KINIKSA

Every Second Counts!™

FDA Approval of ARCALYST® (rilonacept) for Recurrent Pericarditis

March 18, 2021

Agenda

Welcome| *Mark Ragosa, Chief Financial Officer*

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

ARCALYST Label & Data| *John F. Paolini, MD, PhD, FACC, Chief Medical Officer*

Commercialization Strategy| *Ross Moat, ARCALYST General Manager*

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" 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 corporate strategy; product development and prospects; potential indications; potential market opportunities and competitive position; potential impact of FDA approval of ARCALYST in recurrent pericarditis for patients and Kiniksa; our commercial strategy; potential impact of our commercialization activities; commercial launch timing; timing for establishing payer coverage; on going, planned and potential clinical trials and other studies; timing and potential impact of clinical data; mechanisms of action and potential of our product candidates; 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: our inexperience as a company commercializing therapeutic products; our limited experience as a company establishing sales, marketing, distribution and general infrastructure either directly and/or through agreements with third parties for therapeutic products; the potential for ARCALYST to not gain market acceptance by physicians, patients, or third-party payers for recurrent pericarditis; the potential delay or failure of ARCALYST to obtain or maintain coverage and adequate reimbursement for the treatment of recurrent pericarditis; potential for a smaller target patient population for ARCALYST in recurrent pericarditis; our reliance on third parties for manufacturing our product candidates and the supply of drug substance, including Regeneron as the sole source of supply of ARCALYST; drug substance and/or drug product shortages; our reliance on third parties for conducting clinical trials, research and other studies; the impact of the COVID-19 pandemic and measures taken in response thereto on our business and operations as well as the those of the third parties with whom we conduct business or otherwise engage; 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; impact of additional data from us or other companies; potential undesirable side effects caused by our product candidates; potential changes in our strategy, corporate priorities, operating plan and funding requirements; substantial new or existing competition; potential for applicable regulatory authorities to not accept our regulatory filings or to delay or deny approval of any of our product candidates or to require additional trials to support any such approval; complications in coordinating requirements, regulations and guidelines of regulatory authorities across jurisdictions for our clinical trials; and our ability to attract and retain qualified personnel. These and the other important factors are discussed under the caption "Risk Factors" in our Annual Report on Form 10-K filed with the Securities and Exchange Commission (the "SEC") on February 25, 2021 and other filings subsequently filed with the SEC. 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.

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

NOW FDA APPROVED

Arcalyst®

(rilonacept) For Injection

ARCALYST Label and Data

Dr. John F. Paolini
Chief Medical Officer

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Recurrent Pericarditis Episodes are Painful, Debilitating and Disruptive to Quality of Life

"I cannot work, walk to the mailbox, or go up/down stairs without a great deal of pain and shortness of breath. Many referred visits to the ER because of pain, where ER docs accuse me of drug seeking for pain. It's humiliating and scary." [1]

Pericarditis Recurrences are Burdensome for Patients…

- Significant pain with similar symptoms as heart attack that **drive patients to the ER**[1,2,5]
- After acute pain resolves, **residual pain** and other effects can last weeks to months[1,2]
- Elevated **risk for major complications**, such as cardiac tamponade and constrictive pericarditis[4,6]
- Results **in hospitalization and ER visits** for large proportion of patients[1,4,6,7,8]
- Increased **absenteeism** driven by pain and anxiety[1,2]

"I have gained a great deal of weight from steroids and inactivity. Exercise sets off more events, so am afraid to exercise. Pain is there constantly, just not as intense as it is during an event. [My] quality of life [is] greatly diminished." [1]

…And the Burden of the Disease Persists Even After the Acute Episode Resolves

- Testimonials reveal **negative impact on quality of life (QoL)** (anxiety, loss of sleep, lifestyle change, physical activity)[1,2,5]
- Between flares, 48% of patients report their level of **fear of pericarditis** as "quite a bit" or "very much"[9]
- Corticosteroids have well known **safety and tolerability issues**, and increase recurrence rates with taper[1,2,4,5,6,7]
- Significantly **worse QoL than general population** - Ph2 PROMIS physical and mental health[3]
- Increased **depression and anxiety** diagnoses seen in claims data following initial pericarditis event[4]
- 98% of patients express **need for additional therapies** that reduce the likelihood of another recurrence[1]

1) Patient ATU Research 2019 W1; 2) Patient & Physician Emotive Journey Qual 2019 Q2; 3) Ph2 Pilot study; 4) IQVIA PharMetrics Plus Claims Analyses; 5) Putnam Patient Journey Research 2018 Q1; 6) External Publications (Cremer, P "Complicated Pericarditis", ESC Guidelines), 7) HCP ATU Research 2019 W1; 8) ER ATU Research 2019 W1; 9) LeWinter (2020 ISPOR Abstract)

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ARCALYST Label

ARCALYST is a patient-administered once-weekly subcutaneous therapy

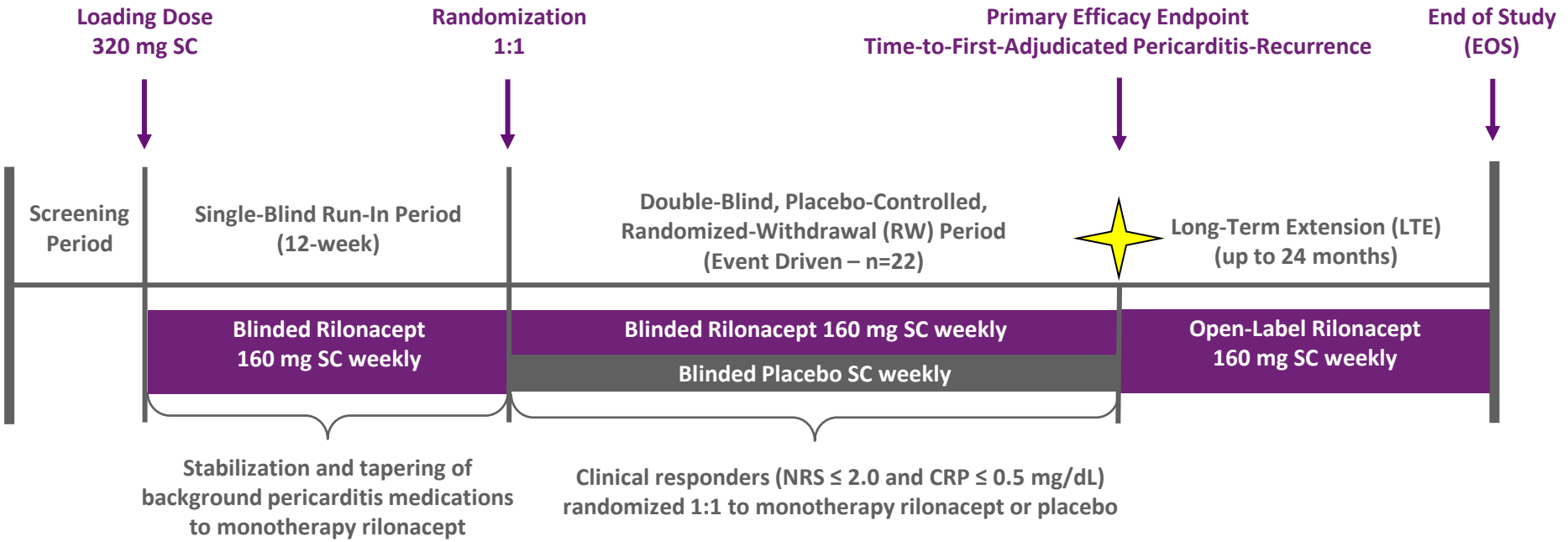
ADULTS (18 years and older)	**ADOLESCENTS** (12 to 17 years)
Loading dose: **320 mg** delivered as two 160 mg (2 mL) injections	**Loading dose:** **4.4 mg/kg** delivered up to a maximum of 320 mg, delivered as 1 or 2 injections (not to exceed 2 mL/injection)
Weekly maintenance dose: **160 mg** delivered once weekly as a 2 mL injection	**Weekly maintenance dose:** **2.2 mg/kg** delivered up to a maximum of 160 mg (2 mL) injection, once weekly

The first injection of ARCALYST should be performed under the supervision of a healthcare professional.

ARCALYST is supplied in sterile, single-use, 20-mL glass vials

- Each vial contains 220 mg ARCALYST, a sterile, white to off-white lyophilized powder
- Reconstitution with 2.3 mL of preservative-free Sterile Water for Injection is required prior to subcutaneous administration of the drug
- The reconstituted ARCALYST is a viscous, clear, colorless to pale yellow, free from particulates, 80-mg/mL preservative-free solution

Pivotal Phase 3 Trial of ARCALYST in Recurrent Pericarditis



Inclusion Criteria:

- All etiologies except infection and malignancy
- Present at screening with at least a third pericarditis episode, defined as at least 1 day with **NRS pain of ≥ 4** and **CRP value ≥ 1 mg/dL** within the 7-day period prior to first study drug administration
- Concomitant NSAIDs and/or colchicine and/or oral corticosteroid treatment in any combination

Primary Efficacy Endpoint :

- Time-to-first-adjudicated pericarditis-recurrence in the RW period

Major Secondary Efficacy Endpoints (16-weeks):

- Proportion of subjects who maintained Clinical Response
- Percentage of days with no or minimal pain
- Proportion of subjects with absent or minimal pericarditis symptoms

CEC Adjudication Criteria:

- Typical pericarditis pain (≥ 1 pain **NRS recording ≥4**) AND elevated **CRP (≥1.0 mg/dL)**, same day or ≤ 7 days
- Typical pericarditis pain (≥ 1 pain **NRS recording ≥4**) AND abnormal **CRP (>0.5 mg/dL),** same day or ≤ 7 days AND **≥ 1 supportive evidence** of pericarditis
- Typical pericarditis pain (BUT pain **NRS recording ≤ 4**) AND elevated **CRP (≥1.0 mg/dL)**, AND **≥ 1 supportive evidence** of pericarditis

CRP = C-reactive protein; NRS = Numerical Rating Scale; NSAIDs = nonsteroidal anti-inflammatory drugs; CEC = Clinical Endpoint Committee

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.

ARCALYST Initiation Resulted in Rapid Resolution of Pericarditis Episodes

Pivotal Phase 3 RHAPSODY Data

RHAPSODY

Rapid and sustained reductions in both reported pain and inflammation as early as after the first dose of ARCALYST

Median time to pain response = 5.0 days; Median time to CRP normalization = 7.0 days

Secondary endpoints that were assessed during the run-in period

5 days

Time to treatment response (median; 95% CI: 4, 7)*

97%

Treatment response* rate

7.9 weeks

Time to ARCALYST monotherapy (median; 95% CI: 7, 8)

*Time to treatment response was defined as the time from the first dose to the first day when pericardial pain was NRS ≤2 and CRP ≤0.5 mg/dL (measured within 7 days before or after the pain response). During the 12-week run-in period, 77 of 79 patients demonstrated a treatment response.

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.
ARCALYST (rilonacept) prescribing information 2021

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ARCALYST demonstrated a steroid-sparing treatment effect

Pivotal Phase 3 RHAPSODY Data

RHAPSODY

Patients treated with ARCALYST discontinued corticosteroids

In the run-in period of the Phase 3 trial RHAPSODY, patients receiving corticosteroids at baseline were transitioned to ARCALYST monotherapy in 7.9 weeks

Each patient treated with corticosteroids at baseline achieved clinical response with ARCALYST monotherapy

- 44.3% (27 of 61) of patients received corticosteroids at baseline
- None of the patients treated with corticosteroids at baseline and randomized to ARCALYST monotherapy experienced a recurrence while on therapy

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.
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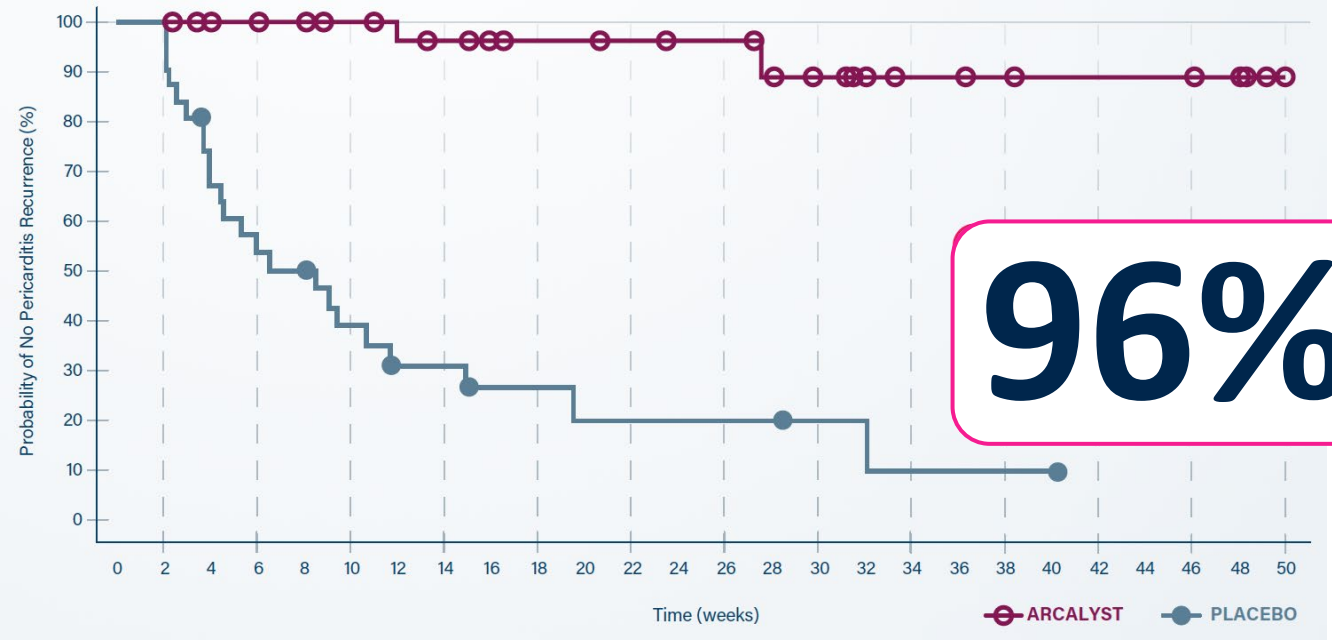
96% Reduction in Risk of Pericarditis Recurrence

Pivotal Phase 3 RHAPSODY Data

RHAPSODY

ARCALYST reduced the risk of pericarditis recurrence

The primary efficacy endpoint was time to first adjudicated pericarditis recurrence in the randomized withdrawal period.



The median time to recurrence on ARCALYST could not be estimated due to the low number of recurrences

- 2 of 30 of patients treated with ARCALYST had a recurrence
- The 2 pericarditis recurrences with ARCALYST occurred during temporary interruptions of 1 to 3 doses of ARCALYST

96% **reduction in the risk of recurrent pericarditis (hazard ratio: 0.04; *p*<0.0001)**

The median time to recurrence on placebo was 8.6 weeks (95% CI: 4.0, 11.7)

- 74% (23 of 31) of patients treated with placebo experienced a recurrence at the time that the event-driven portion of the trial was closed
- Consistent with the expected washout pharmacokinetics of once-weekly ARCALYST at steady state

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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.
ARCALYST (rilonacept) prescribing information 2021

92% of Trial Days of No/Minimal Pain

Pivotal Phase 3 RHAPSODY Data

RHAPSODY

Patients on ARCALYST had significantly more trial days with no/minimal pain vs placebo

Secondary efficacy endpoint was assessed during the randomized withdrawal period

92% of days

Patients reported no/minimal (NRS≤2) pericarditis pain

Compared with 40% of trial days in patients on placebo ($p<0.0001$) at the secondary endpoint assessed at Week 16 of the randomized withdrawal period.

At Week 16 of the randomized withdrawal period:

- A majority (81%) of patients maintained a clinical response measured at Week 16 of the randomized withdrawal period compared with 20% of patients on placebo ($p=0.0002$)

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.
ARCALYST (rilonacept) prescribing information 2021

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Most common ARCALYST adverse reactions: Injection-site reactions and upper respiratory tract infections

RHAPSODY

Adverse experiences in RHAPSODY

	RUN-IN PERIOD	RANDOMIZED-WITHDRAWAL PERIOD				TOTAL (N=86)
EVENT	Rilonacept (N=86)	Rilonacept, Including Bailout (N=30)	Placebo, Including Bailout (N=31)	Rilonacept, Before Bailout (N=30)	Placebo, Before Bailout (N=31)	
			number of patients with event (percent)			
Any adverse event	69 (80)	24 (80)	22 (71)	24 (80)	13 (42)	74 (86)
Adverse events according to maximum severity†						
Mild	52 (60)	16 (53)	17 (55)	16 (53)	9 (29)	47 (55)
Moderate	15 (17)	8 (27)	5 (16)	8 (27)	4 (13)	25 (29)
Severe	2 (2)	0	0	0	0	2 (2)
Serious adverse event	1 (1)	1 (3)	3 (10)	1 (3)	1 (3)	5 (6)
Adverse event leading to death	0	0	0	0	0	0
Adverse event leading to dose interruption	0	1 (3)	0	1 (3)	0	1 (1)
Adverse event leading to discontinuation of rilonacept or placebo	4 (5)	0	0	0	0	4 (5)
Cancer‡	0	1 (3)	0	1 (3)	0	1 (1)
Injection-site reaction	28 (33)	6 (20)	2 (6)	5 (17)	0	29 (34)
Infection or infestation	14 (16)	12 (40)	7 (23)	12 (40)	3 (10)	29 (34)
Upper respiratory tract infection	12 (14)	7 (23)	2 (6)	7 (23)	0	19 (22)

*Patients with multiple events were counted once in each appropriate category

†Counted once, according to the maximum severity of the adverse event.

‡Cancer was an event of special interest.

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.

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ARCALYST Use in Clinical Practice

Average Duration of Recurrent Pericarditis is 2 Years[1]

- The presence of certain baseline characteristics may identify patients who may benefit from longer-term treatment
- The mean duration of disease in RHAPSODY in patients prior to enrollment was 2.4 years

Median treatment duration in RHAPSODY was 9 months, with a range up to 14 months, at the close of the randomized period

- ARCALYST treatment was associated with a 96% reduction in risk for pericarditis recurrence
- Patients on ARCALYST experienced none/minimal pericarditis pain for 92% of trial days[2]
- 74/75 patients continued into LTE for longer-term therapy, demonstrating a desire to continue to a duration of up to 24 months

Data support treatment duration tailored to duration of autoinflammation

- Registry data indicate patients treated for 6 months have worse outcomes compared to patients treated for 9 months[3]
- The only events in the ARCALYST arm in the randomized period of RHAPSODY took place in the setting of temporary drug interruptions of 1-3 doses
- Continued ARCALYST treatment resulted in continued treatment response.

Additional data anticipated from LTE, in which patients are assessed at 18 months (including imaging) for possible treatment cessation under observation[4]

1) D. Lin, et al.; Recurrence Burden in Recurrent Pericarditis: A US-Based Retrospective Study of Administrative Healthcare Claims; Quality of Care and Outcomes Research (QCOR) 2020 Scientific Sessions; 2) Compared to 40% of trial days in patients treated with placebo; 3) M. Imazo, et al.; *Anakinra for corticosteroid-dependent and colchicine-resistant pericarditis: The IRAP (International Registry of Anakinra for Pericarditis) study.* European Journal of Preventative Cardiology 2019; 4) A. Klein, et al.; 2020 AHJ Reference for Phase 3 design; LTE = long-term extension

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Commercialization Strategy

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.

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

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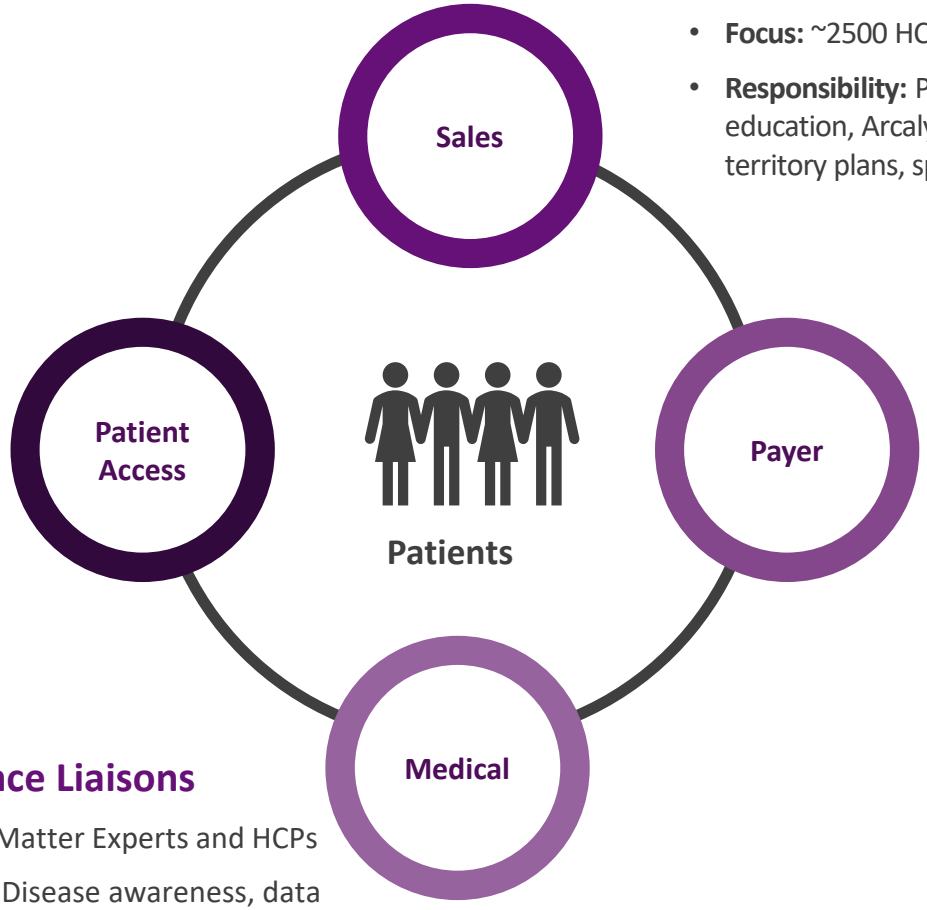
Collaborative Field Force to Drive Awareness, Overcome Access Barriers and Help Ensure Positive Patient and Physician Experience

Clinical Sales Specialists

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

Patient Access Leads

- **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



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

HCP = health care provider

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

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

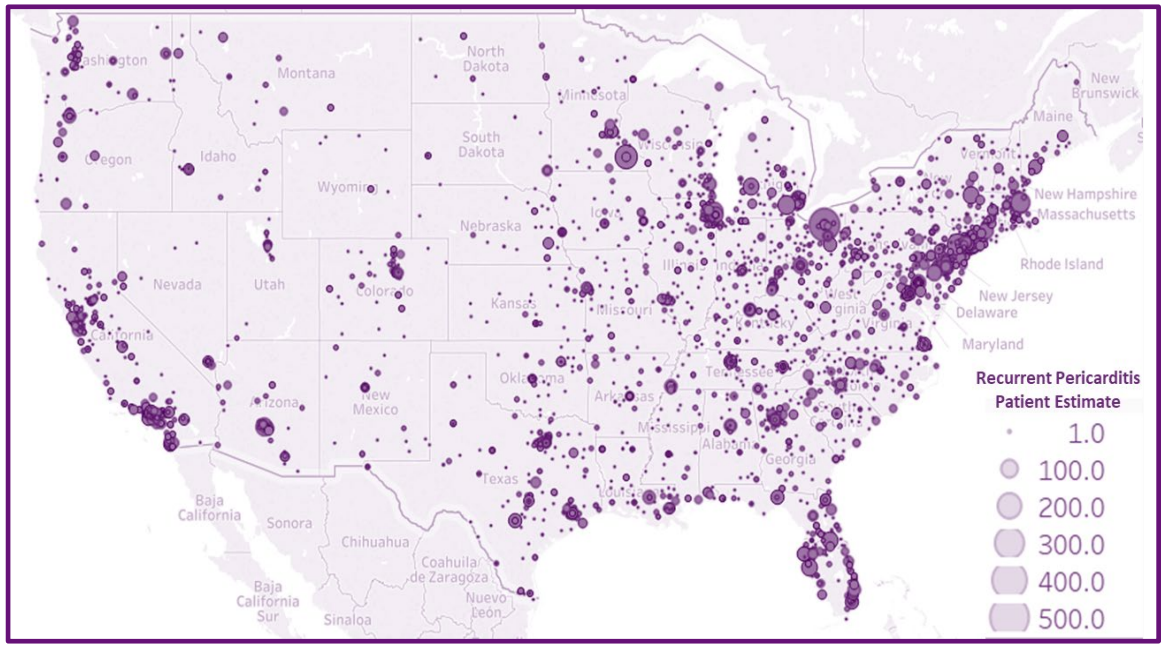
Clear Call to Action: ~14K Patients

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.

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

Estimated Recurrent Pericarditis Patients by Account

Strategy Targeting



Focused & Targeted Sales Execution

National	Territory Level	
Initial launch focus on top tier accounts ~**45%** of RP patients nationally ~**350** accounts nationally	**First 3 months**	**Within the First Year**
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

HCP = health care physicians

COVID-19: Strategic Response and Tools to Help Ensure a Successful Launch

Enabled Tools to Support Effective Remote Detailing

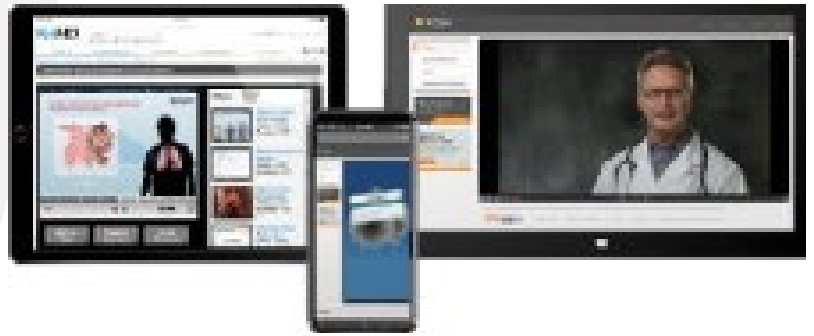
- Support convenient, impactful and compliant virtual content sharing
- Mitigate COVID-19 risk of physical access restriction

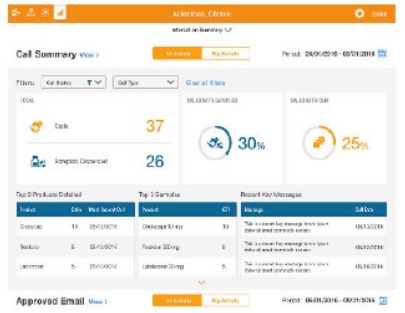
Representative-Triggered Approved Emails

- Improve quality of email reach with more tailored messages
- Drive engagement rates due to a known cardiovascular sales representative

Field Force Build

- Extensive Cardiology, Biologic and Rare Disease experience
- Previous experience with multiple drug launches and familiarity with virtual selling





Building to and Supporting a Successful Launch

Disease Educational Programs

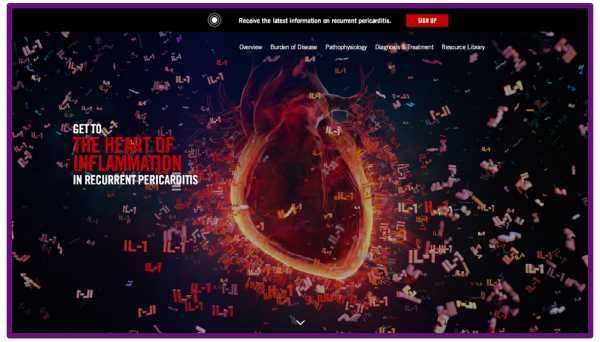
- Whatispericarditis.com; co-created with patients to provide support and self-advocacy including doctor discussion guides
- Heartofinflammation.com; targeted for healthcare professional disease knowledge
- Webcast series focused on recurrent pericarditis disease understanding

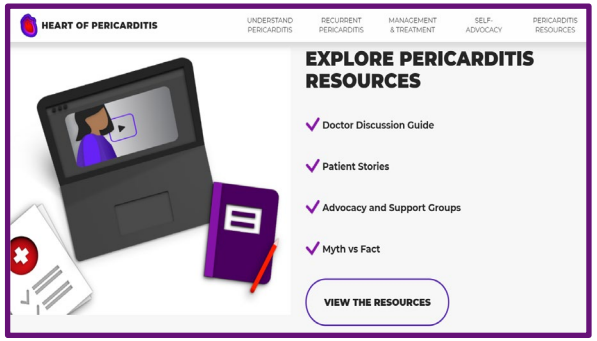
Promotional Engagements

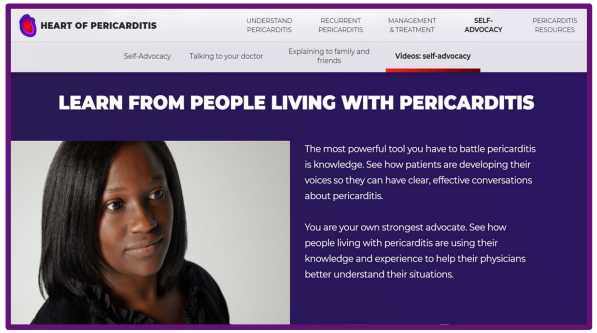
- Launch meetings in top accounts during early weeks of launch
- Treatment focused patient webcasts
- Peer-to-Peer speaker programs
- Key congresses in 2021

Continued Patient Advocacy

- Pericarditis Alliance
- Myocarditis Foundation
- Autoinflammatory Alliance







PERICARDITIS ALLIANCE

MYOCARDITIS FOUNDATION

American Heart Association.

AUTOINFLAMMATORY ALLIANCE formerly known as The NOMID Alliance

AMERICAN COLLEGE of CARDIOLOGY

>1,000 Patients & Caregivers Registered with Kiniksa

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Pricing, Access and Distribution Considerations

Pricing

- Kiniksa maintains the already established list price for ARCALYST **of $20,000 per month**

  Based on first and only FDA-approved therapy for recurrent pericarditis, in-line with specialty biologics with Breakthrough Therapy and Orphan Drug designation.

- Helping to ensure patient affordability and access to treatment is one of our core principles and to this end, we offer a suite of programs to support affordability to eligible patients who are prescribed ARCALYST.

Access

- Kiniksa's goal is to enable rapid and broad access to ARCALYST for patients with Recurrent Pericarditis, CAPS, and DIRA.
- Payer mix for ARCALYST is largely **commercial (60%) and Medicare (25%)**.
- Early payer engagement has increased awareness of recurrent pericarditis and the differentiated value of ARCALYST (145 meetings and 24 clinical presentations)*
- **Kiniksa One Connect** is a personalized treatment support program for patients prescribed ARCALYST

Distribution

- ARCALYST is distributed through a closed network of **5 specialty pharmacies and the Veterans Affairs**.
- The distribution network for ARCALYST was developed to provide a high and consistent level of patient support with broad access. Network pharmacies provide customized services to support patients.

**Estimated through end of March*
CAPS = cryopyrin-associated periodic syndromes ; DIRA = deficiency of IL-1 receptor antagonist

Comprehensive Support for Patients Through Kiniksa One Connect

KINIKSA oneconnect™
support made simple.

The Patient Access Lead provides one-on-one support, including:

- ✓ Insurance coverage determination
- ✓ Explanation of benefits verification
- ✓ Assistance with prior authorizations and appeals
- ✓ Virtual or hybrid model 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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Closing Remarks

Sanj K. Patel

Chief Executive Officer and Chairman of the Board

Summary of ARCALYST Profit Share Arrangement with Regeneron[1]

ARCALYST Net Sales (CAPS + DIRA + Recurrent Pericarditis)[2]

Minus 100% of Cost of Goods Sold[3]

Minus 100% of Field Force Expenses

Minus Marketing & Commercial Expenses (Subject to Specified Limits)

Minus 100% of Regulatory & Certain Other Expenses

Calculated ARCALYST Operating Profit to be Shared

Minus 50% of Shared ARCALYST Operating Profit (Booked as a separate line item within Opex)

Minus R&D Expenses for Additional Indications or Other Studies Required for Approval

Minus Marketing & Commercial Expenses that Exceeded Specified Limits (if any)

Kiniksa Operating Income from ARCALYST

- Upfront payment: $5 million
- Regulatory milestones: $27.5 million in aggregate
- Kiniksa covers 100% of development expenses related to approval of additional indications
- In the U.S. and Japan, the initial license covers all indications other than CAPS[4], DIRA[5], oncology, and local application for eye and inner ear
- Kiniksa has rights to develop and commercialize ARCALYST in our field worldwide, with the exception of MENA[6]
- The BLA[7] for ARCALYST in CAPS transferred to Kiniksa following highly statistically significant Phase 3 clinical data
- The scope of the license expanded to include CAPS and DIRA in the U.S. and Japan upon the approval for recurrent pericarditis. Kiniksa is responsible for the sales and distribution of ARCALYST across all approved indications
- Profits on sales of ARCALYST will be equally split after deducting certain commercialization expenses subject to specified limits

1) Subject to description contained in definitive agreement; 2) Global net sales for CAPS, DIRA and recurrent pericarditis recognized as revenue on Kiniksa's income statement; 3) Including cost of product purchased from Regeneron as well as relevant Kiniksa overhead; 4) CAPS = Cryopyrin-Associated Periodic Syndromes; 5) DIRA = deficiency of the interleukin-1 receptor antagonist; 6) MENA = Middle East and North Africa; 7) BLA = Biologics License Application

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Portfolio of Four Immune-Modulating Assets

Program & Target	Preclinical	Phase 1	Phase 2	Phase 3	Commercial	Commercial Rights
ARCALYST[1] IL-1α & IL-1β	Recurrent Pericarditis					**Worldwide** (Excluding MENA)
	CAPS					**U.S. & Japan**
	DIRA					**U.S. & Japan**
Mavrilimumab[2] GM-CSFRα	Giant Cell Arteritis					**Worldwide**
	COVID-19 Pneumonia & Hyperinflammation					**Worldwide**
Vixarelimab[3] OSMRβ	Prurigo Nodularis					**Worldwide**
KPL-404 CD40	Severe Autoimmune Diseases					**Worldwide**

1) The FDA granted Breakthrough Therapy designation to ARCALYST for recurrent pericarditis in 2019 and Orphan Drug designation to ARCALYST for 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; IL-1α = interleukin-1α ; IL-1β = interleukin-1β; GM-CSFRα = granulocyte macrophage colony stimulating factor receptor alpha; OSMRβ = oncostatin M receptor beta; CAPS = Cryopyrin-Associated Periodic Syndromes; DIRA = deficiency of the interleukin-1 receptor antagonist; MENA = Middle East and North Africa

Q&A Session

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