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Rilonacept Interim Phase 2 Clinical Data and Pivotal Phase 3 Clinical Trial Initiation

December 11, 2018

Introduction

Mark Ragosa

Investor Relations

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 subsidiary, together, unless context otherwise requires, 'Kiniksa," "we" or our). In some cases, you can identify forward looking statements by terms such as "may," "will," "should," "expect," "plan," "anticipate," "could," "intend," "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 the potential for rilonacept to be the first approved therapy for patients suffering from recurrent pericarditis, its potential impact, and our stage in the process for providing such an approved therapy; our conclusions from the Phase 2 interim clinical trial data and expected timing for reporting the completed dataset; and statements regarding objectives of the design of our Phase 3 clinical trial for rilonacept.

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 the important factors discussed under the caption "Risk Factors" in our Quarterly Report on Form 10-Q filed with the Securities and Exchange Commission ("SEC") on November 6, 2018 and our other reports 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.

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.

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.

Kiniksa Overview

Sanj K. Patel

Chief Executive Officer & Chairman of the Board

KINIKSA

A company with a sequential pipeline, FIRMLY rooted in strong biologic rationale or validated mechanisms, potential for multiple indications, and designed to deliver near-, mid- and long-term value

Rilonacept Interim Phase 2 Data & Pivotal Phase 3 Initiation

John Paolini

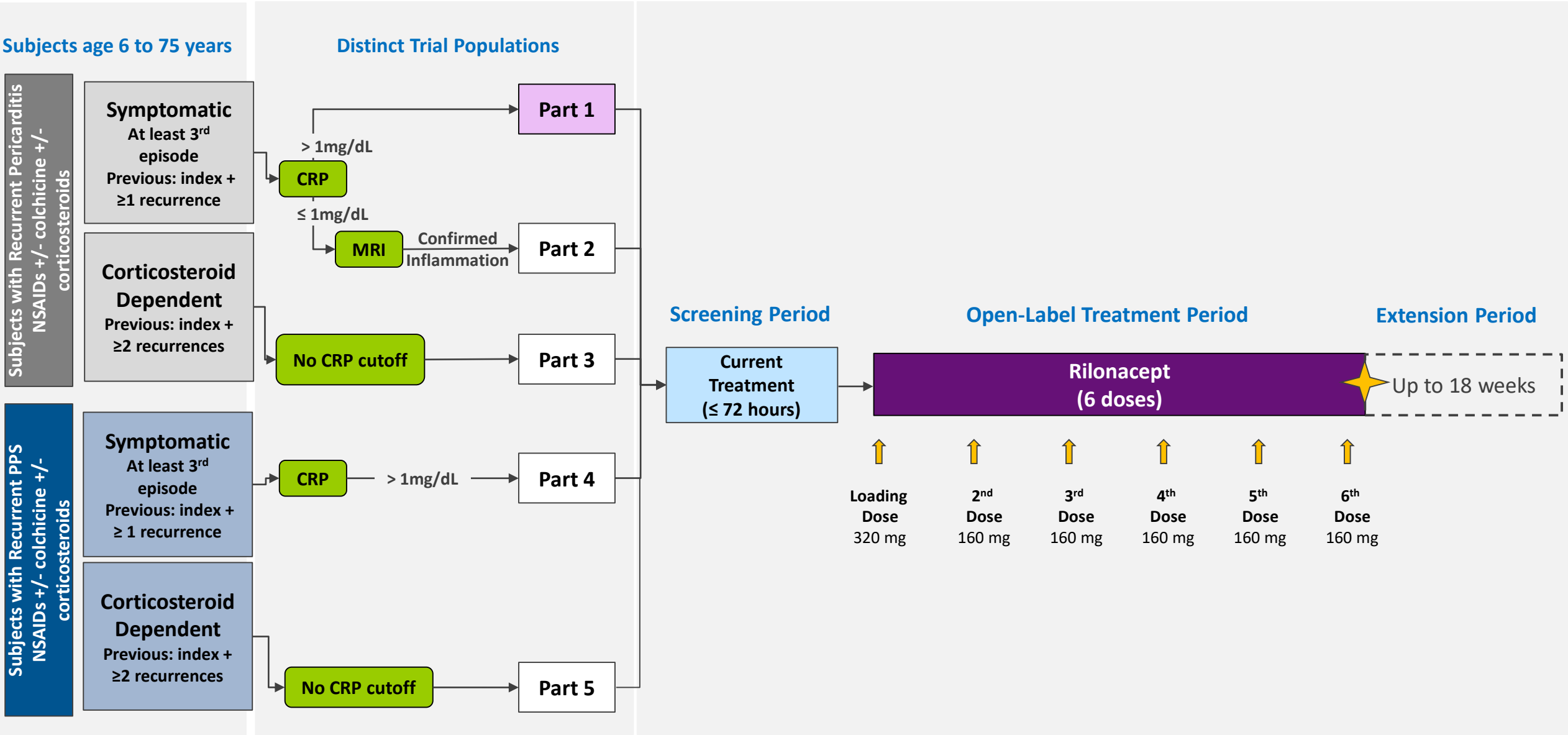
Chief Medical Officer

Rilonacept provides a potential opportunity to address an inflammatory cardiovascular disease with no currently approved therapies

Lead Indication	• For the treatment of Recurrent Pericarditis
Patient Population[1]	• ~90k patients in the US experience an acute incident of pericarditis per year • ~3k refractory patients and ~9k who are not well managed on existing therapies in the US
Mechanism of Action[2]	• A cytokine trap that blocks IL-1α and IL-1β signaling by acting as a soluble decoy receptor preventing IL-1α and IL-1β interaction with cell surface receptors
Competition[3]	• No currently-approved therapies for Recurrent Pericarditis; differentiated from both existing marketed IL-1 targeted therapeutics (dosing frequency and mechanism of action)
Clinical Development	• External Proof of concept: Interim Phase 2 pilot study data show reduced pain/inflammation • Pivotal Phase 3 trial, RHAPSODY, actively recruiting and screening subjects
Rights	• Worldwide excluding Middle East and North Africa • BLA transfers to Kiniksa upon FDA approval in Recurrent Pericarditis

1) UptoDate, Trinity Partners, Mayo Clin Proc. 2010 ;85 (6): 572-593; New Diagnostic Criteria for Acute Pericarditis: A Cardiac MRI Perspective, 2015 American College of Cardiology; 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.

Open-label Phase 2 pilot study of rilonacept in pericarditis populations



Note: Trial design inspired by investigator led study AIRTRIP, published in JAMA (Brucato et al., JAMA, 2016;316(18): 1906-1912)

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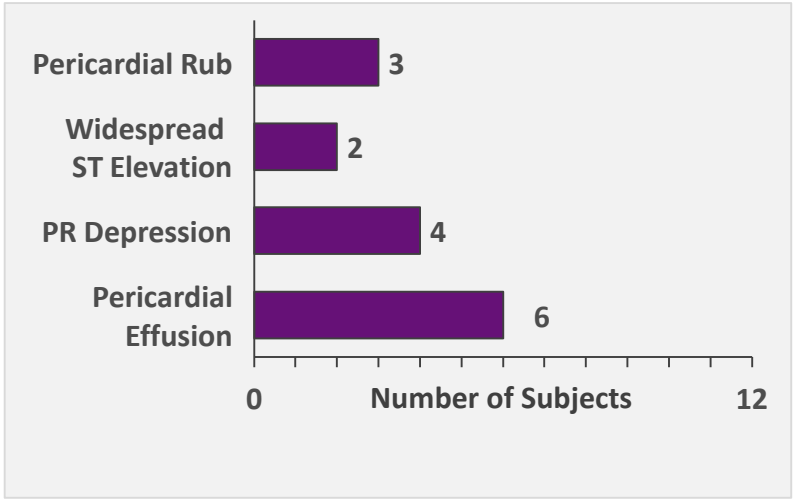
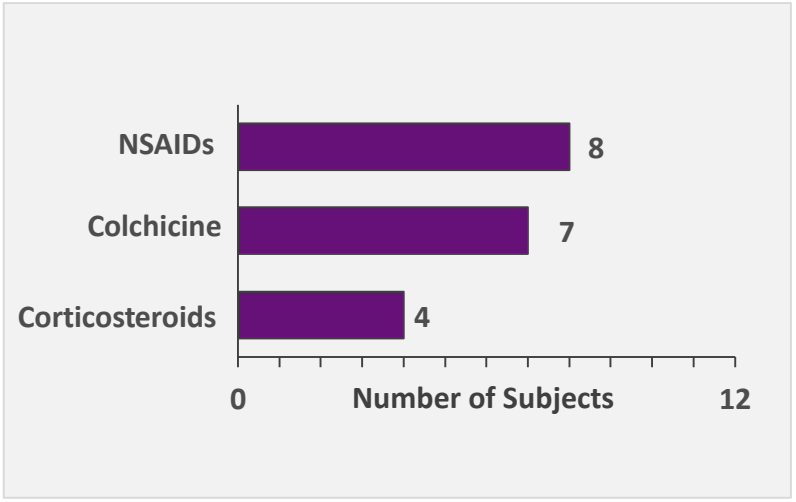
Baseline characteristics: symptomatic recurrent pericarditis subjects (Part 1)

General Characteristics	Data
Number of Subjects Enrolled *As of Nov 1, 2018*	12
Age (yrs)	26-52
Sex (Male/Female)	3/9
Episodes Prior to Qualifying Episode	2-5

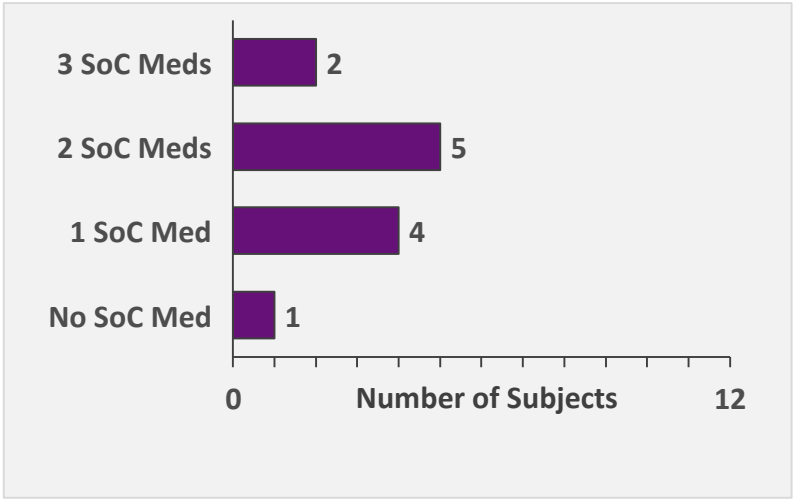
Clinical Findings at Baseline



SoC Received at Baseline



Combination Therapy at Baseline



Notes: Preliminary data from on-going study (Part1) as of Nov 1st, 2018; SoC = Standard of Care; SoC Medications =Colchicine, NSAIDs, Corticosteroids

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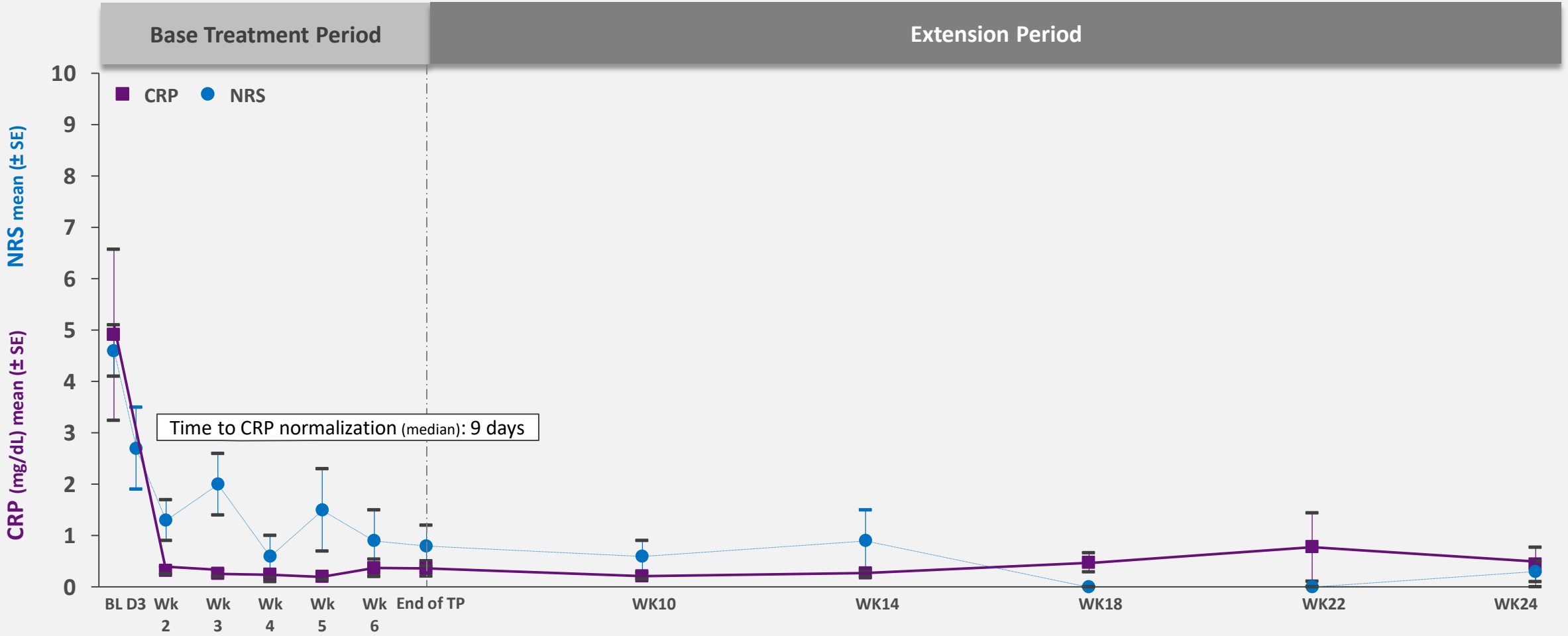
Reductions in both CRP and NRS after the first dose; persistent response



		BL	D3	Wk 2	Wk 3	Wk 4	Wk 5	Wk 6	EoTP	Wk10	Wk14	Wk18	Wk22	Wk24
CRP	# of Subjects	12		9	11	10	9	4	10	7	7	3	2	4
CRP	CRP mean	4.91		0.32	0.26	0.24	0.20	0.37	0.36	0.21	0.27	0.47	0.78	0.44
Pain	# of Subjects	12	10	12	12	7	12	8	10	7	7	5	6	4
Pain	Pain mean	4.6	2.7	1.3	2.0	0.6	1.5	0.9	0.8	0.6	0.9	0.0	0.0	0.3

Notes: Preliminary data from on-going study (Part1) as of Nov 1st, 2018; Baseline = rilonacept 320mg loading dose; Week 1 through Week 6= rilonacept 160mg; EoTP= End of Treatment Period

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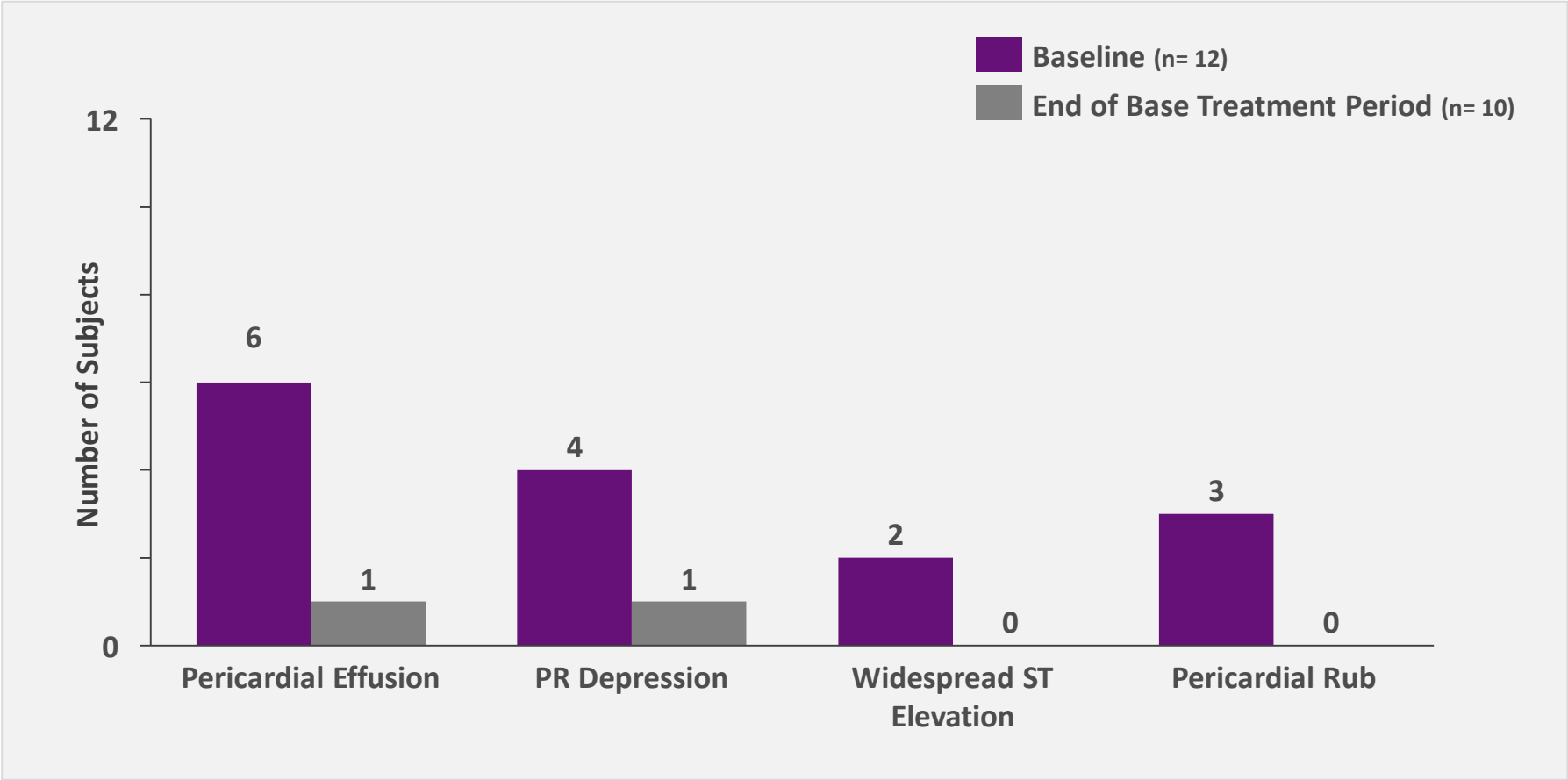
Pericardial signs resolved during rilonacept 6-week base treatment period



Note: Preliminary data from on-going study (Part1) as of Nov 1st, 2018

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Summary of adverse events

- Rilonacept was generally well-tolerated
- 7/12 subjects experienced at least one treatment-related adverse event during the treatment period
- The most common adverse event was mild transient injection site reaction
- One patient discontinued from the study due to a Treatment-Emergent Serious Adverse Event, skin abscess

Treatment-Related and Non-Treatment-Related TEAEs

Category	Subjects (n=12)
Subjects with at least one TEAE	12
Subjects with at least one treatment-related TEAE	7
Subjects with at least one serious TEAE	2
Subjects with a serious Treatment-Related TEAE	1
Subjects with at least one TEAE leading to treatment discontinuation[1]	1
Subjects with at least one TEAE leading to death	0

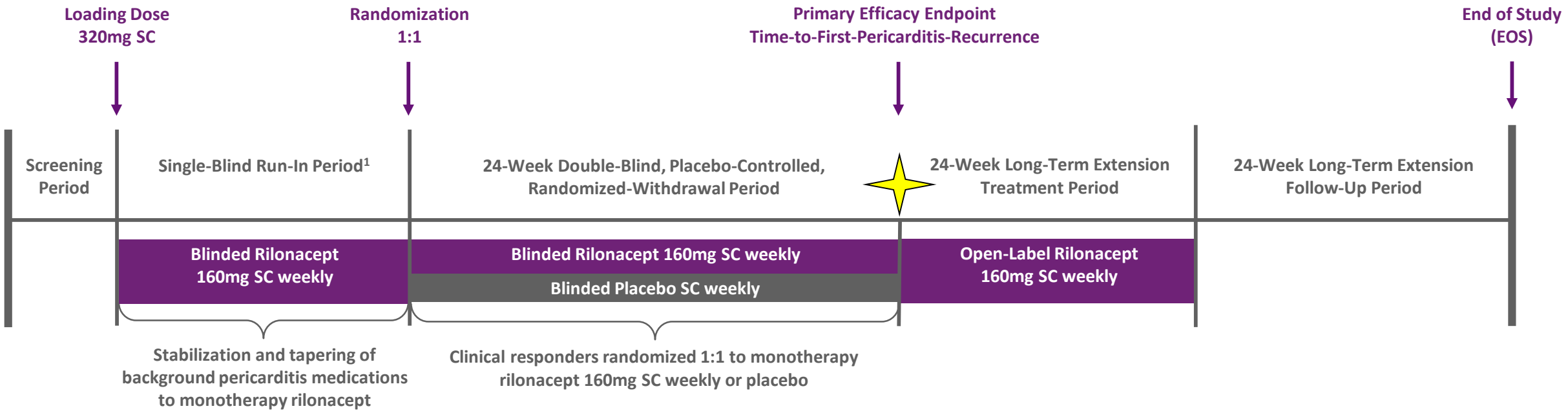
AEs Occurring at Least Once (by Affected Organ System)

Organ System	Preferred Term	Part 1 (n=12)
Number (%) of subjects who had at least one AE		12 (100%)
Gastrointestinal disorders		6 (50%)
General disorders and administration site conditions		5 (41.7%)
Infections and Infestations		5 (41.7%)
Investigations		5 (41.7%)
	Liver function test increased	2 (16.7%)
	Blood Cholesterol increased	1 (8.3%)
	Blood creatine phosphokinase increased	1 (8.3%)
	HDL increased	1 (8.3%)
Musculoskeletal and connective tissue disorders		2 (16.7%)

Notes: Preliminary data from on-going study (Part1) as of 1/1/2018; AE = Adverse Event; TEAE = Treatment Emergent Adverse Event

1) 1 patient discontinued due to SAE of skin abscess (occurred after the Nov 1st data cutoff)

Pivotal Phase 3 clinical trial of rilonacept for recurrent pericarditis



Inclusion Criteria:

- Present at screening with at least a third pericarditis episode, defined as at least 1 day with NRS pain of ≥ 4 within the 7-day period prior to first study drug administration
- 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 Outcome Measure (24 weeks):

- Time to pericarditis recurrence

Secondary Outcome Measures (24-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
- Proportion of subjects with adverse events

[1]Duration of the run-in period undisclosed in order to maintain study subjects blinded to the start of the randomized-withdrawal period.

Every Second Counts!™