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): **April 2, 2024**

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 Stock Market LLC (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 Disclosure.

On April 2, 2024, Kiniksa Pharmaceuticals, Ltd. (the "Company") issued a press release (the "Press Release") announcing, among other things, (i) its plans to initiate a Phase 2b clinical trial of abiprubart in Sjogren's Disease, including clinical trial design for such trial, (ii) topline data from Cohort 4 of its Phase 2 clinical trial of abiprubart in rheumatoid arthritis and (iii) that it expected to remain cash flow positive on an annual basis within its current operating plan. In connection with such announcement, the Company posted an investor presentation (the "Investor Presentation") containing information related to the above, among other things, to its website at investors.kiniksa.com.

A copy of the Press Release and the Investor Presentation are furnished with this Current Report on Form 8-K as Exhibit 99.1 and Exhibit 99.2, respectively.

Item 9.01. Financial Statements and Exhibits.

(d) Exhibits

Exhibit No.	Description
99.1	Press Release issued by Kiniksa Pharmaceuticals, Ltd., dated April 2, 2024
99.2	Kiniksa Pharmaceuticals, Ltd. Investor Presentation
104	Cover Page Interactive Data File (embedded within the inline XBRL document)

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: April 2, 2024

By: /s/ Madelyn Zeylikman
Madelyn Zeylikman
Senior Vice President, General Counsel and Secretary

Kiniksa Pharmaceuticals Announces Development Indication for Abiprubart

– Abiprubart Phase 2b trial in Sjogren's Disease planned to initiate in 2H 2024 –
– Abiprubart Phase 2 Cohort 4 rheumatoid arthritis data further validate biological activity –
– Abiprubart development in Sjogren's Disease fully funded through Phase 3 –
– Company expects to remain cash flow positive on an annual basis within current operating plan –

HAMILTON, BERMUDA – April 2, 2024 – Kiniksa Pharmaceuticals, Ltd. (Nasdaq: KNSA) (Kiniksa), a commercial-stage biopharmaceutical company with a pipeline of immune-modulating assets designed to target a spectrum of cardiovascular and autoimmune diseases, today announced plans to initiate a Phase 2b trial with abiprubart in Sjogren's Disease. Additionally, the company announced data from Cohort 4 of the Phase 2 clinical trial of abiprubart in rheumatoid arthritis. Abiprubart is an investigational humanized anti-CD40 monoclonal antibody designed to inhibit CD40-CD154 (CD40 ligand) interaction.

"We believe abiprubart has the potential to provide meaningful benefit to patients suffering from Sjogren's Disease, a debilitating disorder with no current FDA-approved therapies. Based on the clear biological activity demonstrated by abiprubart, potential for convenient subcutaneous administration, and external proof-of-concept of inhibition of the CD40-CD154 interaction, we plan to initiate a Phase 2b trial of abiprubart in Sjogren's Disease in the second half of 2024," said Sanj K. Patel, Chairman and Chief Executive Officer of Kiniksa. "Supported by our robust ARCALYST revenue growth and current cash position, we continue to execute a strategic and disciplined capital allocation approach in areas we believe provide the best opportunity to drive long-term value. Based on our current operating plan, which includes advancement of abiprubart through Phase 3 development in Sjogren's Disease, we expect to remain cash flow positive on an annual basis."

Phase 2b Clinical Trial of Abiprubart in Sjogren's Disease

Kiniksa is planning to initiate a randomized, double-blind, placebo-controlled Phase 2b trial designed to evaluate the treatment response of chronic subcutaneous (SC) administration of abiprubart in patients with Sjogren's Disease.

The placebo-controlled portion of the trial will randomize approximately 201 patients in a 1:1:1 ratio to receive abiprubart 400 mg SC biweekly, 400 mg SC monthly, or placebo over a period of 24 weeks. The primary endpoint will be change from baseline in EULAR Sjogren's Syndrome Disease Activity Index (ESSDAI) versus placebo at Week 24. Subsequently, patients will enter a long-term extension in which all patients will receive active treatment for an additional 24 weeks. The trial is expected to initiate in the second half of 2024.

Abiprubart Phase 2 Rheumatoid Arthritis Data

The Phase 2 rheumatoid arthritis trial uses a randomized, double-blind, placebo-controlled design to evaluate pharmacokinetics (PK), safety, and efficacy of chronic SC administration of abiprubart and to provide optionality to evaluate abiprubart across a range of autoimmune diseases. The trial enrolled patients with active rheumatoid arthritis who had an inadequate response or were intolerant to a Janus kinase inhibitor (JAKi) or at least one biologic disease-modifying anti-rheumatic drug (bDMARD).

Following previously reported topline data, Kiniksa today announced final data from the first three cohorts of the clinical trial:

- In Cohorts 1 and 2, multiple doses of abiprubart were well-tolerated and enabled the proof-of-concept portion of the study.
- In the Cohort 3 abiprubart 5 mg/kg SC weekly dose group (n=27), the Least Squares (LS) mean change [95% confidence interval (CI)] from baseline in Disease Activity Score of 28 Joints Using C-reactive Protein (DAS28-CRP) at Week 12 was -2.17 [-2.60, -1.74] points, compared to -1.61 [-2.04, -1.17] points in placebo recipients (n=26), (LS Mean Difference = -0.57, p=0.0470).
- In the Cohort 3 abiprubart 5 mg/kg SC biweekly dose group (n=25), the LS mean change [95% CI] from baseline in DAS28-CRP at Week 12 was -1.96 [-2.40, -1.52] points, compared to -1.61 [-2.04, -1.17] points in placebo recipients (n=26), (LS Mean Difference = -0.36, p=0.2124).
- There was a statistically significant reduction of over 40% in Rheumatoid Factor, a clinical marker of disease activity and an autoantibody pharmacodynamic marker of CD40 target engagement, in both Cohort 3 abiprubart dose groups (p<0.0001).
- Abiprubart was well-tolerated, with no dose-related adverse experiences observed.

Today, Kiniksa announced topline data from the fourth cohort of the clinical trial:

- In the Cohort 4 abiprubart 400 mg SC monthly dose group (n=31), the LS mean change [95% CI] from baseline in DAS28-CRP at Week 12 was -1.87 [-2.54, -1.21] points, compared to -1.30 [-1.98, -0.62] points in placebo recipients (n=20), (LS Mean Difference = -0.58, p=0.109).
- There was a statistically significant reduction of approximately 40% in Rheumatoid Factor in the abiprubart group (p=0.0003).
- As in the first three cohorts, abiprubart was well-tolerated, and no dose-related adverse experiences were observed.

Additionally, Kiniksa today announced a post-hoc analysis of data pooled from the Cohort 3 and Cohort 4 abiprubart and placebo groups:

- In the pooled abiprubart group (n=83), the LS mean change [95% CI] from baseline in DAS28-CRP at Week 12 was -2.04 [-2.34, -1.74] points, compared to -1.52 [-1.88, -1.16] points in placebo recipients (n=46), (LS Mean Difference = -0.52, nominal p=0.010).

“These Phase 2 data demonstrate that abiprubart is a potentially efficacious and well-tolerated therapeutic approach for multiple autoimmune diseases, including Sjogren’s Disease,” said John F. Paolini, MD, PhD, Chief Medical Officer of Kiniksa. “The comparable magnitude of reduction in Rheumatoid Factor observed across weekly, biweekly, and monthly dosing and supportive post-hoc analysis of pooled efficacy data reinforce our confidence that abiprubart is highly active. We look forward to initiating the Phase 2b trial in Sjogren’s Disease in the second half of this year.”

Financial Guidance

Kiniksa expects to remain cash flow positive on an annual basis within its current operating plan, which includes advancement of abiprubart through Phase 3 development in Sjogren’s Disease.

About Kiniksa

Kiniksa is a commercial-stage biopharmaceutical company focused on discovering, acquiring, developing, and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. Kiniksa’s immune-modulating assets, ARCALYST, abiprubart, and mavrilimumab, are based on strong biologic rationale or validated mechanisms, target a spectrum of underserved cardiovascular and autoimmune conditions, and offer the potential for differentiation. For more information, please visit www.kiniksa.com.

About Abiprubart

Abiprubart is an investigational humanized monoclonal antibody that binds to CD40 and is designed to inhibit the CD40-CD154 (CD40 ligand) interaction, a key T-cell co-stimulatory signal critical for B-cell maturation and immunoglobulin class switching and Type 1 immune responses. Kiniksa believes disrupting the CD40-CD154 co-stimulatory interaction is an attractive approach to addressing multiple autoimmune disease pathologies.

About the Phase 2 Clinical Trial of Abiprubart in Rheumatoid Arthritis

The ongoing Phase 2 rheumatoid arthritis trial uses a randomized, double-blind, placebo-controlled design to evaluate PK, safety, and efficacy of chronic SC administration of abiprubart and to provide optionality to evaluate abiprubart across a range of autoimmune diseases. This trial enrolled patients with active rheumatoid arthritis who had an inadequate response or were intolerant to a JAKi or at least one bDMARD.

The multiple ascending-dose PK lead-in portion randomized 8 patients each in a 3:1 ratio to receive abiprubart 2 mg/kg or placebo (Cohort 1) or 5 mg/kg or placebo (Cohort 2), administered subcutaneously biweekly over a period of 12 weeks. The primary objective of this part of the trial was to evaluate PK, safety, and tolerability over 12 weeks. The secondary efficacy endpoint was change from baseline in DAS28-CRP versus placebo.

The first part of the proof-of-concept portion of the trial (Cohort 3) randomized 78 patients in a 1:1:1 ratio to receive abiprubart 5 mg/kg SC weekly, abiprubart 5 mg/kg SC biweekly, or placebo over a period of 12 weeks. The final part of the proof-of-concept portion of the trial (Cohort 4) randomized 51 patients in a 3:2 ratio to receive a fixed 600 mg loading dose on Day 1 followed by 400 mg SC every four weeks or placebo over a period of 12 weeks. The primary efficacy endpoint of the proof-of-concept portion of the trial was change from baseline in DAS28-CRP versus placebo.

Forward-Looking Statements

This press release contains forward-looking statements. 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 plan to initiate a Phase 2b clinical trial of abiprubart in Sjogren's Disease in the second half of 2024; our expectation that abiprubart development in Sjogren's Disease will remain fully funded through Phase 3 development; our expectation to advance abiprubart through Phase 3 development in Sjogren's Disease; our expectation to remain cash flow positive on an annual basis within our current operating plan; our expectation that our capital allocation will provide the best opportunity to drive long-term value; our expectations around the possibility of achieving meaningful effect and differentiation via subcutaneous administration with abiprubart; future clinical trial design, including the design of our planned Phase 2b trial of abiprubart in Sjogren's Disease; our beliefs about the mechanisms of our product candidates and potential impact of their approach, including that using abiprubart to disrupt the CD40-CD154 co-stimulatory interaction is an attractive approach to address multiple autoimmune disease pathologies; and our belief that all of our product candidates offer the potential for differentiation.

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: delays or difficulty in enrollment of patients in, and activation or continuation of sites for, our clinical trials; delays or difficulty in completing our clinical trials as originally designed; potential for changes between final data and any preliminary, interim, top-line or other data from clinical trials; our inability to replicate results from our earlier clinical trials or studies; impact of additional data from us or other companies, including the potential for our data to produce negative, inconclusive or commercially uncompetitive results; potential undesirable side effects caused by our products and product candidates; our inability to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities; potential for applicable regulatory authorities to not accept our filings, delay or deny approval of any of our product candidates or require additional data or trials to support approval; our reliance on third parties as the sole source of supply of the drug substance and drug product used in our product candidates; raw material, important ancillary product and drug substance and/or drug product shortages; our reliance on third parties to conduct research, clinical trials, and/or certain regulatory activities for our product candidates; complications in coordinating requirements, regulations and guidelines of regulatory authorities across jurisdictions for our clinical trials; changes in our operating plan, business development strategy or funding requirements; and existing or new competition.

These and other important factors discussed in our filings with the U.S. Securities and Exchange Commission, including under the caption "Risk Factors" contained therein, 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. Except as required by law, we disclaim any intention or obligation to update or revise any forward-looking statements. 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. All other trademarks are the property of their respective owners.

Every Second Counts! ®

Kiniksa Investor and Media Contact
Rachel Frank
(339) 970-9437
rfrank@kiniksa.com

KINIKSA

Corporate Presentation

APRIL 2024

Forward Looking Statements

This presentation (together with any other statements or information that we may make in connection herewith) contains forward-looking statements with respect to Kiniksa Pharmaceuticals, Ltd. (and 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," "sho "expect," "plan," "anticipate," "could," "intend," "goal," "design," "target," "project," "contemplate," "believe," "estimate," "predict," "potential," "strategy," or "continue" or the negative of these terms or othe 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 value drivers; potential indications; potential market opportunities and competitive positi ongoing, 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 commercial activities; expected run rate for our cash, cash equivalents and short-term investments; expected funding of our operating plan; financial guidance; 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 earl trials and studies; our reliance on third parties for manufacturing and conducting clinical trials, research and other studies; risks arising from our technology transfer of ARCALYST drug substance manufacturing; our ability to realize value from our licensing and collaboration arrangements; our ability to source sufficient drug product, as needed, to meet our clinical and commercial requirements; inability to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities; potential for applicable regulatory authorities to not accept our filings or to delay or deny approval of a our product candidates or to require additional data or trials to support any such approval or authorization; delays, difficulty or inability to successfully execute on our commercial strategy for ARCALYS potential changes in our strategy, clinical trial priority, operating plan, business development strategy or funding requirements; raw materials, important ancillary product and drug substance and/or dru product shortages; substantial new or existing competition; risks arising from political and economic instability; and our ability to attract and retain qualified personnel.

These and the important factors discussed in our filings with the U.S. Securities and Exchange Commission, including under the caption "Risk Factors" contained therein could cause actual results to d materially from those indicated by the forward-looking statements made in this presentation. These forward-looking statements reflect various assumptions of Kiniksa's management that may or may n 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 otherwis 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, except as required by law.

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 re 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 simi 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

Portfolio of Immune-Modulating Assets

Program	Target	Preclinical	Phase 1	Phase 2	Phase 3	Commercial
CARDIOVASCULAR FRANCHISE						
ARCALYST® (rilonacept)[1,2,3] IL-1α & IL-1β	*Recurrent Pericarditis*					
Mavrilimumab[4] GM-CSFRα	*Evaluating Potential Partnership Opportunities*					
AUTOIMMUNE FRANCHISE						
Abiprubart CD40/CD154	*Sjogren's Disease*					

Program	Licensee	Exclusive Licensed Territory
OUT-LICENSING AGREEMENTS		
ARCALYST (rilonacept) IL-1α & IL-1β	*Huadong Medicine*	*Asia Pacific Region, Excluding Japan*
Mavrilimumab GM-CSFRα	*Huadong Medicine*	*Asia Pacific Region, Excluding Japan*
Vixarelimab OSMRβ	*Roche and Genentech*	*Worldwide*

1) Approved in the U.S.; ARCALYST is also approved in the U.S. for cryopyrin-associated periodic syndromes (CAPS) and deficiency of the interleukin-1 receptor antagonist (DIRA); 2) The FDA granted Breakthrough Therapy designation to ARCALYST for recurrent pericarditis in 2019; the FDA granted Orphan Drug exclusivity to ARCALYST in March 2021 for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and pediatric patients 12 years and older. The European Commission granted Orphan D designation to ARCALYST for the treatment of idiopathic pericarditis in 2021; 3) Kiniksa has worldwide rights, excluding the Middle East and North Africa; Kiniksa granted Huadong Medicine exclusive rights in the Asia Pacific Region, excluding Japan; 4) Phase 2 clini trials of mavrilimumab in rheumatoid arthritis and giant cell arteritis achieved their primary and secondary endpoints with statistical significance; Kiniksa granted Huadong Medicine exclusive rights in the Asia Pacific Region, excluding Japan
IL-1α = interleukin-1α; IL-1β = interleukin-1β; GM-CSFRα = granulocyte macrophage colony stimulating factor receptor alpha; OSMRβ = oncostatin M receptor beta

KINIKSA

ARCALYST®

Arcalyst (rilonacept) For Injectio

IL-1α AND IL-1β CYTOKINE TRAP

DISEASE AREA: Recurrent pericarditis[1]; painful and debilitating auto-inflammatory cardiovascular disease

COMPETITION[2]: First and only FDA-approved therapy for recurrent pericarditis

REGULATORY: U.S. Orphan Drug exclusivity for treatment of and reduction in risk of recurrence of recurrent pericarditis; European Commission Orphan Drug designation in idiopathic pericarditis

STATUS: FDA-Approved

ECONOMICS: 50/50 split on profit and third-party proceeds

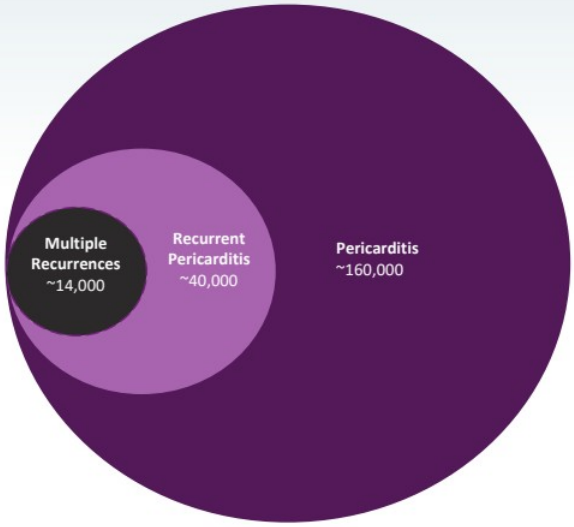
RIGHTS: Kiniksa has worldwide rights[3] (excluding MENA) for all indications outside those in oncology and local administration to the eye or ea

KINIKSA

1) ARCALYST is also approved and marketed for Cryopyrin-Associated Periodic Syndromes (CAPS) and maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA) in the United States;
2) 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; 3) Kiniksa granted Huadong Medicine exclusive rights in the Asia Pacific Region, excluding Japan;
IL-1α = interleukin-1α ; IL-1β = interleukin-1β; MENA = Middle East North Africa

Pericarditis Epidemiology

Of the 14,000 target population with multiple recurrences there is a high turnover of ~50% of patients each year, meaning ongoing opportunities to ensure diagnosis and targeted treatment



All figures annual period prevalence

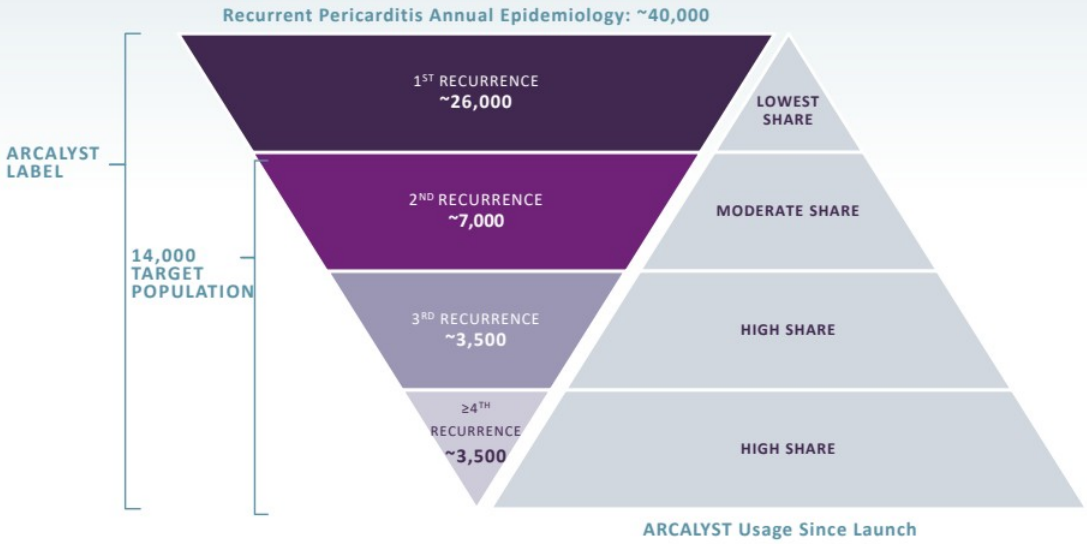
Approximately 14,000 recurrent pericarditis patients in the U.S. suffer fro
persistent underlying disease, with multiple recurrences and inadequate response to conventional therapy[1]

- **~160,000:** Epidemiological analysis using large national surveillance databases to calculate the pooled annualized prevalence of pericarditis ***(Basis for Orphan Drug Designation approval)[2]***
- **~40,000:** Up to 30% experience at least one recurrence; some recur over multiple years[6,7]
- **~14,000:** Nearly 50% annual turnover with ~7,000 patients entering into the pool each year[8]

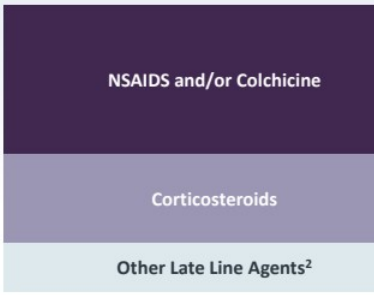
KINIKSA

1) Cremer et al. American Journal of Cardiology. 2016:2311-2328; 2) DOF, Kiniksa Pharmaceuticals, Ltd.; 3) Brucato A, Maestroni S, Cumetti D, et al. Autoimmun Rev. 2008; 8:44-47; 4) Lange R, Hills L. N Engl J Med. 2004; 351: 2195-2202; 5) Imazio M, Cecchi E, Demichelis B, et al. Circulation. 2007; 115: 2739-2744; 6) Imazio et al. Circulation. 2005;112:2012-2016; 7) Adler et al. Circulation. 1998;97:2183-2185; 8) Klein A, Cremer P, Kontzias A, et al. US database study of clinical burden and unmet need in recurrent pericarditis. J Am Heart Assoc. 2021; 10:e018950. doi:10.1161/JAHA. 120.018950

Treated Patients Since Launch Are Closely Associated to the 14,000 Targ
Population, While Prescribers Can Utilize ARCALYST Earlier in the Diseas



ARCALYST PATIENTS BY PRIOR PRODU



ARCALYST PATIENTS BY FLARE STATUS @ INITIATION[1]

Klein A, Cremer P, Kontzias A, Furqan M, Tubman R, Roy M, Magestro M. Annals of Epidemiology. 2019;36:71; 2) Lin D, Majeski C, DerSarkissian M, Magestro M, Cavanaugh C, Laliberte F, Lejune D, Mahendran M, Duh M, Klein A, Cremer P, Kontzias A, Furqan M, Tubman R, Roy M, Mage. (Nov, 2019). Real-World Clinical Characteristics and Recurrence Burden of Patients Diagnosed with Recurrent Pericarditis in the United States. Poster session presented at the American Heart Association, Philadelphia, PA.; 3) ClearView Forecasting Analysis 2019 Q1

Source: 1) Kiniksa Pharmaceuticals data on file 2024. 2) Other late line agents include anakinra, azathioprine, methotrexate

ARCALYST Commercial Growth in 2023: By the Numbers

~78% annual growth vs Q4 2022

~90% annual growth vs full-year 2022

~80% annual growth vs end of Q4 2022

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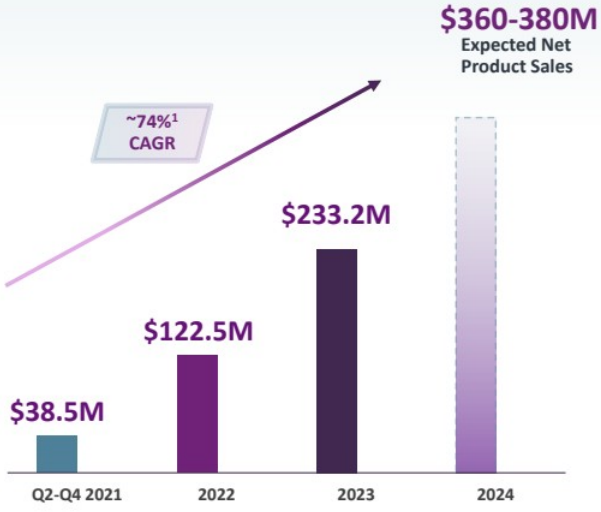
Long-Term Growth Potential Through Commercialization Maturation

Accelerating Revenue with Long-Term Growth Horizon



Total Prescribers	**>1,700**
Repeat Prescribers (% of Total)	**~24%**
Payer Approval (% of Completed Cases)	**>90%**
Average Total Duration of Therapy	**~23 months**
Patient Compliance	**>85%**

ARCALYST Collaboration Operating Profit

2021	2022	2023
($8.0M)[2]	$36.2M	$113.0M

KINIKSA

[1]Implied 2022-2024 Compound Annual Growth Rate assuming midpoint of projected 2024 net product sales
[2]The ARCALYST collaboration achieved profitability in the fourth quarter of 2021, following three quarters of commercial availability for recurrent pericarditis

Key Executional Priorities to Drive Greater Patient and Physician Adopt

Identify appropriate patients and drive a proactive mindset with physicians and patients

Close the ARCALYST **knowledge gap** with physicians

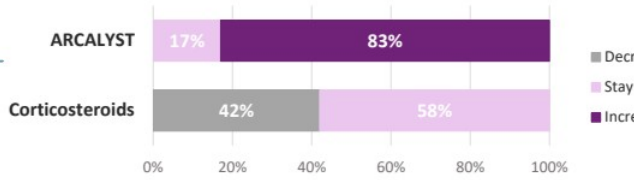
Advance the **treatment paradigm**

Educate on **duration of disease** and treatment

Externally: Thought leaders are introducing treatment paradigm recurrent pericarditis that recommend IL-1 antagonists, such as ARCALYST, be used ahead of corticosteroids[1]

Our Aim: Continue to drive the evolution of this treatment para

Intended Future Use Among Target Healthcare Providers[2]



- Of target physicians who have knowledge of ARCALYST, they overwheln expect to **increase their prescribing of ARCALYST in next 6 months**
- The biggest barriers for physicians to prescribing ARCALYST are **limited knowledge about the product and/or perception of the payer approva process**

1) Dong, Klein, Wang. Paradigm Shift in Diagnosis and Targeted Therapy in Recurrent Pericarditis. Springer Nature. 2023.; Klein, Cremer, Kafil. Recurrent Pericarditis A Promising Future for IL-1 Blockers in Autoinflammatory Phenotypes. Journal of the American College of Cardiology, Editorial Comment. 2023.; Thomas, Bonaventura, Vecchié, et al. Interleukin-1 blockers for the treatment of recurrent pericarditis: pathophysiology, patient reported outcomes and perspectives. Journal of Cardiovascular Pharmacology. 2023.; Imazio, Mardigyan, Andreis, et al. New developments in the management of recurrent pericarditis. Canadi Journal of Cardiology. 2023.; Kumar, Khubber, Reyaldeen, et al. Advances in Imaging and Targeted Therapies for Recurrent Pericarditis. JAMA Cardiology Review. 2022.; Sushil, Cremer, Raisinghani.
2) HCP Market Research, Q3 2023; Kiniksa Data on File.

Average Total Duration of ARCALYST Therapy: ~23 Months[1]

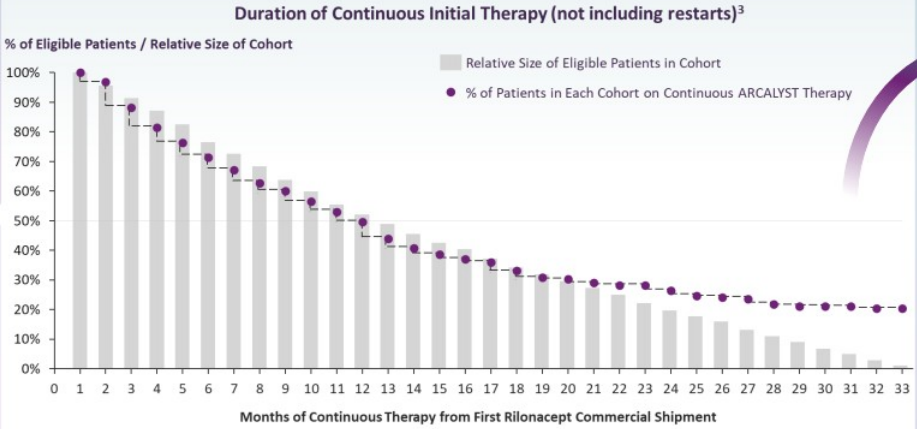
Advancing the treatment paradigm to treat continuously throughout disease duration (median 3 years[2])

Average *Initial* Duration of Therapy

~15 Months[1]

Median *Initial* Duration of Therapy

~12 Months[1]



~45%

Of Patients Restarted Therapy Following Initial Discontinuation

(Within ~8 weeks)

~23 Months Average ***Total*** Duration of Therapy After Accounting for Patient Restarts

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1) As of Q4 2023; 2) Lin D, Laliberté F, Majeski C, et al. Disease and economic burden associated with recurrent pericarditis in a privately insured United States population. Adv Ther. 2021;38(10):5127-5143. doi:10.1007/s12325-021-01868-7; 3) Initial continuous therapy is determined to have ended if greater than 28 days elapses beyond the exhaustion date of a patient's most recent days supplied without an observed refill of ARCALYST

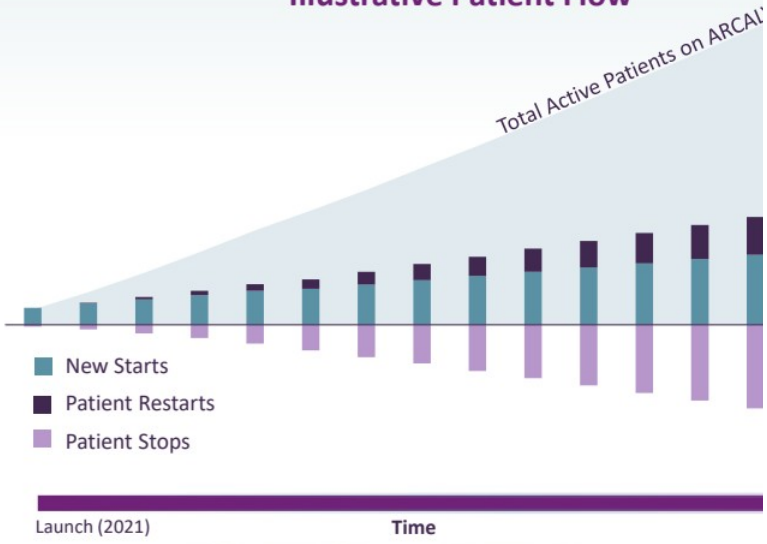
Growth in Total Patients on ARCALYST Therapy

Acceleration in new-to-brand and restart patients offset higher patient stops over time

ARCALYST Patient Flow

New to Brand Patients	Strong sequential growth: >1,700 unique prescribers; ~24% of which are repeat prescribers
−	
Patient Stops	Increases over time as base of active ARCALYST patients grows with Initial Starts and Restarts
+	
Patient Restarts	Increases over time as patient stops increase; currently ~45% after ~8 weeks
=	
Active Patients	Increases over time driven by new-to-brand and restart growth; as of Q4 2023, ~9% of 14K multiple recurrence target patient population

Illustrative Patient Flow



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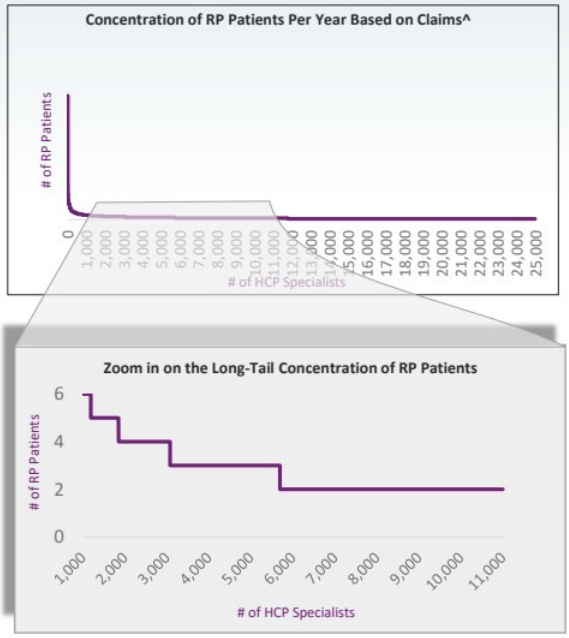
Evolving ARCALYST Field Strategy

Targeting an increased number of top and mid-tier physicians

The recurrent pericarditis population is widely dispersed



*Including targets, prospects, and opportunistic calls to non-targets

^Internal analysis based on Komodo Claims Data; includes patients with at least 1 recurrence

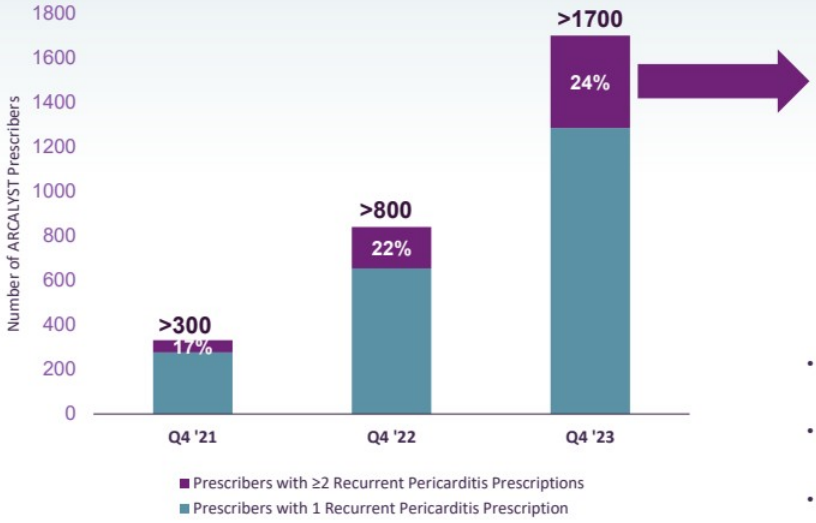
Data driven expansion to field sales team

Q4 2022	Q4 2023
Prior expansion to create greater reach & frequency	New expansion to provide greater frequency on top tier physicians and improved coverage to the mid tier
~50 Specialty Cardiology Reps	**~85** Specialty Cardiology Reps
Reaching:	Reaching:
~6,000 top and mid tier prescribers	**~11,000** top and mid tier prescribers
~70%* of RP patients nationally	**~85%*** of RP patients nationally

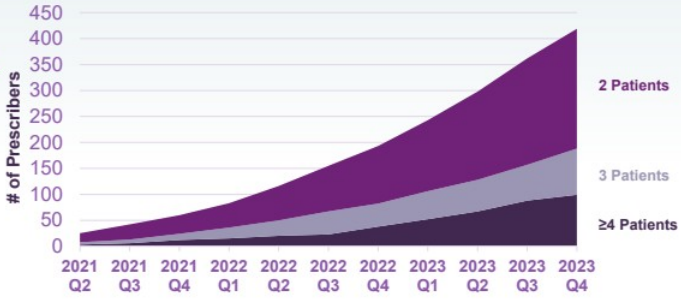
- In any given year, the 14,000 multiple recurrent pericarditis patients may present to of the >20,000 cardiologists and >5,000 rheumatologists in US
- With our field expansion, we expect to accelerate coverage and frequency among t tier as well as the long tail of physicians who may identify recurrent pericarditis pat
  - Data-driven decisions ensured continued growth in collaboration profitability following the prior expansion
  - With the new expansion, we have the opportunity to meaningfully increase frequency on prior field targets and to reach new health care providers that ha prior field interactions

Opportunity for Continued ARCALYST Growth Remains High

Total and Repeat Prescribers of ARCALYST for Recurrent Pericarditis Patients



The Growing Repeat Prescriber Base is Delivering ~40% of All New Patient Prescriptions



- Strong sequential growth in **both new and repeat prescribers**, underscoring the dispersed patent population
- Both physicians and patients are gaining **positive experiences with ARCALYST** as t first and only approved therapy for recurrent pericarditis
- Cardiologist market research shows a steady **increase in their level of comfort wit prescribing biologics**
- **Greater than 40% of all new prescriptions in 2023 came from repeat prescribers**

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Pricing, Access and Distribution Considerations

Pricing

- ARCALYST list price of $22,603 per month

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

- Helping to ensure **patient affordability** and access to treatment is one of our core principles and to this end, we offer a suite of programs to support affordability to eligible patients who are prescribed ARCALYST; eligible patients are able to get ARCALYST for a copay of as low as $0

Access

- Kiniksa's goal is to maintain rapid and broad access to ARCALYST for patients with Recurrent Pericarditis, CAPS, and DIRA
- Payer mix for ARCALYST is largely **commercial (~70%)**
- Payer engagement has increased awareness of recurrent pericarditis and the differentiated value of ARCALYST
- The **Kiniksa OneConnect™** program is a personalized treatment support program for patients prescribed ARCALYST

Distribution

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

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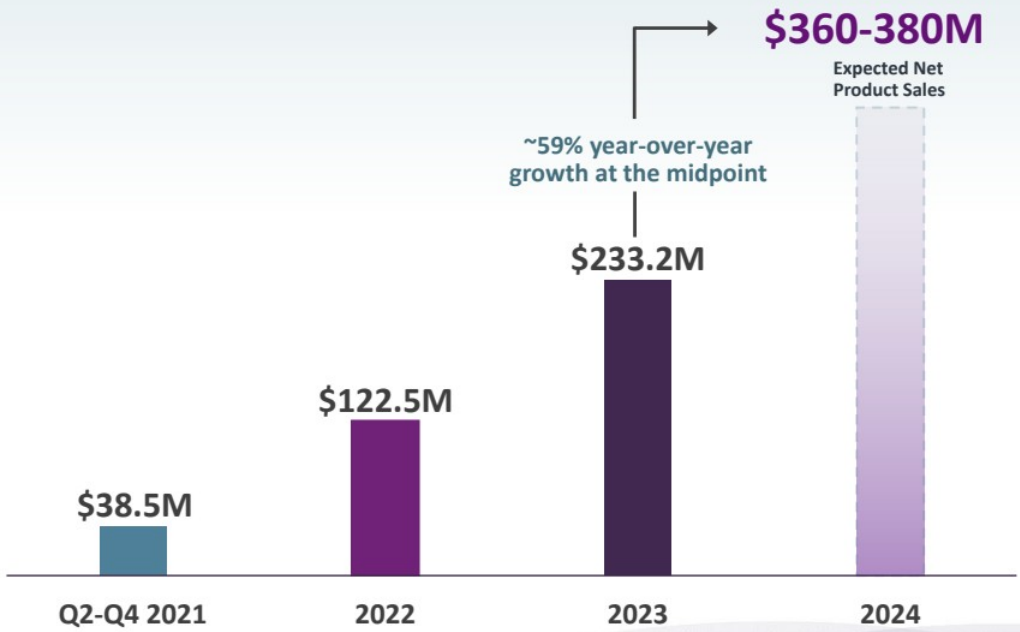
CAPS = Cryopyrin-Associated Periodic Syndromes ; DIRA = Deficiency of IL-1 Receptor Antagonist

2024 ARCALYST Net Product Sales Guidance

Well-positioned to expand the breadth and depth of ARCALYST in recurrent pericarditis



Summary of ARCALYST Profit Share Arrangement with Regeneron[1]

- Kiniksa is responsible for sales and distribution of ARCALYST in all approved indications in the United States.
- Kiniksa's license to ARCALYST includes worldwide rights*, excluding MENA, for all applications other than those in oncology and local administration to the eye or ear.
- Kiniksa covers 100% of development expenses related to approval of additional indications.
- Kiniksa evenly splits profits on ARCALYST sales and licensing proceeds with Regeneron

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1) Subject to description contained in definitive agreement; 2) Global net sales for CAPS, DIRA and recurrent pericarditis recognized as revenue on Kiniksa's income statement; 3) Profit Split-Eligible Cost of Goods Sold = to
cost of goods sold - amortization of Regeneron milestone payment
*Kiniksa exclusively licensed rights for the development and commercialization of ARCALYST in APAC (ex-Japan) to Huadong Medicine
CAPS = Cryopyrin-Associated Periodic Syndromes; DIRA = Deficiency of the Interleukin-1 Receptor Antagonist; MENA =Middle East and North Africa; APAC = Asia Pacific Region

ABIPRUBART

ANTI-CD40 MONOCLONAL ANTIBODY INHIBITOR OF THE CD40-CD154 CO-STIMULATORY INTERACTION

DISEASE AREA: Sjogren's Disease, an immune system disease characterized by autoimmune-driven destruction of the salivary and tear glands as well as arthritis, kidney, and lung dysfunction

SCIENTIFIC RATIONALE[1,2]: Attractive target for blocking T-cell dependent, B-cell–mediated autoimmunity; external proof-of-concept previously establishe broad range of autoimmune diseases: Sjogren's Disease, systemic lupus, solid organ transplant and Graves' Disease[3,4]

STATUS: Plan to initiate a Phase 2b trial in Sjogren's Disease in the second half of 2024

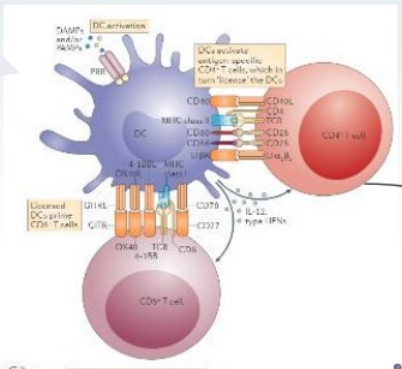
ECONOMICS: Negligible clinical and regulatory milestones and royalty on annual net sales

RIGHTS: Worldwide

Sources: 1) Elgueta, et al. Immunol Rev 2009, 229 (1), 152-172; 2) Peters, et al. Semin Immunol 2009, 21 (5) 293-300 3) Muralidharan et al. Preclinical immunopharmacologic assessment of KPL-404, a novel, humanized, nc depleting antagonistic anti-CD40 monoclonal antibody. J Pharmacol Exp Ther. 2022, 381(1):12-21. 4) Samant M, Ziemniak J, Paolini JF. First-in-Human Phase 1 Randomized Trial with the Anti-CD40 Monoclonal Antibody KP 404: Safety, Tolerability, Receptor Occupancy, and Suppression of T-Cell-Dependent Antibody Response. J Pharmacol Exp Ther. 2023 Dec;387(3):306-314.

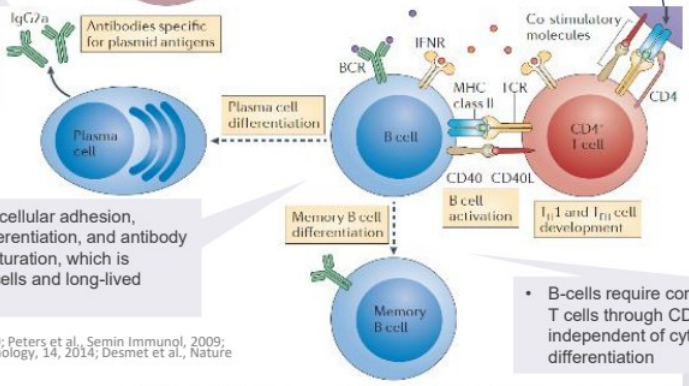
CD40/CD154 Interaction: Essential Immune Pathway for T-Cell Priming and T-Cell Depende B-Cell Responses

- CD40 is expressed on the surface of dendritic cells, B-cells, antigen-presenting cells and non-immune cell types
- Its ligand, CD40L (CD154), is expressed by activated T-cells, platelets, and other cell types



- CD40 ligation on DCs induces cell maturation by promoting ant presentation and enhancing their costimulatory activity
- Mature DCs stimulate activated T-cells to increase IL-2 product that facilitates T-helper cells (Th) and cytolytic T-Lymphocyte (C expansion
- CD40-stimulated DCs also secrete cytokines favoring Th1 cell differentiation and promoting Th cell migration to sites of inflammation
- CD40 ligation also provides a pro-inflammatory signal within the mononuclear phagocyte system

- Humoral immunity is dependent on a thriving B cell population and activation by Th cells; blockade of CD40-CD40L interaction has been shown to completely ablate primary and secondary TDAR response



- CD40 engagement triggers B-cell intercellular adhesion, sustained proliferation, expansion, differentiation, and antibody isotype switching leading to affinity maturation, which is essential for generation of memory B cells and long-lived plasma cells

- B-cells require contact-dependent stimulus from T cells through CD40-CD40L interaction independent of cytokines to trigger growth and differentiation

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Sources: Elgueta et al., Immunol Rev, 2009; Peters et al., Semin Immunol, 2009; Kambayashi et al., Nature Reviews: Immunology, 14, 2014; Desmet et al., Nature Reviews: Immunology, 12, 2012

Abiprubart Has Potential to Provide Meaningful Benefit to Patients with Sjogren's Disease

Unmet Need for Patients: No FDA-Approved Therapies

Sjogren's Disease is a debilitating disease characterized by autoimmune-driven destruction of the salivary and tear glands as well as arthritis, kidney, and lung dysfunction

Biological Rationale for CD40 Inhibition in Sjogren's Disease

There is substantial **external proof-of-concept** that the inhibition of the CD40-CD154 co-stimulatory interaction could be an efficacious therapeutic approach for Sjogren's Disease

Abiprubart Differentiation Potential

The **clear biological activity** and **favorable pharmacokinetics** of abiprubart have enabled **convenient chronic subcutaneous dosing** and could provide significant differentiation versus other assets in development for Sjogren's Disease

~50% of these patients are believed to be addressable with biologic therapies[2]

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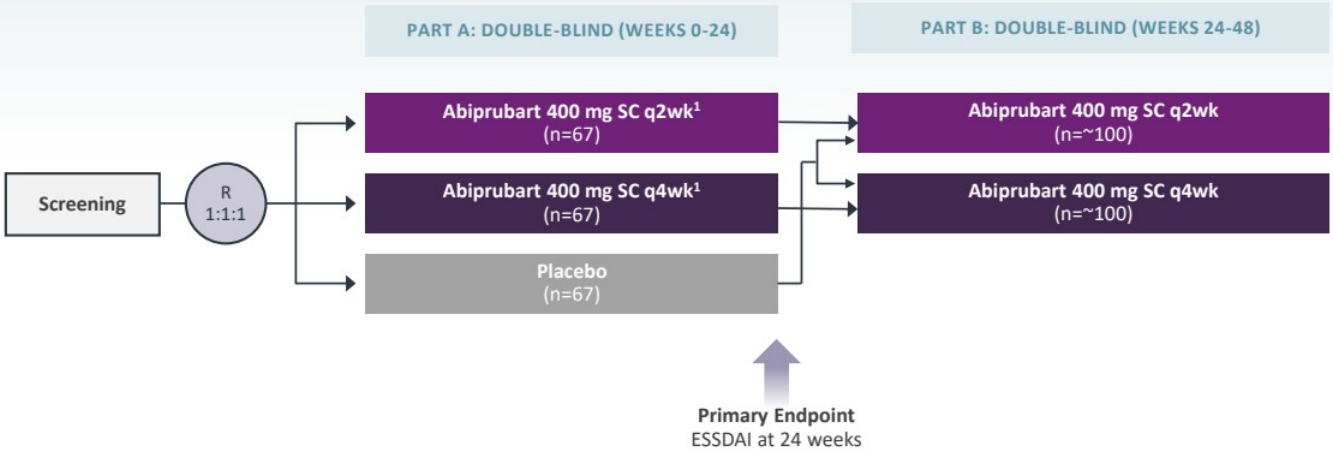
1) Maciel, G., Crowson, C.S., Matteson, E.L. and Cornec, D. (2017), Prevalence of Primary Sjögren's Syndrome in a US Population-Based Cohort. Arthritis Care & Research, 69: 1612-1616. https://doi.org/10.1002/acr.23173
2) Kiniksa primary market research

Planned Abiprubart Phase 2b Trial in Sjogren's Disease

Trial is expected to initiate in the second half of 2024



- Patients randomized to abiprubart groups in Part A will continue the same treatment assignment in Part B (without unblinding to prior treatment assignment)
- Patients randomized to Placebo in Part A will also be randomized 1:1 to an abiprubart treatment arm in Part B (without unblinding to prior treatment assignment)

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1) Both abiprubart dosing groups include an 800mg loading dose on Day 1
SC = subcutaneous; q2wk = every other week; q4wk = every four weeks; R = Randomization; ESSDAI = EULAR Sjogren's Syndrome Disease Activity Index

Abiprubart Phase 2 Trial in Rheumatoid Arthritis

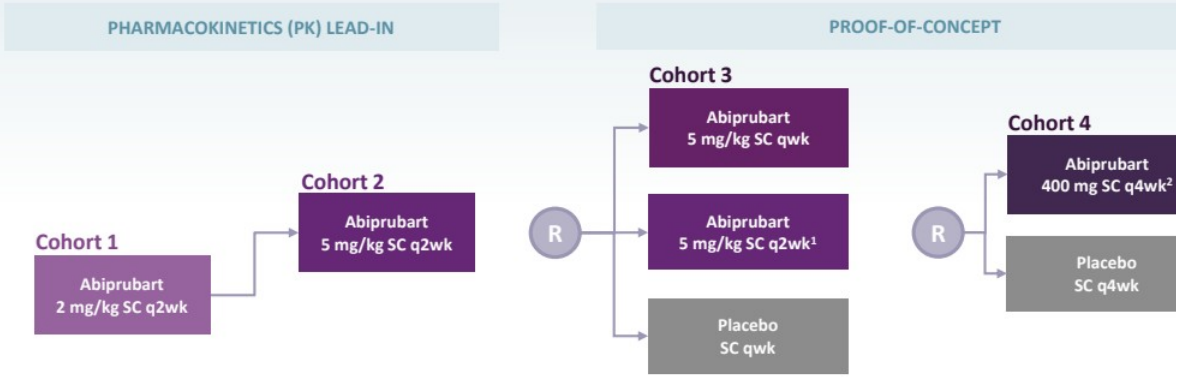
Study to evaluate the efficacy, dose response, PK, and safety of chronic SC dosing over a 12-week treatment du

PATIENT POPULATION:

- Patients with active RA who have been treated with a biological disease-modifying anti-rheumatic drug (bDMARDs) AND/OR Janus kinase inhibitor (JAKi) therapy for RA for ≥ 3 months and who have had inadequate response or have had to discontinue bDMARD and/or JAKi therapy due to intolerance or toxicity, regardless of treatment duration.

DISEASE CRITERIA:

- Six or more swollen joints and ≥ 6 tender joints at screening and baseline line visits; levels of high sensitivity C-reactive protein ≥ 5 mg/L; seropositivity for serum RF and/or ACPA at screening.



PK Lead-In: Cohorts 1-2

- Each cohort sequentially randomized 8 patients in a 3:1 (active:placebo) ratio; placebo recipients from Cohorts 1 and 2 were pooled
- Primary Endpoints:
  - Incidence of treatment-emergent adverse events (TEAEs)
  - Pharmacokinetics (C_{max}, $AUC_{(0-t)}$)
- Secondary Efficacy Endpoint:
  - Change from baseline in DAS28-CRP at Week 12

Proof of Concept: Cohorts 3-4

- Cohort 3 randomized 78 patients in a 1:1:1 ratio (n~26/arm)
- Cohort 4 randomized 51 patients in a 3:2 ratio (n=~20-30/arm)
- Primary Efficacy Endpoint:
  - Change from baseline in DAS28-CRP at Week 12
- Secondary Endpoints :
  - Incidence of treatment-emergent adverse events (TEAEs)
  - Pharmacokinetics (C_{max}, $AUC_{(0-t)}$)

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1) The 5 mg/kg SC q2wk group will receive weekly administrations of alternating active investigational product and matching blinded placebo
2) The Cohort 4 Abiprubart 400mg SC q4wk group includes a 600mg loading dose on Day 1

SC = subcutaneous; qwk = every week; q2wk = every other week; q4wk = every four weeks; AUC = Area Under the Curve; RF = Rheumatoid Factor; ACPA = anti-citrullinated protein antibodies, PD = Pharmacodynamics; PK = Pharmacokinetics; R = Randomization

Baseline Demographics (Cohort 3)[1]

	Abiprubart 5 mg/kg SC qwk (n=27)	Abiprubart 5 mg/kg SC q2wk (n=25)	Placebo (n=26)	Total (n=78)
Mean Age (Years)	58.5	60.0	57.6	58.7
Sex % (Male/Female)	18.5/81.5	20.0/80.0	7.7/92.3	15.4/84.6
Race				
White %; (n)	92.6 (n=25)	92.0 (n=23)	92.3 (n=24)	92.3 (n=72)
Black or African American %; (n)	3.7 (n=1)	8.0 (n=2)	7.7 (n=2)	6.4 (n=5)
Asian %; (n)	3.7 (n=1)	0	0	1.3 (n=1)
Country[2]				
United States %; (n)	29.6 (n=8)	28.0 (n=7)	38.5 (n=10)	32.1 (n=25)
Bulgaria %; (n)	0	4.0 (n=1)	11.5 (n=3)	5.1 (n=4)
Czechia %; (n)	11.1 (n=3)	4.0 (n=1)	3.8 (n=1)	6.4 (n=5)
Georgia %; (n)	7.4 (n=2)	12.0 (n=3)	11.5 (n=3)	10.3 (n=8)
Hungary %; (n)	18.5 (n=5)	4.0 (n=1)	3.8 (n=1)	9.0 (n=7)
Poland %; (n)	25.9 (n=7)	28.0 (n=7)	19.2 (n=5)	24.4 (n=19)
South Africa %; (n)	7.4 (n=2)	20.0 (n=5)	11.5 (n=3)	12.8 (n=10)

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1) Modified Intention to Treat (mITT) analysis population (all randomized subjects who received at least one dose of study drug and had a baseline assessment and at least one post-baseline assessment for the primary efficacy endpoint); 2) Cohorts 1 and 2 were conducted entirely in the United States

Baseline Disease Characteristics Balanced Across Treatment Arms (Cohort

	Abiprubart 5 mg/kg SC qwk (n=27)	Abiprubart 5 mg/kg SC q2wk (n=25)	Placebo (n=26)	Total (n=78)
DAS28-CRP Score				
DAS28-CRP[2]	5.58	5.92	5.98	5.82
Tender Joint Count-28[2]	13.4	16.1	15.4	14.9
Swollen joints-28[2]	10.1	12.2	12.0	11.4
Patient Global Assessment[2]	6.68	6.49	6.73	6.64
C-Reactive Protein (mg/L)[2]	16.00	18.72	26.74	20.45
Mean Duration of Rheumatoid Arthritis (years)	12.24	13.50	15.47	13.72
Rheumatoid factor (IU/mL)[2]	165.21	183.45	154.62	167.53
Anti-Cyclic Citrullinated Peptide %; (n)				
Positive	74.1 (n=20)	80.0 (n=20)	76.9 (n=20)	76.9 (n=60)
Negative	22.2 (n=6)	20.0 (n=5)	23.1 (n=6)	21.8 (n=17)
Intermediate	3.7 (n=1)	0	0	1.3 (n=1)

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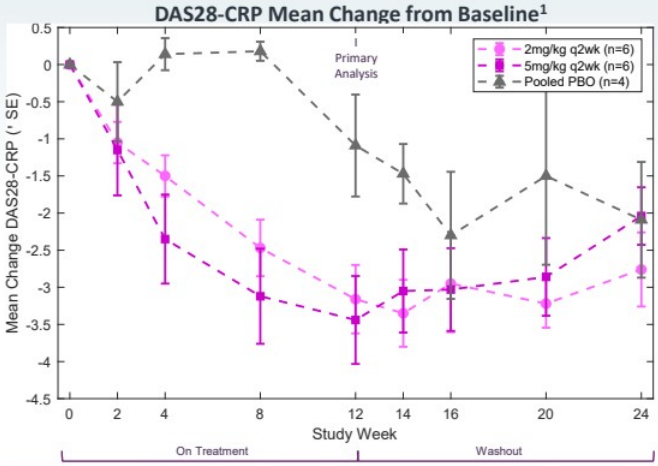
1) Modified Intention to Treat (mITT) analysis population (all randomized subjects who received at least one dose of study drug and had a baseline assessment and at least one post-baseline assessment for the primary efficacy endpoint); 2) Mean

Phase 2 Clinical Trial of Abiprubart in Rheumatoid Arthritis Met Primary Efficacy Endpoint (Change from Baseline in DAS28-CRP vs Placebo at Week 1

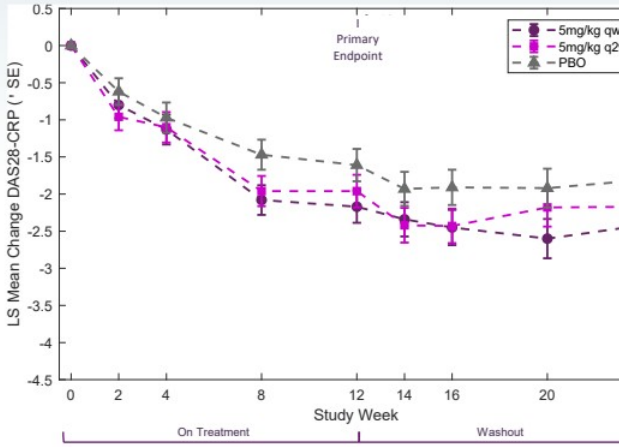
Cohorts 1 & 2
DAS28-CRP Mean Change from Baseline[1]



Cohort 1: in the abiprubart 2 mg/kg SC biweekly dosing group (n=6), mean change from baseline in DAS28-CRP at Week 12 was -3.16 points, compared to -1.09 points in pooled placebo recipients (n=4), (Mean Difference = -2.07, p=0.0312)

Cohort 2: in the abiprubart 5 mg/kg SC biweekly dosing group (n=6), mean change from baseline in DAS28-CRP at Week 12 was -3.44 points, compared to -1.09 points in pooled placebo recipients (n=4), (Mean Difference = -2.35, p=0.0338)

Cohort 3
DAS28-CRP LS Mean Change from Baseline[1,2]



In the abiprubart 5 mg/kg SC weekly dosing group (n=27), LS mean change [95% CI] baseline in DAS28-CRP at Week 12 was -2.17 [-2.60, -1.74] points, compared to -1. [-2.04, -1.17] points in placebo recipients (n=26), (LS Mean Difference = -0.57, p=0.0

In the abiprubart 5 mg/kg SC biweekly dosing group (n=25), LS mean change [95% CI] from baseline in DAS28-CRP at Week 12 was -1.96 [-2.40, -1.52] points, compared to -1.61 [-2.04, -1.17] points in placebo recipients (n=26), (LS Mean Difference = -0.36, p=0.2124)

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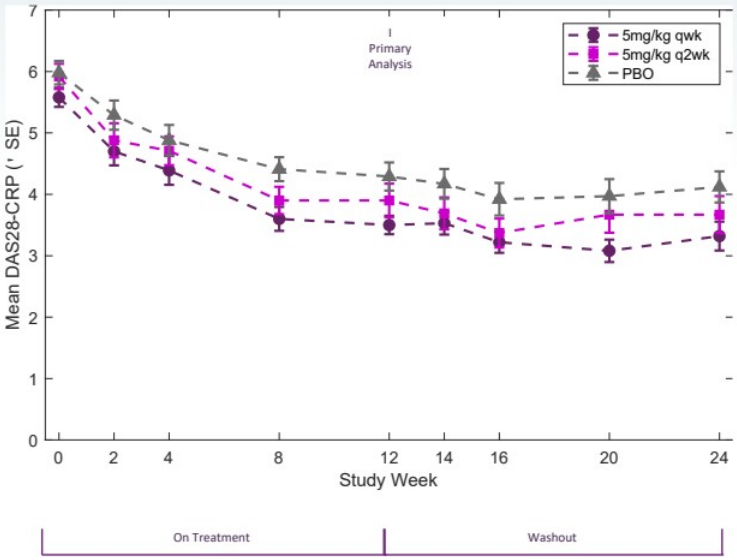
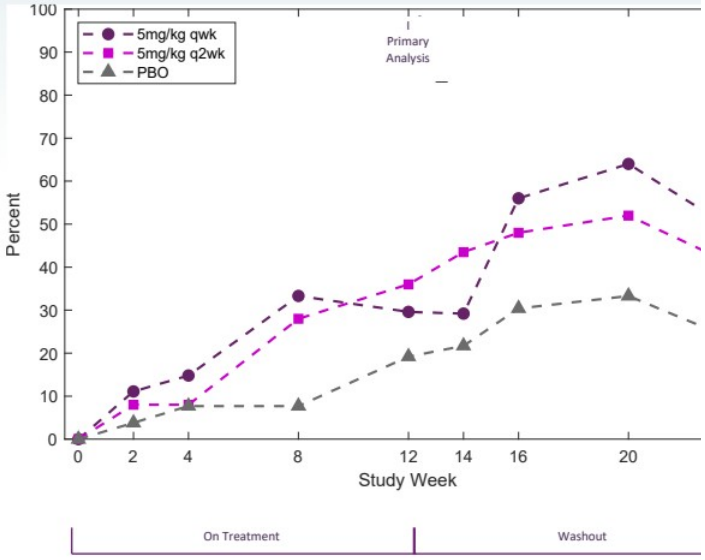
1) Final data; 2) Modified Intention to Treat (mITT) analysis population (all randomized subjects who received at least one dose of study drug and had a baseline assessment and at least one post-baseline assessment for the primary efficacy endpoint)
DAS28-CRP = Disease Activity Score of 28 Joints Using C-reactive Protein; SC = Subcutaneous; LS = Least Squares; CI = Confidence Interval

DAS28-CRP Scores Over Time (Cohort 3)[1]

DAS28-CRP Score Through Week 24



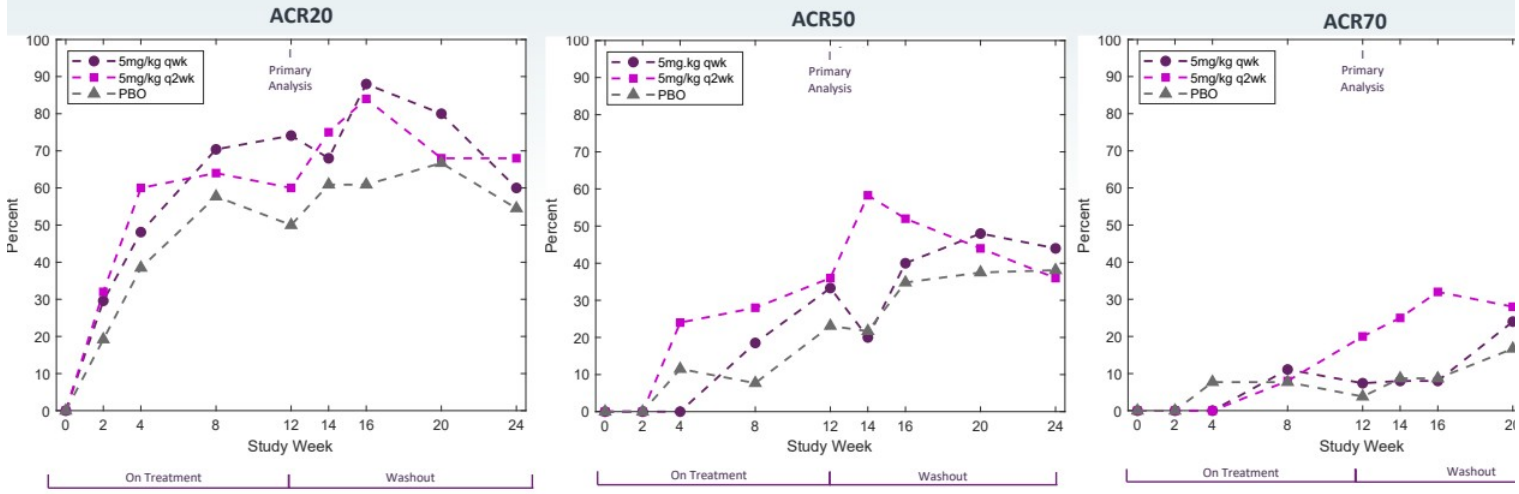
Patients Achieving DAS28 Low Disease Activity or Remission



1) Final data; Modified Intention to Treat (mITT) analysis population (all randomized subjects who received at least one dose of study drug and had a baseline assessment and at least one post-baseline assessment for the primary efficacy endpoint)

DAS28-CRP = Disease Activity Score of 28 Joints Using C-reactive Protein; Low Disease Activity = patients achieving DAS28-CRP low disease activity (≥2.6 and < 3.2); Remission = patients achieving DAS28-CRP remission (< 2.6)

ACR Responders Over Time (Cohort 3)[1]



1) Final data; Modified Intention to Treat (mITT) analysis population (all randomized subjects who received at least one dose of study drug and had a baseline assessment and at least one post-baseline assessment for the primary efficacy endpoint)
ACR20 = a composite measure defined as an improvement of 20% in the number of tender and swollen joints and a 20% improvement in three of the following five criteria: patient global assessment, physician global assessment, functional ability measure [most often Health Assessment Questionnaire (HAQ)], visual analog pain scale, and erythrocyte sedimentation rate or C-reactive protein (CRP); ACR50 and ACR70 = the same instruments as ACR20 with improvement levels defined as 50% and 70%, respectively, versus 20% for ACR20.

Abiprubart was Well-Tolerated in Phase 2 RA Trial (Cohort 3 Data)[1]

Category[2]	Abiprubart 5 mg/kg SC qwk (n=27)	Abiprubart 5 mg/kg SC q2wk (n=25)	Placebo (n=26)
Treatment Emergent Adverse Events (TEAEs)[3]	44.4 (n=12)	24.0 (n=6)	30.8 (n=8)
Drug Related TEAE[4]	7.4 (n=2)	8.0 (n=2)	7.7 (n=2)
TEAEs by Maximum severity[5]	44.4 (n=12)	24.0 (n=6)	30.8 (n=8)
Mild	29.6 (n=8)	12.0 (n=3)	15.4 (n=4)
Moderate	14.8 (n=4)	12.0 (n=3)	15.4 (n=4)
Severe	0	0	0
Potentially Life Threatening	0	0	0
Fatal	0	0	0
Serious TEAEs (SAE)	3.7 (n=1)[5]	0	3.8(n=1)
Drug-Related SAEs[3]	0	0	0
TEAEs Leading to Death	0	0	0
TEAEs Leading to Dose Interruption	3.7 (n=1)	0	3.8 (n=1)
TEAEs Leading to Treatment Discontinuation	0	0	0
TEAEs of Special Interest	0	4.0 (n=1)	0
Injection Site Reaction	3.7 (n=1)	4.0 (n=1)	0

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1) Safety Population: All randomized subjects who received at least one dose of study drug; 2) all categories are represented in percentages; 3) Defined as any event not present before exposure to study drug or any event already present that worsens in either intensity or frequency after exposure to study drug during treatment period; 4) Definitely related or possibly related, as assessed by the investigator; 5) Each subject has only been represented with the maximum severity; 5) Monaural deafness at Week 12, not related, resolved with pulse-dose steroids

Baseline Demographics (Cohort 4)[1]

	Abiprubart 400 mg SC q4wk (n=31)	Placebo (n=20)	Total (n=51)
Mean Age (Years)	58.8	58.3	58.6
Sex % (Male/Female)	19.4/80.6	25.0/75.0	21.6/78.4
Race			
White %; (n)	83.9 (n=26)	85.0 (n=17)	84.3 (n=43)
Black or African American %; (n)	9.7 (n=3)	5.0 (n=1)	7.8 (n=4)
Asian %; (n)	6.5 (n=2)	10.0 (n=2)	7.8 (n=4)
Country			
United States %; (n)	32.3(n=10)	20.0 (n=4)	27.5 (n=14)
Bulgaria %; (n)	6.5 (n=2)	0	3.9 (n=2)
Czechia %; (n)	16.1 (n=5)	20.0 (n=4)	17.6 (n=9)
Georgia %; (n)	9.7 (n=3)	15.0 (n=3)	11.8 (n=6)
Hungary %; (n)	22.6 (n=7)	15.0 (n=3)	19.6 (n=10)
Poland %; (n)	3.2 (n=1)	5.0 (n=1)	3.9 (n=2)
South Africa %; (n)	9.7 (n=3)	25.0 (n=5)	15.7 (n=8)

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1) Modified Intention to Treat (mITT) analysis population (all randomized subjects who received at least one dose of study drug and had a baseline assessment and at least one post-baseline assessment for the primary efficacy endpoint); the Phase 2 study of abiprubart in rheumatoid arthritis is ongoing, this topline analysis includes all patients having reached Week 12, and follow-up to Week 24 is ongoing;

Baseline Disease Characteristics: Balanced Across Treatment Arms (Cohort

	Abiprubart 400 mg SC q4wk (n=31)	Placebo (n=20)	Total (n=51)
DAS28-CRP Score			
DAS28-CRP[2]	5.65	5.89	5.75
Tender Joint Count-28[2]	13.6	15.2	14.2
Swollen joints-28[2]	9.30	11.9	10.30
Patient Global Assessment[2]	6.88	6.59	6.77
C-Reactive Protein (mg/L)[2]	22.65	22.75	22.69
Mean Duration of Rheumatoid Arthritis (years)	11.70	10.77	11.34
Rheumatoid factor (IU/mL)[2]	117.43	210.57	153.96
Anti-Cyclic Citrullinated Peptide %; (n)			
Positive	74.2 (n=23)	85.0 (n=17)	78.4 (n=40)
Negative	25.8 (n=8)	15.0 (n=3)	21.6 (n=11)
Intermediate	0	0	0

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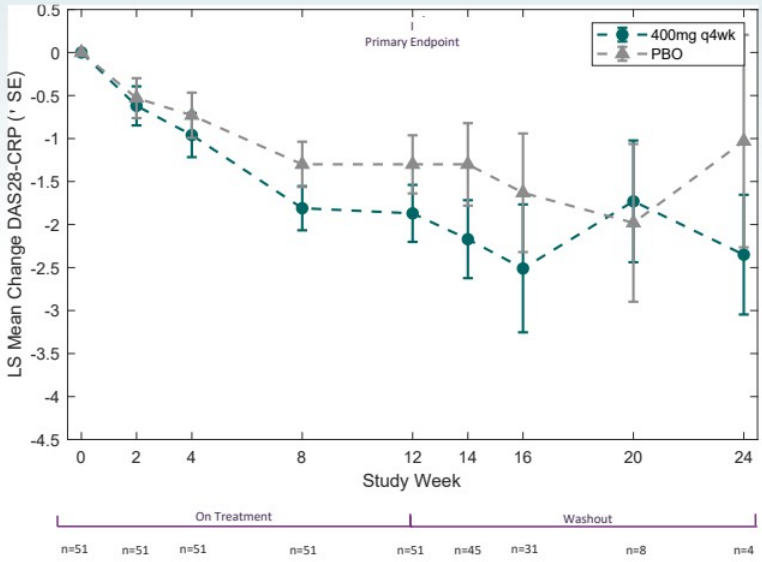
1) Modified Intention to Treat (mITT) analysis population (all randomized subjects who received at least one dose of study drug and had a baseline assessment and at least one post-baseline assessment for the primary efficacy endpoint); the Phase 2 study of abiprubart in rheumatoid arthritis is ongoing, this topline analysis includes all patients having reached Week 12, and follow-up to Week 24 is ongoing; 2) Mean

DAS28-CRP Scores Over Time (Cohort 4)[1]

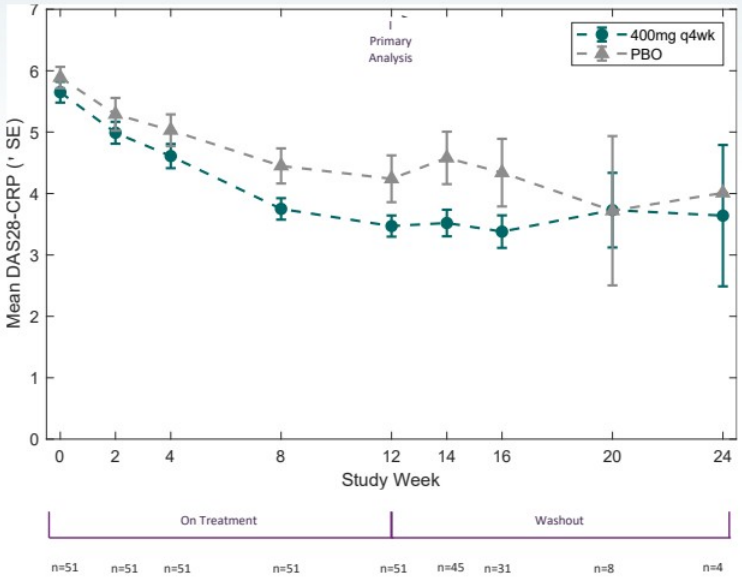


In the abiprubart 400 mg SC monthly dose group (n=31), the LS mean change [95% CI] from baseline in DAS28-CRP at Week 12 was -1.87 [-2.54, -1.21] points, compared to -1.30 [-1.98, -0.62] points in placebo recipients (n=20), (LS Mean Difference = -0.58, p=0.109)

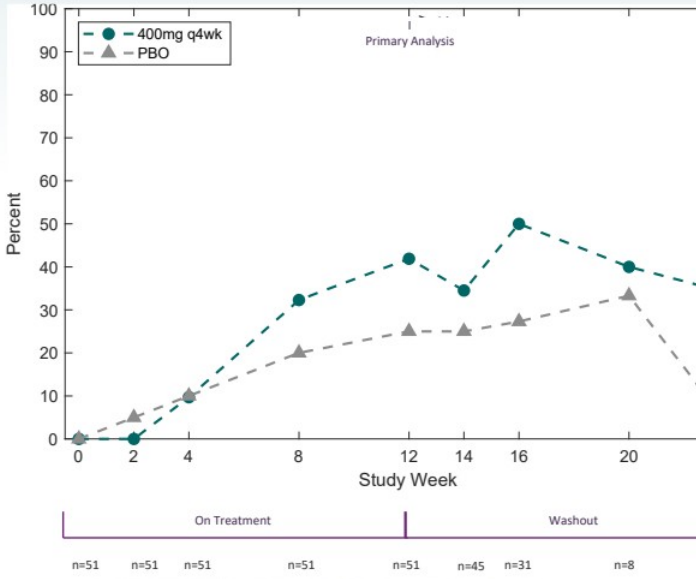
1) Topline data; Modified Intention to Treat (mITT) analysis population (all randomized subjects who received at least one dose of study drug and had a baseline assessment and at least one post-baseline assessment for the primary efficacy endpoint); the Phase 2 study of abiprubart in rheumatoid arthritis is ongoing, this topline analysis includes all patients having reached Week 12, and follow-up to Week 24 is ongoing

DAS28-CRP Scores Over Time (Cohort 4)[1]

DAS28-CRP Score Through Week 24

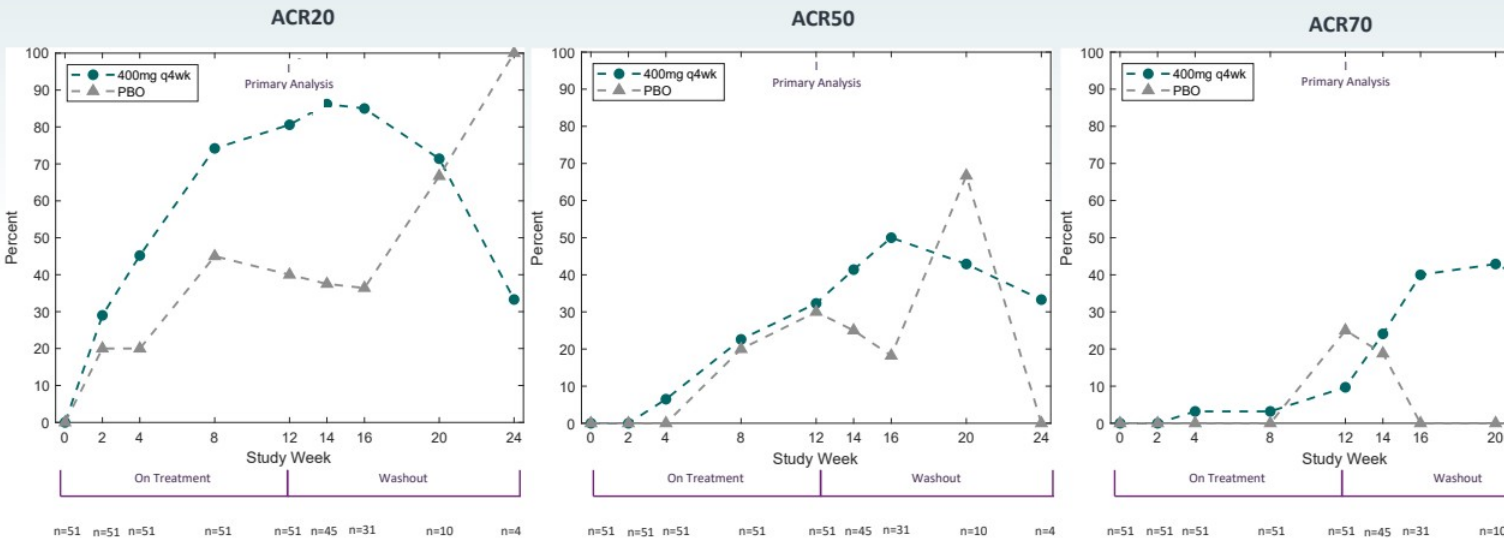


Patients Achieving DAS28 Low Disease Activity or Remission



1) Topline data; Modified Intention to Treat (mITT) analysis population (all randomized subjects who received at least one dose of study drug and had a baseline assessment and at least one post-baseline assessment for the primary efficacy endpoint); the Phase 2 study of abiprubart in rheumatoid arthritis is ongoing, this topline analysis includes all patients having reached Week 12, and follow-up to Week 24 is ongoing
DAS28-CRP = Disease Activity Score of 28 Joints Using C-reactive Protein; Low Disease Activity = patients achieving DAS28-CRP low disease activity (≥2.6 and < 3.2); Remission = patients achieving DAS28-CRP remission (< 2.6

ACR Responders Over Time (Cohort 4)[1]



1) Topline data; Modified Intention to Treat (mITT) analysis population (all randomized subjects who received at least one dose of study drug and had a baseline assessment and at least one post-baseline assessment for the primary efficacy endpoint); the Phase 2 study of abiprubart in rheumatoid arthritis is ongoing, this topline analysis includes all patients having reached Week 12, and follow-up to Week 24 is ongoing
ACR20 = a composite measure defined as an improvement of 20% in the number of tender and swollen joints and a 20% improvement in three of the following five criteria: patient global assessment, physician global assessment, functional ability measure [most often Health Assessment Questionnaire (HAQ)], visual analog pain scale, and erythrocyte sedimentation rate or C-reactive protein (CRP); ACR50 and ACR70 = the same instruments as ACR20 with improvement levels defined as 50% and 70%, respectively, versus 20% for ACR20.

Abiprubart was Well-Tolerated in Phase 2 RA Trial (Cohort 4 Data)[1]

Category[2]	Abiprubart 400mg SC q4wk (n=31)	Placebo (n=20)
Treatment Emergent Adverse Events (TEAEs)[3]	25.8 (n=8)	40.0 (n=8)
Drug Related TEAE[4]	9.7 (n=3)	5.0 (n=1)
TEAEs by Maximum severity[5]	25.8 (n=8)	40.0 (n=8)
Mild	12.9 (n=4)	25.0 (n=5)
Moderate	12.9 (n=4)	15.0 (n=3)
Severe	0	0
Potentially Life Threatening	0	0
Fatal	0	0
Serious TEAEs (SAE)	0	0
Drug-Related SAEs[3]	0	0
TEAEs Leading to Death	0	0
TEAEs Leading to Dose Interruption	0	0
TEAEs Leading to Treatment Discontinuation	3.2 (n=1)	5.0 (n=1)
TEAEs of Special Interest	0	0
Injection Site Reaction	6.5 (n=2)	0

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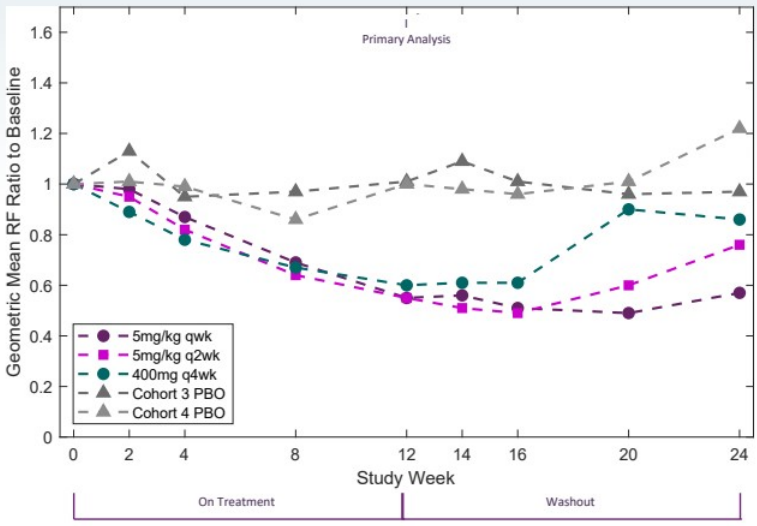
1) Safety Population: All randomized subjects who received at least one dose of study drug; 2) all categories are represented in percentages; 3) Defined as any event not present before exposure to study drug or any event already present that worsens in either intensity or frequency after exposure to study drug during treatment period; 4) Definitely related or possibly related, as assessed by the investigator; 5) Each subject has only been represented with the maximum severity; 5) Monaural deafness at Week 12, not related, resolved with pulse-dose steroids

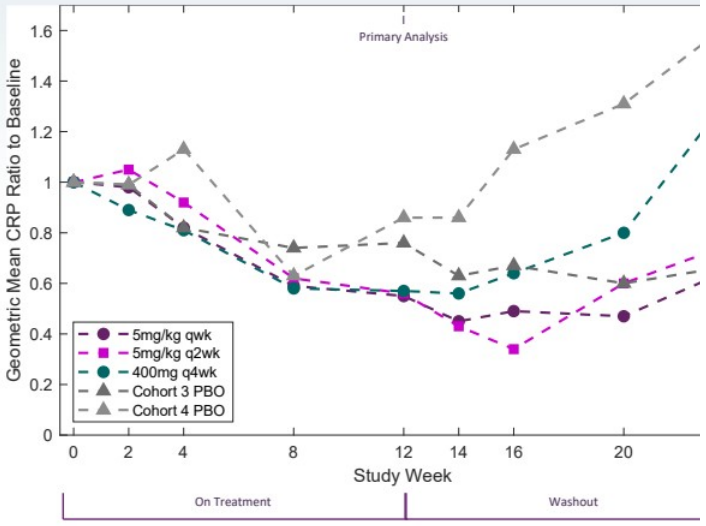
Abiprubart Significantly Reduced Disease-Related Inflammatory Markers (Cohorts 3 & 4)[1]

Rheumatoid Factor
Geometric Mean Ratio to Baseline[2]



Cohort 3 n's	n=78	n=78	n=78	n=78	n=70	n=74	n=74	n=72
Cohort 4 n's	n=51	n=51	n=50	n=47	n=45	n=31	n=11	n=4

C-Reactive Protein
Geometric Mean Ratio to Baseline



Cohort 3 n's	n=78	n=78	n=78	n=78	n=70	n=74	n=74
Cohort 4 n's	n=51	n=51	n=50	n=47	n=45	n=31	n=11

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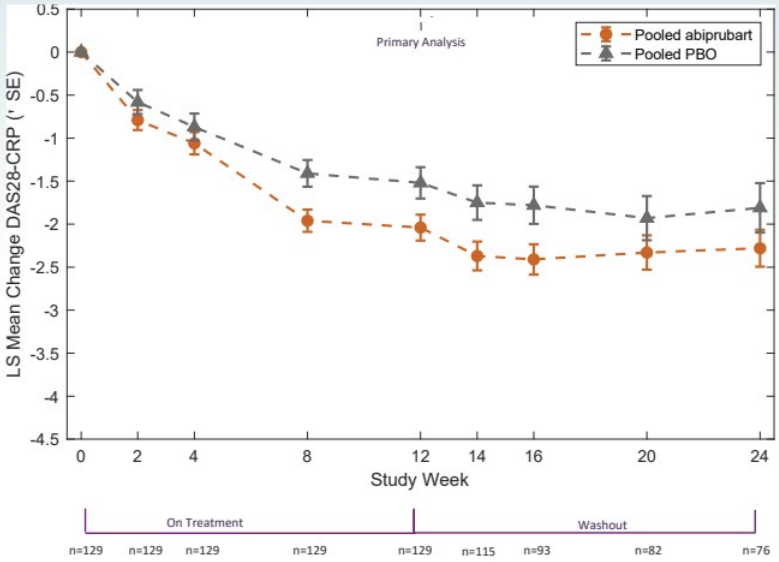
1) Modified Intention to Treat (mITT) analysis population (all randomized subjects who received at least one dose of study drug and had a baseline assessment and at least one post-baseline assessment for the primary efficacy endpoint); 2) In both Cohort 3 abiprubart dose groups (5 mg/kg SC weekly and 5 mg/kg SC biweekly) ($p<0.0001$); in the Cohort 4 abiprubart dose group (400 mg SC monthly) ($p=0.0003$).

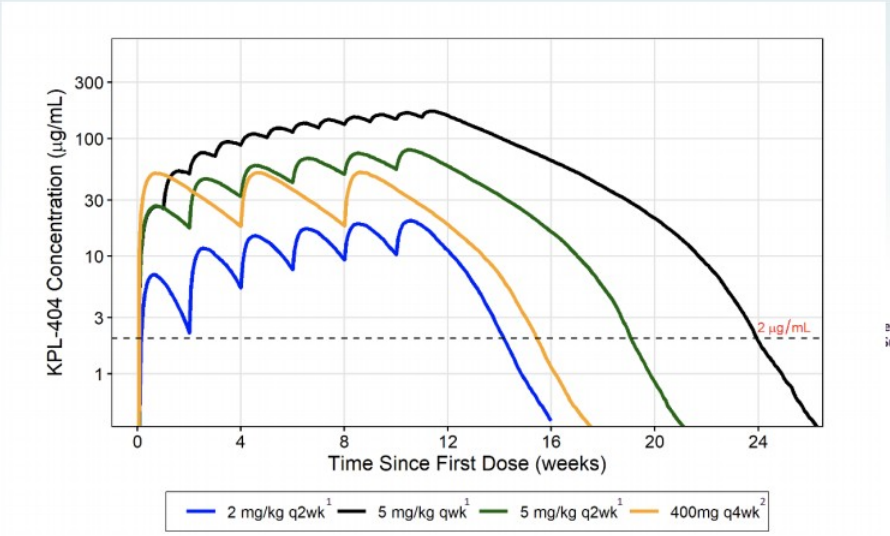
DAS28-CRP Scores Over Time in Pooled Abiprubart and Placebo Groups (Cohorts 3 & 4)[1]



In the pooled abiprubart group (n=83), the LS mean change [95% CI] from baseline in DAS28-CRP at Week 12 was -2.04 [-2.34, -1.74] points, compared to -1.52 [-1.88, -1.16] points in placebo recipients (n=46), (LS Mean Difference = -0.52, nominal p=0.010)

1) Modified Intention to Treat (mITT) post-hoc analysis population (all randomized subjects who received at least one dose of study drug and had a baseline assessment and at least one post-baseline assessment for the primary efficacy endpoint)

PK-Modeling From the Phase 2 Rheumatoid Arthritis Trial (Cohorts 1-4)



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iuppression

Modeling data generated based on PK data from Cohorts 1-4 of the abiprubart Phase 2 trial in rheumato arthritis as well as Phase 1 data from healthy volunteers

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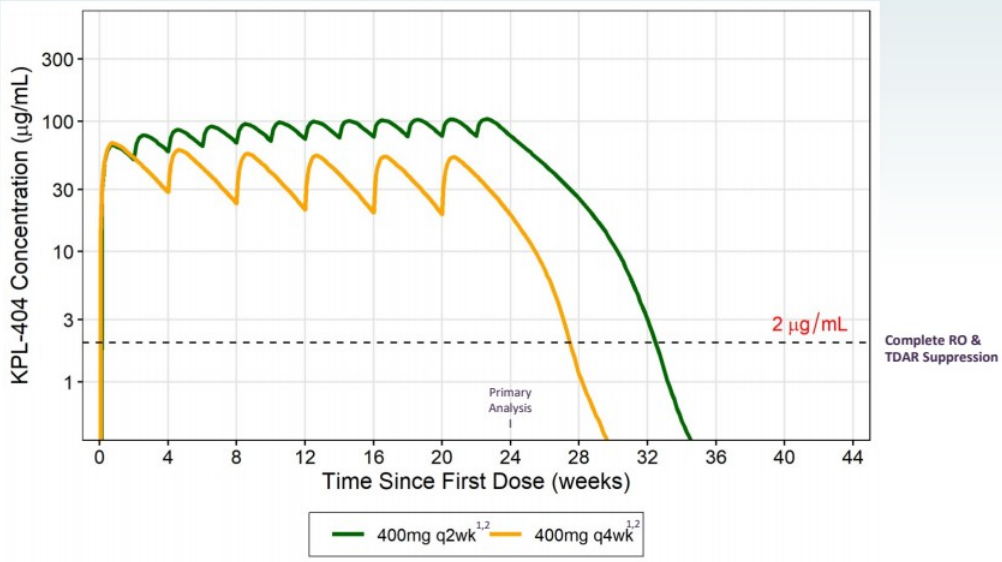
1) All doses are subcutaneous; 2) The Cohort 4 abiprubart 400mg SC q4wk group includes a 600mg loading dose on Day 1
RO = receptor occupancy; TDAR = T-Cell Dependent Antibody Response

PK-Modeling and Dose Simulations for the Phase 2b Sjogren's Trial



Modeling data generated based on PK data from Cohorts 1-4 of the abiprubart Phase 2 trial in rheumato arthritis as well as Phase 1 data from healthy volunteers

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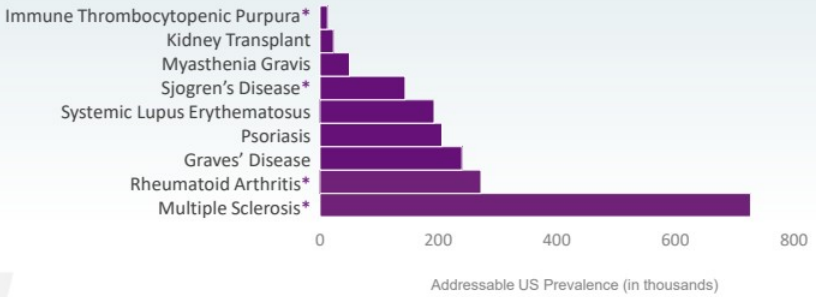
1) All doses are subcutaneous; 2) Both abiprubart dosing groups include an 800mg loading dose on Day 1
RO = receptor occupancy; TDAR = T-Cell Dependent Antibody Response

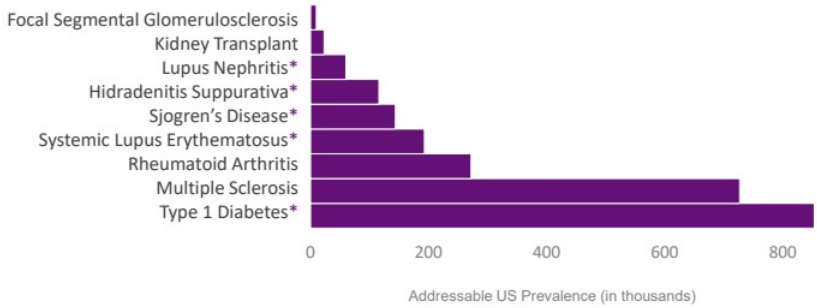
Potential for Evaluation of Abiprubart in a Range of Autoimmune Diseases

CD40/CD154 interaction has been implicated in a number of devastating diseases

Indications with Published Data



Indications with Pending Data & Trials Ongoing



INDICATION SELECTION CRITERIA

- Robust data or proof-of-concept supporting mechanism
- Differentiation vs. competitors
- Commercial attractiveness

*Indications evaluated with subcutaneous administration

Sources: 2019 numbers: https://unos.org/data/transplant-trends/; Hunter et al. Prevalence of rheumatoid arthritis in the United States adult population in healthcare claims databases, 2004-2014; Rheumatol Int. 2017 Sep;37(9):1551-1557; Overall Prevalence: Maciel et al, Arthritis Care Res (Hoboken) 2017; Qin et al, Ann Rheum Dis 2015; UpToDate; Baldini et al. Prevalence of Severe Extra-Glandular Manifestations in a Large Cohort of Patients with Primary Sjögren's Syndrome; 2012 ACR/ARHP Annual Meeting, ABSTRACT NUMBER: 2185; Wallin et al. The prevalence of MS in the United States A population-based estimate using health claims data, Neurology, March 5, 2019; Somers et al.; Prevalence of Systemic Lupus Erythematosus in the United States: Preliminary Estimates from a Meta-Analysis of the Centers for Disease Control and Prevention Lupus Registries; 2019 ACR/ARP Annual Meeting ABSTRACT NUMBER: 2886; Garg et al. JAMA Dermatol. 2017;153(8):760-764. doi:10.1001/jamadermatol.2017.0201 Sex- and Age-Adjusted Population Analysis of Prevalence Estimates for Hidradenitis Suppurativa in the United States; MayoClinic.org; Yale J Biol Med. 2013 Jun; 86(2): 255–260. N Engl J Med 2016;375:2570-81; https://www.diabetesresearch.org/diabetes-statistics; Nephcure.org; Kitiyakara C, Eggers P, Kopp JB. Twenty-one-year trend in ESRD due to focal segmental glomerulosclerosis in the United States Am J Kidney Dis. 2004 Nov;44(5):815-25; Rachakonda et al. J Am Acad Dermatol . 2014 Mar;70(3):512-6. doi: 10.1016/j.jaad.2013.11.013. Epub 2014 Jan 2. Psoriasis prevalence among adults in the United States; Yeung et al. Psoriasis severity and the prevalence of major medical co-morbidities: a population-based study; JAMA Dermatol. 2013 Oct 1; 149(10): 1173–1179; Hoover et al. Kidney Int. 2016 Sep; 90(3): 487–492. Insights into the Epidemiology and Management of Lupus Nephritis from the U.S. Rheumatologist's Perspective.

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Financials Fourth Quarter and Full-Year 2023

Fourth Quarter and Full-Year 2023 Financial Results

Income Statement	Three Months Ended December 31,		Year Ended December 31,	
	2023	2022	2023	2022
Product Revenue	$71.2M	$39.9M	$233.2M	$122.5M
License and Collaboration Revenue	$12.2M	$21.9M	$37.1M	$97.7M
Total Revenue	**$83.4M**	**$61.9M**	**$270.3M**	**$220.2M**
Cost of Goods Sold	$9.6M	$6.7M	$33.4M	$22.9M
Collaboration Expenses[1]	$16.9M	$7.5M	$56.5M	$24.1M
Research and Development	$20.1M	$14.4M	$76.1M	$65.5M
Selling, General and Administrative	$36.7M	$27.2M	$129.4M	$98.0M
Total Operating Expenses	**$83.3M**	**$55.8M**	**$295.5M**	**$210.4M**
Income Tax Benefit (Provision)	$22.8M	($2.4M)	$30.7M	$172.3M
Net Income	**$25.2M**	**$4.5M**	**$14.1M**	**$183.4M**

Collaboration Expenses[1]	Three Months Ended December 31,		Year Ended December 31	
	2023	2022	2023	2
ARCALYST Net Sales	**$71.2M**	**$39.9M**	**$233.2M**	**$1**
Profit Split-Eligible Cost of Goods Sold[2]	($9.3M)	($6.5M)	($32.4M)	($2
Commercial, Marketing, Regulatory and Other Expenses	($28.0M)	($18.4M)	($87.7M)	($6
ARCALYST Collaboration Operating Profit	**$33.9M**	**$15.0M**	**$113.0M**	**$3**
ARCALYST Licensing Proceeds	$0.0M	$0.0M	$0.0M	$6
Collaboration Expenses[1]	**$16.9M**	**$7.5M**	**$56.5M**	**$2**

Balance Sheet	December 31, 2023	December 31 2022
Cash, Cash Equivalents and Short-term Investments	**$206.4M**	**$190.6M**

Kiniksa expects to remain cash flow positive on an annual basis within its current operating plan[3]

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1) Subject to the terms of the definitive agreements between Kiniksa and Regeneron; 50% of ARCALYST Collaboration Operating Profit plus 50% of ARCALYST Licensing Proceeds;
2) Profit Split-Eligible Cost of Goods Sold = total cost of goods sold - amortization of Regeneron milestone payment
3) Financial guidance as of April 2024

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Appendix Out-Licensing Agreements

Out-Licensing Agreements

Partnership with Huadong Medicine Gives Kiniksa Opportunity to Expand Footprint into Asia Pacific Region (Excluding Japan)

- In February 2022, Kiniksa announced a strategic collaboration with Huadong to develop and commercialize ARCALYST and mavrilimumab in Greater China, South Korea, Australia and 18 other countries, excluding Japan
- Kiniksa received a $22M upfront payment and is eligible to receive up to approximately $640M in specified development, regulatory and sales-based milestone along with tiered royalty payments
- Collaboration provided non-dilutive capital, cost-sharing, and additional resources to help accelerate development and commercialization efforts

License Agreement with Roche Genentech for Global Rights to Develop and Commercialize Vixarelimab

- Kiniksa has received $100 million in upfront and near-term payments:
  - $80 million, which was received following the transaction's closing in Q3 2022
  - $20 million, which was received following Kiniksa's last delivery of certain drug supplies to Genentech in Q1 2023
- Kiniksa is eligible to receive up to approximately $600 million in certain clinical, regulatory, and sales-based milestones, before fulfilling upstream financial obligations, of which approximately $575 million remains
- Kiniksa is also eligible to receive royalties on annual net sales ranging from low-double digits to mid-teens, before fulfilling upstream financial obligations
- Proceeds from the transaction to help grow cardiovascular franchise and build autoimmune franchise

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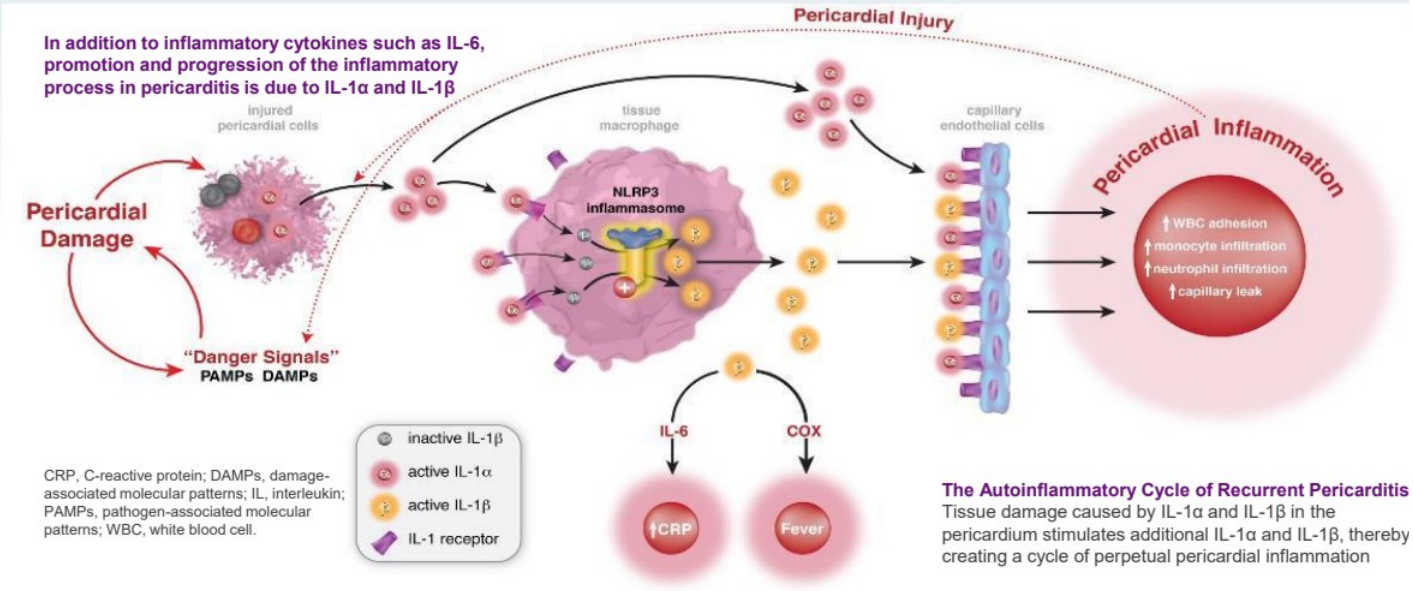
Appendix
ARCALYST (rilonacept)

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



CRP, C-reactive protein; DAMPs, damage-associated molecular patterns; IL, interleukin; PAMPs, pathogen-associated molecular patterns; WBC, white blood cell.

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

Brucato A, et al. Int Emerg Med 2018 https://doi.org/10.1007/s11739-018-1907-x
Dinarello CA, et al. Nat Rev Drug Discov 2012;11:633-652

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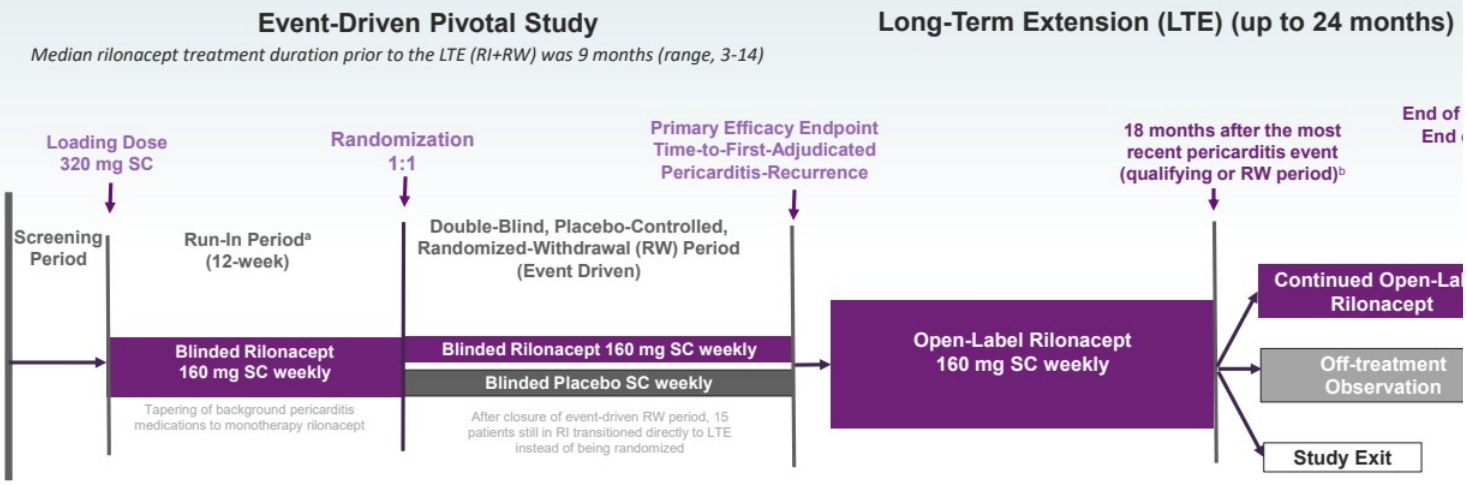
RHAPSODY Design



[a] The duration of the run-in period was concealed from patients, so that they were blinded to the timing of randomization

[b] For each patient in the LTE, **a decision was made 18 months after the most recent pericarditis recurrence (Qualifying or RW period)** based on clinical status and one of the following actions was taken at the investigator's discretion:

- Continue rilonacept on-study

OR

- Suspend rilonacept treatment and remain on-study for observation (rilonacept rescue for recurrence allowed)

OR

- Discontinue the LTE completely (no further observation)

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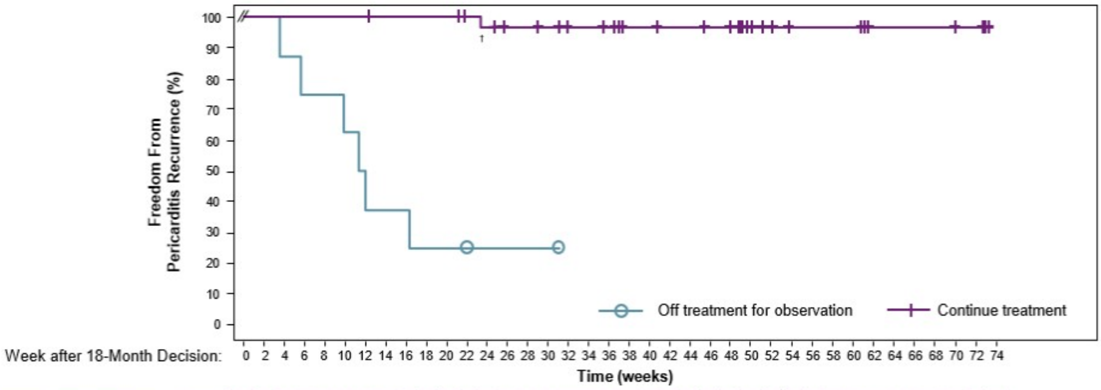
Adapted from: Imazio M, Klein AL, et al. Prolonged Rilonacept Treatment in Rhapsody Long-term Extension Provided Persistent Reduction of Pericarditis Recurrence Risk. Poster 2223 (Presented at AHA Scientific Sessions 2022)

RHAPSODY Long-Term Extension Data Demonstrated Rilonacept Treatm Beyond 18 months Resulted in Continued Treatment Response[1]



Continued Rilonacept Treatment, Patients at Risk, n	33	33	33	33	33	33	33	32	32	32	32	30	29	27	27	25	24	23	22	18	18	17	17	16	16	11	9	7	7	7	7	4	4	4	4	4	3	0
Off Treatment for Observation, Patients at Risk, n	8	8	7	6	6	6	4	3	3	2	2	2	1	1	1	1	0																					

†The patient with a recurrence at 23.4 weeks had interrupted rilonacept treatment ~4 weeks prior.

Hazard ratio = 0.02
Log-rank *P*<0.0001
Risk reduction = 98%

	N	Patients with Recurrence,[a] n (%)	W Rec Medi
Continued rilonacept treatment	**33**	**1 (3)**	**NE**
Off treatment for observation	**8**	**6 (75)**	**11.8**

[a]After 18-month decision.
CI, confidence interval; NE, not estimable.

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1) Imazio M, Klein AL, et al. Prolonged Rilonacept Treatment in Rhapsody Long-term Extension Provided Persistent Reduction of Pericarditis Recurrence Risk. Poster 2223 (Presented at AHA Scientific Sessions 2022)

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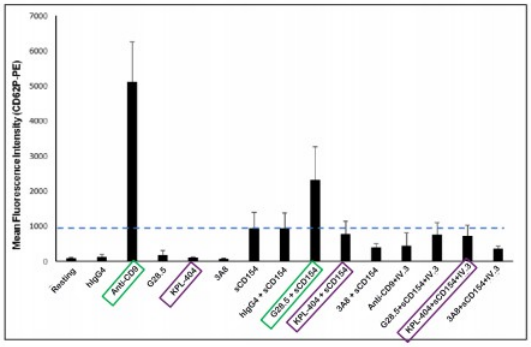
Appendix
Abiprubart

Abiprubart Does Not Cause Platelet Activation or Aggregation *in vitro*

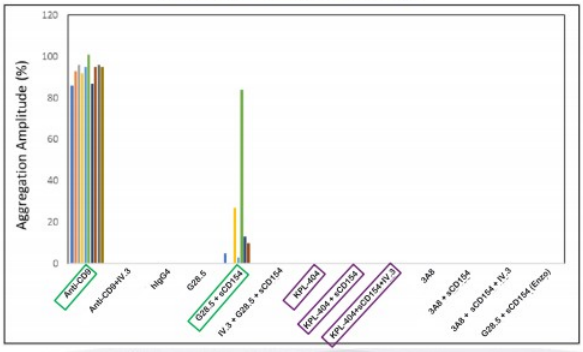
- At least three first-generation IgG1 anti-CD154 mAbs* were associated with thromboembolic events in humans and NHPs[1]
- **Mechanism:** Activation of platelets through cross-linking mediated by IgG-Fc/FcγRIIa interaction
  - Platelet activation observed *in vivo* with anti-CD154 mAbs with active Fc region
  - Platelet activation *in vitro* by anti-CD40 mAbs requires presence of sCD154 and active Fc region
  - Absence of an active Fc-region prevents platelet activation[1,2]

Abiprubart did not cause upregulation of the cell-surface platelet activation marker CD62P
Abiprubart did not induce platelet aggregation in the presence (or absence) of soluble CD154[3]

Abiprubart Alone and in Combination with sCD154 does not increase CD62P Expression on the Platelet Surface



Abiprubart Alone and in Combination with sCD154 does not increase Platelet Aggregation Amplitude (%)



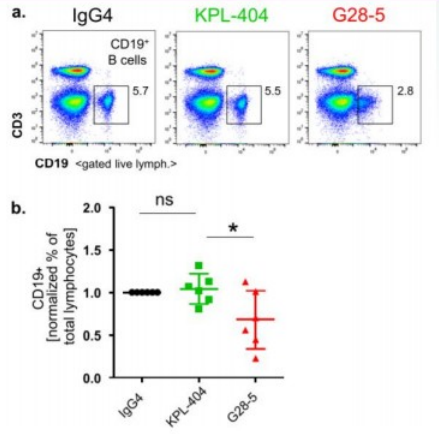
Positive controls:
- G28.5: anti-CD40 causes sCD40L-de platelet activatio (Langer et al., Thromb Haem 1137-1146)
- Anti-CD9: mAb– c sCD40L-independ activation
- IV.3 - anti-FcγRII

*ruplizumab/hu5c8, toralizumab/IDEC-131, ABI793

Sources: 1) Law & Grewal, Advances in Experimental Medicine and Biology, vol 647. Springer; 2) Shock et al., Arthritis Research & Therapy 17, Article Number: 234 (2015); 3) KNSA in-house data

Abiprubart Does Not Reduce B cell Numbers, Activate B Cells, or Induce B Cell Proliferation *in vitro*

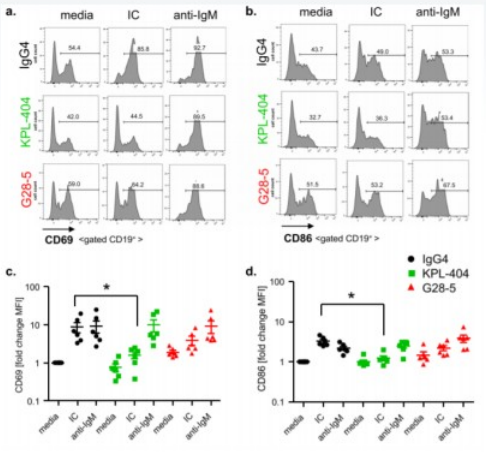
Abiprubart does not reduce B cell numbers in activated PBMCs *in vitro*



G28.5: agonistic aCD40 mAb

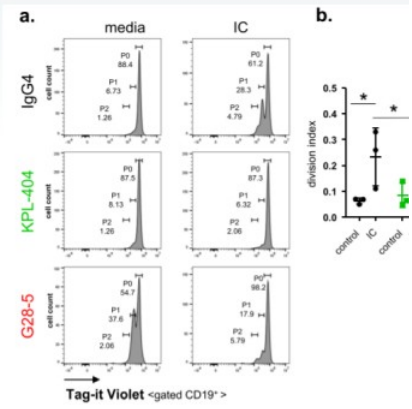
PBMCs were cultured in the presence of 10 µg/ml IgG4 isotype control or anti-CD40 Abs Abiprubart, or the agonistic aCD40 mAb, G28-5 (16–18 h of cell culture)

Abiprubart does not induce B cell activation



PBMCs were cultured in the presence of 10 µg/ml IgG4 isotype control or anti-CD40 Abs Abiprubart, or G28-5 (16–18 h of cell culture). Cells were left unstimulated (media control) or stimulated with CD3/CD28 cross-linker IC or F(ab')2 goat anti-human IgM (anti-IgM)

Abiprubart does not induce B cell proliferation *in vitro*



PBMCs were labeled with a cell proliferation tracker d
it Violet) and cultured for 5 days in the presence of 10
IgG4 isotype control Ab or anti-CD40 Abs—Abiprubar
G28-5. Cells were left untreated (media control) or sti
with anti-CD3/CD28 cross-linking reagent ImmunoCul

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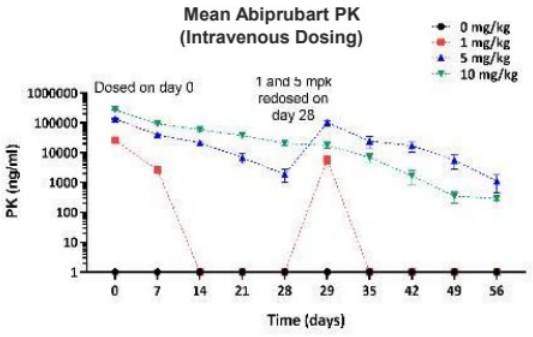
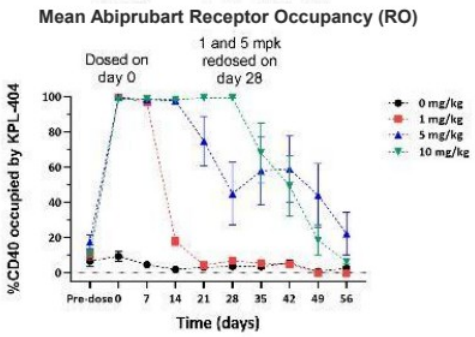
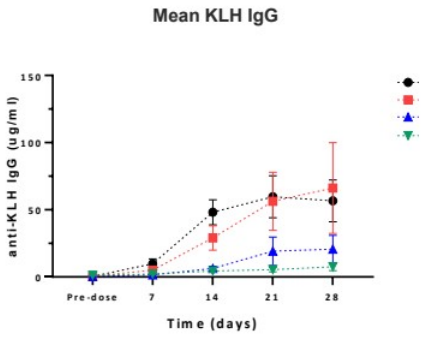
*Marken et al, Arthritis Res Ther. 2021 Jan 21;23(1):36. doi: 10.1186/s13075-021-02425-x.

Abiprubart Demonstrated Prolonged Suppression of TDAR Response in a Non-Human Primate Model







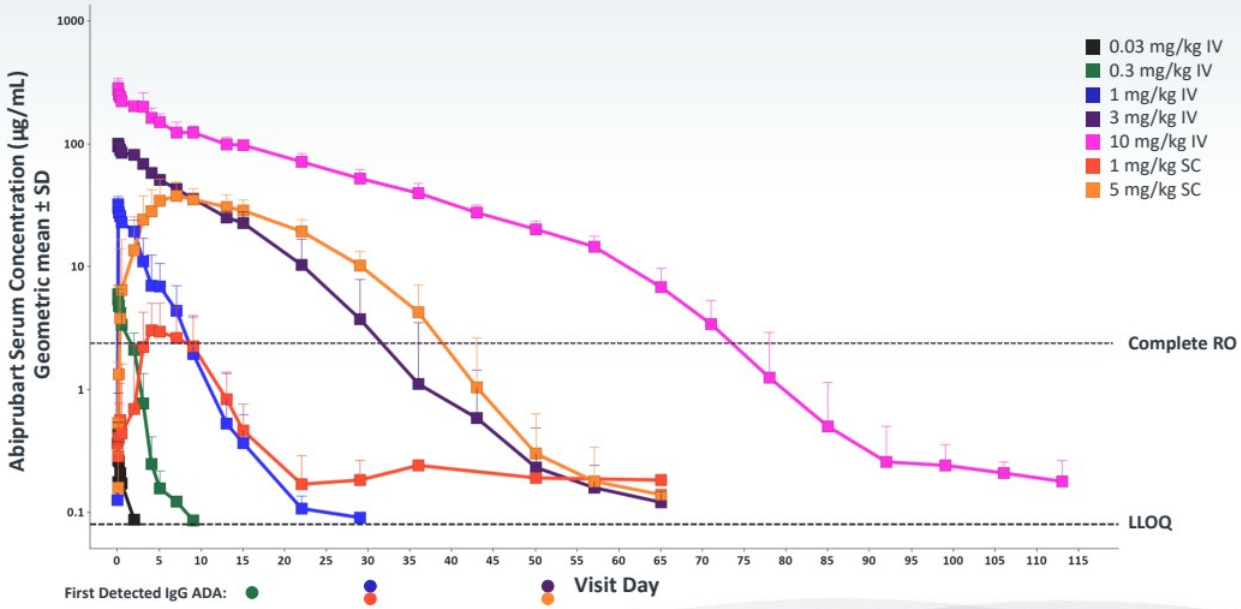
Showed linear pharmacokinetic profile with low variability between non-human primate subjects (n=7)	*Abiprubart achieved 100% receptor occupancy for 2 weeks in all animals at 5mg/kg and 4 weeks in all animals at 10mg/kg*	*Complete suppression of primary T-cell dep antigen response correlated with 100% rec occupancy*

Source: Muralidharan et al. Preclinical immunopharmacologic assessment of KPL-404, a novel, humanized, non-depleting antagonistic anti-CD40 monoclonal antibody. J Pharmacol Exp Ther. 2022, 381(1):12-21

TDAR = T-cell dependent antibody response; KLH = keyhole limpet hemocyanin

Final Data from Abiprubart Single-Ascending-Dose Phase 1 Study

Pharmacokinetic profiles for abiprubart

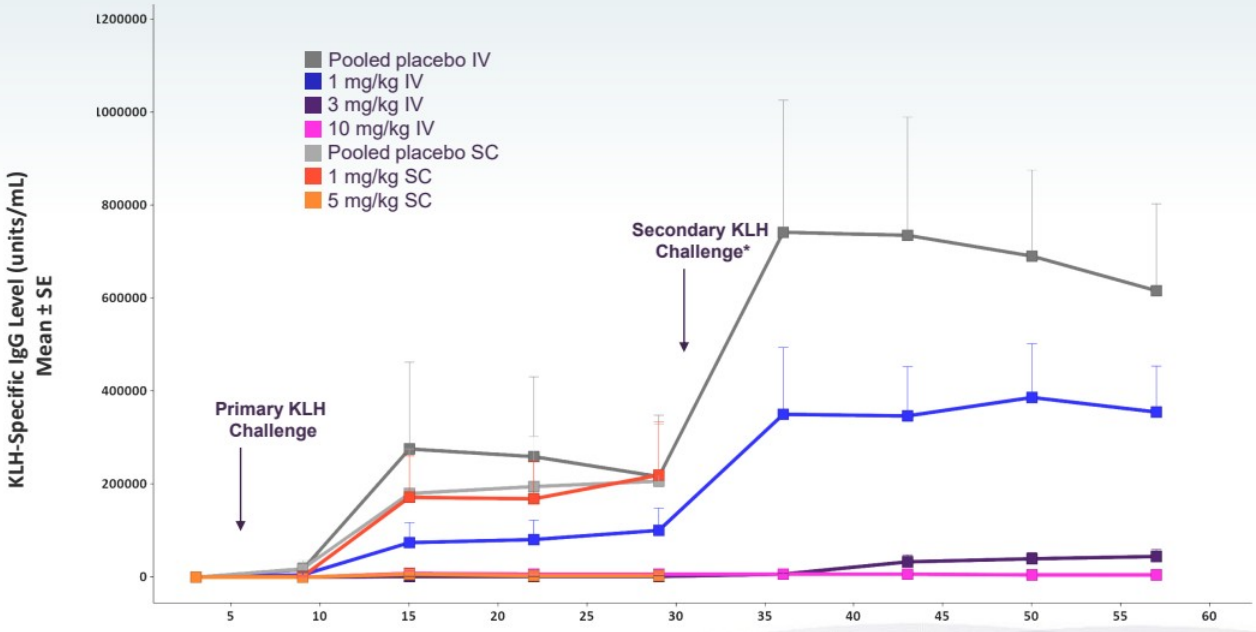


Source: Samant M, Ziemniak J, Paolini JF. First-in-Human Phase 1 Randomized Trial with the Anti-CD40 Monoclonal Antibody KPL-404: Safety, Tolerability, Receptor Occupancy, and Suppression of T-Cell-Dependent Antibody Response. J Pharmacol Exp Ther. 2023 Dec;387(3):306-314.

SD = standard deviation (upward bars depicted); IV = intravenous; SC = subcutaneous; LLOQ = lower limit of quantitation; ADA = anti-drug antibody

Final Data from Abiprubart Single-Ascending-Dose Phase 1 Study

T-Cell Dependent Antibody Response (TDAR) for KLH antigen challenge



*Only IV cohorts were rechallenged with KLH on day 29

Source: Samant M, Ziemniak J, Paolini JF. First-in-Human Phase 1 Randomized Trial with the Anti-CD40 Monoclonal Antibody KPL-404: Safety, Tolerability, Receptor Occupancy, and Suppression of T-Cell-Dependent Antibody Response. J Pharmacol Exp Ther. 2023 Dec;387(3):306-314.

KLH = keyhole limpet hemocyanin

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Corporate Presentation

APRIL 2024