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Second Quarter 2021 Financial Results and Recent Corporate and Pipeline Activity

AUGUST 3, 2021

Agenda

Introduction | *Sanj K. Patel, CEO and Chairman of the Board*

ARCALYST® Launch Quarter | *Ross Moat, Head of ARCALYST Franchise*

Portfolio Update | *John F. Paolini, MD, PhD, FACC, Chief Medical Officer*

Second Quarter 2021 Financial Results | *Mark Ragosa, Chief Financial Officer*

Closing Remarks | *Sanj K. Patel, CEO and Chairman of the Board*

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; 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; 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.

Introduction

Sanj K. Patel

Chief Executive Officer and Chairman of the Board

Developing Life-Changing Medicines For The Patients Who Need Them Most

BY THE NUMBERS

1 FDA-approved therapy

3 Clinical-stage assets in multiple indications

3 Orphan Drug designations

2 Breakthrough designations

40 active and completed global clinical studies to date

200+ employees and growing

2015 Company founded

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

LOCATIONS

MORE STRENGTHS

- Passionate employees
- A robust pipeline of product candidates for debilitating diseases
- Find and deliver novel treatments for patients with a significant unmet need
- Focus on immune modulation
- Strong biologic rationale and validated mechanisms
- In-house research team and lab
- Kiniksa manufacturing for early-stage programs

DISEASE AREAS

- Recurrent Pericarditis
- Cryopyrin-Associated Periodic Syndromes (CAPS)
- Deficiency of IL-1 Receptor Antagonist (DIRA)
- Giant Cell Arteritis (GCA)
- COVID-19-Related Acute Respiratory Distress Syndrome (ARDS)
- Prurigo Nodularis
- Rheumatoid Arthritis

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ARCALYST Launch Quarter

Ross Moat
ARCALYST General Manager

ARCALYST: First and Only FDA-Approved Therapy for Recurrent Pericarditis

Third indication for ARCALYST underscores utility in IL-1 mediated diseases

Arcalyst® (rilonacept) Injection for Subcutaneous Use

Arcalyst® (rilonacept) For Injection

2008	2020	2021
CAPS FDA Approved	DIRA FDA Approved	Recurrent Pericarditis FDA Approved

KINIKSA oneconnect™ support made simple.

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

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

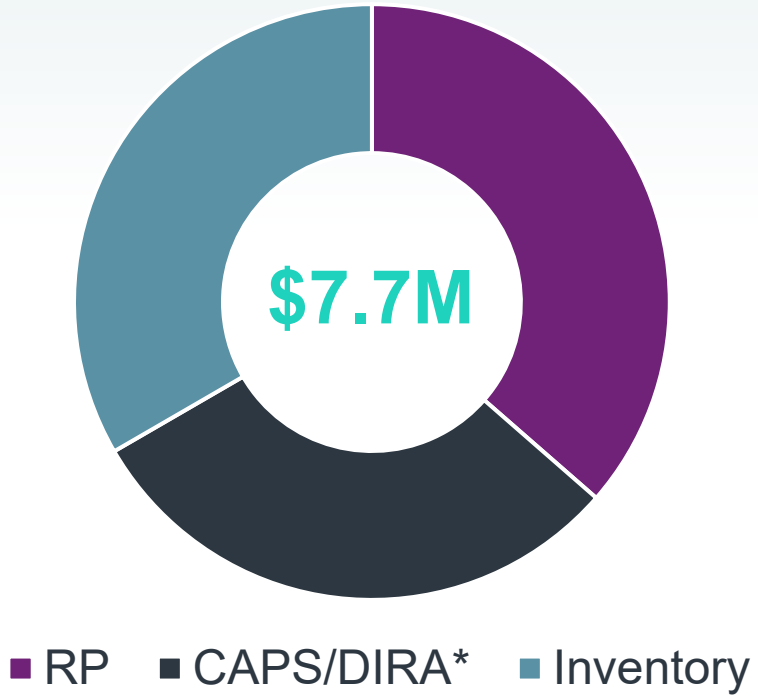
Net Revenue

- $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

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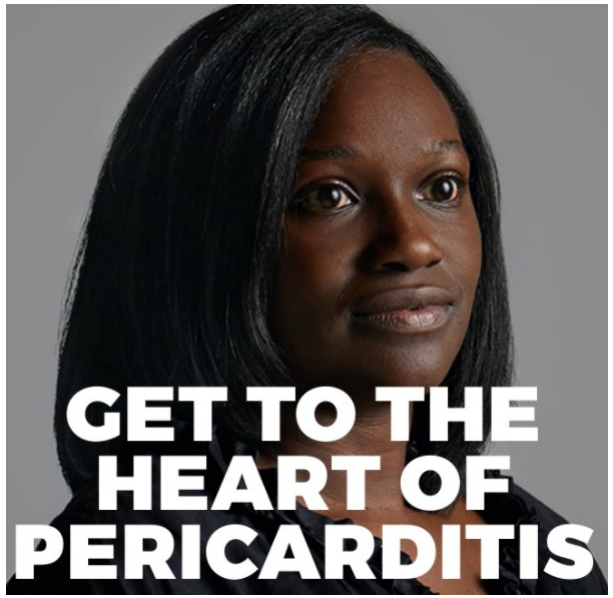
*Includes prescriptions for other indications

Strong Execution Generated Patient Demand, Physician Breadth and Early Payer Approvals in Recurrent Pericarditis

Arcalyst® (rilonacept) For Injection



Early broad physician adoption

- >65% of all recurrent pericarditis prescriptions came from HCPs practicing outside of Phase 3 RHAPSODY clinical trial sites
- >100 physicians who did not participate in RHAPSODY prescribed ARCALYST to at least one recurrent pericarditis patient

RHAPSODY Conversions

- Greater than 70% of US RHAPSODY patients from the LTE remained on therapy in the commercial setting

Strong Early Payer Experience

- >90% approval rate among RP patients with completing the Prior Authorization / Appeal process
- Expectation remains that most payers will establish formal coverage policies within six months of launch

Compliance and Duration

- While early, refills are being filled on time; expectation is compliance will be similar to benchmarks for injectables

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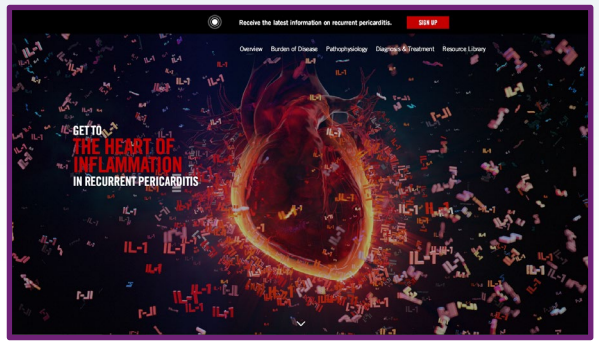
Specialty Cardiology Salesforce Expected to Reach ~70% of U.S. Recurrent Pericarditis Patients

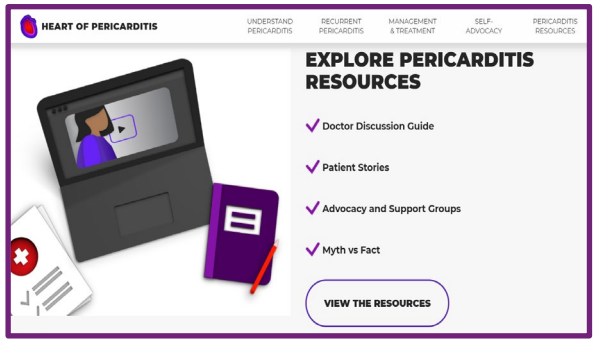
Focused & Targeted Sales Execution

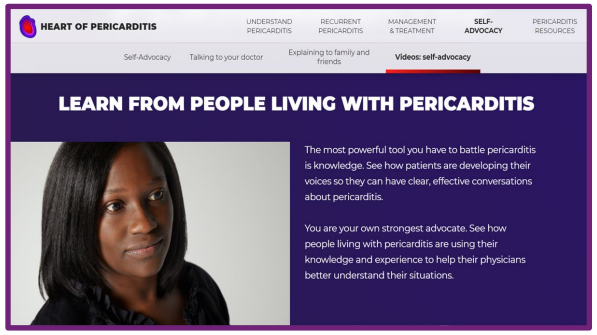
National	Territory Level	
Initial launch focus on top tier accounts **~45%** of RP patients nationally **~350** accounts nationally	**Early Launch**	**Mid-Term Expansion**
Following adoption, moving into next deciles to **~70%** of RP patients nationally **~800** accounts nationally (20% of total accounts)	**10-15** accounts **~60** high value HCPs	**30** accounts **~100** high value HCPs

Specialty cardiology sales force of ~30 reps

Disease Awareness and ARCALYST promotion







Patient Advocacy Support

PERICARDITIS ALLIANCE

AUTOINFLAMMATORY ALLIANCE formerly known as The NOMID Alliance

MYOCARDITIS FOUNDATION

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Comprehensive Support for Patients Through Kiniksa OneConnect™

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

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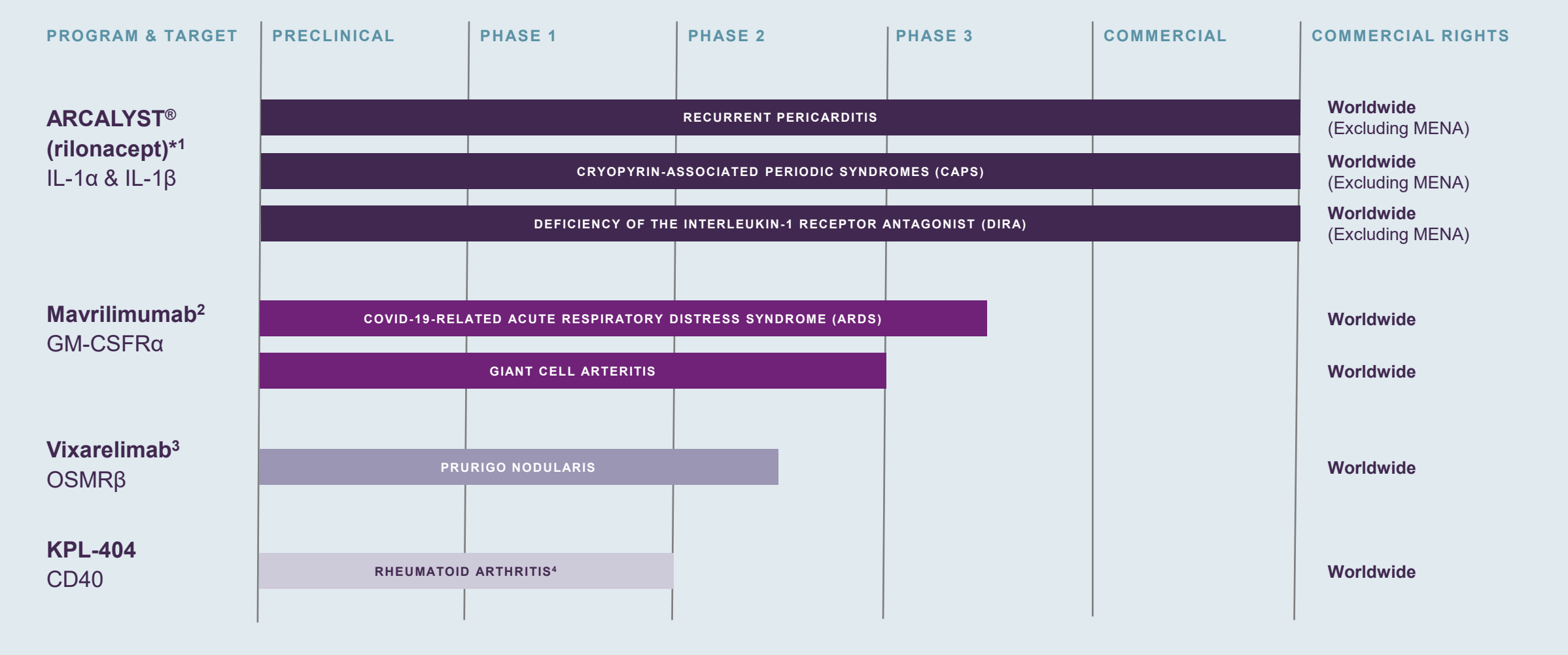
Portfolio Update

Dr. John F. Paolini
Chief Medical Officer

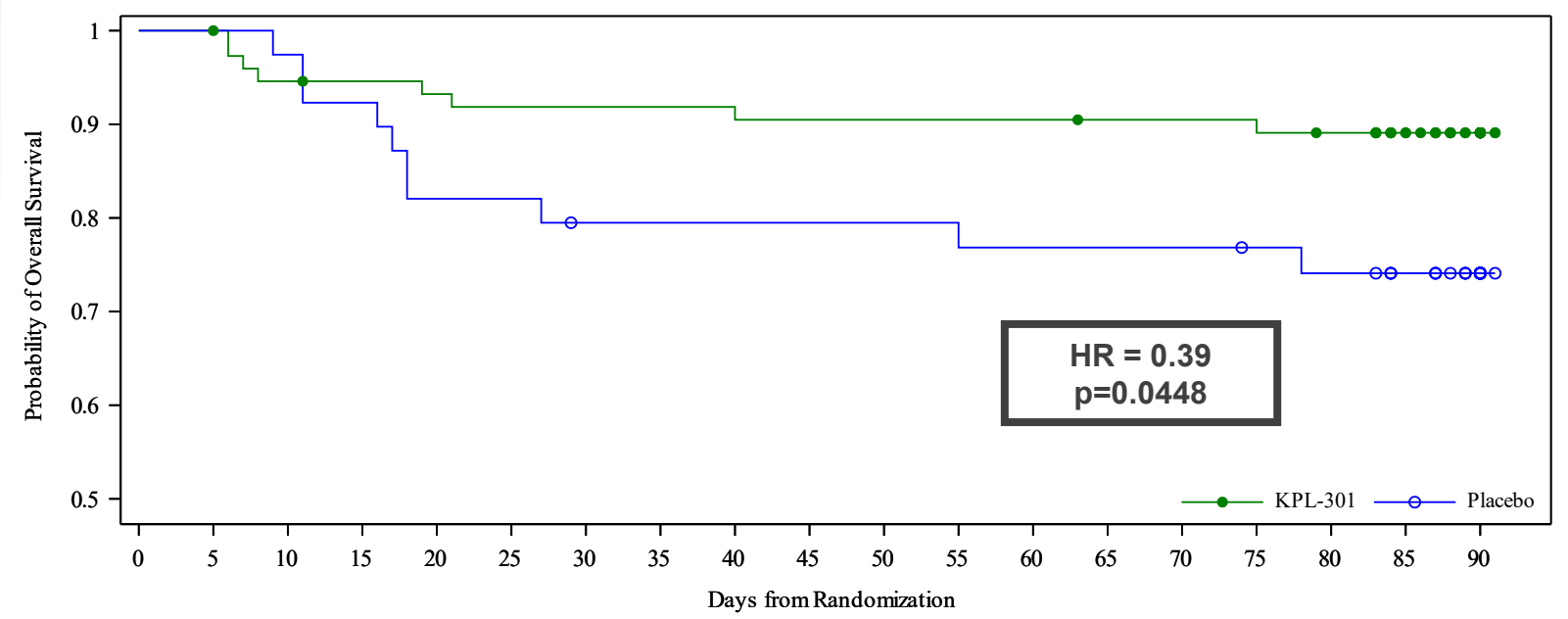
Portfolio of Four Immune-Modulating Assets



* 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 mavrilimumab 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

Mavrilimumab Demonstrated Persistent Clinical Effect Through Day 90

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



KPL-301	75	75	70	69	68	67	67	67	67	66	66	66	66	65	65	65	63	55	43
Placebo	39	39	38	36	32	32	30	30	30	30	30	30	29	29	29	28	27	23	16

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Second Quarter 2021 Financials

Mark Ragosa
Chief Financial Officer

Q2 2021 Financial Results

Income Statement	Three Months Ended June 30,	
	2021	2020
Total Revenue	$7.7M	N/A
Cost of Goods Sold	$2.5M	N/A
Research and Development Expenses	$23.9M	$22.3M
Selling, General and Administrative Expenses	$21.8M	$9.5M
Total Operating Expenses	$48.3M	$31.9M
Net Loss	($41.6M)	($37.5M)

Balance Sheet	June 30, 2021	December 31, 2020
Cash, Cash Equivalents and Short-term Investments	$225.9M	$323.5M

Q2 2021 Cash Reserves Expected to Fund Current Operating Plan into 2023

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Closing Remarks

Sanj K. Patel

Chief Executive Officer and Chairman of the Board

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

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