Every Second Counts!™

Corporate Presentation

September 2019

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," "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 and corporate goals, potential acquisitions and collaborations, product development activities, clinical trials and other studies, regulatory and other applicable authority submissions, applications and approvals, our pre-commercial efforts, potential value drivers for the company, potential market opportunities and competitive position, and plans for 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 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 August 13, 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.

- ✓ Passionate Employees
- ✓ Sequential Pipeline
- ✓ Autoimmune and Autoinflammatory Diseases
- ✓ Strong Biologic Rationale or Validated Mechanisms
- ✓ Potential for Multiple Indications

Building a fully-integrated global biopharmaceutical company

Focusing on strong biologic rationale and/or validated mechanisms

Targeting underserved pockets of unmet medical need

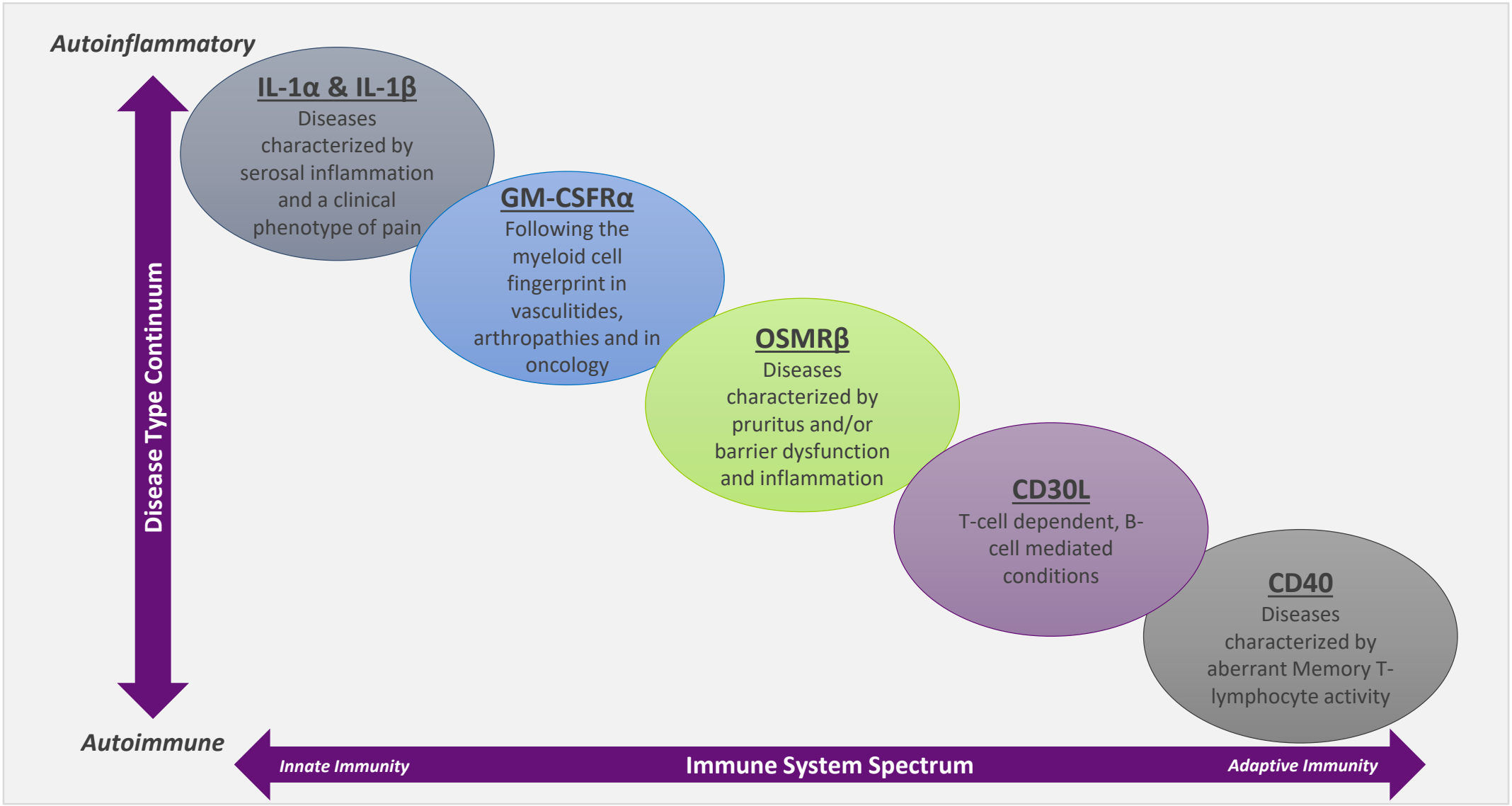
Acquiring/discovering molecules aimed at modulating central control nodes of the immune system

Allocating capital across the portfolio relative to the opportunity

Executing on communicated timelines
Every Second Counts!™

Discovering ◆ *Acquiring* ◆ *Developing* ◆ *Commercializing*

Development strategy focused on modulating central nodes of the immune system



Pipeline of product candidates across various stages of development

Program & Target	Indication	Preclinical / Phase 1 / Phase 2 / Phase 3	Status	Rights
Rilonacept[1] IL-1α & IL-1β	Recurrent Pericarditis		• Enrolling single, pivotal Phase 3 trial	Worldwide (excluding MENA)
Mavrilimumab GM-CSFRα	Giant Cell Arteritis (GCA)		• Enrolling global Phase 2 proof-of-concept trial	Worldwide
KPL-716 OSMRβ	Prurigo Nodularis (PN)		• Enrolling Phase 2a trial in PN	Worldwide
	Multiple Diseases Characterized by Chronic Pruritus[2]		• Enrolling exploratory Phase 2 study in diseases characterized by chronic pruritus	
KPL-404 CD40	Autoimmune		• Plan to file IND in 2H 2019	Worldwide
KPL-045 CD30L	Autoimmune		• Preclinical activities	Worldwide

1) Rilonacept (ARCALYST®) is approved and marketed for cryopyrin-associated periodic syndrome, in the United States by Regeneron Pharmaceuticals, Inc. We will assume the rights to this indication upon receiving approval for rilonacept in the recurrent pericarditis indication; 2) Chronic Idiopathic Pruritus, Chronic Idiopathic Urticaria, Plaque Psoriasis, Lichen Simplex Chronicus, Lichen Planus

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Initial indications are based on validated mechanisms and/or strong biologic rationale

Mechanism of Action	Rationale	Initial Indication
Rilonacept **IL-1α and IL-1β cytokine trap**	IL-1α and IL-1β are cytokines that have been shown to play a key role in inflammatory diseases[1]	Interim data from Phase 2 open-label study in subjects with **recurrent pericarditis** showed reduction in CRP and reported pain as well as increase in quality of life scores
Mavrilimumab **monoclonal antibody inhibitor blocking GM-CSFR signaling**	Reported data suggest GM-CSF is a key growth factor and cytokine in autoinflammation and autoimmunity[2]	GM-CSF and GM-CSFRα are both highly expressed in biopsies of **giant cell arteritis** patients vs. normal healthy controls
KPL-716 **monoclonal antibody inhibiting signaling through OSMRβ**	IL-31 and oncostatin M are two key cytokines implicated in inflammation, pruritus and fibrosis[3]	IL-31, OSM and OSMRβ mRNA are all upregulated in lesional biopsies of **prurigo nodularis** subjects vs. normal healthy controls

1) Dinarello CA, et al. Nat Rev Drug Discov 2012;11:633-652 and Brucato A, et al. Int Emerg Med 2018; 13:839–844; 2) Wicks, Roberts, Nature Review Immunology, 2015; Hamilton, Expert Review of Clinical Immunology, 11:4, 457-465; 3) Feeney et al., 2015 ACR/ARHP Annual Meeting, Abstract #1914; Ruzicka et al., N Engl J Med, 2017

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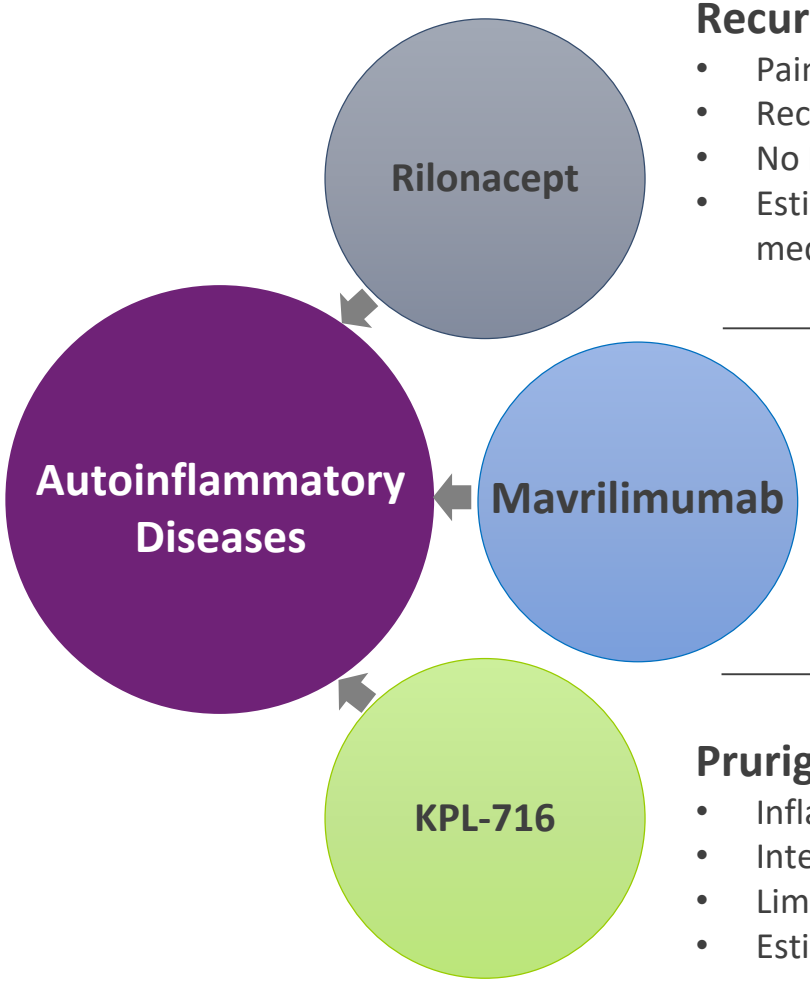
Targeted exploration of attractive commercial opportunities



Recurrent Pericarditis

- Painful inflammatory cardiovascular disease
- Recurrence burden impacts morbidity and impairs quality of life
- No FDA-approved therapies
- Estimated U.S. prevalence ~40K patients seeking and receiving medical treatment

Giant Cell Arteritis

- Chronic inflammation of medium-large blood vessels
- Acute events include permanent vision loss
- Only one FDA-approved therapy, but unmet need remains
- Estimated U.S. prevalence ~75K-150K patients

Prurigo Nodularis

- Inflammatory skin disease characterized by pruritic lesions
- Intense desire to scratch results in a decrease in quality of life
- Limited and ineffective treatment options
- Estimated U.S. prevalence ~300K patients

Expansion Potential

IL-1 mediated inflammatory cardiovascular conditions

Vasculitides and inflammatory cardiomyopathies

Chronic pruritic conditions where inflammation and fibrosis may be present

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Rilonacept – Phase 3

(IL-1α and IL-1β cytokine trap)

RHAPSODY

Rilonacept Mavrilimumab KPL-716 KPL-404 KPL-045

Opportunity in an inflammatory cardiovascular disease with no currently-approved therapies

Mechanism of Action[1]	IL-1α and IL-1β cytokine trap
Lead Indication	Recurrent Pericarditis (approved in the U.S. for CAPS[4], a rare autoinflammatory disease)
Addressable Population[2]	~14k patients in the U.S. (~3k refractory, ~6k poorly-controlled or steroid-dependent, ~5K steroid-intolerant)
Competition[3]	No FDA-approved therapies for recurrent pericarditis; differentiated from other marketed IL-1 agents
Clinical Development	Enrolling a global, pivotal Phase 3 clinical trial (RHAPSODY)
Rights	Worldwide (excluding MENA); BLA transfers to Kiniksa upon approval in recurrent pericarditis

1) Brucato et al. JAMA. 2016, 316 (18): 1906-1912; Arcalyst Prescribing Information; 2) IQVIA PharMetrics Plus Claims Data 1/1/2013-3/31/2018; ClearView Analysis, 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; 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; 4) Rilonacept (ARCALYST®) is approved and marketed for cryopyrin-associated periodic syndrome, in the United States by Regeneron Pharmaceuticals, Inc. We will assume the rights to this indication upon receiving approval for rilonacept in the recurrent pericarditis indication.

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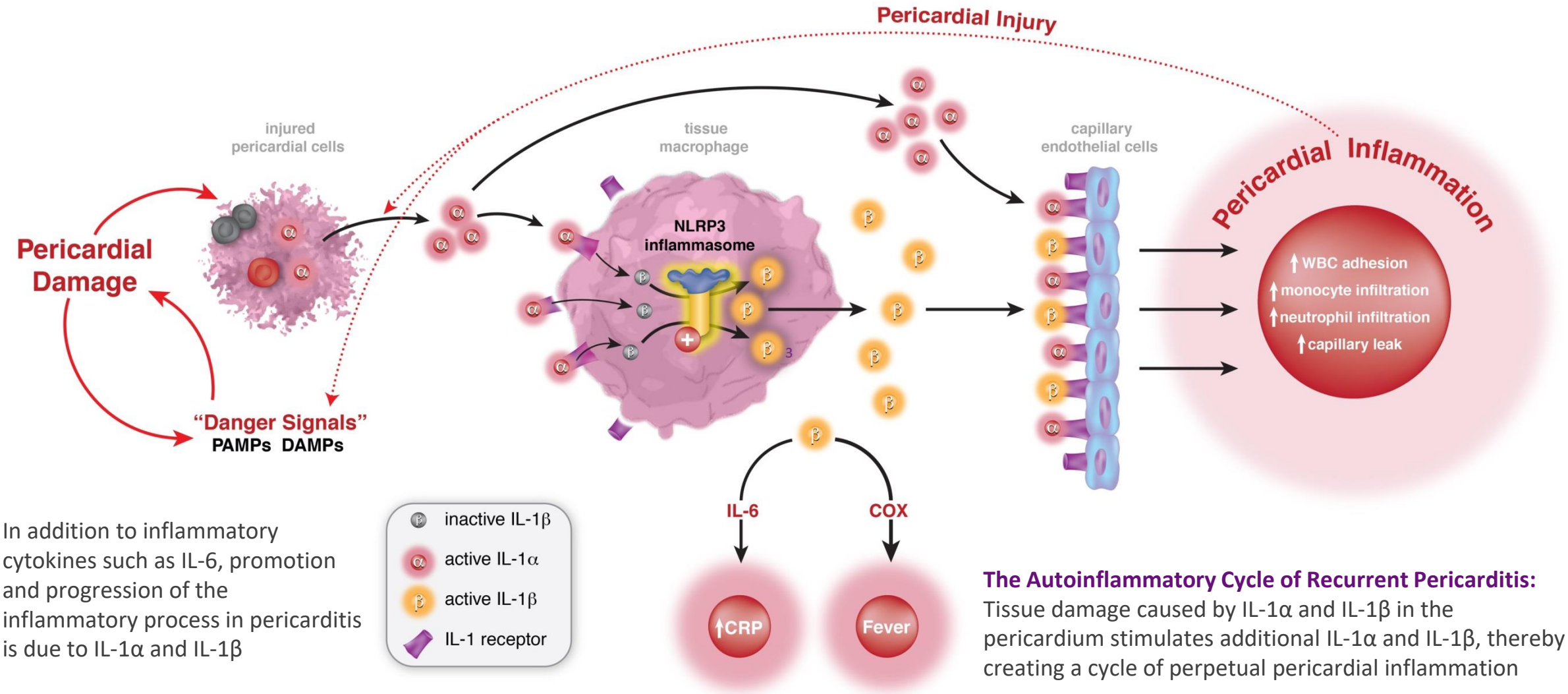
Role of IL-1α and IL-1β in the autoinflammatory cycle of recurrent pericarditis



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

The Autoinflammatory Cycle of Recurrent Pericarditis:
Tissue damage caused by IL-1α and IL-1β in the pericardium stimulates additional IL-1α and IL-1β, thereby creating a cycle of perpetual pericardial inflammation

CRP, C-reactive protein; DAMPs, damage-associated molecular patterns; IL, interleukin; PAMPs, pathogen-associated molecular patterns; WBC, white blood cell.

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

Recurrent pericarditis is a debilitating disease with no currently 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 and PR depression); (3) 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 Recurrent Idiopathic Pericarditis:

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

Recurrent pericarditis causes significant impairment of quality of life

Acute Episodes Have Favorable Prognosis:

- For most patients, acute pericarditis episodes last less than a few weeks and resolve on their own

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 surgery

1) Maish et al European Heart Journal 2004, 25, 587-610; 2) Alder et Al. European Heart Journal, 2015 ESC guidelines

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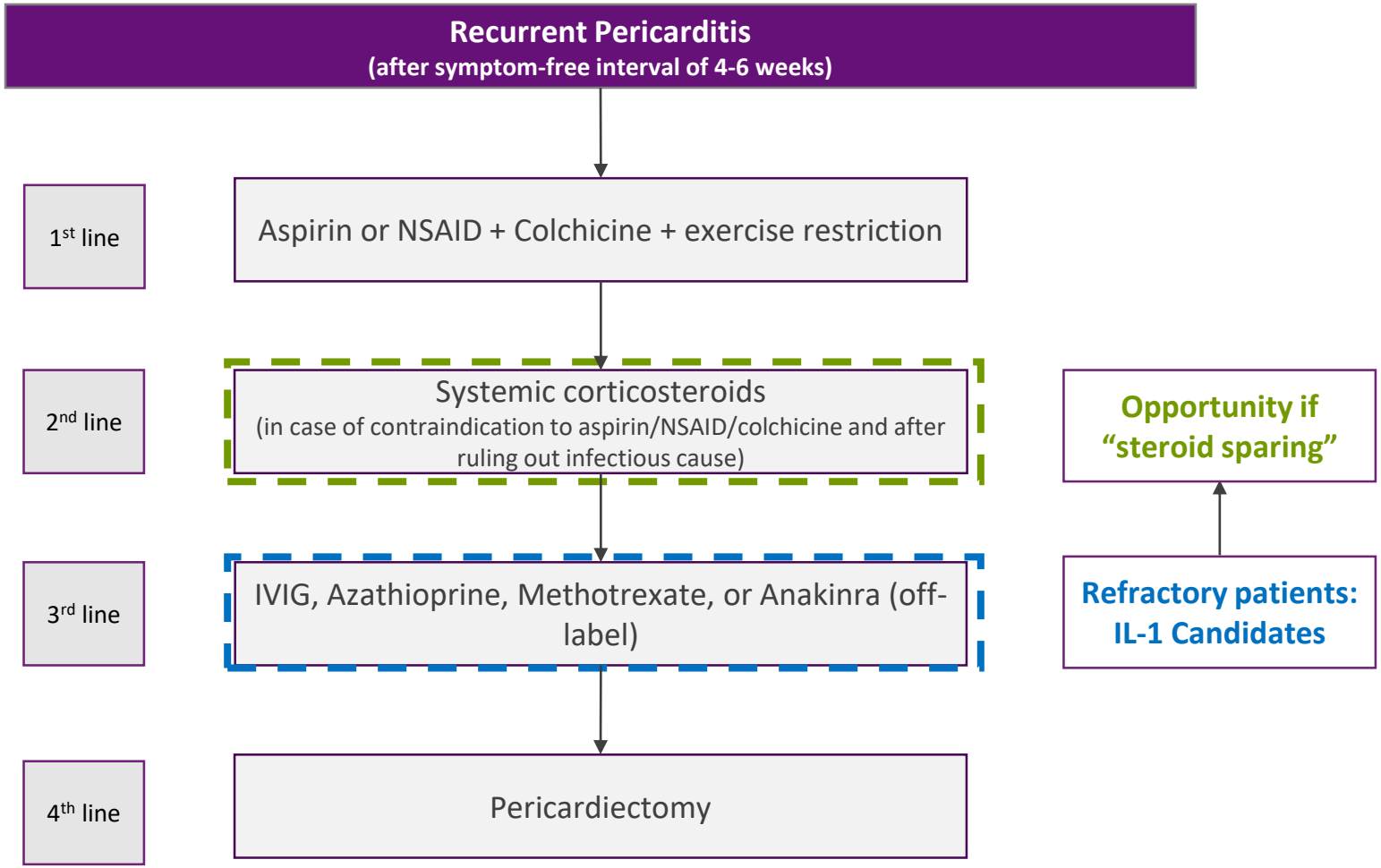
Refractory patients are left with few treatment options; mitigating the dangers of long-term steroid use is an important unmet medical need



Sources: UptoDate, Trinity Partners, Kiniksa Analysis

Patients with recurrent pericarditis have a high burden of disease that significantly impacts their overall health and quality of life

Impact of Pericarditis

30-40% of **refractory** and **steroid-dependent patients** experience ≥2 recurrences per year, significantly higher than the broad recurrent population

8% of refractory and steroid dependent patients experienced **cardiac tamponade** and 6% experienced **constrictive pericarditis** over the last 2 years

75% of refractory patients and 81% of steroid-dependent patients take **opioids** to deal with the intense pain associated with their disease

Unpredictability of disease activity causes **significant anxiety and depression**, resulting in disruption to day-to-day activities

Based on multiple claims data

Source: IQVIA PharMetrics Plus Claims Data 1/1/2013 – 3/31/2018; ClearView Analysis.

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Recurrent pericarditis prevalence in the U.S. estimated to be ~40k patients*

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

~3k

Refractory Patients

+

~6k

Poorly-Controlled or Steroid-Dependent Patients

+

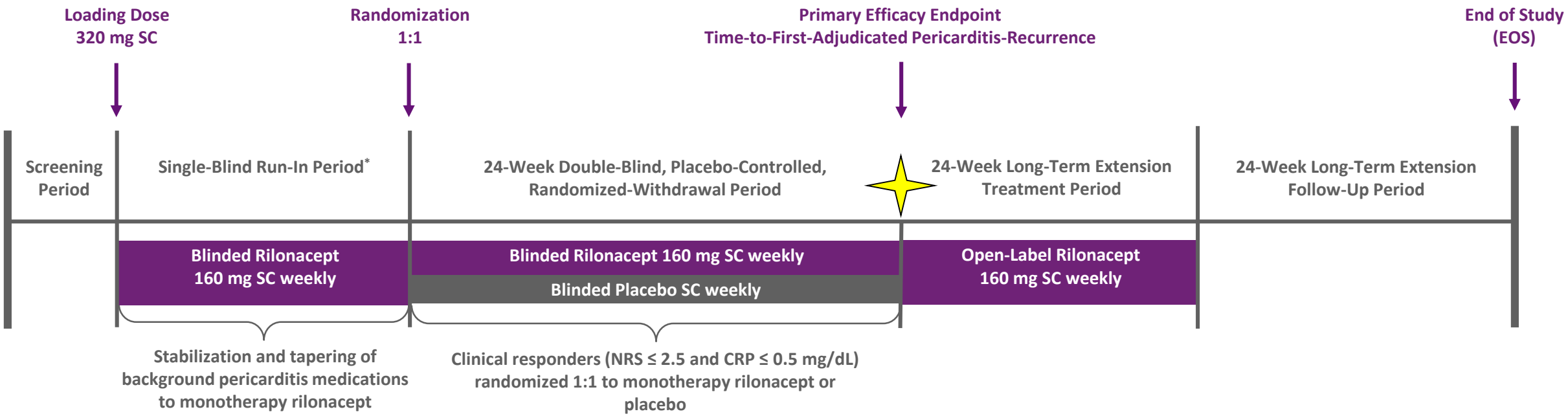
~5k

Steroid-Intolerant Patients Refractory to NSAIDs and Colchicine

Based on multiple claims data

* Estimates based upon the diagnosed and treated patients in the healthcare system per IQVIA PharMetrics Plus Claims Data 1/1/2013 – 3/31/2018; ClearView Analysis.

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):

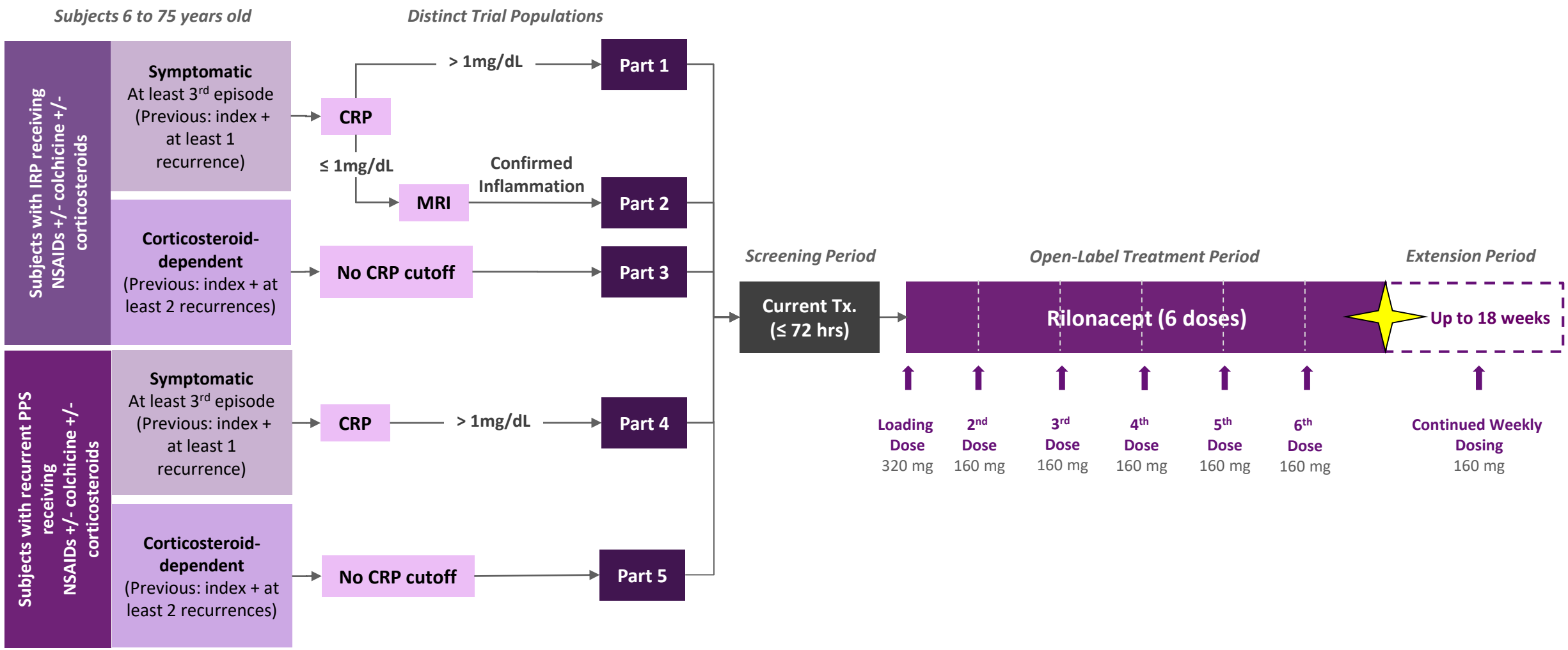
- Time-to-first-adjudicated pericarditis-recurrence in the RW period

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

* Duration of the run-in period undisclosed in order to maintain study subjects blinded to the start of the randomized-withdrawal period.

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



Open-label interim Phase 2 baseline demographic and clinical characteristics

Characteristic	Part 1	Part 2	Part 3	Part 4	Part 5	Total
Number of patients	12	3	6	1	3	25
Mean (SD) age, y	39.6 (10.2)	42.7 (15.0)	51.3 (7.8)	34.0	42.0 (7.2)	42.8 (10.5)
Female sex, n (%)	9 (75.0)	3 (100.0)	2 (33.3)	0	1 (33.3)	15 (60.0)
Race, n (%)						
White	10 (83.3)	2 (66.7)	6 (100.0)	1 (100.0)	3 (100.0)	22 (88.0)
Black/African American	2 (16.7)	1 (33.3)	0	0	0	3 (12.0)
Mean (SD) BMI, kg/m	30.2 (5.4)	40.0 (12.1)	31.1 (4.1)	29.3	24.7 (2.1)	30.9 (6.7)
Mean (SD) pain rating, NRS[a]	4.6 (1.7)	4.3 (2.5)	1.2 (0.8)	4.0	2.0 (2.7)	3.4 (2.2)
Mean (SD) baseline CRP, mg/dL	4.9 (5.8)	2.8[b] (4.0)	0.2 (0.1)	1.1	0.1 (0.04)	2.8 (3.3)
Pericarditis medications, n (%)						
Aspirin	0	0	2 (33.3)	1 (100.0)	0	3 (12.0)
NSAIDs	6 (50.0)	1 (33.3)	3 (50.0)	0	1 (33.3)	11 (44.0)
Colchicine	8 (66.7)	3 (100.0)	6 (100.0)	1 (100.0)	2 (66.7)	20 (80.0)
Corticosteroids	4 (33.3)	2 (66.7)	6 (100.0)	0	3 (100.0)	15 (60.0)

Note: Interim data from ongoing study as of January 23rd, 2019; BMI, body mass index; CRP, C-reactive protein; CS, corticosteroid; NRS, numeric rating scale; NSAID, nonsteroidal anti-inflammatory drug.

[a]11-point numeric scale, ranging from zero (0, no pain) to ten (10, pain as bad as possible); [b]CRP levels of patients enrolled in Part 2 were ≤1 mg/dL at screening; an error in the study database resulted in a mean number of 2.8 mg/dL and will be corrected at time of final data analysis.

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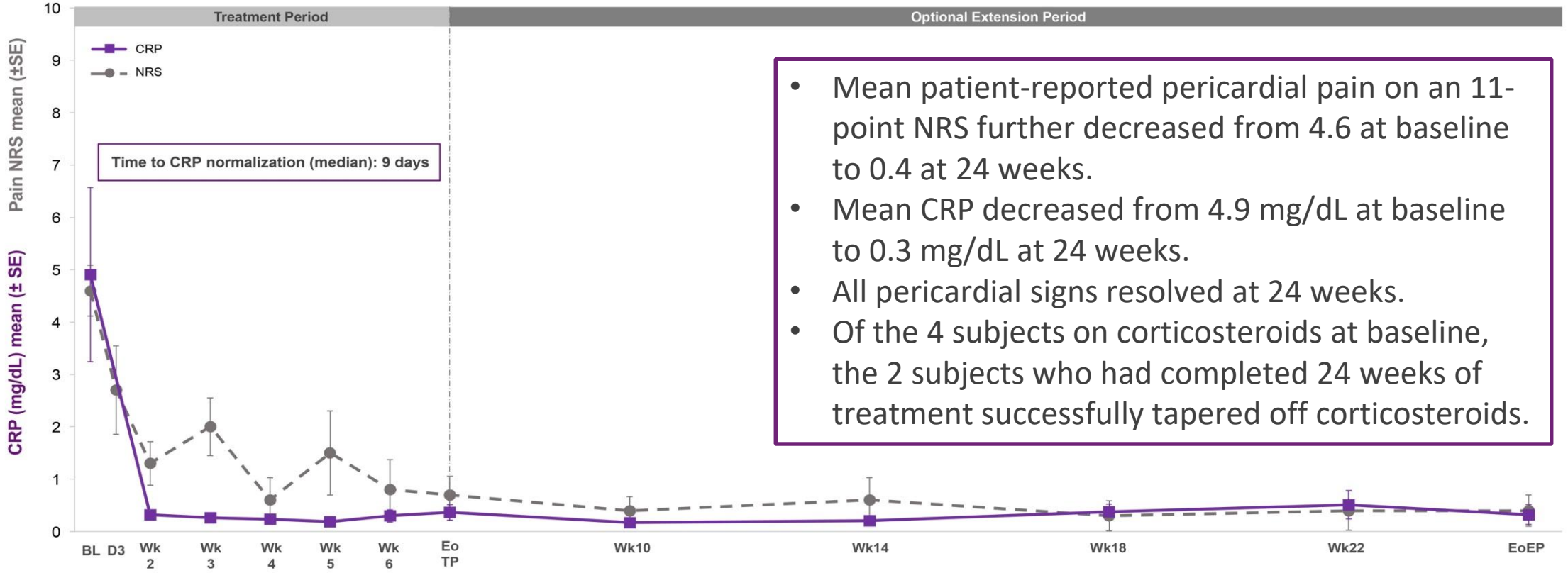
Open-label interim Phase 2 baseline demographic and clinical characteristics (cont'd)

Characteristic	Part 1	Part 2	Part 3	Part 4	Part 5	Total
Pericarditis medication categories, n (%)						
0	3 (25.0)	0	0	0	0	3 (12.0)
1	2 (16.7)	0	0	0	0	2 (8.0)
2	5 (41.7)	3 (100.0)	1 (16.7)	1 (100.0)	3 (100.0)	13 (52.0)
≥3	2 (16.7)	0	5 (83.3)	0	0	7 (28.0)
Number of previous pericarditis recurrences						
Mean	2.7	3.0	3.2	9.0	3.7	3.2

Note: Interim data from ongoing study of January 23rd, 2019

Open-label interim Phase 2 data: Part 1 showed reduction in both the inflammation biomarker (CRP) and reported pain (NRS)

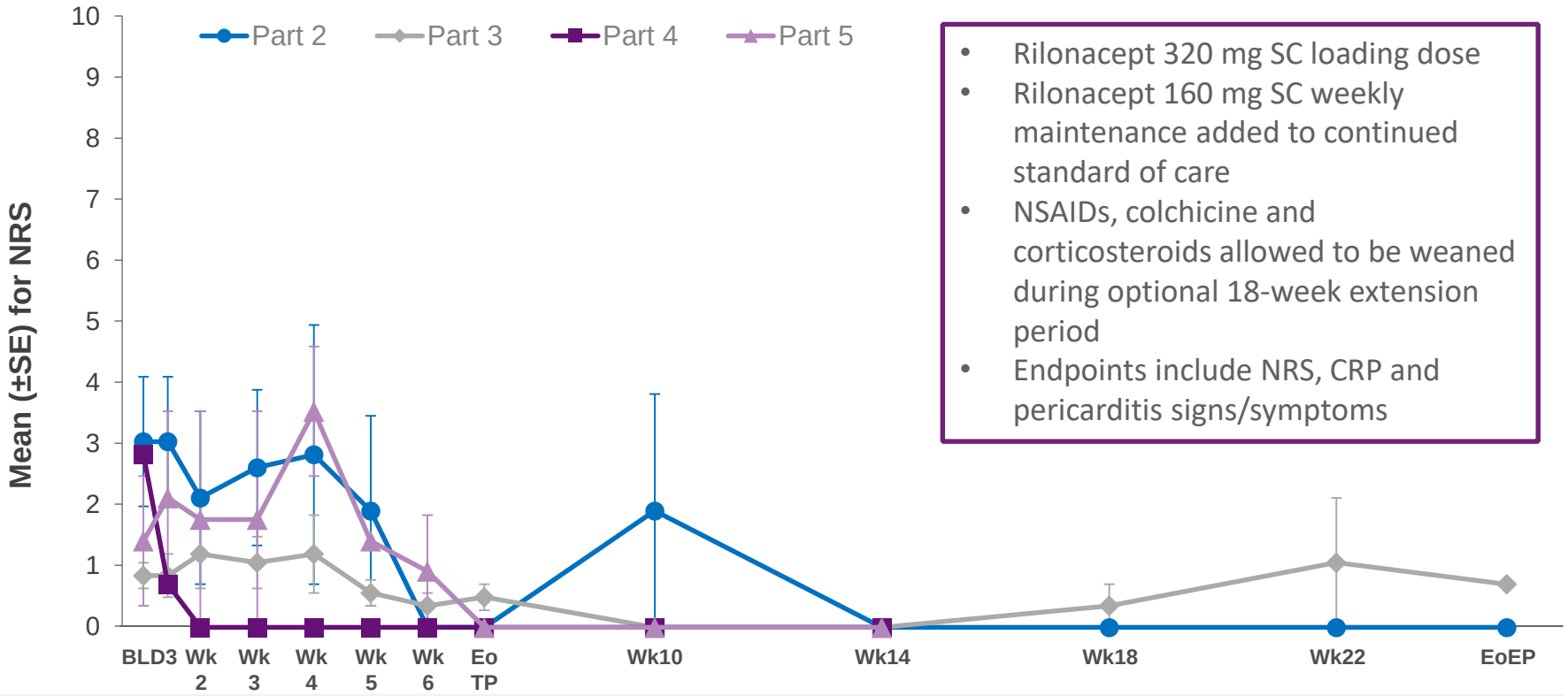


- Mean patient-reported pericardial pain on an 11-point NRS further decreased from 4.6 at baseline to 0.4 at 24 weeks.
- Mean CRP decreased from 4.9 mg/dL at baseline to 0.3 mg/dL at 24 weeks.
- All pericardial signs resolved at 24 weeks.
- Of the 4 subjects on corticosteroids at baseline, the 2 subjects who had completed 24 weeks of treatment successfully tapered off corticosteroids.

		BL	D3	Wk 2	Wk 3	Wk 4	Wk 5	Wk 6	EoTP	Wk10	Wk14	Wk18	Wk22	EoEP
CRP	# of Patients	12		9	11	10	10	6	10	10	11	7	5	7
CRP	CRP mean	4.91		0.32	0.26	0.24	0.19	0.30	0.36	0.17	0.21	0.38	0.51	0.32
Pain NRS	# of Patients	12	10	12	12	7	12	9	12	10	10	7	8	7
Pain NRS	Pain mean	4.6	2.7	1.3	2	0.60	1.5	0.8	0.7	0.4	0.6	0.3	0.4	0.4

Notes: Interim data from on-going study (Part1) as of Jan 23rd, 2019; Baseline (BL) = rilonacept 320mg loading dose; Week 1 through Week 6= rilonacept 160mg; EoEP = End of Extension Period; EoTP= End of Treatment Period; CRP = C-reactive protein; NRS = numeric rating scale

Open-label interim Phase 2 data: Parts 2 through Part 5 showed reduction in reported pain (NRS)

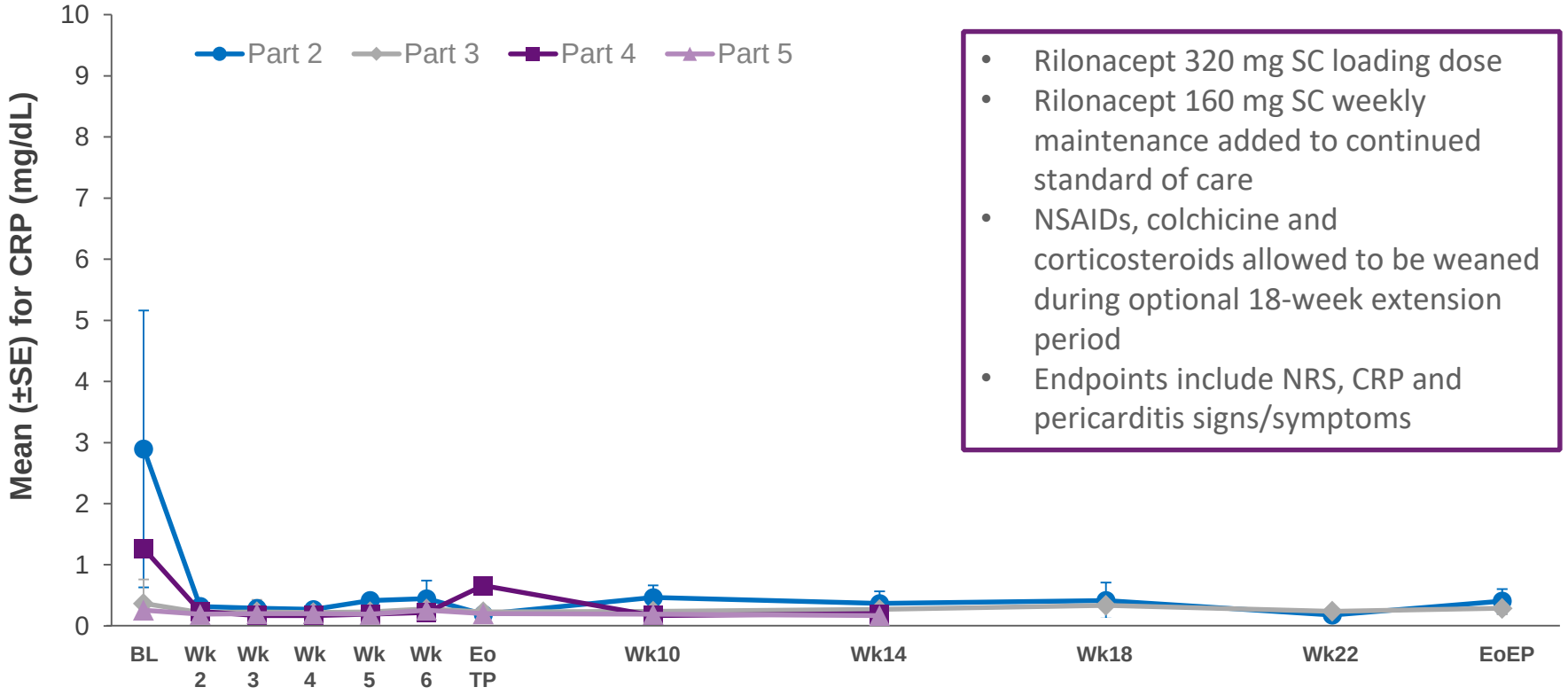


- Rilonacept 320 mg SC loading dose
- Rilonacept 160 mg SC weekly maintenance added to continued standard of care
- NSAIDs, colchicine and corticosteroids allowed to be weaned during optional 18-week extension period
- Endpoints include NRS, CRP and pericarditis signs/symptoms

		BL	D3	Wk 2	Wk 3	Wk 4	Wk 5	Wk 6	EoTP	Wk10	Wk14	Wk18	Wk22	EoEP
Part 2	# of Patients	3	3	3	3	2	3	2	2	3	2	2	2	2
	NRS mean	4.3	4.3	3.0	3.7	4.0	2.7	0	0	2.7	0	0	0	0
Part 3	# of Patients	6	6	6	6	3	6	6	6	5	4	2	2	2
	NRS mean	1.2	1.2	1.7	1.5	1.7	0.8	0.5	0.7	0	0	0.5	1.5	1.0
Part 4	# of Patients	1	1	1	1	1	1	1	1	1	1	0	0	0
	NRS mean	4.0	1.0	0	0	0	0	0	0	0	0	0	0	0
Part 5	# of Patients	3	2	2	2	1	3	3	3	2	1	0	0	0
	NRS mean	2.0	3.0	2.5	2.5	5.0	2.0	1.3	0	0	0	N/A	N/A	N/A

Notes: Interim data from on-going study (Parts 2-5) as of Jan 23rd, 2019; Baseline (BL) = rilonacept 320mg loading dose; Week 1 through Week 6= rilonacept 160mg; EoEP = End of Extension Period; EoTP= End of Treatment Period; CRP = C-reactive protein; NRS = numeric rating scale

Open-label interim Phase 2 data: Parts 2 through Part 5 showed reduction in the inflammation biomarker (CRP)



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- Endpoints include NRS, CRP and pericarditis signs/symptoms

		BL	Wk 2	Wk 3	Wk 4	Wk 5	Wk 6	EoTP	Wk10	Wk14	Wk18	Wk22	EoEP
Part 2	# of Patients	3	3	3	3	3	2	1	3	2	2	1	2
	CRP mean	2.8	0.2	0.2	0.1	0.3	0.3	0.1	0.3	0.2	0.3	0	0.3
Part 3	# of Patients	6	6	5	5	6	3	6	5	5	2	2	2
	CRP mean	0.2	0.1	0.1	0.1	0.1	0.1	0.1	0.1	0.1	0.2	0.1	0.2
Part 4	# of Patients	1	1	1	1	1	1	1	1	1	0	0	0
	CRP mean	1.1	0.1	0	0	0.1	0.1	0.5	0	0.1	N/A	N/A	N/A
Part 5	# of Patients	2	2	2	1	2	1	2	2	1	0	0	0
	CRP mean	0.1	0.1	0.1	0.1	0.1	0.1	0.1	0	0	N/A	N/A	N/A

Notes: Interim data from on-going study (Parts 2-5) as of Jan 23rd, 2019; Baseline (BL) = rilonacept 320mg loading dose; Week 1 through Week 6= rilonacept 160mg; EoEP = End of Extension Period; EoTP= End of Treatment Period; CRP = C-reactive protein; NRS = numeric rating scale

Open-label Phase 2 data: resolution of pericardial signs

Time Point	Part 1 n/N (%)	Part 2 n/N (%)	Part 3 n/N (%)	Part 4 n/N (%)	Part 5 n/N (%)
Baseline					
Widespread ST elevation	2/12 (16.7)	0/3	0/6	0/1	0/3
PR depression	3/12 (25.0)	0/3	0/6	0/1	0/3
Pericardial rub	2/12 (16.7)	0/3	0/6	0/1	0/2
Fever	0/12	0/3	0/6	0/1	0/3
Pericardial effusion on ECHO	7/12 (58.3)	0/3	2/6 (33.3)	0/1	0/2
End of TP (visit 7)					
Widespread ST elevation	0/12	0/2	0/6	0/1	0/3
PR depression	1/12 (8.3)	0/2	0/6	0/1	0/3
Pericardial rub	0/11	0/2	0/6	0/1	0/3
Fever	0/12	0/2	0/6	0/1	0/3
Pericardial effusion on ECHO	1/12 (8.3)	0/2	1/6 (16.7)	0/1	0/3
Final visit					
Widespread ST elevation	0/7	0/2	0/2	0/0	0/0
PR depression	0/7	0/2	0/2	0/0	0/0
Pericardial rub	0/7	0/2	0/2	0/0	0/0
Fever	0/7	0/2	0/2	0/0	0/0
Pericardial effusion on ECHO	0/7	0/1	0/2	0/0	0/0

Notes: Interim data from ongoing study as of January 23rd, 2019; ECHO, echocardiography; TP, treatment period.

Open-label interim Phase 2 data: quality of life improvement as assessed by PROMIS questionnaire

Domain	Part 1 (n=12)	Part 2 (n=3)	Part 3 (n=6)	Part 4 (n=1)	Part 5 (n=3)
Global Physical Health, mean (SD)					
Baseline	41.3 (8.6)	36.2 (12.1)	43.7 (6.5)	34.9	42.3 (0.0)
End of TP (visit 7)	51.0 (8.1)	57.7 (0.0)	45.2 (4.8)	42.3	44.0 (1.5)
Final visit	50.5 (7.2)	58.0 (5.5)	42.2 (21.9)	N/A	N/A
Global Mental Health, mean (SD)					
Baseline	46.8 (9.5)	41.4 (14.2)	47.7 (9.2)	31.3	43.5 (0.0)
End of TP (visit 7)	50.9 (10.6)	56.2 (4.0)	49.3 (6.2)	28.4	45.1 (2.8)
Final visit	51.6 (10.6)	63.3 (6.1)	48.6 (10.5)	N/A	N/A

Notes: Interim data from ongoing study as of January 23rd, 2019; PROMIS, Patient-Reported Outcomes Measurement Information System; TP, treatment period. N/A: not available; corresponding data collection is ongoing.

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Open-label interim Phase 2 data: changes in concomitant corticosteroids and treatment - retreatment with rilonacept

Changes in concomitant corticosteroids (CS) for recurrent pericarditis during the study

15/25 patients received CS at baseline; of these 15 patients, 5 completed 24 weeks of treatment and successfully tapered and discontinued CS:

- 1 patient discontinued CS early during the 6-week base treatment period and remained off CS throughout the study
- 4 additional patients discontinued CS in the 18-week extension period (2 in Part 1 and 2 in Part 3)

Treatment-Retreatment with rilonacept during the study

- One Part 1 patient, who completed the 6-week base TP treatment period and the 18-week extension period (symptom-free, normalized CRP), experienced recurrence of pericarditis symptoms requiring addition of celecoxib approximately 8 weeks after completing rilonacept treatment.
- The patient subsequently experienced a frank recurrence of pericarditis with tamponade physiology and re-enrolled into the study, resulting in rapid reductions in CRP and pericardial pain after re-initiation of rilonacept treatment.

Notes: Interim data from on-going study as of Jan 23rd, 2019

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Open-label Phase 2 data: summary of adverse events

Treatment-Related and Non-Treatment-Related TEAEs

Category	Part 1 (n=12)	Total (N=25)
Patients with ≥1 AE, n (%)	12 (100.0)	23 (92.0)
Patients with ≥1 TEAE, n (%)	12 (100.0)	23 (92.0)
Patients with ≥1 treatment-related TEAE, n (%)	9 (75.0)	17 (68.0)
Patients with ≥1 serious TEAE, n (%)	2 (16.7)	2 (8.0)
Patients with ≥1 treatment-related serious TEAE, n (%)	1 (8.3)	1 (4.0)
Patients with ≥1 TEAE leading to treatment discontinuation, n (%)	1 (8.3)	1 (4.0)
Patients with ≥1 TEAE leading to death, n (%)	0	0
Patients with TEAEs by severity, n (%)		
Mild	9 (75.0)	18 (72.0)
Moderate	2 (16.7)	4 (16.0)
Severe	1 (8.3)	1 (4.0)

Notes: Interim data from on-going study as of Jan 23rd, 2019 AE, adverse event; TEAE, treatment-emergent adverse event.

AEs Occurring at Least Once (by Affected Organ System)

System Organ Class	Part 1 (n=12)	Total (N=25)
General disorders and administration site conditions, n (%)	6 (50.0)	15 (60.0)
Infections and infestations, n (%)	5 (41.7)	7 (28.0)
Musculoskeletal and connective tissue disorders, n (%)	3 (25.0)	7 (28.0)
Gastrointestinal disorders, n (%)	6 (50.0)	6 (24.0)
Investigations, n (%)	2 (16.7)	6 (24.0)
Respiratory, thoracic, and mediastinal disorders, n (%)	0	3 (12.0)
Ear and labyrinth disorders, n (%)	2 (16.7)	2 (8.0)
Skin and subcutaneous tissue disorders, n (%)	0	2 (8.0)
Cardiac disorders, n (%)	0	1 (4.0)
Eye disorders, n (%)	1 (8.3)	1 (4.0)
Nervous system disorders, n (%)	1 (8.3)	1 (4.0)
Unspecified, n (%)	1 (8.3)	1 (4.0)

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Summary of rilonacept profit share arrangement with Regeneron[1]

Rilonacept Net Sales (CAPS + Recurrent Pericarditis)[2]

Minus 100% of Cost of Goods Sold[3]

Minus 100% of Certain Maintenance Costs

Minus 100% of Field Force Costs

Minus Marketing & Certain Other Commercial Expenses (Subject to Specified Limits)

Calculated Rilonacept Operating Profit to be Shared

Minus 50% of Shared Rilonacept Operating Profit (Booked as COGS on P&L)

Minus R&D Expenses for Additional Indications or Other Studies Req'd for Approval

Minus Marketing & Commercial Expenses That Exceeded Specified Limits (if Any)

Kiniksa Operating Income from Rilonacept

- 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 CAPS[4], DIRA[5], 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 MENA[6]
- Upon receipt of positive Phase 3 clinical data, the BLA[7] 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

1) Subject to description contained in definitive agreement; 2) Global net sales for CAPS and recurrent pericarditis recognized as revenue on Kiniksa's income statement; 3) Including cost of product purchased from Regeneron; 4) CAPS = Cryopyrin-Associated Periodic Syndromes; 5) DIRA = deficiency of the interleukin-1 receptor antagonist 6) MENA = Middle East and North Africa; 7) BLA = Biologics License Application

Mavrilimumab – Phase 2

(monoclonal antibody inhibitor targeting GM-CSFRα)

Rilonacept | **Mavrilimumab** | KPL-716 | KPL-404 | KPL-045

Mechanistic rationale for focusing on high unmet need vasculitides & inflammatory cardiomyopathies

Mechanism of Action[1]	Monoclonal antibody inhibitor targeting GM-CSFRα; a key mediator of inflammation and autoimmunity
Lead Indication	Giant Cell Arteritis (GCA)
Addressable Population[2]	~75k - 150k prevalent in the U.S.; similar prevalence in other major markets
Competition[3]	Only one FDA-approved therapy for GCA and unmet needs remain
Clinical Development	Enrolling a global Phase 2 proof-of-concept clinical trial
Rights	Worldwide

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

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GCA is a serious condition characterized by inflammation of medium-large blood vessels; it can lead to bilateral blindness if left untreated

1 Chronic Inflammation of Medium-Large Blood Vessels

- GCA is characterized by inflammation of medium-large blood vessels 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**; treatment with high-dose steroids effectively prevents complications

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

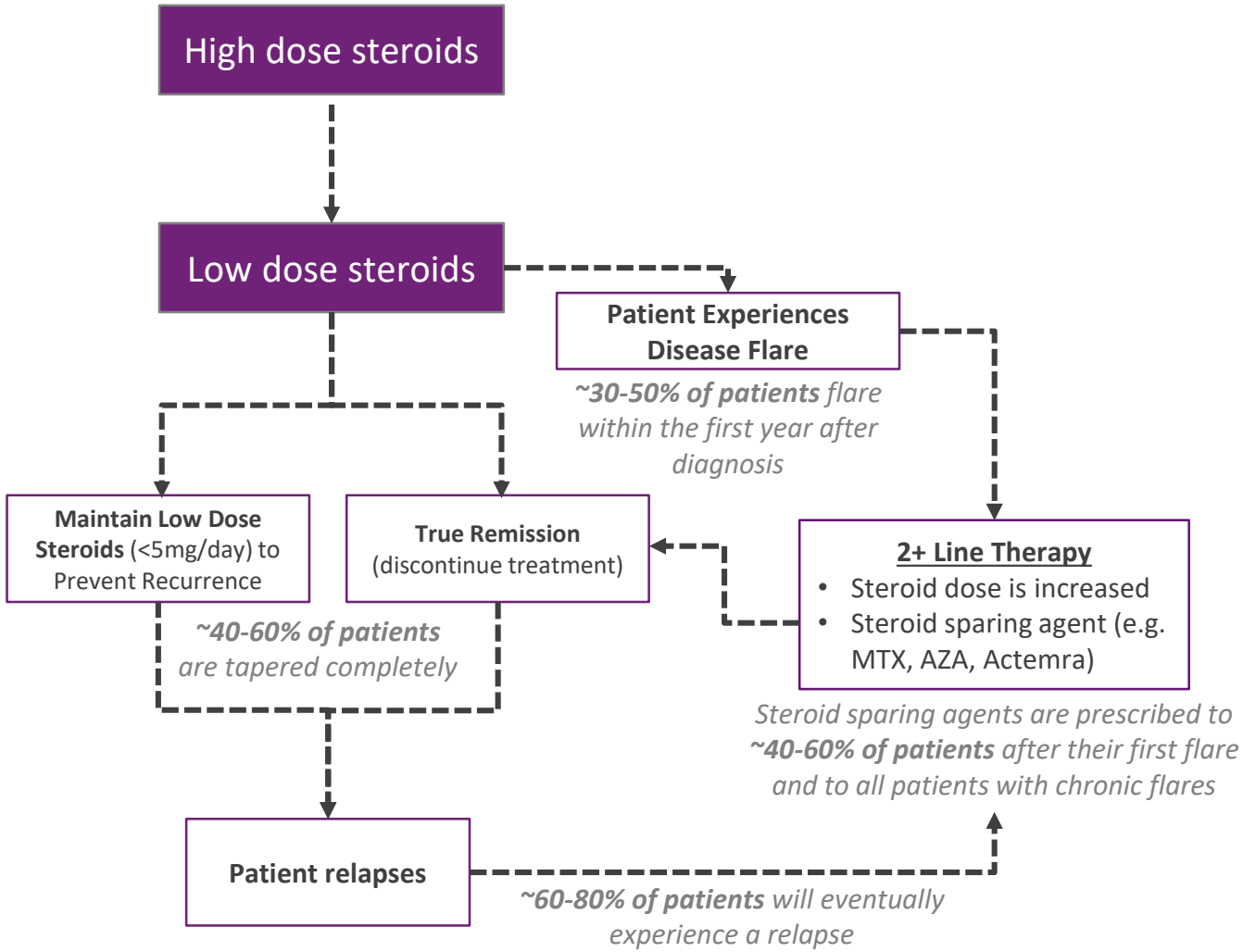
> *"There is an urgency of treatment with these patients, compared to other conditions it's serious."* – Rheumatologist

> *"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 to go blind."* – GCA Patient

Sources: Medcape; Trinity Partners primary market research

Current treatment paradigm for GCA involves high-dose steroids for all patients upon clinical suspicion



All Patient Receive High-Dose Steroids:

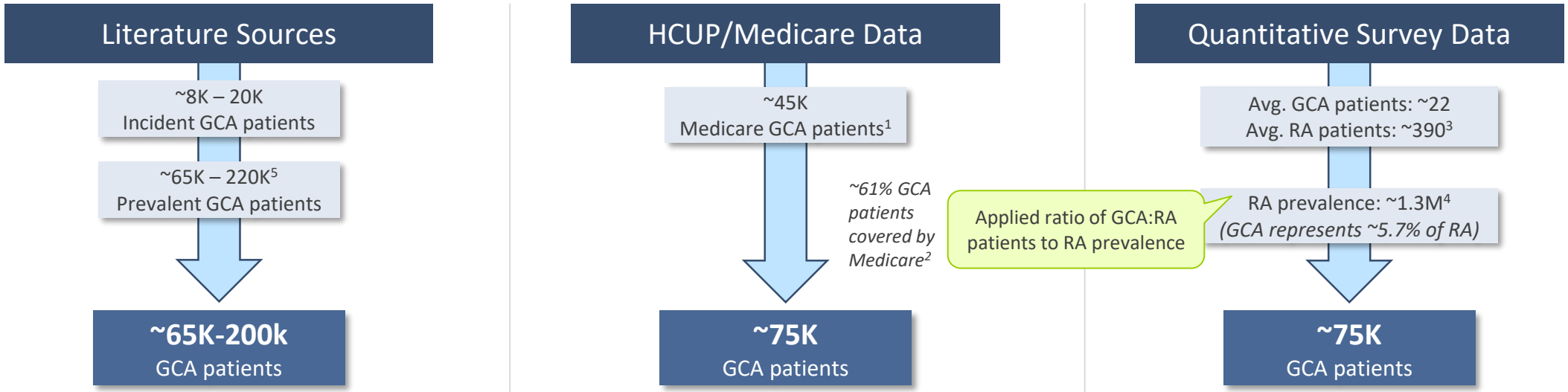
- High-dose steroids are **effective at preventing disease related complications;** however, **they may lead to life altering side-effects** like osteoporosis and diabetes

No Algorithmic Treatment Approach:

- A few treaters initiate **steroid sparing agents** early on 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

Source: Trinity Partners Primary Market Research (n=10 Rheumatologists)

GCA prevalence in the U.S. estimated to be between 75k-150k



Key Considerations to Market Sizing Approach

Wide range

- **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 ~75k-150k (less than that of purely Northern Europeans, but more than estimates from Asian countries)

Under-representation

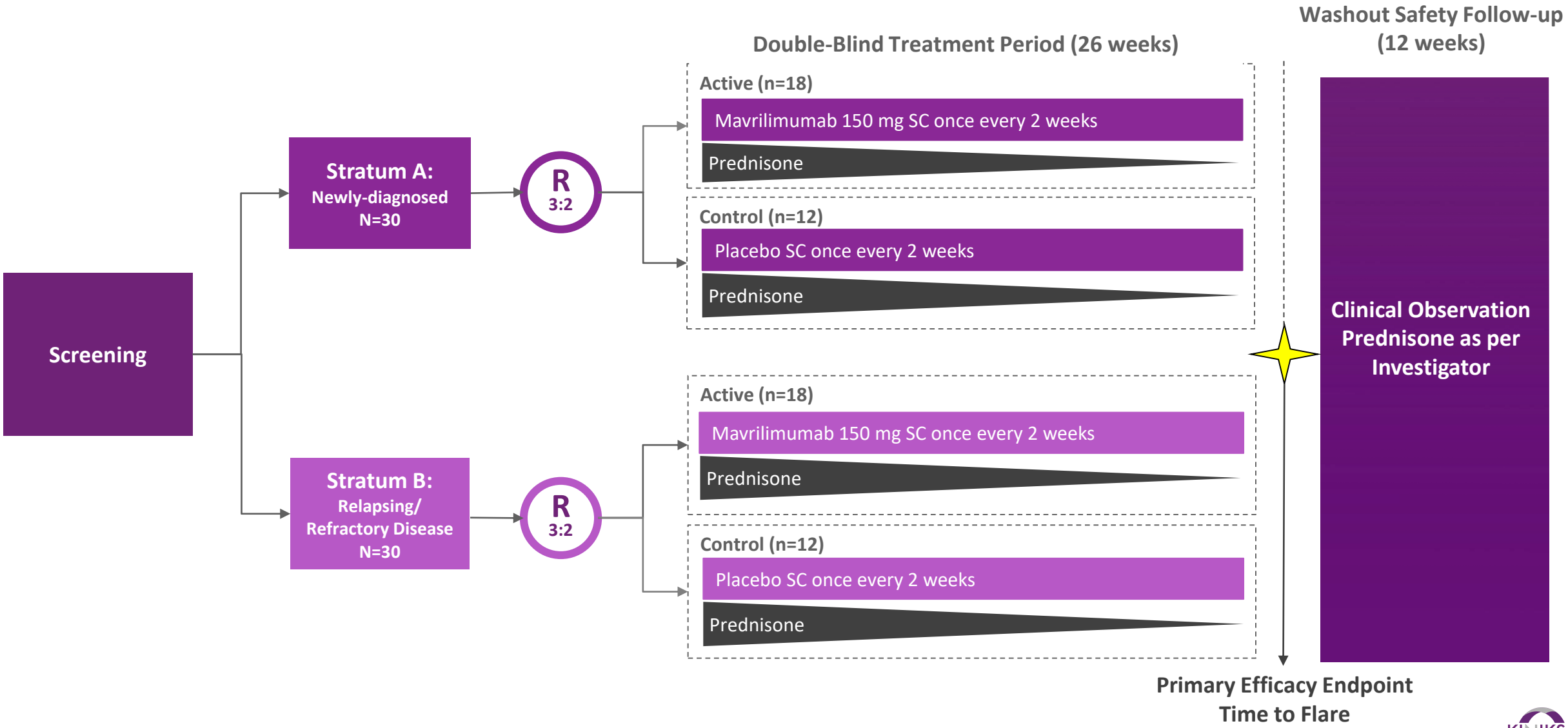
- **Represents Actively Managed Patients:** Medicare analysis does not capture GCA patients who were not actively managed within a given year; thus, the estimate from this analysis will exclude some remission patients or patients likely to relapse

Under-representation

- **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

Sources: 1.) Medicare analysis conducted 1/2018 2.) Trinity Partner's Quantitative Primary Market Research (n=74) 3.) Trinity Partner's Quantitative Primary Market Research (n=196) (includes data from screener portion of survey) 4.)Prevalence of rheumatoid arthritis in the United States adult population in healthcare claims databases, 2004-2014, Hunter et al. 2017, 5.) Crowson et. al, 2017

Randomized, double-blind, placebo-controlled Phase 2 study of mavrilimumab in GCA



GM-CSF is a key growth factor believed to be involved in the pathology of GCA

1	**GM-CSF and GM-CSFRα are overexpressed in GCA lesions**	• Both the receptor[3] and the GM-CSF[4] are expressed in the lesion vs. normal healthy controls
2	**GCA Lesions are heavily comprised of giant cells & non-classical macrophages**	• GM-CSF signaling plays a role in the generation and maturation of giant cells[5] and non-classical macrophages (CD16+)[6] • GM-CSF has been shown to induce endothelial cell migration and proliferation[7] • Inhibition of GM-CSF signaling by mavrilimumab could reduce the number and/or activity of these cells in the vessel wall
3	**Multiple key cytokines driving GCA are downstream of GM-CSF signaling**	• Relevant downstream cytokines in GCA are IL-6, IFNγ and IL-17/23[8] • Inhibiting GM-CSF signaling with mavrilimumab could reduce the relevant pathways involved in both new-onset disease and refractory disease maintenance
4	**Mavrilimumab P2 Trial Underway**	• First-in-class mechanism with the potential to treat both newly diagnosed and refractory patient subsets • Global, P2 proof-of-concept trial ongoing with strata for both patient populations

1) Chandran et al., Scand J Rheumatol, 2015; Trinity Consulting – HCUP/Medicare Data, Quantitative Survey (n=102 rheumatologists); 2) Alba et. al., Medicine 2014
3.) Unpublished Kiniksa Data; 4.) Weyand et al., Ann. Int. Med. 1994; 5.) Yoshihara et al., Immunology 2003; 6.) van Sleen et al., Sci Reports, 2017; 7.) Bussolino et al., Letters to Nature 1989; 8.) Samson et al Autoimm. Rev. 2017

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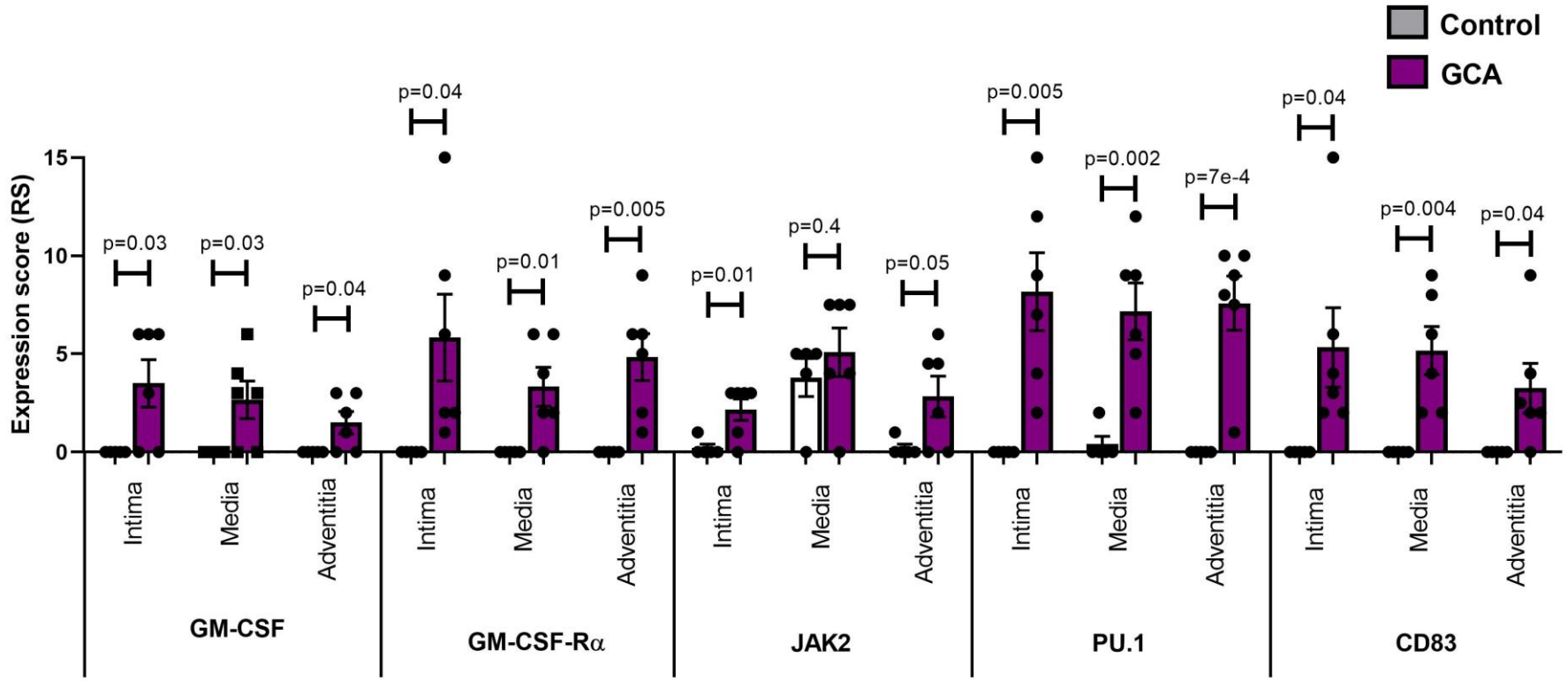
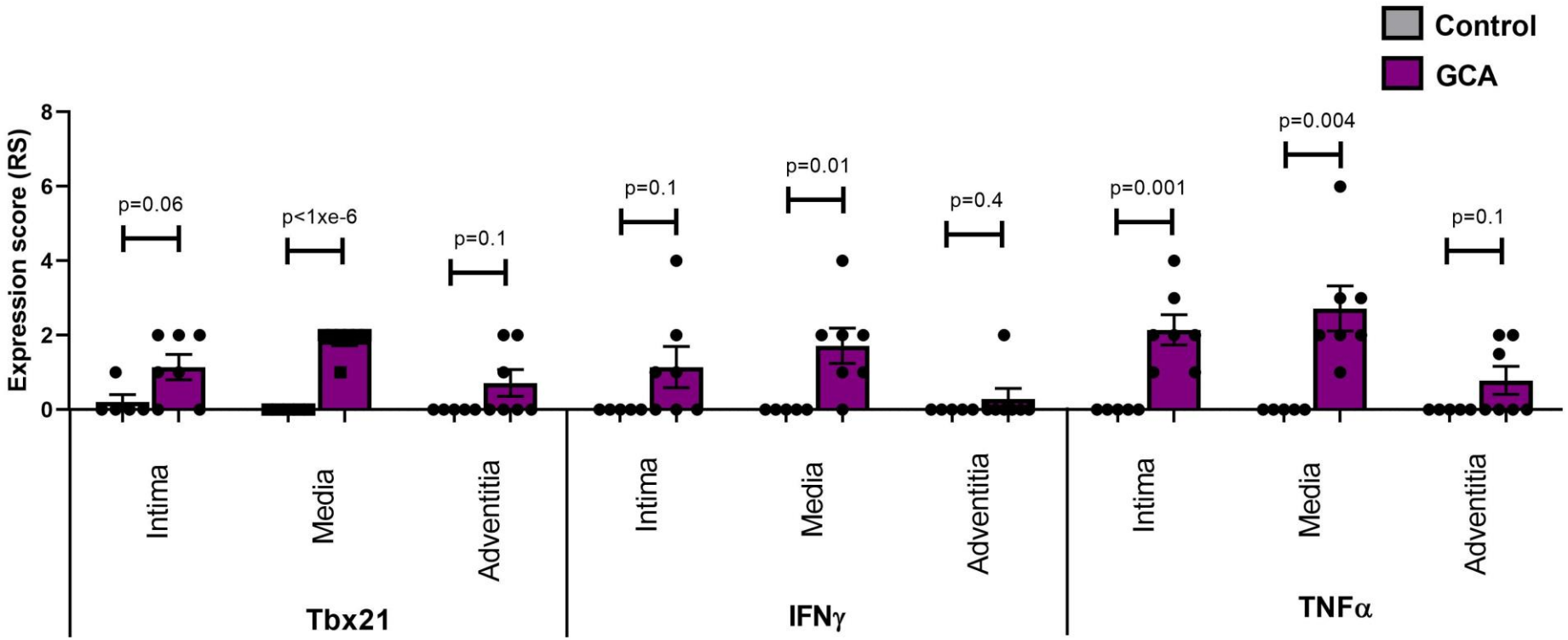
Transcriptomic analysis showed elevated mRNA expression of genes associated with GM-CSF-Rα pathway



Data from RNAscope experiments

Poster Presentation at European Congress of Rheumatology 2019 (EULAR): *GM-CSF Pathway Signature Identified in Temporal Artery Biopsies of Patients With Giant Cell Arteritis*
Maria C. Cid, Rohan Gandhi, Marc Corbera-Bellalta, Nekane Terrades-Garcia, Sujatha Muralidharan, John F. Paolini

mRNA expression of multiple genes associated with T_H1 pathway was elevated in GCA arteries



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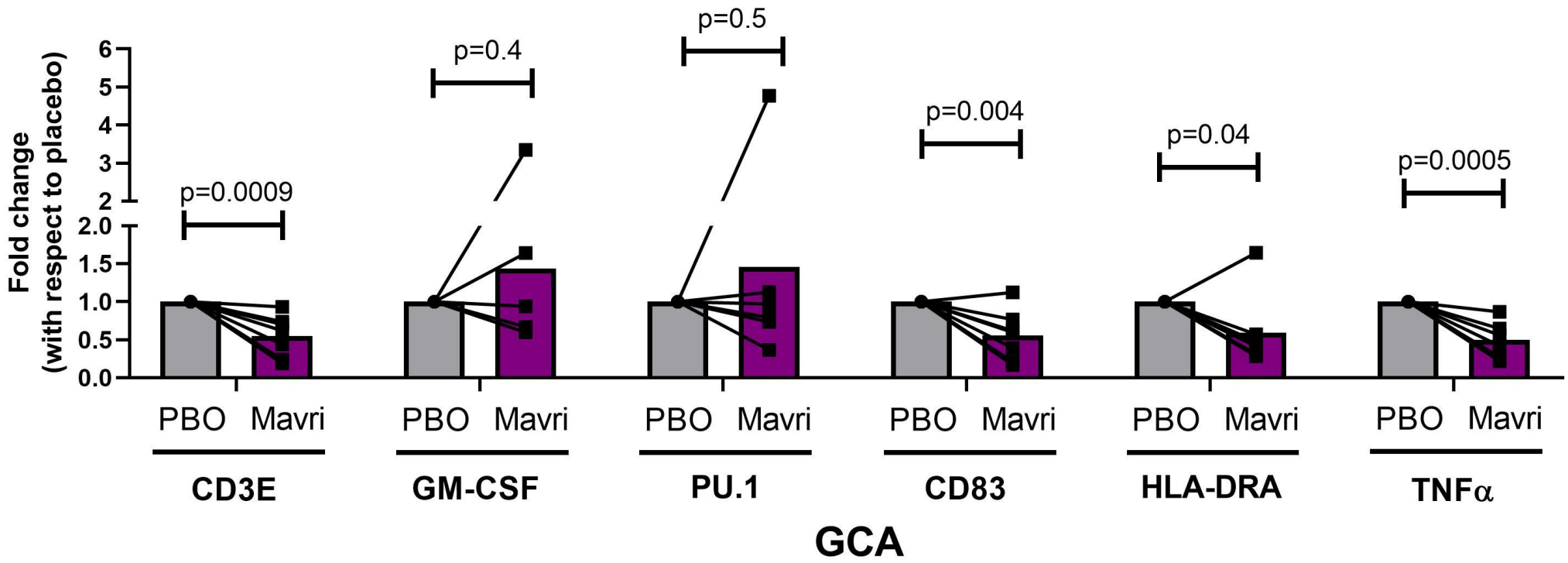
Mavrilimumab shown to suppress the expression of genes associated with immune cell infiltration, inflammation and GM-CSF pathway in cultured GCA arteries



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KPL-716 – Phase 2

(monoclonal antibody inhibitor targeting OSMRβ)

Rilonacept | Mavrilimumab | **KPL-716** | KPL-404 | KPL-045

Differentiated molecule with potential to treat variety of pruritic, inflammatory and fibrotic indications

Mechanism of Action[1]	Monoclonal antibody inhibitor targeting OSMRβ; a key receptor subunit shared by IL-31 and Oncostatin M
Lead Indication	Prurigo nodularis
Addressable Population[2]	~300k prevalent in the U.S.
Competition	No FDA-approved therapies for prurigo nodularis
Clinical Development	Enrolling a Phase 2a clinical trial in prurigo nodularis and an exploratory Phase 2 study in diseases characterized by chronic pruritus
Rights	Worldwide

1) Trinity Qualitative Interviews; 2) 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., British Journal of Dermatology, 2001

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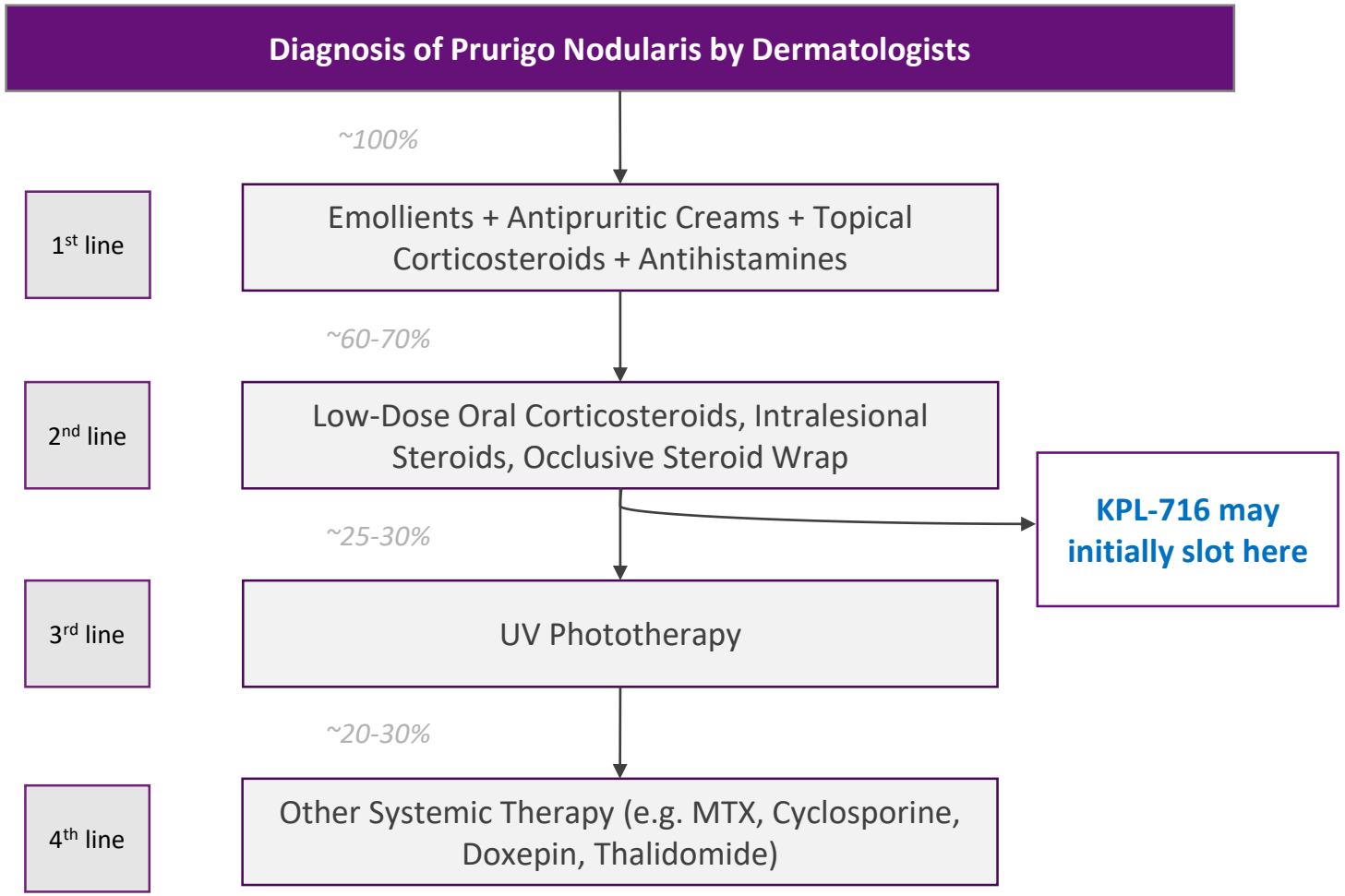
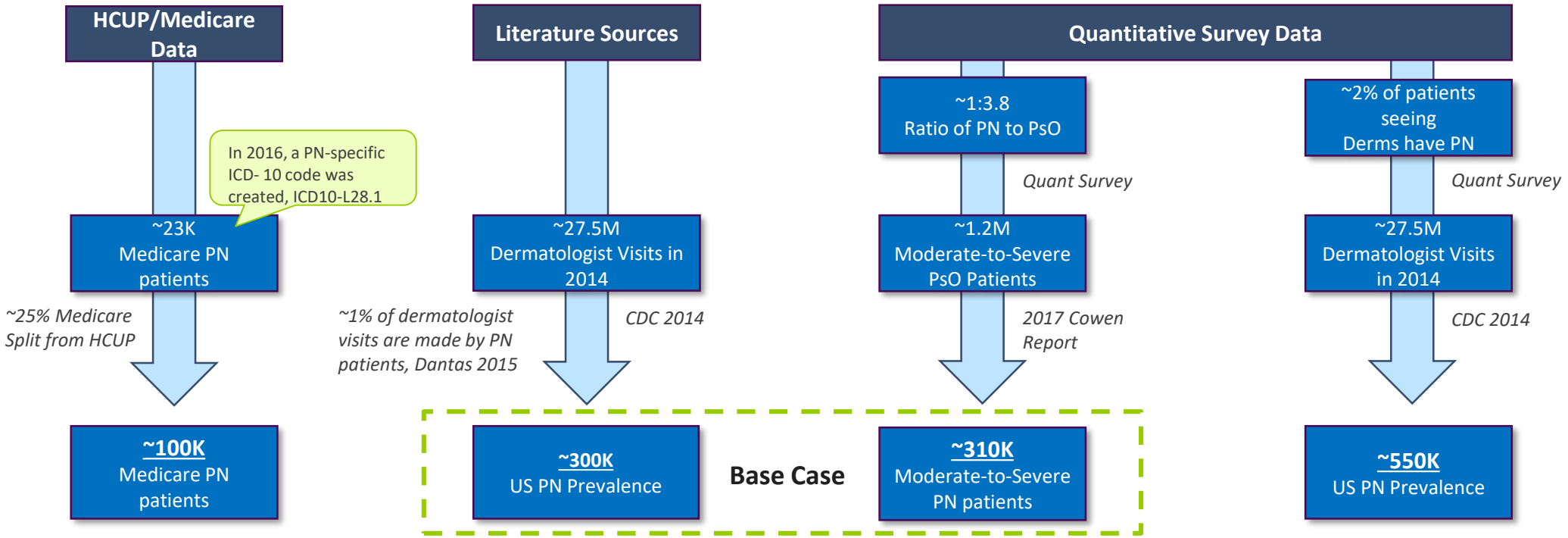
Prurigo nodularis is typically treated by dermatologists through a combination of medications and behavioral therapies; treatment is usually unsuccessful



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

Sources: 1. Medscape, 2. Trinity Qualitative Research

The prevalence of prurigo nodularis is estimated at ~300K in the U.S.



Sources: CDC 2014: National Ambulatory Medical Care Survey: 2014 State and National Summary Tables <https://www.cdc.gov/nchs/data/ahcd/namcs_summary/2014_namcs_web_tables.pdf>; Cowen and Company, Therapeutic Categories Outlook: Comprehensive Study September 2017; Primary Market Research; 3. Dantas, 2015, "Prevalence of dermatoses in dermatologic evaluation requests from patients admitted to a tertiary hospital for 10 years"

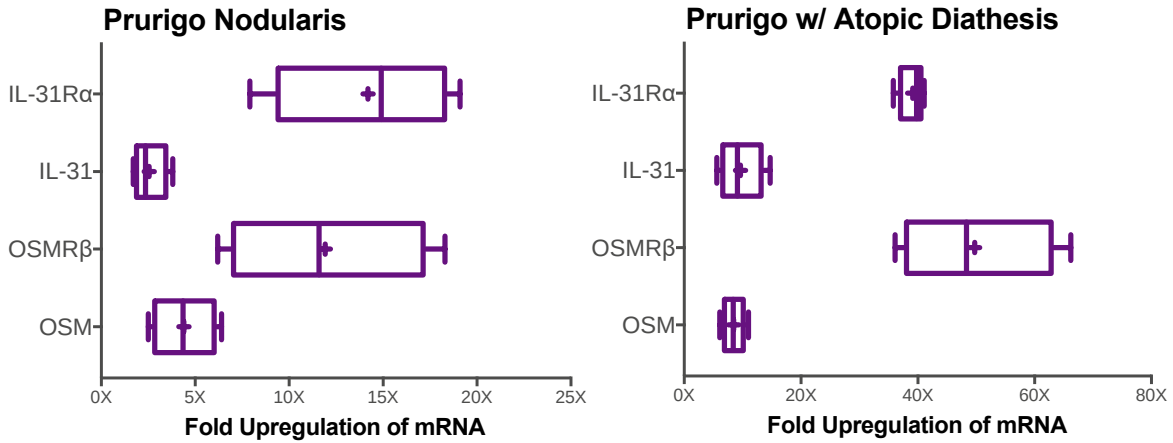
IL-31 and OSM are implicated in the pathology of prurigo nodularis

Quantitative Real-time PCR Analysis of IL-31 mRNA in Human Skin

- **IL-31 significantly overexpressed in pruritic skin vs. non-pruritic skin**
  - Highest levels of IL-31 were detected in PN, one of the most pruritic forms of chronic skin inflammation
- **In PN lesions there is a 50-fold upregulation of IL-31 mRNA vs. normal skin and a 4.5-fold upregulation vs. lesional atopic dermatitis**
  - While there was some variability in IL-31 mRNA levels seen among PN patients, levels in all patients were significantly elevated compared with healthy controls

Source: Sonkoly et al., J Allergy Clin Immunol, 2006; Internal Kiniksa Data (Generated by the Loser Lab)

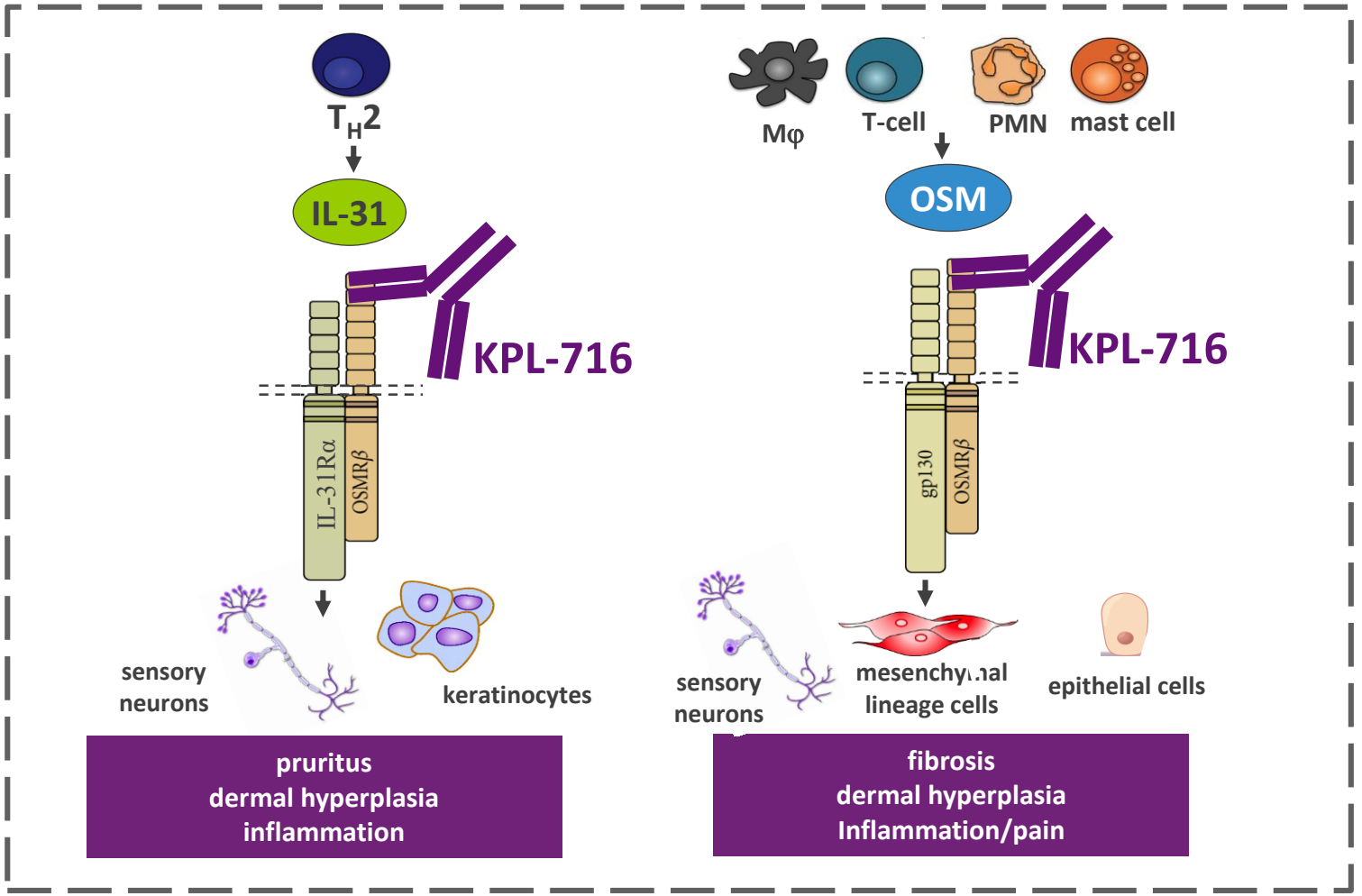
Dual-targeting of OSM and IL-31 though OSMRβ blockade has the potential to be disease modifying

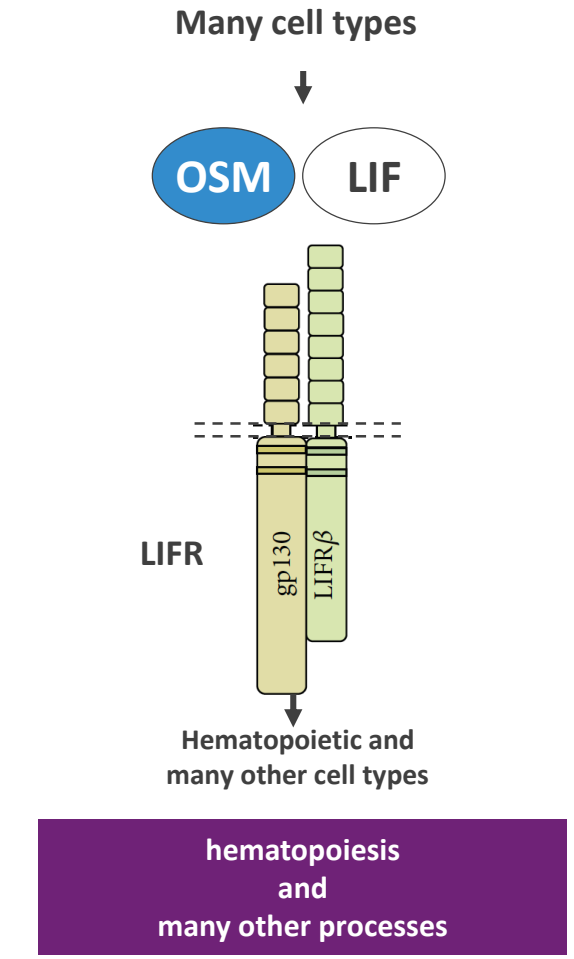


- Messenger RNA levels of IL-31, OSM and their receptor subunits (IL-31Rα and OSMRβ) are significantly elevated in lesions of prurigo nodularis, implicating them as major drivers of pruritus, leading to disease pathophysiology
- This phenotype is even more evident in the case of patients with prurigo nodularis that have an atopic diathesis since their receptor subunits are even more highly up-regulated than in prurigo nodularis alone
- These data provide strong mechanistic rationale to target both IL-31 and OSM by blocking OSMRβ

Source: Internal Kiniksa Data (Generated by the Loser Lab)

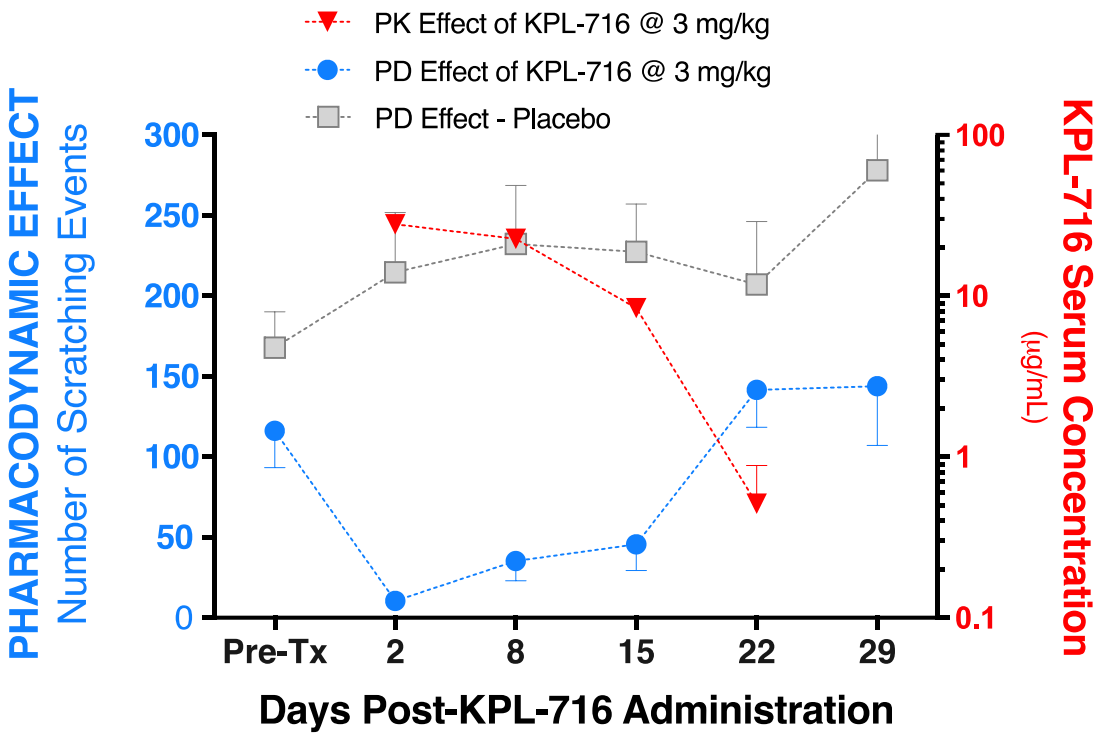
KPL-716 inhibited IL-31 & OSM signaling through OSMRβ but avoided inhibiting signaling critical to hematopoiesis through OSM/LIFR in *in vitro* studies





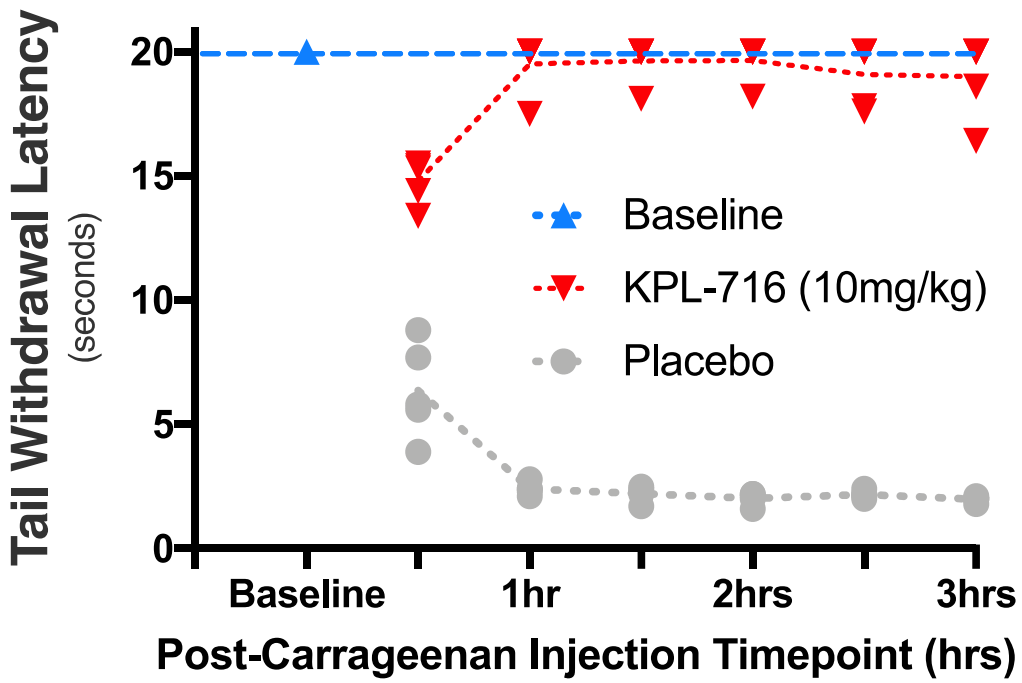
KPL-716 showed signs of potential efficacy in two validated non-human primate models of pruritus and inflammation after a single dose

NHP Model of Pruritus*



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

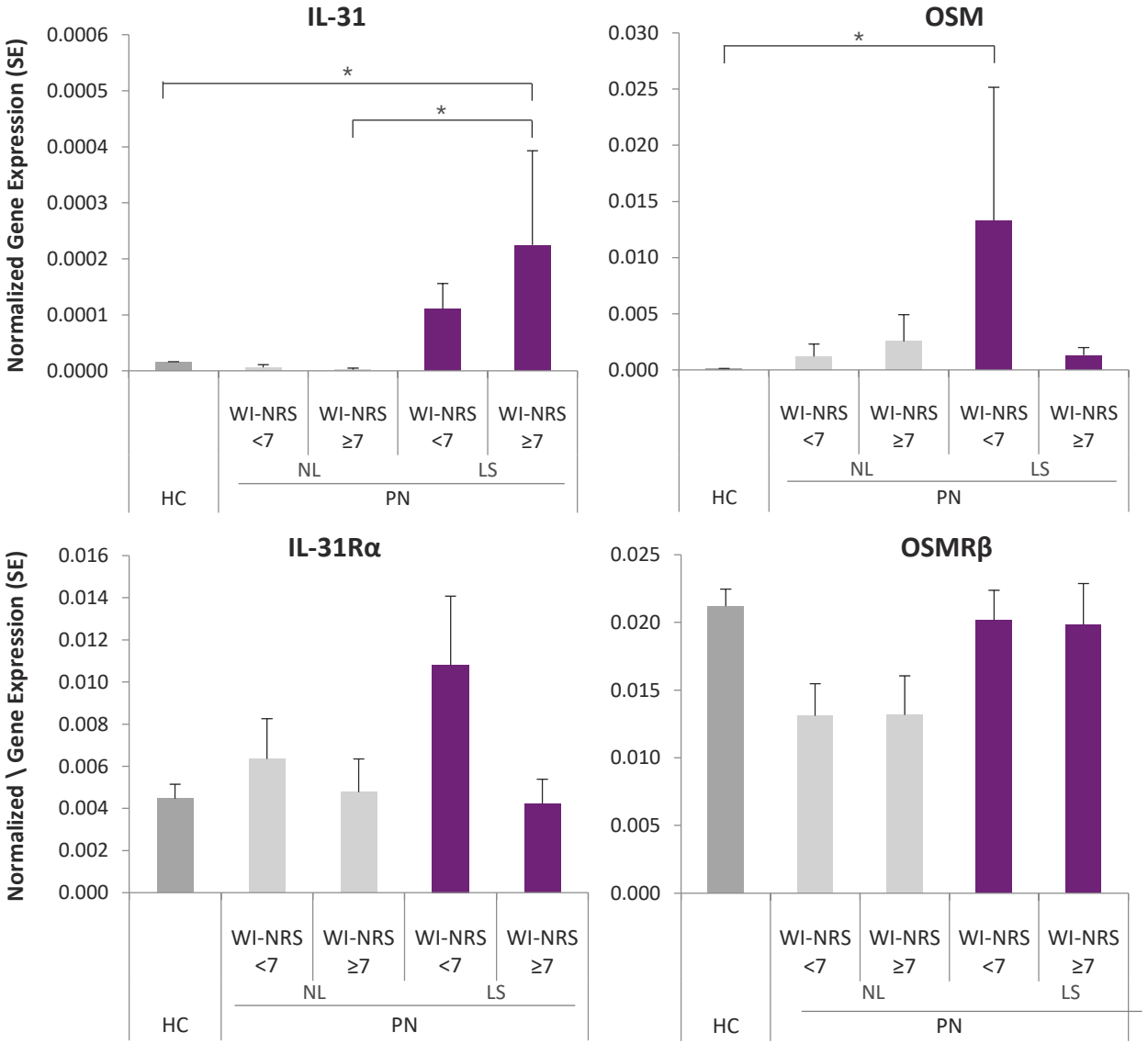
NHP Model of Inflammation*



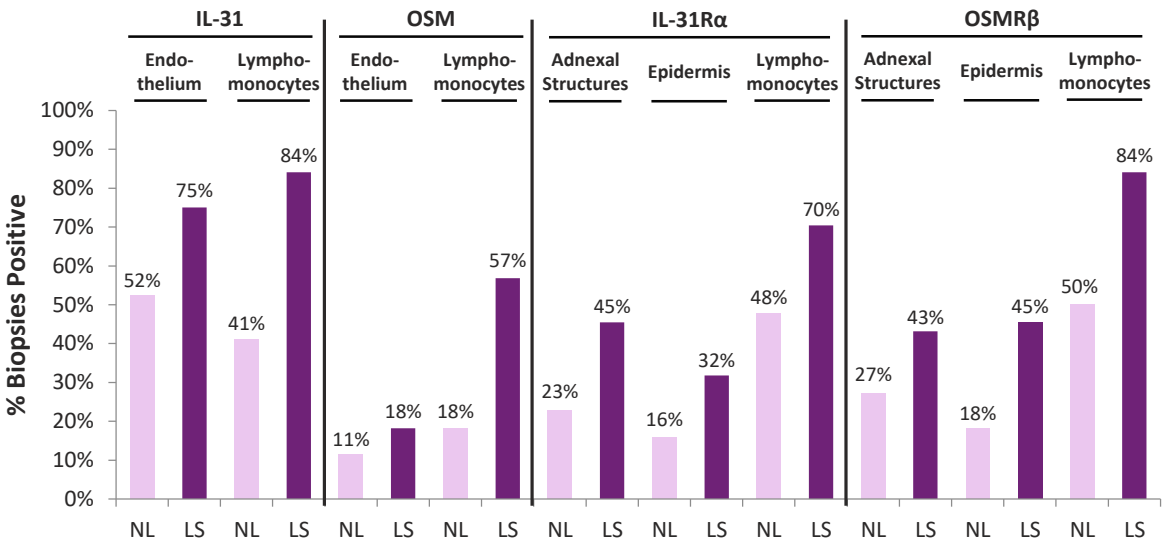
A single dose of KPL-716 at 10mg/kg increased tail withdrawal latency; implicates OSMRβ in the inflammatory response

All components of the Type II OSMRβ signaling complex show upregulation in lesional skin of PN patents; 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 OSMRβ 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, OSMRβ, IL-31, and IL-31Rα mRNA expression was higher in lesional (LS) PN biopsies compared with non-lesional (NL) biopsies; all components except for OSMRβ, which is known to be constitutively expressed, showed elevation compared to healthy controls (HC)
  - LS samples from PN patients with WI-NRS≥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 OSMRβ signaling proteins shows upregulation in LS vs NL biopsies of PN patients' skin

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

HC, healthy volunteers; IL-31Rα, 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"). *$P<0.05$

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

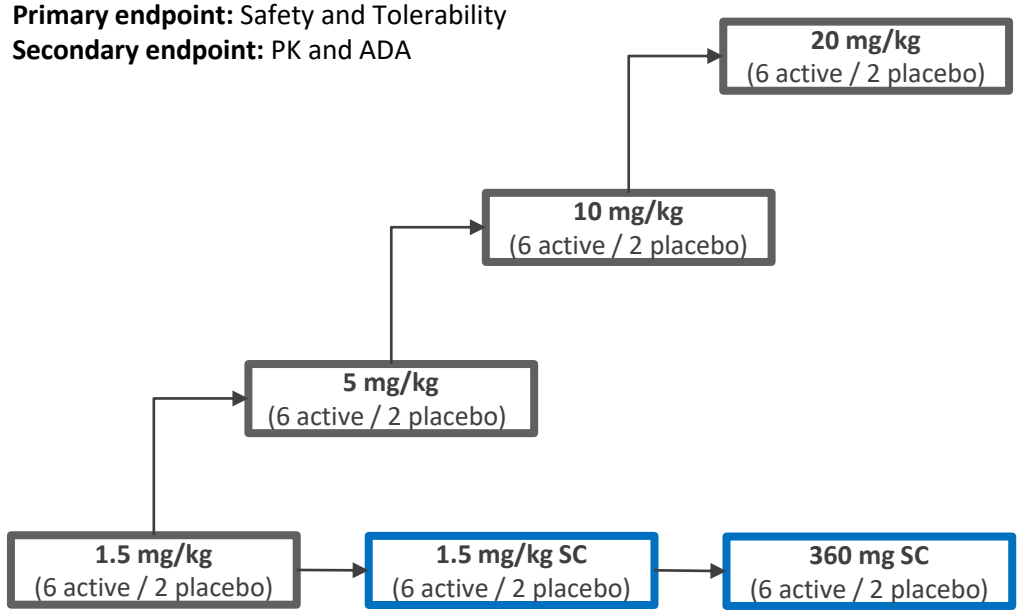
Phase 1a: Normal Healthy Volunteer (n=50)

Dose Groups

Single IV Dose

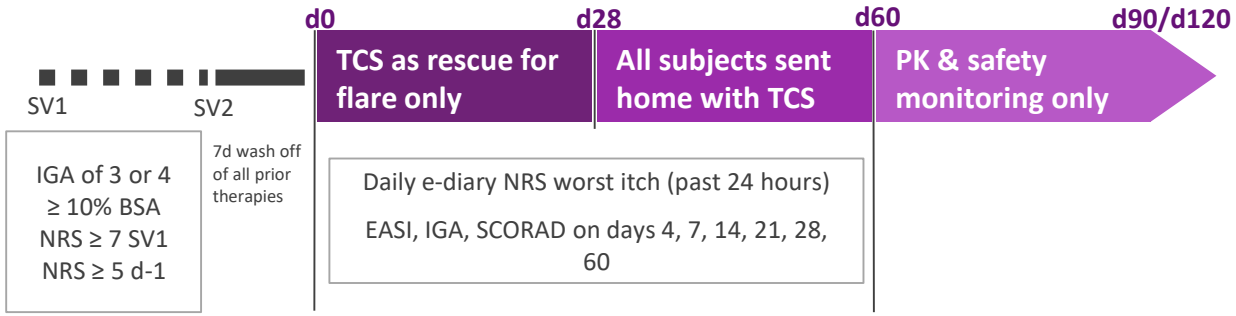
Single SC Dose

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



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

Patient Experience

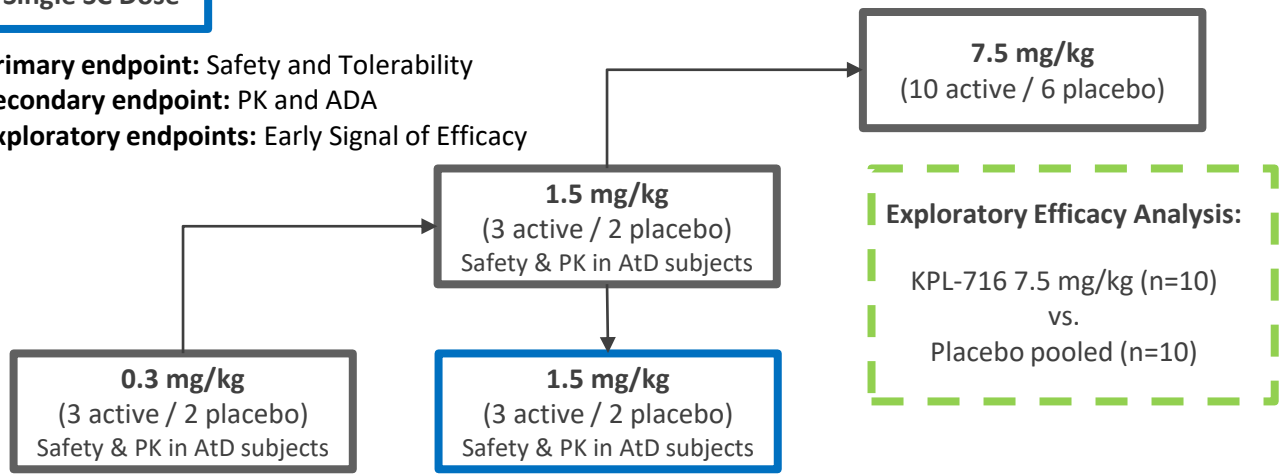


Dose Groups

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)

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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 sequalae

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.

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)**

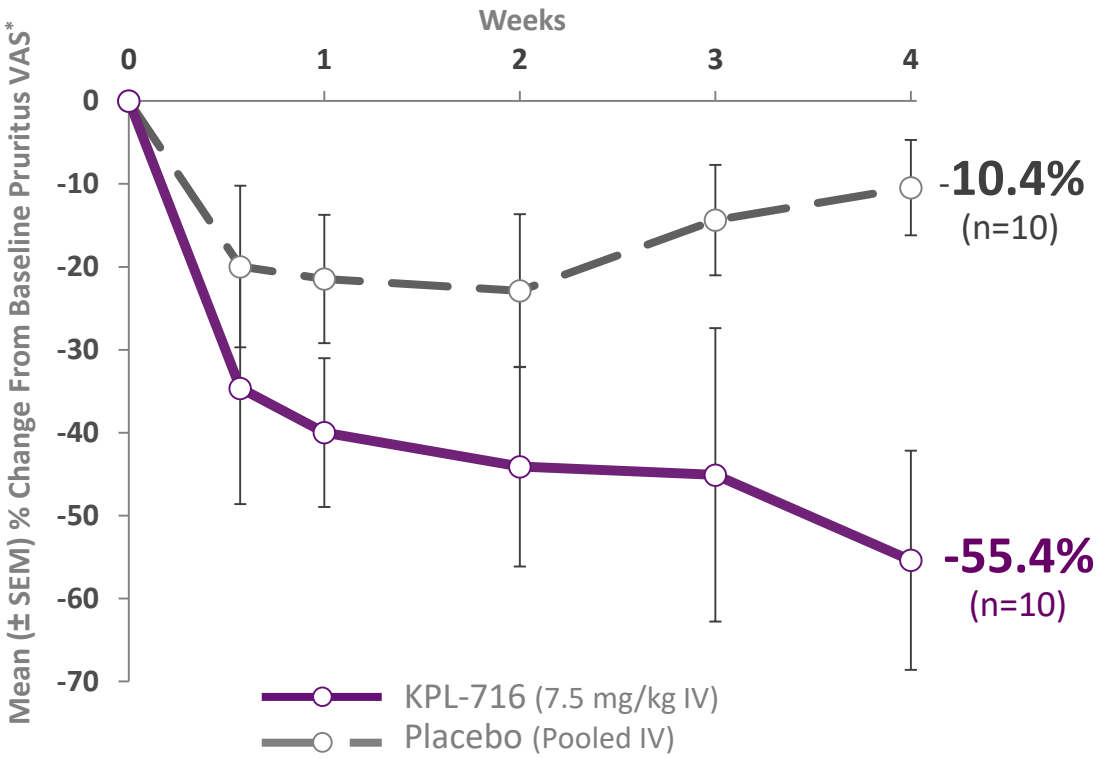
Post Hoc Efficacy Analysis

- **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**

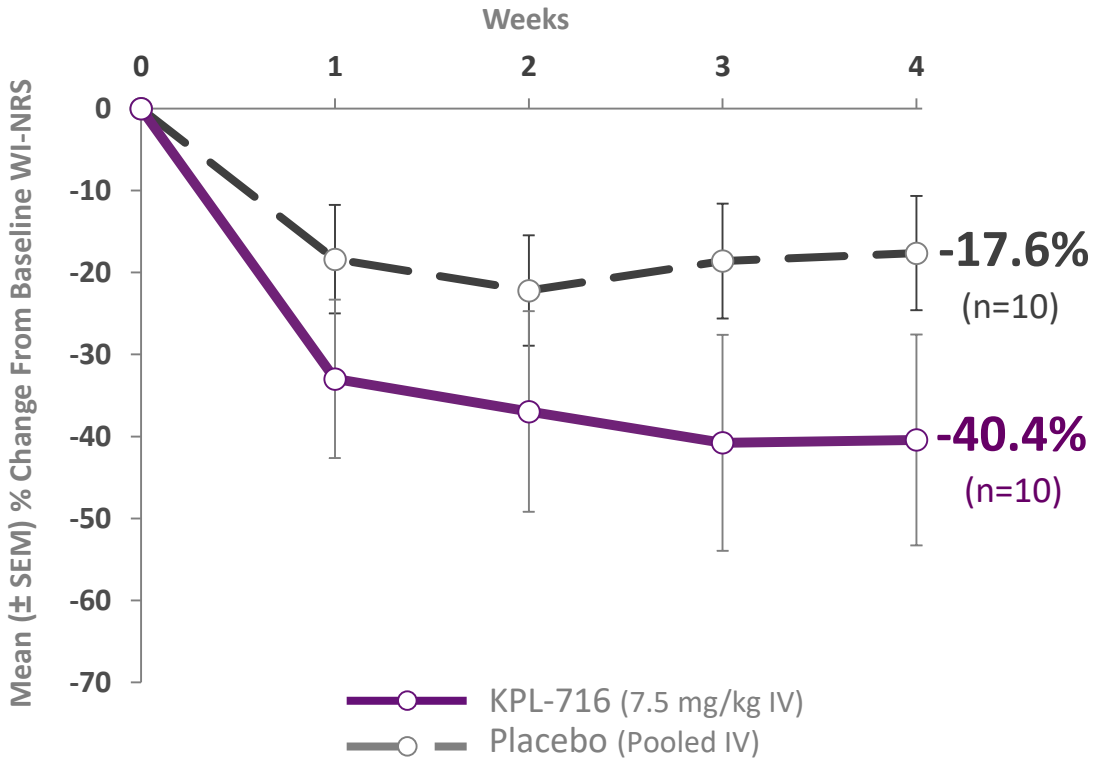
AD = atopic dermatitis, IV = intravenous, WI-NRS = Worst Itch Numerical Rating Scale, EASI = Eczema Area and Severity Index, SCORAD = Scoring atopic dermatitis (severity scale)

Single doses in Phase 1a/1b provided early evidence indicative of target engagement and showed reduction in pruritus over the 28-day monotherapy period

Pruritus Visual Analog Scale (VAS)*



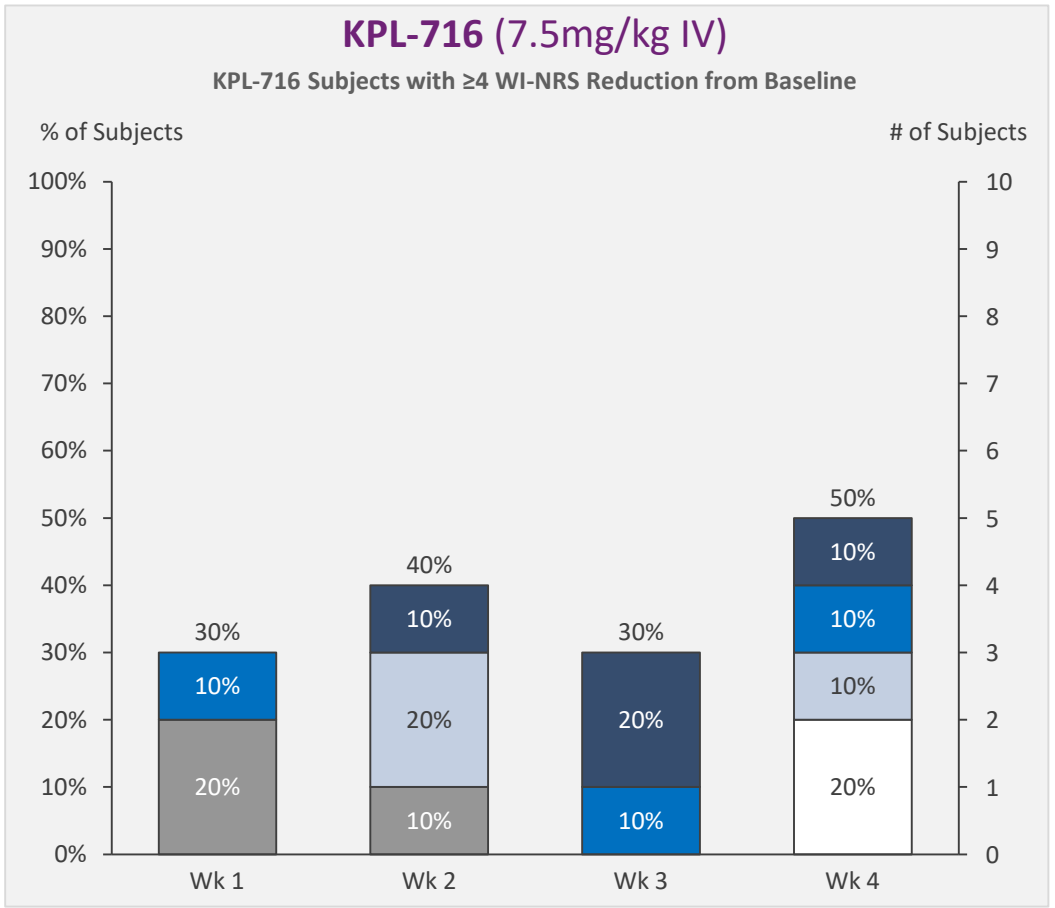
Weekly Average Worst Itch Numerical Rating Scale (WI-NRS)

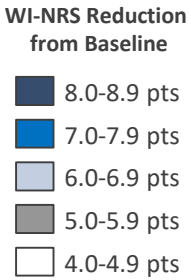


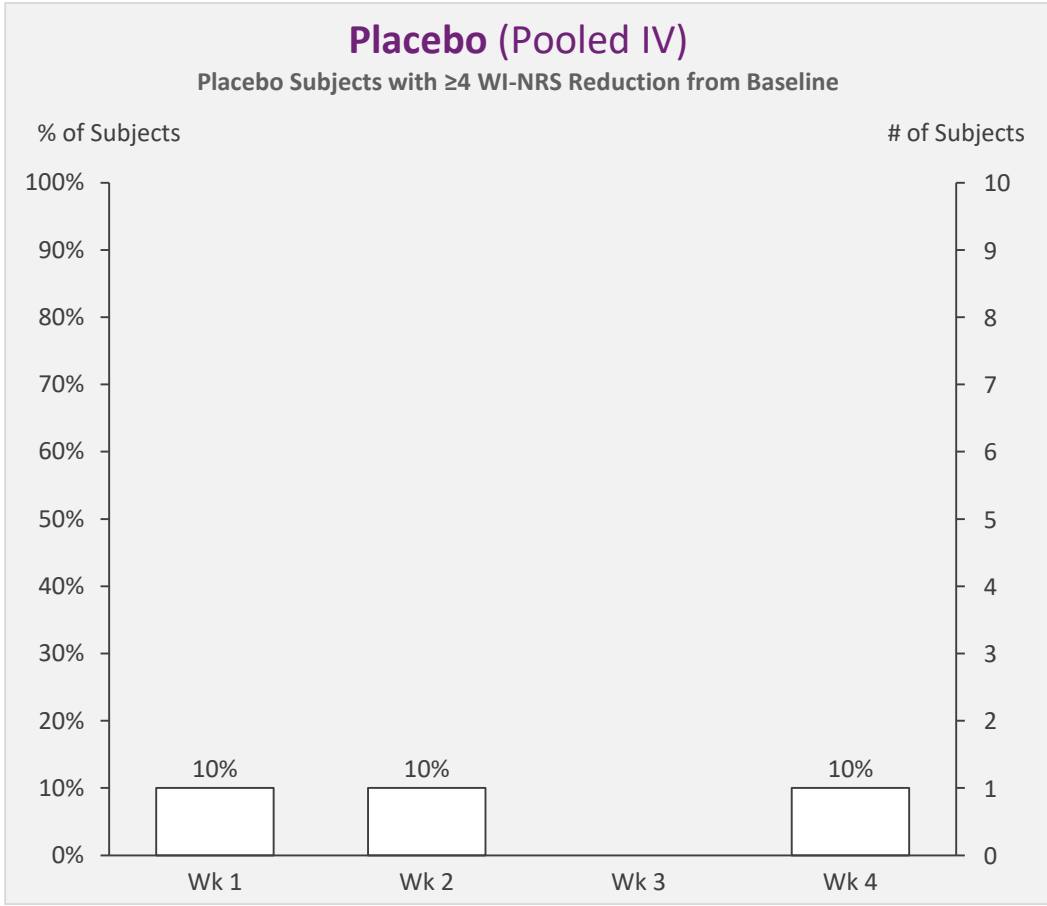
In subjects who received rescue medication, last observation was carried forward. Two KPL-716 recipients (d15, d21) and three placebo recipients (d3, d14, d26)

* VAS = Visual Analog Scale and a component of SCORAD (Scoring atopic dermatitis) severity scale

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





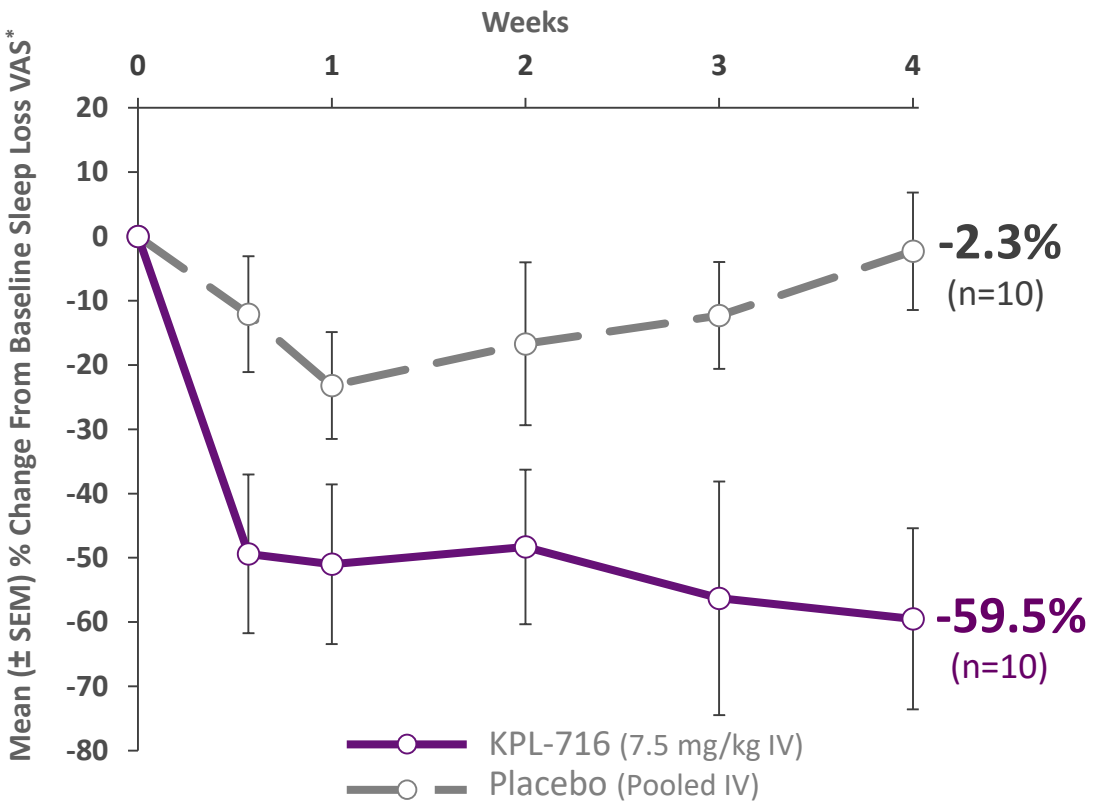


Subjects were treated as non-responders after rescue. Two KPL-716 recipients (d15, d 21) and three placebo recipients (d3, d14, d26).

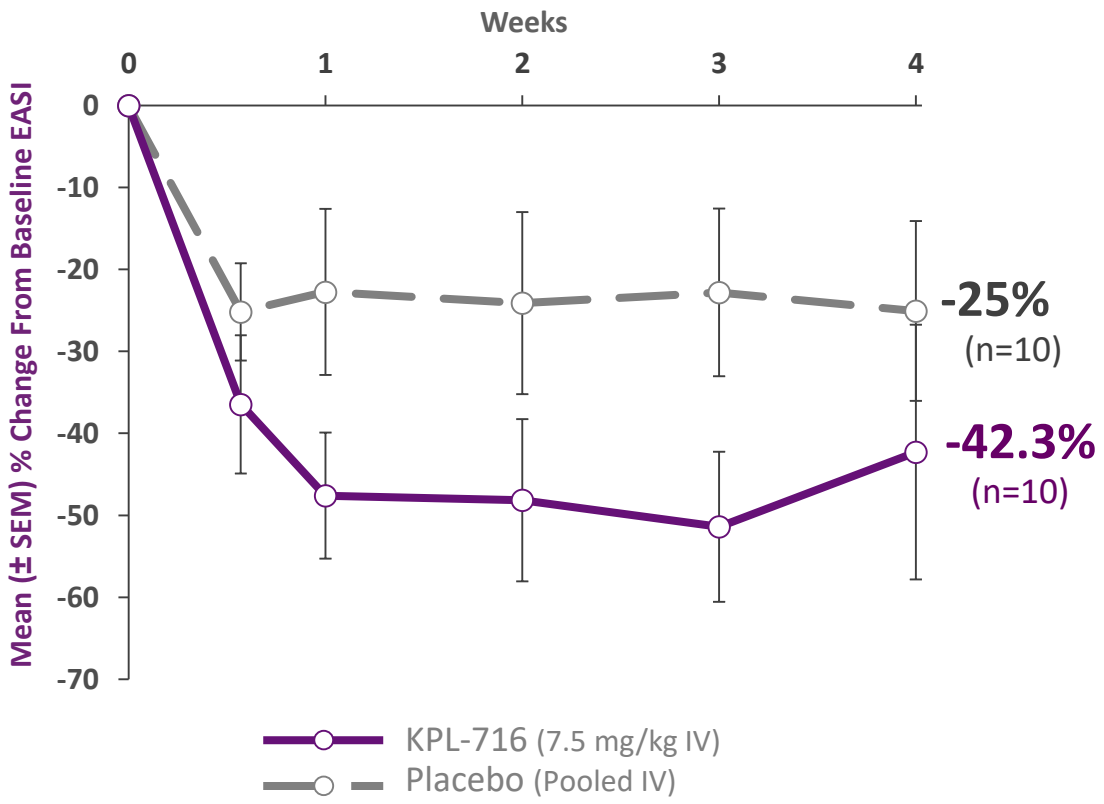
WI-NRS = Worst Itch Numerical Rating Scale, TCS = topical corticosteroids

Single doses in Phase 1a/1b showed reduction in sleep loss and disease severity over the 28-day monotherapy period

Sleep Loss VAS*



Eczema Area and Severity Index (EASI)

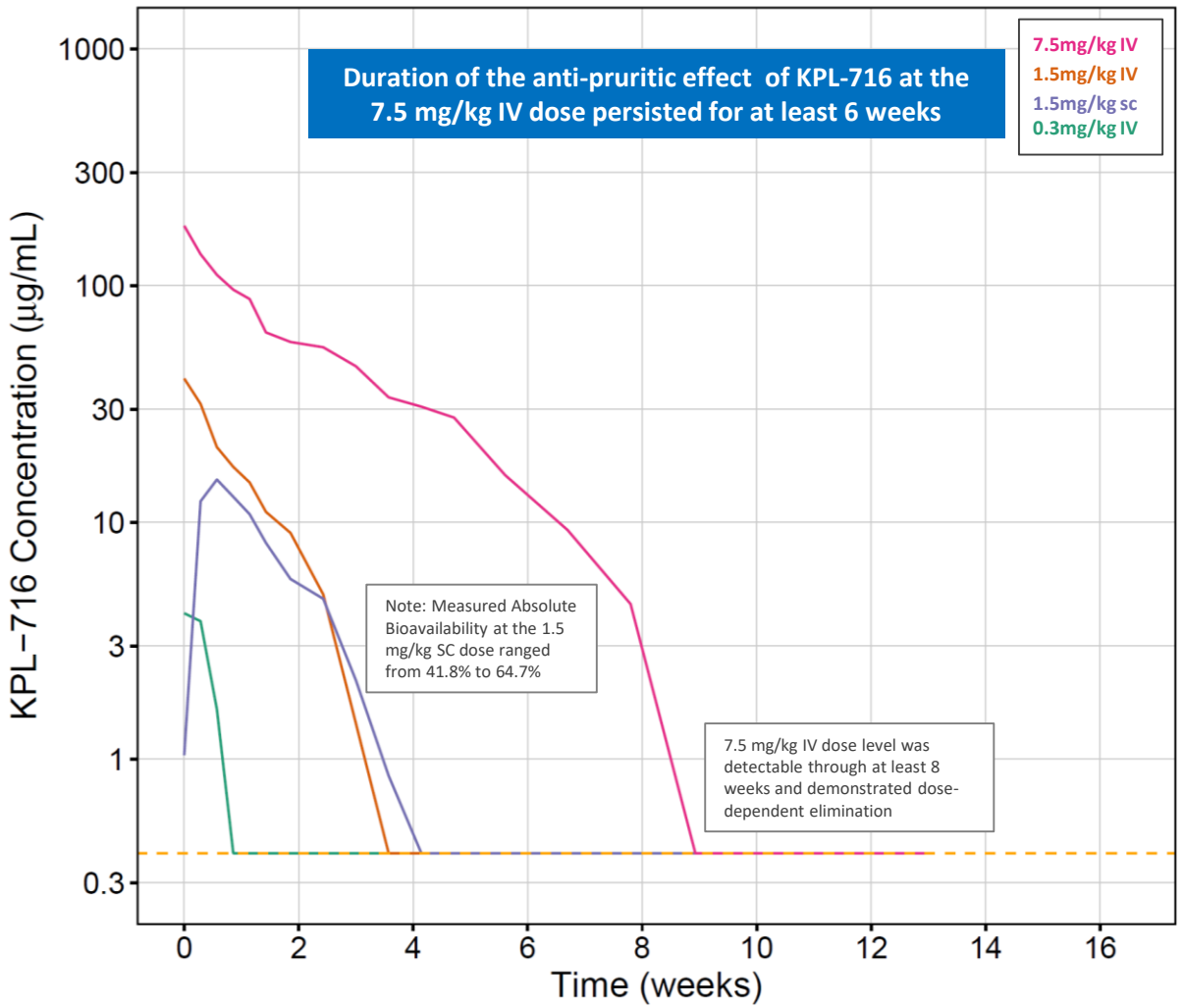


In subjects who received rescue medication, last observation was carried forward. Two KPL-716 recipients (d15, d21) and three placebo recipients (d3, d14, d26)

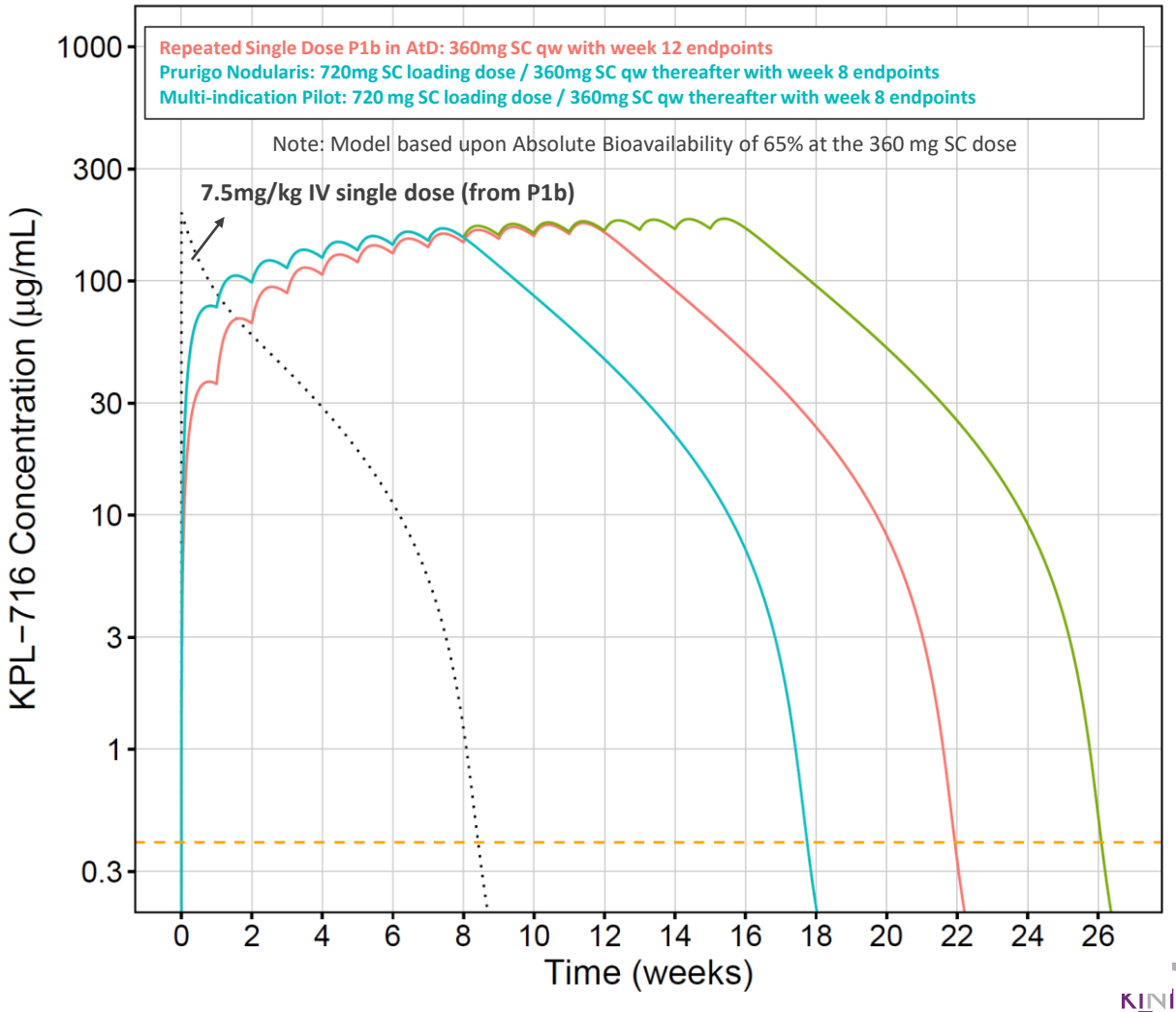
* VAS = Visual Analog Scale and a component of SCORAD (Scoring atopic dermatitis) severity scale

PK/PD model predicts that weekly SC dosing provides sufficient/high exposures for current POC studies as well as studying alternate dosing regimens in future dose-finding studies (e.g., q2w and/or qm)

Measured KPL-716 PK From P1b Single Dose



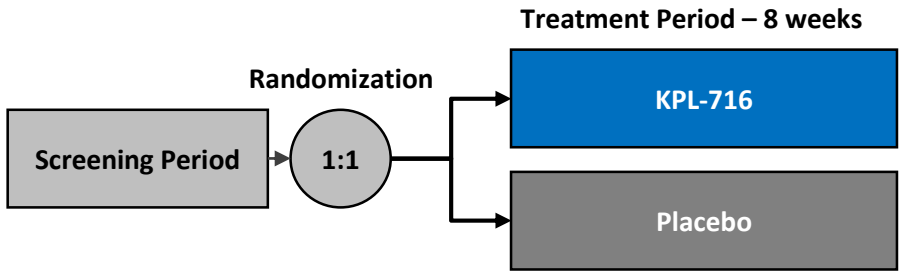
Phase 1b data used to build predictive PK/dosing model for currently ongoing multiple-dose studies (RSD, PN, Chronic pruritus pilot)



KPL-716 Phase 2a/2b trial in prurigo nodularis

Ph2a Proof-of-Concept (POC) Segment

- **Objective:** Assess pruritus reduction
- **Sample size:** n=100
- **Dose:** 720 mg SC loading dose --> 360 mg single SC QW thereafter



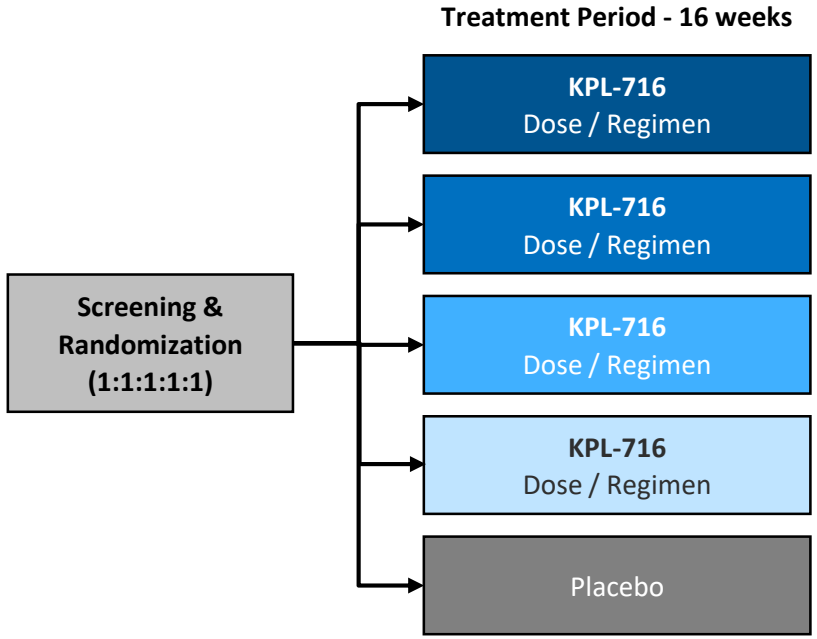
Primary Endpoint:
- % change from baseline in weekly average Worst Itch-Numeric Rating Scale (WI-NRS)

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

Ph2b Dose Range-Finding Segment:

- **Objective:** Define optimal KPL-716 dose/regimen on pruritus endpoint
- **Sample size:** n=300 (anticipated)
- **Doses/Interval:** TBD



Primary Endpoint:
- Likely identical to Ph2a, but will be adjusted if needed based on Ph2a data

Secondary Endpoints:
- Will be determined based on observations from Ph2a

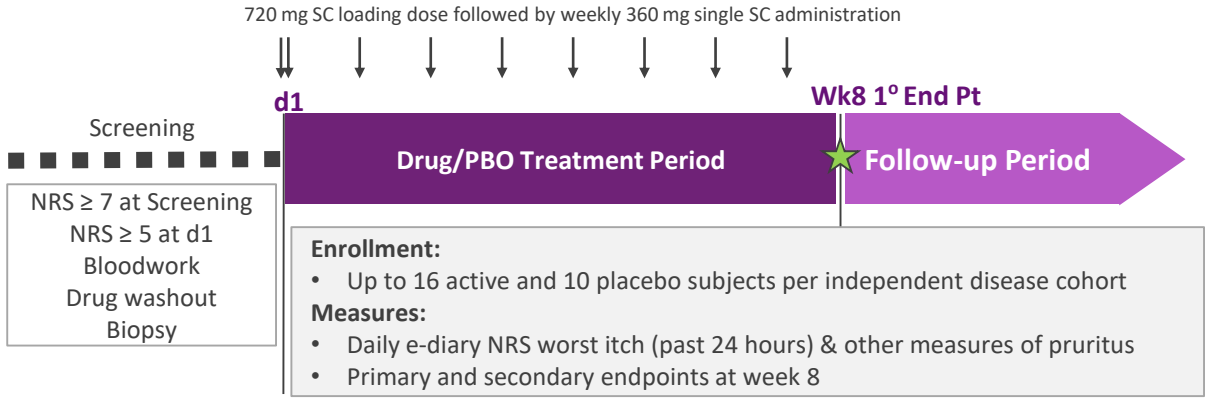
KPL-716 exploratory Phase 2 study in diseases characterized by chronic pruritus

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

Chronic Idiopathic Urticaria (CIU)	• **US Prevalence:** ~2-3 M[1,2] • **Pruritus Burden:** ~1-in-3 experience pruritus refractory to conventional therapies; ~15-20% treated with Xolair continue to experience pruritus[3]
Chronic Idiopathic Pruritus (CIP)	• **US Prevalence:** Treating physicians report ~1 CIP patient for every 3 atopic dermatitis patients[3,4,] • **Pruritus Burden:** ~50% experience symptoms lasting for >1-yr; ~1-in-3 treated patients experience refractory pruritus[3]
Lichen Planus (LP)	• **US Prevalence:** ~0.5 M+[5] • **Pruritus Burden:** ~1-in-3 treated patients experience refractory pruritus[3]
Lichen Simplex Chronicus (LSC)	• **US Prevalence:** Treating physicians report ~1 LSC patient for every PN patient[3] (~0.3 M addressable in the US)[6,7] • **Pruritus Burden:** ~40% of treated patients experience refractory pruritus[3]
Plaque Psoriasis	• **US Prevalence:** ~12 M[8,9] • **Pruritus Burden:** ~2-3 M patients in US with moderate-to-severe pruritus[9]

Subject Experience in Each Disease Cohort



Note: US prevalence figures are estimates based on references which may include only a single EU country and/or based on primary market research where physicians were asked to relate the estimated number of patients they treat with the target disease in relation to another disease they treat where the prevalence estimates are more well known

1) Gaig et al., Epidemiology of urticaria in Spain, J Investig Allergol Clin Immunol. 2004 | 2) Saini, Chronic Spontaneous Urticaria, Immunology & Allergy Clinics, 2014 | 3) Kiniksa survey data (n=83 dermatologists, n=38 allergists) | 4) Weisshaar et al., European Guideline on Chronic Pruritus; Acta Derm Venereol 2012 | 5) Cleach & Chosidow, Lichen Planus, NEJM 2012 | 6) Dantas, 2015, Prevalence of dermatoses in dermatologic evaluation requests from patients admitted to a tertiary hospital for 10 years, An Bras Dermatol. 2015 | 7) HCUP/Medicare Data 2012/2013 | 8) Michalek et al., A systematic review of worldwide epidemiology of psoriasis, J Eur Acad Dermatol Venereol. 2017 | 9) Menlo Tx Company Presentation June 2018

Interim KPL-716 repeated-single-dose Phase 1b summary

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

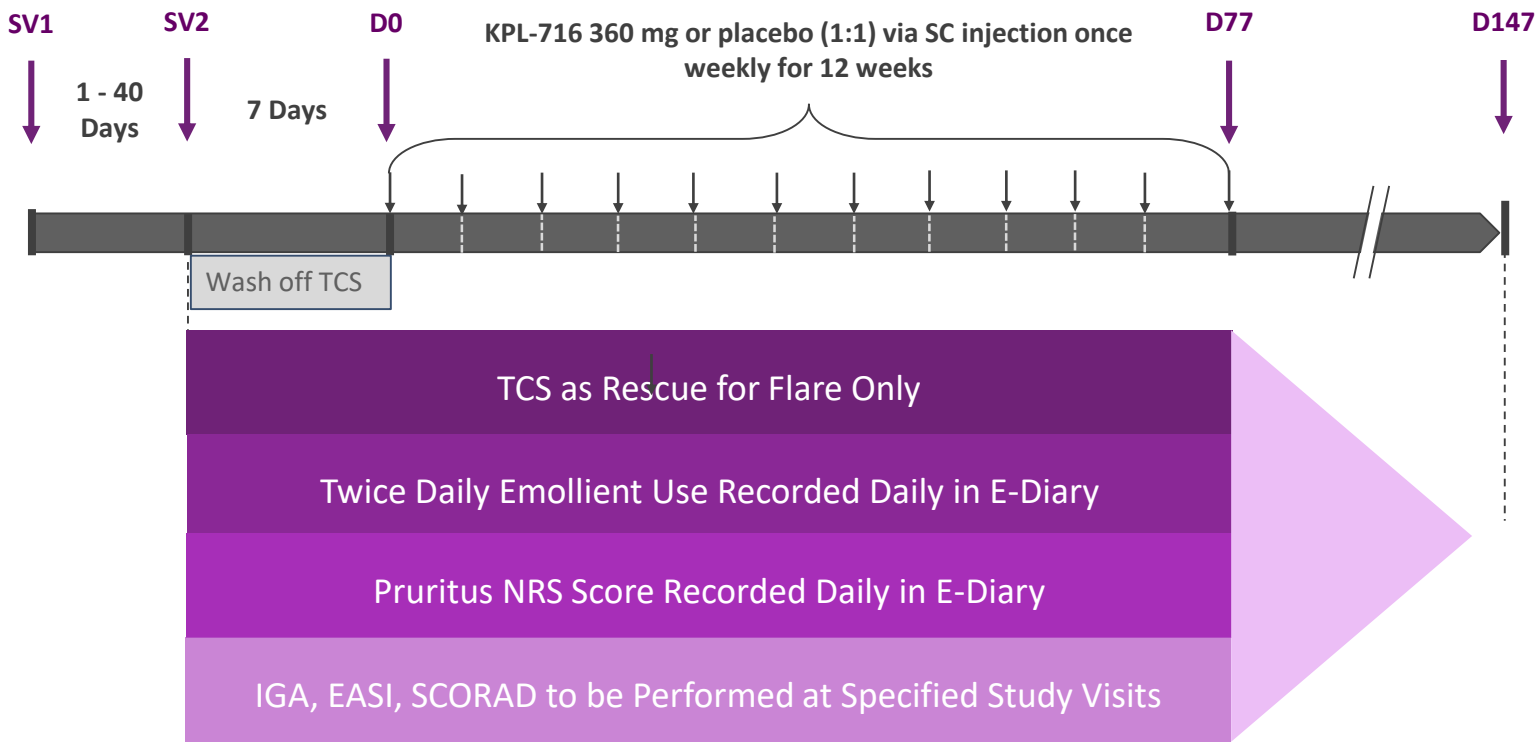
Topline Observations:

- 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

KINIKSA

KPL-716 placebo-controlled repeated-single-dose Phase 1b study design in patients with moderate-to-severe atopic dermatitis

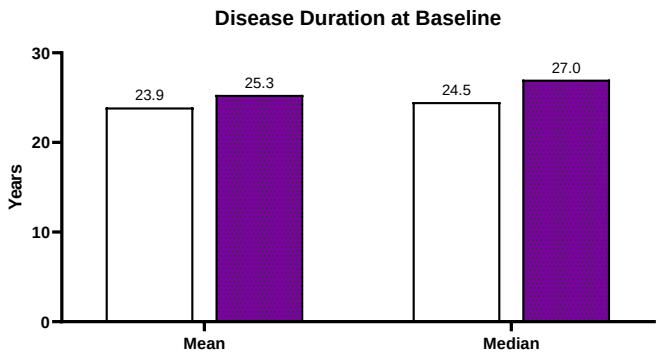
Key Inclusion Criteria:
- IGA of 3 or 4
- BSA ≥ 10%
- EASI ≥ 12
- NRS ≥7 at SV1
- NRS ≥5 at d0

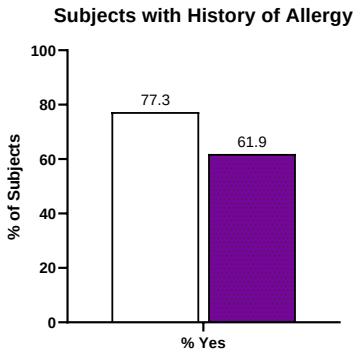


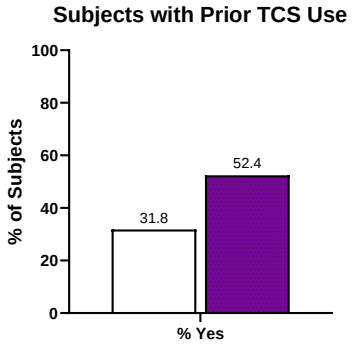
Baseline subject and disease characteristics

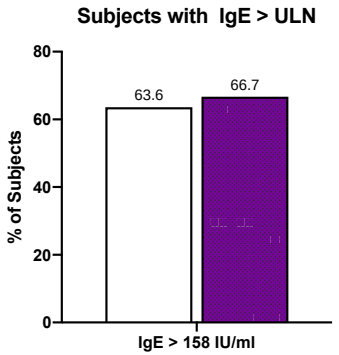
PBO (All Subjects)
KPL-716 (All Subjects)

Baseline Subject Characteristics

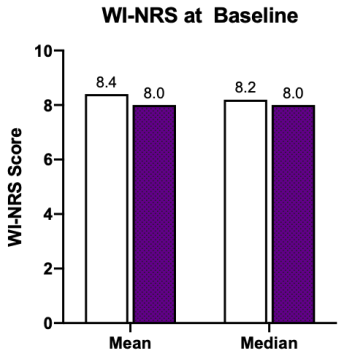


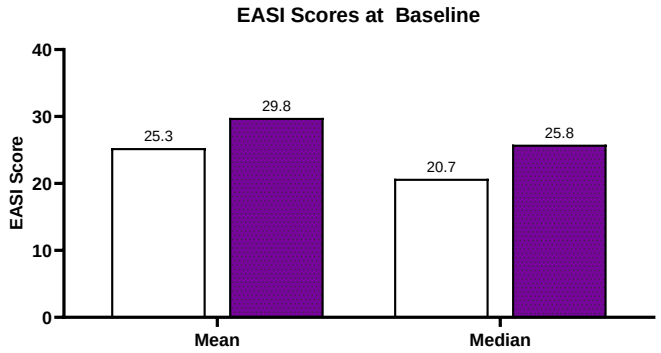


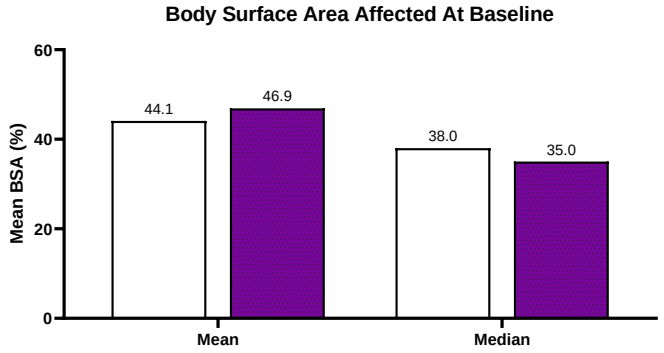


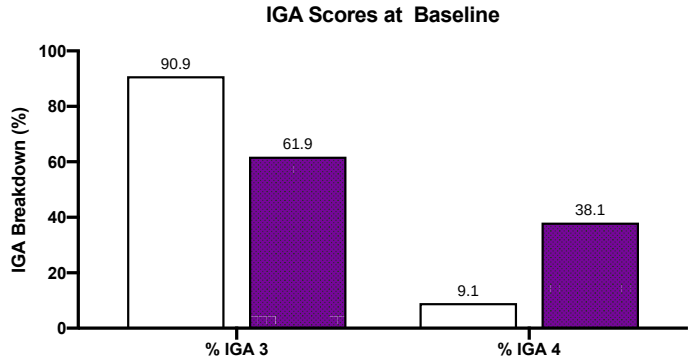


Baseline Disease Characteristics









ULN: Upper Limit of Normal; **PBO**: Placebo; **TCS:** Topical corticosteroid

Overview of treatment-emergent adverse events (TEAE) through 12-week treatment period

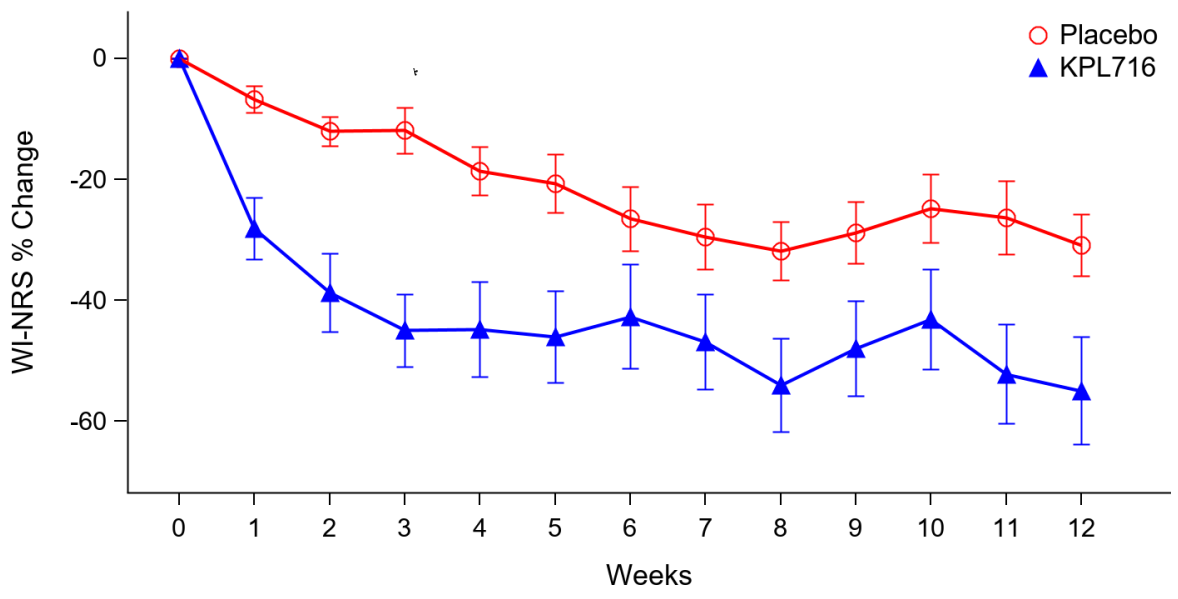
	Placebo (N=22)	KPL-716 (N=21)
Any TEAE	12 (54.5%)	18 (85.7%)
Any Drug-Related TEAE	4 (18.2%)	8 (38.1%)
Any Moderate or Severe TEAE	6 (27.3%)	11 (52.4%)
Any Drug-Related Moderate or Severe TEAE	0	2 (9.5%)
Any Treatment-Emergent Serious AE	0	0
Any Drug-Related Serious TEAE	0	0
Any Atopic Dermatitis Flare-Related 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%)

KPL-716 showed rapid and sustained reduction in pruritus versus placebo despite more flares in the active treatment arm

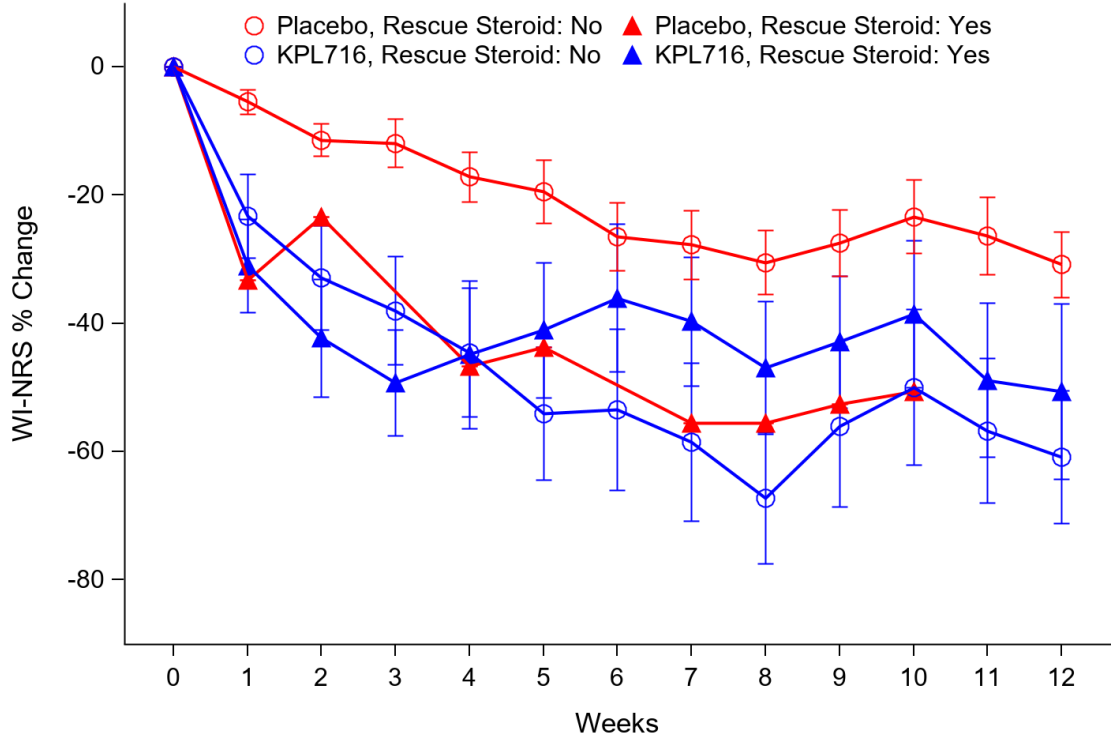
Percent Change in Weekly Average WI-NRS from Baseline
As Observed



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
RSC: PLB		1	1	1	1	1	1	1	1	1	1	1	1
RSC: KPL			1	2	2	2	4	4	7	9	9	9	9

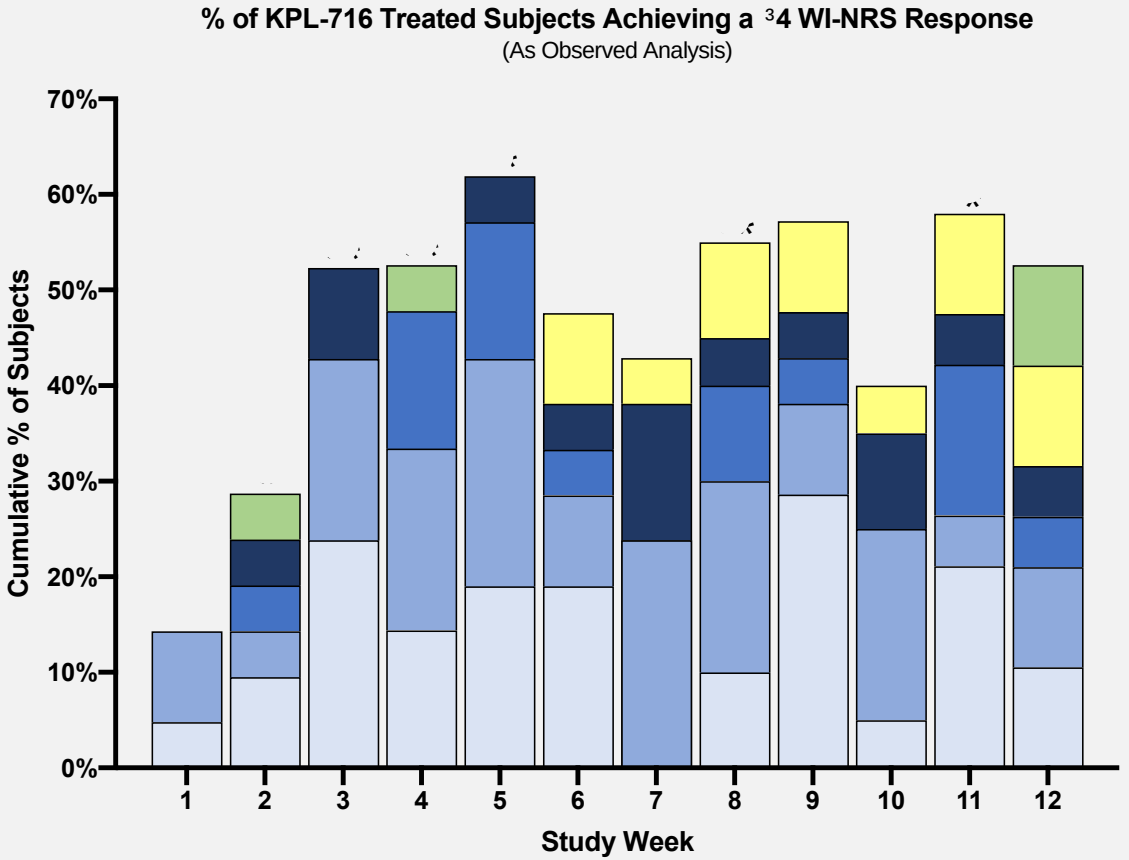
RSC: PLB - Rescue TCS used in placebo arm
RSC: KPL – Rescue TCS used in KPL-716 arm
Note: Based on full interim data set as of 1st database lock

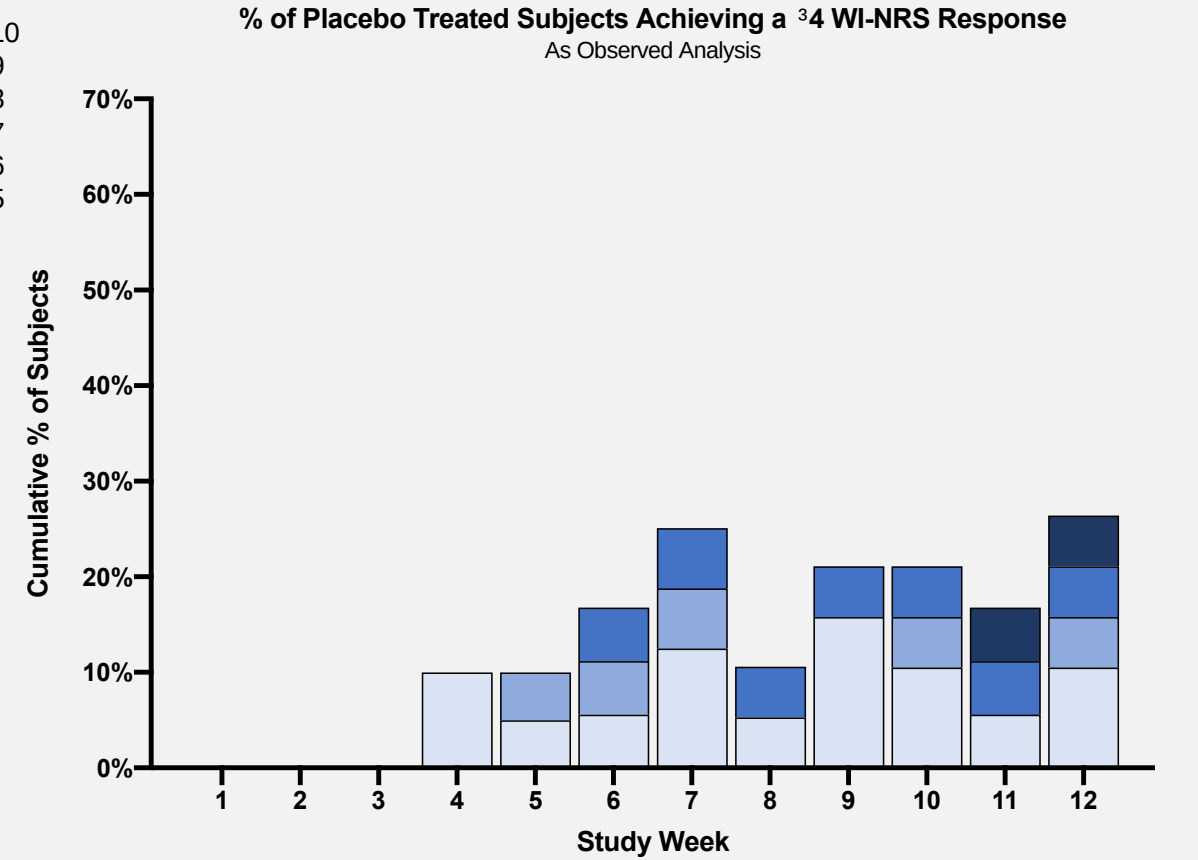
Percent Change in Weekly Average WI-NRS from Baseline
As Observed



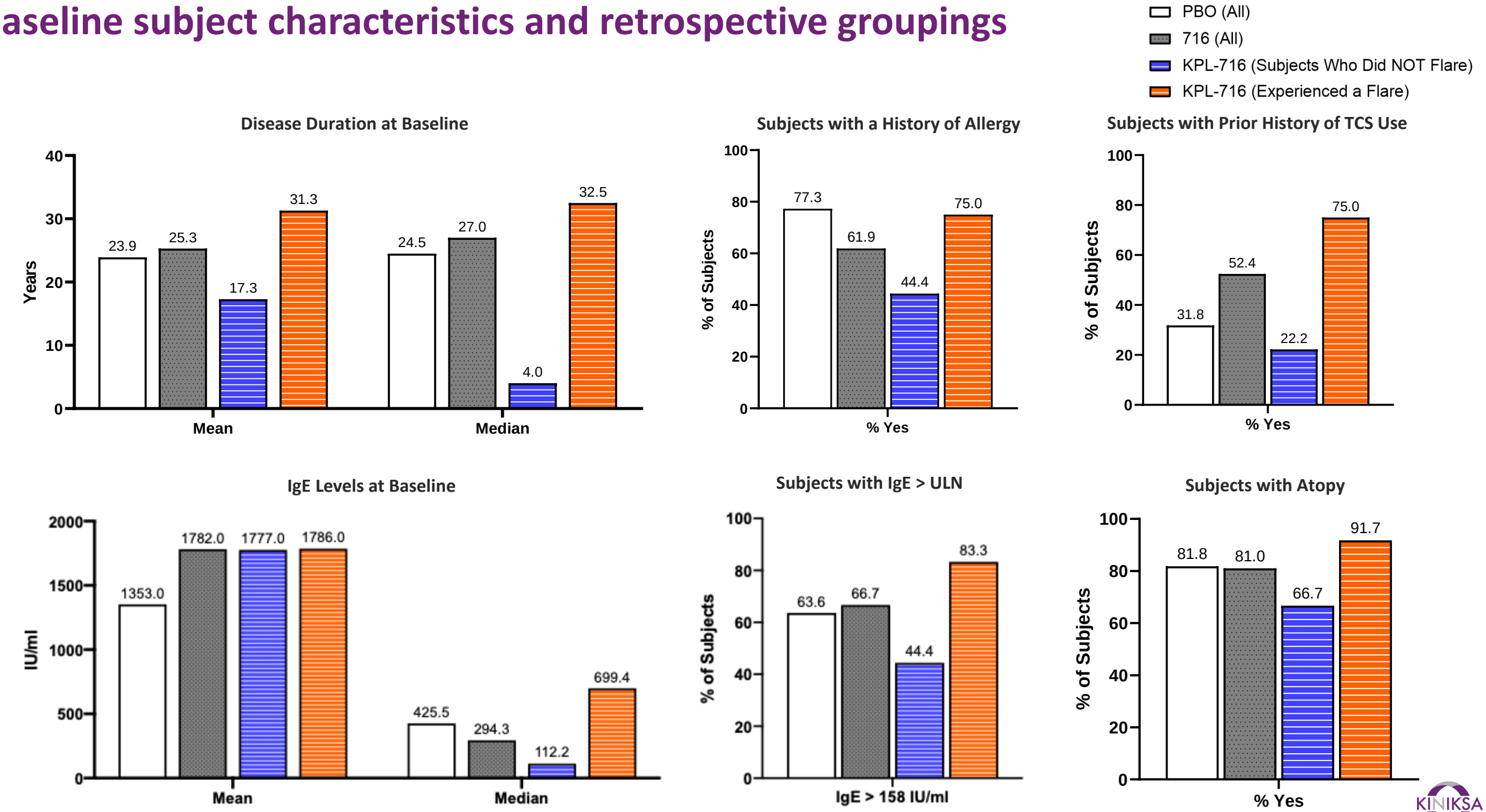
Note: Placebo TCS rescue curve is n=1
Note: y-axis is different from chart on the left

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



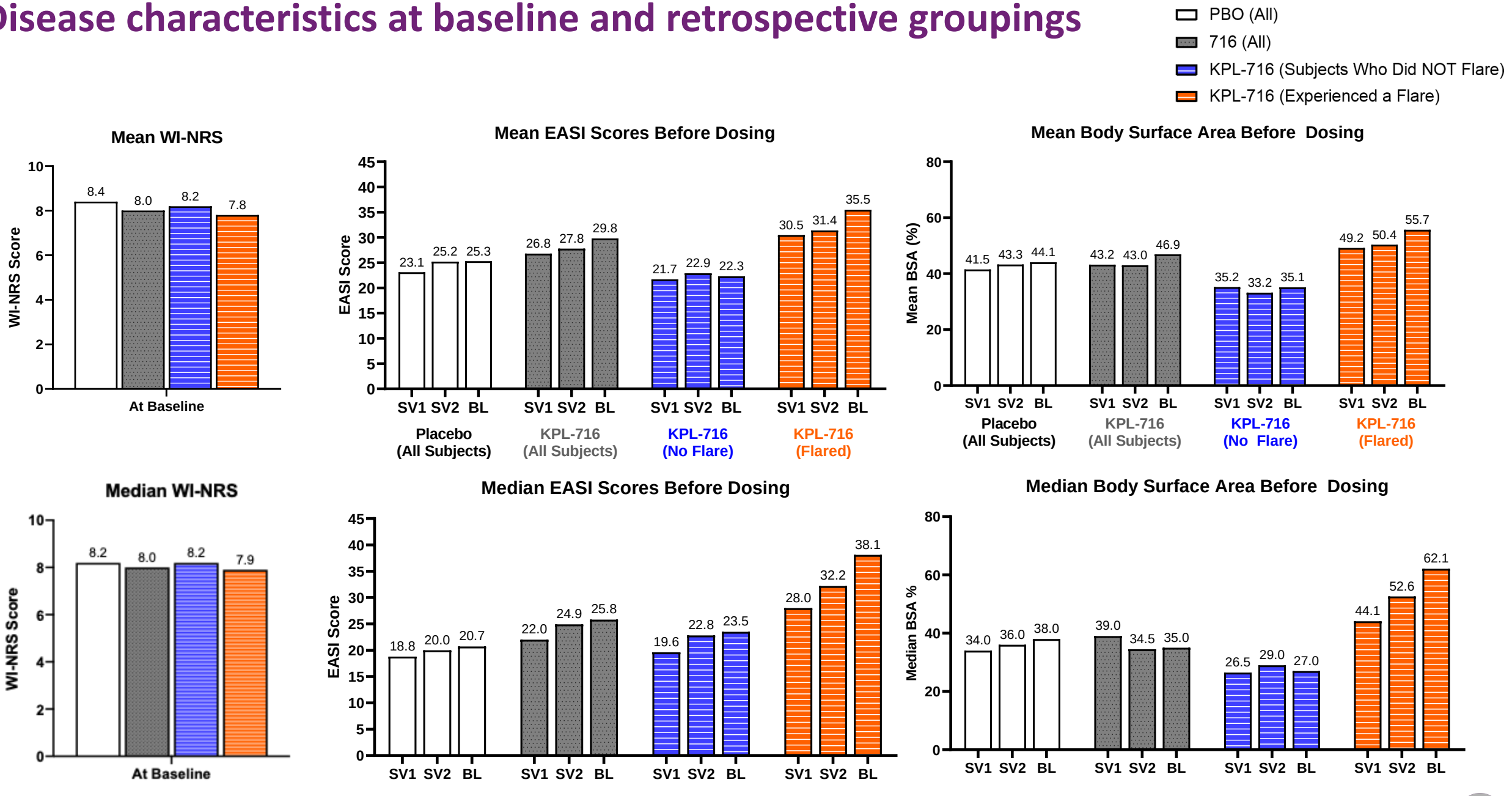


Baseline subject characteristics and retrospective groupings



Note: Based on full interim data set as of 1st database lock **ULN**: Upper Limit of Normal; **PBO**: Placebo; **TCS:** Topical corticosteroid

Disease characteristics at baseline and retrospective groupings



Note: Based on full interim data set as of 1st database lock
SV1: Screening Visit 1; **SV2**: Screening Visit 2; **BL**: Baseline on Day 0 before first dose of KPL-716; **TCS**: Topical Corticosteroid; **PBO:** Placebo

KPL-404 – Preclinical

(monoclonal antibody targeting CD40)

Rilonacept | Mavrilimumab | KPL-716 | **KPL-404** | KPL-045

Humanized monoclonal antibody inhibitor of signaling between CD40L and CD40

- CD40/CD40L interaction between B & T-cells are required for humoral responses
- Antigen presenting cells express and require signaling through CD40 for activation
- Proof-of-mechanism established in non-human primates
- Plan to file IND in 2H 2019

KPL-045 – Preclinical

(monoclonal antibody targeting CD30L)

Rilonacept | Mavrilimumab | KPL-716 | KPL-404 | **KPL-045**

Fully-human monoclonal antibody inhibitor of signaling between CD30 and CD30L

- Involved in T-effector memory function, humoral response & T_H2 immunity
- CD30L is expressed at high levels on activated T cells
- Proof-of-mechanism established in mice and non-human primates
- Preclinical activities

Anticipated 2019-2020 milestones for rilonacept, mavrilimumab and KPL-716

Program	Milestone	Anticipated Timing
Rilonacept	Final data from Phase 2 trial in different pericarditis populations	2H 2019
	Top-line data from Phase 3 RHAPSODY trial in recurrent pericarditis	2H 2020
Mavrilimumab	Provide data from non-clinical and biomarker studies on the role of GM-CSF in GCA	2H 2019
	Announce additional investigational indication for mavrilimumab	2H 2019
	Top-line data from global Phase 2 trial in GCA	2H 2020
KPL-716	Provide data from non-clinical and biomarker studies of IL-31 and OSM in prurigo nodularis and atopic dermatitis	2H 2019
	Present top-line data from Phase 2a trial in PN	1H 2020
	Present top-line data from exploratory Phase 2 study in diseases characterized by chronic pruritus	Throughout 2020

Kiniksa at a glance

Corporate Highlights

Bermuda-Based Corporate Entity

5
Pipeline Programs

>180
Issued Patents

Financial Highlights

~$287M
Cash & Short-Term Investments[1]

54.9M
Shares Outstanding[1]

Capital Allocation to High Value Opportunities Across Existing Portfolio, Internal R&D and Business Development

1) As of 6/30/19

Every Second Counts!™