KINIKSA

Every Second Counts!™

Rilonacept Analyst Day

September 29, 2020

Welcome
Mark Ragosa
Investor Relations and Finance

Agenda

Building Value at Kiniksa | *Sanj K. Patel, Chairman of the Board and CEO*

Burden of Recurrent Pericarditis| *Patient Video: Nadine's Story*

Recurrent Pericarditis Burden and Pathophysiology | *Paul Cremer, MD, Cleveland Clinic*

Review of Phase 3 RHAPSODY Data | *John F. Paolini, MD, PhD, Chief Medical Officer*

Pericarditis Epidemiology | *Matt Magestro, Value and Access*

Commercial Strategy | *Qasim Rizvi, MD, Chief Commercial Officer*

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

Q&A Session

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; potential value drivers; potential indications; potential market opportunities and competitive position; potential commercial launch strategy and pre-commercial activities for rilonacept in recurrent pericarditis; potential pricing of rilonacept; the potential for rilonacept to be the first approved product for recurrent pericarditis; potential safety and efficacy of our products; on-going, planned and potential clinical trials and other studies; timing and potential impact of clinical data; regulatory and other submissions, applications and approvals; 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 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 drug substance and/or drug product and conducting clinical trials, research and other studies; potential changes in our strategy, 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 other important factors are discussed under the caption "Risk Factors" in our Quarterly Report on Form 10-Q filed with the Securities and Exchange Commission (the "SEC") on August 4, 2020 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.

Building Value at Kiniksa

Sanj K. Patel

Chairman of the Board and CEO

Building Value at Kiniksa

Focus on strong biologic rationale and/or validated mechanisms

Acquire well-designed molecules aimed at various central control nodes of the immune system

Target pockets of unmet need that are ripe for innovation

Allocate capital across the portfolio relative to the opportunity

Build credibility through solid execution on communicated timelines

2015

Kiniksa incorporated

2016

Acquired **vixarelimab** from Biogen as pre-clinical stage asset

2017

Licensed **KPL-404** from Primatope as pre-clinical stage asset

Licensed rilonacept from Regeneron; approved in the U.S. for CAPS

Licensed **mavrilimumab** from MedImmune; developed through Phase 2b in RA

2018

Initiated Ph 2 trial of rilonacept in recurrent pericarditis

Kiniksa Initial Public Offering

Initiated Ph 2 trial of **mavrilimumab** in GCA

Reported interim Ph 2 rilonacept data in recurrent pericarditis

Initiated Ph 3 trial of rilonacept in recurrent pericarditis

2019

Kiniksa public offering and private placement

Acquired Primatope and IP related to **KPL-404**

Initiated Ph 2 trial of **vixarelimab** in diseases characterized by chronic pruritus

Initiated Ph 2 trial of **vixarelimab** in PN

Reported final Ph 2 rilonacept data in recurrent pericarditis

Initiated Ph 1 study of **KPL-404** in healthy volunteers

BTD granted for rilonacept in recurrent pericarditis

2020

Reported Ph 2a data from **vixarelimab** in PN

Reported data from **mavrilimumab** in COVID-19 pneumonia and hyperinflammation open-label treatment protocol

Reported Ph 2 data from **vixarelimab** in diseases characterized by chronic pruritus

Kiniksa public offering and private placement

Reported highly statistically significant Ph 3 rilonacept data in recurrent pericarditis

ODD granted for rilonacept in pericarditis

Initiated Ph 2/3 trial of **mavrilimumab** in COVID-19 pneumonia and hyperinflammation

Kiniksa public offering and private placement

ODD granted for **mavrilimumab** in GCA

RA = Rheumatoid Arthritis; GCA = Giant Cell Arteritis; PN = Prurigo Nodularis; BTD = Breakthrough Therapy designation; ODD = Orphan Drug designation

Product Candidates and Clinical Status

Disease Area	Program & Target	Preclinical	Phase 1	Phase 2	Phase 3	Status
Recurrent Pericarditis[1]	**Rilonacept[2]** IL-1α & IL-1β					Phase 3 Data Reported in Q3 2020
Giant Cell Arteritis	**Mavrilimumab** GM-CSFRα					Phase 2 Data Expected in Q4 2020
COVID-19 Pneumonia & Hyperinflammation	**Mavrilimumab** GM-CSFRα					Adaptive Design Phase 2/3 Initiated in Q3 2020
Prurigo Nodularis	**Vixarelimab** OSMRβ					Phase 2b Initiation Expected in Q4 2020
Severe Autoimmune Diseases	**KPL-404** CD40					Phase 1 Data Expected in Q4 2020

1) The FDA granted Breakthrough Therapy designation to rilonacept for recurrent pericarditis in 2019 and Orphan Drug designation to rilonacept for pericarditis in 2020; 2) Rilonacept (ARCALYST®) is approved and marketed for cryopyrin-associated periodic syndrome, in the United States by Regeneron Pharmaceuticals, Inc.; IL-1α = interleukin-1α ; IL-1β = interleukin 1β; GM-CSFRα = granulocyte macrophage colony stimulating factor receptor alpha; OSMRβ = oncostatin M receptor beta

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Rilonacept

Disease Area: Recurrent pericarditis[1]; painful and debilitating auto-inflammatory cardiovascular disease

Mechanism of Action[2]: IL-1α and IL-1β cytokine trap

Competition[3]: No FDA-approved therapies for recurrent pericarditis

Regulatory: U.S. Orphan Drug designation in pericarditis; Breakthrough Therapy designation in recurrent pericarditis

Status: sBLA being submitted to the FDA in recurrent pericarditis in 2020

Economics: 50/50 profit split on the approved indications in the U.S.

1) Rilonacept (ARCALYST®) is approved and marketed for cryopyrin-associated periodic syndrome (CAPS), in the United States by Regeneron Pharmaceuticals, Inc.; rilonacept in recurrent pericarditis is an investigational therapy; 2) Brucato et al. JAMA. 2016, 316 (18): 1906-1912; Arcalyst Prescribing Information; 3) Drugs@FDA: Arcalyst Prescribing Information, Ilaris Prescribing Information, Kineret Prescribing Information; Kaiser et al. Rheumatol Int (2012) 32:295–299; Theodoropoulou et al. Pediatric Rheumatology 2015, 13(Suppl 1):P155 ; Fleischmann et al, 2017 ACR/ARHP Abstract 1196; Kosloski et al, J of Clin Pharm 2016, 56 (12) 1582-1590; Cohen et al. Arthritis Research & Therapy 2011, 13:R125; Cardiel et al. Arthritis Research & Therapy 2010, 12:R192; Hong et al. Lancet Oncol 2014, 15: 656-666

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Burden of Recurrent Pericarditis
Nadine’s Story

Recurrent Pericarditis Disease Overview

Paul Cremer, MD

Cardiovascular Medicine, Cleveland Clinic

Associate Program Director,
Cardiovascular Training Program
Cardiovascular Imager
Cleveland Clinic Foundation

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Recurrent Pericarditis: The Need for Novel Therapies for a Debilitating Autoinflammatory Disease

Paul Cremer MD

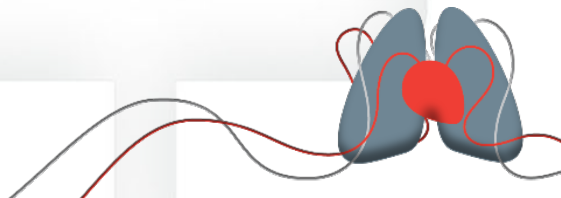
Associate Program Director,

Cardiovascular Training Program

Cardiovascular Imager

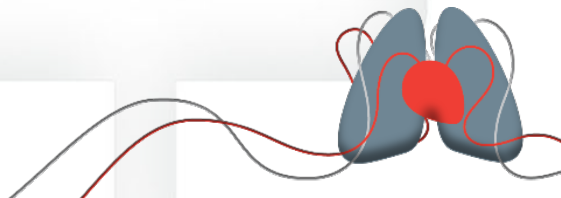
Cleveland Clinic Foundation

September 29, 2020



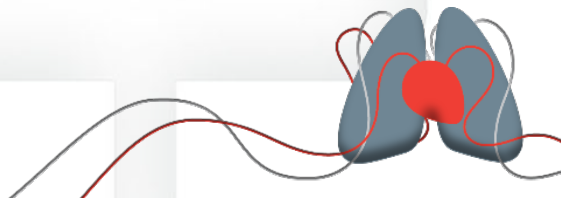
Disclosures

- Scientific advisory committee and consultation: Sobi Pharmaceuticals, Kiniksa Pharmaceuticals
- Core Imaging Lab: Cleveland Clinic, C5 Research
- The views expressed are my own and not necessarily those of the Cleveland Clinic

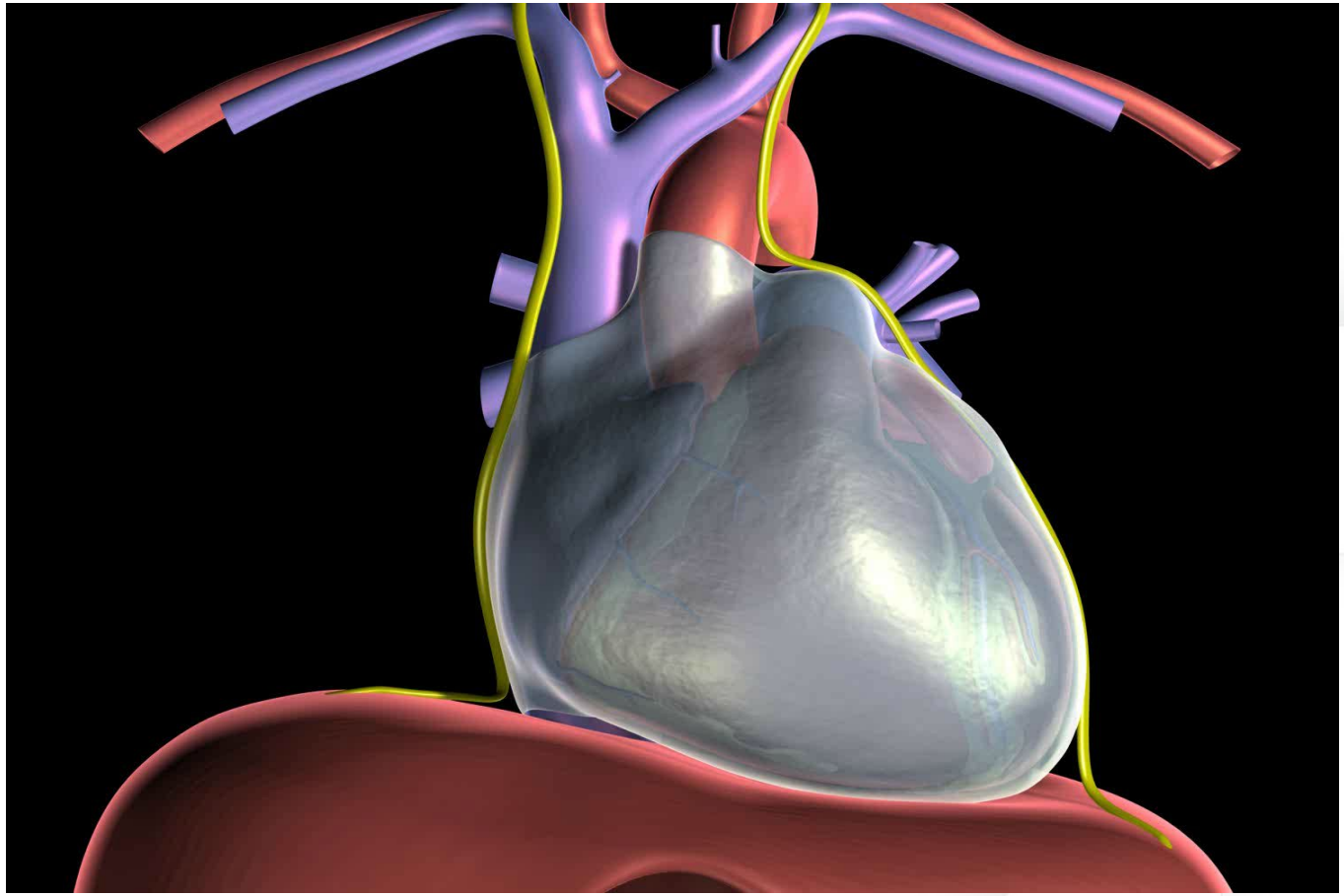


Overview

- Pericardial anatomy and histology
- Burden of disease
- Current treatment paradigm
- Pathophysiology of pericarditis (Role of IL-1)
- Need for targeted therapies

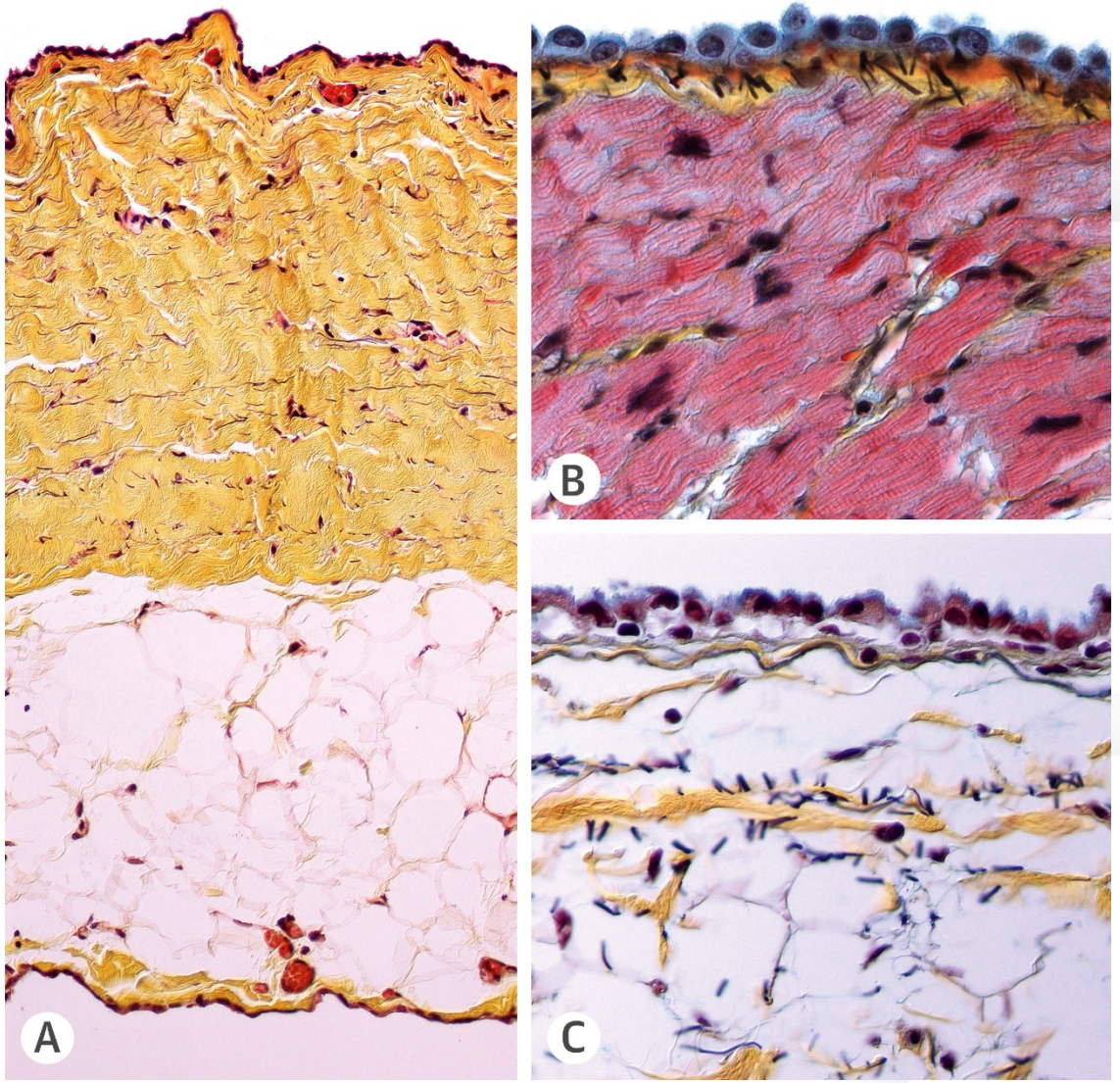


The Pericardium



Cremer et al. J Am Coll Cardiol 2016;68:2311-2328.

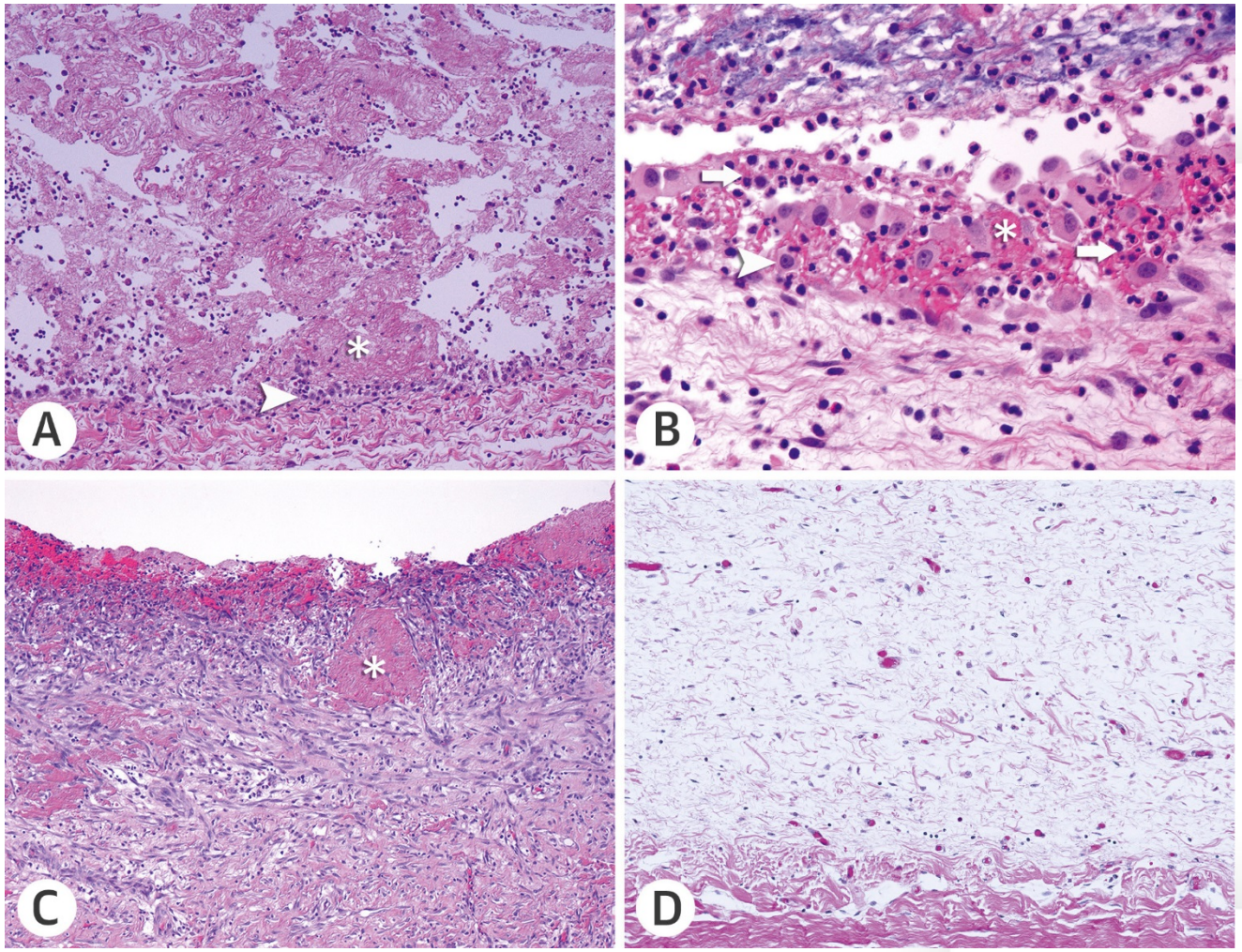
Normal Pericardial Histology



Cremer et al. J Am Coll Cardiol 2016;68:2311-2328.

Progression of Pericarditis (Histology)

1. Deposition of fibrinous material and recruitment of inflammatory cells



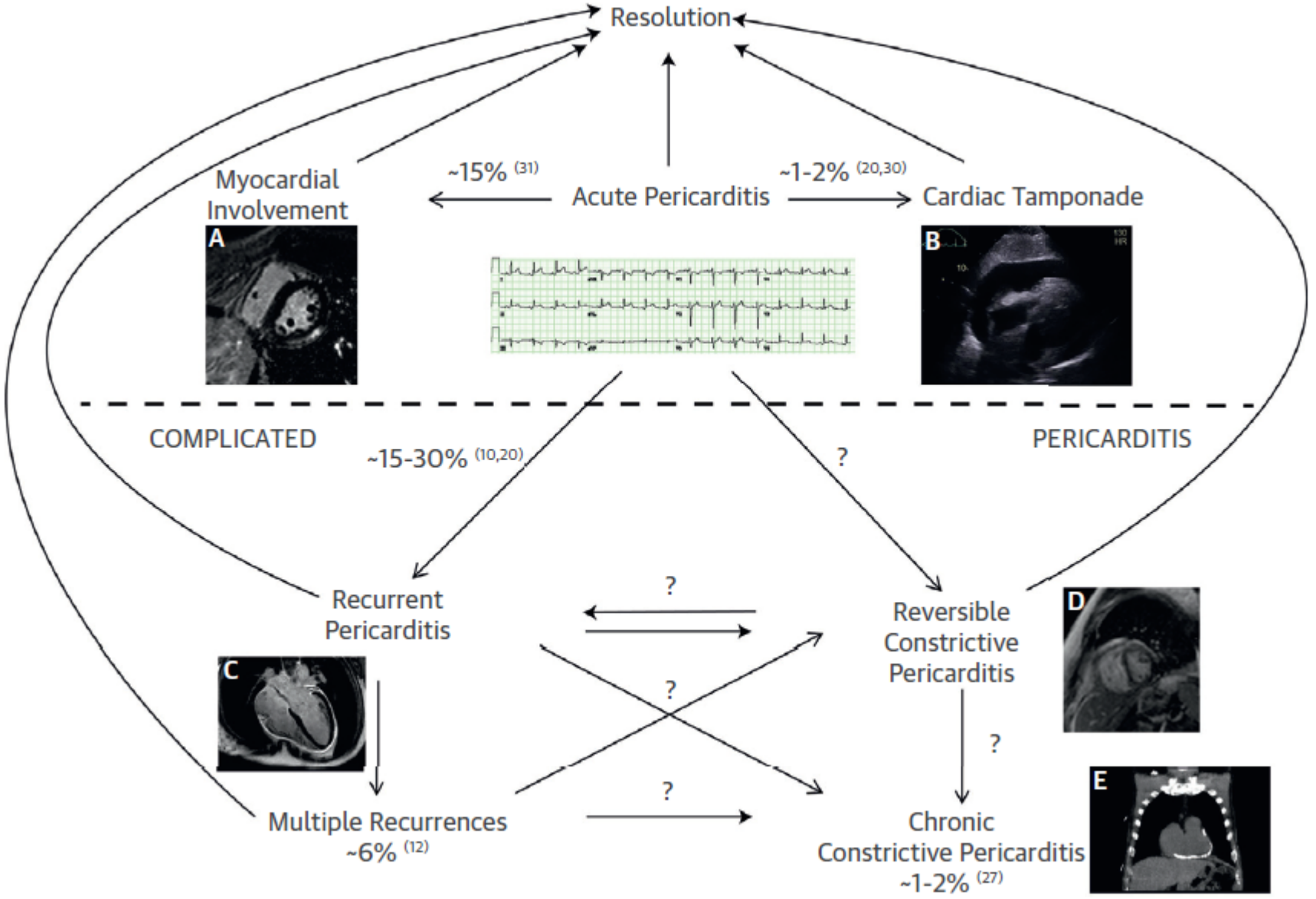
2. Disintegration of mesothelial cells

3. Organization with neo-vascularization and ingrowth of fibroblasts

4. Loose fibrous adhesions in healed pericarditis

Cremer et al. J Am Coll Cardiol 2016;68:2311-2328.

Who are the patients?

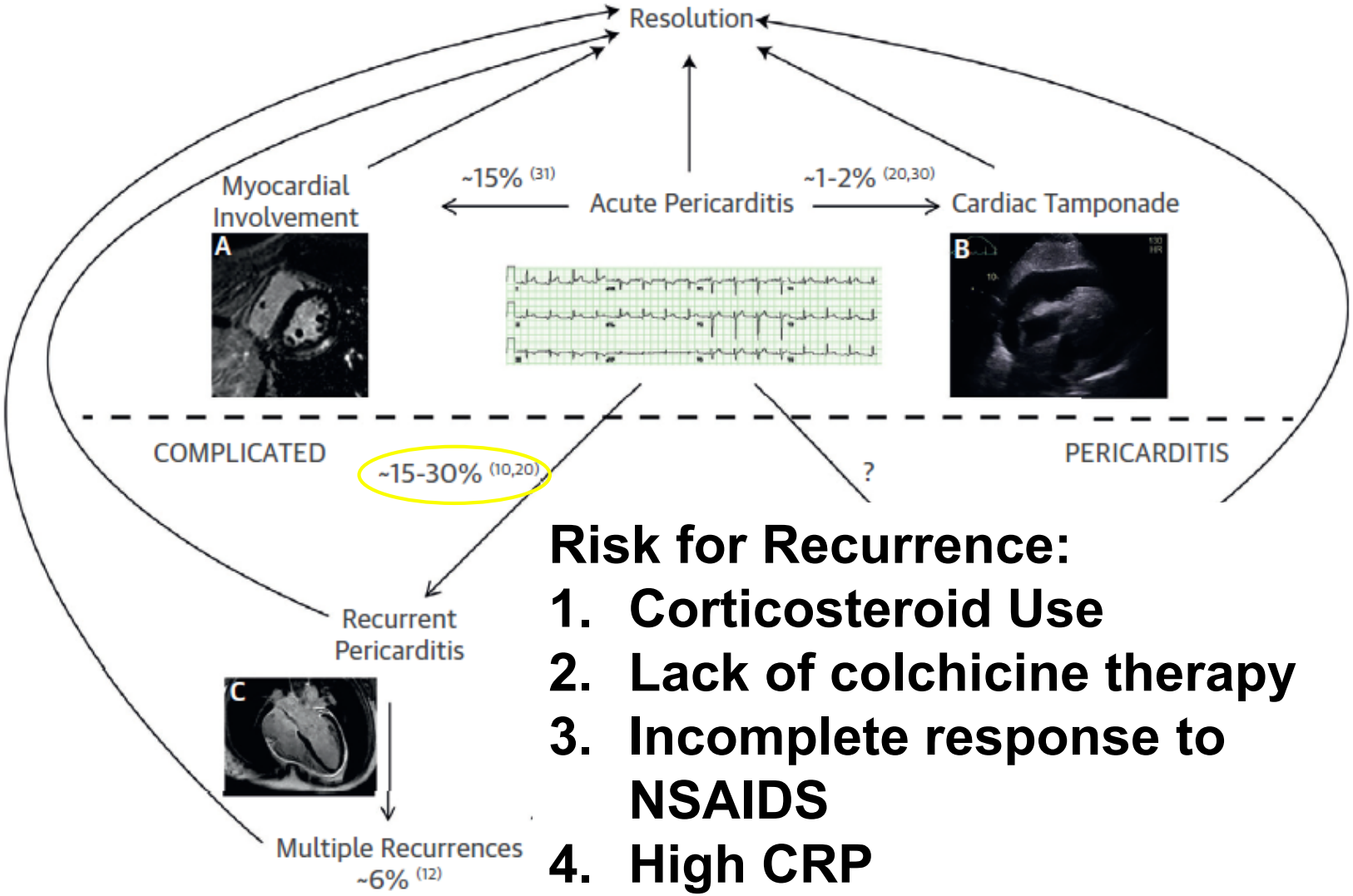


Cremer et al. J Am Coll Cardiol 2016;68:2311-2328.

Estimated Burden of Disease in US

- Acute pericarditis: 27.7 new cases per 100,000 per year
- Recurrent pericarditis: 15-30%
- Cardiac tamponade: 1-16%
- Constriction: 0.5-14.3%

Risks for Recurrence



Risk for Recurrence:

1. **Corticosteroid Use**
2. **Lack of colchicine therapy**
3. **Incomplete response to NSAIDS**
4. **High CRP**

Cremer et al. J Am Coll Cardiol 2016;68:2311-2328.

Treatment for Pericarditis

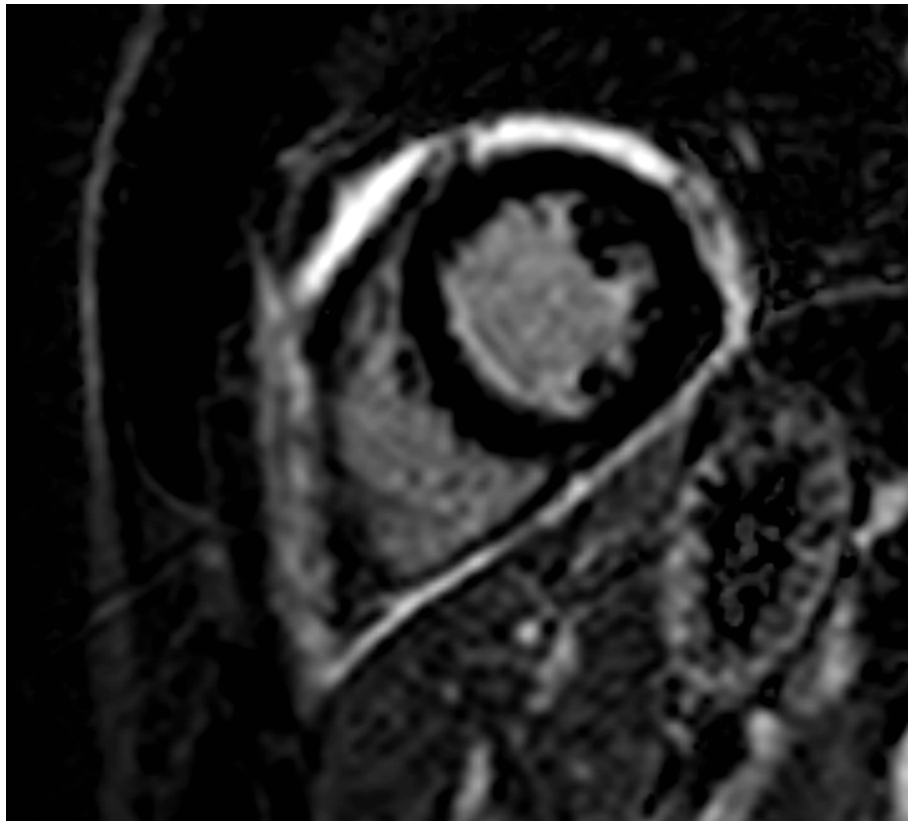
- Acute Pericarditis: NSAID (ibuprofen or ASA) for weeks and colchicine for 3 months
- First Recurrence: NSAID for weeks to months and colchicine for > 6 months
- Second Recurrence: NSAIDs, colchicine, prednisone, ? Steroid-sparing agent
- Third Recurrence: NSAIDs, colchicine, prednisone, steroid-sparing agent (azathioprine, methotrexate, IL-1 antagonist)

Define the phenotype to inform treatment and duration

Cremer et al. J Am Coll Cardiol 2016;68:2311-2328.

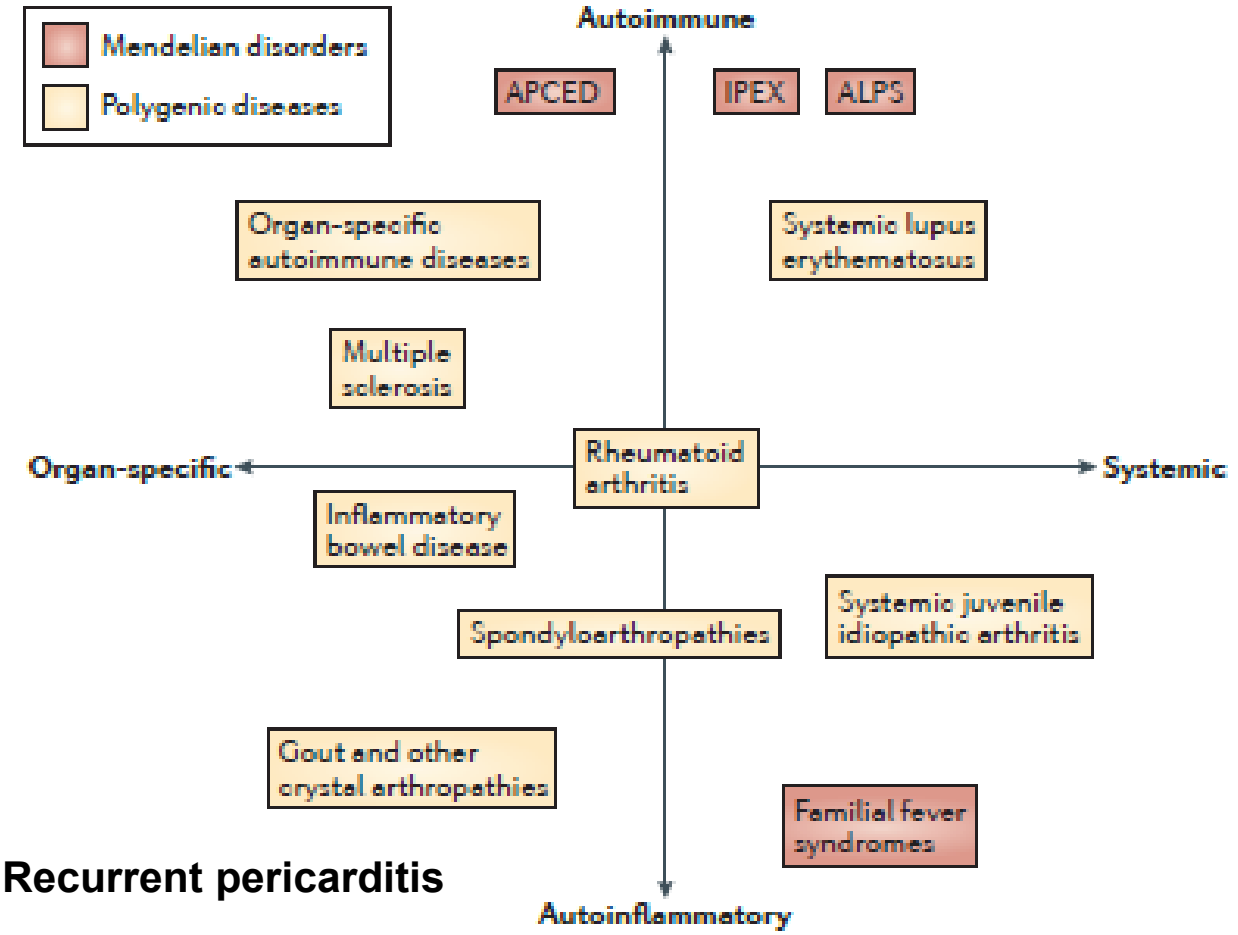
How much inflammation?

DHE



>12 months of therapy

AUGUST 2012 | VOLUME 12
www.nature.com/reviews/immunol



Recurrent pericarditis

- Autoinflammation is driven by endogenous danger signals and is perpetuated by inflammasome induced IL-1 and IL-18 production
- Autoimmunity involves activation of T and B cells and characterized by type I interferon signatures
- Efficacy of biologic agents is distinct

REPLY: Acute and Recurrent Pericarditis

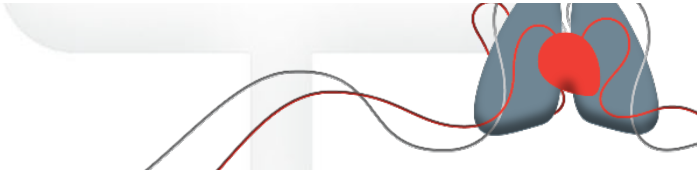
Still Idiopathic?

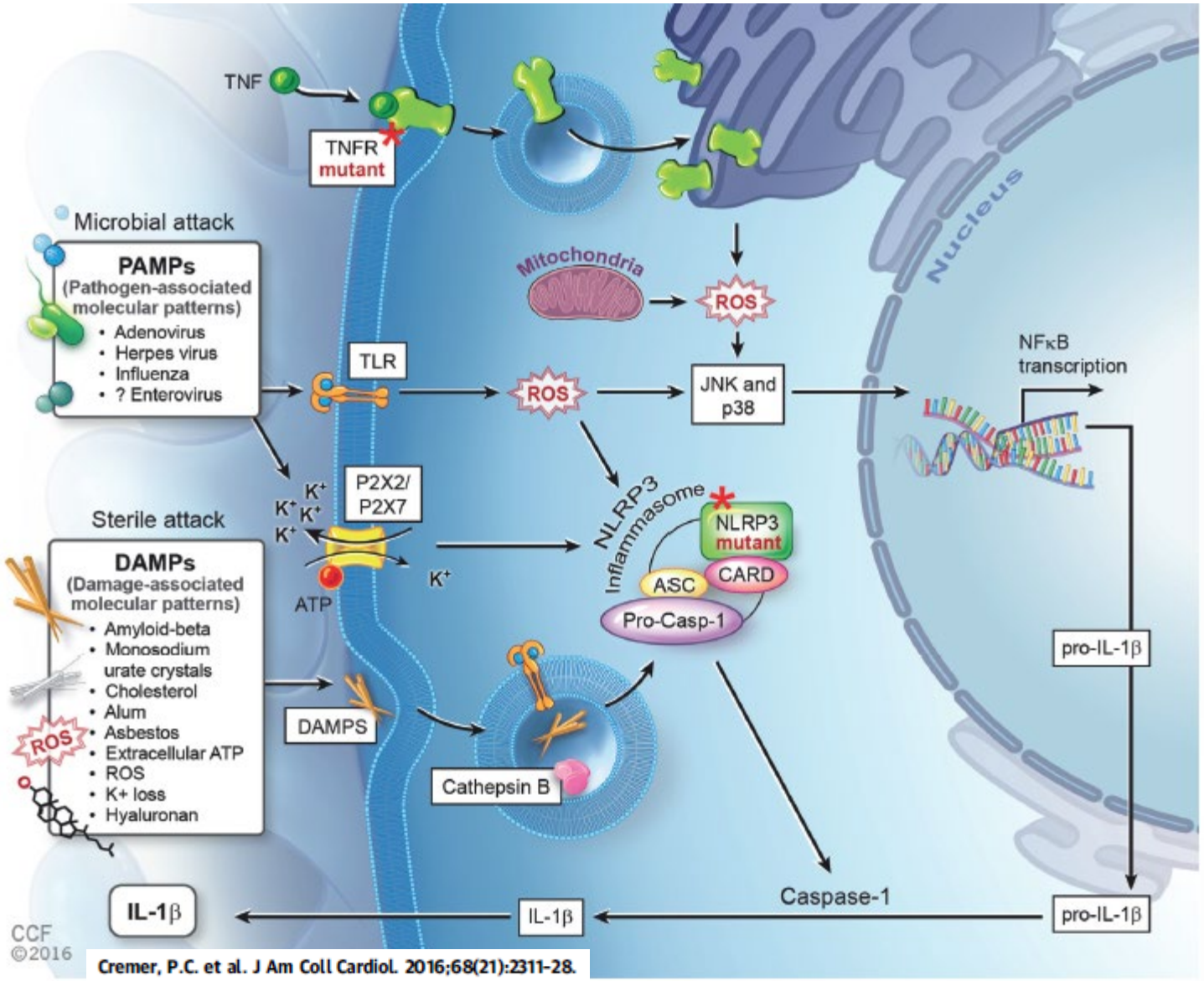
JACC VOL. 69, NO. 22, 2017
JUNE 6, 2017:2769–76

IM - REVIEW

Recurrent pericarditis: still idiopathic? The pros and cons of a well-honoured term

Antonio Brucato[1] · Massimo Imazio[2] · Paul C. Cremer[3] · Yehuda Adler[4] · Bernhard Maisch[5] · George Lazaros[6] · Marco Gattorno[7] · Alida L. P. Caforio[8] · Renzo Marcolongo[9] · Giacomo Emmi[10] · Alberto Martini[7,11] · Allan L. Klein[3]

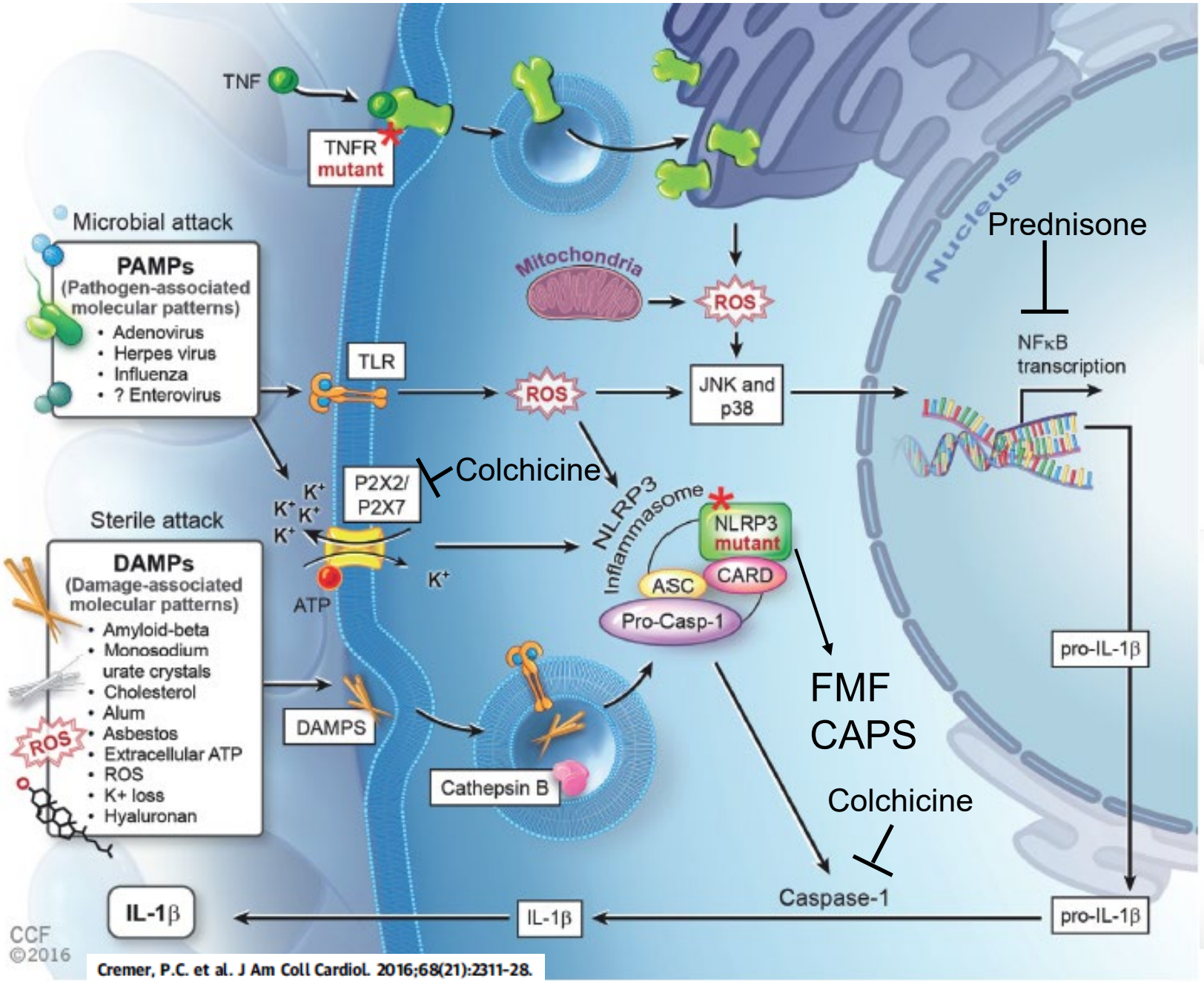




Cremer, P.C. et al. J Am Coll Cardiol. 2016;68(21):2311-28.

Activation of the Inflammasome

- Binding of PAMPs and DAMPs to receptors leads to translocation of NFk-B to the nucleus and transcription of pro-IL-1β and NLRP3 inflammasome
- Second signal recruits ASC to activate NLRP3 inflammasome
- NLRP3 inflammasome converts pro-Caspase to Caspase which in turn converts pro-IL-1β to IL-1β



Cremer, P.C. et al. J Am Coll Cardiol. 2016;68(21):2311-28.

Letters to the Editor

COLCHICINE FOR RECURRENT PERICARDITIS

SIR,—It is not known why pericarditis is self-limiting in some patients and recurrent in others. One suggestion is that an immune phenomenon may sometimes be involved. Recurrences may spread over many years, and they often follow withdrawal of anti-inflammatory drugs or dose reduction. The best treatment for recurrent pericarditis—once secondary causes such as malignant disease or renal insufficiency[1,2] have been eliminated—is not known. A non-steroidal anti-inflammatory drug, especially aspirin, is the first choice but corticosteroids are often used when recurrences become frequent or severe. If recurrences persist despite steroids, or when excessive and prolonged doses are needed, there are few alternatives. Some recommend a pericardiectomy,[3,4] and success with immunosuppressive drugs has been reported.[5]

Three patients, two men and one woman aged 28, 34, and 41, with recurrent pericarditis that was idiopathic in two and due to systemic lupus erythematosus in one were on high dose steroids (60 mg daily) but had frequent recurrences when the dose was reduced below 20 mg daily. Pericardiectomy or immunosuppressive therapy was being considered. On the basis of the reported efficacy of colchicine in the recurrent polyserositis seen in familial Mediterranean fever[6,7] this drug was tried at a dose of 1 mg per day. The response was spectacular: the patients have been recurrence-free for 15, 24 and 36 months, and steroid treatment was withdrawn 2 months after colchicine was started. The maintenance dose of colchicine has been reduced to 0·5 mg daily. No side-effects have been observed.

These encouraging results should be taken with caution since the treatment has been tried in only three patients so far.

A. RODRÍGUEZ DE LA SERNA
J. GUINDO SOLDEVILA
V. MARTÍ CLARAMUNT
A. BAYÉS DE LUNA

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Hospital de la Santa Creu i Sant Pau,
08025 Barcelona, Spain

Why colchicine?

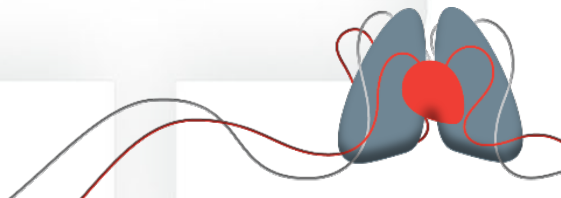
"On the basis of reported efficacy of colchicine in the recurrent polyserositis seen in familial Mediterranean fever"

Efficacy of Intermittent Colchicine Therapy in Familial Mediterranean Fever

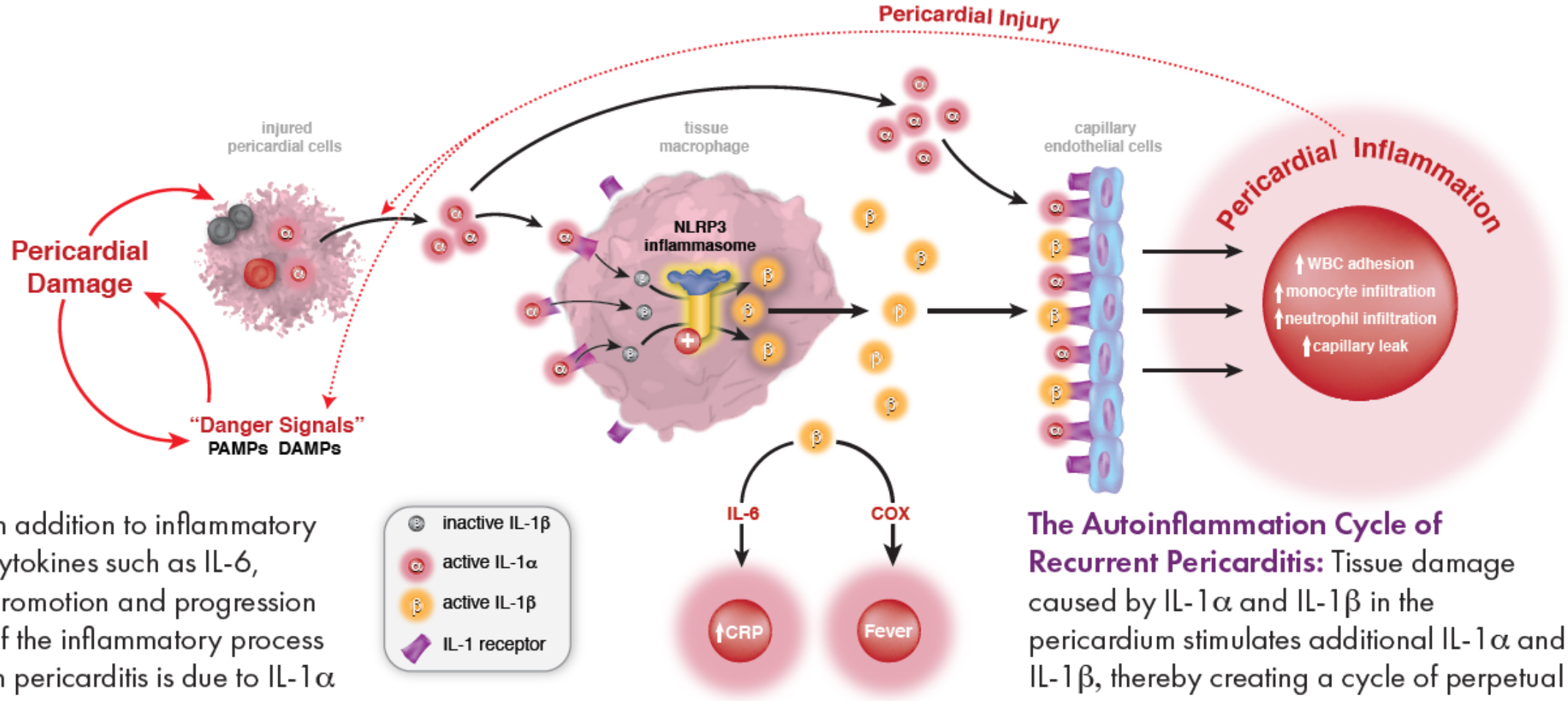
DANIEL G. WRIGHT, M.D.; SHELDON M. WOLFF, M.D., F.A.C.P.; ANTHONY S. FAUCI, M.D.; and DAVID W. ALLING, M.D., Ph.D.; Bethesda, Maryland

Annals of Internal Medicine 86:162-165, 1977

"Patients can recognize the prodrome of their FMF attacks and some patients can consistently abort their attacks with short course of colchicine"



Role of IL-1α and IL-1β in Autoinflammatory Cycle of Recurrent Pericariditis

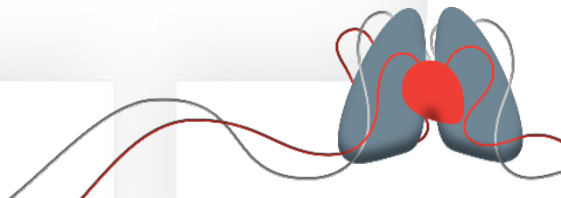


In addition to inflammatory cytokines such as IL-6, promotion and progression of the inflammatory process in pericarditis is due to IL-1α and IL-1β

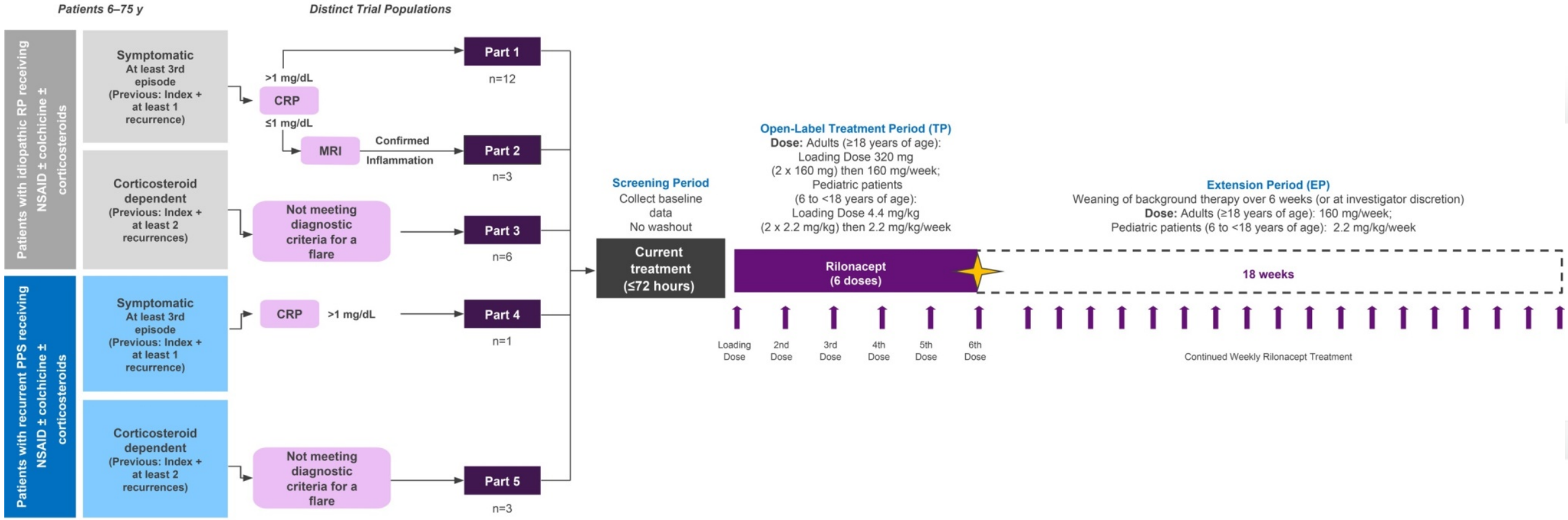
The Autoinflammation Cycle of Recurrent Pericarditis: Tissue damage caused by IL-1α and IL-1β in the pericardium stimulates additional IL-1α and IL-1β, thereby creating a cycle of perpetual pericardial inflammation

Targeted Therapies

- Is this an autoinflammatory phenotype?
  - Recurrences characterized by elevated CRP and systemic symptoms
    - IL-1 antagonism to avoid prolonged course of corticosteroids
  - Recurrences occur with tapering of corticosteroids
    - IL-1 antagonism to facilitate tapering of corticosteroids

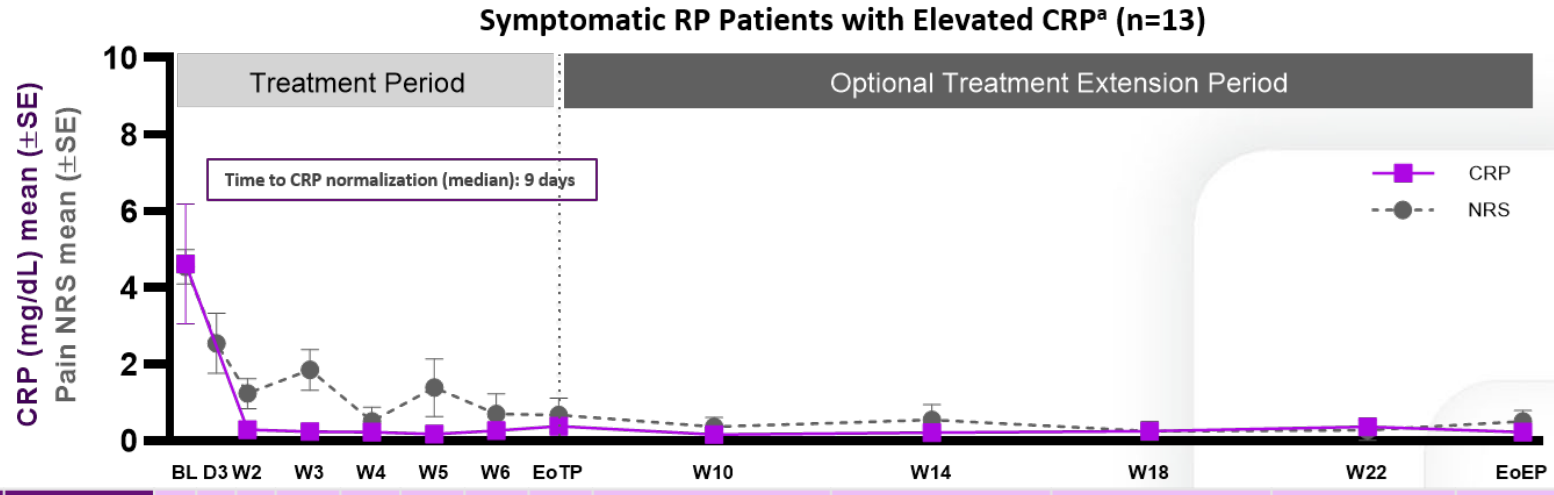


Efficacy and Safety of Rilonacept for Recurrent Pericarditis: Results from a Phase II Clinical Trial



- 25 patients (active recurrence or corticosteroid-dependent without active recurrence)
- Primary outcome: decrease in pain and CRP, disease activity after corticosteroid taper

Efficacy and Safety of Rilonacept for Recurrent Pericarditis: Results from a Phase II Clinical Trial

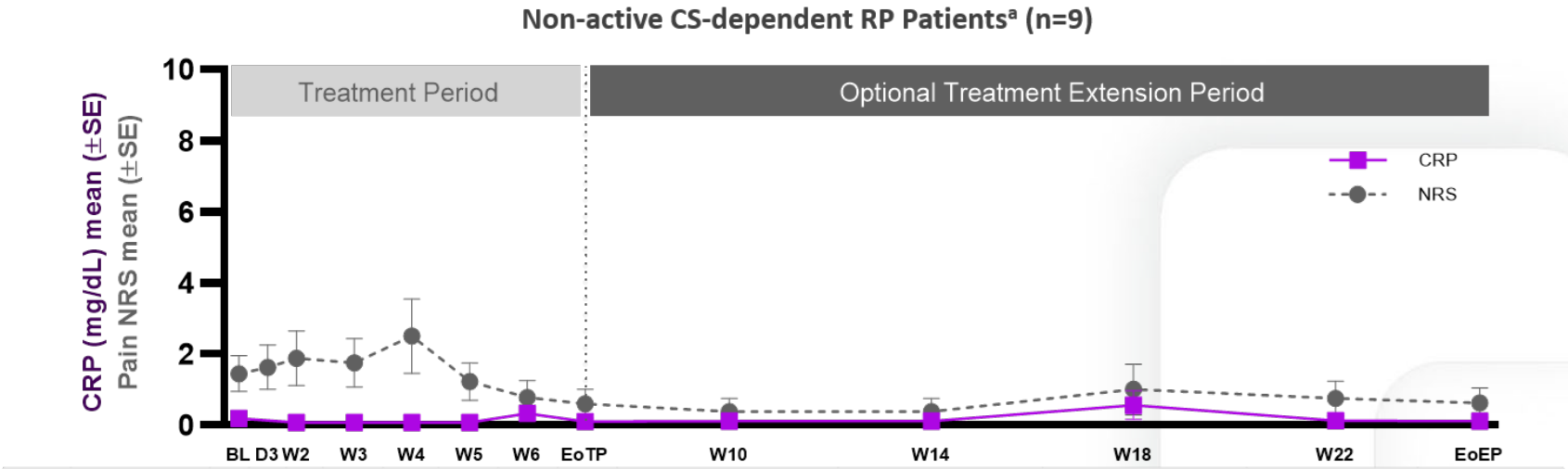


		BL	D3	W2	W3	W4	W5	W6	EoTP	W10	W14	W18	W22	EoEP
CRP	# of Patients	13		11	13	8	12	7	12	11	10	7	11	12
	CRP Mean	4.62		0.29	0.23	0.22	0.17	0.27	0.38	0.16	0.22	0.25	0.37	0.22
Pain NRS	# of Patients	13	11	13	13	8	13	10	9	11	11	8	11	12
	Pain Mean	4.5	2.5	1.2	1.8	0.5	1.4	0.7	0.7	0.4	0.5	0.3	0.3	0.5

[a]Parts 1 and 4 combined. Patients with elevated CRP and symptomatic disease (Parts 1 and 4) are most representative of real-world recurrent pericarditis. Inclusion and exclusion criteria for the ongoing Phase 3 study RHAPSODY align with this patient population (clinicaltrials.gov/NCT03737110).
EoTP, end of treatment period; EoEP, end of extension period;

Median time to CRP normalization: 9 days

Efficacy and Safety of Rilonacept for Recurrent Pericarditis: Results from a Phase II Clinical Trial



		BL	D3	W2	W3	W4	W5	W6	EoTP	W10	W14	W18	W22	EoEP
CRP	# of Patients	9		8	9	5	9	5	9	7	8	4	8	8
	CRP Mean	0.19		0.07	0.07	0.07	0.07	0.33	0.09	0.10	0.11	0.56	0.12	0.12
Pain NRS	# of Patients	9	8	8	8	4	9	9	5	8	8	4	8	8
	Pain Mean	1.4	1.6	1.9	1.8	2.5	1.2	0.8	0.6	0.4	0.4	1.0	0.8	0.6

[a]Part 3 and Part 5 combined
EoTP, end of treatment period; EoEP, end of extension period;

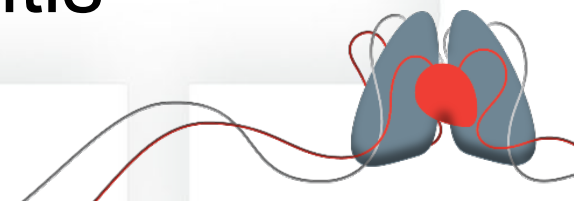
- 11 patients discontinued corticosteroids, 2 tapered
- No recurrences during follow-up

Efficacy and Safety of Rilonacept for Recurrent Pericarditis: Results from a Phase II Clinical Trial

- Mean quality of life scores (PROMIS) improved: physical 39.9 to 51.3, mental health 44.5 to 50.5
- Pericardial effusion improved in 6 of 7 patients
- Among 8 patients with pericardial DHE, improved or resolved in 6 patients
- Annualized incidence of pericarditis recurrence from 3.9 to 0.18 episodes per year before and after Rilonacept

Conclusions

- Recurrent pericarditis as an autoinflammatory disease
- Patients develop debilitating recurrences
- IL-1 has been shown to play a key role in driving inflammation
  - Corticosteroids are non-specific and have numerous adverse effects
  - Targeted immunomodulatory approaches blocking IL-1 signaling show promise in recurrent pericarditis



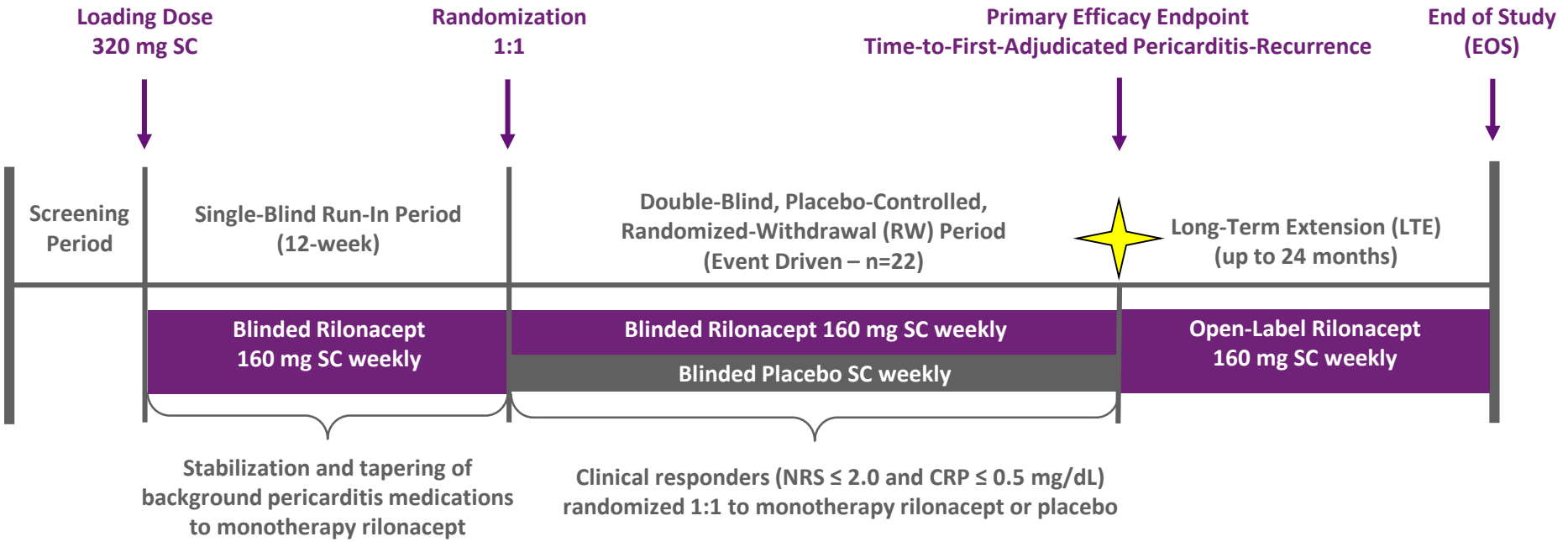
Review of Phase 3 RHAPSODY Data

John F. Paolini, MD PhD

Chief Medical Officer

The Phase 3 RHAPSODY data were previously posted on the Kiniksa website on June 29, 2020

Design of Pivotal Phase 3 Trial of Rilonacept 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 A et al. Am Heart J. 2020 Oct;228:81-90

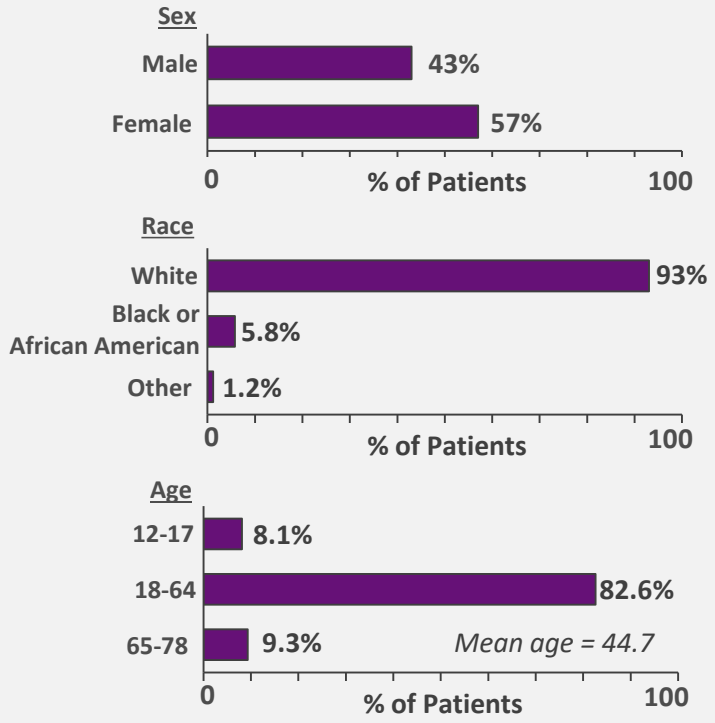
The Phase 3 RHAPSODY data were previously posted on the Kiniksa website on June 29, 2020

Baseline Demographics and Clinical Characteristics

Pivotal Phase 3 Rilonacept Data

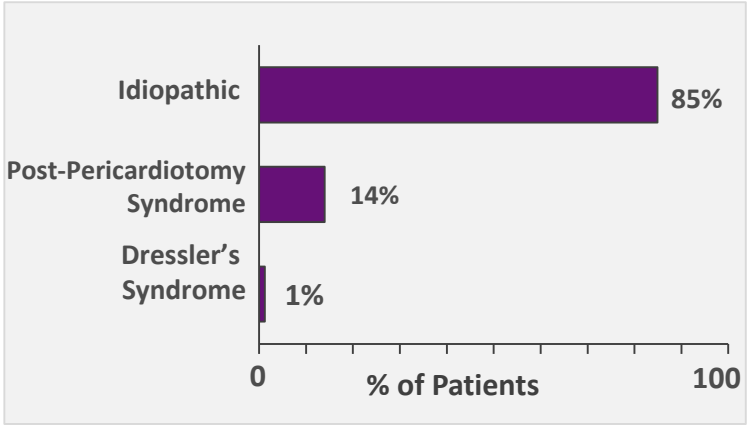
RHAPSODY

Baseline Demographics (n=86)

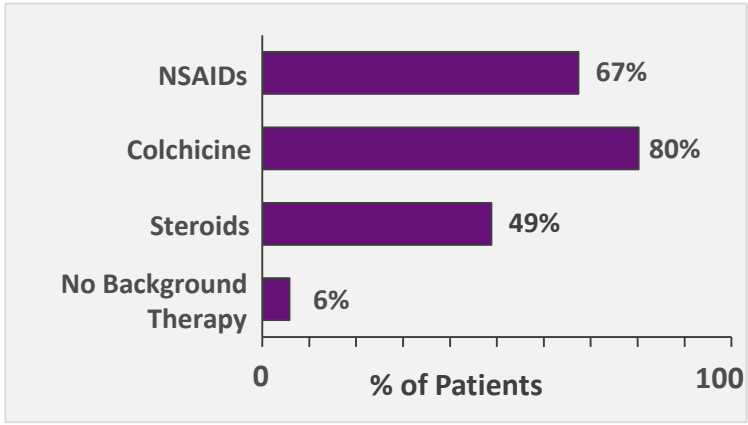


Total Number of Episodes Including Index and Qualifying Episodes	Run-in Period (n=86)
Mean	4.7

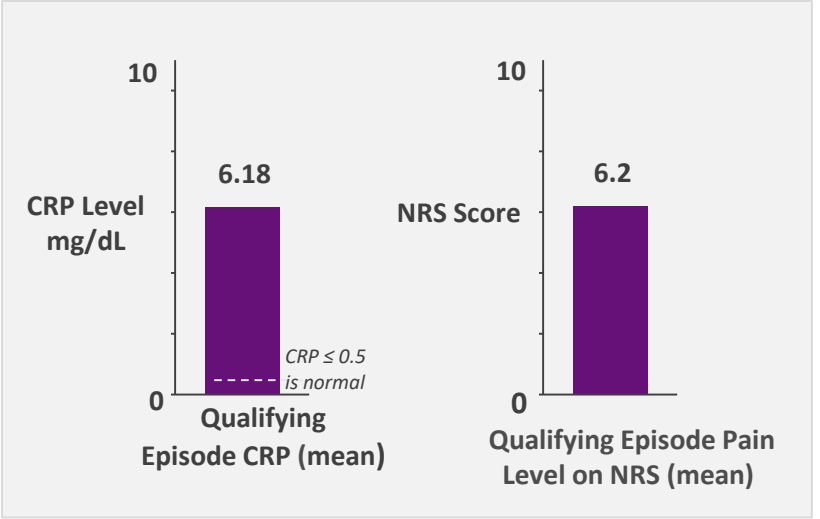
Prior Pericarditis History at Baseline (n=86)



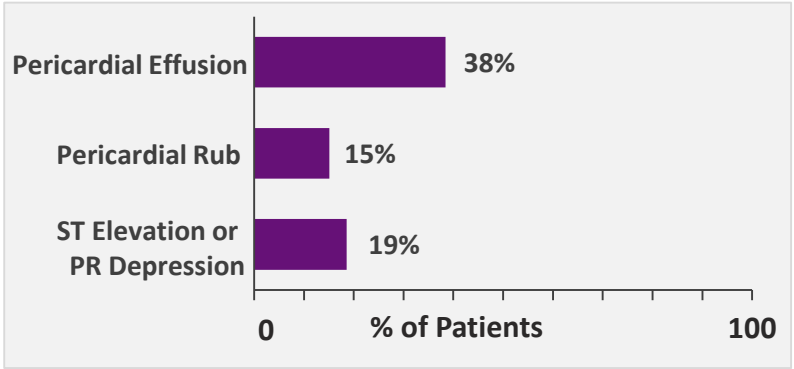
SoC Received at Qualifying Episode (n=86)



Qualifying Episode CRP & NRS (n=86)



Pericarditis Manifestations at Qualifying Episode (n=86)



CRP = C-reactive protein; NRS = Numerical Rating Scale; SoC = Standard of Care; NSAIDs = nonsteroidal anti-inflammatory drugs;
The Phase 3 RHAPSODY data were previously posted on the Kiniksa website on June 29, 2020

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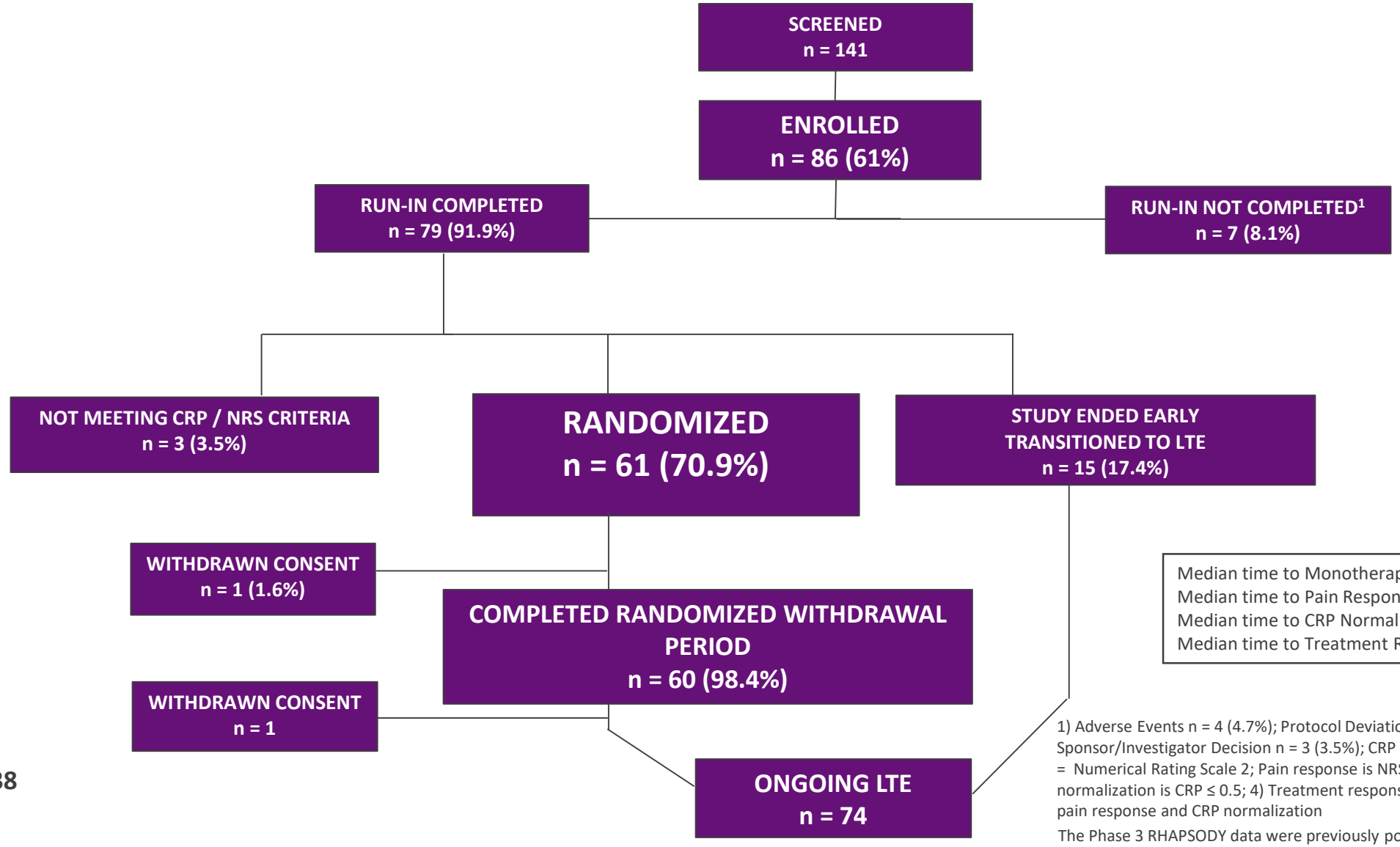
Subject Disposition

Pivotal Phase 3 Rilonacept Data



Median time to Monotherapy Rilonacept = 7.9 weeks (7.0, 8.1)
Median time to Pain Response[2] = 5.0 days (4.0, 6.0)
Median time to CRP Normalization[3] = 7.0 days (5.0, 8.0)
Median time to Treatment Response[4] = 5.0 days (4.0, 7.0)

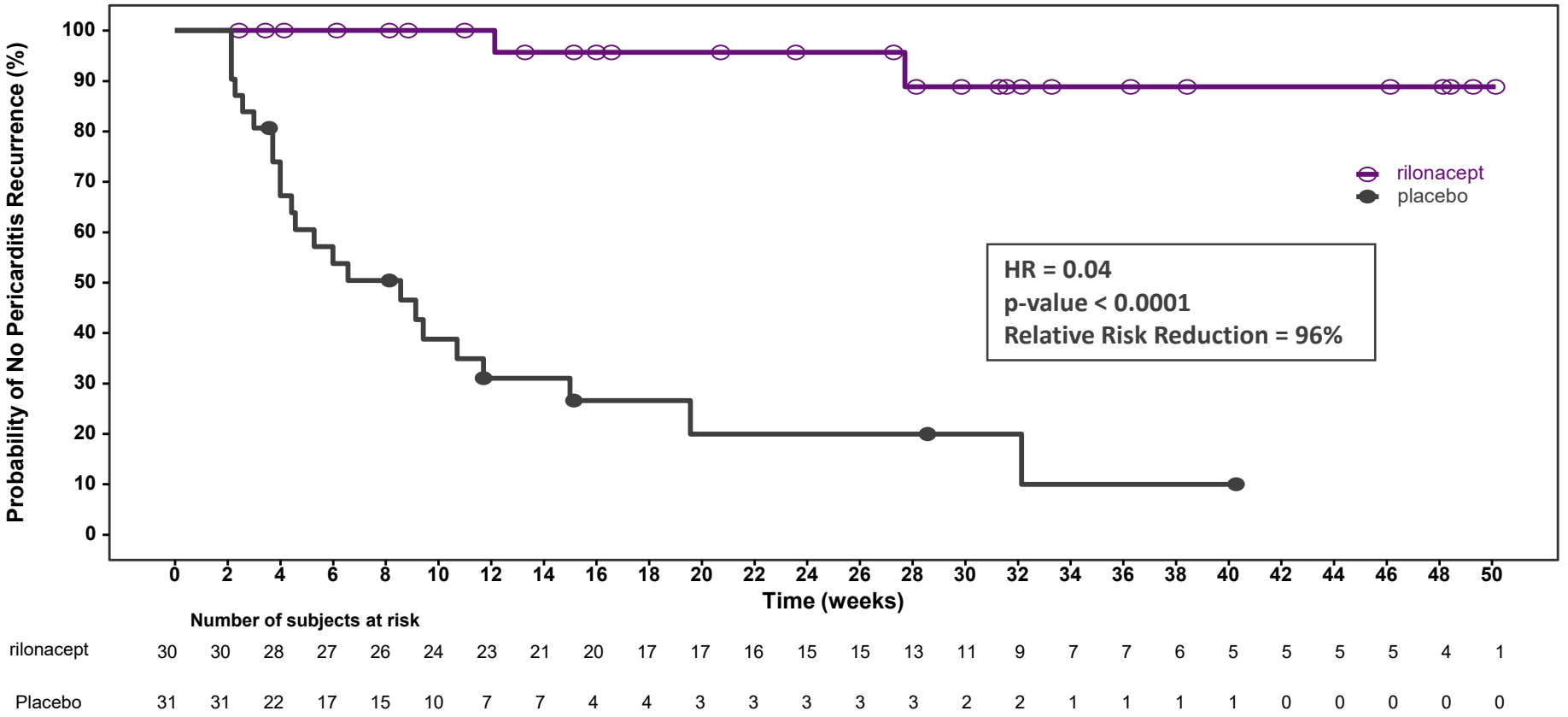
1) Adverse Events n = 4 (4.7%); Protocol Deviation /Withdrawn Consent / Sponsor/Investigator Decision n = 3 (3.5%); CRP = C-reactive protein; NRS = Numerical Rating Scale 2; Pain response is NRS ≤ 2; 3) CRP normalization is CRP ≤ 0.5; 4) Treatment response is the combination of pain response and CRP normalization

The Phase 3 RHAPSODY data were previously posted on the Kiniksa website on June 29, 2020

Primary Efficacy Endpoint: Time-to-First Adjudicated Pericarditis Recurrence

Pivotal Phase 3 Rilonacept Data

RHAPSODY



Pericarditis Recurrence Categories, n (%)	Rilonacept (N=30)	Placebo (N=31)
Number of Subjects with Events (Adjudicated Pericarditis Recurrence), n(%)	2 (6.7)	23 (74.2)
Time to First Adjudicated Pericarditis Recurrence; Median, 95% CI (Weeks)	NE (NE, NE)	8.6 (4.0, 11.7)

NE = Not Estimable

The Phase 3 RHAPSODY data were previously posted on the Kiniksa website on June 29, 2020

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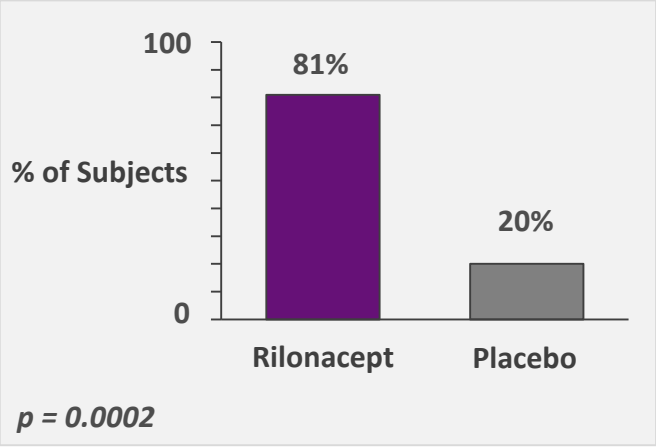
Secondary Endpoints at Week 16 of the Randomized Withdrawal Period

Pivotal Phase 3 Rilonacept Data

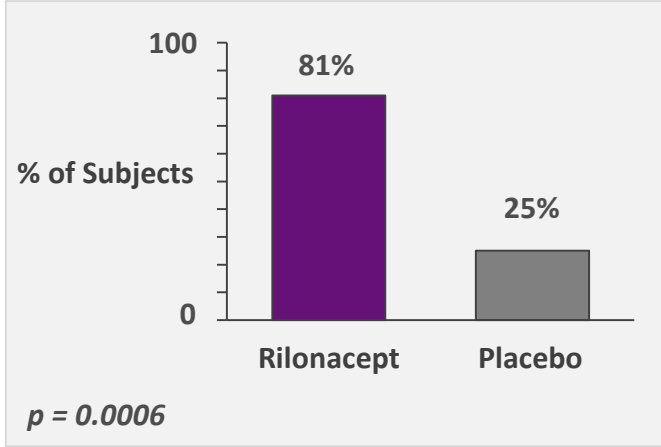
RHAPSODY

Proportion of Subjects Who Maintained Clinical Response [1]



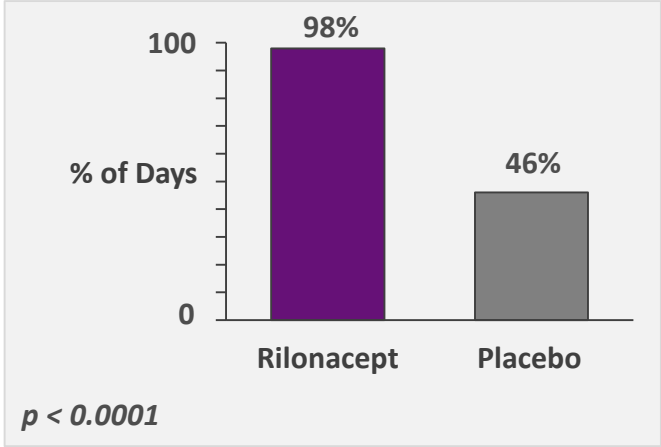
Data at Weeks 8 and 24 were consistent and statistically significant
(Week 8, p < 0.0001; Week 24, p=0.0022)

Proportion of Subjects with Absent/Minimal Pericarditis Symptoms based on the 7-point PGIPS [2]



Data at Weeks 8 and 24 were consistent and statistically significant
(Week 8, p < 0.0001; Week 24, p=0.0002)

Percent of Days with No or Minimal Pain in First 16 Weeks (ITT Week 16) [3]



Data at Weeks 8 and 24 were consistent and statistically significant
(Week 8, p < 0.0001; Week 24, p < 0.0001)

1) Clinical Response is defined as a weekly average of daily pericarditis pain of ≤2.0 on the 11-point NRS, CRP level ≤0.5 mg/dL, and on monotherapy of randomized study drug in that week. Subjects who had recurrence, or used bailout rilonacept, or used rescue medication, discontinued double-blinded treatment, or lost to follow-up before the week will be considered as non-responders;
2) PGIPS = Patient Global Impression of Pericarditis Severity baseline;
3) No or minimal pain is defined as non-missing daily NRS ≤ 2. The percentage of days with no or minimal pain in the first 24, 16, and 8 weeks is calculated for each subject using 24x7, 16x7, 8x7, respectively, as the denominator. Missing values in pain diary will be counted as 0 day with no or minimal pain. On days of using ORT or corticosteroid, count as 0 day with no or minimal pain. If bailout rilonacept was used, each administration (loading dose or not) will be counted as 7 days without qualifying no or minimal pain.

The Phase 3 RHAPSODY data were previously posted on the Kiniksa website on June 29, 2020

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Summary of Adverse Events

Pivotal Phase 3 Rilonacept Data

RHAPSODY

	Run-In Period	Randomized Withdrawal Period	
Category [1]	Rilonacept (N=86) n (%)	Rilonacept Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)
All Adverse Events	69 (80.2)	24 (80.0)	13 (41.9)
TEAEs [2]	69 (80.2)	24 (80.0)	13 (41.9)
TEAEs by Maximum severity [3]			
Mild	52 (60.5)	16 (53.3)	9 (29.0)
Moderate	15 (17.4)	8 (26.7)	4 (12.9)
Severe	2 (2.3)	0	0
Drug-Related TEAEs [4]	46 (53.5)	10 (33.3)	1 (3.2)
Serious TEAEs (SAE)[5]	1 (1.2)	1 (3.3)	1 (3.2)
TEAEs Leading to Death	0	0	0
Drug-Related SAE [4]	0	0	0
TEAEs Leading to Dose Interruption	0	1 (3.3)	0
TEAEs Leading to Study Drug Discontinuation	4 (4.7)[6]	0	0
TEAEs of Special Interest (Malignancy) [7]	0	1 (3.3)	0
TEAEs of Injection Site Reaction	28 (32.6)	6 (20.0)	0
TEAEs of Injections and Infestations	14 (16.3)	12 (40.0)	3 (9.7)

	Run-In Period	Randomized Withdrawal Period	
Category [1]	Rilonacept (N=86) n (%)	Rilonacept Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)
Bronchitis	0	1 (3.3)	0
Conjunctivitis	0	1 (3.3)	0
Ear infection	0	0	0
Gastroenteritis	0	0	1 (3.2)
Gastroenteritis viral	0	0	0
Gastroenteritis viral infection	0	1 (3.3)	1 (3.2)
Hordeolum	1 (1.2)	0	0
Influenza	1 (1.2)	0	1 (3.2)
Nasopharyngitis	6 (7.0)	2 (6.7)	0
Oral herpes	1 (1.2)	1 (3.3)	0
Otitis media	0	1 (3.3)	0
Pharyngitis	1 (1.2)	0	0
Pharyngitis streptococcal	0	0	0
Rhinitis	1 (1.2)	0	0
Sinusitis	1 (1.2)	3 (10.0)	0
Subcutaneous abscess	1 (1.2)	0	0
Upper respiratory tract infection	2 (2.3)	1 (3.3)	0
Urinary tract infection	1 (1.2)	3 (10.0)	0
Vaginal infection	0	1 (3.3)	0
Viral upper respiratory tract infection	2 (2.3)	1 (3.3)	0

1) Subjects with multiple events are counted once in the same category; 2) A Treatment-emergent adverse events (TEAEs) are defined as AEs that start or increase in severity on or after the date of first dose and before 6 weeks after the last dose of study drug; 3) Each subject has only been represented with the maximum severity; 4) Related or possibly related or missing, as assessed by the investigator; 5) SAEs (all unrelated to study drug) - Run in Period: CVA (carotid dissection); RW Period: Chest fluttering after alcohol (on PBO), and Pyrexia, Squamous cell Carcinoma, and post-operative ileus (on rilonacept); 6) alopecia, allergic alveolitis (related to other factors), erythema, and systemic allergic reaction (hypersensitivity); 7) Includes malignancy, excluding basal cell carcinoma of the skin

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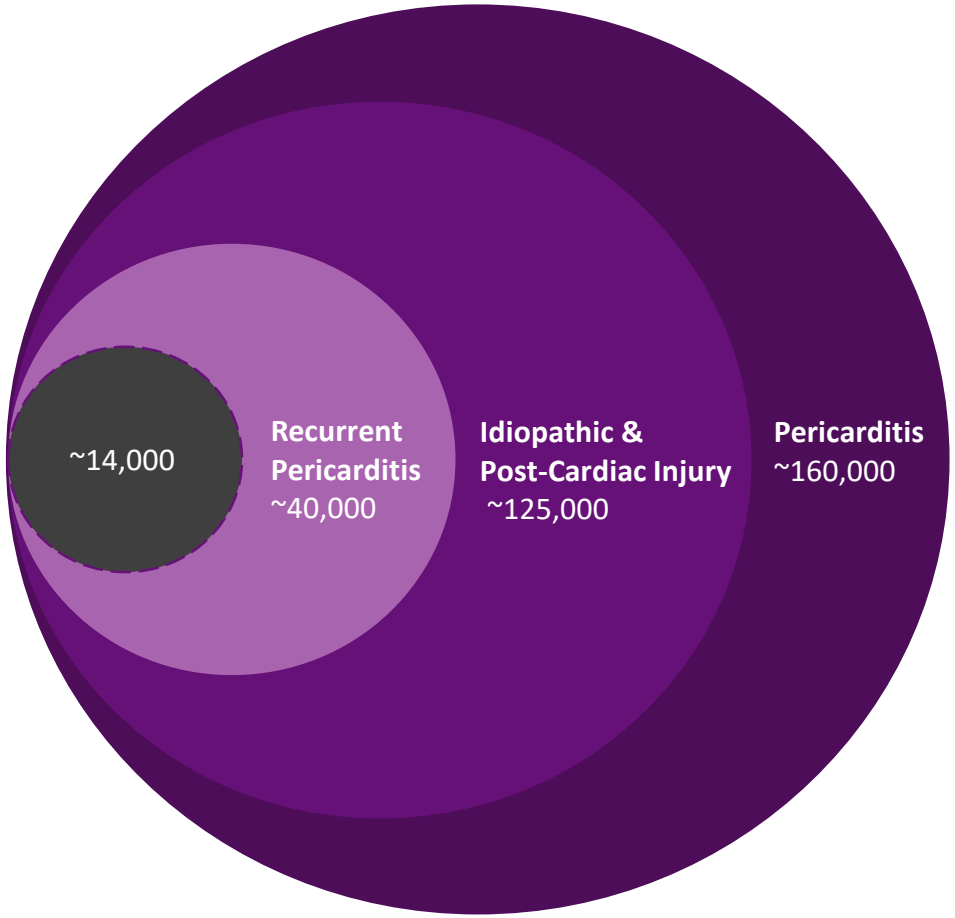
The Phase 3 RHAPSODY data were previously posted on the Kiniksa website on June 29, 2020

Pericarditis Epidemiology

Matt Magestro

Value and Access

Pericarditis Epidemiology



All figures annual period prevalence

Approximately 14,000 recurrent pericarditis patients suffer from persistent underlying disease, with multiple recurrences and inadequate response to conventional therapy[1]

- **~ 160,000:** Epidemiological analysis using large national surveillance databases to calculate the pooled annualized prevalence of pericarditis
  (Basis for Orphan Drug Designation approval)[2]
- **~125,000:** Approximately 75-80% are considered idiopathic (thought to be post-viral) and post cardiac injury[3-5]
- **~40,000:** Up to 30% experience at least one recurrence; some recur over multiple years[6,7]
- **~14,000:** Nearly 50% annual turnover with ~7,000 patients coming into the pool each year[8]

1) Cremer et al. American Journal of Cardiology. 2016;2311-2328; 2) DOF, Kiniksa Pharmaceuticals, Ltd.; 3) Brucato A, Maestroni S, Cumetti D, et al. Autoimmun Rev. 2008; 8:44-47; 4) Lange R, Hills L. N Engl J Med. 2004; 351: 2195-2202; 5) Imazio M, Cecchi E, Demichelis B, et al. Circulation. 2007; 115: 2739-2744; 6) Imazio et al. Circulation. 2005;112:2012-2016; 7) Adler et al. Circulation. 1998;97:2183-2185; 8) DOF, Kiniksa Pharmaceuticals, Ltd.

Commercial Strategy

Qasim Rizvi, MD

Chief Commercial Officer

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

Refractory[1,2]	Patient fails to respond, or is intolerant, to NSAIDs, colchicine, and steroids (~3K) or patient fails to respond to NSAIDs and colchicine and not suitable for steroids (~5K)	**~8K**
Multiple Relapsing[1,2]	Patient previously responding to NSAIDs, colchicine, and/or steroids, but who continues to experience multiple recurrences	**~5K**
Steroid-Dependent[1,2]	Patient is unable to be tapered off steroids without experiencing subsequent recurrences	**~1K**

Represented by the patient population studied in RHAPSODY

Highest unmet needs:

- **Resolution of episodes**
- **Prevention of future episodes**
- **Steroid-sparing disease control**
- **Quality of life**

Physicians indicated an interest to treat across all three subgroups[3]

Potential to Broaden Utilization Over Time: ~3K Patients

First Recurrence, High Risk[1,2]	Patient identified during first recurrence as having high risk features predictive of multiple future recurrences (very large effusions, tamponade, etc.)	**~3K**

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

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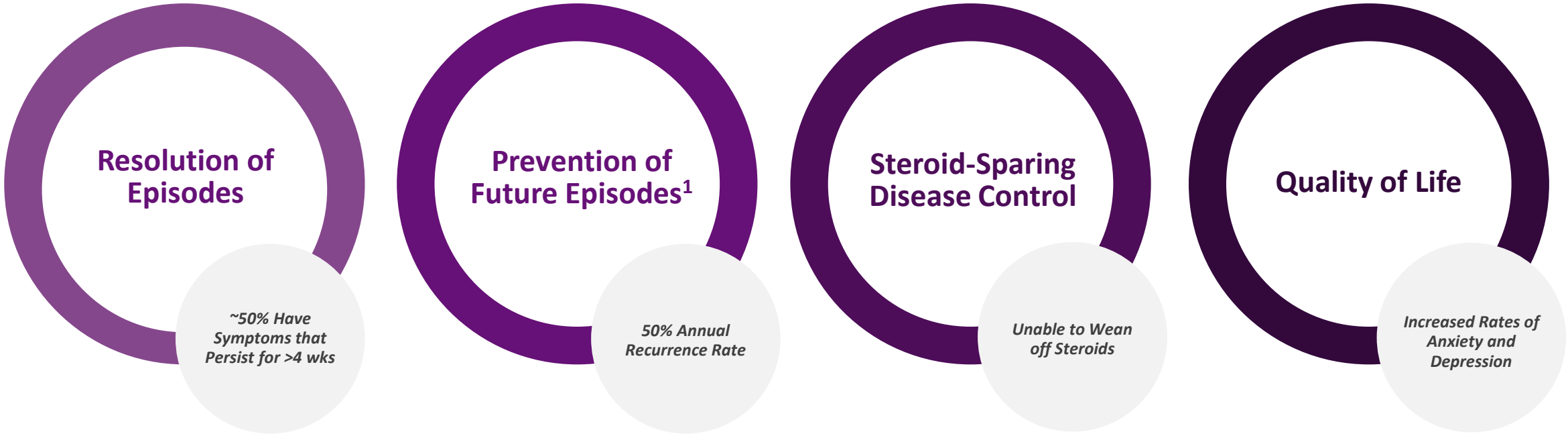
Key Areas of Unmet Need in Patients with Recurrent Pericarditis

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



Source: Kiniksa Pharmaceuticals data on file 2019; 1) Prevention of future episodes while on treatment

Strategic Imperatives for Commercial Launch of Rilonacept in Recurrent Pericarditis

Successful Execution to Physician Adoption of Rilonacept

	Unmet Need	Standard of Care	Payer Reimbursement	Patient Support
	Recurrent pericarditis is viewed as a serious, debilitating disease mediated primarily by IL-1	**Aim for rilonacept to be the product of choice for the treatment and prevention of recurrent pericarditis**	**Broad patient access at a price that reflects rilonacept's value as a first-in-class IL-1 inhibitor of inflammation**	**Optimize the patient and customer experience with rilonacept and Kiniksa**
Strategy	• Drive awareness and understanding of recurrent pericarditis and the role of inappropriate IL-1 production • Characterize and communicate burden of recurrent pericarditis on patients	• Help ensure there is an understanding of the benefit/risk of rilonacept • Demonstrate scientific evidence that rilonacept targets the primary mediator of recurrent pericarditis disease pathophysiology (IL-1 overproduction)	• Demonstrate product benefits, establish rapid payer coverage, and navigate potential access barriers • Implement scalable operations to support customers	• Establish robust patient support programs • Provide a seamless experience for patients starting on rilonacept & support ongoing adherence
Tactics	• 'Heart of Inflammation' disease awareness campaign and website • Continued presence at scientific congresses • Virtual patient advisory boards, podcasts and videos	• Specialty cardiovascular sales force • Efficient digital marketing • Peer to Peer speaker program • Patient support network • Scientific Congress Exhibits and Symposia (ACC, ESC, AHA)	• Compelling value proposition and supportive tools (value dossier and budget impact model) • Comprehensive payer engagement plan • Specialty pharmacy network distribution network	• High-touch patient support, reimbursement services, patient financial assistance, initiation support (Quick Start), injection training • Partner with the pericarditis community to improve advocacy, education and support for affected patients

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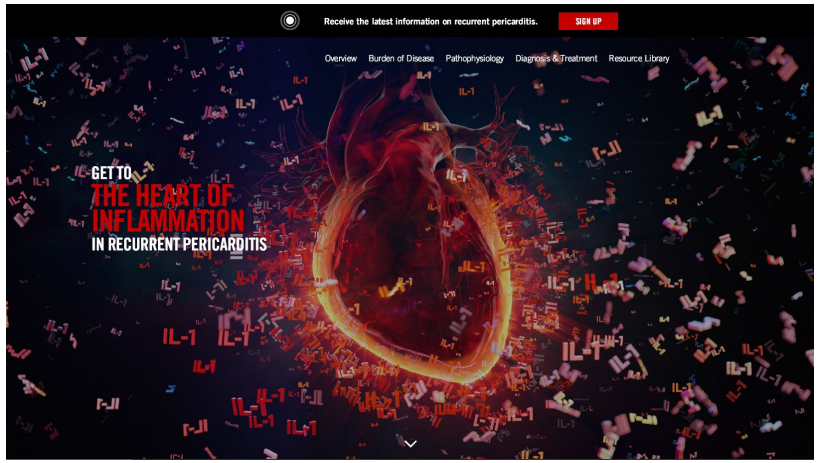
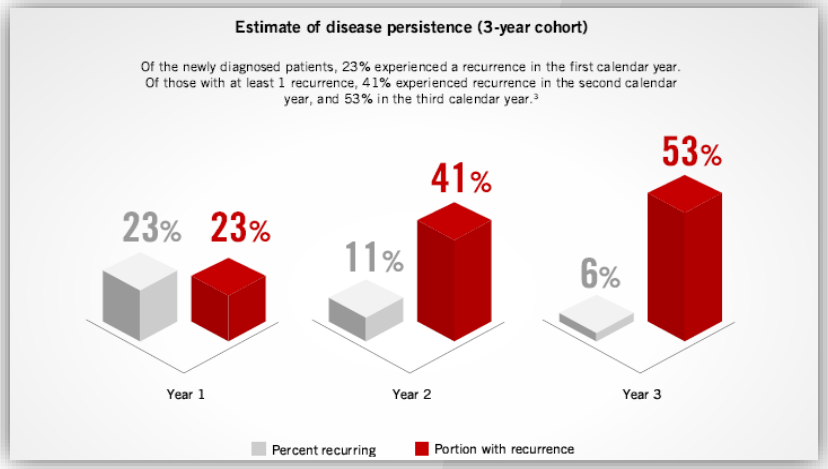
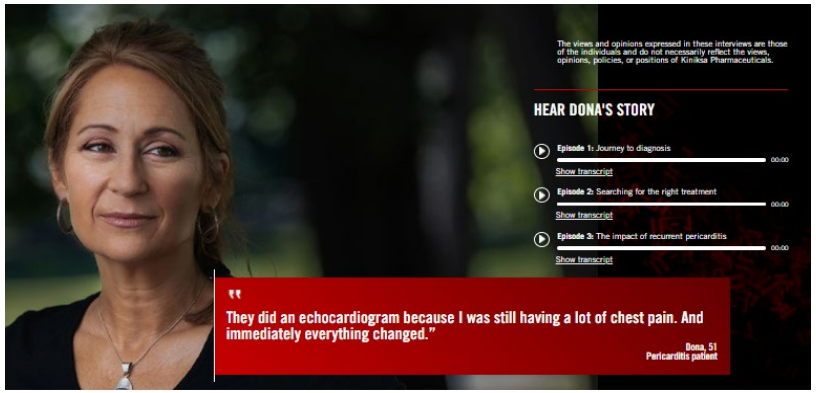
Unmet Need: ‘Heart of Inflammation’ Disease Awareness Campaign

Advancing Physicians’ Perceptions of Recurrent Pericarditis and Treatment Behaviors







‘Heart of Inflammation’

- Disease Overview
  - Epidemiology
  - Disease progression
  - Risk factors
- Burden of Disease
  - Patient podcasts
  - Health-related quality of life
- IL-1 Pathophysiology
  - Role of IL-1
- Diagnostics & Treatment
- Resource Library

www.heartofinflammation.com

Unmet Need: Multi-Channel Patient Support Network

Providing Resources to Help Ensure an Understanding of the Benefit/Risk of Treatment

Peer Videos & Testimonials

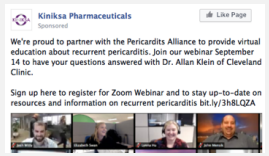




Social Communication & Engagement

Patient Education

MedicineX
STORYTELLING BY DOCTORS FOR PATIENTS



Patient Opt-In

Access to Advocacy

Pericarditis Alliance

MYOCARDITIS FOUNDATION

AUTOINFLAMMATORY ALLIANCE formerly known as The NOMID Alliance

"We are grateful to Kiniksa for taking interest in this disease and developing resources to help us. As people suffering from recurrent pericarditis, we feel alone and overlooked. We are looking to you for hope - hope that this won't last forever, hope that your drug will help us, hope that others won't have to suffer like we have. ***We are no longer in the dark. We now have your research and support to help us!****" – Patient Advisor*

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Standard of Care: Kiniksa One

Building a Collaborative Field Force to Help Enable Rilonacept to be the Product of Choice for the Treatment and Prevention of Recurrent Pericarditis

Medical Science Liaisons

- **Focus:** Subject matter experts and HCPs
- **Responsibility:** Disease awareness with a scientific and clinical focus, advocacy development, account and payer support, speaker management

Field Sales

- **Focus:** ~2500 HCPs across ~800 accounts
- **Responsibility:** Physician accounts, disease education, Arcalyst promotion (at approval), account and territory plans, speaker program planning, in-office injection training

Patient Services

- **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

Payer Team

- **Focus:** ~350 payers and 5 Specialty Pharmacies
- **Responsibility:** Payer/specialty pharmacy relationship, strategic account planning, support sales team

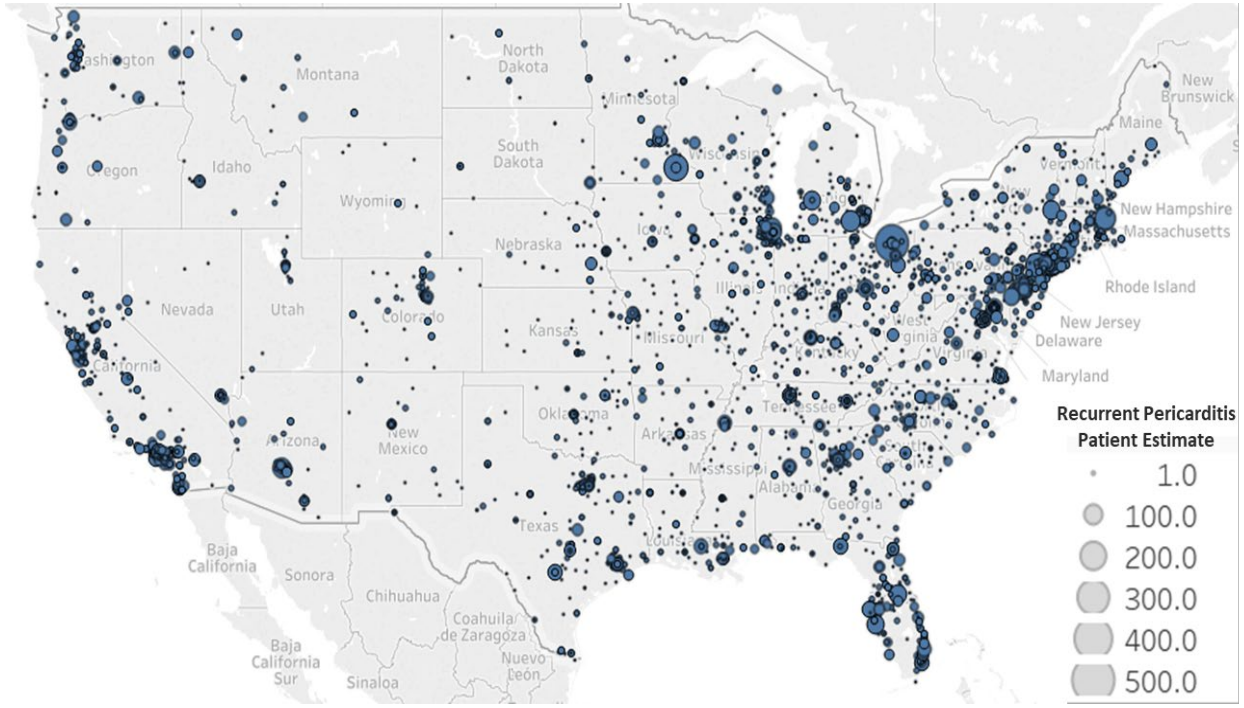
Note: Field teams will be trained to ensure compliance throughout all interactions.

HCP = health care provider

Standard of Care: Specialty Cardiovascular Sales Force

Planning to Reach ~70% of Recurrent Pericarditis Patients at Top ~800 Accounts

Estimated Recurrent Pericarditis Patients by Account



Focused & Targeted Sales Execution

National	Territory Level	
	First 3 months	Within the First Year
Initial launch focus on top tier accounts **~45%** of RP patients nationally **~350** accounts nationally	**10-15** accounts **~60** high value HCPS	**30** accounts **~100** high value HCPS
Following adoption, moving into next deciles to **~70%** of RP patients nationally **~800** accounts nationally (20% of total accounts)		

HCP = health care provider

COVID-19: Impact on Launch Strategy and Providing Tools to Ensure Success in Any Environment

Virtual Meeting Tools Providing Remote Detailing

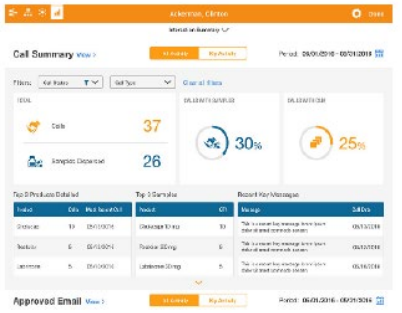
- Improved customer engagement during pandemic with convenient and compliant virtual content sharing
- Increased productivity and reach by using video to connect with HCPs regardless of physical access restrictions and geography
- Maintain compliance by ensuring field teams use only approved and existing closed loop marketing content.
- Provide specialized training focused on creating high-performers working in a virtual environment

Approved Emails

- Increased the frequency and quality of email reach with more tailored messages selected for physicians with recent interactions, and broader outreach to low-see and no-see customers
- Higher engagement rates driven by a known cardiovascular sales rep in the “From” line; results in open rates more than 10 times higher than mass email

Field Force Build

- Emphasis placed on prior cardiology experience, when possible, to leverage existing relationships



HCP = health care provider

Payer Reimbursement: Three-Phased Payer Engagement Plan

Facilitating Broad Access and Affordability

1. **Introduce Kiniksa**
2. **Disease Awareness**
3. **Support Payer Reviews**

Desired Outcomes

- **Shortest Time Payer Review and Coverage Policy Update**
- **Most Favorable Formulary Tier Means the Lowest Patient Cost Burden**
- **Least Burdensome Coverage Restrictions**

Goal will be to avoid criteria that requires prior use of corticosteroids and documented levels of C-reactive protein

Patient Support: Programs to Help Provide a Seamless Experience

Facilitating Initiation, Adherence and Retention

- Enrollment Form & Patient Consent Management
- Adherence Program
- Patient Assistance Program
- Online Tools
- Reimbursement, Prior Auth. and Appeal Support
- Non-Commercial Dispensing Pharmacy
- Specialty Pharmacy
- Copay Card Support
- Partner Integrations
- Case Management
- Quick Start
- Data & Reporting
- Injection Training
- Bridge Program

Pricing and Treatment Duration Considerations

*The gross **price** for rilonacept (ARCALYST) in CAPS is $20,000 per month based on the weekly administration; in-line with specialty biologics with Breakthrough Therapy designation, Orphan Drug designation and high unmet need*

*Our expectations are consistent with the RHAPSODY dataset; our initial assessment is that patients could be treated for at least 6 to 9 months, and the appropriate **treatment duration** for some patients may be 12 months or longer*

Inputs to Determining Optimal Treatment Duration

- **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
- **Median treatment duration in RHAPSODY was 9 months, with a range up to 15 months**
  - Rilonacept treatment was associated with a 96% reduction in risk for pericarditis recurrence
  - Patients on rilonacept experienced none/minimal pericarditis pain for 98% of trial days
  - 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 longer treatment duration: continued treatment resulted in continued treatment response**
  - Registry data indicate patients treated for 6 months have worse outcomes compared to patients treated for 9 months[2]
  - The only events in the rilonacept arm in RHAPSODY took place in the setting of temporary drug interruptions of 1-3 weeks.
  - All patients in the placebo arm who received bailout rilonacept did not experience a recurrence through the end of the RW period
- **Additional data will be available through LTE where patients are assessed for observed treatment cessation (with imaging) at 18 months**[3]

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) 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; 3) A. Klein, et al.; 2020 AHJ Reference for Phase 3 design

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Closing Remarks

Sanj K. Patel

Chairman of the Board and CEO

Recurrent pericarditis is a debilitating IL-1 driven disease with significant unmet need

RHAPSODY provided data on clinically meaningful outcomes associated with recurrent pericarditis

Clear call to action: **~14K patients with multiple recurrences** annually

Initially expect **treatment duration** of at least 6 to 9 months and 12 months or longer for some patients

The **current gross price for rilonacept** in CAPS is $20k per month based on weekly administration

Potential **launch of rilonacept** in the first half of 2021[1]

1) Timeline assumes priority review

Continued Execution Across our Pipeline

Indication	Program & Target	Preclinical	Phase 1	Phase 2	Phase 3	Status
Recurrent Pericarditis[1]	**Rilonacept[2]** IL-1α & IL-1β					Phase 3 Data Reported in Q3 2020
Giant Cell Arteritis	**Mavrilimumab** GM-CSFRα					Phase 2 Data Expected in Q4 2020
COVID-19 Pneumonia & Hyperinflammation	**Mavrilimumab** GM-CSFRα					Adaptive Design Phase 2/3 Initiated in Q3 2020
Prurigo Nodularis	**Vixarelimab** OSMRβ					Phase 2b Initiation Expected in Q4 2020
Severe Autoimmune Diseases	**KPL-404** CD40					Phase 1 Data Expected in Q4 2020

1) The FDA granted Breakthrough Therapy designation to rilonacept for recurrent pericarditis in 2019 and Orphan Drug designation to rilonacept for pericarditis in 2020; 2) Rilonacept (ARCALYST®) is approved and marketed for cryopyrin-associated periodic syndrome, in the United States by Regeneron Pharmaceuticals, Inc.; IL-1α = interleukin-1α ; IL-1β = interleukin 1β; GM-CSFRα = granulocyte macrophage colony stimulating factor receptor alpha; OSMRβ = oncostatin M receptor beta

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Every Second Counts!™