



JP Morgan Conference

JANUARY 2023

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 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; expected run rate for our cash, cash equivalents and short-term investments; expected funding of our operating plan; financial guidance; third-party collaborations and licensing; 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 realize value from our licensing and collaboration arrangements; our ability to source sufficient drug product, as needed, to meet our clinical and commercial requirements; our ability 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 any of our product candidates or to require additional data or trials to support any such approval or authorization; delays, difficulty or inability to successfully execute on our commercial strategy for ARCALYST; potential changes in our strategy, clinical trial priority, operating plan and funding requirements; raw materials, important ancillary product and drug substance and/or drug product shortages; substantial new or existing competition; potential impact of the COVID-19 pandemic or any subsequent pandemic, and measures taken in response to such pandemics, 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; risks related to the ongoing war in Ukraine; risks arising from global political and economic instability; 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.



Building a Generational Company by Acquiring, Developing and Commercializing Assets



Business development is a key part of our strategy, and we continue to focus on augmenting or rationalizing our portfolio



Consistent execution

Portfolio of Immune-Modulating Assets

Cardiovascular Franchise

PROGRAM & TARGET	PRECLINICAL	PHASE 1	PHASE 2	PHASE 3	COMMERCIAL	KINIKA RIGHTS
ARCALYST® (rilonacept)^{1,2} IL-1α & IL-1β	RECURRENT PERICARDITIS					Worldwide⁴ (Excluding MENA)
Mavrimumab³ GM-CSFRα	EVALUATING DEVELOPMENT IN RARE CARDIOVASCULAR DISEASES					Worldwide⁴

Autoimmune Franchise

KPL-404 CD40/CD154	RHEUMATOID ARTHRITIS					Worldwide
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Vixarelimab
OSMRβ

Roche and Genentech

GLOBAL RIGHTS FOR ALL INDICATIONS⁵



1) Approved in the U.S.; ARCALYST is also approved for cryopyrin-associated periodic syndromes (CAPS) and deficiency of the interleukin-1 receptor antagonist (DIRA); 2) The FDA granted Breakthrough Therapy designation to ARCALYST for recurrent pericarditis in 2019; the FDA granted Orphan Drug exclusivity to ARCALYST in March 2021 for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and pediatric patients 12 years and older. The European Commission granted Orphan Drug designation to ARCALYST for the treatment of idiopathic pericarditis in 2020; 3) Phase 2 clinical trials of mavrilimumab in rheumatoid arthritis and giant cell arteritis achieved their primary and secondary endpoints with statistical significance; 4) Kiniksa granted Huadong Medicine exclusive rights in the Asia Pacific Region, excluding Japan; 5) In September 2022, Kiniksa granted Genentech and Roche exclusive global rights to develop and commercialize vixarelimab; 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

ARCALYST is Shaping the Treatment Paradigm of Recurrent Pericarditis, Leading to Launch Success

- Riloncept proved to be highly effective in reducing the risk of recurrent pericarditis in the pivotal Phase 3 study, RHAPSODY¹
- Long-term extension data presented at the American Heart Association (AHA) Scientific Sessions 2022; showed treatment with riloncept for longer than 18 months resulted in a continued treatment response²
- ARCALYST continues to shift the treatment paradigm for recurrent pericarditis



1) Klein AL, Imazio M, Cremer P, et al. Phase 3 trial of interleukin-1 trap riloncept in recurrent pericarditis. N Engl J Med. 2021;384(1):31-4; 2) Imazio M, Klein AL, et al. Prolonged Riloncept Treatment in Rhapsody Long-term Extension Provided Persistent Reduction of Pericarditis Recurrence Risk. Poster 2223 (Presented at AHA Scientific Sessions 2022)

Steady Growth into 14,000 Multiple Recurrent Addressable Population

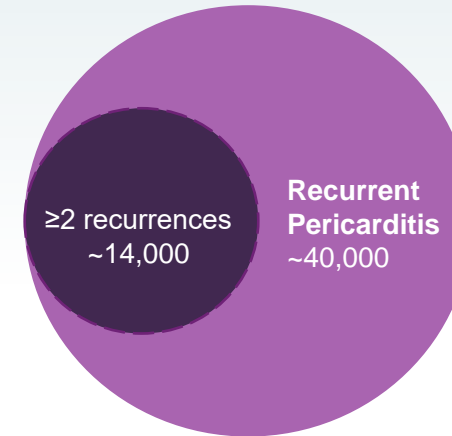
Opportunity in and Beyond our Target Population

- 14,000 target patients who suffer from 2 or more recurrent pericarditis events every year
- 26,000 patients who are on their first recurrence; given the broad label, already seeing some physicians prescribe for those patients

Field Force Expansion

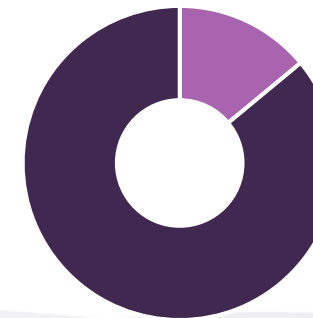
- Frequent call activity has been a major driver of prescriber understanding of the disease burden and ARCALYST adoption
- Expanded team of ~50 reps (from ~30 reps) enables greater call frequency with our higher decile doctors as well as further broadens our reach, allowing us to help even more recurrent pericarditis patients
- Foundation for future commercial growth

14,000 TARGET PATIENTS WITH UPSIDE IN FIRST RECURRENCE POOL



All figures annual period prevalence

RECURRENCES PRIOR TO ARCALYST INITIATION



■ 1 Recurrence ■ 2+ Recurrences

Continued Execution Resulting in Steady ARCALYST Growth

\$39.9M

Net revenue for Q4 2022 (unaudited); representing ~20% sequential growth vs Q3 2022

\$122.5M

*Full-year 2022 net revenue (unaudited)
2022 full-year guidance: **\$115-130M***

~5%

Of the target 14,000 multiple-recurrent pericarditis patients were actively on ARCALYST treatment as of the end of Q4 2022

Key Drivers of ARCALYST Revenue Growth in Q4 2022

Physician Growth

- Since launch >800 unique prescribers have written ARCALYST for recurrent pericarditis
- 22% of the total prescribing base have written for 2 or more patients

Payer Access

- In Q4, greater than 90% approval rate of completed cases

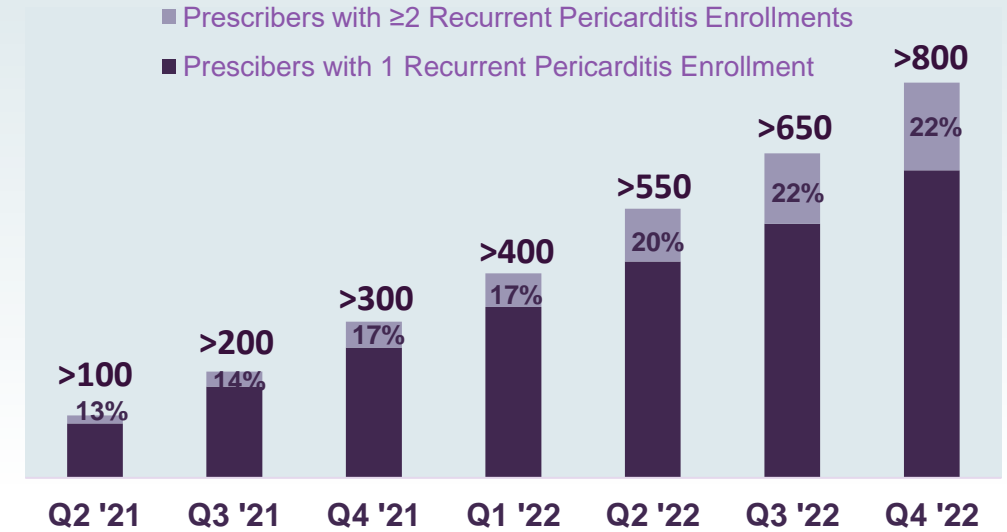
Duration of Therapy

- Long-term extension data shown at AHA 2022 further supports that patients need to be treated throughout the course of their disease and that continued treatment results in continued treatment response
- Commercial setting: duration of initial therapy was, on average, approximately 12 months¹

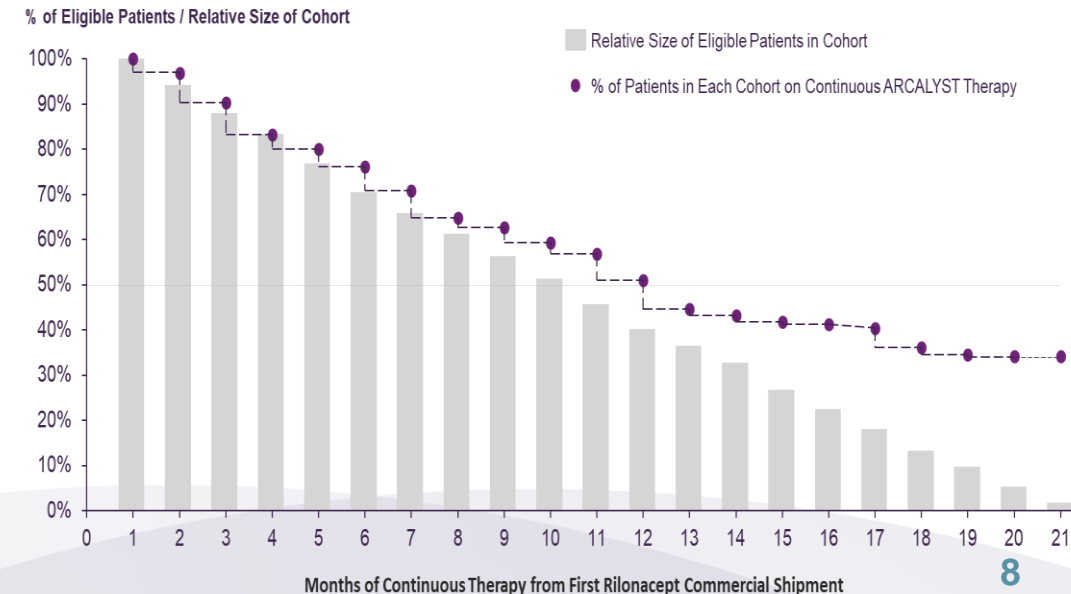
Patient Restarts

- Of those patients that stopped initial therapy, around 45% went back on ARCALYST, and the majority of those restarted within 8 weeks

Total and Repeat Prescriber Growth per Quarter



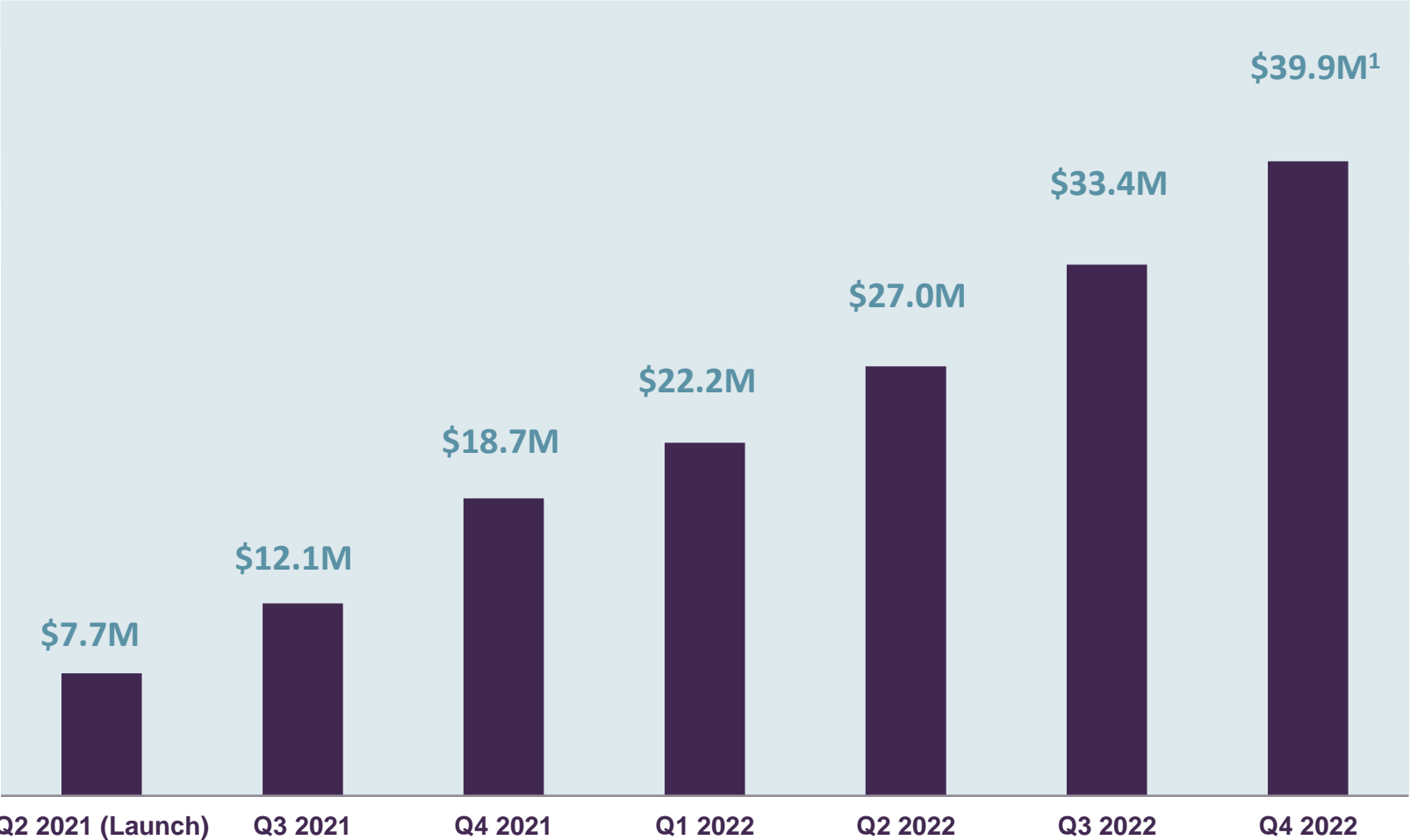
Duration of Continuous Initial Therapy (not including restarts)^{1,2}



1) Initial continuous therapy is determined to have ended if greater than 28 days elapses beyond the exhaustion date of a patient's most recent days supplied without an observed refill of ARCALYST; 2) Patients restarting after an initial therapy lapse as of 12/8/2022 (patient restarts are not included in the chart)

Robust Commercial Execution Led to Steady Sequential Growth

ARCALYST Net Revenue of \$161.0M Since Recurrent Pericarditis Launch



>100% increase in 2022 net product revenue versus our first 12 months on the market (unaudited)

Plan to provide full-year 2023 net revenue guidance with Q4 2022 financial results

Compliance and quality are Kiniksa's core strengths



1) Unaudited

KPL-404: Potentially Best-in-Class, Subcutaneously Delivered Monoclonal Antibody Inhibitor of the CD40/CD154 Interaction

KPL-404 is a humanized, IgG4 monoclonal antibody that binds to and antagonizes signaling through CD40 (high concentration liquid formulation)

CD40 antagonism has been clinically validated as a key regulator of cellular and humoral adaptive immunity across multiple disease states

KPL-404 has been well-tolerated to date, avoiding liabilities with previous generations of pathway antagonists

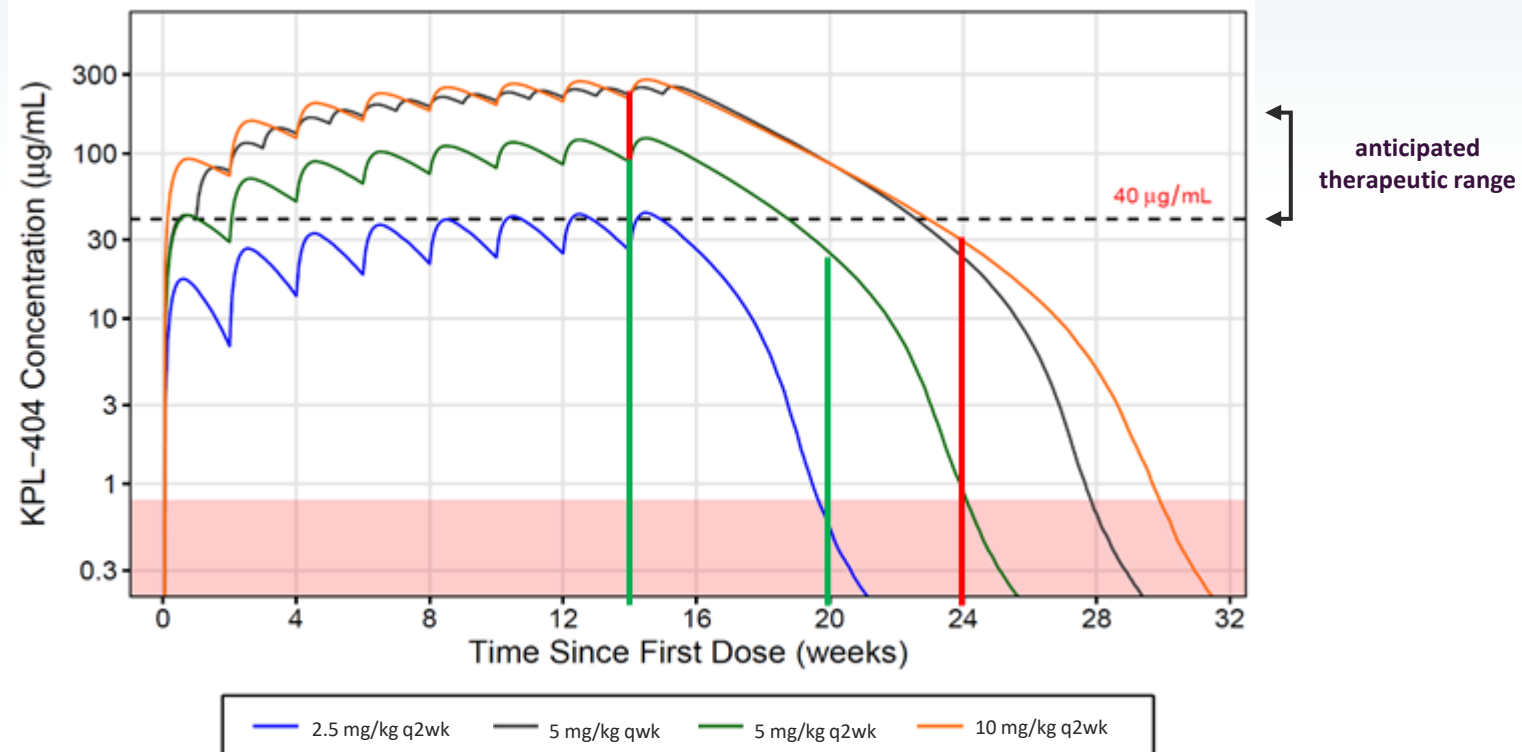
KPL-404 drug product is formulated in a high concentration liquid formulation that enables subcutaneous-administration

KPL-404 non-clinical and clinical data generated to date suggest it is well positioned against competitors

- In-licensed KPL-404 in 2019 as a pre-clinical stage asset through acquisition of Primatope Therapeutics
- Quickly took asset into Phase 1; data support testing of longer-term subcutaneous administration in patients with autoimmune disease
- KPL-404 now in multiple-ascending-dose Phase 2 study
- Kiniksa owns the vast majority of the economics for KPL-404

PK-Modeling and Dose Simulations for KPL-404 Dosing in Phase 2

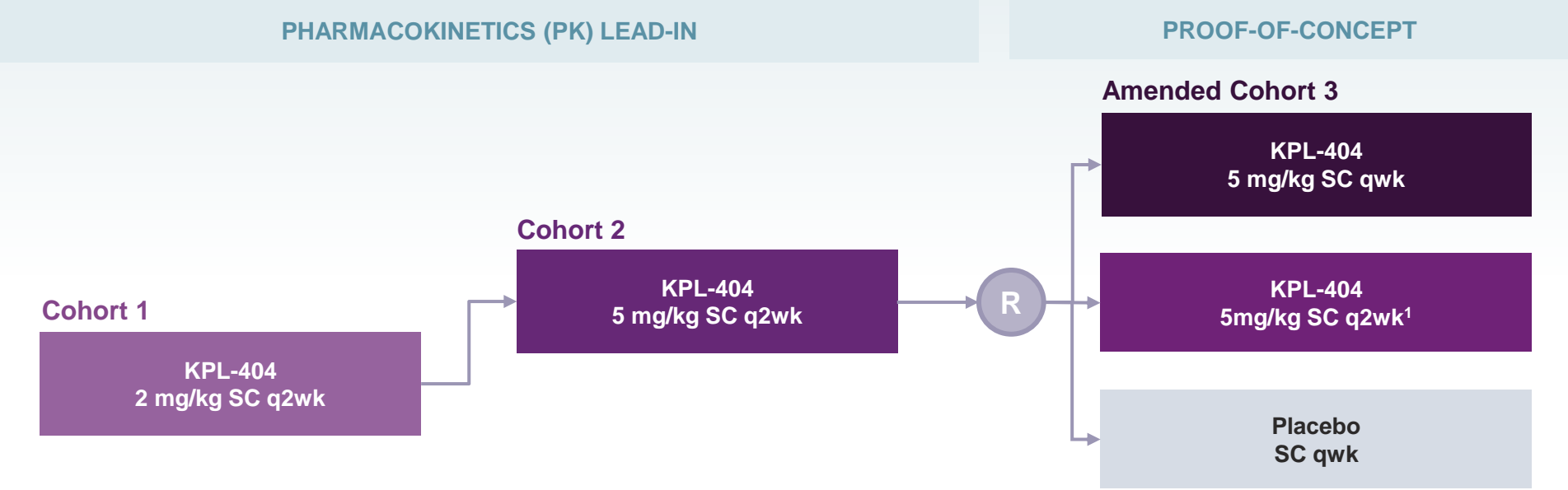
Pharmacokinetic Simulations



- High concentration liquid formulation translates into potential ability to deliver large amounts of drug through subcutaneous dosing
- The PK/PD data show potential to reach plasma concentrations we believe necessary to see efficacy in the clinic
- KPL-404 has the potential to be a best-in-class therapeutic

KPL-404 Phase 2 Trial in Rheumatoid Arthritis

Multiple-ascending-dose study that evaluates PK and safety and then transitions into a parallel dose efficacy portion



PATIENT POPULATION:

- Active RA who have an inadequate response to or are intolerant to a Janus kinase inhibitor (JAKi) or at least one biologic disease-modifying anti-rheumatic drug (bDMARD). Subjects who have failed both bDMARD and JAKi are excluded from the study.

DISEASE CRITERIA:

- Six or more swollen joints and ≥ 6 tender joints at screening and baseline line visits; levels of high sensitivity C-reactive protein ≥ 5 mg/L; seropositivity for serum RF and/or ACPA at screening.

COHORTS 1-2 (PK Lead-In)

- Each cohort will sequentially randomize 8 patients
- Primary Endpoints:
 - Incidence of treatment-emergent adverse events (TEAEs)
 - Pharmacokinetics (C_{max}, AUC_(0-t))
- Secondary Endpoint:
 - Change from baseline in DAS28-CRP at Week 12

AMENDED COHORT 3 (Proof of Concept)

- Cohort 3 will randomize up to 75 patients
- Primary Endpoint:
 - Change from baseline in DAS28-CRP at Week 12
- Secondary Endpoints :
 - Incidence of treatment-emergent adverse events (TEAEs)
 - Pharmacokinetics (C_{max}, AUC_(0-t))

Objectives: Evaluate safety, efficacy, and PD compared with placebo across the estimated therapeutic range and to characterize PK across varying dose levels of KPL-404

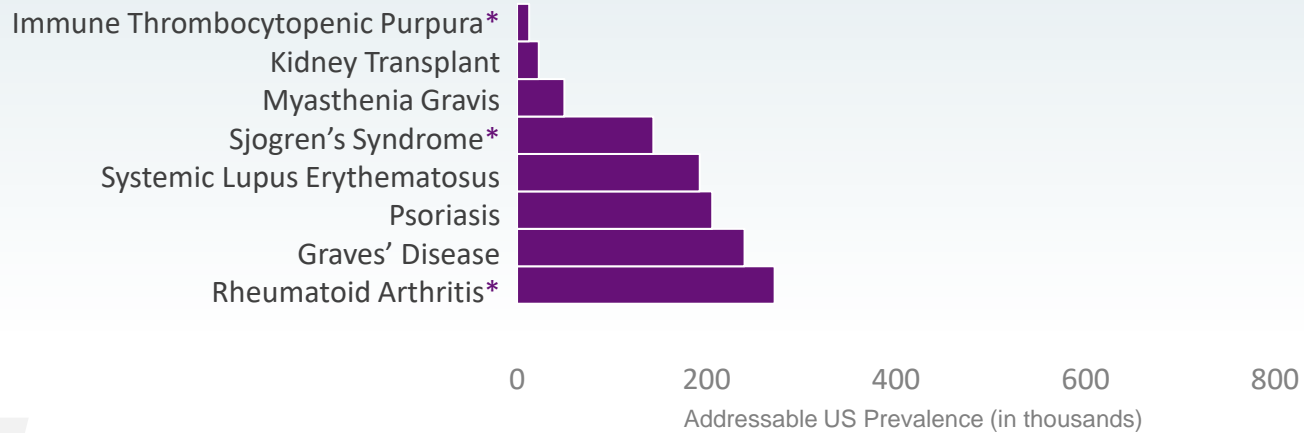


1) The 5 mg/kg SC q2wk group will receive weekly administrations of alternating active investigational product and matching blinded placebo
 SC = subcutaneous; q2wk = every other week; AUC = Area Under the Curve; RF = Rheumatoid Factor; ACPA = anti-citrullinated protein antibodies, PD = Pharmacodynamics;
 PK = Pharmacokinetics; R = Randomization

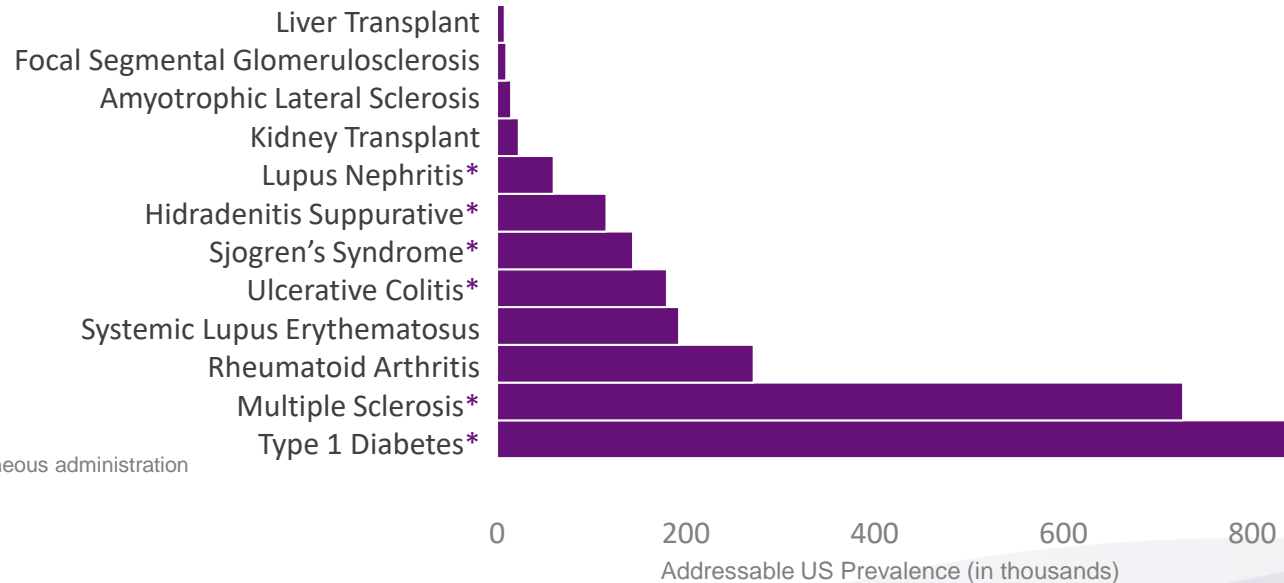
Potential for Evaluation of KPL-404 in a Broad Range of Autoimmune Diseases

CD40/CD154 interaction has been implicated in a number of devastating diseases

Indications with Published Data



Indications with Pending Data & Trials Ongoing



*Indications evaluated with subcutaneous administration

INDICATION SELECTION CRITERIA

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

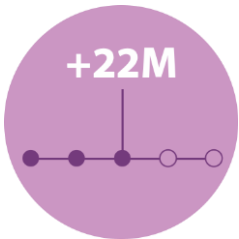


Sources: 2019 numbers: <https://unos.org/data/transplant-trends/>; Hunter et al. Prevalence of rheumatoid arthritis in the United States adult population in healthcare claims databases, 2004-2014; Rheumatol Int. 2017 Sep;37(9):1551-1557; Overall Prevalence: Maciel et al, Arthritis Care Res (Hoboken) 2017; Qin et al, Ann Rheum Dis 2015; UpToDate; Baldini et al. Prevalence of Severe Extra-Glandular Manifestations in a Large Cohort of Patients with Primary Sjogren's Syndrome; 2012 ACR/ARHP Annual Meeting, ABSTRACT NUMBER: 2185; Wallin et al. The prevalence of MS in the United States A population-based estimate using health claims data. Neurology, March 5, 2019; Somers et al.; Prevalence of Systemic Lupus Erythematosus in the United States: Preliminary Estimates from a Meta-Analysis of the Centers for Disease Control and Prevention Lupus Registries; 2019 ACR/ARP Annual Meeting ABSTRACT NUMBER: 2886; Garg et al. JAMA Dermatol. 2017;153(8):760-764. doi:10.1001/jamadermatol.2017.0201 Sex- and Age-Adjusted Population Analysis of Prevalence Estimates for Hidradenitis Suppurativa in the United States; MayoClinic.org; Yale J Biol Med. 2013 Jun; 86(2): 255-260. N Engl J Med 2016;375:2570-81; <https://www.diabetesresearch.org/diabetes-statistics>; Nephcare.org; Kitiyakara C, Eggers P, Kopp JB. Twenty-one-year trend in ESRD due to focal segmental glomerulosclerosis in the United States. Am J Kidney Dis. 2004 Nov;44(5):815-25; Rachakonda et al. J Am Acad Dermatol. 2014 Mar;70(3):512-6. doi: 10.1016/j.jaad.2013.11.013. Epub 2014 Jan 2. Psoriasis prevalence among adults in the United States; Yeung et al. Psoriasis severity and the prevalence of major medical co-morbidities: a population-based study; JAMA Dermatol. 2013 Oct 1; 149(10): 1173-1179; Hoover et al. Kidney Int. 2016 Sep; 90(3): 487-492. Insights into the Epidemiology and Management of Lupus Nephritis from the U.S. Rheumatologist's Perspective.

Partnership with Huadong Medicine Gives Kiniksa Opportunity to Expand Footprint into Asia Pacific Region (Excluding Japan)



In February 2022, Kiniksa announced a strategic collaboration with Huadong to develop and commercialize **ARCALYST** and **mavrilimumab** in Greater China, South Korea, Australia and 18 other countries, excluding Japan



Kiniksa received a \$22M upfront payment and is eligible to receive up to approximately \$640M in specified development, regulatory and sales-based milestone along with tiered royalty payments

- Huadong has filed a Biologic License Application for CAPS in mainland China and if approved, Kiniksa would receive additional non-dilutive capital.



Collaboration provided non-dilutive capital, cost-sharing, and additional resources to help accelerate development and commercialization efforts

License Agreement with Roche Genentech for Global Rights to Develop and Commercialize Vixarelimab

Kiniksa to receive \$100 million in upfront and near-term payments:

- \$80 million, which was received following the transaction's closing
- \$20 million, which Kiniksa is eligible to receive following Kiniksa's delivery of certain drug supplies to Genentech



Kiniksa is eligible to receive up to approximately \$600 million in certain clinical, regulatory, and sales-based milestones, before fulfilling upstream financial obligations

Kiniksa is also eligible to receive royalties on annual net sales ranging from low-double digits to mid-teens, before fulfilling upstream financial obligations



\$100 million in non-dilutive proceeds from the transaction to help grow cardiovascular franchise and build autoimmune franchise

Executed Across Commercial and Clinical-Stage Portfolio in 2022

Setting the stage for continued success in 2023 and beyond

2022 – Consistent Execution

- ✓ Emerging leader in immune-modulating therapies
- ✓ Strong commercial launch of ARCALYST in recurrent pericarditis
- ✓ Multiple ascending dose portion (Cohort 1 & 2) of KPL-404 Phase 2 trial in rheumatoid arthritis enrolled
- ✓ License agreement with Genentech for global rights to vixarelimab
- ✓ Strategic collaboration with Huadong Medicine for Asia Pacific Region

2023

ARCALYST revenue growth opportunity by continuing to broaden reach with recurrent pericarditis physicians and patients through focus on high decile physicians

Enroll patients in proof-of-concept portion (Cohort 3) of Phase 2 trial of KPL-404 in rheumatoid arthritis

Continue to evaluate potential value-creating business development opportunities through in-licensing and out-licensing

Remain well capitalized

- **Year end 2022 \$190.4M cash reserves expected to fund current operating plan into at least 2025¹**

Continue to help patients and build value



1) As used herein the term, "Cash Reserves" means our cash, cash equivalents and short-term investments (unaudited) as of December 31, 2022



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