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): **January 13, 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)

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

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

☐ 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 2.02. Results of Operations and Financial Condition.

On January 13, 2020, Kiniksa Pharmaceuticals, Ltd. (the "Company") issued a press release that included an announcement of the Company's cash, cash equivalents and short-term investments as of December 31, 2019. A copy of the press release is furnished with this Current Report on Form 8-K as Exhibit 99.1.

The information contained in this Item 2.02 of this Current Report on Form 8-K and Exhibit 99.1 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 (the "Securities Act"), 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 7.01. Regulation FD Disclosure.

As previously announced, the Company will present at the 38th Annual J.P. Morgan Healthcare Conference (the "JPM HC Conference") on January 13, 2020, which will be webcast live. A copy of the presentation is furnished with this Current Report on Form 8-K as Exhibit 99.2.

The information contained in this Item 7.01 of this Current Report on Form 8-K and Exhibit 99.2 shall not be deemed "filed" for purposes of Section 18 of 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 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

Exhibit No.	Description
99.1	Press Release issued by Kiniksa Pharmaceuticals, Ltd. dated January 13, 2020
99.2	Presentation by Kiniksa Pharmaceuticals, Ltd. for the JPM HC Conference dated January 13, 2020

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: January 13, 2020

By: /s/ Thomas Beetham
Thomas Beetham
Chief Legal Officer

Exhibit 99.1

Kiniksa Announces Pipeline Progress and Reiterates 2020 Clinical Data Readouts

- Enrollment target achieved for rilonacept Phase 3 trial in recurrent pericarditis; top-line data expected in 2H 2020 -

- Enrollment target achieved for mavrilimumab Phase 2 trial in giant cell arteritis; top-line data expected in 2H 2020 -

- Top-line data from KPL-716 Phase 2a trial in prurigo nodularis expected in 1H 2020 -

- Interim data from cohorts of KPL-716 Phase 2 trial in diseases characterized by chronic pruritus expected in 1H 2020 -

- Top-line data from KPL-404 first-in-human study with antigen challenge TDAR expected in 2H 2020 -

HAMILTON, BERMUDA – January 13, 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, today announced recent pipeline progress and upcoming 2020 clinical milestones. Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa will provide further detail in a corporate presentation at the 38th Annual J.P. Morgan Healthcare Conference today, Monday, January 13, 2020 at 2:00 p.m. Pacific Time / 5:00 p.m. Eastern Time at the Westin St. Francis Hotel in San Francisco, California.

“Kiniksa’s clinical-stage assets, including rilonacept, mavrilimumab, KPL-716 and KPL-404, are on track to generate clinical data this year,” said Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa. “Each readout has the potential to be an important value-driving event and to help inform our portfolio strategy and capital allocation decisions. We have the capital to achieve multiple clinical data readouts and project that our 2019 year-end cash reserves of approximately $233 million will fund our operating plan into the second half of 2021.”

Recent Pipeline Progress and Expected 2020 Clinical Data Readouts
Rilonacept (IL-1α and IL-1β cytokine trap)

- Kiniksa has achieved its enrollment target for RHAPSODY, a global, randomized-withdrawal design, pivotal Phase 3 trial of rilonacept in patients with recurrent pericarditis. The company expects top-line data in the second half of 2020.
- Kiniksa recently announced the U.S. Food and Drug Administration (FDA) granted Breakthrough Therapy designation for rilonacept for the treatment of recurrent pericarditis. Kiniksa’s Breakthrough Therapy application was based on final data from an open-label Phase 2 clinical trial of rilonacept in a range of recurrent pericarditis populations.

Mavrilimumab (monoclonal antibody inhibitor targeting GM-CSFRα)

- Kiniksa has achieved its enrollment target for a global Phase 2 proof-of-concept trial of mavrilimumab in patients with giant cell arteritis (GCA). The company expects top-line data in the second half of 2020.
- Kiniksa and Kite, a Gilead company (Kite), recently announced a clinical collaboration evaluating the investigational combination of Yescarta® (axicabtagene ciloleucel) and mavrilimumab in relapsed or refractory large B-cell lymphoma. The objective of the study is to determine the effect of mavrilimumab on the safety of Yescarta. Preclinical evidence shows the potential for interruption of granulocyte macrophage colony stimulating factor (GM-CSF) signaling to disrupt chimeric antigen receptor T (CAR T) cell-mediated inflammation without disrupting anti-tumor efficacy.

KPL-716 (monoclonal antibody inhibitor of signaling through OSMRβ)

- Kiniksa expects top-line data from a Phase 2a clinical trial of KPL-716 in patients with prurigo nodularis in the first half of 2020.
- Kiniksa expects interim data from cohorts of a Phase 2 clinical trial of KPL-716 in diseases characterized by chronic pruritus in the first half of 2020.

KPL-404 (monoclonal antibody inhibitor of signaling between CD40 and CD40L)

- Kiniksa is enrolling and dosing subjects in a single-ascending-dose Phase 1 clinical trial of KPL-404 in healthy volunteers. The first-in-human trial will provide safety data and pharmacokinetics as well as receptor occupancy and T-cell Dependent Antibody Response (TDAR). Top-line data are expected in the second half of 2020.

KPL-045 (monoclonal antibody inhibitor of the CD30L co-stimulatory molecule)

- Based on preclinical data in the context of Kiniksa's portfolio, the company no longer plans to progress KPL-045 into clinical development.

Financial Guidance

Kiniksa ended 2019 with approximately $233 million in cash, cash equivalents and short-term investments (unaudited). The company expects that these reserves will fund its operating plan into the second half of 2021.

Presentation at the 38th Annual J.P. Morgan Healthcare Conference

Kiniksa will webcast its corporate presentation at the 38th Annual J.P. Morgan Healthcare Conference today, Monday, January 13, 2020 at 2:00 p.m. Pacific Time / 5:00 p.m. Eastern Time. A live webcast of Kiniksa's presentation will be accessible through the Investors & Media section of the company's website (www.kiniksa.com). A replay of the webcast will be available on Kiniksa's website for 14 days following the conference.

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 has a pipeline of product candidates across various stages of development, focused on autoinflammatory and autoimmune conditions. For more information, please visit www.kiniksa.com.

About Rilonacept
Rilonacept is a weekly, subcutaneously-injected, recombinant fusion protein that blocks interleukin-1 alpha (IL-1α) and interleukin-1 beta (IL-1β) signaling. 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 Cryopyrin-Associated Periodic Syndromes (CAPS), which includes Familial Cold Autoinflammatory Syndrome and Muckle-Wells Syndrome. 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. Kiniksa exclusively licensed rilonacept from Regeneron for recurrent pericarditis and certain other indications. Rilonacept in recurrent pericarditis is an investigational drug. The FDA has granted Breakthrough Therapy designation to rilonacept for recurrent pericarditis.

About Mavrilimumab
Mavrilimumab is an investigational fully-human monoclonal antibody that is designed to antagonize GM-CSF signaling by binding to the alpha subunit of the GM-CSF receptor. Kiniksa's lead indication for mavrilimumab is GCA, an inflammatory disease of medium to large arteries. Additionally, Kiniksa and Kite have a clinical collaboration to evaluate mavrilimumab in combination with Yescarta® (axicabtagene ciloleucel) in patients with relapsed or refractory large B-cell lymphoma.

About KPL-716
KPL-716 is an investigational fully-human monoclonal antibody that targets oncostatin M receptor beta (OSMRβ), which mediates signaling of interleukin-31 (IL-31) and oncostatin M (OSM), two key cytokines implicated in pruritus, inflammation and fibrosis. Kiniksa believes KPL-716 to be the only monoclonal antibody in development that targets both pathways simultaneously.

About KPL-404
KPL-404 is an investigational humanized monoclonal antibody that is designed to inhibit CD40-CD40 ligand (CD40L) interaction, a key T-cell co-stimulatory signal critical for B-cell maturation and immunoglobulin class switching. Kiniksa believes disrupting CD40-CD40L interaction is an attractive approach for blocking T-cell mediated, B-cell driven responses, drivers of multiple autoimmune disease pathologies such as Sjogren's syndrome, systemic lupus erythematosus, rheumatoid arthritis, solid organ transplant and Graves' disease.

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 expectations for fiscal year 2020; plans and timing of enrollment of our clinical trials; proposed indications for the investigation of our product candidates; our beliefs about the approach of our product candidates and potential impact; our clinical collaboration with Kite evaluating the combination of Yescarta® and mavrilimumab; the potential for mavrilimumab with respect to CAR T cell mediated inflammation and otherwise; plans and timing to report or present preliminary, interim and final top-line clinical trial data and the potential impact of that data, including on our portfolio strategy and capital allocation; expected cash, cash equivalents and short-term investments at fiscal year-end 2019; projected timeframe for funding our operating plan with current cash, cash equivalents and short-term investments; and our expectations around not needing to raise additional capital prior to delivering our anticipated 2020 clinical trial data readouts.

These forward-looking statements are based on management’s current plans, estimates or 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: potential delays or difficulty in enrollment of patients in, and activation of sites for, our clinical trials; potential complications in coordinating among requirements, regulations and guidelines of regulatory authorities across a number of jurisdictions for our global clinical trials; potential amendments to our clinical trial protocols initiated by us or required by regulatory authorities; potential delays or difficulty in completing our clinical trials, including as a result of our clinical trial design; potential for lower accrual of events in our clinical trials; potential undesirable side effects caused by our product candidates; our potential inability to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities or otherwise producing negative, inconclusive or commercially uncompetitive results; potential for changes between final data and any preliminary and interim “top-line” data we announce; impact of additional data from us or other companies; our potential inability to replicate in later clinical trials positive results from our earlier pre-clinical and clinical trials; drug substance and/or drug product shortages caused by issues at our third-party manufacturers’ facilities; our reliance on certain third parties as the sole source of supply of the drug substance and drug products used in our product candidates; our reliance on third parties to conduct our research, pre-clinical studies, clinical trials, and other trials for our product candidates; changes in our operating plan and funding requirements; changes in the capital markets; market reaction to our anticipated 2020 clinical trial data readouts; substantial 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 November 5, 2019 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 plans, estimates, or expectations 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.

ARCALYST® is a registered trademark of Regeneron Pharmaceuticals, Inc. and Yescarta® is a registered trademark of Gilead Sciences, Inc., or its related companies.

Every Second Counts!™

Kiniksa Investor and Media Contact
Mark Ragosa
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mragosa@kiniksa.com

Exhibit 99.2


KINIKSA
Every Second Counts!™
J.P. Morgan Healthcare Conference
January 13, 2020

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 market opportunities and competitive position; clinical trials and other studies; timing and potential impact of clinical data; regulatory and other submissions; commercial strategy and pre-commercial activities; expected cash, cash equivalents and short-term investments for FY 2019; 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; substantial new or existing competition; and our ability to attract and retain qualified personnel. These and 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 5, 2019 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

KINIKSA

Every Second Counts!™

- Focused on unmet need in autoimmune and autoinflammatory diseases
- Product candidates based on validated mechanisms and/or strong biologic rationale
- Target underserved conditions and offer potential differentiation
- Allocate capital across portfolio relative to opportunity

Pipeline: Multiple Clinical Stage Assets

1st Indication	Program & Target	Preclinical	Phase 1	Phase 2	Phase 3	Stage
Recurrent Pericarditis *Autoinflammatory cardiovascular disease*	**Rilonacept[1]** IL-1α & IL-1β					Pivotal Phase 3 Study (RHAPSODY)
Giant Cell Arteritis (GCA) *Chronic inflammatory arterial disease*	**Mavrilimumab (Mavri)** GM-CSFRα					Global Phase 2 Study
Prurigo Nodularis (PN) *Chronic inflammatory skin disease*	**KPL-716** OSMRβ					Phase 2 Study
Multiple Diseases Characterized by Chronic Pruritus[2]	**KPL-716** OSMRβ					Phase 2 Study
Severe Autoimmune Diseases	**KPL-404** CD40					Phase 1 Study

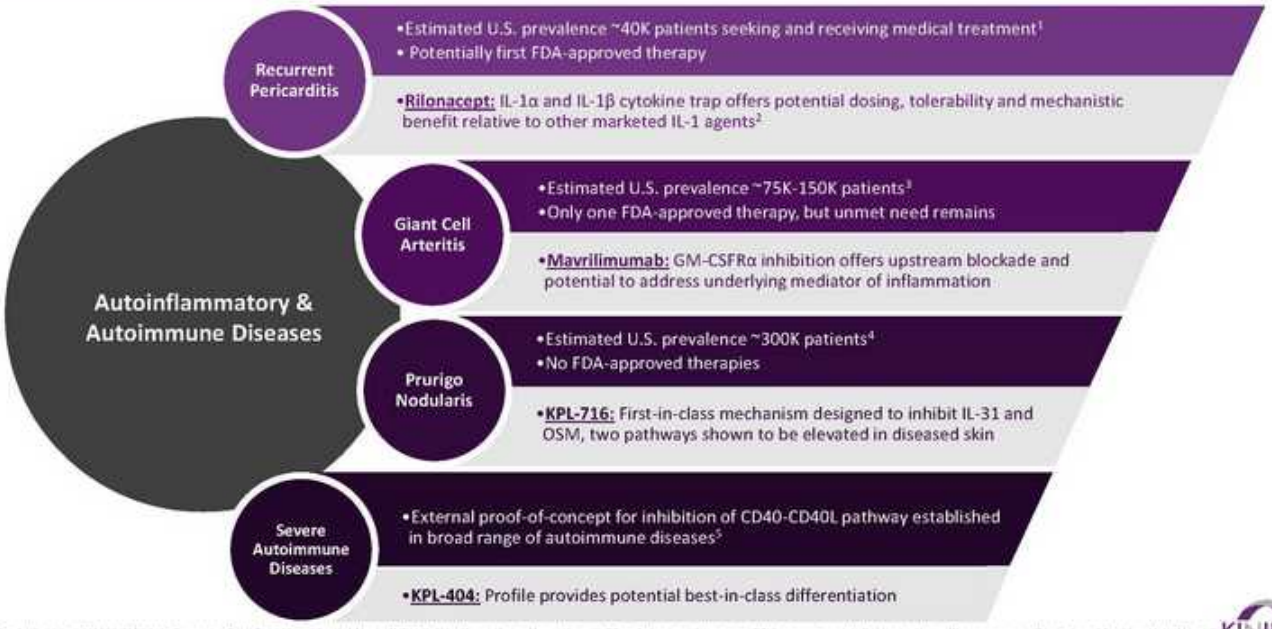
1) Rilonacept (ARCALYST®) is approved and marketed for cryopyrin-associated periodic syndrome, in the United States by Regeneron Pharmaceuticals, Inc. BLA transfers to Kiniksa after receipt of positive Phase 3 clinical data; 2) Chronic Idiopathic Pruritus, Chronic Idiopathic Urticaria, Plaque Psoriasis, Lichen Simplex Chronicus, Lichen Planus; IL-1® = interleukin-1®; IL-13 = interleukin 13; GM-CSFRα = granulocyte macrophage colony stimulating factor alpha; OSMRβ = oncostatin M receptor beta

Clinical-Stage Assets Based on Validated Mechanisms and/or Strong Biologic Rationale

Mechanism of Action	Rationale	Initial Indication
Rilonacept IL-1α and IL-1β cytokine trap	IL-1α and IL-1β cytokines shown to play key role in inflammatory diseases[1]	Phase 2 data in **recurrent pericarditis** showed resolution of pericarditis episodes, reduction in recurrences while on treatment, and tapering/discontinuation of corticosteroids[6]
Mavrilimumab monoclonal antibody inhibitor blocking GM-CSFRα signaling	GM-CSF is a key growth factor and cytokine in autoinflammation and autoimmunity[2]	GM-CSF and GM-CSFRα are highly expressed in biopsies of **giant cell arteritis** patients vs. normal healthy controls[7]
KPL-716 monoclonal antibody inhibitor targeting OSMRβ	IL-31 and oncostatin M are key cytokines implicated in prurigo nodularis[3]	IL-31, OSM and OSMRβ mRNA are upregulated in lesional biopsies of **prurigo nodularis** subjects vs. normal healthy controls[3]
KPL-404 monoclonal antibody inhibitor of CD40 / CD40L interaction	CD40-CD40L interaction is an attractive mechanism for targeting T-cell mediated, B-cell driven autoimmune diseases[4,5]	External proof-of-concept for inhibition of pathway has been established in a broad range of **autoimmune diseases**[8]

1) Dinarello CA, et al. Nat Rev Drug Discov 2012;11:633-652 and Brucato A, et al. Int Emerg Med 2018; 13:839–844; 2) Wicks, Roberts, Nature Review Immunology, 2015; Hamilton, Expert Review of Clinical Immunology, 11:4, 457-465; 3) Poster presentation at the 28th European Academy of Dermatology and Venereology (EADV): *IL-31 is Implicated in the Pathogenesis of Prurigo Nodularis, a Chronic Pruritic Skin Disease that can Exist Irrespective of Co-morbid Conditions (LOTUS-PN Study)*; 4) Elgueta, et al. Immunol Rev 2009. 229 (1), 152-172; 5) Peters, et al. Semin Immunol 2009, 21 (5) 293-300; 6) Final open-label Phase 2 data - Poster presentation at American Heart Association (AHA) Scientific Sessions 2019: Efficacy and Safety of Rilonacept in Recurrent Pericarditis: A Multicenter Phase 2 Clinical Trial; 7) Poster presentation at European Congress of Rheumatology 2019 (EULAR): *GM-CSF Pathway Signature Identified in Temporal Artery Biopsies of Patients With Giant Cell Arteritis*; 8) National Center for Biotechnology Information - *Targeting the CD40-CD154 Signaling Pathway for Treatment of Autoimmune Arthritis*: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6721639/

Clinical-Stage Assets Target Underserved Diseases and Offer Potential Differentiation



Multiple Clinical Data Readouts Expected in 2020

KPL-716 – Phase 2 (monoclonal antibody inhibitor targeting OSMRβ)	**Prurigo Nodularis** (Top-line Phase 2 Data)	**1H 2020**
KPL-716 – Phase 2 (monoclonal antibody inhibitor targeting OSMRβ)	**Diseases Characterized by Chronic Pruritus** (Interim Phase 2 Data in Limited Cohorts)	**1H 2020**
Rilonacept – Phase 3 (IL-1α and IL-1β cytokine trap) RHAPSODY	**Recurrent Pericarditis** (Pivotal Top-line Phase 3 Data)	**2H 2020**
Mavrilimumab – Phase 2 (monoclonal antibody inhibitor targeting GM-CSFRα)	**Giant Cell Arteritis** (Top-line Phase 2 Data)	**2H 2020**
KPL-404 – Phase 1 (monoclonal antibody inhibitor of CD40-CD40L interaction)	**Healthy Subjects** (Top-line Phase 1 Data)	**2H 2020**

KINIKSA

Rilonacept – Phase 3

RHAPSODY

Rilonacept | Mavrilimumab | KPL-716 | KPL-404

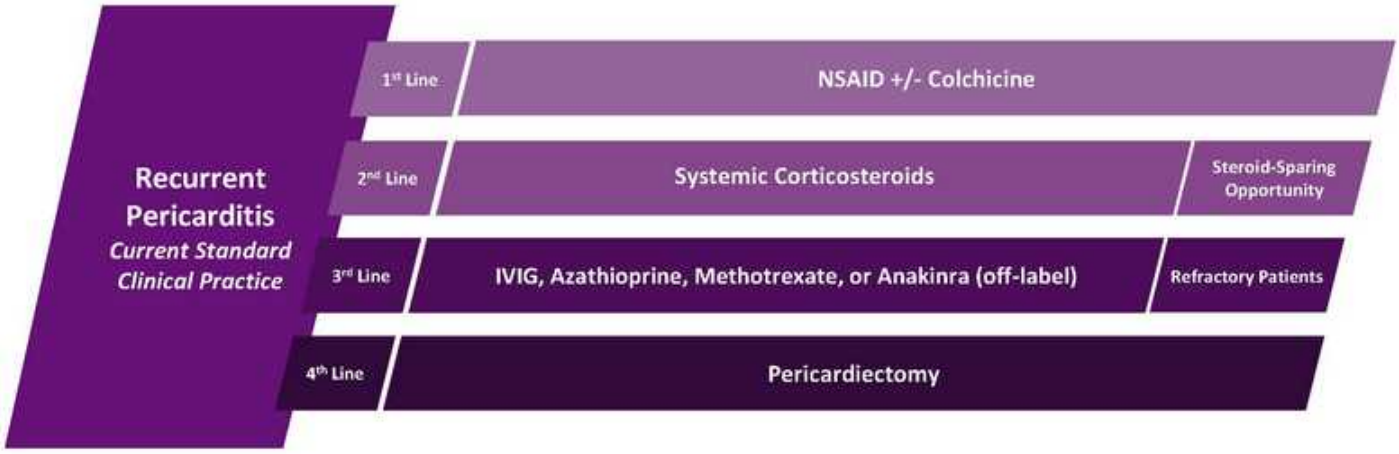
First Indications	**CAPS**[4]: already approved; **Recurrent Pericarditis**: Painful autoinflammatory cardiovascular disease
Mechanism of Action[1]	IL-1α and IL-1β cytokine trap
Scientific Rationale[1]	IL-1α and IL-1β are cytokines shown to play key role in inflammatory diseases
Prevalence[2]	~40k prevalent in U.S.; addressable opportunity of ~14k in U.S.
Competition[3]	No FDA-approved therapies for recurrent pericarditis
Status	**Breakthrough Therapy designation granted**; target enrollment achieved in pivotal Phase 3 clinical trial; developed by Regeneron and approved for CAPS[4]
Rights	Worldwide (excluding MENA); BLA transfers to Kiniksa after receipt of positive Phase 3 clinical data

1) Brucato et al. JAMA. 2016, 316 (18): 1906-1912; Arcalyst Prescribing Information; 2) 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; 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; Cardel et al. Arthritis Research & Therapy 2010, 12:R192; Hong et al. Lancet Oncol 2014, 15: 656-666; 4) Rilonacept (ARCALYST®) is approved and marketed for cryopyrin-associated periodic syndrome (CAPS), in the United States by Regeneron Pharmaceuticals, Inc. We will assume the rights to this indication upon receiving approval for rilonacept in the recurrent pericarditis indication.

KINIKSA

Recurrent Pericarditis Patients Currently Have Limited Treatment Options

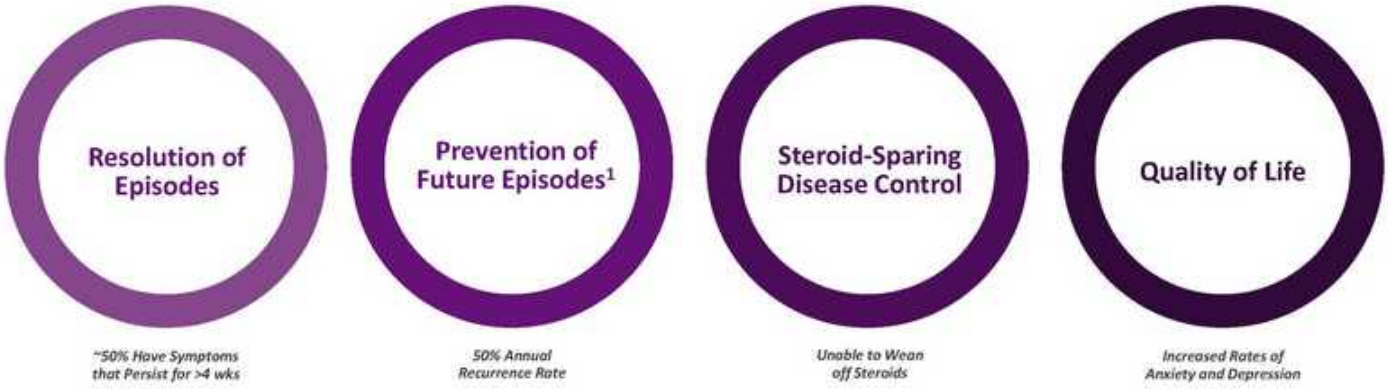
Patients deemed recurrent if symptomatic after symptom-free period of 4-6 weeks; 20-30% recurrent



Sources: 2015 ESC Guidelines for the Diagnosis and Management of Pericardial Diseases. Eur Heart J 2015; Aug 29; Trinity Partners, Kiniksa Analysis

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

Key areas of unmet need patients are seeking to address



Physicians and patients often resort to opioids to manage the pain

"The worst thing about pericarditis is its unpredictability and its chronicity. It's a permanent condition, so it has the potential to impact everything…work, exercise, family plans, travel." - Patient quote, 2019

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

Clinical Development Plan for Rilonacept in Recurrent Pericarditis

Designed to generate data on clinically meaningful outcomes

Phase 2 ✓

- Open-label, 5-part clinical trial with rilonacept in range of pericarditis populations
- Provided first evidence that rilonacept treatment improved clinically meaningful outcomes in study[1]
- Rilonacept was well-tolerated in study, with safety profile consistent with FDA-approved label for CAPS[2]

Completed

Phase 3 (RHAPSODY)

- **Target enrollment achieved**
- Pivotal clinical trial of rilonacept for treatment of recurrent pericarditis
- 24-week, double-blind, placebo-controlled, randomized-withdrawal (RW) study with open-label extension
- Primary efficacy endpoint is time-to-first-adjudicated pericarditis-recurrence in the RW period
- Continuing to enroll for limited time to help facilitate the number of primary efficacy endpoint events

Top-line data expected 2H 2020

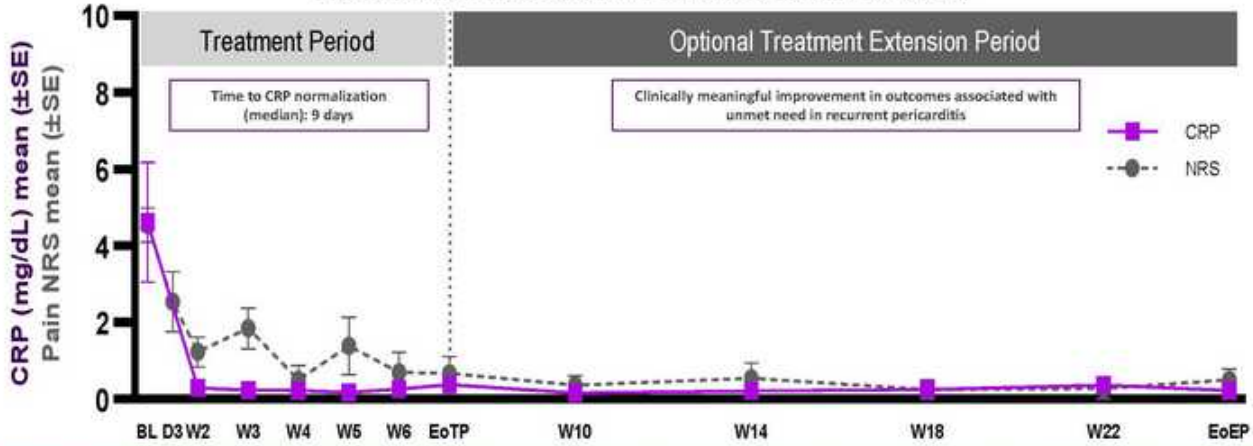
Rilonacept in Recurrent Pericarditis is for Investigational Use Only; Kiniksa Clinical Protocols, Phase 2: KPL-914-C001, Phase 3: KPL-914-C002; NCT03737110

1) Final open-label Phase 2 data - Poster presentation at American Heart Association (AHA) Scientific Sessions 2019: *Efficacy and Safety of Rilonacept in Recurrent Pericarditis: A Multicenter Phase 2 Clinical Trial*; 2) CAPS = Cryopyrin-Associated Periodic Syndromes

Final Phase 2 Rilonacept Data: Resolution of Pericarditis Episodes in Symptomatic Patients

Rapid and sustained reduction in reported pain and inflammation after first dose; persistent and clinically meaningful response throughout 6-month study

Symptomatic Recurrent Pericarditis Patients with Elevated CRP[1] (n=13)



		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

1] 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; CRP = C-Reactive Protein; NRS = Numeric Rating Scale

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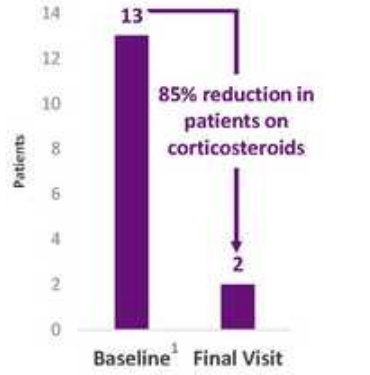
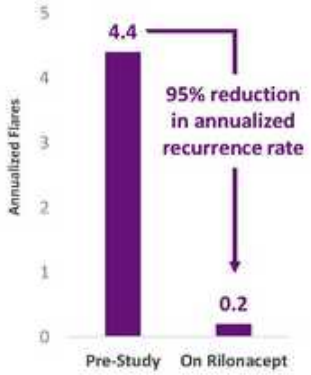
Phase 2 Rilonacept Data: Discontinuation of Corticosteroids, Decrease in Incidence of Pericarditis Episodes While on Treatment and Improvement in Quality of Life Scores

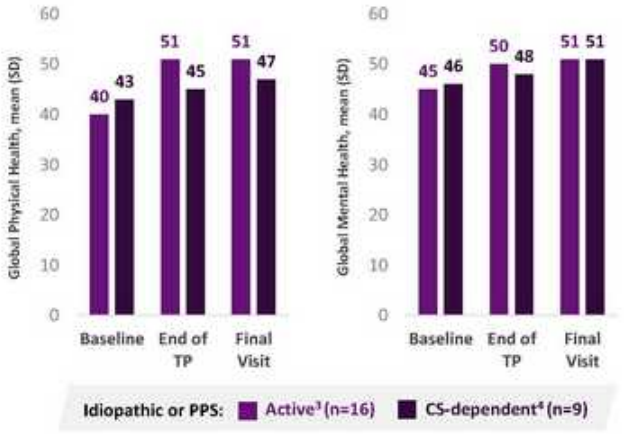
Discontinuation of Corticosteroids Without Pericarditis Recurrence



Decrease in Annualized Incidence of Pericarditis Episodes While on Treatment



Improved Quality of Life Scores[2]



1) 15 recurrent pericarditis patients on corticosteroids at baseline enrolled in the 6-week base treatment period, and 13 continued into the optional 18-week extension treatment period and completed 24 weeks of treatment; 2) PROMIS® - Patient Reported Outcomes Measurement Information System. The higher the score, the better global health is. US national average score for Global Physical and Mental Health is 50 (SD 10); 3) Parts 1, 2, and 4; 4) Parts 3 and 5

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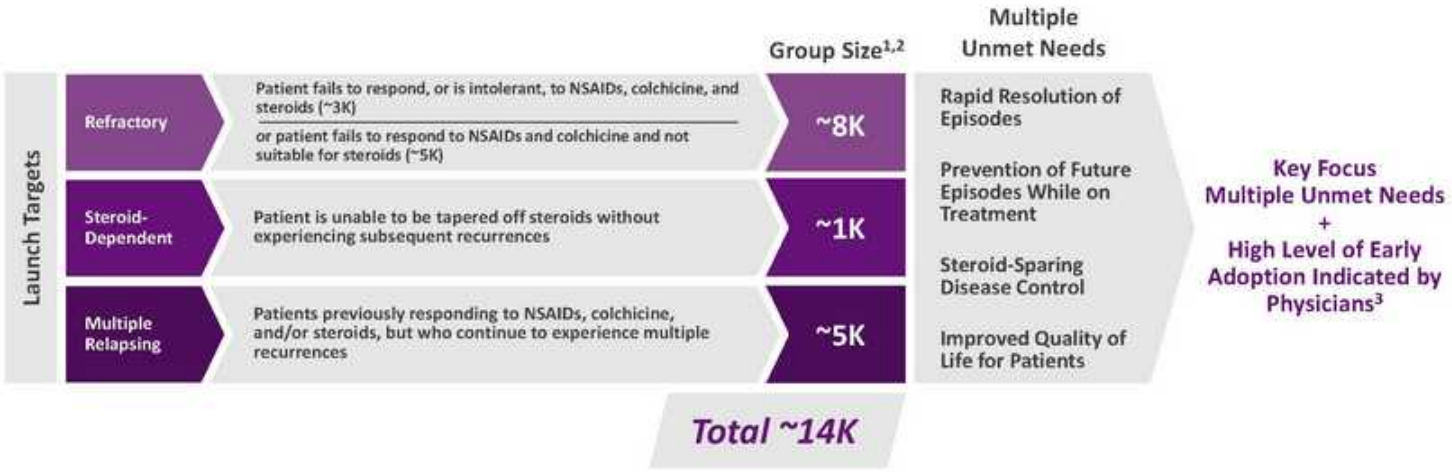
Commercial Opportunity: U.S. Prevalence Estimated to be ~40K RP Patients

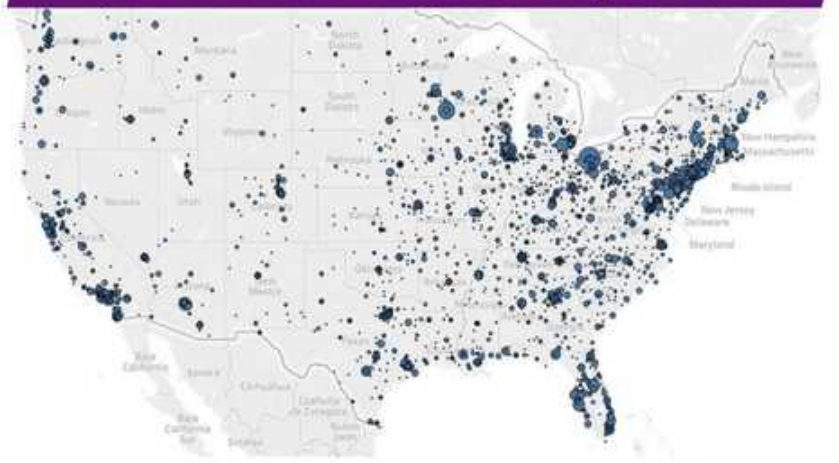
Addressable U.S. opportunity for rilonacept estimated to be ~14K patients



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, Der Sarkissian M, Magestro M, Cavanaugh C, LaBlanc F, Lejuez 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

Commercial Strategy
Focus on high-volume specialists

Recurrent Pericarditis Patient Volume by Account



Commercialization Plan Linked to Opportunity

- Specialty cardiology sales force of ~30 reps to call on high volume specialists
- Supported by current Kiniksa MSL team
- Efficient digital marketing to educate lower volume specialists
- Robust patient services capabilities to maintain appropriate patients on therapy
- Market research: duration of therapy expected to be at least 6-12 months
- Pricing in-line with high unmet need in rare disease

Source: IQVIA HPD Targeting Data, Komodo INTERGRITY Data, & Decision Resources Group RWD Data.

Mavrilimumab – Phase 2

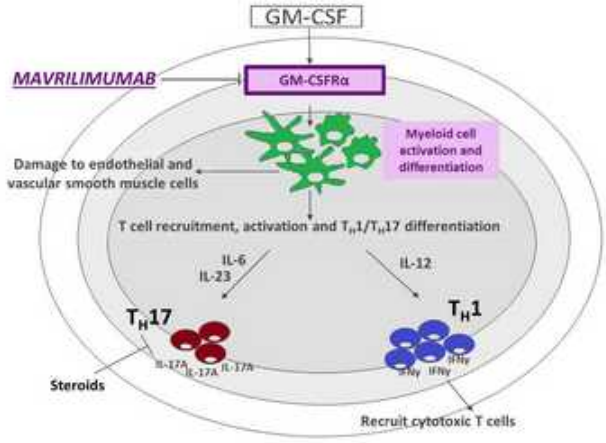
Rilonacept | **Mavrilimumab** | KPL-716 | KPL-404

First Indication	**Giant Cell Arteritis:** Chronic inflammatory disease of medium-large arteries (can lead to blindness)
Mechanism of Action[1]	Monoclonal antibody inhibitor targeting GM-CSFRα
Scientific Rationale[2,3]	Reported data implicate GM-CSF is key growth factor and cytokine in GCA
Prevalence[4]	~75k - 150k prevalent in U.S.; similar prevalence in other major markets
Competition[5]	Only one FDA-approved therapy for GCA and unmet needs remain
Status	Enrollment target achieved in global Phase 2 clinical trial; collaboration with Kite Gilead in R/R LBCL[6]
Rights	Worldwide

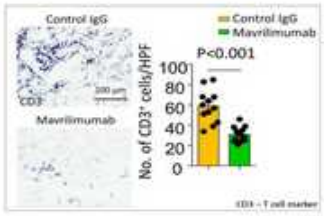
1) Sources: Wicks, Roberts, Nature Review Immunology, 2015; Hamilton, Expert Review of Clinical Immunology, 11:4, 457-465; 2) Lemaire et al. Journal of Leukocyte Biology, 1996; 60(4):500-18; 3) Wicks & Roberts. Nature Reviews. Rheumatology, 2016; 12(1):37-48; 4) Chandran et al., Scand J Rheumatol, 2015; Trinity Consulting – HCUP/Medicare Data, Quantitative Survey (n=102 rheumatologists); 5) Cortellis.; UpToDate; Correspondence. Trial of Tocilizumab in Giant-Cell Arteritis, NEJM, 2017; 6) relapsed or refractory large B-cell lymphoma

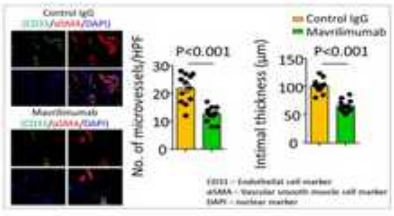
2019 Preclinical Data Support the Mechanistic Rationale of Targeting GM-CSF in GCA

EULAR June 2019: GM-CSF and its receptor shown to be elevated in GCA biopsies compared to control[1]

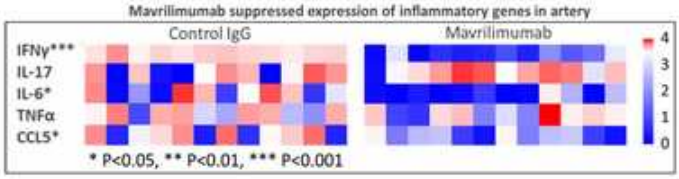


ACR Nov 2019: Mavrilimumab reduced arterial inflammation in an *in vivo* model of vasculitis compared to control[2]





Mavrilimumab suppressed expression of inflammatory genes in artery



1) Poster presentation at European Congress of Rheumatology 2019 (EULAR): *GM-CSF Pathway Signature Identified in Temporal Artery Biopsies of Patients With Giant Cell Arteritis*. Maria C. Cid, Rohan Ganthi, Marc Cádiera-Bellalta, Rekaris Terrades-García, Sujatha Muralidharan, John F. Paolini. 2) Presentation at 2019 American College of Rheumatology (ACR): *GM-CSF is a Pro-Inflammatory Cytokine in Experimental Vasculitis of Medium and Large Arteries*. Ryu Wakahata, Hui Zhang, Toshihisa Maeda, Mitsuhiro Akiyama, Rohan Ganthi, John F. Paolini, Gerald J. Berry, Cornelia M. Weyand

Clinical Development Plan for Mavrilimumab

Phase 2
Giant Cell Arteritis

- **Target enrollment achieved**
- 26-week, double-blind, randomized, placebo-controlled clinical trial of mavrilimumab with a corticosteroid taper in subjects with new-onset or refractory GCA
- Primary efficacy endpoint involves measuring GCA flares during 26-week treatment period
- Continuing to enroll for limited time to help facilitate the number of primary efficacy endpoint events

Top-line data expected 2H 2020

Phase 2
Relapsed/Refractory Large B-Cell Lymphoma

- **Clinical collaboration with Kite, a Gilead Company**
- Study of mavrilimumab with Yescarta® (axicabtagene ciloleucel) in patients with relapsed or refractory large B-cell lymphoma
- Objective: Determine whether combination therapy can help control chimeric antigen receptor T (CAR T) cell mediated inflammation without disrupting anti-tumor efficacy

Timeline TBD

KPL-716 – Phase 2

Rilonacept | Mavrilimumab | **KPL-716** | KPL-404

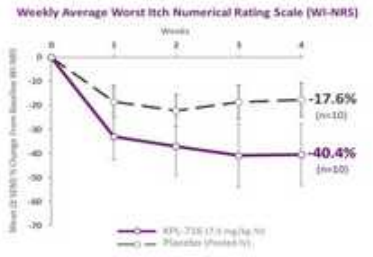
First Indication	**Prurigo Nodularis:** Chronic inflammatory skin disease with pruritic lesions
Mechanism of Action[1]	Monoclonal antibody inhibitor targeting OSMRβ
Scientific Rationale[2]	OSMRβ is a key receptor subunit shared by IL-31 and OSM; cytokines implicated in prurigo nodularis
Prevalence[3]	~300k prevalent in U.S.
Competition[4]	No FDA-approved therapies for prurigo nodularis
Status	Enrolling Phase 2a clinical trial in prurigo nodularis and exploratory Phase 2 study in diseases characterized by chronic pruritus
Rights	Worldwide

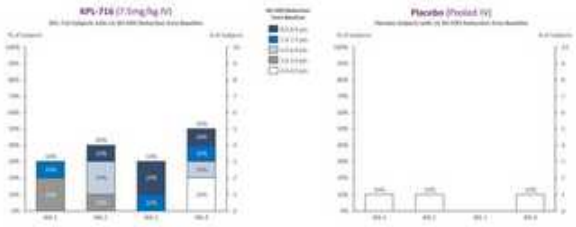
1) Trinity Qualitative Interviews; 2) Dillon SR, Sprecher C, Hammond A, Bilsborough J, Rosenfeld-Franklin M, Presnell SR, et al. Interleukin 31, a cytokine produced by activated T cells, induces dermatitis in mice. Nat Immunol. 2004; 5(7):752-60; Weigelt N, et al. J Cutan Pathol. 2010;37:578-86. 3) Trinity Consulting - HCUP/Medicare Data 2012/2013; Quantitative Survey (n=100 dermatologists); Dantas, 2015, "Prevalence of dermatoses in dermatologic evaluation requests from patients admitted to a tertiary hospital for 10 years"; Mortz et al., British Journal of Dermatology, 2001; 4) Journal of the American Academy of Dermatology - Analysis of Real-World Treatment Patterns in Patients with Prurigo Nodularis: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6721639/

Preclinical, Clinical and External Data Support Further Development of KPL-716

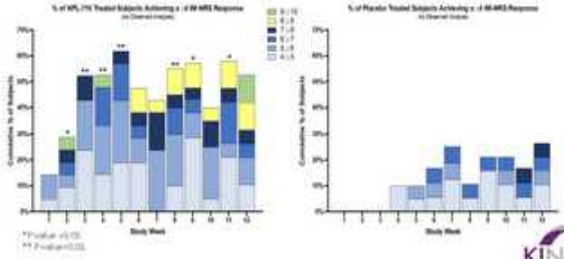
Phase 1b data showed rapid and sustained anti-pruritic effect

Single-Dose Phase 1b[1]





Repeated-Single-Dose Phase 1b[2]



1) Oral presentation at the 27th European Academy of Dermatology and Venereology (EADV) Congress: *First-in-Human Study of KPL-716, Anti-Oncostatin M Receptor Beta Monoclonal Antibody, in Healthy Volunteers and Subjects with Atopic Dermatitis*

2) Interim data results 8/12/19 – available through Investors & Media section of Kiniksa's website at www.kiniksa.com

Clinical Development Plan for KPL-716

Phase 2
Prurigo Nodularis

- Enrolling 8-week, double-blind, randomized, placebo-controlled clinical trial of KPL-716 in subjects with prurigo nodularis
- Primary efficacy endpoint is percent change from baseline in weekly average Worst-Itch Numeric Rating Scale (WI-NRS) at 8 weeks

Top-line data expected 1H 2020

Phase 2
Multiple Chronic Pruritic Diseases

- Enrolling 8-week, double-blind, randomized, placebo-controlled clinical trial of KPL-716 in subjects with chronic idiopathic urticaria, chronic idiopathic pruritus, lichen planus, lichen simplex chronicus and plaque psoriasis
- Primary efficacy endpoint is percent change from baseline in weekly average WI-NRS at 8 weeks

Interim data from select number of cohorts expected 1H 2020

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KPL-404 – Phase 1

Rilonacept | Mavrilimumab | KPL-716 | **KPL-404**

Autoimmune Diseases[1]	External proof-of-concept previously established in broad range of autoimmune diseases: Sjogren's disease, systemic lupus, rheumatoid arthritis, solid organ transplant and Graves' disease[1]
Mechanism of Action[2]	Monoclonal antibody inhibitor of CD40-CD40L interaction
Scientific Rationale[3,4]	Attractive target for blocking T-cell mediated, B-cell driven
Status	Enrolling and dosing subjects in first-in-human clinical study with antigen challenge TDAR[5]
Rights	Worldwide

1) National Center for Biotechnology Information - Targeting the CD40-CD154 Signaling Pathway for Treatment of Autoimmune Arthritis: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6721639/; 2) Poster presentation at the Keystone Symposia: Antibodies as Drugs: New Horizons in the Therapeutic Use of Engineered Antibodies: *KPL-404, a CD40 antagonist, blocked antigen-specific antibody responses in an in vivo NHP model and demonstrated strong PK/PD correlation*; 3) Elgueta, et al. Immunol Rev 2009, 229 (1), 152-172; 4) Peters, et al. Semin Immunol 2009, 21 (5) 293-300; 5) TDAR, T-cell Dependent Antibody Response

Multiple Key Clinical Data Readouts Expected in 2020

KPL-716 – Phase 2 (monoclonal antibody inhibitor targeting OSMRβ)	**Prurigo Nodularis** (Top-line Phase 2 Data)	**1H 2020**
KPL-716 – Phase 2 (monoclonal antibody inhibitor targeting OSMRβ)	**Diseases Characterized by Chronic Pruritus** (Interim Phase 2 Data in Limited Cohorts)	**1H 2020**
Rilonacept – Phase 3 (IL-1α and IL-1β cytokine trap) RHAPSODY	**Recurrent Pericarditis** (Pivotal Top-line Phase 3 Data)	**2H 2020**
Mavrilimumab – Phase 2 (monoclonal antibody inhibitor targeting GM-CSFRα)	**Giant Cell Arteritis** (Top-line Phase 2 Data)	**2H 2020**
KPL-404 – Phase 1 (monoclonal antibody inhibitor of CD40-CD40L interaction)	**Healthy Subjects** (Top-line Phase 1 Data)	**2H 2020**

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Rare Diseases with **Unmet Medical Need**

Potentially **Attractive Commercial Opportunities**

YE 2019 **~$233M** Cash Balance **Extends** into **2H 2021**[1]

Multiple **Clinical Data Readouts** Expected in **2020**

Autoimmune and **Autoinflammatory** Pipeline

1) Cash, Cash Equivalents and short-term investments as of 12/31/19 (unaudited)


KINIKSA
Every Second Counts!™