



*Every Second Counts!™*

**J.P. Morgan Healthcare Conference**  
*January 11, 2021*

# 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 corporate priorities and strategy; product development and prospects; potential value drivers; prospects for 2021; potential indications; potential market opportunities and competitive position; on going, planned and potential clinical trials and other studies; timing and potential impact of clinical data; mechanisms and potential of our product candidates; regulatory and other submissions, applications and approvals; commercial strategy, pre-commercialization activities and commercial launch timing; expected run rate for our cash, cash equivalents and short-term investments; 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 or startup thereof; 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; impact of additional data from us or other companies; potential undesirable side effects caused by our product candidates; our reliance on third parties for manufacturing and conducting clinical trials, research and other studies; potential changes in our strategy, corporate priorities, operating plan and funding requirements; drug substance and/or drug product shortages; substantial new or existing competition; potential for applicable regulatory authorities to not accept our regulatory filings or to delay or deny approval of any of our product candidates or to require additional trials to support any such approval; complications in coordinating requirements, regulations and guidelines of regulatory authorities across jurisdictions for our clinical trials; potential impact of the COVID-19 pandemic, and measures taken in response to the pandemic, on our business and operations as well as the business and operations of our manufacturers, CROs upon whom we rely to conduct our clinical trials, and other third parties with whom we conduct business or otherwise engage, including the FDA and other regulatory authorities; and our ability to attract and retain qualified personnel. These and the other important factors are 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, 2020 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 Patient-Centric Leadership in Immune-Modulating Therapies

Leveraging internal & external expertise to drive growth

**4 Product Candidates; First  
PDUFA Date in 1Q 2021<sup>1</sup>**

**Validated Mechanisms or  
Strong Biologic Rationale**



**Targeting Debilitating Diseases  
with Unmet Medical Need**



**Pipeline-in-a-Molecule  
Potential Across the Portfolio**

# Worldwide Commercial Rights to Four Immune-Modulating Product Candidates

Program & Target	Preclinical	Phase 1	Phase 2	Phase 3	Regulatory <sup>2</sup>	Commercial Rights
<b>Rilonacept<sup>1</sup></b> IL-1α & IL-1β	<b>Recurrent Pericarditis</b>				<b>PDUFA: 03/21/21; Orphan Drug Designation &amp; Breakthrough Therapy Designation</b>	<b>Worldwide (Excluding MENA)</b>
<b>Mavrilimumab</b> GM-CSFRα	<b>Giant Cell Arteritis</b>				<b>Orphan Drug Designation</b>	<b>Worldwide</b>
	<b>COVID-19 Pneumonia &amp; Hyperinflammation</b>					<b>Worldwide</b>
<b>Vixarelimab</b> OSMRβ	<b>Prurigo Nodularis</b>				<b>Breakthrough Therapy Designation</b>	<b>Worldwide</b>
<b>KPL-404</b> CD40	<b>Severe Autoimmune Diseases</b>					<b>Worldwide</b>

1) Rilonacept (ARCALYST®) is approved and marketed for cryopyrin-associated periodic syndrome (CAPS) and maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA) in the United States by Regeneron Pharmaceuticals, Inc.; 2) The FDA granted Breakthrough Therapy designation to rilonacept for recurrent pericarditis in 2019 and Orphan Drug designation to rilonacept for pericarditis in 2020; The FDA granted Orphan Drug designation to mavrilimumab for giant cell arteritis in 2020; The FDA granted Breakthrough Therapy designation to vixarelimab for the treatment of pruritus associated with prurigo nodularis in 2020; IL-1α = interleukin-1α ; IL-1β = interleukin-1β; GM-CSFRα = granulocyte macrophage colony stimulating factor receptor alpha; OSMRβ = oncostatin M receptor beta; PDUFA = Prescription Drug User Fee Act ; MENA = Middle East North Africa

# Product Candidates Based on Validated Mechanisms and with Attractive Commercial Prospects

	Indication	Validated Mechanism	U.S. Current Prevalence	U.S. Current Addressable
<b>Rilonacept</b>	<b>Recurrent Pericarditis</b>	✓	~40k <sup>1</sup>	~14-17k <sup>1</sup>
<b>Mavrilimumab</b>	<b>Giant Cell Arteritis</b>	✓	~75-150k <sup>2</sup>	~45-65k <sup>3</sup>
<b>Vixarelimab</b>	<b>Prurigo Nodularis</b>	✓	~300k <sup>4</sup>	~75-105k <sup>5</sup>
<b>KPL-404</b>	<b>Severe Autoimmune Diseases</b>	✓	TBD	TBD

1) 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 2) Chandran AK, Udayakumar PD, Crowson CS, Warrington KJ, Matteson EL. The incidence of giant cell arteritis in Olmsted County, Minnesota, over a 60-year period 1950–2009. Scand J Rheumatol. 2015; 44(3):215–8. Labarca C, Koster MJ, Crowson CS, et al. Predictors of relapse and treatment outcomes in biopsy-proven giant cell arteritis: a retrospective cohort study. *Rheumatology (Oxford)*. 2016;55(2):347-356. Medcape; Trinity Lifesciences primary market research; Trinity Lifesciences analysis of Integrated 2016-2019 Medicare FFS & 2016-2019 IBM MarketScan Commercial & Medicare Supplemental data 3) Trinity Life Sciences –Trinity Life Sciences – EvidenceFirst Database Analysis, HCUP/Medicare Data, Quantitative Survey (n=102 rheumatologists) 4) Trinity Life Sciences – 2020 Analysis; Trinity Life Sciences - HCUP/Medicare Data; 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, 200 5) Trinity Life Sciences Analysis; Moderate/Severe Patients inadequately controlled by topical corticosteroids

# Strong Execution in 2020 Sets the Stage for a Transformational 2021

	2020	2021
<b>Rilonacept</b>	✓ Positive Phase 3 data in RP	PDUFA goal date: 3/21/21; potential commercial launch in 1H 2021 <sup>1</sup>
<b>Mavrimumab</b>	✓ Positive Phase 2 data in GCA; encouraging data in COVID-19	Next steps for program in 1H 2021; COVID-19 data in 1H 2021
<b>Vixarelimab</b>	✓ Positive Phase 2a data in PN	Phase 2b study in PN evaluating a range of once-monthly dose regimens
<b>KPL-404</b>	✓ Encouraging preliminary Phase 1 data	Final Phase 1 data in 1H 2021

# Rilonacept

IL-1 $\alpha$  and IL-1 $\beta$  cytokine trap

**Disease Area:** Recurrent pericarditis<sup>1</sup>; painful and debilitating auto-inflammatory cardiovascular disease

**Competition<sup>2</sup>:** No FDA-approved therapies for recurrent pericarditis

**Regulatory:** U.S. Orphan Drug designation in pericarditis; Breakthrough Therapy designation in recurrent pericarditis

**Status:** sBLA accepted with priority review; PDUFA goal date of March 21, 2021

**Economics:** 50/50 profit split on the approved indications in the U.S.

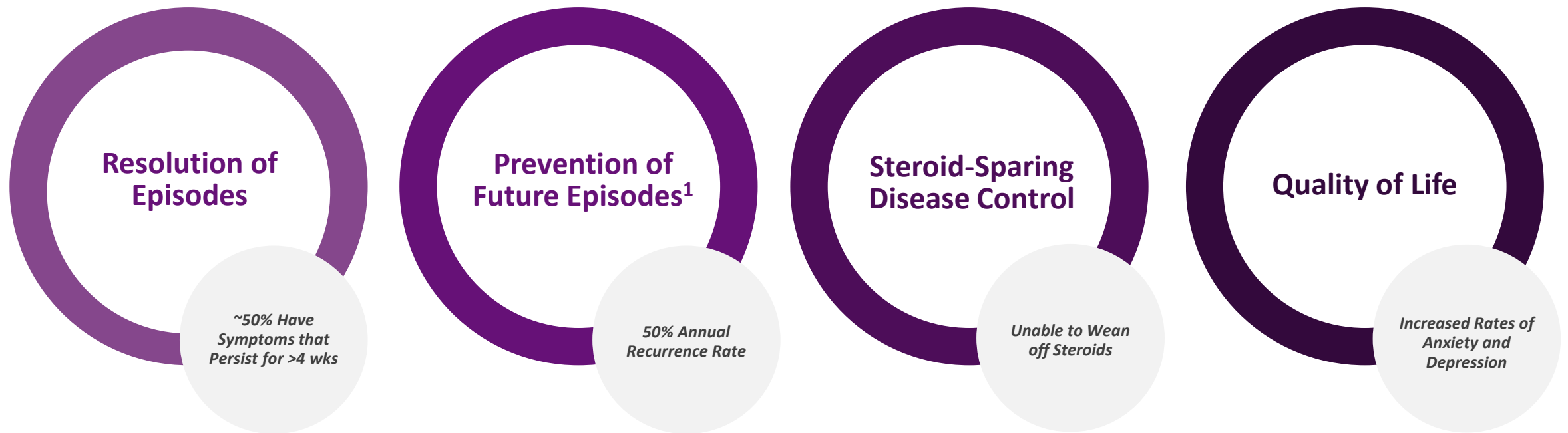
**Rights:** Upon approval Kiniksa has the rights to recurrent pericarditis worldwide (excluding MENA)

1) Rilonacept (ARCALYST®) is approved and marketed for cryopyrin-associated periodic syndrome (CAPS) and maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA) in the United States by Regeneron Pharmaceuticals, Inc; 2) 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; IL-1 $\alpha$  = interleukin-1 $\alpha$  ; IL-1 $\beta$  = interleukin-1 $\beta$ ;PDUFA = Prescription Drug User Fee Act; sBLA = supplemental Biologics License Application; MENA = Middle East North Africa



# Key Areas of Unmet Need in Patients with Recurrent Pericarditis

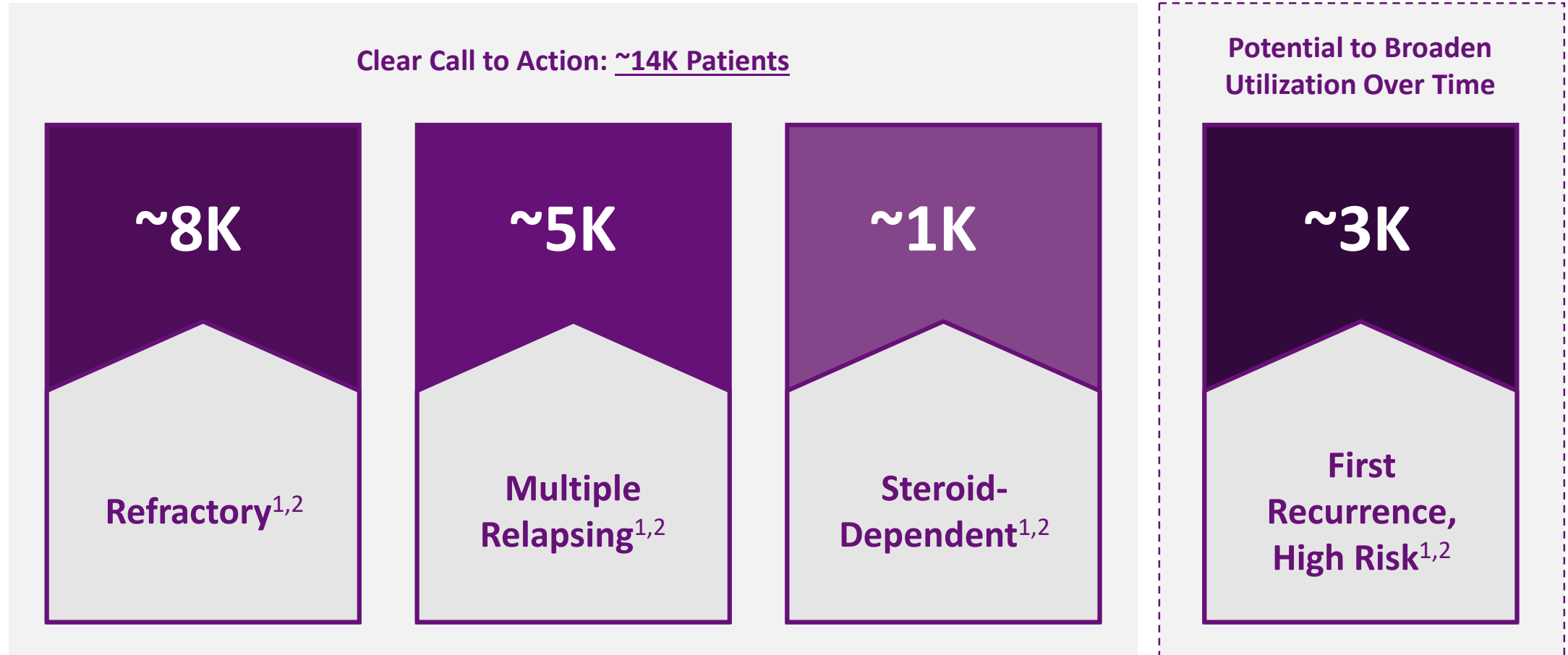
Recurrent pericarditis episodes: painful, debilitating and disruptive to quality of life





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

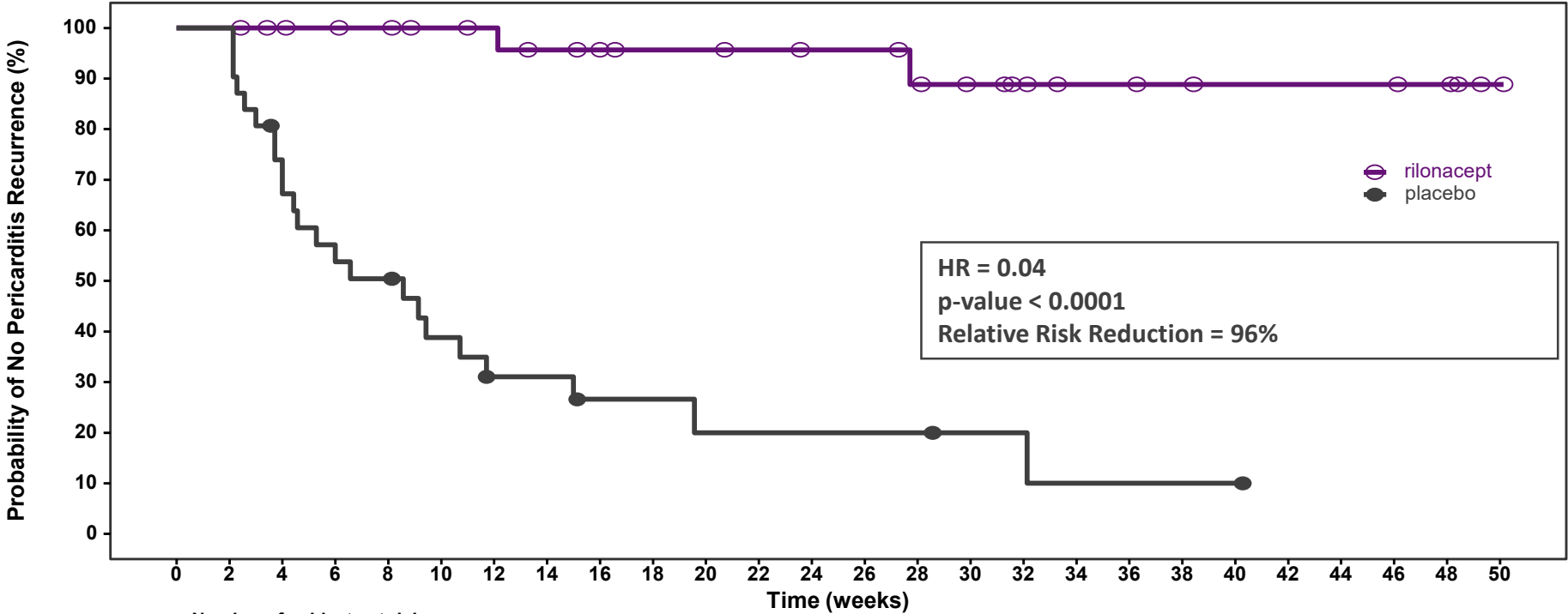
~14K patients with inadequate response to conventional therapy and persistent underlying disease



1) Klein A, Cremer P, Kontzias A, Furqan M, Tubman R, Roy M, Magestro M. Annals of Epidemiology. 2019;36:71; 2) Lin D, Majeski C, DerSarkissian M, Magestro M, Cavanaugh C, Laliberte F, Lejune D, Mahendran M, Duh M, Klein A, Cremer P, Kontzias A, Furqan M, Tubman R, Roy M, Mage. (Nov, 2019). *Real-World Clinical Characteristics and Recurrence Burden of Patients Diagnosed with Recurrent Pericarditis in the United States*. Poster session presented at the American Heart Association, Philadelphia, PA.; 3) ClearView Forecasting Analysis 2019 Q1

# Highly Statistically Significant Primary Efficacy Endpoint: Time-to-First Adjudicated Pericarditis Recurrence

Pivotal Phase 3 Rilonacept Data



	Number of Patients with Recurrence <sup>1</sup> n (%)	Number of Weeks to Recurrence <sup>1</sup> Median (95% CI)
Rilonacept	2 (6.7)	NE (NE, NE)
Placebo	23 (74.2)	8.6 (4.0, 11.7)

	0	2	4	6	8	10	12	14	16	18	20	22	24	26	28	30	32	34	36	38	40	42	44	46	48	50
rilonacept	30	30	28	27	26	24	23	21	20	17	17	16	15	15	13	11	9	7	7	6	5	5	5	5	4	1
Placebo	31	31	22	17	15	10	7	7	4	4	3	3	3	3	3	2	2	1	1	1	1	0	0	0	0	0

Annualized incidence of pericarditis recurrence decreased from 4.42 episodes per year prior to the study to 0.15 episodes per year while on rilonacept treatment.

1) Mean (median, range); NE = Not Estimable; HR = hazard ratio  
Klein, et. al. New England Journal of Medicine. Nov 2020



# Strategic Imperatives for Potential Commercial Launch of Rilonacept in Recurrent Pericarditis

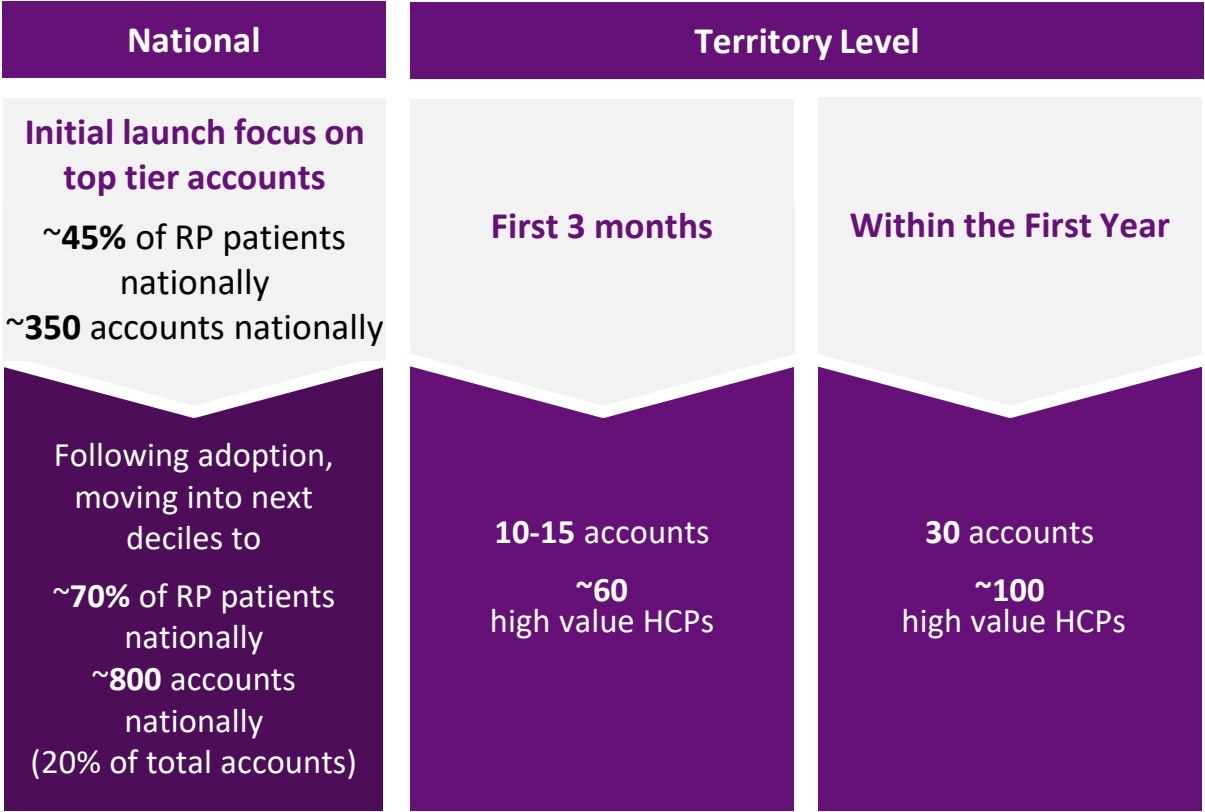
Unmet Need	Standard of Care	Payer Reimbursement	Patient Support
<p>Drive awareness and understanding of recurrent pericarditis and the role of inappropriate IL-1 production</p> <ul style="list-style-type: none"> <li>• ‘Heart of Inflammation’ disease awareness campaign and website</li> <li>• Continued presence at scientific congresses</li> <li>• Advocacy engagement, podcasts and videos</li> </ul>	<p>Aim for rilonacept to be the product of choice for the treatment and prevention of recurrent pericarditis</p> <ul style="list-style-type: none"> <li>• Specialty cardiovascular sales force</li> <li>• Efficient digital marketing</li> <li>• Peer to Peer speaker program</li> <li>• Patient support network</li> <li>• Scientific Congress Exhibits and Symposia (ACC, ESC, AHA)</li> </ul>	<p>Demonstrate product benefits, establish rapid payer coverage, &amp; navigate potential access barriers</p> <ul style="list-style-type: none"> <li>• Compelling value proposition and supportive tools (value dossier and budget impact model)</li> <li>• Comprehensive payer engagement plan</li> <li>• Specialty pharmacy network distribution</li> </ul>	<p>Optimize the patient and customer experience with rilonacept and Kiniksa</p> <ul style="list-style-type: none"> <li>• High-touch patient support, reimbursement services, patient financial assistance, initiation support (Quick Start), injection training</li> <li>• Partner with the pericarditis community to improve advocacy, education and support for affected patients</li> </ul>

# Specialty Cardiology Salesforce Expected to Reach ~70% of U.S. Recurrent Pericarditis Patients

Estimated Recurrent Pericarditis Patients by Account



Focused & Targeted Sales Execution



Specialty cardiology sales force of ~30 reps

# Mavrilimumab

Monoclonal antibody inhibitor targeting GM-CSFR $\alpha$

**Disease Areas:** Giant Cell Arteritis (GCA): chronic inflammatory disease of medium-to-large arteries; COVID-19 Pneumonia and Hyperinflammation

**Competition<sup>1</sup>:** Only one FDA-approved therapy for GCA, but unmet needs remain

**Regulatory:** U.S. Orphan Drug designation in GCA

**Status:** Positive Phase 2 data in GCA reported in Q4 2020; Phase 2 data from Phase 2/3 in severe COVID-19 pneumonia and hyperinflammation expected in 1H 2021

**Economics:** Clinical, regulatory and sales milestones; tiered royalty on annual net sales

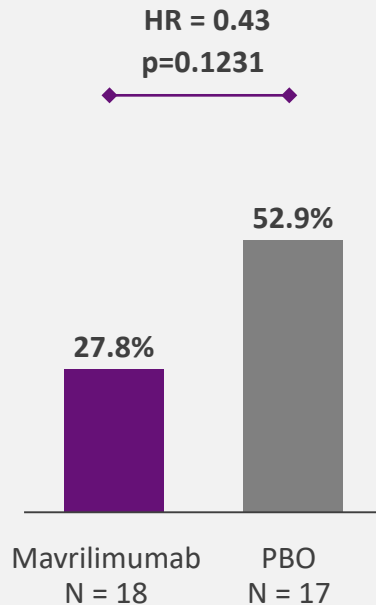
**Rights:** Worldwide

# Unmet Need and Commercial Opportunity for Safe and Effective GCA Therapies

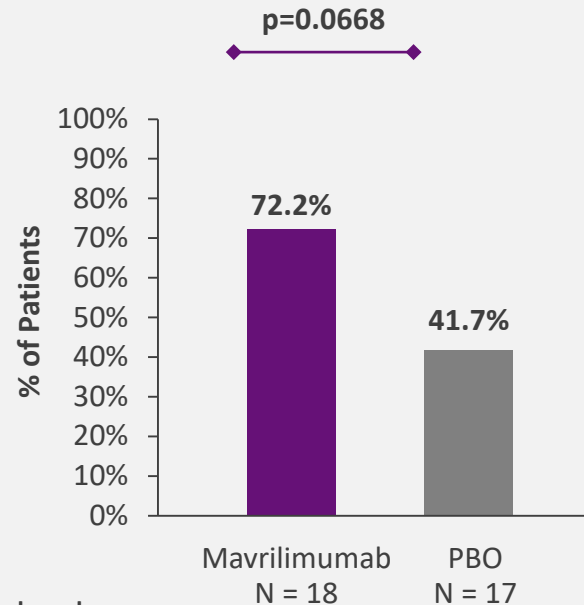
Mavrilimumab Phase 2 giant cell arteritis data<sup>1</sup>

## Relapsing/Refractory Cohort

### Patients With Flare by Week 26



### Sustained Remission at Week 26



\*Nominal p values

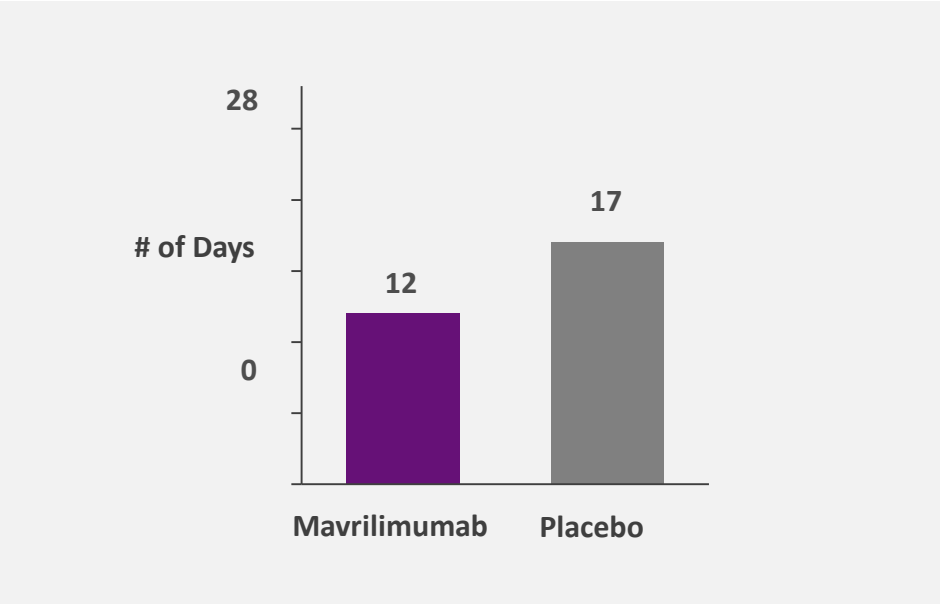
## Remaining Unmet Need

- Cumulative U.S. GCA prevalence expected to grow 50% by 2035<sup>2</sup>
- ~50% of relapse / refractory patients are unable to achieve sustained remission within 1-year of starting treatment with approved biologics<sup>3</sup>
- Mechanistic (GM-CSFR $\alpha$  vs. IL-6) and administrative (Q2WK vs QWK) differentiation
- Well-tolerated safety profile particularly important given large elderly patient population

# Encouraging Trends of Decreased Mortality and Duration of Mechanical Ventilation

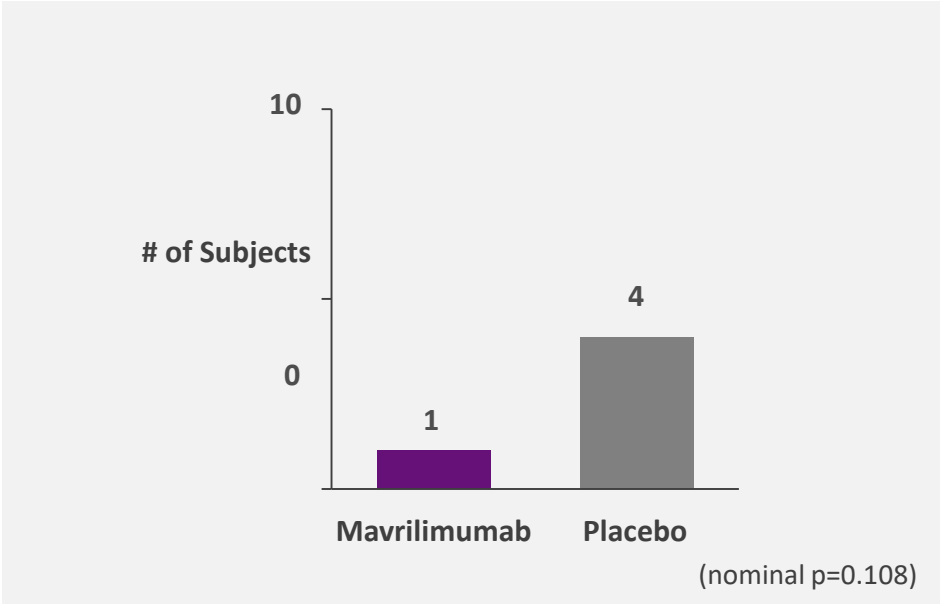
U.S. investigator-initiated study in patients with severe COVID-19 pneumonia and hyperinflammation<sup>1</sup>

Median (IQR) Duration of Mechanical Ventilation



- 4 of the 5 patients who progressed to mechanical ventilation in the mavrilimumab arm had recovered by Day 28.
- All 4 patients in the placebo arm who progressed to mechanical ventilation had died by Day 28.

Death by Day 60



- There was 1 death (4.8%) in the mavrilimumab arm by Day 28, compared to 3 deaths (15.8%) in the placebo arm (nominal p=0.222). By Day 60 there was 1 death (4.8%) in the mavrilimumab arm, compared to 4 deaths (21.1%) in the placebo arm (nominal p=0.108).

1) There was a 20.5% relative increase in the primary efficacy endpoint, the proportion of patients alive and off supplemental oxygen at Day 14 (mavrilimumab: 57.1% [n=21]; placebo: 47.4% [n=19]; nominal p=0.536). There was a 20.7% relative increase in the secondary efficacy endpoint, the proportion of patients alive and without respiratory failure at Day 28 (mavrilimumab: 95.2%; placebo: 78.9%; nominal p=0.172). There was no difference in serious adverse events between the mavrilimumab arm and the placebo arm.



# Potential Broad Utility

Next steps for development of mavrilimumab expected in 1H 2021

## Mavrilimumab Data Across 3 Indications:

### Giant Cell Arteritis

Phase 2 trial of mavrilimumab in giant cell arteritis achieved both the primary and secondary efficacy endpoints with statistical significance

### Severe COVID-19 Pneumonia and Hyperinflammation

Encouraging and similar trends in mortality shown in 28-day clinical outcomes data from the open-label treatment protocol in Italy and U.S. IIS

### Rheumatoid Arthritis

Mavrilimumab was dosed in over 550 patients with rheumatoid arthritis through Phase 2b clinical studies in Europe and achieved prospectively-defined primary and secondary efficacy endpoints

*Mavrilimumab has been shown to be well-tolerated in giant cell arteritis, severe COVID-19 pneumonia and hyperinflammation, and rheumatoid arthritis clinical trials*



# Vixarelimab

Monoclonal antibody inhibitor targeting OSMR $\beta$

**Disease Area:** Prurigo Nodularis (PN); chronic inflammatory skin disease with pruritic nodules

**Competition<sup>1</sup>:** No FDA-approved therapies for PN

**Regulatory:** U.S. Breakthrough Therapy designation for the treatment of pruritus associated with prurigo nodularis

**Status:** Enrolling and dosing in Phase 2b clinical trial in Q4 2020 evaluating a range of once-monthly dose regimens

**Economics:** Clinical, regulatory and sales milestones; tiered royalty on annual net sales

**Rights:** Worldwide

1) Journal of the American Academy of Dermatology - Analysis of Real-World Treatment Patterns in Patients with Prurigo Nodularis: [https://www.jaad.org/article/S0190-9622\(19\)32744-6/pdf](https://www.jaad.org/article/S0190-9622(19)32744-6/pdf) ; OSMR $\beta$  = oncostatin M receptor beta

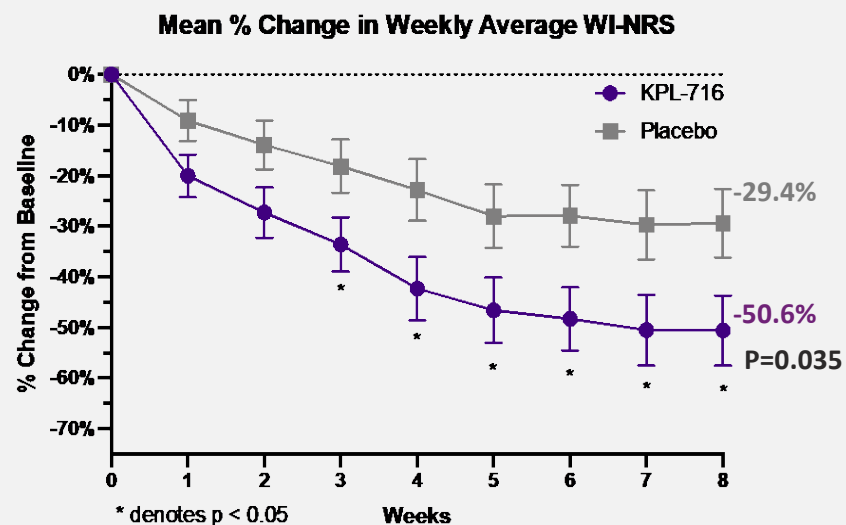
# Dual Mechanism Offers Potential Pruritus Relief and Nodule Improvement

## Vixarelimab Phase 2a prurigo nodularis data

Vixarelimab is the only mAb targeting OSMR $\beta$ , which mediates signaling of key cytokines (IL-31 & OSM)

### Primary Efficacy Endpoint

Mean change in weekly-average WI-NRS at Week 8 was -50.6% in vixarelimab recipients compared to -29.4% in placebo recipients (p=0.035).



### Secondary Efficacy Endpoint

30.4% of vixarelimab recipients achieved a PN-IGA score of 0/1 at Week 8 compared to 7.7% of placebo recipients (p=0.032).

Subject 1



Subject 2



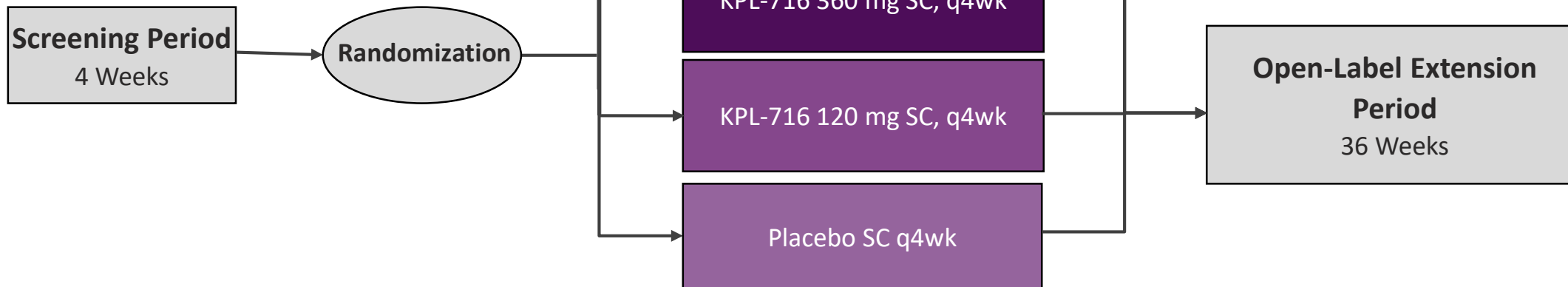
Representative Treatment Response

# Vixarelimab Phase 2b Dose-Ranging Study in Prurigo Nodularis

Enrollment and dosing of patients commenced in Q4 2020

## Expected to enroll approximately 180 patients

- Moderate-to-severe prurigo nodularis experiencing severe pruritus.
- Patients are required to stop antihistamines and topical treatments, including corticosteroids, for at least two weeks prior to dosing.
- Prurigo nodularis treatments, other than study drug, are not allowed except for rescue.



## Primary Efficacy Endpoint (Week 16):

- WI-NRS (% change from baseline in weekly average)

## Key Secondary Efficacy Endpoints (Week 16):

- WI-NRS, weekly avg. (proportion of subjects achieving  $\geq 6$ -point reduction from baseline)
- WI-NRS, weekly avg. (proportion of subjects achieving  $\geq 4$ -point reduction from baseline)
- PN-IGA-Stage (proportion of subjects achieving 0 or 1 from baseline)

# KPL-404

## Monoclonal antibody inhibitor interaction between CD40 and CD40L

**Disease Area:** 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<sup>1</sup>

**Scientific Rationale<sup>2,3</sup>:** Attractive target for blocking T-cell dependent, B-cell-mediated autoimmunity

**Status:** RO and TDAR suppression shown through Day 29 at 3mg/kg IV in Phase 1; Data to-date support subsequent study in patients, including potential monthly IV or SC monthly administration; Final data from all cohorts expected in 1H 2021

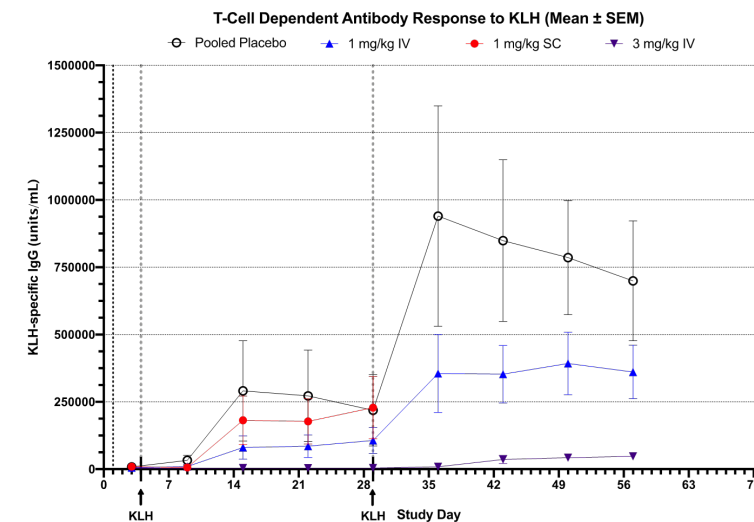
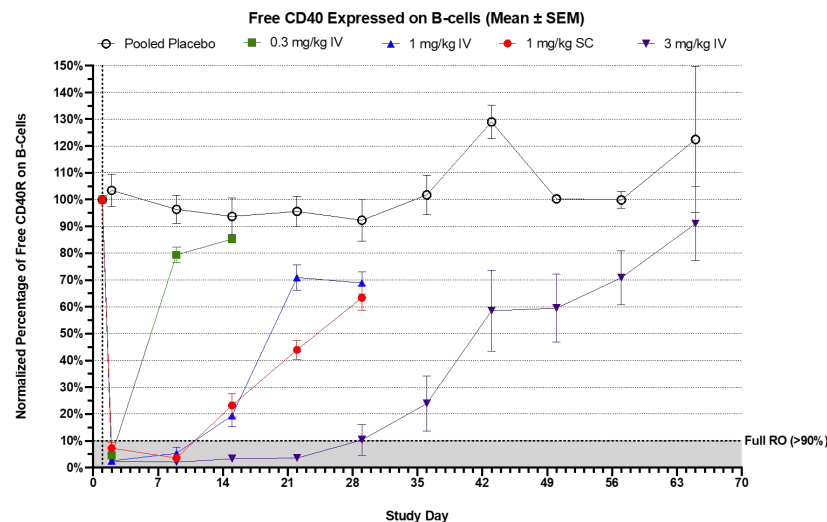
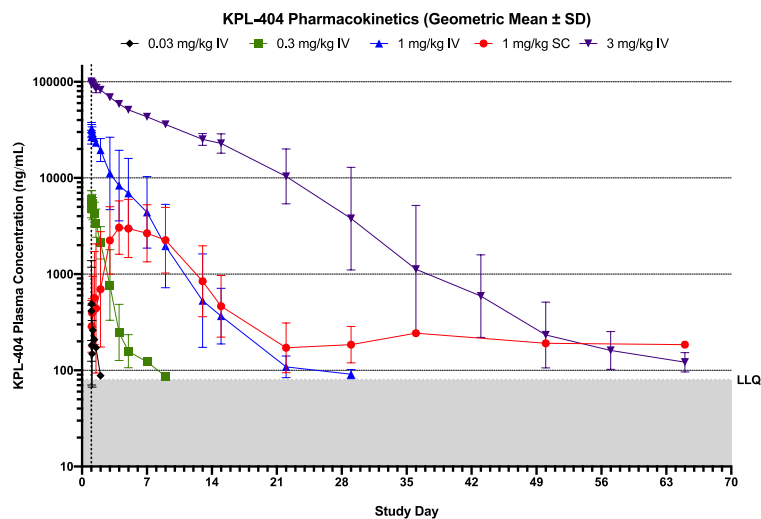
**Economics:** Clinical and regulatory milestones and royalty on annual net sales

**Rights:** Worldwide

1) Poster presentation at the Keystone Symposia: Antibodies as Drugs: New Horizons in the Therapeutic Use of Engineered Antibodies: KPL-404, a CD40 antagonist, blocked antigen-specific antibody responses in an in vivo NHP model and demonstrated strong PK/PD correlation; 2) Elgueta, et al. Immunol Rev 2009, 229 (1), 152-172; 3) Peters, et al. Semin Immunol 2009, 21 (5) 293-300; CD40L = CD40 ligand; RO = receptor occupancy; TDAR = T-cell Dependent Antibody Response

# RO and TDAR Suppression Shown Through Day 29 at 3mg/kg IV

## Preliminary KPL-404 Phase 1 data

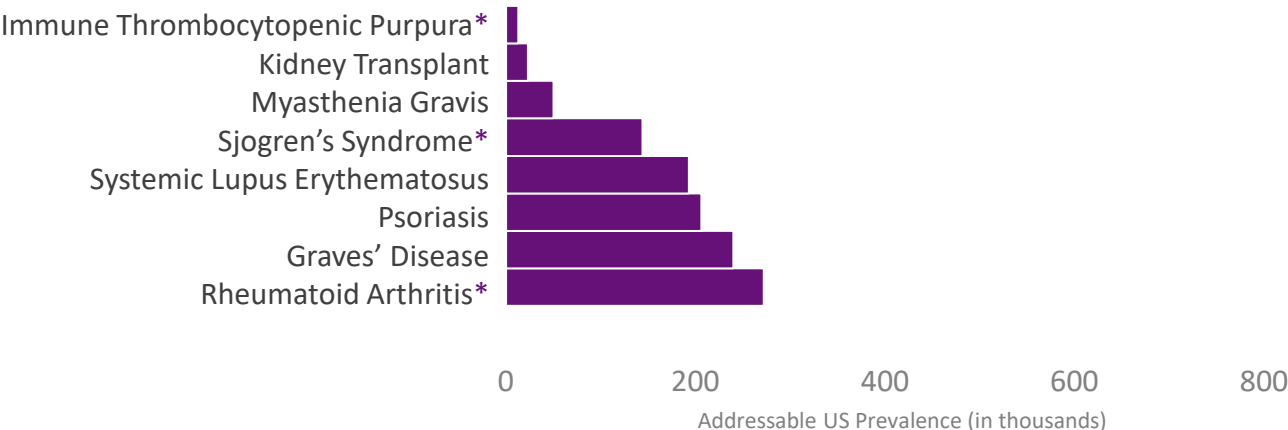


*Preliminary data support subsequent study in patients, including potential monthly intravenous or subcutaneous administration*

*Final data from all cohorts expected in 1H 2021*

# Potential for Evaluation of KPL-404 in a Broad Range of Autoimmune Diseases

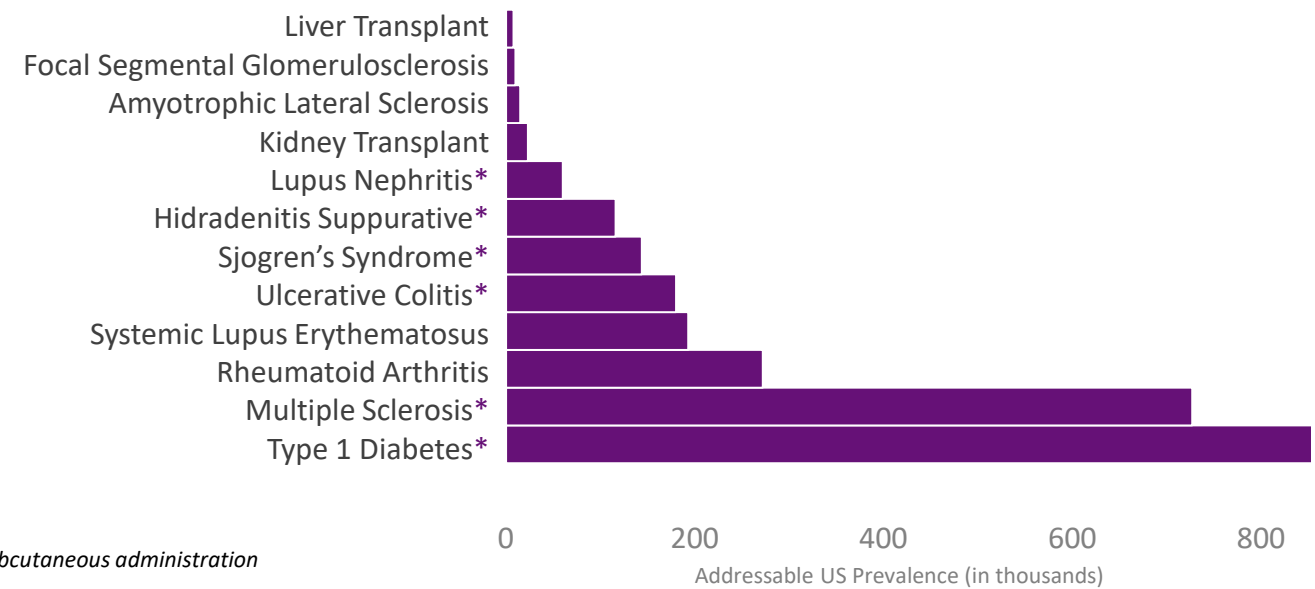
## Indications with Published Data<sup>1</sup>



## Indication Selection Criteria

- Robust Data or proof-of-concept supporting mechanism
- Differentiation vs. Competitors
- Commercial Attractiveness

## Indications with Pending Data & Trials Ongoing<sup>1</sup>



\*Indications evaluated with subcutaneous administration

1) With the CD40 mechanism



# Building Value at Kiniksa

## 2021 Corporate Priorities

<b>Rilonacept</b>	<i>PDUFA goal date: 3/21/21; potential commercial launch in 1H 2021<sup>1</sup></i>
<b>Mavrilimumab</b>	<i>Next steps for program in 1H 2021 and COVID-19 data in 1H 2021</i>
<b>Vixarelimab</b>	<i>Phase 2b study in PN evaluating a range of once-monthly dose regimens</i>
<b>KPL-404</b>	<i>Final Phase 1 data in 1H 2021</i>

***Year End 2020 ~\$323M Cash Reserves Expected to Fund Current Operating Plan into 2023<sup>2</sup>***



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