Fourth Quarter and Full-Year 2021 Financial Results and Corporate Update

FEBRUARY 22, 2022

Agenda

Introduction | *Sanj K. Patel, Chief Executive Officer*

ARCALYST® Commercial Execution | *Ross Moat, Chief Commercial Officer*

Portfolio Review | *Eben Tessari, Chief Operating Officer*

Fourth Quarter and Full-Year 2021 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 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; delays, difficulty or inability 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, 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.

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Introduction

Sanj K. Patel
Chief Executive Officer

Executed Across Commercial and Clinical-Stage Portfolio in 2021

Setting the stage for continued success in 2022 and beyond

2021

- Strong initial commercial launch of ARCALYST in recurrent pericarditis
- Generated Phase 2 and 3 data for mavrilimumab in COVID-19-related ARDS
- Enrolled patients in a Phase 2b dose-ranging study of vixarelimab in prurigo nodularis
- Initiated Phase 2 study of KPL-404 in rheumatoid arthritis

2022

- **Continued commercial execution for ARCALYST**
- **Phase 2b data for vixarelimab expected in 2H 2022**
- **Evaluating development of mavrilimumab in cardiovascular diseases where GM-CSF mechanism has been implicated**
- **Enrolling and dosing patients in Phase 2 study of KPL-404**
- **Strategic collaboration with Huadong Medicine**

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ARCALYST Commercial Execution

Ross Moat
Chief Commercial Officer

Continued ARCALYST Growth in Q4 2021

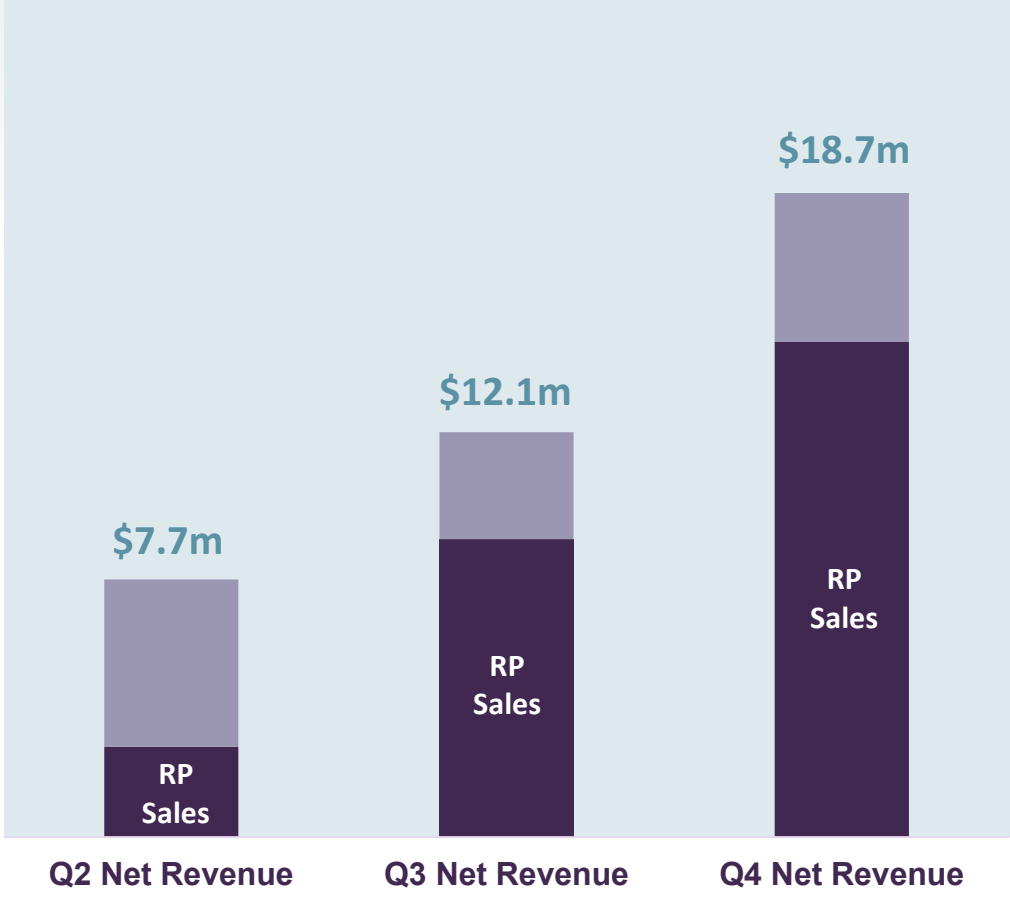
Net Revenue

- ARCALYST net revenue for Q4 2021 was $18.7 million; total 2021 net revenue since launch on April 1, 2021 was $38.5 million
- ARCALYST collaboration achieved profitability in Q4 2021 only three quarters after launch

Revenue Drivers

- Strong recurrent pericarditis demand was the primary growth driver with accelerated new patient initiations, strong adherence and compliance.
- CAPS and DIRA patient demand remained stable and broadly consistent with the previous quarters.
- Growth rate represents continued uptake and adoption of ARCALYST from physicians, payers and patients in this previously unmet and debilitating autoinflammatory cardiovascular disease.

Revenue Since Launch (April 2021): $38.5m



Kiniksa is expecting 2022 ARCALYST net revenue of $115-130 million

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

Continued Execution and Early Patient Experiences Have Driven Desired Results and Set ARCALYST Up for Strong Future Growth

Continued Broad Prescriber Adoption with Growing Depth

- More than 300 prescribers have prescribed ARCALYST for recurrent pericarditis since approval
- More than 50 physicians have prescribed ARCALYST for two or more recurrent pericarditis patients

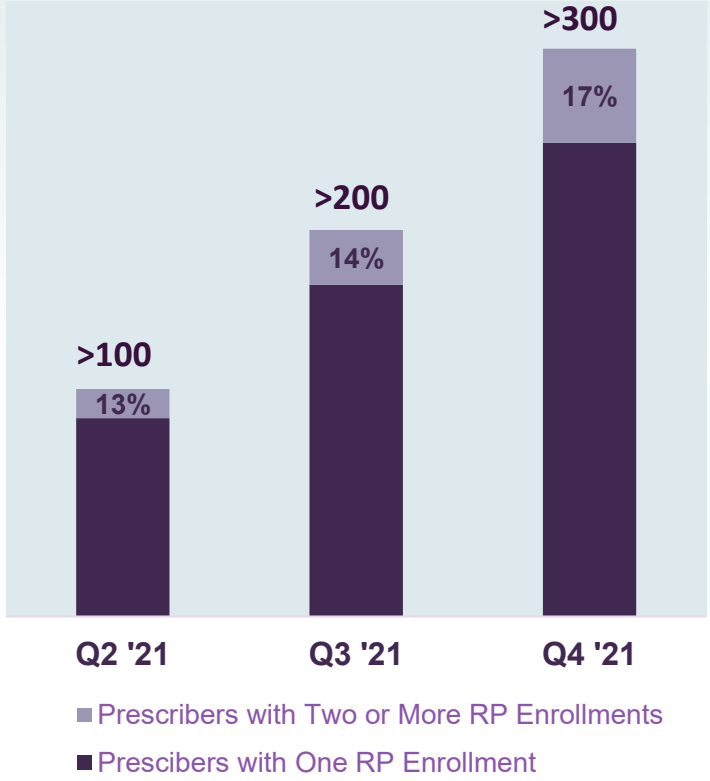
Strong Payer Experience

- In Q4, 95% of completed patient enrollment cases for recurrent pericarditis were approved for coverage
- Since launch, the median time prior to requiring re-authorization from payers is one year

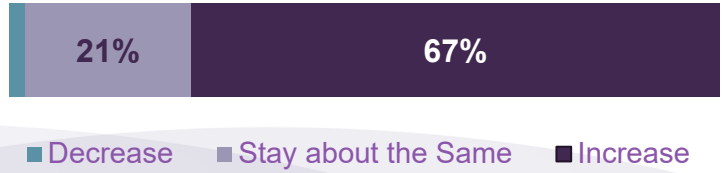
Adherence and Duration

- Adherence to ARCALYST in recurrent pericarditis has been strong, with refills generally happening on time
- Two thirds of initial ARCALYST prescriptions for recurrent pericarditis have been written for 12 months of therapy
- Approximately 70% of recurrent pericarditis patients who started ARCALYST in the second quarter of 2021 remained on therapy at the end of 2021

BREADTH AND DEPTH OF PRESCRIBER ADOPTION

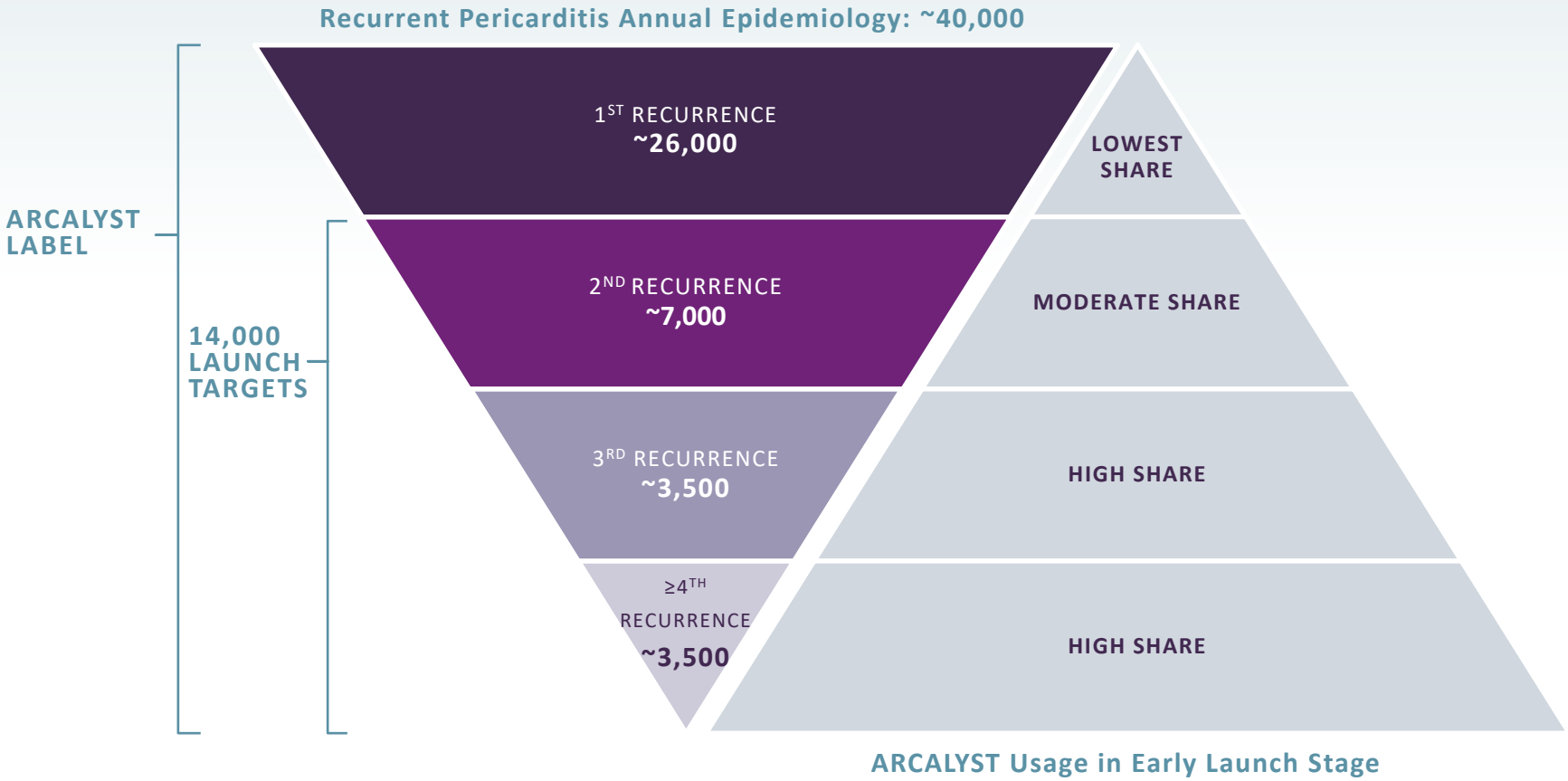


ANTICIPATED FUTURE ARCALYST PRESCRIBING[1]

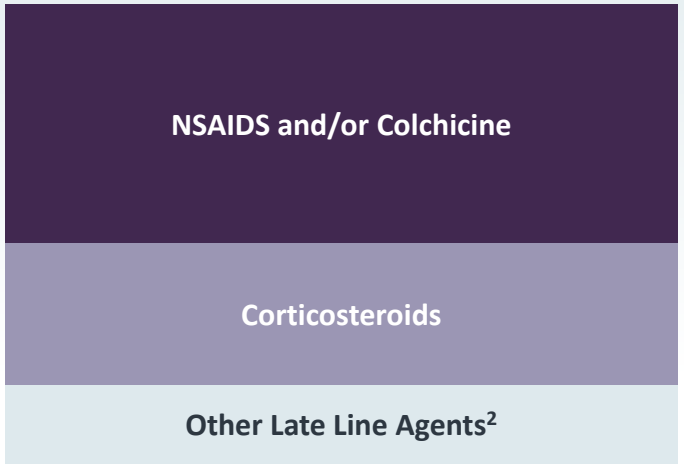


1: Among Cardiologists aware of ARCALYST. Data on file.

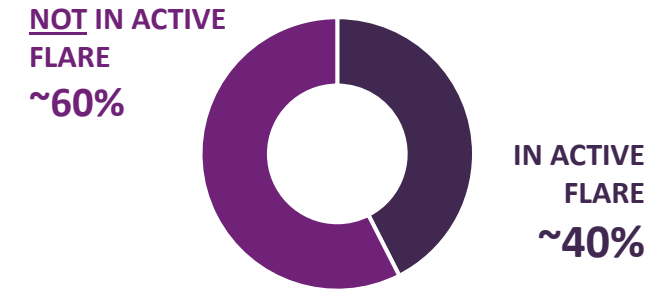
Early Treated Patients Are Closely Associated to the Launch Target Population, While Prescribers Can Utilize ARCALYST Earlier in the Disease



ARCALYST PATIENTS BY PRIOR PRODUCT[1]



ARCALYST PATIENTS BY FLARE STATUS @ INITIATION[1]

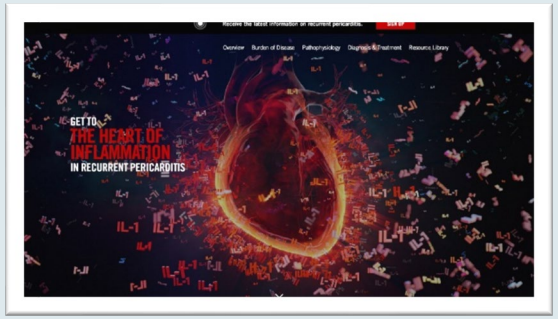


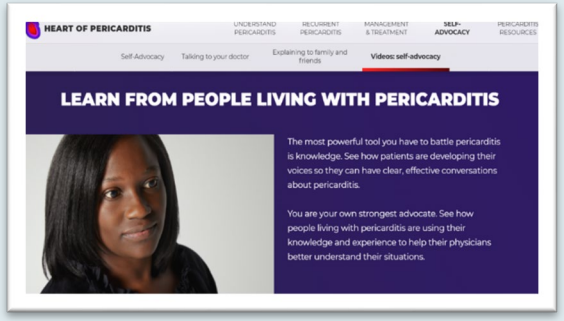
Klein A, Cremer P, Kontzias A, Furqan M, Tubman R, Roy M, Magestro M. Annals of Epidemiology. 2019;36:71; 2) Lin D, Majeski C, DerSarkissian M, Magestro M, Cavanaugh C, Laliberte F, Lejune D, Mahendran M, Duh M, Klein A, Cremer P, Kontzias A, Furqan M, Tubman R, Roy M, Mage. (Nov, 2019). Real-World Clinical 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

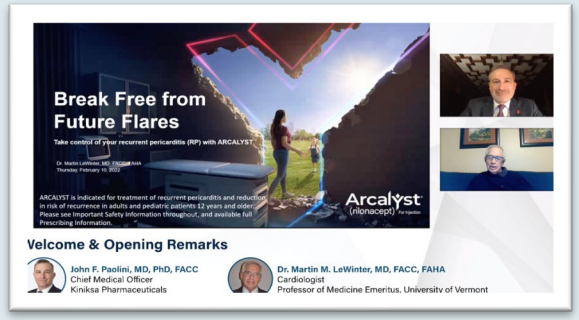
Source: 1) Kiniksa Pharmaceuticals data on file 2021. 2) Other late line agents include anakinra, azathioprine, methotrexate

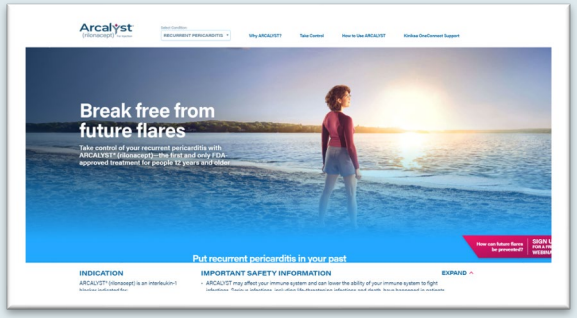
Connecting with Patients by Increasing Disease and Treatment Awareness

Educational Webinars, Search Optimization, Social Media, Patient Advocacy and Targeted Advertising is Driving Awareness and Database Opt-Ins









~2,300

Pericarditis Patients & Caregivers Currently Opted-In to Kiniksa Database

- **Tailored communication plan developed to educate and support patients**
- **Messaging individualized to each stage of the patient journey (diagnosis, initial recurrence, subsequent recurrence)**
- **Empowers patients to take action and discuss ARCALYST with their healthcare professional**

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Portfolio Review

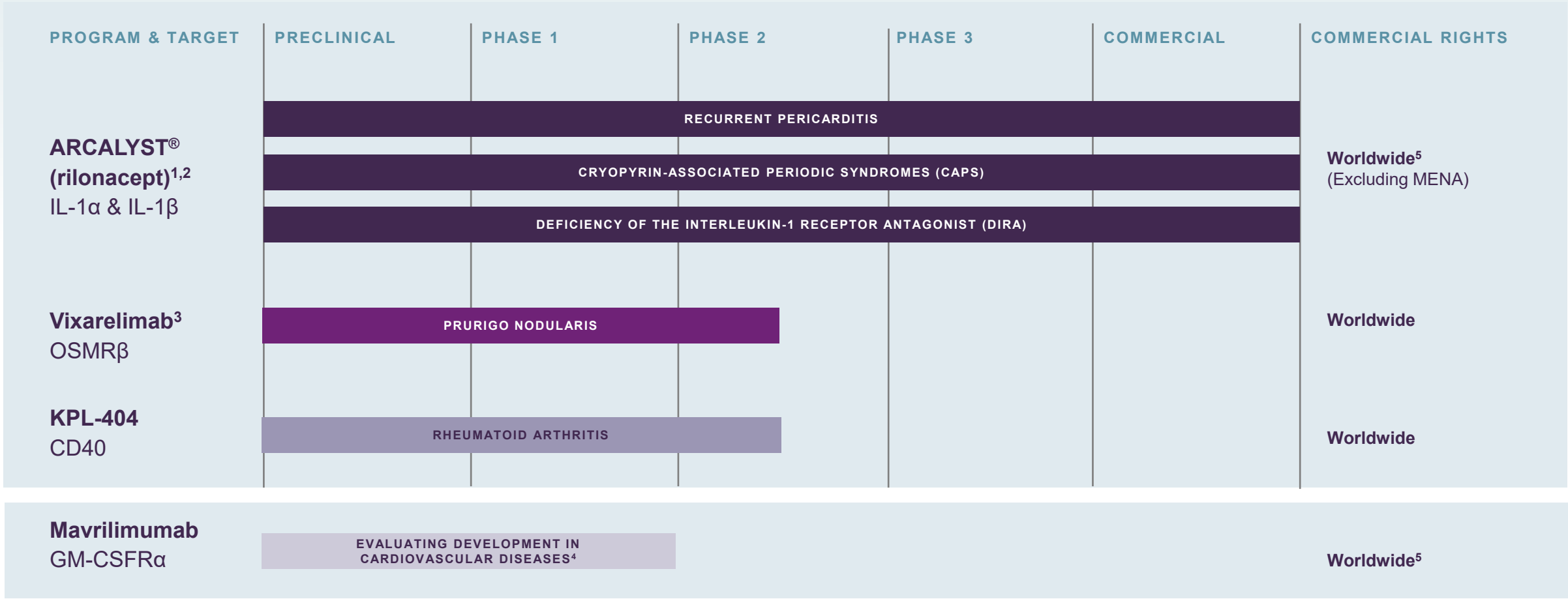
Eben Tessari
Chief Business Officer

Strategic Collaboration with Huadong Medicine for Asia Pacific Region

- Collaboration includes exclusive rights to develop and commercialize ARCALYST and mavrilimumab in the Asia Pacific Region (excluding Japan).
- Kiniksa will receive $22 million upfront and is eligible to receive up to approximately $640 million in specified development, regulatory and sales-based milestones.
  - The upfront payment includes $12 million for the territory license of ARCALYST and $10 million for the territory license of mavrilimumab.
- Kiniksa is also eligible to receive tiered royalties ranging from the low-teens to the low-twenties on annual net sales.
- Kiniksa will retain all existing development and commercialization rights for both assets.

Collaboration provides non-dilutive capital and resources to expand our clinical trials into the Asia Pacific Region to accelerate Kiniksa's drug development and commercialization efforts

Portfolio of Four 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) The FDA granted Breakthrough Therapy designation to vixarelimab for the treatment of pruritus associated with prurigo nodularis in 2020; 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

Enrolling and Dosing Patients in a Randomized Phase 2b Study of Vixarelimab in Prurigo Nodularis Across Range of Once-Monthly Dosing Regimens

EXPECTED TO ENROLL APPROX. 180 PATIENTS

- Prurigo nodularis could potentially be a meaningful opportunity
- Currently no FDA-approved therapies for prurigo nodularis and limited therapeutic options.
- Prevalence of around 300k in the US
- Market could sustain multiple entrants, especially one with differentiated mechanism

PRIMARY EFFICACY ENDPOINT (WEEK 16):

- Percent change from baseline in weekly average Worst-Itch NRS (WI-NRS) at week 16; Objective is to define a minimum effective dose level with practical subcutaneous dosing
- For each comparison, 38 subjects in each arm will provide 80% power to demonstrate a treatment effect vs placebo, assuming treatment effect of 23% difference in weekly average WI-NRS reduction from baseline at Week 16, standard deviation of 35%, and two-sided alpha of 0.05

KEY SECONDARY EFFICACY ENDPOINTS (WEEK 16):

- Proportion of patients achieving a score of 0 or 1 in PN-IGA at week 16
- Proportion of subjects achieving a ≥4-point reduction in weekly average WI-NRS from baseline

WI-NRS = Worst-Itch Numeric Rating Scale; PN-IGA = prurigo nodularis-investigator's global assessment

Data expected in 2H 2022

Vixarelimab has Potential to Deliver a Differentiated Profile Based on its Mechanistic Targets, Dosing and Presentation

PRODUCT	MECHANISM	MAGNITUDE OF ITCH RESPONSE AT STUDY END	SPEED OF ITCH RESPONSE	NODULE RESOLUTION AT WEEK 8	SPEED OF NODULE RESOLUTION	POTENTIAL DOSING AND ADMINISTRATION	PRESENTATION
VIXARELIMAB	IL-31 & OSM Inhibitor[1]	+++	+++	+++	+++	Subcutaneous Once Monthly[5]	APFS
NEMOLIZUMAB[2]	IL-31 Inhibitor	+++	+++	++	++	Subcutaneous Once Monthly	Vials for Reconstitution
DUPILUMAB[3]	IL-4 & IL-13 inhibitor	++	+	+	+	Subcutaneous Every 2 Weeks	APFS & Pre-filled Pen
NALBUPHINE ER[4]	mu opioid and kappa opioid antagonist	+	+	+	+	Oral Twice Daily	Tablet

Key: '+' designations are imputed from public data
APFS = Accessorized Pre-Filled Syringe
1) By binding to OSMRβ vixarelimab inhibits signaling through the IL-31 receptor and the Type II OSM receptor; 2) https://www.sanofi.com/-/media/Project/One-Sanofi-Web/Websites/Global/Sanofi-COM/Home/media-room/press-releases/2021/2021-10-22-07-00-00-2318876-en.pdf; 3) N Engl J Med 2020; 382:706-716 DOI: 10.1056/NEJMoa1908316; 4) https://www.sec.gov/Archives/edgar/data/1563880/000156459021015506/trvi-10k_20201231.htm; 5) Patients in the Phase 2a trial of vixarelimab recieved a loading dose of vixarelimab or placebo followed by vixarelimab or placebo weekly. The current Phase 2b trial is evaluating vixarelimab across a range of once-monthly dosing regimens.

Fourth Quarter and Full-Year 2021 Financials

Mark Ragosa
Chief Financial Officer

Fourth Quarter and Full-Year 2021 Financial Results

Income Statement	Three Months Ended December 31,		Year Ended December 31,	
	2021	2020	2021	2020
Total Revenue	$18.7M	N/A	$38.5M	N/A
Cost of Goods Sold	$3.9M	N/A	$9.1M	N/A
Collaboration Expenses	$0.8M	N/A	$0.8M	N/A
Research and Development Expenses	$27.4M	$37.4M	$99.3M	$112.0M
Selling, General and Administrative Expenses	$22.7M	$15.5M	$85.9M	$45.3M
Total Operating Expenses	$54.9M	$52.9M	$195.2M	$157.4M
Net Loss	($36.3M)	($53.7M)	($157.9M)	($161.4M)

Balance Sheet	December 31, 2021	December 31, 2020
Cash, Cash Equivalents and Short-term Investments	$182.2M	$323.5M

Q4 2021 Cash Reserves Expected to Fund Current Operating Plan into 2024

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Closing Remarks

Sanj K. Patel

Chief Executive Officer

Kiniksa is an Emerging Leader in the Development of Immune-Modulating Therapies

BY THE NUMBERS

1 FDA-approved therapy

3 Clinical-stage assets

3 Orphan Drug designations

2 Breakthrough Therapy designations

2015 Company founded

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

DISEASE AREAS

- **Recurrent Pericarditis**
- **Cryopyrin-Associated Periodic Syndromes (CAPS)**
- **Deficiency of IL-1 Receptor Antagonist (DIRA)**
- **Prurigo Nodularis**
- **Rheumatoid Arthritis**
- **Evaluating development in cardiovascular diseases**

- Commercialized **ARCALYST** and brought this therapeutic to patients in need only 3.5 years after in-licensing it.
- Highly encouraged by the potential of **vixarelimab** in prurigo nodularis and **KPL-404** in a range of autoimmune diseases.
- Evaluating development of **mavrilimumab** in cardiovascular diseases where the GM-CSF mechanism has been implicated and with synergies with the company's existing commercial infrastructure.
- Determined to continue to help patients in need and fulfill plan of becoming a generational company.

Cash reserves expected to fund our current operating plan into 2024

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Fourth Quarter and Full-Year 2021 Financial Results and Corporate Update

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