

# Cantor Conference Presentation

SEPTEMBER 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," "strategy," 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 commercial and clinical strategy; potential value drivers; potential indications; potential market opportunities and competitive position; ongoing, planned and potential clinical trials and other studies; timing and potential impact of clinical data; regulatory and other submissions, applications and approvals; commercial strategy and commercial activities; achievement of commercial milestones; 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; potential inability to demonstrate safety or efficacy or otherwise producing negative, inconclusive or uncompetitive results; potential for changes in final data from preliminary or interim data; potential inability to replicate in later clinical trials positive results from earlier trials and studies; our reliance on third parties for manufacturing and conducting clinical trials, research and other studies; our ability to source sufficient drug product, as needed, to meet our clinical and commercial requirements; our inability to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities; potential for applicable regulatory authorities to not accept our filings or to delay or deny approval of, or emergency use authorization for, any of our product candidates or to require additional data or trials to support any such approval or authorization; impediments delaying or preventing us from successfully executing on our commercial strategy for ARCALYST; potential changes in our strategy, clinical trial priority, operating plan and funding requirements; drug substance and/or drug product shortages; substantial new or existing competition; 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.

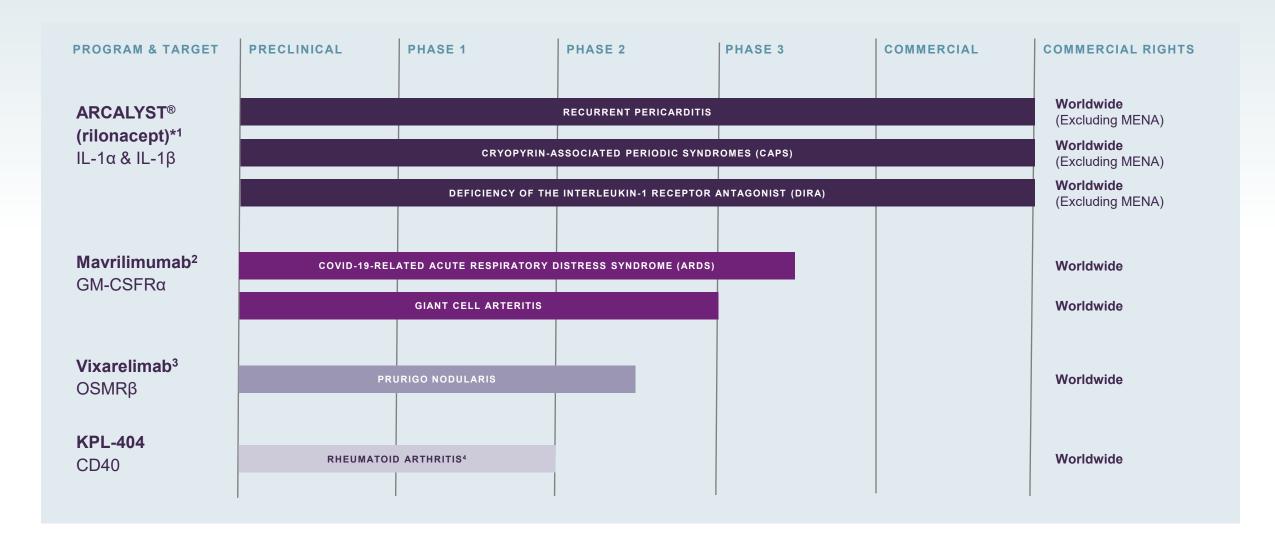
These and the important factors discussed in our filings with the U.S. Securities and Exchange Commission, including under the caption "Risk Factors" contained therein, could cause actual results to differ materially from those indicated by the forward-looking statements made in this presentation. These 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, except as required by law.

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.

ARCALYST is a registered trademark of Regeneron Pharmaceuticals, Inc. All other trademarks are the property of their respective owners.



# **Portfolio of Four Immune-Modulating Assets**





<sup>\*</sup> Approved in the U.S.

1) The FDA granted Breakthrough Therapy designation to ARCALYST for recurrent pericarditis in 2019 and Orphan Drug designation to ARCALYST for pericarditis in 2020. The European Commission granted Orphan Drug designation to ARCALYST for the treatment of idiopathic pericarditis in 2020; 2) The FDA granted Orphan Drug designation to mavrillimumab for giant cell arteritis in 2020; 3) The FDA granted Breakthrough Therapy designation to vixarelimab for the treatment of pruritus associated with prurigo nodularis in 2020; 4) Kiniksa plans to initiate a Phase 2 proof-of-concept trial in patients in the fourth quarter of 2021. The planned trial will provide safety and characterization of chronic administration as well as the potential to evaluate KPL-404 across a range of other autoimmune diseases; IL-1α = interleukin-1α; IL-1β = interleukin-1β; GM-CSFRα = granulocyte macrophage colony stimulating factor receptor alpha; OSMRβ = oncostatin M receptor beta; MENA = Middle East and North Africa

# **Execution Across Portfolio of In-Licensed Immune Modulating Assets**

### **ARCALYST**

Strong commercial launch in recurrent pericarditis driven by broad physician and patient adoption, and viable reimbursement conditions ahead of payers establishing coverage policy; Q3 net revenue expected to be \$9-10 million

# **MAVRILIMUMAB**

Broad utility demonstrated across multiple indications; potential best-in-class in reducing risk of death in patients with severe COVID-19-related ARDS, Phase 3 trial ongoing with data expected in Q1 2022; clear path to Phase 3 development in GCA

### **VIXARELIMAB**

First-in-class mechanism; Phase 2b dose-ranging trial in patients with prurigo nodularis enrolling

# **KPL-404**

Potential best-in-class treatment option for a broad range of autoimmune diseases; expected to initiate Phase 2 proof-of-concept trial in RA in Q4 2021

#### BY THE NUMBERS

1	FDA-approved therapy	40	Active and completed global clinical studies to date
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3	Clinical-stage assets in multiple indications	200+	Passionate and dedicated employees
_	multiple malcations		employees

3	Orphan Drug designations	2015	Compani founded

Breakthrough designations	2021	Commercial availability of first and only FDA- approved therapy for recurrent pericarditis in the US: ARCALYST® (rilonacept)
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# **ARCALYST®**



# IL-1α AND IL-1β CYTOKINE TRAP

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

**COMPETITION**<sup>2</sup>: First and only FDA-approved therapy for recurrent pericarditis

**REGULATORY:** U.S. Orphan Drug designation in pericarditis; U.S. Breakthrough Therapy designation in recurrent pericarditis; European Commission Orphan Drug designation in idiopathic pericarditis

**STATUS:** FDA-Approved

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

RIGHTS: Kiniksa has the worldwide rights (excluding MENA) to recurrent pericarditis, CAPS and DIRA



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

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

# **CLEAR CALL TO ACTION: ~14K PATIENTS**





# Key Areas of Unmet Need in Patients with Recurrent Pericarditis





# Collaborative Field Force to Drive Awareness, Overcome Access Barriers and Help Ensure Positive Patient and Physician Experience



**PATIENT** 

**ACCESS** 

#### **CLINICAL SALES SPECIALISTS**

- Focus: ~2500 HCPs across ~800 accounts
- Responsibility: Physician accounts, disease education, Arcalyst promotion, account and territory plans, speaker program planning

### STRATEGIC ACCOUNTS

- Focus: ~350 payers and 5 Specialty Pharmacies
- Responsibility: Payer/specialty pharmacy relationship, strategic account planning, support sales team

### **MEDICAL SCIENCE LIAISONS**

- Focus: Subject Matter Experts and HCPs
- Responsibility: Disease awareness, data dissemination, advocacy development, account and payer support, speaker management

# KINIKSA ONECONNECT™

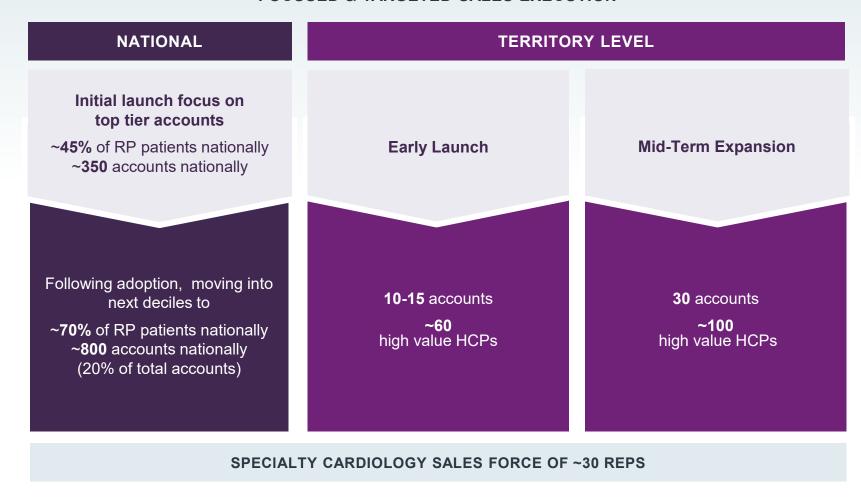
- Focus: Patients and caregivers, HCPs seeking reimbursement support for their patients
- **Responsibility:** Optimize patient and customer experience with Arcalyst and Kiniksa, provide seamless initiation, reimbursement, and adherence support



HCP = health care provider

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

### **FOCUSED & TARGETED SALES EXECUTION**



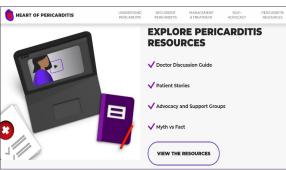


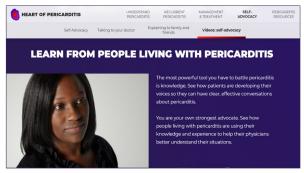
# Comprehensive Support for Patients Through Kiniksa OneConnect™

#### **DISEASE AWARENESS AND ARCALYST PROMOTION**









### PATIENT ADVOCACY SUPPORT







# THE PATIENT ACCESS LEADS PROVIDE ONE-ON-ONE SUPPORT, INCLUDING:

- Insurance coverage determination
- Explanation of benefits verification
- Assistance with prior authorizations and appeals
- Injection training support and education with ARCALYST Nurse Educators
- Identification of possible sources of financial assistance
- Help with ARCALYST shipment and delivery



# First Launch Quarter Resulted in the Successful Transition of Existing Patients and Strong Demand in Recurrent Pericarditis (RP)

#### **NET REVENUE**

\$7.7 million

#### **REVENUE DRIVERS**

- Q2 revenue relatively evenly split between RP, CAPS/DIRA, and initial channel inventory build
- Solid execution led to robust CAPS and DIRA patient continuation of therapy with demand at/near historical levels
- Q2 ending inventory weeks on hand was higher than is expected in subsequent quarters
- Strong RP demand is the primary growth driver with high conversion rate of RHAPSODY patients and new to brand patients

# **Q2 ARCALYST NET REVENUE**



KINIKSA IS EXPECTING Q3 ARCALYST REVENUE OF \$9.0-10.0M

Driven by robust anticipated growth in RP demand



# MAVRILIMUMAB

#### MONOCLONAL ANTIBODY INHIBITOR TARGETING GM-CSFRα

**DISEASE AREA:** COVID-19-related acute respiratory distress syndrome (ARDS); Giant Cell Arteritis (GCA): chronic inflammatory disease of medium-to-large arteries

**COMPETITION¹:** Therapeutic options for patients hospitalized with COVID-19-related ARDS are limited; 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; Data from Phase 3 trial in severe COVID-19-related ARDS expected in Q1 2022

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

**RIGHTS:** Worldwide

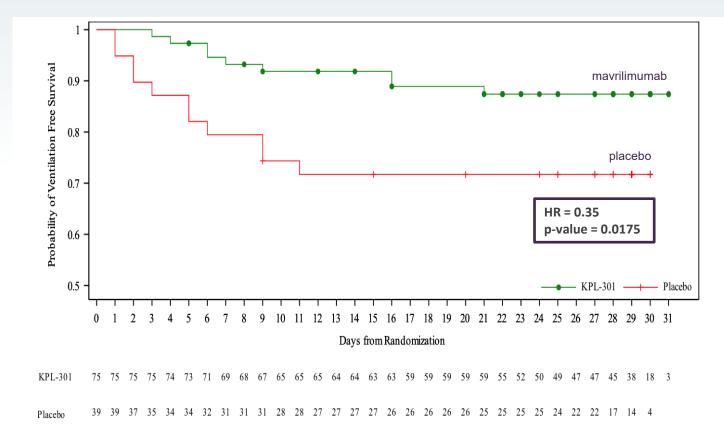


# Mavrilimumab Reduced the Risk of Mechanical Ventilation or Death by 65% Versus Placebo at Day 29 Pooled Across Dose Levels

# Phase 2 data from the Phase 2/3 trial of Mavrilimumab in COVID-19-related ARDS

# **Baseline Demographics & Characteristics:**

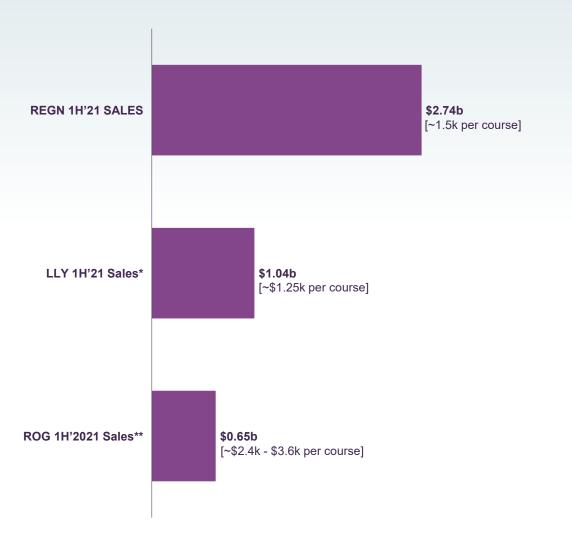
- 43% non-whites, 57% males enrolled
- 96% received corticosteroids/dexamethasone
- 29% received antivirals/remdesivir
- Randomized number of patients by country
  - Brazil (37.72%)
  - United States (31.58%)
  - South Africa (27.19%)
  - Peru (2.63%)
  - Chile (0.88%)



Note: Time to ventilation or death by Day 29 is defined as time (in days) from randomization to the date of death or start date of using mechanical ventilation (NIAID <= 2) by Day 29. All subjects who never had NIAID <= 2 by Day 29 will be censored at last assessment date of NIAID 8-point ordinal scale.



# **Significant Market Opportunity for COVID-19 Treatments**



"Regeneron's COVID-19 antibodies rake in \$2.6B, reflecting their market dominance and blowing away estimates"

Fierce Pharma 8/5/2021

"Eli Lilly Profit Rises, Helped by COVID-19 Drug Sales"

The Wall Street Journal 1/29/2021

"Roche warns of global Actemra shortage as delta variant drives huge spike in demand for COVID-19 patients"

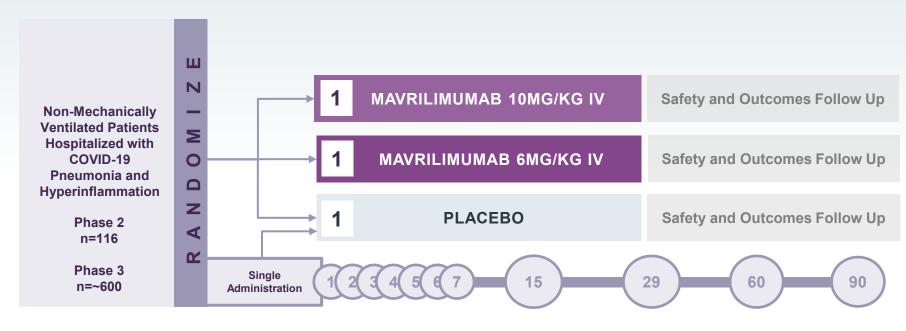
Fierce Pharma 8/17/2021



# Phase 3 Clinical Trial of Mavrilimumab in COVID-19-Related ARDS

#### **KEY INCLUSION CRITERIA**

- Positive COVID-19 test within 14 days prior to randomization
- Hospitalized for COVID-19
- Bilateral pneumonia on chest x-ray or computed tomography
- Active fever or recently documented fever within 72 hours prior to randomization
- Clinical laboratory results indicative of hyper-inflammation
- Non-ventilated; requiring supplemental oxygen to maintain oxygen saturation (SpO2) ≥ 92% and not-intubated
- All patients should receive best standard of care, including steroids and antivirals, according to investigator judgement



STUDY FOLLOW UP (DAYS)

#### PRIMARY EFFICACY ENDPOINT:

Proportion of patients alive and without mechanical ventilation at Day 29

# **SECONDARY EFFICACY ENDPOINTS:**

- Mortality rate at Day 29
- Ventilation-free survival (time to ventilation or death) by Day 29
- Overall survival by Day 29

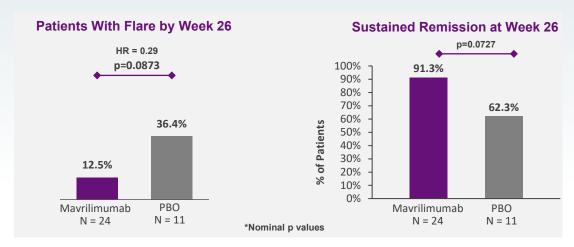


ARDS = acute respiratory distress syndrome

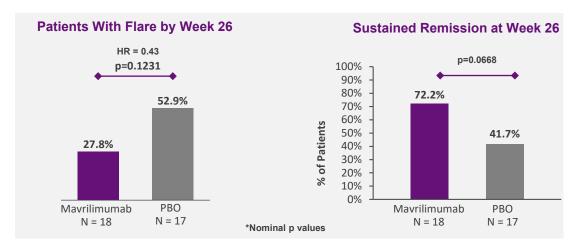
# **Unmet Need and Commercial Opportunity for Safe and Effective GCA Therapies**

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

#### **NEW-ONSET GCA**



### RELAPSING/REFRACTORY GCA



### **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 one year of starting treatment with approved biologics<sup>3</sup>
- Mechanistic (GM-CSFRα vs IL-6) and administrative (Q2WK vs QWK) differentiation
- Well-tolerated safety profile particularly important given large elderly patient population



# **VIXARELIMAB**

#### MONOCLONAL ANTIBODY INHIBITOR TARGETING OSMRB

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 a Phase 2b clinical trial, evaluating a range of once-monthly dose regimens

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

**RIGHTS:** Worldwide



# **Dual Mechanism Offers Potential Pruritus Relief and Nodule Improvement**

Vixarelimab Phase 2a prurigo nodularis data

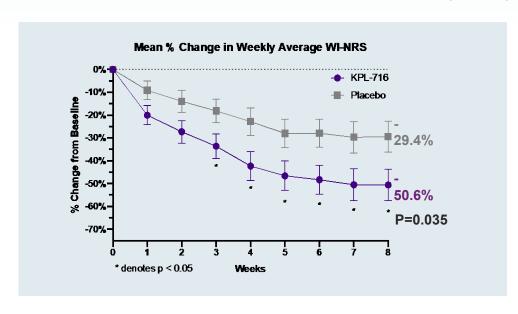
# Vixarelimab is the only mAb targeting OSMRβ, which mediates signaling of key cytokines (IL-31 & OSM)

#### 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).





Representative Treatment Response



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

Enrollment and dosing of patients commenced in Q4 2020

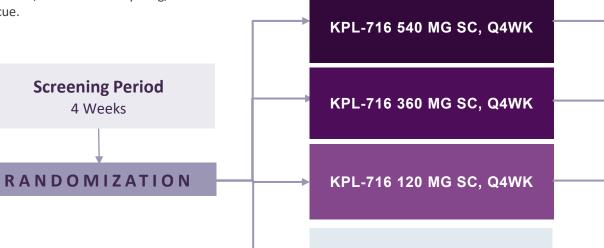
**Screening Period** 

4 Weeks

#### **EXPECTED TO ENROLL APPROX. 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.

# **Double-Blind Period** 16 Weeks



PLACEBO SC Q4WK

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

**Open-Label Extension Period** 36 Weeks



# **KPL-404**

#### MONOCLONAL ANTIBODY INHIBITOR INTERACTION BETWEEN CD40 AND CD40L

**DISEASE AREA:** Rheumatoid Arthritis; a chronic inflammatory disorder affecting many joints; External proof-of-concept previously established in broad range of autoimmune diseases: Sjogren's disease, systemic lupus, 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:** Phase 1 single-ascending-dose study in healthy volunteers completed and supports further development in patients with optionality for testing SC and/or IV dosing; Expect to initiate Phase 2 proof-of concept trial in patients in Q4 2021

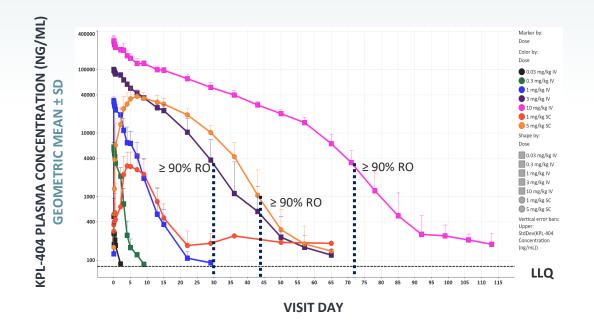
**ECONOMICS:** Clinical and regulatory milestones and royalty on annual net sales

**RIGHTS:** Worldwide



# Final Data from KPL-404 Single-Ascending-Dose Phase 1 Study

PK profiles for KPL-404 & T-Cell Dependent Antibody Response (TDAR) for KLH antigen challenge

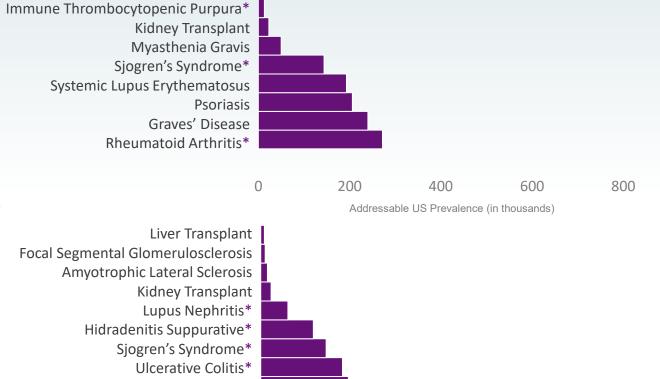






# Potential for Evaluation of KPL-404 in a Broad Range of Autoimmune Diseases

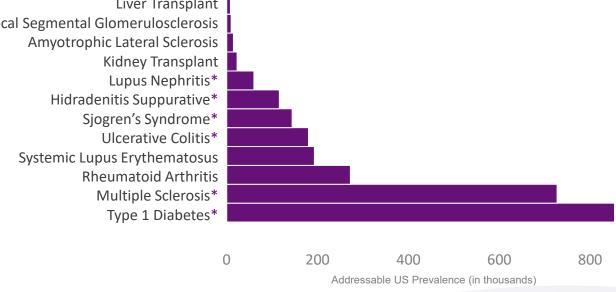
**INDICATIONS WITH** PUBLISHED DATA<sup>1</sup>



### **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>





<sup>\*</sup>Indications evaluated with subcutaneous administration 1) With the CD40 mechanism

# **Building Value at Kiniksa**

**Corporate Priorities** 

# **ARCALYST**

Commercial launch in recurrent pericarditis (April 2021)

# **MAVRILIMUMAB**

Phase 3 COVID-19-related ARDS data expected Q1 2022

# **VIXARELIMAB**

Phase 2b study in prurigo nodularis evaluating a range of once-monthly dose regimens

# **KPL-404**

Final Phase 1 data (May 2021); plan to initiate Phase 2 proof-of-concept trial in rheumatoid arthritis in Q4 2021

CASH, CASH EQUIVALENTS AND SHORT-TERM INVESMENTS EXPECTED TO FUND OUR CURRENT OPERATING PLAN INTO 2023





# **Corporate Presentation**

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