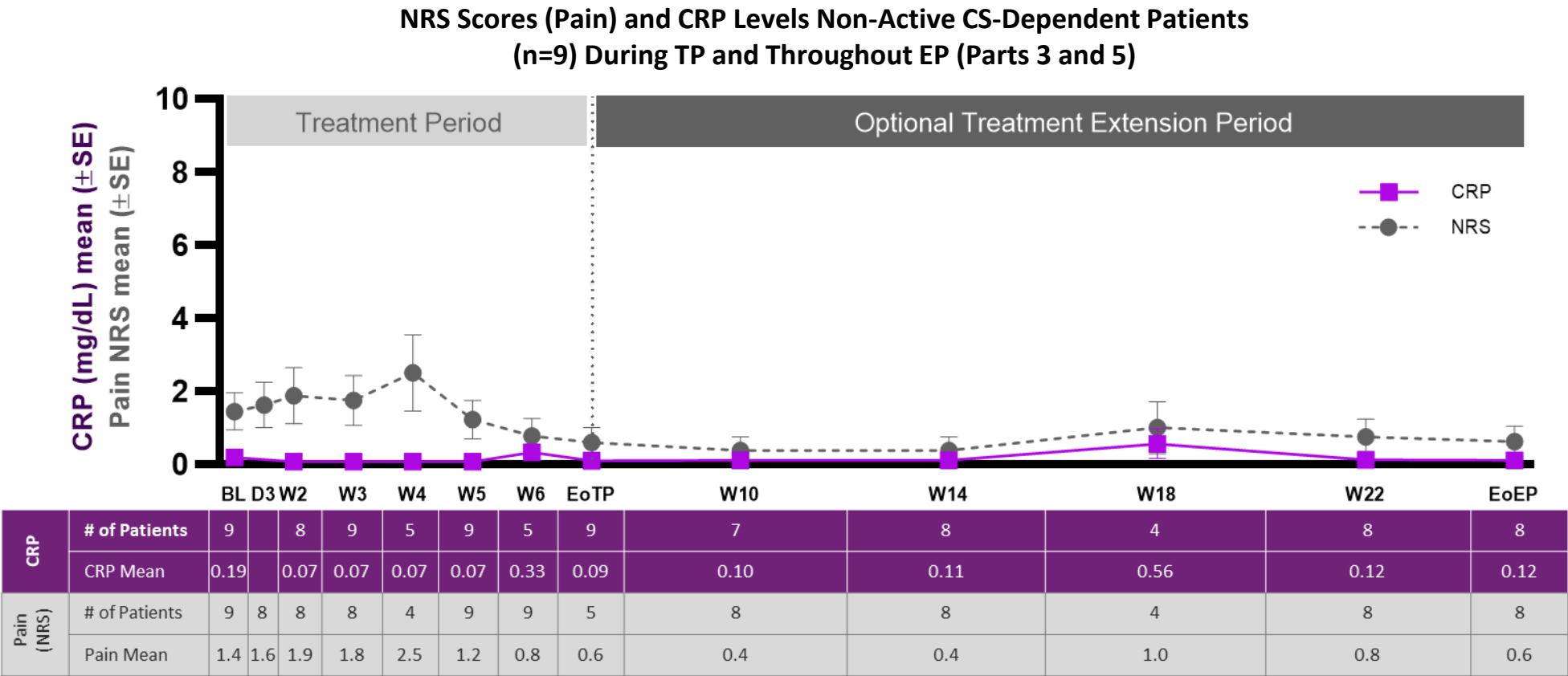
Corticosteroid-Dependent Patients (Parts 3 and 5): Pericarditis Medications During TP and EP Combined

n/N (%)	<u>Medications</u>					
	At least 1	Analgesics	Aspirin	NSAIDs	Colchicine	CS
Dose stopped	7/8 (87.5)	0/0	0/1	2/5 (40.0)	1/7 (14.3)	7/8 (87.5)
Dose decreased	4/8 (50)	0/0	1/1 (100)	2/5 (40)	1/7 (14.3)	1/8 (12.5)
Dose increased	0/8	0/0	0/1	0/5	0/7	0/8
Starting new	0/8	0/8	0/8	0/8	0/8	0/8

CS, corticosteroid; NSAID, nonsteroidal anti-inflammatory drugs

Phase 2 Rilonacept Data

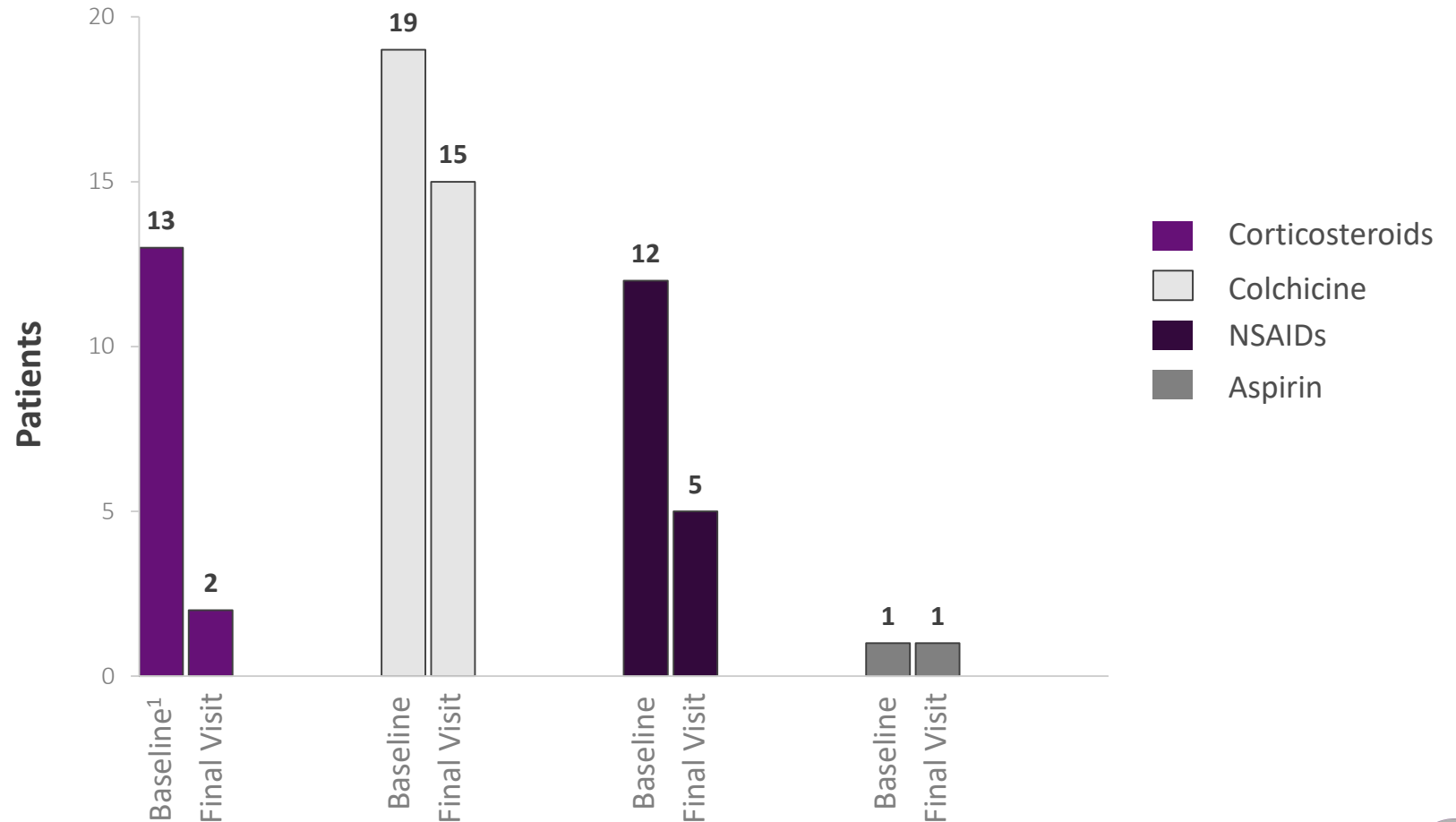
Pericarditis pain scores and CRP in corticosteroid-dependent patients (Parts 3 and 5)



Phase 2 Rilonacept Data

All patients on corticosteroids (CS) at baseline who completed 24 weeks of treatment stopped or tapered CS during rilonacept treatment without experiencing a recurrence

No patients had pericarditis recurrence in investigators' judgement after stopping concomitant pericarditis medication while on rilonacept treatment



Phase 2 Rilonacept Data

Of 13 patients on corticosteroids (CS) at baseline who completed 24 weeks of treatment, 11 discontinued CS and the CS dose was successfully tapered in the remaining 2 patients

	Idiopathic			PPS		Idiopathic or PPS
Disease Status:	Active ¹	Active ²	CS-dep ³	Active ⁴	CS-dep ⁵	All ¹⁻⁵
CRP requirement (mg/dL):	>1	≤1	N/A	>1	N/A	N/A
N:	12	3	6	1	3	25
Baseline						
Patients on prednisone ⁶ , n	4	2	6	0	3	15
Mean dose (mg/day)	8.4	40.0	8.9	0	7.7	12.7
Min	1.0	30.0	2.5	0	3.0	1.0
Max	12.5	50.0	30	0	15.0	50.0
Corticosteroid Changed During TP and EP Combined						
Prednisone dose decreased ^{7,8}	0/3	1/2 (50.0)	1/5 (20.0)	0/0	0/3	2/13 (15.4)
Prednisone stopped ^{8,9}	3/3 (100)	1/2 (50.0)	4/5 (80.0)	0/0	3/3 (100)	11/13 (84.6)
Prednisone dose increased ⁷	0/3	0/2	0/5	0/0	0/3	0/13
Prednisone initiated ⁹	0/11	0/3	0/5	0/1	0/3	0/23

1) Part 1; 2) Part 2; 3) Part 3; 4) Part 4; 5) Part 5; 6) 2 patients on prednisone at baseline did not enter EP (one in Part 1 and in Part 3) 7) Refers to patients who entered the study on prednisone; 8) 1 patient decreased prednisone dose in TP, and 1 stopped prednisone in TP (both in Part 2); 9) Refers to all patients in EP; CRP= C-reactive protein; CS-dep = corticosteroid-dependent; PPS = post-pericardiotomy syndrome; TP = treatment period; EP = extension period

Phase 2 Rilonacept Data

Annualized incidence of pericarditis episodes decreased during rilonacept treatment in the study

Disease Status: CRP requirement (mg/dL): N:	Idiopathic			PPS	
	Active ¹	Active ²	CS-dep ³	Active ⁴	CS-dep ⁵
	>1	≤1	N/A	>1	N/A
	12	3	6	1	3
Prior to the study ⁶					
Pericarditis episodes per year, mean (SD)	4.4 (4.68)	2.0 (1.75)	4.5 (2.58)	1.3 (N/A)	3.7 (3.02)
During the study ⁷					
Patients with pericarditis episodes, n	1 ^h	0	0	0	0
Pericarditis episodes per year, mean (SD)	0.18 (0.62)	0	0	0	0

1) Part 1; 2) Part 2; 3) Part 3; 4) Part 4; 5) Part 5; 6) Episodes at enrollment include index, prior recurrences, and current episode; 7) Episodes during the study include recurrences during TP and EP combined. Pericarditis recurrence during the study was based on Investigator’s judgement; ^hPatient had a mild pericarditis recurrence in TP, 5 days duration, with NRS pain increase from 0 to 2, CRP 0.10 mg/dL, not requiring addition of new medication to treat pericarditis; CRP = C-reactive protein; CS-dep = corticosteroid-dependent; PPS = post-pericardiotomy syndrome



Phase 2 Rilonacept Data

Rilonacept treatment resulted in improvement of quality of life scores¹

	Idiopathic or PPS	
	Active ¹ (n=16)	CS-dependent ² (n=9)
Global Physical Health, mean (SD)		
Baseline	39.94 (8.941)	43.3 (5.311)
End of TP	51.35 (7.962)	45.09 (4.057)
Final Visit	51.32 (6.564)	46.81 (9.266)
Global Mental Health, mean (SD)		
Baseline	44.5 (10.484)	46.49 (7.767)
End of TP	50.13 (11.325)	47.91 (5.509)
Final Visit	50.54 (10.995)	50.66 (6.299)

Phase 2 Rilonacept Data

Summary of adverse events

Disease Status:	Idiopathic			PPS		Idiopathic or PPS		
	Active ¹	Active ²	CS-dep ³	Active ⁴	CS-dep ⁵	Active ^{1,2,4}	CS-dep ^{3,5}	All ¹⁻⁵
CRP requirement (mg/dL):	>1	≤1	N/A	>1	N/A	N/A	N/A	N/A
N:	12	3	6	1	3	16	9	25
≥1 TEAE, n (%)	12 (100)	3 (100)	6 (100)	1 (100)	3 (100)	16 (100)	9 (100)	25 (100)
≥1 treatment-related TEAE, n (%)	9 (75)	2 (66.7)	3 (50)	1 (100)	2 (66.7)	12 (75)	5 (55.6)	17 (68)
≥1 serious TEAE, n (%)	2 (16.7)	0	0	0	0	2 (12.5)	0	2 (8)
≥1 treatment-related serious TEAE, n (%)	1 (8.3)	0	0	0	0	1 (6.3)	0	1 (4)
≥1 TEAE leading to treatment discontinuation, n (%)	1 (8.3)	0	0	0	0	1 (6.3)	0	1 (4)
≥1 TEAE leading to death, n (%)	0	0	0	0	0	0	0	0
TEAEs by severity, n (%)								
Mild	9 (75)	3 (100)	4 (66.7)	1 (100)	2 (66.7)	13 (81.3)	6 (66.7)	19 (76)
Moderate	2 (16.7)	0	2 (33.3)	0	0	2 (12.5)	2 (22.2)	4 (16)
Severe	1 (8.3)	0	0	0	1 (33.3)	1 (6.3)	1 (11.1)	2 (8)
Reactions at injection site ⁶ , n (%)	5 (41.7)	1 (33.3)	3 (50)	1 (100)	2 (66.7)	7 (43.8)	5 (55.6)	12 (48)

- There were 2 serious treatment-emergent AEs reported in Part 1, both of which resolved
 - 1 patient with subcutaneous abscess (possibly related to study drug) that resolved with medical management discontinued rilonacept treatment
 - 1 patient with atypical chest pain (not related to study drug) continued rilonacept treatment
- AEs observed with rilonacept treatment are consistent with the known safety profile of rilonacept
- The most common AEs were observed in the general disorders and administration site conditions (injection site reactions), infections and infestations, and musculoskeletal and connective tissue disorders classes

Case Study: Treatment/Retreatment of Recurrent Pericarditis with Rilonacept

- **Patient**

- 50-year-old female with idiopathic pericarditis and 1 prior recurrence, enrolled in Part 1 during her third episode (pain NRS 6/10; CRP 8.85 mg/dL; pericardial effusion on echocardiography) while receiving colchicine 0.6 mg bid.

- **Pain and CRP Reduction During the Study**

- Addition of rilonacept to colchicine background rapidly reduced pain (week 2 pain NRS 1/10; week 24 pain NRS 0/10), decreased CRP (week 2 CRP 0.66 mg/dL; week 24 CRP 0.09 mg/dL), and resolved pericardial effusion.

- **Safety**

- Mild, transient injection site reactions occurred for 21 of 24 rilonacept injections; the patient also had reported mild AEs of heartburn, common cold, worsening of elevated LFTs, elevated cholesterol, elevated HDL, intermittent chest discomfort and elevated CK

- **After Completing the EP**

- Approximately 8 weeks after rilonacept discontinuation, while continuing on colchicine 0.6 mg bid, the patient presented with pericarditis symptoms requiring addition of celecoxib 200 mg/day. Ten weeks later the patient developed frank pericarditis recurrence (pain NRS 7/10; CRP 23.1 mg/dL) and cardiac tamponade requiring pericardiocentesis. The patient was re-enrolled in the study.

- **Pain and CRP Normalized and Pericardial Effusion Resolved with Rilonacept Retreatment**

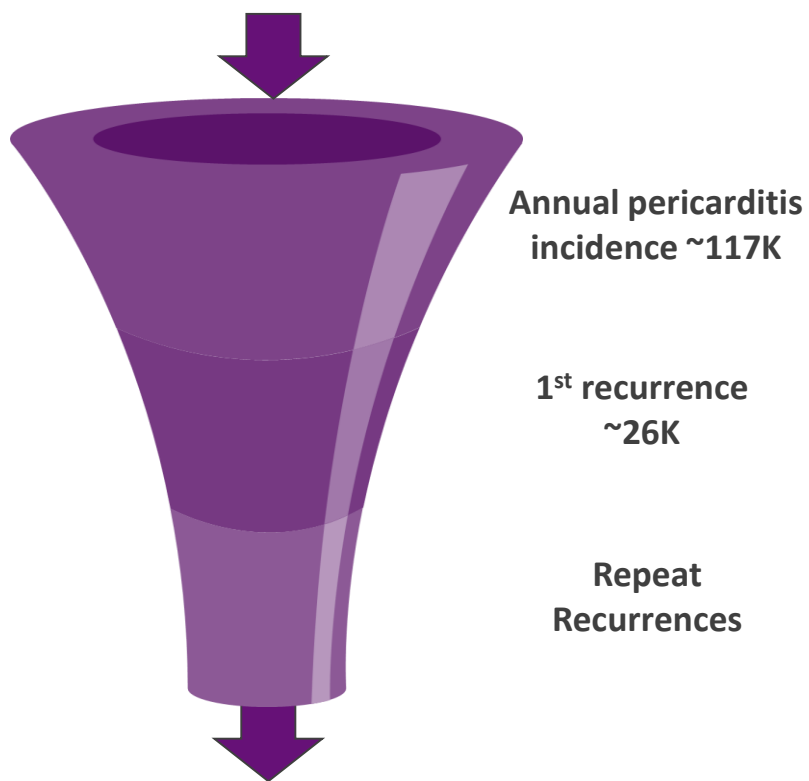
- Rapid improvements in pain and CRP were observed after the first rilonacept administration (week 2 pain NRS 0/10; CRP 0.57 mg/dL). At the week 7 visit, NRS pain was 1/10, CRP was 0.09 mg/dL, and there was no evidence of pericardial effusion on echocardiography. At the last study evaluation available (1 month EP), NRS pain was 0/10 and CRP remained normal (0.08 mg/dL). At the Final Visit NRS pain was 0/10 and CRP remained normal (0.14 mg/dL).

- **Safety**

- Mild, transient injection site reactions occurred in 17 out of 24 rilonacept administrations; the patient also developed mild AEs of hypokalemia, decreased WBC count, and increased lipids.

Addressable U.S. Opportunity of Rilonacept Estimated to be ~14K Patients

~7K new patients with multiple recurrences enter target pool annually



Year	-4	-3	-2	-1	0
Incident case of acute pericarditis (1 st episode) ¹	117K	117K	117K	117K	117K
Incidence of initial RP patients (1 st recurrence) ²	26K	26K	26K	26K	26K
Ongoing recurrent from year-1 ³				7K	
Ongoing recurrent from year-2 ³			7K	3.5K	
Ongoing recurrent from year-3 ³		7K	3.5K	1.8K	
Ongoing recurrent from year-4 ³	7K	3.5K	1.8K	0.9K	
Ongoing recurrent from year-5 ³	3.5K	1.8K	0.9K	0.5K	
Ongoing recurrent from year-6 ³	1.8K	0.9K	0.5K	0.2K	
Ongoing recurrent from year-7 ³					0.1K

Addressable Opportunity in U.S.

1: Prevalence estimate from Imazio, et al. (2008); includes all etiologies (~80% idiopathic)
 2: Mid point of 15-30% of initial recurrence rate published in ESC Guidelines given higher colchicine use today
 3: Estimate for recurrence rate of subsequent recurrences from ESC Guidelines and Claims Analysis

Summary of Rilonacept Profit Share Arrangement with Regeneron¹



- Upfront payment: \$5 million
- Future regulatory milestones: \$27.5 million in aggregate
- Kiniksa covers 100% of development expenses related to approval of additional indications
- In the U.S. and Japan, the initial license covers all indications other than CAPS⁴, DIRA⁵, oncology, and local application for eye and inner ear
- Kiniksa has rights to develop and commercialize rilonacept in our field worldwide, with the exception of MENA⁶
- After receipt of positive Phase 3 clinical data, the BLA⁷ for rilonacept transfers to Kiniksa
- Upon approval for a new indication, the scope of the license expands to include CAPS and DIRA in the US and Japan, and we will assume the sales and distribution of rilonacept in these additional indications
- Profits on sales of rilonacept will be equally split after deducting certain commercialization expenses subject to specified limits



Every Second Counts!™

Appendix – Mavrilimumab

GCA is a Serious Condition Characterized by Inflammation of Medium-to-Large Arteries

1

Chronic inflammation of medium-to-large arteries

- GCA is characterized by inflammation of medium-to-large arteries with **predisposition for the cranial branches of the carotid artery** and is typically found in patients over 50 years old
- Due to the impact on the carotid arteries, GCA is **often characterized by temporal specific symptoms like headaches**, jaw claudication and scalp tenderness

2

If left untreated, GCA can cause serious complications

- While the onset of symptoms tends to be subacute, patients can experience **acute events including permanent vision loss (~10-20% of patients)** and/or aneurysms/dissections (~1-6% of patients)
- Due to the threat of these more serious complications, giant cell arteritis is **considered a medical emergency**

3

GCA variants associated with unique presentations

- **LV-GCA**, characterized by the involvement of the aorta and its major proximal branches, is estimated to be involved in anywhere from **~30-80% of patients**
- **~40-50%** of GCA patients **suffer from polymyalgia rheumatica**, a rheumatic disease characterized by widespread aching and stiffness; symptoms are relieved immediately upon starting on low-dose steroids

“There is an urgency of treatment with these patients, compared to other conditions it’s serious.”

– Rheumatologist

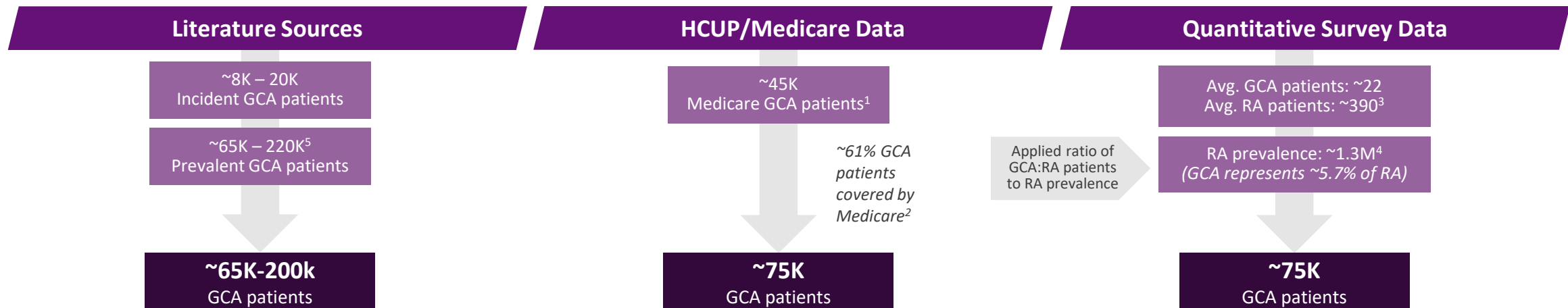
“There are people out there that need to get this disease under control, but they never receive the correct treatment, this is life threatening!”

– Rheumatologist

“I hate steroids, the long –term side effects are sometimes worse than the disease but, I definitely don’t want patients to go blind.”

– Rheumatologist

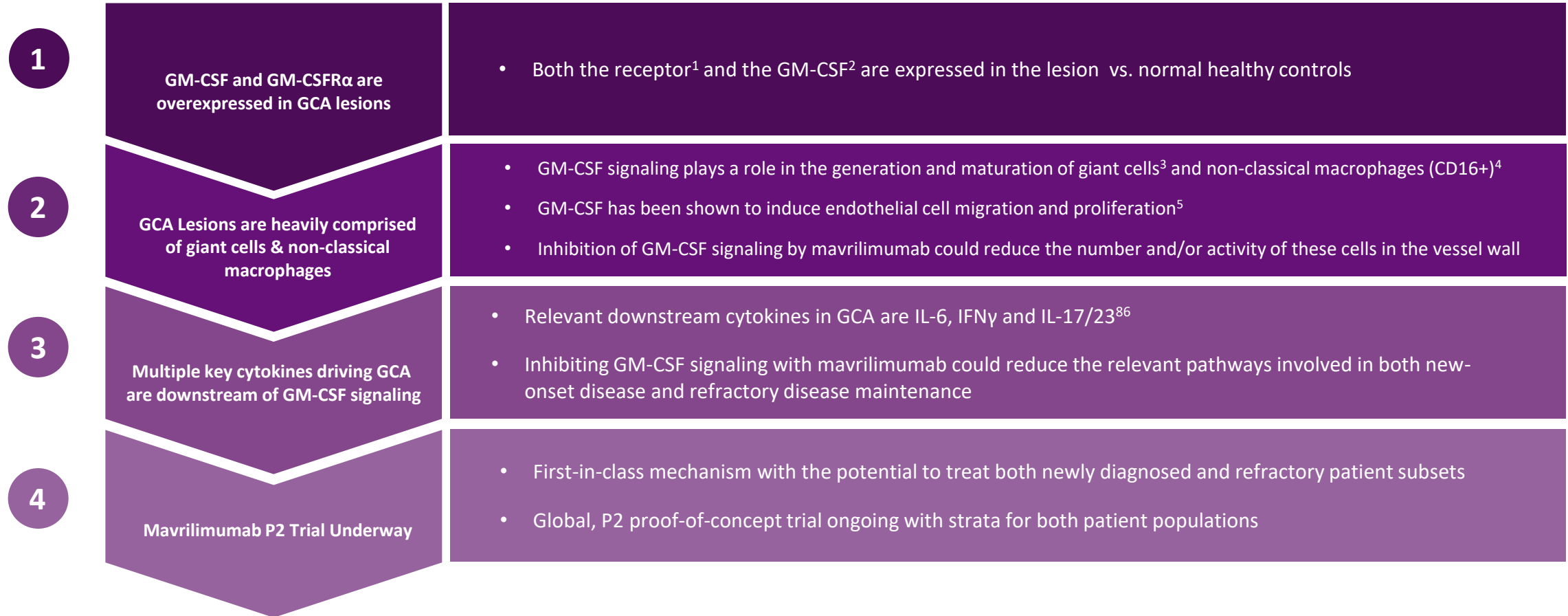
GCA U.S. Prevalence Estimated to be ~75-150k Patients



Key Considerations to Market Sizing Approach

Wide Range	Under-Representation	Under-Representation
High geographic variation GCA prevalence estimates vary across geographies with Northern European populations showing the highest rates and Asian populations the lowest Weighted by US demographics Given the demographic breakdown of the US, prevalence of GCA is likely ~75-150k (less than that of purely Northern Europeans, but more than estimates from Asian countries)	Represents Actively Managed Patients Medicare analysis does not capture GCA patients who were not actively managed within a given year; thus, the estimate from this analysis will exclude some remission patients or patients likely to relapse	Represents patients actively seen by a Rheum Rheumatologists reported the number of GCA patients they manage. Patients who are not actively managed would likely be excluded from these estimates

GM-CSF is a Key Growth Factor Believed to be Involved in the Pathology of GCA

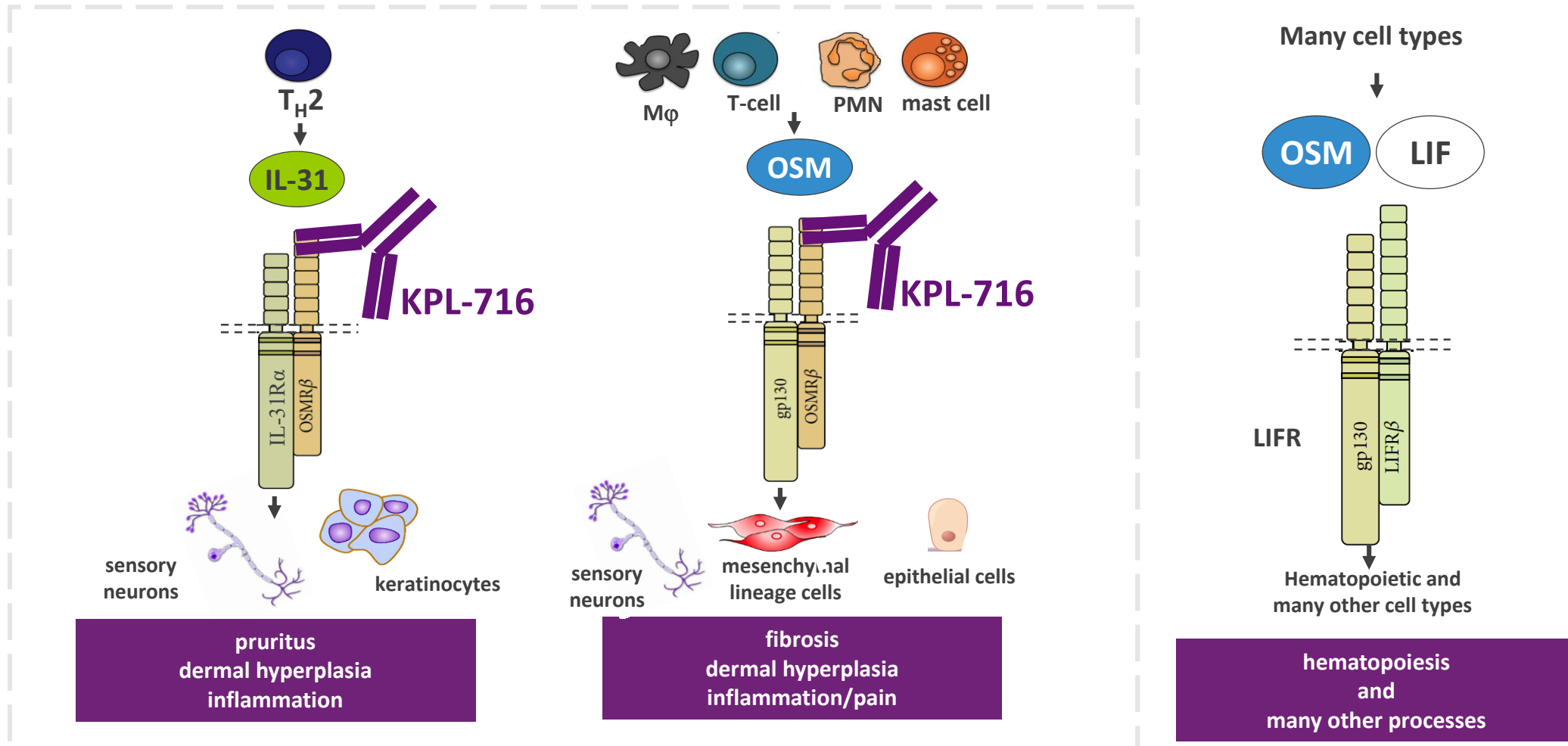




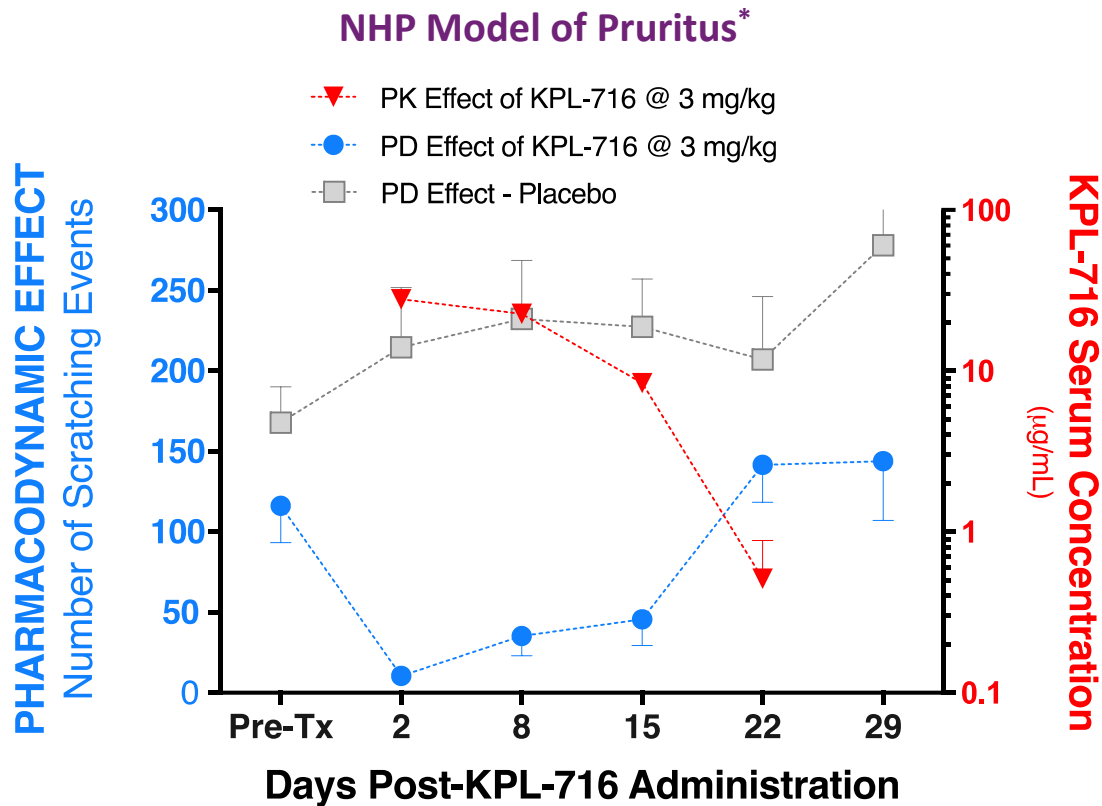
Every Second Counts!™

Appendix – KPL-716

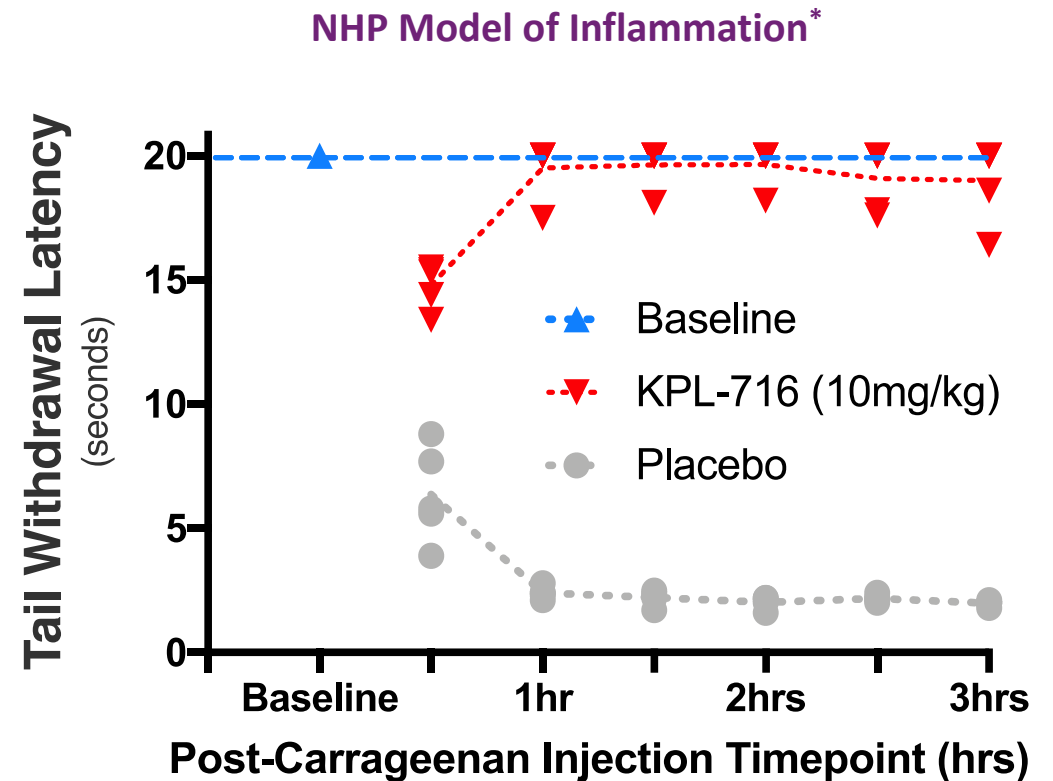
KPL-716 Inhibits IL-31 & OSM Signaling Through OSMR β but Avoids Inhibiting Signaling Critical to Hematopoiesis Through OSM/LIFR *in vitro* Studies



KPL-716 Inhibited Pruritic Response and Pain Reflex in Two Validated Non-Human Primate Models of Pruritus and Inflammation After a Single Dose

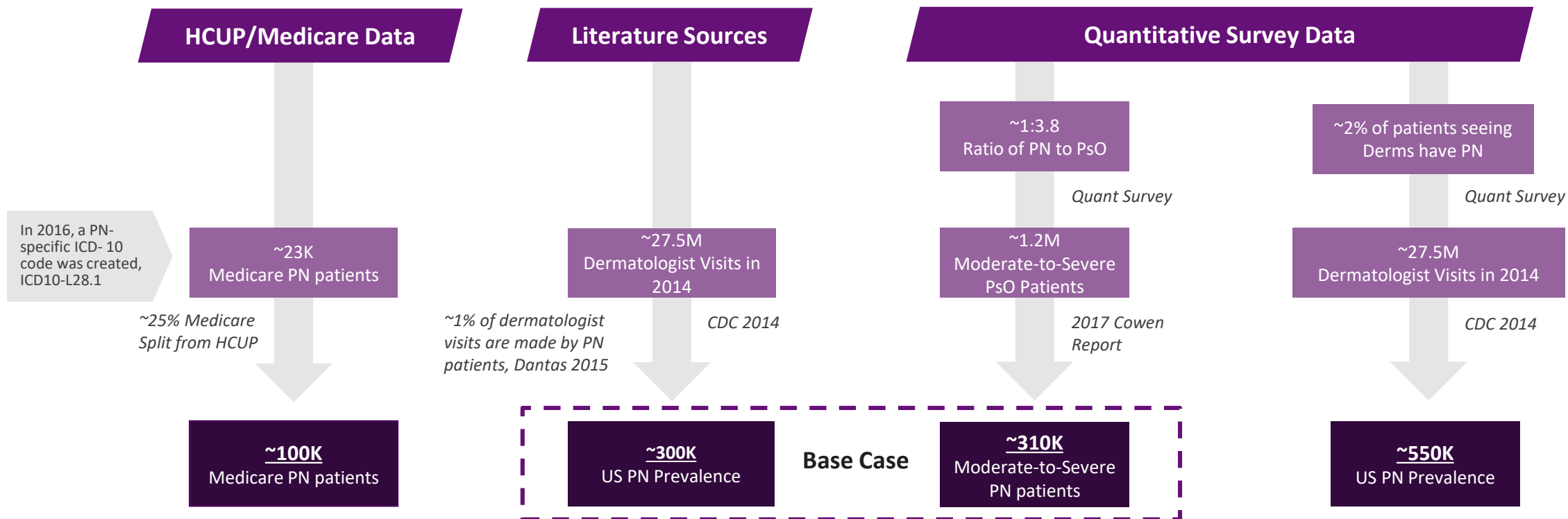


A single dose of KPL-716 at 3mg/kg inhibited pruritic response driven by supraphysiologic levels of IL-31 for over 2 weeks



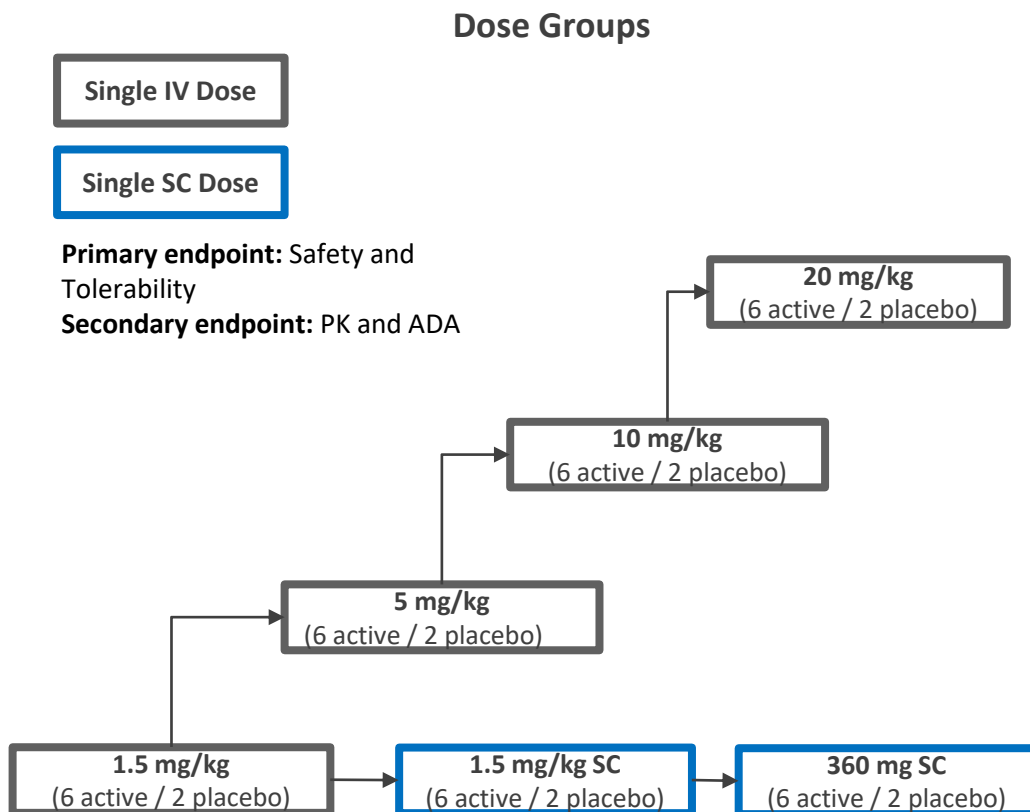
A single dose of KPL-716 at 10mg/kg increased tail withdrawal latency; implicates OSMR β in the inflammatory response

Prurigo Nodularis U.S. Prevalence Estimated to be ~300K Patients

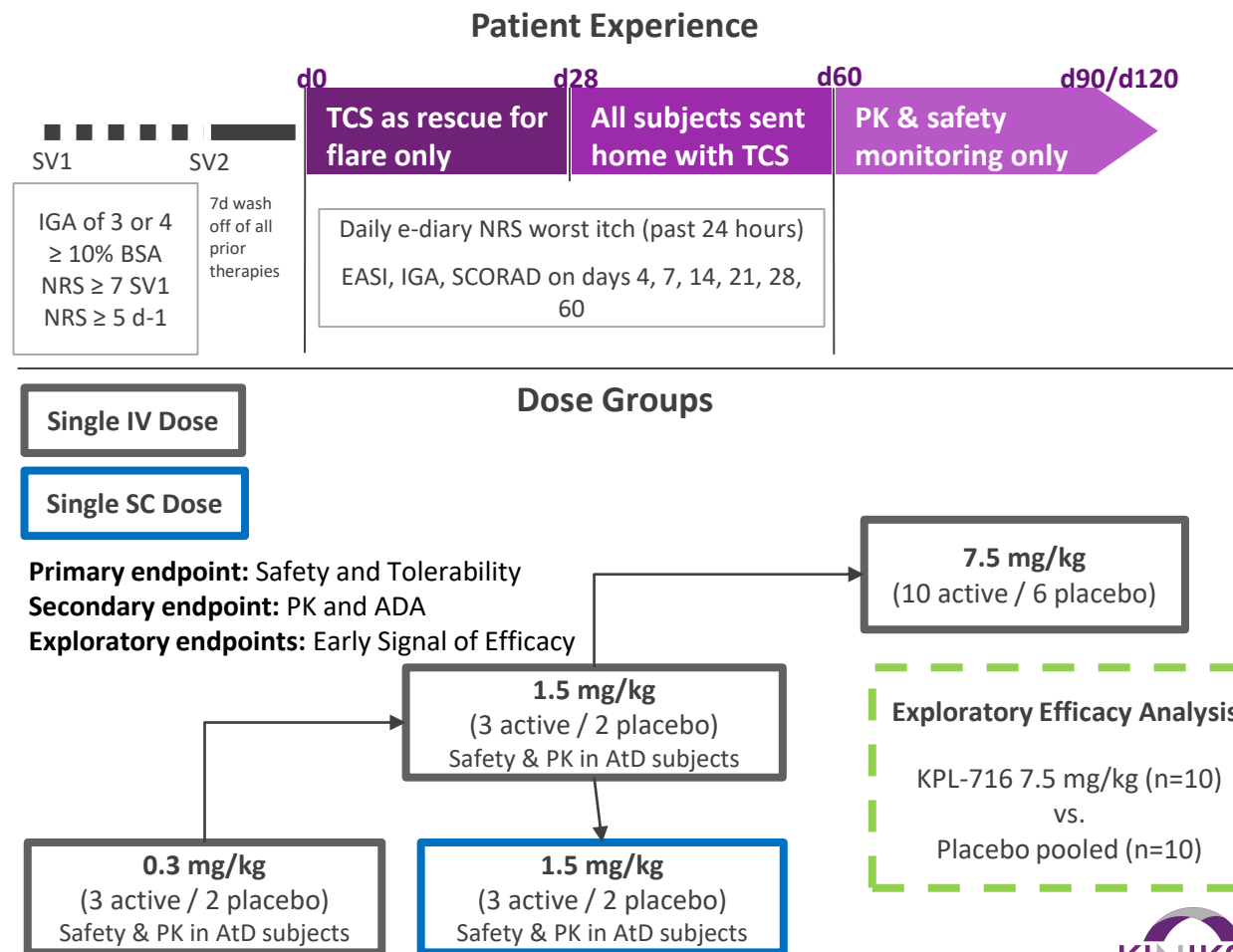


KPL-716 Placebo-Controlled, Single-Ascending-Dose Phase 1a/1b Study Design

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



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



KPL-716 was Well-Tolerated in Single-Dose Phase 1a/1b Study

- 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
- All resolved without sequelae

Normal Healthy Volunteers

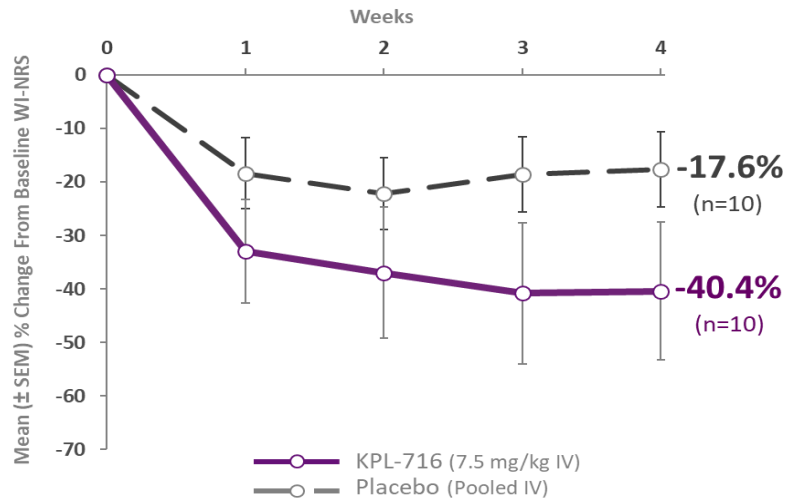
AE	KPL-716 (IV)					Placebo (IV)		KPL-716 (SC)		Placebo (SC)	
	1.5 mg/kg n=6	5 mg/kg n=6	10 mg/kg n=6	20 mg/kg n=6	Pooled n=8			1.5 mg/kg n=6	360 mg n=7	Pooled n=5	
DR-TEAE	0	Mild headache (n=1)	0	0	0			Mild flushing (n=1)	Mild anemia (n=1)	0	

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		

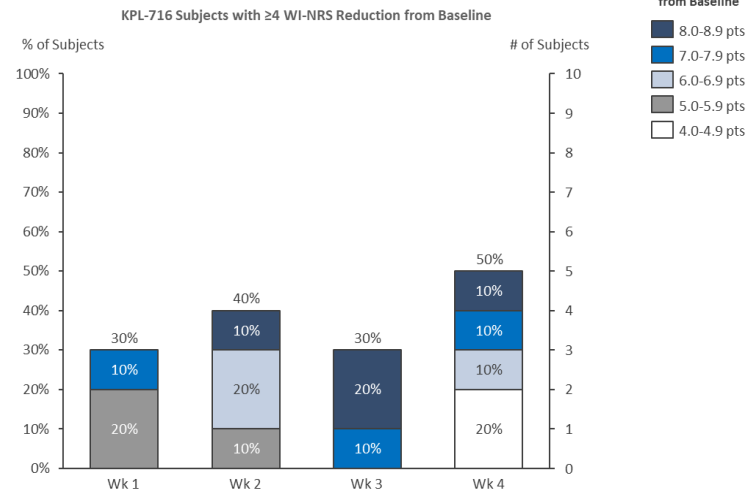
Single Doses of KPL-716 Provided Early Evidence Indicative of Target Engagement and Showed Reduction in Pruritus Over 28-Day Monotherapy Period¹

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

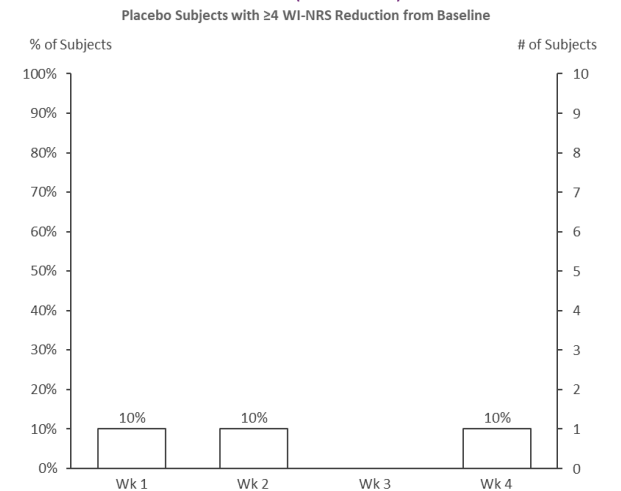


Mean % change in WI-NRS decreased by 40.4% in KPL-716 recipients compared to 17.6% decrease in placebo recipients at Day 28 in the absence of concomitant TCS

KPL-716 (7.5mg/kg IV)



Placebo (Pooled IV)

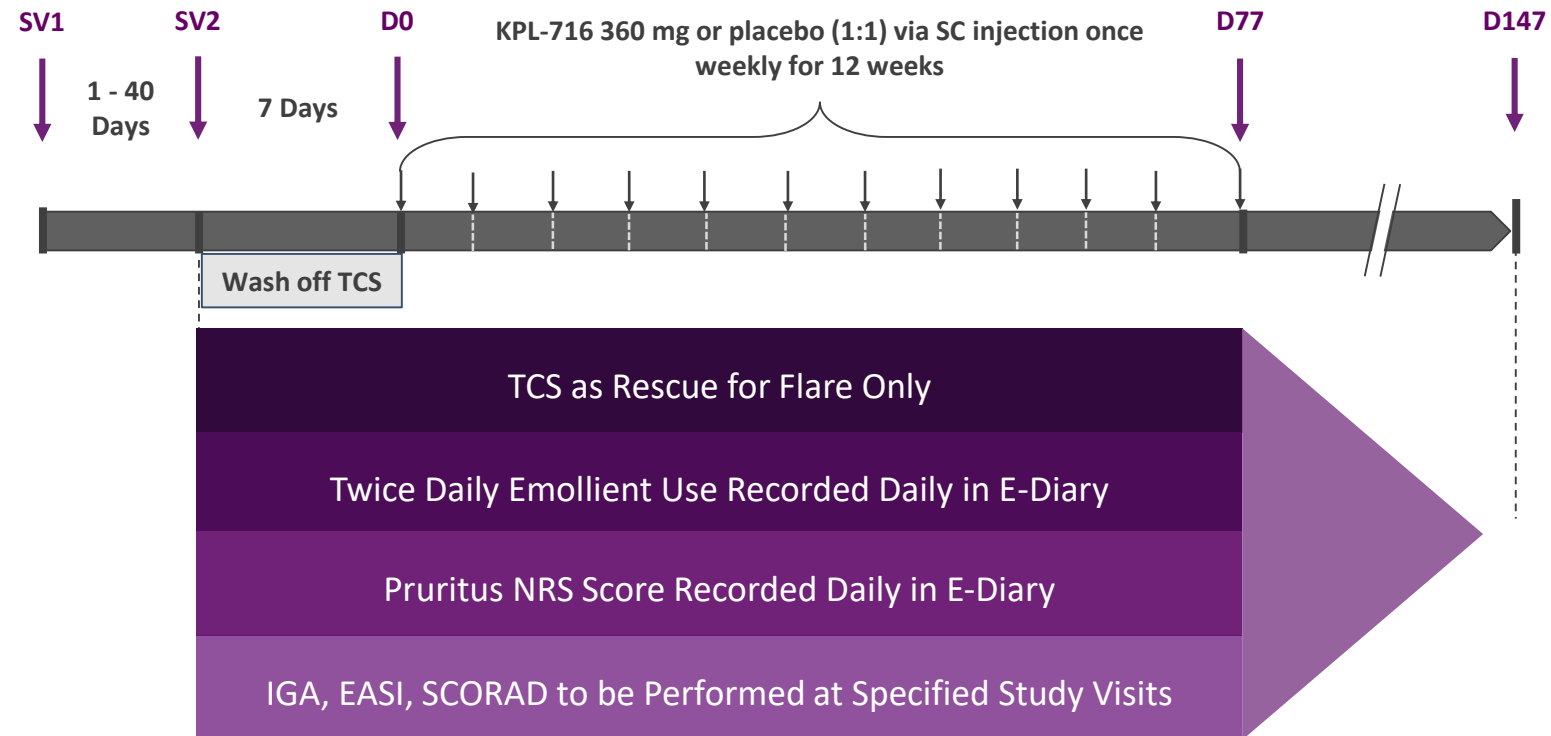


50% of KPL-716 recipients demonstrated a ≥ 4-point reduction in WI-NRS compared to 10% of placebo recipients at Day 28 in the absence of TCS

KPL-716 Placebo-Controlled Repeated-Single-Dose Phase 1b Study Design in Patients with Moderate-to-Severe Atopic Dermatitis

Key Inclusion Criteria:

- IGA of 3 or 4
- BSA $\geq 10\%$
- EASI ≥ 12
- NRS ≥ 7 at SV1
- NRS ≥ 5 at d0



Summary of Interim KPL-716 Phase 1b Repeated-Single-Dose Data

Enrolled 43 Subjects with Moderate-to-Severe Atopic Dermatitis Experiencing Moderate-to-Severe Pruritus

- Randomized 1:1 between weekly subcutaneous (SC) injections of either placebo or 360mg of KPL-716 for 12 weeks
- Interim data includes all subjects through the 12-week treatment period

Primary Endpoint: safety and tolerability of KPL-716

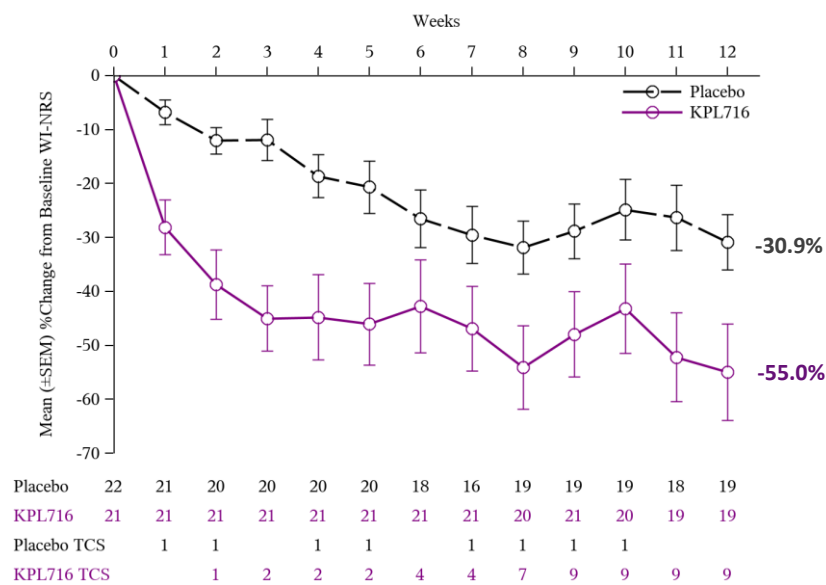
Exploratory Endpoints

- Worst-Itch Numerical Rating Score (WI-NRS) as recorded in a daily e-diary
- Measures of atopic dermatitis disease severity

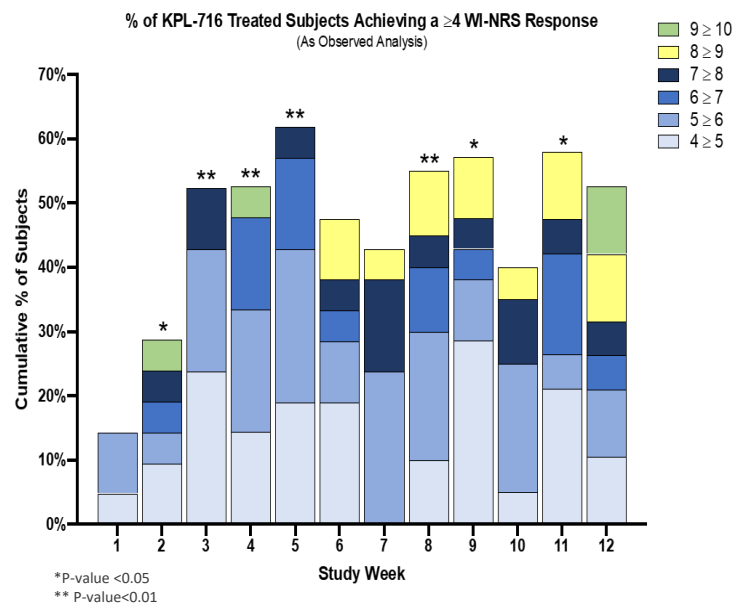
Topline Observations

- KPL-716 showed rapid and sustained reductions in pruritus versus placebo for the duration of the treatment period
 - The mean change from baseline in weekly-average WI-NRS at Week 1 was -28.1% in KPL-716 recipients compared to -6.8% in placebo recipients
 - The mean change from baseline in weekly-average WI-NRS at Week 12 was -55.0% in KPL-716 recipients compared to -30.9% in placebo recipients
 - 52.6% of KPL-716 recipients demonstrated a ≥ 4 -point reduction in weekly-average WI-NRS at Week 12 compared to 26.3% of placebo recipients
- There were no meaningful benefits of repeated-single-doses of KPL-716 on other efficacy endpoints specific to atopic dermatitis, including Eczema Area and Severity Index (EASI) and Scoring Atopic Dermatitis (SCORAD)
- There were no serious adverse events. However, there were more atopic dermatitis flares in KPL-716 recipients compared to placebo recipients (47.6% for the KPL-716 arm vs. 4.5% for the placebo arm) through the 12-week treatment period. KPL-716 was otherwise well-tolerated

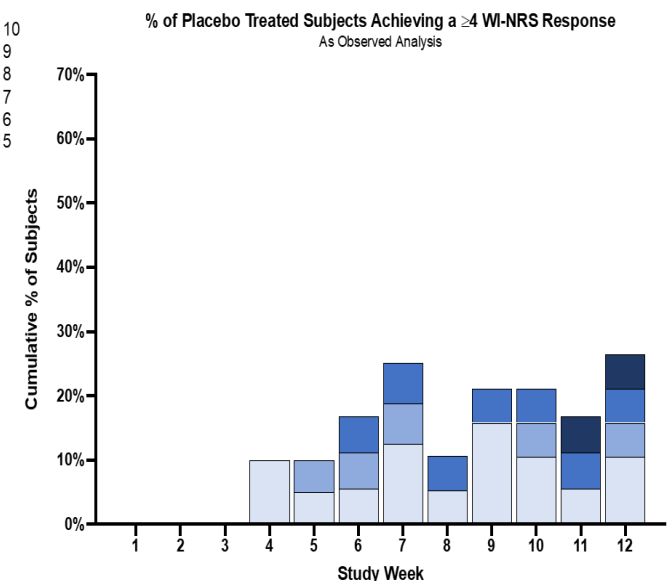
Repeated-Single-Doses of KPL-716 Showed Rapid and Sustained Reduction in Pruritus Versus Placebo¹



Mean % change in WI-NRS decreased by 55.0% in KPL-716 recipients compared to 30.9% decrease in placebo recipients at Week 12



A larger percentage of subjects in the KPL-716 arm achieved a ≥4-point change in weekly average WI-NRS versus placebo



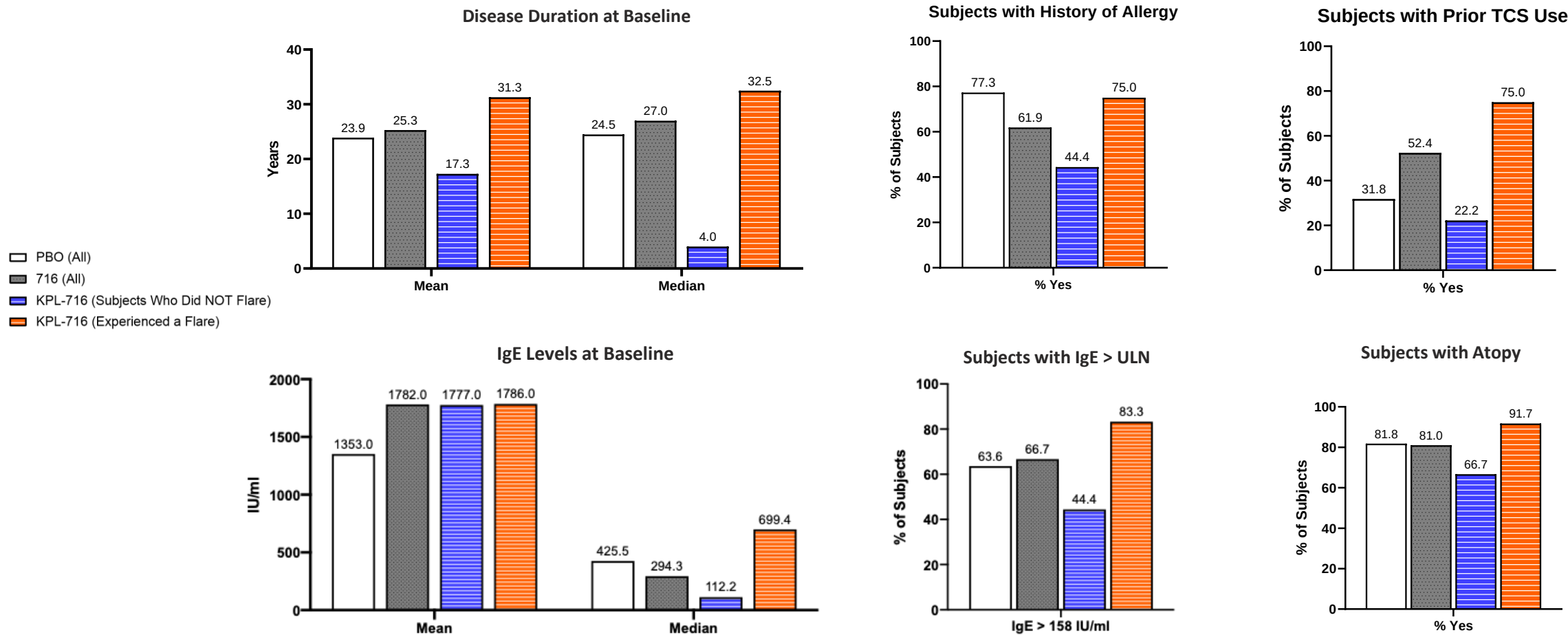
Overview of Treatment-Emergent Adverse Events (TEAE) Through 12-Week Treatment Period

	Placebo (N=22)	KPL-716 (N=21)
Any TEAE	12 (54.5%)	18 (85.7%)
Any Drug-Related TEAE	4 (18.2%)	8 (38.1%)
Any Moderate or Severe TEAE	6 (27.3%)	11 (52.4%)
Any Drug-Related Moderate or Severe TEAE	0	2 (9.5%)
Any Treatment-Emergent Serious AE	0	0
Any Drug-Related Serious TEAE	0	0
Any Atopic Dermatitis Flare-Related TEAE	1 (4.5%)	10 (47.6%)
Any Injection Site Reaction	2 (9.1%)	3 (14.3%)
Any TEAE Led to Dose Interruptions	1 (4.5%)	2 (9.5%)
Any TEAE Led to Study Drug Discontinuation	0	2 (9.5%)
Any TEAE Led to Death	0	0

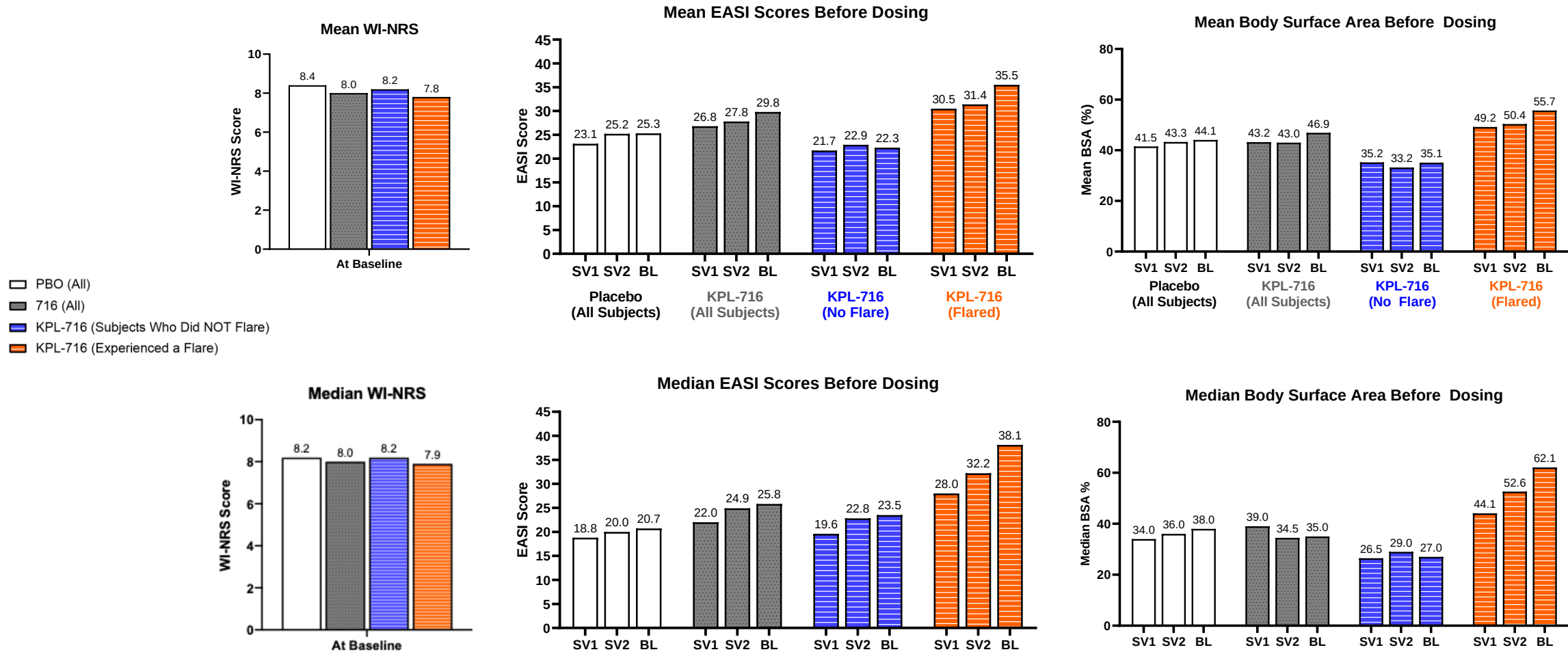
Moderate / Severe Drug-Related TEAE

	Placebo (N=22)	KPL-716 (N=21)
Subjects with At Least 1 Drug-related Moderate or Severe TEAE	0	2 (9.5%)
Infections and infestations	0	1 (4.8%)
Eczema impetiginous	0	1 (4.8%)
Psychiatric disorders	0	1 (4.8%)
Depression	0	1 (4.8%)
Skin and subcutaneous tissue disorders	0	1 (4.8%)
Dermatitis atopic	0	1 (4.8%)

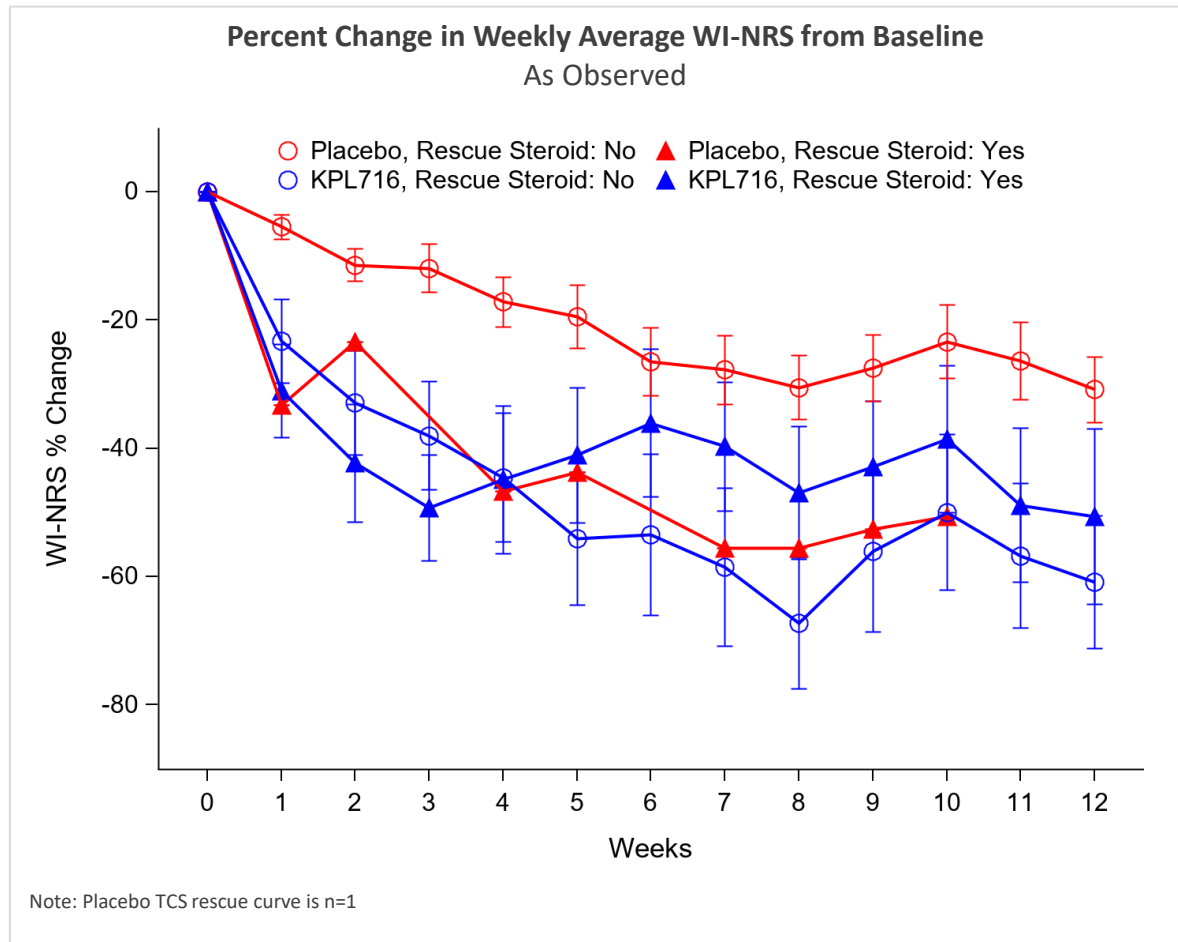
Baseline Subject Characteristics and Retrospective Groupings



Disease Characteristics at Baseline and Retrospective Groupings



KPL-716 Showed Rapid and Sustained Reduction in Pruritus in Patients Who Did Not Receive Topical Corticosteroid Rescue¹





Every Second Counts!™