UNITED STATES
SECURITIES AND EXCHANGE COMMISSION
WASHINGTON, D.C. 20549

FORM 8-K

CURRENT REPORT

Pursuant to Section 13 or 15(d) of the Securities Exchange Act of 1934

Date of Report (Date of earliest event reported): **June 29, 2020**

Kiniksa Pharmaceuticals, Ltd.
(Exact name of Registrant as Specified in Its Charter)

Bermuda	**001-730430**	**98-1327726**
(State or other jurisdiction of incorporation or organization)	(Commission File Number)	(I.R.S. Employer Identification No.)

Kiniksa Pharmaceuticals, Ltd.
Clarendon House
2 Church Street
Hamilton HM11, Bermuda
(808) 451-3453
(Address, zip code and telephone number, including area code of principal executive offices)

Kiniksa Pharmaceuticals Corp.
100 Hayden Avenue
Lexington, MA, 02421
(781) 431-9100
(Address, zip code and telephone number, including area code of agent for service)

N/A
(Former Name or Former Address, if Changed Since Last Report)

Check the appropriate box below if the Form 8-K filing is intended to simultaneously satisfy the filing obligation of the registrant under any of the following provisions

☐ Written communications pursuant to Rule 425 under the Securities Act (17 CFR 230.425)

o Soliciting material pursuant to Rule 14a-12 under the Exchange Act (17 CFR 240.14a-12)

o Pre-commencement communications pursuant to Rule 14d-2(b) under the Exchange Act (17 CFR 240.14d-2(b))

o Pre-commencement communications pursuant to Rule 13e-4(c) under the Exchange Act (17 CFR 240.13e-4(c))

Securities registered pursuant to Section 12(b) of the Act:

Title of each class	**Trading Symbol(s)**	**Name of each exchange on which registered**
Class A Common Shares $0.000273235 par value	KNSA	The Nasdaq Global Select Market

Indicate by check mark whether the registrant is an emerging growth company as defined in Rule 405 of the Securities Act of 1933 (§230.405 of this chapter) or Rule 12b-2 of the Securities Exchange Act of 1934 (§240.12b-2 of this chapter).

Emerging growth company ☒

If an emerging growth company, indicate by check mark if the registrant has elected not to use the extended transition period for complying with any new or revised financial accounting standards provided pursuant to Section 13(a) of the Exchange Act. ☒

Item 7.01. Regulation FD.

On June 29, 2020, Kiniksa Pharmaceuticals, Ltd. (the "Company") issued a press release announcing (a) top-line pivotal data from the global, double blind, placebo controlled, randomized withdrawal design, Phase 3 clinical trial of rilonacept in subjects with recurrent pericarditis, named RHAPSODY, and (b) that the Company will host a webcast conference call at 8:30 a.m. Eastern Time on June 29, 2020 to discuss top-line pivotal Phase 3 data for rilonacept in recurrent pericarditis. Details for the webcast conference call are included in the press release. A copy of the press release and a slide presentation containing data from the trial are furnished with this Current Report on Form 8-K as Exhibit 99.1 and Exhibit 99.2, respectively.

The information contained in this Item 7.01 of this Current Report on Form 8-K and Exhibit 99.1 and Exhibit 99.2 shall not be deemed "filed" for purposes of Section 18 of the Securities Exchange Act of 1934, as amended (the "Exchange Act"), or otherwise subject to the liabilities of that section, nor shall it be deemed incorporated by reference in any filing under the Securities Act of 1933, as amended, or the Exchange Act, regardless of any general incorporation language in such filing and except as expressly provided by specific reference in such filing.

Item 9.01. Financial Statements and Exhibits.

(d) Exhibits

The following exhibits relate to Item 7.01, which shall be deemed to be furnished, and not filed:

Exhibit No.	Description
99.1	Rilonacept Phase 3 Data Press Release issued by Kiniksa Pharmaceuticals, Ltd. dated June 29, 2020
99.2	Kiniksa Pharmaceuticals, Ltd. Phase 3 Data Slide-Presentation

SIGNATURES

Pursuant to the requirements of the Securities Exchange Act of 1934, the registrant has duly caused this report to be signed on its behalf by the undersigned hereunto duly authorized.

KINIKSA PHARMACEUTICALS, LTD.

Date: June 29, 2020

By: /s/ Thomas Beetham
Thomas Beetham
Executive Vice President, Chief Legal Officer

Exhibit 99.1

Kiniksa Announces Positive Data from Phase 3 Trial of Rilonacept in Recurrent Pericarditis (RHAPSODY)

- *Primary and all major secondary efficacy endpoints were highly statistically significant*
- *Rilonacept treatment resulted in a 96% reduction in risk of recurrent pericarditis events (primary efficacy endpoint: Hazard Ratio = 0.04, p<0.0001)*
- *Safety results consistent with FDA-approved label for CAPS*
- *sBLA submission expected later this year*
- *Conference call and webcast scheduled for 8:30 a.m. EDT today*

HAMILTON, BERMUDA – June 29, 2020 – Kiniksa Pharmaceuticals, Ltd. (Nasdaq: KNSA) ("Kiniksa"), a biopharmaceutical company focused on discovering, acquiring, developing and commercializing therapeutic medicines for patients with significant unmet medical need, reported positive data from RHAPSODY, a pivotal Phase 3 trial of rilonacept, a weekly, subcutaneously-injected, recombinant fusion protein that blocks interleukin-1 alpha (IL-1α) and interleukin-1 beta (IL-1β) signaling, in recurrent pericarditis. RHAPSODY met its prespecified primary and all major secondary efficacy endpoints, showing that rilonacept improved clinically meaningful outcomes associated with the unmet medical need in recurrent pericarditis, a painful and debilitating autoinflammatory disease. The U.S. Food and Drug Administration (FDA) granted Breakthrough Therapy designation to rilonacept for the treatment of recurrent pericarditis in 2019, and Kiniksa expects to submit a Supplemental Biologics License Application (sBLA) later this year.

"We are pleased to announce that RHAPSODY, our pivotal Phase 3 trial of rilonacept in recurrent pericarditis, met its primary and all major secondary efficacy endpoints," said Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa. "Combined with a well-tolerated safety profile and a weekly dosing regimen, these data are an important step forward for patients. We believe rilonacept has the potential to be the first FDA-approved therapy for recurrent pericarditis. We are committed to submitting an sBLA to the FDA later this year and look forward to bringing this potential treatment option to patients as soon as possible."

RHAPSODY is a global, randomized withdrawal design, pivotal Phase 3 clinical trial of rilonacept in recurrent pericarditis. The trial's primary analysis population included 61 actively symptomatic recurrent pericarditis patients who were failing standard of care treatment, including nonsteroidal anti-inflammatory drugs (NSAIDs), colchicine, or corticosteroids, initiated rilonacept treatment during a run-in period, discontinued background medications, and achieved and maintained clinical response (11-point pain Numerical Rating Scale (NRS) ≤ 2.0 and C-reactive protein (CRP) ≤ 0.5 mg/dL) on rilonacept monotherapy. Clinical responders were randomized 1:1 to receive continued weekly rilonacept (n=30) or placebo (n=31) in a blinded manner in the randomized withdrawal period.

The primary efficacy endpoint of time-to-first adjudicated pericarditis recurrence in the randomized withdrawal period was highly statistically significant.

- Median [95% CI] time to pericarditis recurrence for rilonacept recipients in the randomized withdrawal period could not be estimated due to the low number of recurrences in the rilonacept treatment arm. The median time-to-recurrence for placebo recipients was 8.6 [4.0-11.7] weeks (Hazard Ratio = 0.04, $p<0.0001$).

- Rilonacept recipients experienced a 96% reduction in risk of recurrent pericarditis events.

All major secondary efficacy endpoints in the randomized withdrawal period were also highly statistically significant.

- 81% of rilonacept recipients maintained clinical response at Week 16 of the randomized withdrawal period, compared to 20% of placebo recipients ($p=0.0002$). Consistent results were observed at Week 8 and Week 24 and were also highly statistically significant ($p<0.0001$ and $p=0.0022$, respectively).

- The proportion of rilonacept recipients with absent or minimal pericarditis symptoms at Week 16 of the randomized withdrawal period was 81% compared to 25% for placebo recipients ($p=0.0006$). Consistent results were observed at Week 8 and Week 24 and were also highly statistically significant ($p<0.0001$ and $p=0.0002$, respectively).

Rilonacept was well-tolerated in the study, with adverse events consistent with the FDA-approved label for the treatment of Cryopyrin-Associated Periodic Syndromes (CAPS). The most common adverse events were injection site reactions.

“The RHAPSODY data provide hope for patients suffering from recurrent pericarditis,” said John F. Paolini, MD, PhD, Chief Medical Officer of Kiniksa. “In fact, rilonacept patients experienced no or minimal pericarditis pain for nearly 95% of study days through Week 16 compared to less than half of study days for placebo recipients, which was highly statistically significant. We believe that, by treating and preventing disease recurrence, rilonacept has the potential to be a transformational therapeutic advancement in the treatment of patients with recurrent pericarditis and to become the first FDA-approved therapy for this debilitating autoinflammatory disease.”

Additional analyses of the RHAPSODY trial results are ongoing, and Kiniksa plans to present the data at a future medical meeting or in a publication.

Rilonacept was discovered and developed by Regeneron Pharmaceuticals, Inc. (Regeneron) and is approved by the FDA under the brand name ARCALYST® for the treatment of CAPS. Kiniksa licensed rilonacept from Regeneron in 2017 for evaluation in diseases believed to be mediated by both IL-1α and IL-1β, including recurrent pericarditis. The FDA granted Breakthrough Therapy designation to rilonacept for recurrent pericarditis in 2019. Based on the Phase 3 RHAPSODY data announced today, the Biologic License Application (BLA) for CAPS will transfer to Kiniksa, and the company plans to submit an sBLA with the FDA in recurrent pericarditis later this year. Upon receipt of FDA approval for rilonacept in recurrent pericarditis, Kiniksa would assume the sales and distribution of rilonacept for the approved indications in the United States and will evenly split profits on sales with Regeneron.

Conference Call Information

Kiniksa will host a conference call and webcast at 8:30 a.m. Eastern Time on Monday, June 29, 2020 to discuss top-line pivotal Phase 3 data for rilonacept in recurrent pericarditis. Individuals interested in participating in the call should dial (866) 614-0636 (U.S. and Canada) or (409) 231-2053 (international) using conference ID number 2089128. To access the webcast, please visit the Investors and Media section of Kiniksa's website at www.kiniksa.com. The archived webcast will be available on Kiniksa's website for 14 days beginning approximately one hour after the call has completed.

About RHAPSODY

RHAPSODY is the global, randomized withdrawal design, pivotal Phase 3 clinical trial of rilonacept in recurrent pericarditis. Eligible patients presented at screening with at least a third pericarditis episode, defined as at least 1 day with pericarditis pain of ≥ 4 on the 11-point NRS and a CRP value ≥ 1 mg/dL within the 7-day period prior to first study drug administration. Patients could be receiving concomitant NSAIDs and/or colchicine and/or oral corticosteroid treatment in any combination. The study was comprised of 4 periods: a screening period; a single-blind run-in period during which patients received a loading dose of rilonacept 320 mg injected subcutaneously (SC) followed by 160 mg SC weekly while background pericarditis medications were tapered and discontinued; a double-blind, placebo-controlled randomized withdrawal period during which clinical responders to rilonacept were randomized 1:1 and received 160 mg SC weekly rilonacept or placebo; and a long-term extension treatment period with up to 24 months of open-label rilonacept 160 mg SC weekly. The primary efficacy endpoint was time-to-first pericarditis-recurrence in the randomized withdrawal period. The Clinical Endpoint Committee adjudicated all suspected pericarditis recurrences for inclusion in the primary efficacy endpoint analysis. The co-principal investigators are Dr. Allan Klein of Cleveland Clinic and Dr. Massimo Imazio of the University of Torino, Italy. For more information, refer to ClinicalTrials.gov Identifier: NCT03737110.

About Recurrent Pericarditis
Recurrent pericarditis is a painful and debilitating autoinflammatory cardiovascular disease that typically presents with chest pain and is often associated with changes in electrical conduction and sometimes buildup of fluid around the heart, called pericardial effusion. Patients with pericarditis are deemed recurrent if they have an additional episode after a symptom-free period of 4-6 weeks, and chronic if symptoms from any one episode last longer than three months. Recurrent pericarditis symptoms impair qualify of life, limit physical activities, and lead to frequent emergency department visits and hospitalizations. There are currently no FDA-approved treatments for recurrent pericarditis.

About the Rilonacept License Agreement with Regeneron
In 2017, Regeneron granted Kiniksa an exclusive license to develop and commercialize rilonacept worldwide, aside from Israel, Egypt, Turkey and select countries in the Middle East and North Africa. In the United States and Japan, Kiniksa's license is initially for all indications other than those involving local administration to the eye or ear, oncology, deficiency of the interleukin-1 receptor antagonist (DIRA) and CAPS. If Kiniksa is successful in receiving marketing approval for rilonacept in the United States for a new indication, the scope of the license granted to Kiniksa will automatically expand to include DIRA and CAPS in the United States and Japan, and Kiniksa will assume the sales and distribution of rilonacept in these additional indications. Outside the United States and Japan, Kiniksa's license is for all indications other than local application to the eye or ear, oncology, CAPS, DIRA and certain periodic fever syndromes. Kiniksa made an upfront payment of $5.0 million to Regeneron and is obligated to make regulatory milestone payments of up to $27.5 million in the aggregate. Thereafter, Kiniksa and Regeneron will evenly split profits on sales of rilonacept after deducting certain commercialization expenses subject to specified limits.

About Rilonacept
Rilonacept is a weekly, subcutaneously-injected, recombinant fusion protein that blocks IL-1α and IL-1β signaling. Rilonacept was discovered and developed by Regeneron and is approved by the FDA under the brand name ARCALYST® for the treatment of CAPS, which includes Familial Cold Autoinflammatory Syndrome and Muckle-Wells Syndrome. Rilonacept for the treatment of DIRA is currently pending FDA approval following the submission of a supplemental BLA in June 2020. Rilonacept in recurrent pericarditis is an investigational drug. The FDA has granted Breakthrough Therapy designation to rilonacept for recurrent pericarditis.

Important information about ARCALYST® (rilonacept) Injection
IL-1 blockade may interfere with immune response to infections. Serious, life-threatening infections have been reported in patients taking ARCALYST. ARCALYST should be discontinued if a patient develops a serious infection. Taking ARCALYST with TNF inhibitors is not recommended because this may increase the risk of serious infections.

Patients should not receive a live vaccine while taking ARCALYST. It is recommended that prior to initiation of therapy with ARCALYST patients receive all recommended vaccinations, as appropriate, including pneumococcal vaccine and inactivated influenza vaccine. In the initial development program for ARCALYST, six serious adverse reactions were reported by four patients: Mycobacterium intracellular infection, gastrointestinal bleeding and colitis, sinusitis and bronchitis and Streptococcus pneumoniae meningitis. The most commonly reported adverse reactions associated with ARCALYST were injection site reaction and upper respiratory tract infection. Patients should be monitored for changes in their lipid profiles and provided with medical treatment if warranted. Treatment with immunosuppressants, including ARCALYST, may result in an increase in risk of malignancies. Hypersensitivity reactions associated with ARCALYST administration in clinical studies have been rare. If a hypersensitivity reaction occurs, administration of ARCALYST should be discontinued and appropriate therapy initiated.

About Kiniksa
Kiniksa is a biopharmaceutical company focused on discovering, acquiring, developing and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. Kiniksa's clinical-stage product candidates, rilonacept, mavrilimumab, vixarelimab and KPL-404, are based on strong biologic rationale or validated mechanisms, target underserved conditions and offer the potential for differentiation. These pipeline assets are designed to modulate immunological pathways that are implicated across a spectrum of diseases. For more information, please visit www.kiniksa.com.

Forward-Looking Statements
This press release contains forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995. 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 press release that do not relate to matters of historical fact should be considered forward-looking statements, including without limitation, statements regarding: our expectation that the rilonacept BLA will be transferred to Kiniksa pursuant to the Regeneron license agreement; our plan to submit an sBLA for rilonacept; the potential for rilonacept to be an advancement in treatment of pericarditis; our belief that rilonacept has the ability to become the first FDA-approved therapy for recurrent pericarditis; our expectation of presenting additional data from RHAPSODY; and the urgency in bringing an approved therapy to patients suffering from recurrent pericarditis.

These forward-looking statements are based on management's current expectations. These statements are neither promises nor guarantees, but involve known and unknown risks, uncertainties and other important factors that may cause our actual results, performance or achievements to be materially different from any future results, performance or achievements expressed or implied by the forward-looking statements, including without limitation, the following: our potential inability to replicate in later clinical trials the positive final data from our earlier clinical trials or investigator-initiated protocols or studies; impact of additional data from us or other companies; potential undesirable side effects caused by our product candidates; our potential inability to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities; our reliance on third parties to manufacture our product candidates; drug substance and/or drug product shortages; and our reliance on third parties to conduct research, clinical trials, and/or certain regulatory activities for our product candidates; delays or difficulty in activating sites or enrolling patients in our planned clinical trials; potential complications in coordinating among requirements, regulations and guidelines of regulatory authorities across a number of jurisdictions for our planned global clinical trials; the potential impact of the COVID-19 pandemic and measures taken in response to the pandemic; changes in our operating plan and funding requirements; existing or new competition; and our ability to attract and retain qualified personnel.

These and other 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 May 4, 2020 and our other reports subsequently filed with the SEC could cause actual results to differ materially from those indicated by the forward-looking statements made in this press release. Any such forward-looking statements represent management's estimates as of the date of this press release. While we may elect to update such forward-looking statements at some point in the future, we disclaim any obligation to do so, even if subsequent events cause our views to change. These forward-looking statements should not be relied upon as representing our views as of any date subsequent to the date of this press release.

Every Second Counts!™

Kiniksa Investor and Media Contact
Mark Ragosa
(781) 430-8289
mragosa@kiniksa.com

Exhibit 99.2

KINIKSA

Every Second Counts!™

RHAPSODY Top-Line Results

June 2020

KINIKSA

Every Second Counts!™

Welcome

Mark Ragosa
VP of Investor Relations and Finance

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 strategy; potential acquisitions and collaborations; potential value drivers; potential indications; potential market opportunities and competitive position; on going, planned and potential clinical trials and other studies; timing and potential impact of clinical data; regulatory and other submissions, applications and approvals; commercial strategy and pre-commercial activities; 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 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 May 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.

KINIKSA

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Introduction

Sanj K. Patel
CEO and Chairman of the Board

Summary of Pivotal Phase 3 Trial of Rilonacept in Recurrent Pericarditis

RHAPSODY

- Prespecified primary and all major secondary efficacy endpoints were highly statistically significant.
- The primary efficacy endpoint of median time-to-first adjudicated pericarditis recurrence in the randomized withdrawal period was highly statistically significan
  - Median [95% CI] time to pericarditis recurrence for rilonacept recipients in the randomized withdrawal period could not be estimated due to the low number of recurrences in the rilonacept treatment arm. The median time-to-recurrence for placebo recipients was 8.6 [4.0-11.7] weeks (Hazard Ratio = 0.04, p<0.0001).
  - Rilonacept recipients experienced a 96% reduction in risk of recurrent pericarditis events.
- All major secondary efficacy endpoints in the randomized withdrawal period were also highly statistically significant.
  - 81% of rilonacept recipients maintained clinical response at Week 16 of the randomized withdrawal period, compared to 20% of placebo recipients (p=0.0002). Consistent results were observed at Week 8 and Week 24 and were also highly statistically significant (p<0.0001 and p=0.0022, respectively).
  - The proportion of rilonacept recipients with absent or minimal pericarditis symptoms at Week 16 of the randomized withdrawal period was 81% compar to 25% for placebo recipients (p=0.0006). Consistent results were observed at Week 8 and Week 24 and were also highly statistically significant (p<0.000 and p=0.0002, respectively).
  - Rilonacept recipients experienced no or minimal pain for 95% of trial days through Week 16 compared to 47% of trial days for placebo recipients (p<0.0001). Consistent results were observed at Weeks 8 and 24 and were also highly significant (p<0.0001 and p<0.0001, respectively).
- Patients are continuing to receive open label rilonacept in the Long-Term Extension.
- Rilonacept was well-tolerated, with a safety profile consistent with the existing ARCALYST® label.
- Based on the Phase 3 RHAPSODY data, Kiniksa plans to submit an sBLA with the FDA later this year.

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RHAPSODY Top-Line Results

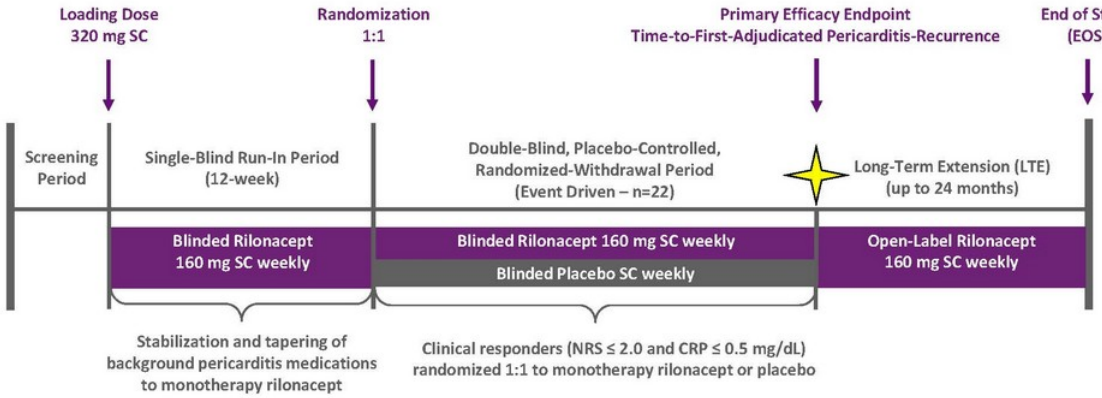
John F. Paolini
Chief Medical Officer

Rilonacept

Indication[1]	**Recurrent Pericarditis:** Painful and debilitating autoinflammatory cardiovascular disease
Mechanism of Action[2]	IL-1α and IL-1β cytokine trap
Scientific Rationale[2]	IL-1α and IL-1β are cytokines shown to play key role in inflammatory diseases
Prevalence[3]	~40k prevalent in U.S.; addressable opportunity of ~14k in U.S.
Competition[4]	No FDA-approved therapies for recurrent pericarditis
Status	Breakthrough Therapy designation granted in December 2019; pivotal Phase 3 data reported in June 2020
Economics	Regulatory milestones; 50/50 profit split upon commercialization excluding certain expenses
Rights	BLA transfers to Kiniksa after receipt of positive Phase 3 clinical data and an acceptable safety profile; upon approval Kiniksa has the rights to recurrent pericarditis worldwide (excluding MENA)

1) Rilonacept (ARCALYST®) is approved and marketed for cryopyrin-associated periodic syndrome (CAPS), in the United States by Regeneron Pharmaceuticals, Inc.; 2) Brucato et al. JAMA. 2016, 316 (18): 1906-1912; Arcalyst Prescribing Information; 3) IQVIA PharMetrics Plus Claims Data 1/1/2013-3/31/2018; ClearView Analysis, 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; 4) 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;

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

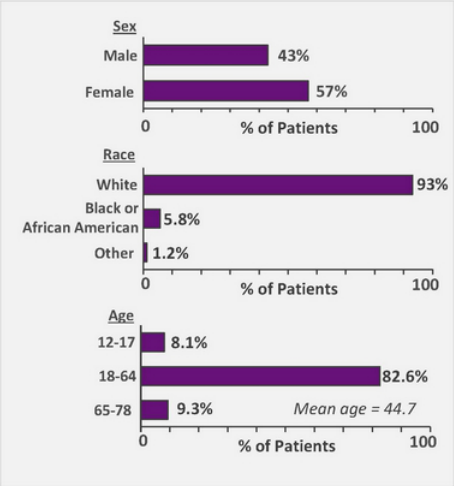
CRP = C-reactive protein; NRS = Numerical Rating Scale

Baseline Demographics and Clinical Characteristics

Pivotal Phase 3 Rilonacept Data

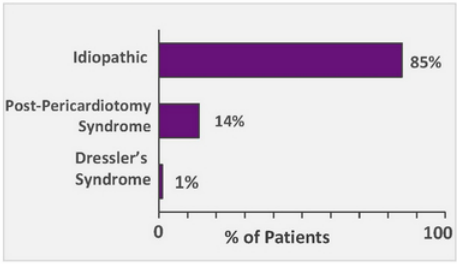
RHAPSODY

Baseline Demographics (n=86)

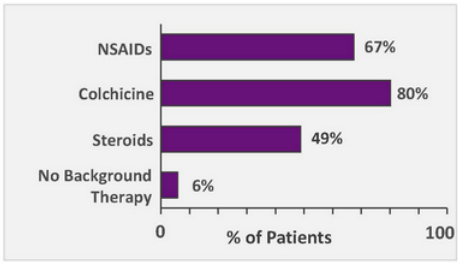


Total Number of Episodes Including Index and Qualifying Episodes	Run-in Period (n=86)
Mean	4.7

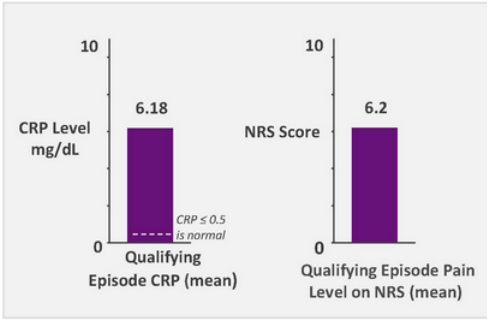
Prior Pericarditis History at Baseline (n=86)



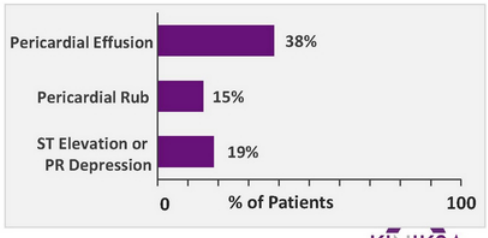
SoC Received at Qualifying Episode (n=86)



Qualifying Episode CRP & NRS (n=86)



Pericarditis Manifestations at Qualifying Episode (n=



KINIKSA

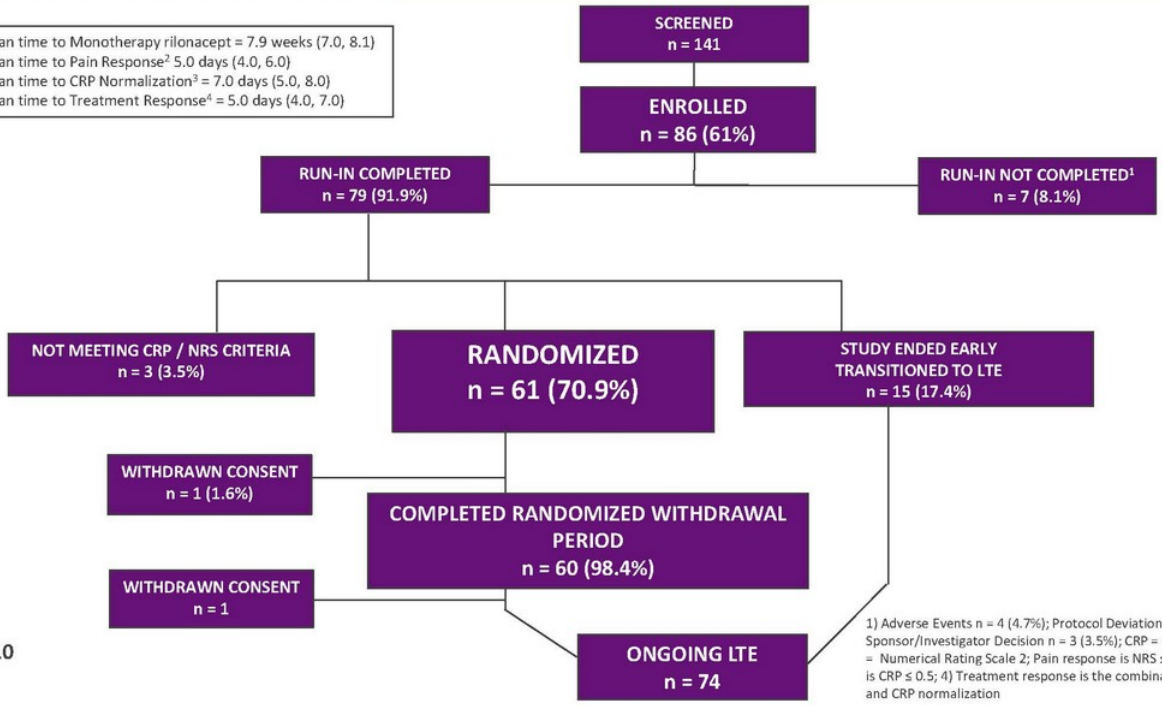
CRP = C-reactive protein; NRS = Numerical Rating Scale

Subject Disposition

Pivotal Phase 3 Rilonacept Data

RHAPSODY

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

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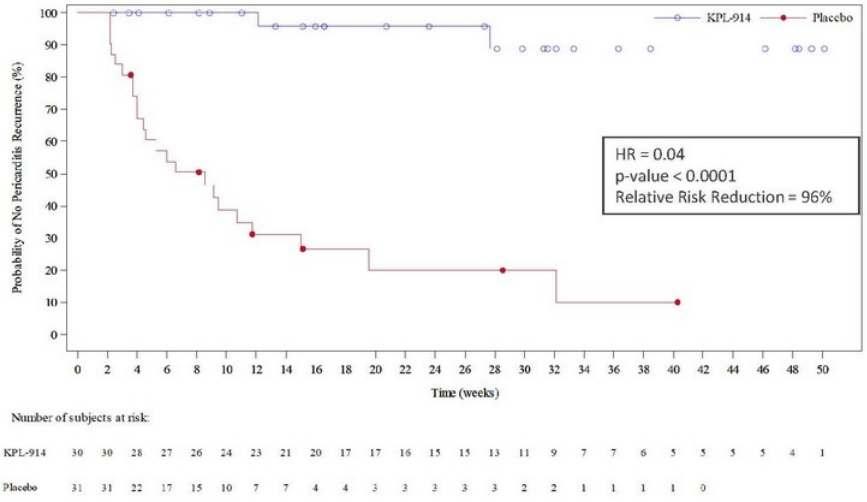
Primary Efficacy Endpoint: Time-to-First Adjudicated Pericarditis Recurrence

Pivotal Phase 3 Rilonacept Data

RHAPSODY

Figure 14.2.1.1 Kaplan-Meier Curves for Time to Pericarditis Recurrence based on CEC Adjudication
ITT Analysis Set



Number of subjects at risk:

KPL-914	30	30	28	27	26	24	23	21	20	17	17	16	15	15	13	11	9	7	7	6	5	5	5	5	4	1
Placebo	31	31	22	17	15	10	7	7	4	4	3	3	3	3	3	2	2	1	1	1	1	0				

Pericarditis Recurrence Categories, n (%)	KPL-914 (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)

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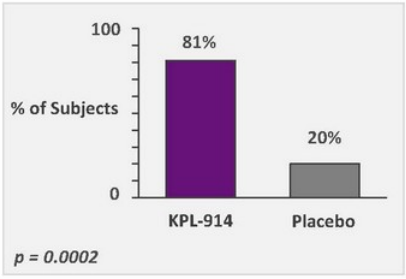
Secondary Endpoints at Week 16 of the Randomized Withdrawal Period

Pivotal Phase 3 Rilonacept Data

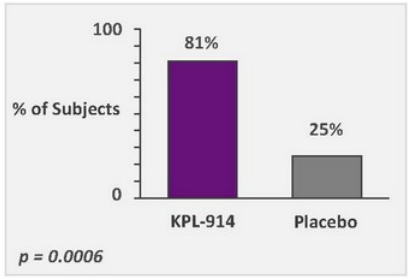
RHAPSODY

Proportion of Subjects Who Maintained Clinical Response [1]



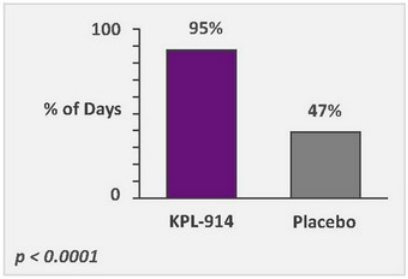
Data at Weeks 8 and 24 were consistent and statistically significant
(Week 8, p < 0.0001; Week 24, p=0.0022)

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.

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Summary of Adverse Events

Pivotal Phase 3 Rilonacept Data

RHAPSODY

	Run-In Period	Randomized Withdrawal Period	
Category [1]	KPL-914 (N=86) n (%)	KPL-914 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]	KPL-914 (N=86) n (%)	KPL-914 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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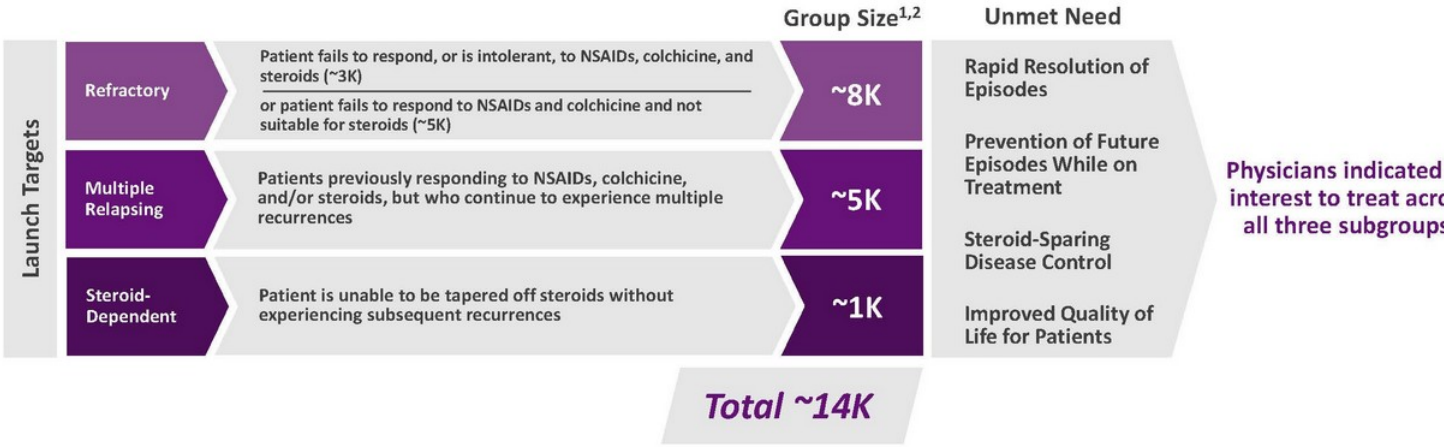
Recurrent Pericarditis U.S. Prevalence Estimated to be ~40K Patients

Addressable U.S. opportunity for rilonacept estimated to be ~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, Lalliberte 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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Product Candidates and Clinical Status

Indication	Program & Target	Preclinical	Phase 1	Phase 2	Phase 3	Status
Recurrent Pericarditis	**Rilonacept**[1] IL-1α & IL-1β					Pivotal Phase 3 Data Announced in June 2020
Giant Cell Arteritis	**Mavrilimumab** GM-CSFRα					Phase 2 Data Expected in Q4 2020
COVID-19 Pneumonia & Hyperinflammation	**Mavrilimumab** GM-CSFRα					Phase 2 Initiation Expected in Q3 2020
CAR T Induced Cytokine Release Syndrome[2]	**Mavrilimumab** GM-CSFRα					Phase 2 Initiation Expected in 2H 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) Rilonacept (ARCALYST®) is approved and marketed for cryopyrin-associated periodic syndrome, in the United States by Regeneron Pharmaceuticals, Inc.; 2) Clinical collaboration with Kite, a Gilead Company, in relapsed or refractory large B-cell lymphoma; 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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Appendix

Summary of Adverse Events

Pivotal Phase 3 Rilonacept Data

RHAPSODY

Category [1]	Run-In Period	Randomized Withdrawal Period	
	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)
Subjects with Any TEAEs	69 (80.2)	24 (80.0)	13 (41.9)
Blood and lymphatic system disorders	2 (2.3)	0	0
Eosinophilia	1 (1.2)	0	0
Lymphadenopathy	1 (1.2)	0	0
Cardiac disorders	5 (5.8)	0	2 (6.5)
Angina pectoris	1 (1.2)	0	0
Aortic valve incompetence	0	0	1 (3.2)
Atrial fibrillation	1 (1.2)	0	0
Cardiac flutter	0	0	1 (3.2)
Palpitations	1 (1.2)	0	0
Sinus tachycardia	1 (1.2)	0	0
Tachycardia	1 (1.2)	0	0
Ventricular dyssynchrony	1 (1.2)	0	0
Ear and labyrinth disorders	1 (1.2)	0	0
Middle ear effusion	0	0	0
Vertigo	1 (1.2)	0	0
Endocrine disorders	0	1 (3.3)	0
Hypothyroidism	0	1 (3.3)	0
Eye disorders	1 (1.2)	0	0
Diplopia	0	0	0
Eye inflammation	1 (1.2)	0	0
Gastrointestinal disorders	14 (16.3)	2 (6.7)	2 (6.5)
Abdominal distension	2 (2.3)	0	0
Abdominal pain	0	0	1 (3.2)
Abdominal tenderness	0	1 (3.3)	0
Aphthous ulcer	0	1 (3.3)	0
Constipation	1 (1.2)	0	0
Diarrhea	5 (5.8)	0	0
Gastric ulcer	1 (1.2)	0	0
Gastritis	1 (1.2)	0	0
Gastrointestinal disorder	1 (1.2)	0	0
Gastrooesophageal reflux disease	1 (1.2)	1 (3.3)	0
Gingival pain	1 (1.2)	0	0
Haemorrhoids	0	0	1 (3.2)
Ileus	0	0	0
Nausea	2 (2.3)	0	0
Tongue ulceration	0	1 (3.3)	0
Vomiting	1 (1.2)	0	0
General disorders and administration site conditions	30 (34.9)	10 (33.3)	1 (3.2)
Asthenia	2 (2.3)	0	0
Chest discomfort	1 (1.2)	1 (3.3)	0
Chills	1 (1.2)	0	0
Fatigue	2 (2.3)	2 (6.7)	0
Feeling abnormal	1 (1.2)	0	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) Includes malignancy excluding basal cell carcinoma of the skin

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Summary of Adverse Events

Pivotal Phase 3 Rilonacept Data

RHAPSODY

Category [1]	Run-In Period	Randomized Withdrawal Period	
	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)
Feeling hot	2 (2.3)	0	0
Injection site bruising	1 (1.2)	0	0
Injection site discolouration	2 (2.3)	0	0
Injection site erythema	18 (20.9)	6 (20.0)	0
Injection site inflammation	1 (1.2)	0	0
Injection site nodule	1 (1.2)	0	0
Injection site pain	4 (4.7)	0	0
Injection site pruritus	5 (5.8)	5 (16.7)	0
Injection site rash	3 (3.5)	0	0
Injection site reaction	2 (2.3)	0	0
Injection site swelling	5 (5.8)	1 (3.3)	0
Non-cardiac chest pain	1 (1.2)	3 (10.0)	1 (3.2)
Oedema peripheral	0	1 (3.3)	0
Pain	1 (1.2)	1 (3.3)	0
Pyrexia	1 (1.2)	0	0
Immune system disorders	1 (1.2)	0	1 (3.2)
Drug hypersensitivity	1 (1.2)	0	0
Hypersensitivity	1 (1.2)	0	0
Seasonal allergy	0	0	1 (3.2)
Infections and infestations	14 (16.3)	12 (40.0)	3 (9.7)
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
Gastrointestinal 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
Injury, poisoning and procedural complications	6 (7.0)	3 (10.0)	1 (3.2)
Epicondylitis	0	1 (3.3)	0
Fall	2 (2.3)	0	0
Humerus fracture	0	0	1 (3.2)

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) Includes malignancy excluding basal cell carcinoma of the skin

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Summary of Adverse Events

Pivotal Phase 3 Rilonacept Data

Category [1]	Run-In Period	Randomized Withdrawal Period	
	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)
Joint injury	0	1 (3.3)	0
Limb injury	0	0	1 (3.2)
Muscle strain	1 (1.2)	0	0
Post procedural contusion	0	1 (3.3)	0
Post-traumatic pain	2 (2.3)	0	0
Procedural dizziness	1 (1.2)	0	0
Investigations	12 (14.0)	7 (23.3)	0
Bacterial test	0	0	0
Blood cholesterol increased	0	1 (3.3)	0
Blood glucose decreased	0	1 (3.3)	0
Blood glucose increased	1 (1.2)	0	0
Blood pressure increased	1 (1.2)	1 (3.3)	0
Blood triglycerides increased	0	1 (3.3)	0
Body temperature decreased	1 (1.2)	0	0
C-reactive protein increased	1 (1.2)	2 (6.7)	0
Eosinophil count increased	1 (1.2)	0	0
Haemoglobin decreased	1 (1.2)	0	0
Heart rate increased	1 (1.2)	1 (3.3)	0
Hepatic enzyme increased	1 (1.2)	1 (3.3)	0
Heart density lipoprotein decreased	1 (1.2)	0	0
Heart density lipoprotein increased	0	3 (10.0)	0
Lipids increased	0	2 (6.7)	0
Liver function test increased	1 (1.2)	0	0
Low density lipoprotein increased	1 (1.2)	0	0
Mean cell volume increased	0	1 (3.3)	0
Smear cervix abnormal	1 (1.2)	0	0
Weight increased	1 (1.2)	0	0
Metabolism and nutrition disorders	0	1 (3.3)	0
Hyperlipidaemia	0	1 (3.3)	0
Musculoskeletal and connective tissue disorders	26 (30.2)	6 (20.0)	4 (12.9)
Arthralgia	8 (9.3)	1 (3.3)	0
Arthritis	0	1 (3.3)	0
Axillary mass	0	1 (3.3)	0
Back pain	3 (3.5)	1 (3.3)	0
Groin pain	1 (1.2)	0	0
Joint stiffness	2 (2.3)	0	0
Musculoskeletal chest pain	3 (3.5)	1 (3.3)	4 (12.9)
Musculoskeletal pain	3 (3.5)	0	0
Myalgia	9 (10.5)	1 (3.3)	0
Neck pain	1 (1.2)	0	1 (3.2)
Osteoarthritis	1 (1.2)	0	0
Pain in extremity	1 (1.2)	0	0
Neoplasms benign, malignant and unspecified (incl cysts and polyps)	1 (1.2)	2 (6.7)	0
Acrochordon	1 (1.2)	0	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) Includes malignancy excluding basal cell carcinoma of the skin

Summary of Adverse Events

Pivotal Phase 3 Rilonacept Data

	Run-In Period	Randomized Withdrawal Period	
Category [1]	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)
Lipoma	0	1 (3.3)	0
Squamous cell carcinoma	0	1 (3.3)	0
Nervous system disorders	14 (16.3)	2 (6.7)	0
Carpal tunnel syndrome	1 (1.2)	0	0
Cerebrovascular accident	1 (1.2)	0	0
Dizziness	2 (2.3)	1 (3.3)	0
Dysgeusia	1 (1.2)	0	0
Head discomfort	0	1 (3.3)	0
Headache	7 (8.1)	0	0
Migraine	1 (1.2)	0	0
Presyncope	1 (1.2)	0	0
Somnolence	1 (1.2)	0	0
Psychiatric disorders	1 (1.2)	0	1 (3.2)
Insomnia	0	0	1 (3.2)
Sleep disorder	1 (1.2)	0	0
Renal and urinary disorders	0	1 (3.3)	1 (3.2)
Nephrolithiasis	0	1 (3.3)	0
Renal colic	0	0	1 (3.2)
Reproductive system and breast disorders	1 (1.2)	1 (3.3)	1 (3.2)
Ovarian cyst	1 (1.2)	0	0
Uterine haemorrhage	0	1 (3.3)	0
Uterine polyp	0	0	1 (3.2)

	Run-In Period	Randomized Withdrawal Period	
Category [1]	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)
Respiratory, thoracic and mediastinal disorders	15 (17.4)	7 (23.3)	1 (3.2)
Alveolitis allergic	1 (1.2)	0	0
Cough	5 (5.8)	1 (3.3)	0
Dysphonia	0	1 (3.3)	0
Dyspnoea	1 (1.2)	1 (3.3)	0
Epistaxis	1 (1.2)	0	0
Nasal congestion	0	0	0
Oropharyngeal pain	1 (1.2)	3 (10.0)	0
Pharyngeal hypoaesthesia	1 (1.2)	0	0
Respiratory tract congestion	2 (2.3)	0	1 (3.2)
Rhinorrhoea	1 (1.2)	0	0
Sinus congestion	2 (2.3)	2 (6.7)	0
Skin and subcutaneous tissue disorders	11 (12.8)	0	1 (3.2)
Acne	1 (1.2)	0	0
Alopecia	1 (1.2)	0	0
Angioedema	1 (1.2)	0	0
Erythema	2 (2.3)	0	0
Pruritus	2 (2.3)	0	0
Pruritus generalised	2 (2.3)	0	1 (3.2)
Rash	1 (1.2)	0	0
Rash macular	3 (3.5)	0	0
Social circumstances	0	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) Includes malignancy excluding basal cell carcinoma of the skin

Summary of Adverse Events

Pivotal Phase 3 Rilonacept Data

	Run-In Period	Randomized Withdrawal Period	
Category [1]	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)
Menopause	0	1 (3.3)	0
Vascular disorders	2 (2.3)	1 (3.3)	1 (3.2)
Hypertension	2 (2.3)	1 (3.3)	1 (3.2)

	Run-In Period	Randomized Withdrawal Period	
Category [1]	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)
Subjects with Any Serious TEAE	1 (1.2)	1 (3.3)	1 (3.2)
Cardiac disorders	0	0	1 (3.2)
Cardiac flutter	0	0	1 (3.2)
Gastrointestinal disorders	0	0	0
Ileus	0	0	0
General disorders and administration site conditions	0	0	0
Pyrexia	0	0	0
Neoplasms benign, malignant and unspecified (incl cysts and polyps)	0	1 (3.3)	0
Squamous cell carcinoma	0	1 (3.3)	0
Nervous system disorders	1 (1.2)	0	0
Cerebrovascular accident	1 (1.2)	0	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) Includes malignancy excluding basal cell carcinoma of the skin