# Third Quarter 2022 Financial Results and Corporate Update

NOVEMBER 1, 2022

# Agenda

- **Introduction** | *Sanj K. Patel, Chief Executive Officer*
- **Clinical Update** | *John F. Paolini, Chief Medical Officer*
- **ARCALYST® Commercial Execution** | *Ross Moat, Chief Commercial Officer*
- **Third Quarter 2022 Financial Results** | *Mark Ragosa, Chief Financial Officer*
- **Closing Remarks** | *Sanj K. Patel, Chief Executive Officer*
- **Q&A Session**

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# 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 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; 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 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.



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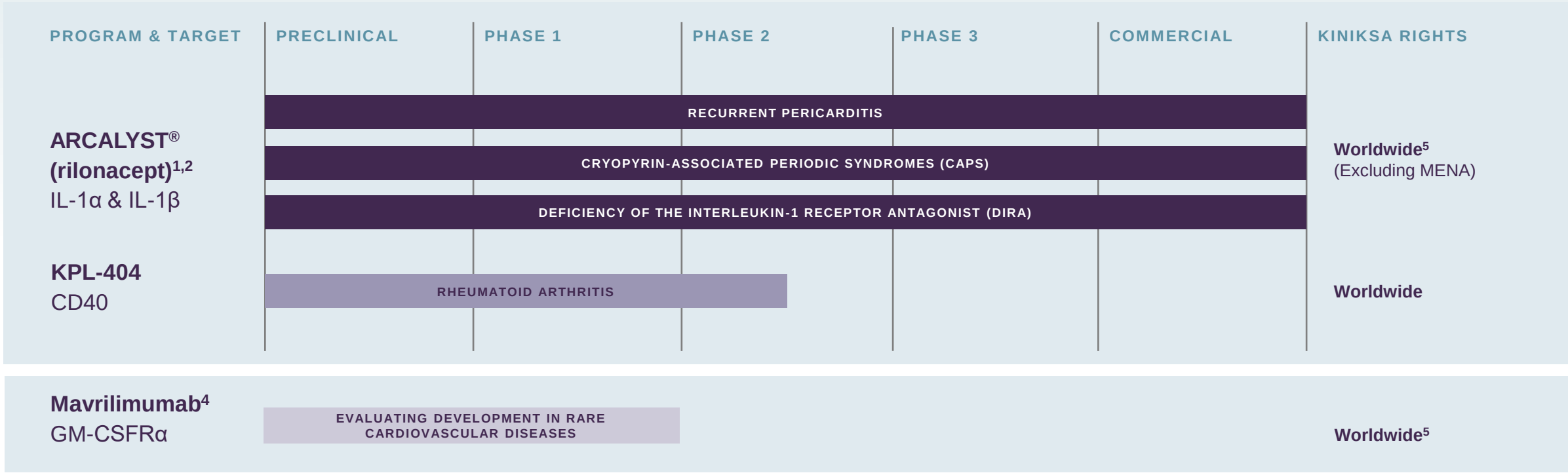
# Introduction

Sanj K. Patel
Chief Executive Officer

# Portfolio of Immune-Modulating Assets




1) Approved in the U.S; 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) In September 2022, Kiniksa granted Genentech and Roche exclusive global rights to develop and commercialize vixarelimab; 4) Phase 2 clinical trials of mavrilimumab in rheumatoid arthritis and giant cell arteritis achieved their primary and secondary endpoints with statistical significance; 5) Kiniksa granted Huadong Medicine exclusive rights in the Asia Pacific Region, excluding Japan; 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

# Clinical Update

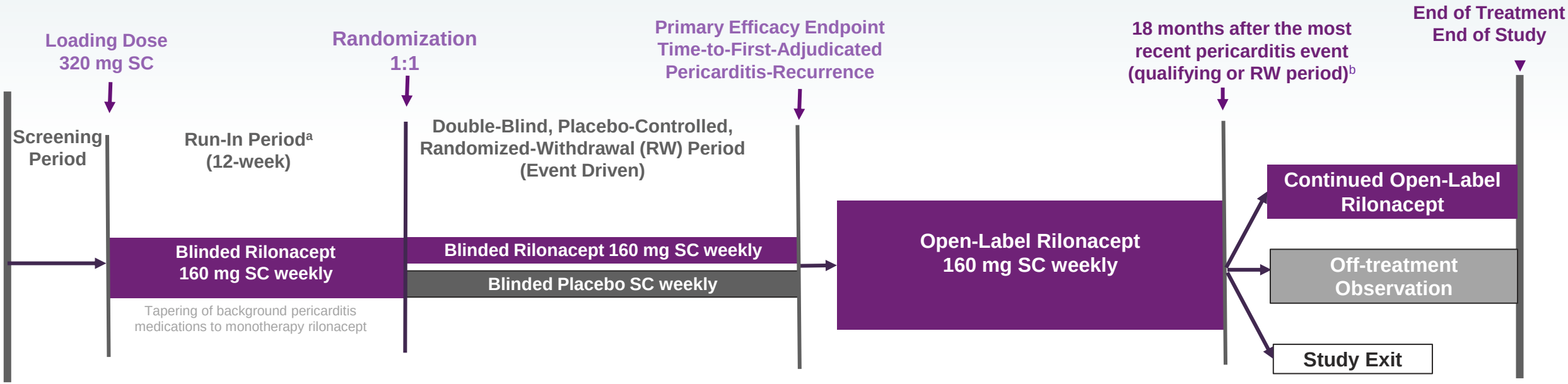
John F. Paolini
Chief Medical Officer

# RHAPSODY Design

## Event-Driven Pivotal Study

*Median rilonacept treatment duration prior to the LTE (RI+RW) was 9 months (range, 3-14)*

## Long-Term Extension (LTE) (up to 24 months)




[a] The duration of the run-in period was concealed from patients, so that they were blinded to the timing of randomization

[b] For each patient in the LTE**, a decision was made 18 months after the most recent pericarditis recurrence (Qualifying or RW period)** based on clinical status and one of the following actions was taken at the investigator's discretion:

- Continue rilonacept on-study

*OR*

- Suspend rilonacept treatment and remain on-study for observation (rilonacept rescue for recurrence allowed)

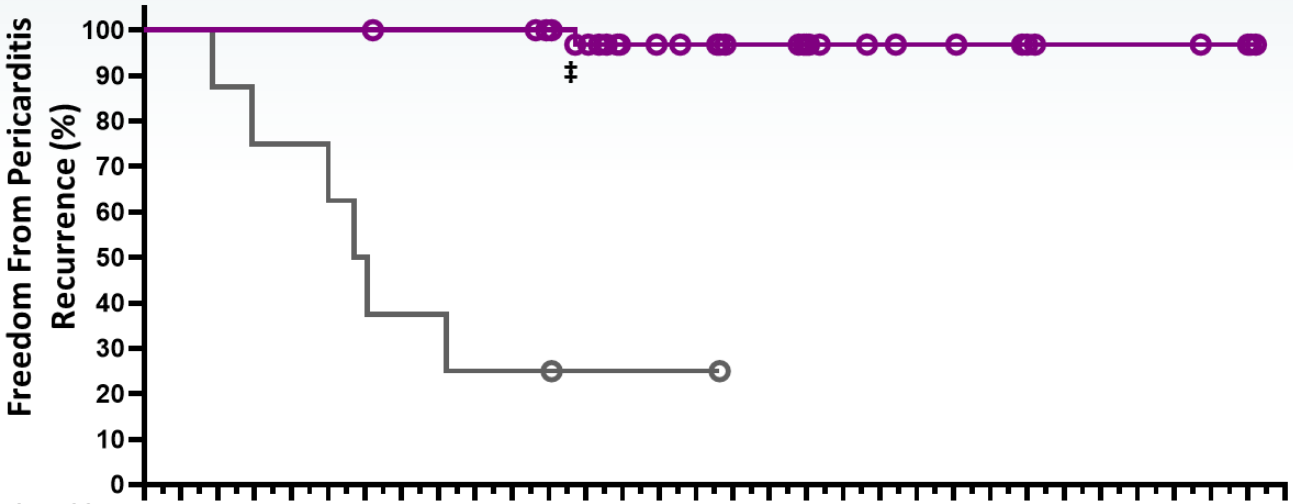
*OR*

- Discontinue the LTE completely (no further observation)

Adapted from: Klein AL, Imazio M, Brucato A, et al. Am Heart J 2020; 228: 81-90 and Klein AL, Imazio M, Cremer P, et al. N Engl J Med 2021; 384:31-41
DOI: 10.1056/NEJMoa2027892

# RHAPSODY Long-Term Extension Data Demonstrated Rilonacept Treatment Beyond 18 months Resulted in Continued Treatment Response[1,2]

**Figure 1. Continued rilonacept treatment beyond 18 months resulted in continued treatment response**




‡ The subject with a recurrence at 23.4 weeks had interrupted rilonacept treatment ~4 weeks prior.

**Hazard Ratio = 0.018**
**Log-rank p-value < 0.0001**
**Risk reduction = 98.2%**

Continued Rilonacept Treatment

Off-treatment for Observation

| | N | Number of Subjects with Recurrence[a] n (%) | Number of Weeks to Recurrence[a] Median (95% CI) |
|---|---|---|---|
| Continued Rilonacept Treatment | 33 | 1 (3) | NE (NE, NE) |
| Off-Treatment for Observation | 8 | 6 (75) | 11.8 (3.7, NE) |

[a]After 18 month decision
NE, not estimable

**Excerpted from Imazio M, Klein AL, et al. Prolonged Rilonacept Treatment in Rhapsody Long-term Extension Provided Persistent Reduction of Pericarditis Recurrence Risk. Circulation 2022; 146 (Suppl_1):A11653 (AHA Scientific Sessions 2022 Published Abstracts)**

1) An abstract containing data from the long-term extension (LTE) of RHAPSODY, the pivotal Phase 3 clinical trial of rilonacept in recurrent pericarditis, was accepted for presentation at the American Heart Association Scientific Sessions 2022 and was published on October 31, 2022. Dr. Massimo Imazio of University Hospital Santa Maria della Misericordia, Udine, will present a poster presentation entitled *Prolonged Rilonacept Treatment in RHAPSODY Long-Term Extension Provided Persistent Reduction of Pericarditis Recurrence Risk* on Sunday, November 6, 2022, from 3:45 to 4:45 p.m. Central Time (4:45 – 5:45 p.m. Eastern Time); 2) Data included in the abstract are as of the abstract submission cutoff date; final data are under embargo until the poster presentation.

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# KPL-404 Phase 2 Trial in Rheumatoid Arthritis

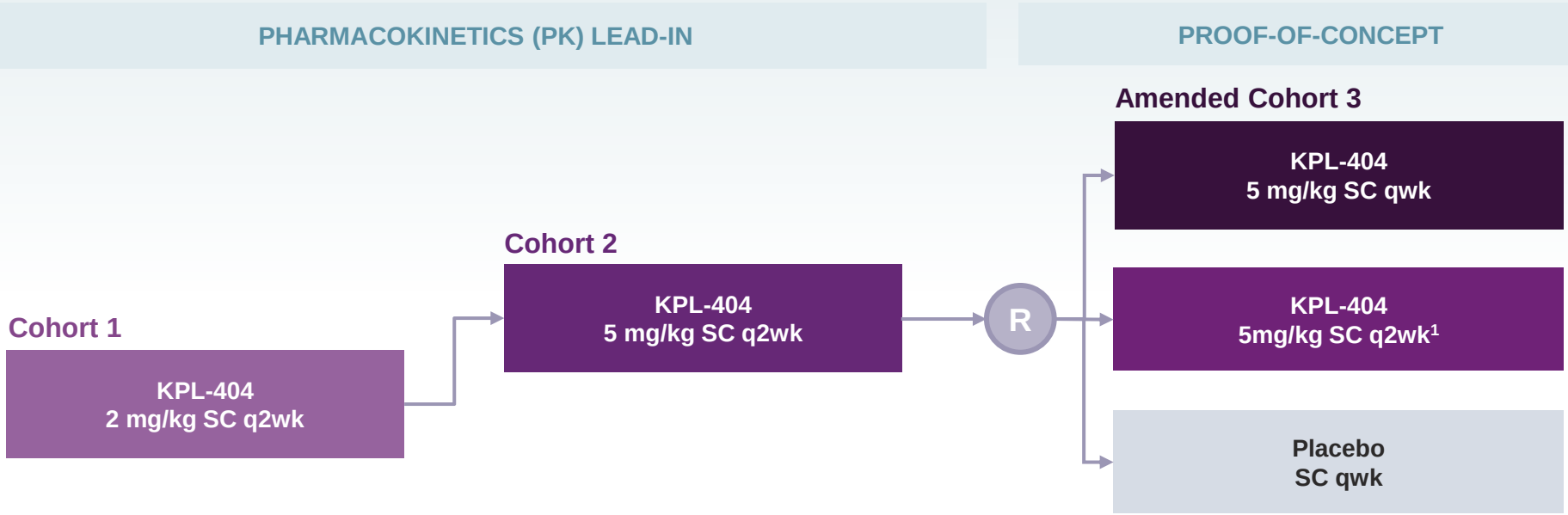
## Designed to provide PK data and early signal of efficacy with chronic (12-week) administration and optionality to evaluate KPL-404 across a range of other autoimmune diseases

**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 each 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; SRC = Safety Review Committee

# ARCALYST Commercial Execution

Ross Moat
Chief Commercial Officer

# Continued Execution Resulting in Steady ARCALYST Growth in Q3 2022; On-Track for Annual Guidance of $115-130M Net Revenue

## Net Revenue

- ARCALYST net revenue for Q3 2022 was $33.4 million
- Represents ~24%, or $6.4M growth versus Q2 2022
- Continued collaboration profitability for the ARCALYST franchise

## Revenue Drivers

- Recurrent Pericarditis continues as the primary growth lever, driven by new patient initiations
- CAPS and DIRA patient demand remained stable and broadly consistent with the previous quarters.
- Growth rate represents continued uptake and adoption of ARCALYST from prescribers, payers and patients in this previously unmet and debilitating autoinflammatory cardiovascular disease.

*$33.4 million in net revenue for the **third quarter of 2022***

*$82.6 million net revenue **year to date***

***Maintaining 2022 guidance** of $115-130 million net revenue*

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CAPS = cryopyrin-associated periodic syndromes; DIRA = deficiency of IL-1 receptor antagonist

# Drivers of Continued ARCALYST Growth in Recurrent Pericarditis

## Physician Growth

- Steady growth of prescriber base with greater than 650 HCPs having prescribed ARCALYST since launch
- Growing repeat prescriber base with 22% having prescribed ARCALYST for 2 or more patients

## Payer Access

- In Q3, payers continued to recognize the value of the first and only approved drug for recurrent pericarditis, with greater than 90% approval rate of completed cases

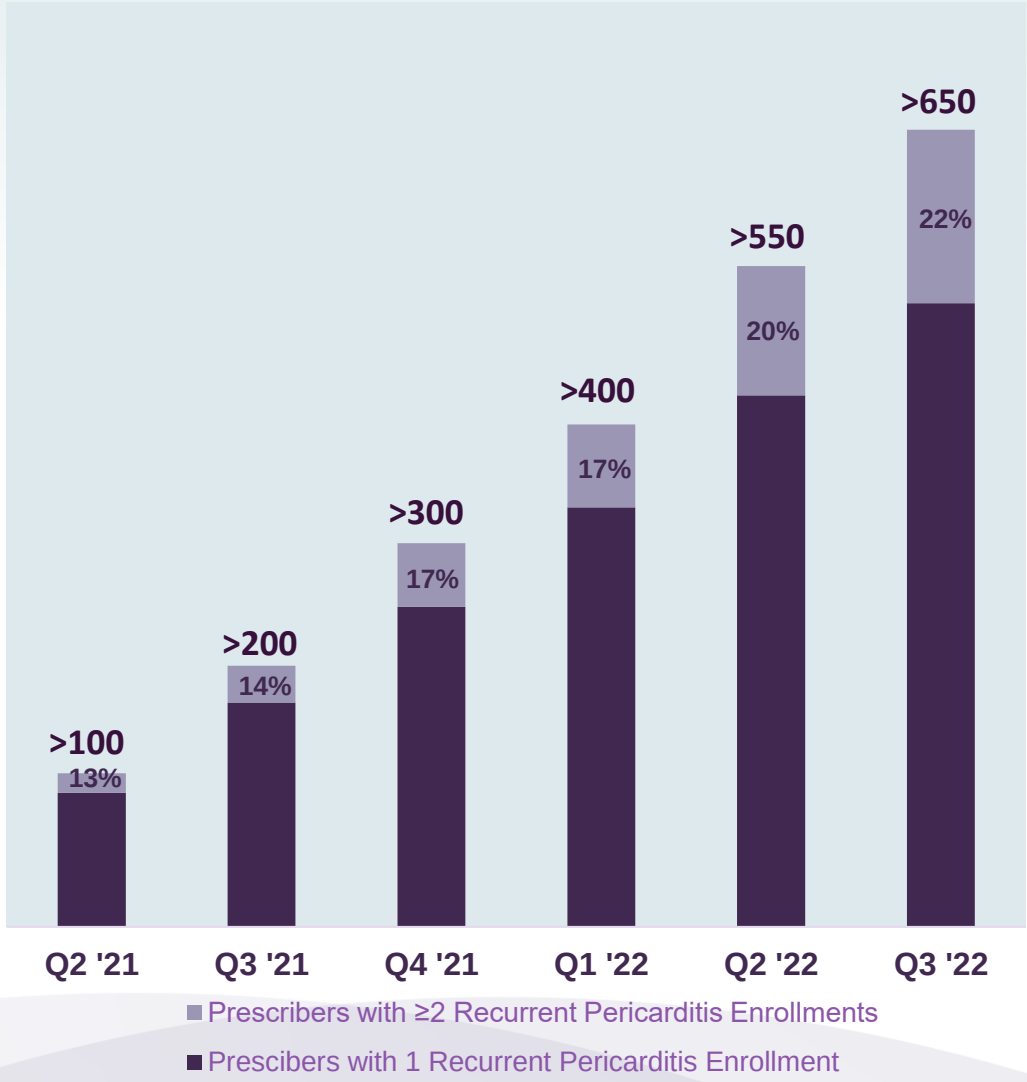
## Duration of Therapy

- As of the end of Q3, average duration of initial therapy in the commercial setting was approximately 12 months[1]

## Patient Restarts

- ~35% of all patients who stop treatment of ARCALYST restart therapy, indicating the need for longer duration of therapy to cover the natural history of the disease

### Total and Repeat Prescriber Growth per Quarter




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.

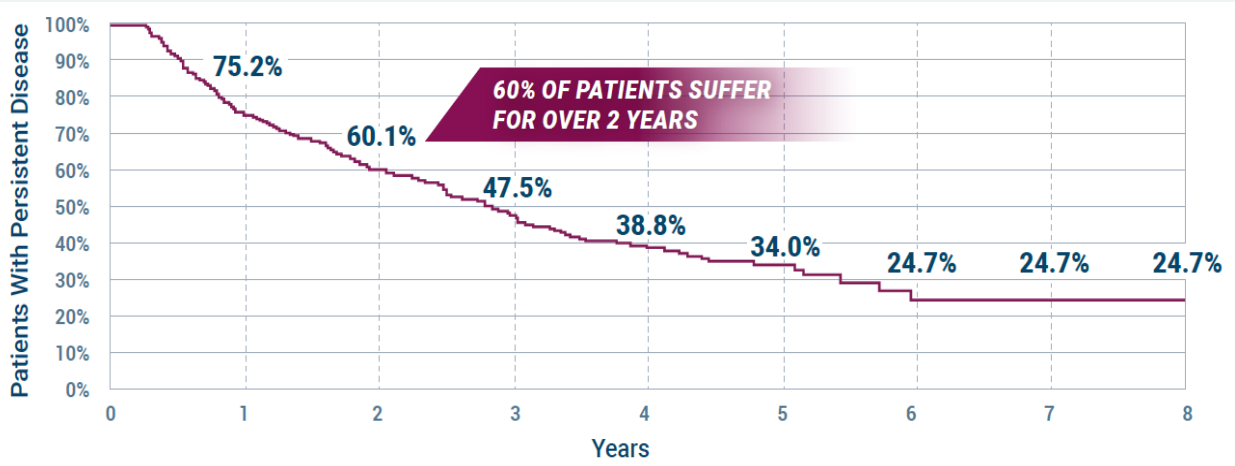
HCPs: Healthcare Professionals

# Considerations for the Duration of ARCALYST Treatment

## RECURRENT PERICARDITIS THOUGHT LEADER POSITIONING OF ARCALYST IN TREATMENT ALGORITHM[1]

| | | |
|---|---|---|
| Acute Pericarditis | Index Episode | • NSAIDs (week(s))<br>• Colchicine (3 months) |
| Recurrent Pericarditis | First Recurrence | • NSAIDs (weeks to months)<br>• Colchicine (≥6 months) |
| | Second (or more) Recurrence | • ARCALYST (rilonacept), including ahead of corticosteroids and other drugs if intolerant |
| Additionally | • ARCALYST can be used in colchicine resistant and/or corticosteroid dependent patients, including while tapering down steroids and other agents | |

## MOST PATIENTS WITH MULTIPLE RECURRENCES SUFFER FOR AT LEAST 2 YEARS[2]




Data from Optum Health Care Solutions, Inc., collected from January 1, 2007, through March 31, 2017, were analyzed for this observational study (N=375 patients with >1 recurrence of RP).

- ARCALYST treatment requires a change in mindset and practice for healthcare professionals compared with durations of traditional, non-specific therapies used for recurrent pericarditis
- The majority (60%) of patients with multiple recurrences of recurrent pericarditis have a disease duration of at least 2 years[2]
- In the Phase 3 study RHAPSODY, ARCALYST has been proven to prevent recurrences as long as there are no interruptions in therapy[3,4]
- Patients have been treated with ARCALYST over the long term[5]

1) Wang and Klein. Current Cardiology Reports (2022) 24:23-30. https://doi.org/10.1007/s11886-021-01621-0; 2) Lin D, Laliberté F, Majeski C, et al. Disease and economic burden associated with recurrent pericarditis in a privately insured United States population. Adv Ther. 2021;38(10):5127-5143. doi:10.1007/s12325-021-01868-7; 3) Other late line agents include anakinra, azathioprine, methotrexate;; 3) ARCALYST. Package insert. Kiniksa Pharmaceuticals (UK), Ltd.; 2021; 4) Klein AL, Imazio M, Cremer P, et al. Phase 3 trial of interleukin-1 trap rilonacept in recurrent pericarditis. N Engl J Med. 2021;384(1):31-41; 5) The median duration of continuous rilonacept therapy for all patients in RHAPSODY, including in the LTE portion of the trial, was 18 months for US patients and 27 months for non-US patients (Imazio M, Klein AL, Abbate A, et al. Prolonged Rilonacept Treatment In Rhapsody Long-term Extension Provided Persistent Reduction Of Pericarditis Recurrence Risk. Circulation 2022; 146 (Suppl_1):A11653

# Strong Foundation for Growth by Supporting Hundreds of Recurrent Pericarditis Patients

- ✓ Growing uptake amongst physicians continues to increase both breadth and depth of prescribing
- ✓ Patients are reporting very high satisfaction on ARCALYST treatment
- ✓ Solid commercial launch laying a strong foundation for future growth
- ✓ Steady, sequential growth quarter-on-quarter since launch
- ✓ Profitable ARCALYST collaboration
- ✓ On-track for $115-130M 2022 net revenue guidance

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**Expanded field team aiming to embed ARCALYST as the standard of care for recurrent pericarditis**

# Third Quarter 2022 Financials

Mark Ragosa
Chief Financial Officer

# Third Quarter 2022 Financial Results

| Income Statement | Q3 2022 | Q3 2021 |
|---|---|---|
| Product Revenue | $33.4M | $12.1M |
| License and Collaboration Revenue | $65.7M | $0.0M |
| **Total Revenue** | **$99.1M** | **$12.1M** |
| Cost of Goods Sold | $6.9M | $2.8M |
| Collaboration Expenses | $4.6M | $0.0M |
| Research and Development | $16.5M | $19.2M |
| Selling, General and Administrative | $24.7M | $20.8M |
| **Total Operating Expenses** | **$52.7M** | **$42.8M** |
| **Income Tax Benefit** | **$177.4M** | **$0.1M** |
| **Net Income (Loss)** | **$224.1M** | **($30.5M)** |

| Balance Sheet | September 30, 2022 | December 31, 2021 |
|---|---|---|
| Cash, Cash Equivalents and Short-term Investments | $200.7M | $182.2M |

**Cash reserves expected to fund operations into at least 2025**

# Closing Remarks

Sanj K. Patel

Chief Executive Officer

# Third Quarter 2022 Financial Results and Corporate Update

NOVEMBER 1, 2022