KINIKSA

Corporate Presentation

OCTOBER 2026

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 International, plc (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," "strategy," 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 value drivers; potential indications; potential market opportunities and competitive position; market trends and dynamics; 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 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, including the data from the interval analysis of our Phase 2 clinical trial of KPL-387 in recurrent pericarditis; 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 products and 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; business development activities and their impact on our financial performance and strategy; changes in our operating plan, business development strategy or funding requirements; existing or new competition; current and future healthcare reforms, including those affecting the delivery of or payment for healthcare products and services; and the impact of global economic policy, including any uncertainty in national and international markets.

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 differ 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 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, 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 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.

ARCALYST® is a registered trademark of Regeneron Pharmaceuticals, Inc. Kiniksa OneConnect™ is a trademark of Kiniksa Pharmaceuticals. All other trademarks are the property of their respective owners.

Addressing Unmet Need is at the Heart of What We Do

Building on our successful foundation by prioritizing development of novel therapies for cardiovascular indications

Arcalyst® (rilonacept) For Injection

Established leadership in recurrent pericarditis market

Continued growth potential with only **~23% penetration** into multiple recurrence population[1]

Expected 2026 ARCALYST revenue of **$1.045 to $1.050B**

Advancing Clinical Portfolio

Conducting **KPL-387 Phase 2/3 trial** in **recurrent pericarditis**

Pivotal Phase 3 trial, PASTORALE, enrolling and dosing patients; data expected in **2H 2027**

KPL-1161 Phase 1 first-in-human trial on track to initiate by **YE 2026**

Maintaining Strong Financial Position

Q3 2026 cash reserves of ~$569M, representing ~$43M of net cash generation for the period

Financial strength provides capacity to **continue investing** in additional **value creation**

KINIKSA

1) As of Q3 2026.

Innovative Portfolio of Commercial and Clinical-Stage Assets

Developing novel therapies for diseases with unmet need, prioritizing cardiovascular indications

Program	Indication	Preclinical	Phase 1	Phase 2	Phase 3	Commercial
SPECIALTY CARDIOVASCULAR						
ARCALYST® (rilonacept)[1-3] IL-1α & IL-1β Trap	***Recurrent Pericarditis***	■	■	■	■	■
	Cardiac Sarcoidosis	*Collaborative Study Agreement with Mayo Clinic & The Johns Hopkins University*				
KPL-387[4] IL-1 Antagonist mAb	***Recurrent Pericarditis***	■	■	■		
KPL-1161 Fc-Modified IL-1 Antagonist mAb	***Undisclosed***	■				
OTHER (NON-CARDIOVASCULAR)						
Abiprubart Anti-CD40 mAb	***Exploring Strategic Alternatives***	■	■	■		

Program	Licensee	Exclusive Licensed Territory
OUT-LICENSING AGREEMENTS		
ARCALYST (rilonacept) IL-1α & IL-1β Trap	***Huadong Medicine***	*Asia Pacific Region, Excluding Japan*
Vixarelimab Anti-OSMRβ mAb	***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 Drug 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) The FDA granted Orphan Drug Designation to KPL-387 for the treatment of pericarditis in 2025.
IL-1α = interleukin-1α; IL-1β = interleukin-1β; IL-1 = interleukin-1; mAb = monoclonal antibody; OSMRβ = oncostatin M receptor beta

ARCALYST®

Arcalyst® (rilonacept) For Injection

IL-1α AND IL-1β CYTOKINE TRAP

DISEASE AREA: Recurrent pericarditis[1]; painful and debilitating autoinflammatory 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

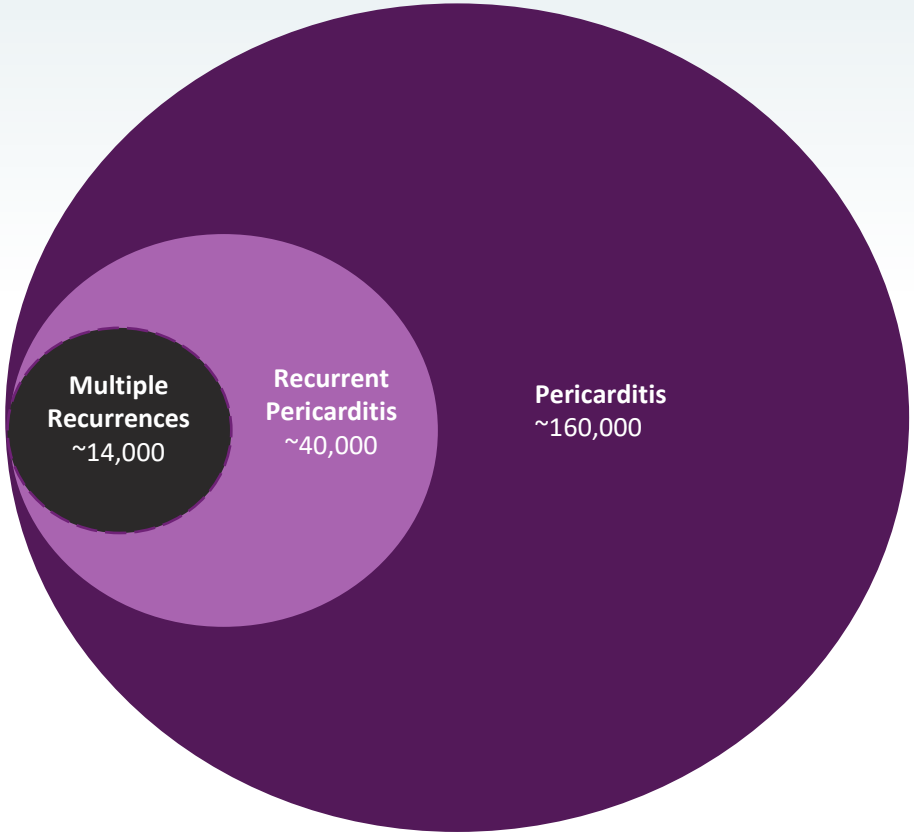
RIGHTS: Kiniksa has worldwide rights[3] (excluding MENA) for all indications outside those in oncology and local administration to the eye or ear

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 Patient Turnover Creates Steady Opportunity for Treatment

Of the 14,000 annual patients 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

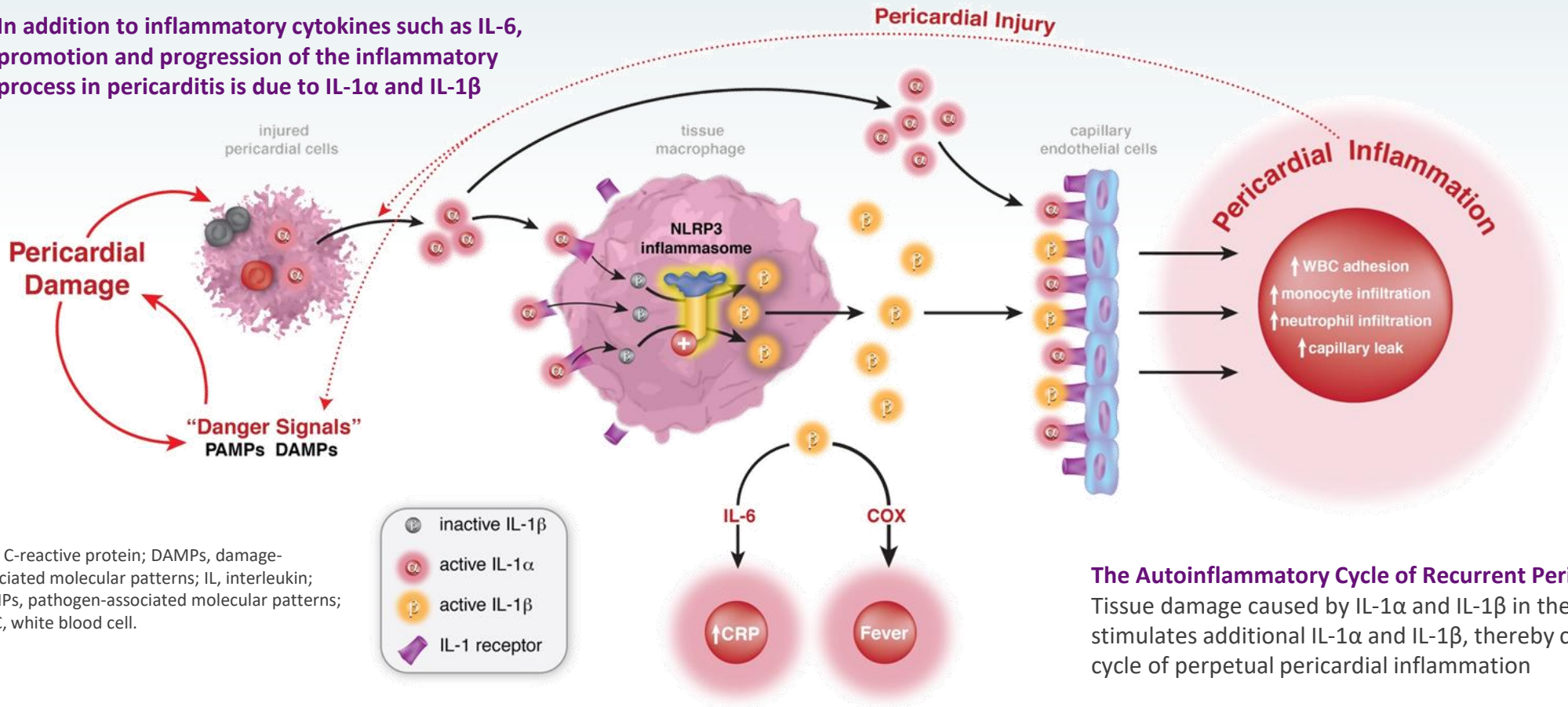
Approximately 14,000 recurrent pericarditis patients in the U.S. suffer from persistent underlying disease, with multiple recurrences and inadequate response to conventional therapy[1]

- **~160,000:** Epidemiological analysis using large national surveillance databases to calculate the pooled annualized prevalence of pericarditis ***(Basis for Orphan Drug Designation)[2]***
- **~40,000:** Up to 30% experience at least one recurrence; some recur over multiple years[3,4]
- **~14,000:** Nearly 50% annual turnover with ~7,000 patients entering into the pool each year[5]

1) Cremer et al. American Journal of Cardiology. 2016;2311-2328; 2) Data on file, Kiniksa Pharmaceuticals; 3) Imazio et al. Circulation. 2005;112:2012-2016; 4) Adler et al. Circulation. 1998;97:2183-2185; 5) 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.

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

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



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

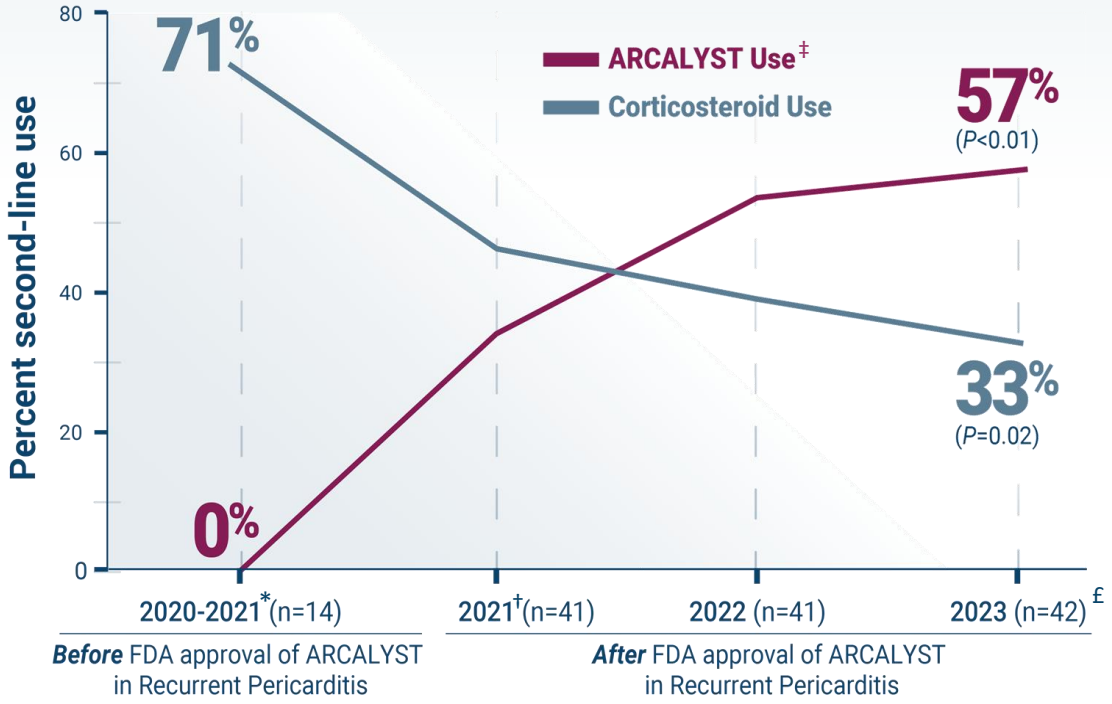
KINIKSA

ARCALYST is Evolving the Treatment Landscape for Recurrent Pericarditis

In the three years following approval, ARCALYST increasingly replaced corticosteroids after NSAIDs and colchicine

RESONANCE RWE: Expert Centers in U.S.

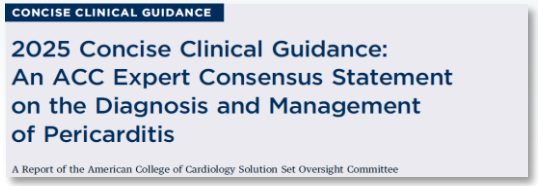
Second-Line use in patients with Recurrent Pericarditis failing NSAIDs/Aspirin/Colchicine[1]



ARCALYST has increasingly replaced corticosteroids in second-line use

ACC Concise Clinical Guidance: First U.S. Formal Guidance

IL-1 pathway inhibition is now the ACC-recommended second-line treatment after initial therapy



Updated Treatment Algorithm

Initial therapy
NSAIDs or aspirin plus colchicine

No Response
(autoinflammatory phenotype)*

Second-line
IL-1 pathway inhibition (e.g., ARCALYST)

Consider corticosteroid

Refractory Cases

Consider radical pericardiectomy
(at high-volume surgical center)

Adapted from Wang TKM, et al. *J Am Coll Cardiol. 2025*
*Autoinflammatory phenotype is defined as patients having fever and/or elevation of CRP and/or CMR imaging evidence of pericardial inflammation.

‡Of 64 patients starting ARCALYST after aspirin/NSAIDs/colchicine, 8 patients utilized steroids as a short-term bridge prior to starting ARCALYST (n=1 in 2021, n=2 in 2022, n=1 in 2023); 8 patients (n=2 in 2021, n=4 in 2023) utilized anakinra as a short-term bridge prior to starting ARCALYST; . £Of those who transitioned from aspirin/NSAIDs/colchicine to second-line ARCALYST, 5% subsequently used corticosteroids for >30 days and <5% subsequently used corticosteroids for <30 days; *1 year prior to ARCALYST approval; †Partial year after ARCALYST commercial availability April 1, 2021 – Dec 31, 2021; £Data were censored at last check-in visit.

1) Cremer, P, Luis, S, Garshick, M. et al. IL-1 Pathway Inhibition in Recurrent Pericarditis Management: Real-World Adoption of Corticosteroid Sparing in RESONANCE. JACC Adv. 2025 Sep, 4 (9). https://doi.org/10.1016/j.jacadv.2025.102050.

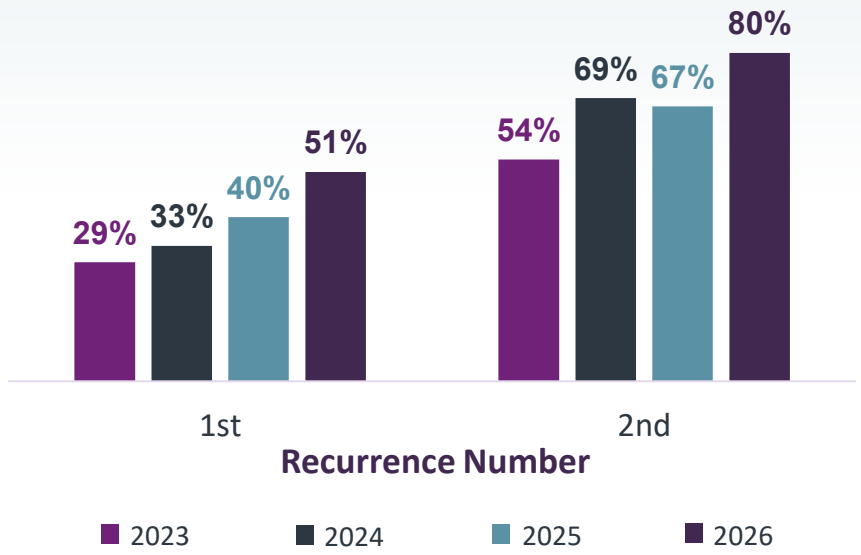
RWE = Real World Evidence; ACC = American College of Cardiology

Key Executional Priorities to Drive Greater Patient and Physician Adoption

- Drive physician awareness of the 2025 **ACC Concise Clinical Guidance**
- Advance direct-to-patient **digital marketing**
- Leverage AI and machine learning to **efficiently target potential prescribers**
- Support creation of an **efficient network of care** with **Pericardial Disease Centers (PDCs)**

% of Physicians Considering ARCALYST by Recurrence[1]

(n= 200 Cardiologists / Rheumatologists)

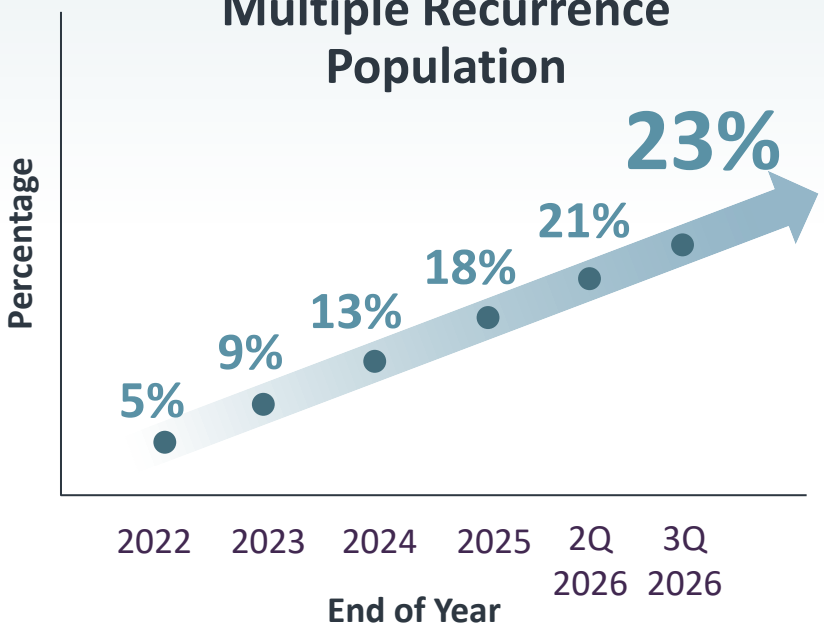


Positive trend in physicians considering ARCALYST earlier in the disease course, including on 1st recurrence

1) HCP market research 2026; Kiniksa data on file
ACC = American College of Cardiology; HCP = Healthcare Professionals

Driving Penetration in Initial Target Population with Upside Opportunity

Increasing Penetration into Multiple Recurrence Population



ARCALYST Label Covers Recurrent Pericarditis

(Annual Epidemiology of Approximately 40,000)

1st Recurrence
~26,000

A sizable portion could be appropriate for biologic treatment

Arcalyst® (rilonacept) For Injection

Additional Opportunity

~20% of patients on ARCALYST were prescribed when they were on their 1st recurrence[1]

≥2nd Recurrence
~14,000

Initial Target Population

~80% of patients on ARCALYST were prescribed when they had 2+ recurrences[1]

While the initial target population focused on patients with multiple recurrences...

...growing adoption of IL-1α & IL-1β inhibition has expanded focus to additional patients earlier in the disease course

Sources: Klein A, Cremer P, Kontzias A, Furqan M, Tubman R, Roy M, Magestro M. Annals of Epidemiology. 2019;36:71; 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.
1) HCP market research 2026; Kiniksa data on file.

Strong Commercial Fundamentals Support Continued ARCALYST Growth

$286.0M
Q3 2026 Net Revenue

*Representing ~**58%** YoY Growth*

>5,450[1]
Total Prescribers

*~**1,640** Repeat Prescribers*

Arcalyst® (rilonacept) For Injection

~3 Years[1]
Average Total Duration of Therapy
In Line with Median Duration of Disease

⅓ of Multiple Recurrence Patients Continue Suffering at 5 Years and ¼ at 8 Years

~23%[2]
Penetration into Multiple Recurrence Population

Increasing Utilization in 1st Recurrence Population

Strong Payer Approval and Patient Compliance

KINIKSA

1) Data since launch through 9/30/2026; 2) as of Q3 2026.

Heart's Home™ DTC Campaign is Showing Strong Signals of Success







Market Research Findings[1]

- **~20%** of patients recall seeing the ARCALYST ad since its launch
- **>50%** of patients seeing the ad indicated they visited the brand website for more information
- **~55%** of patients agreed strongly that the ad made them want to speak with their doctor about ARCALYST

Website Activity Improvements[2]

- **12x** increase in weekly visits to ARCALYST.com relative to pre-DTC period
- **8x** increase in engaged visits where visitor spends more time and/or completes a key site action
- Increases in site traffic observed as new channel partners added to campaign and spend scaled

ARCALYST is prescribed ~80% of the time when requested by a patient[3]

1) Patient market research; Kiniksa data on file; 2) Google analytics data; 3) Kiniksa data on file (HCP survey).

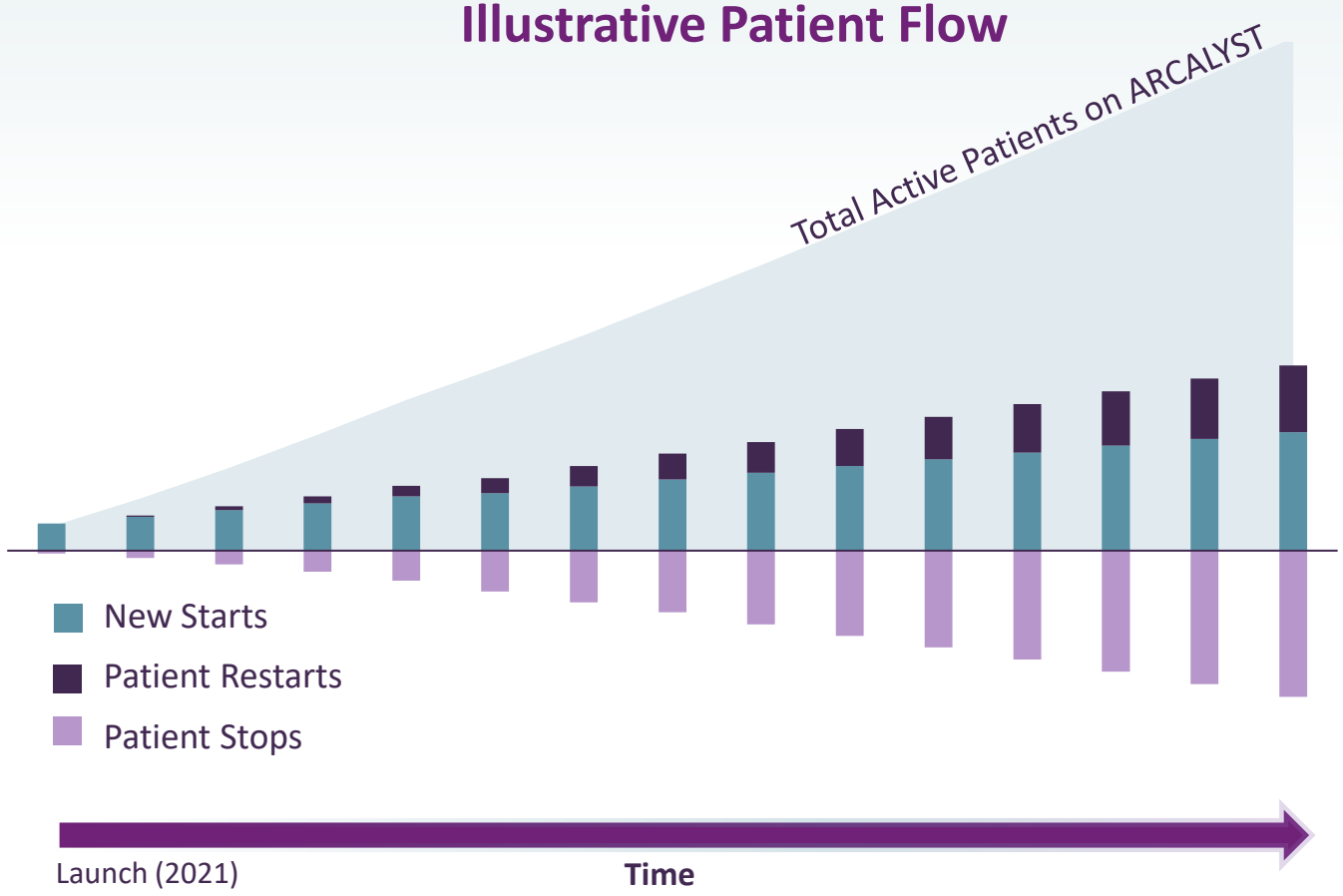
Growth in Total Patients on ARCALYST Therapy

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

ARCALYST Patient Flow

New to Brand Patients	Strong steady growth: >5,450 unique prescribers; ~30% 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 Q3 2026, ~23% of 14K multiple recurrence patient population

Illustrative Patient Flow



ARCALYST Enables Effective Treatment Throughout Duration of Disease

Patients with multiple recurrences often continue suffering from flares for a median of 3 years[1]

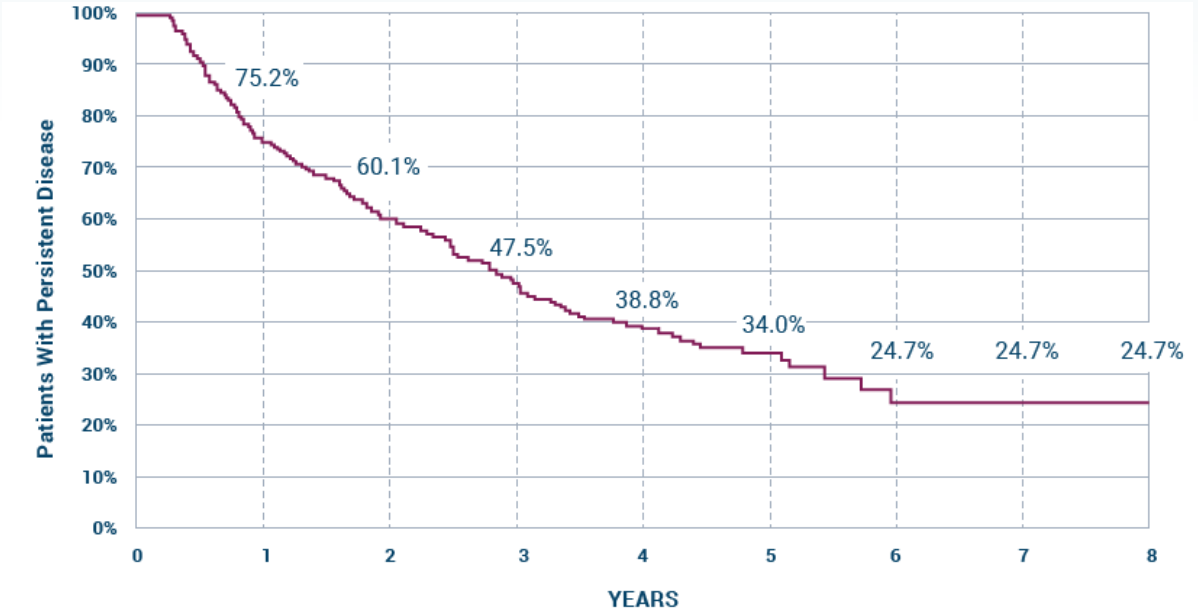
Average Total Duration of Therapy

~3 Years[2]

In Line with Median Duration of Disease

Natural History of Patients with Multiple Recurrences

⅓ of Patients with Multiple Recurrences Suffer at 5 Years and ¼ Continue to Experience Flares at 8 Years[1]

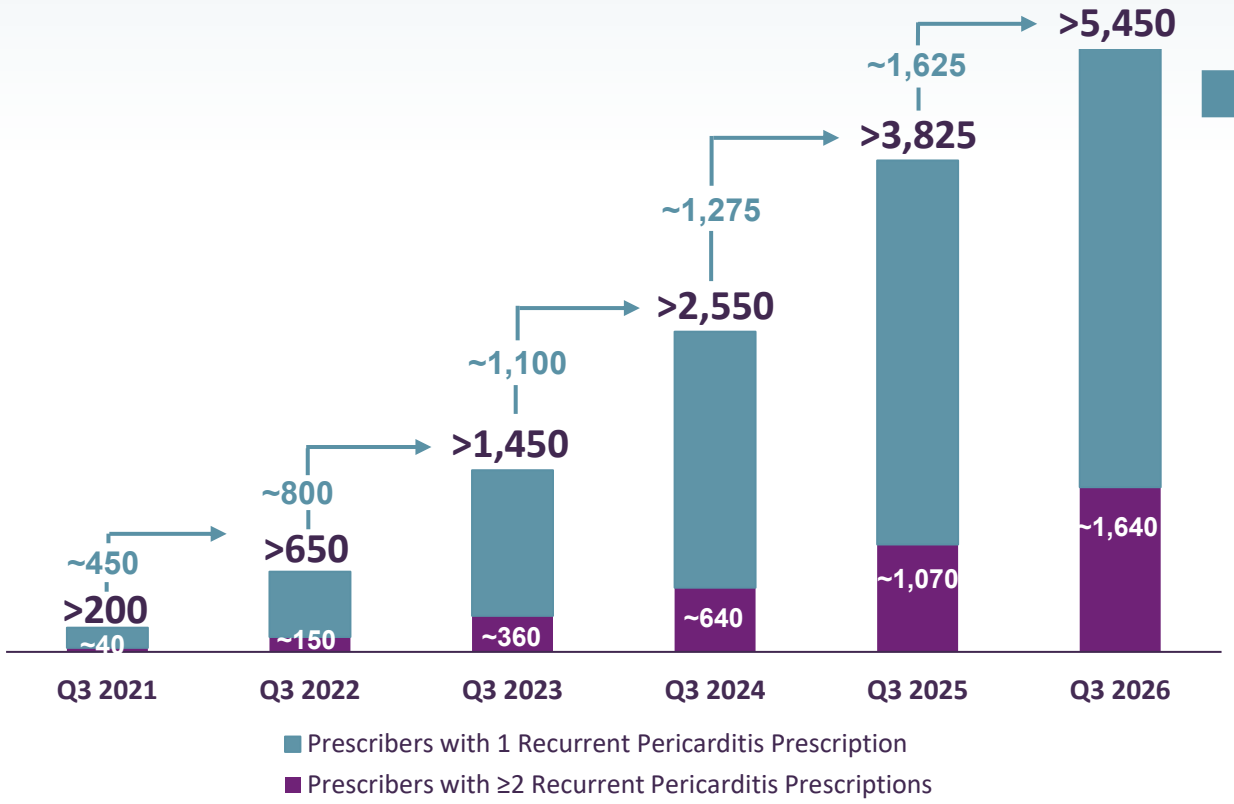


Data from Optum Health Care Solutions, Inc., collected from January 1, 2007, through March 31, 2017, were analyzed for this observational study (N=375 patients with ≥2 recurrences of RP).

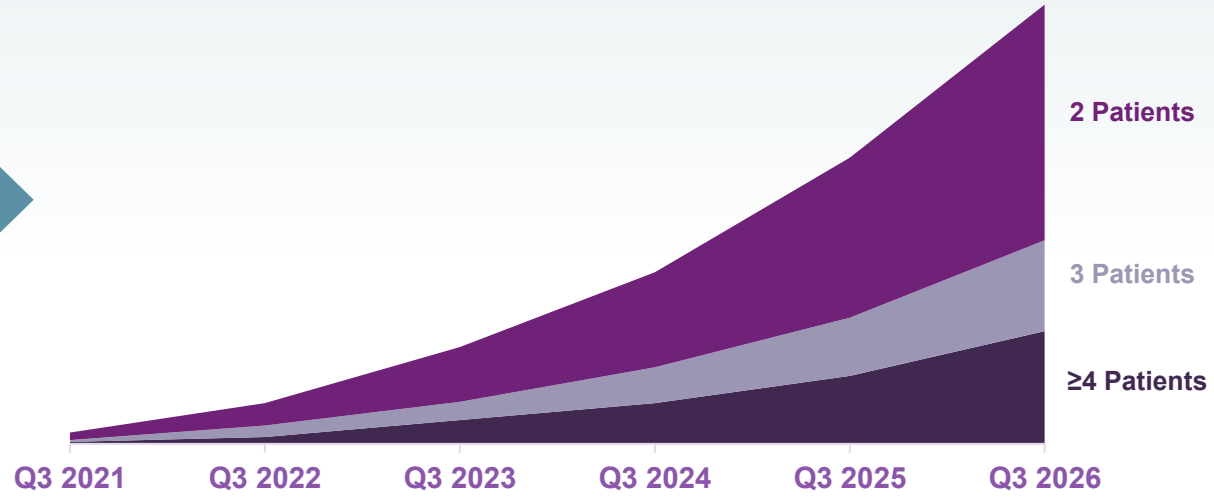
1) Lin D, Laliberté F, Majeski C, et al. Disease and economic burden associated with recurrent pericarditis in a privately insured United States population. Adv Ther. 2021;38(10):5127-5143. doi:10.1007/s12325-021-01868-7; 2) As of Q3 2026.

Expanding Breadth and Depth of ARCALYST Prescribing

Total and Repeat Prescribers of ARCALYST for Recurrent Pericarditis Patients



The Growing Repeat Prescriber Base is Delivering >50% of All New Patient Prescriptions



- Strong, steady growth in **both new and repeat prescribers**, supporting long-term growth-potential
- Both physicians and patients are gaining **positive experiences with ARCALYST** as the first and only approved therapy for recurrent pericarditis
- Cardiologist market research shows a steady **increase in their level of comfort with prescribing biologics**
- **>50% of all new prescriptions in Q3 2026 came from repeat prescribers**

KINIKSA

Pricing, Access, and Distribution Considerations

Pricing

- ARCALYST list price of $25,158 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 (>60%)**
- 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

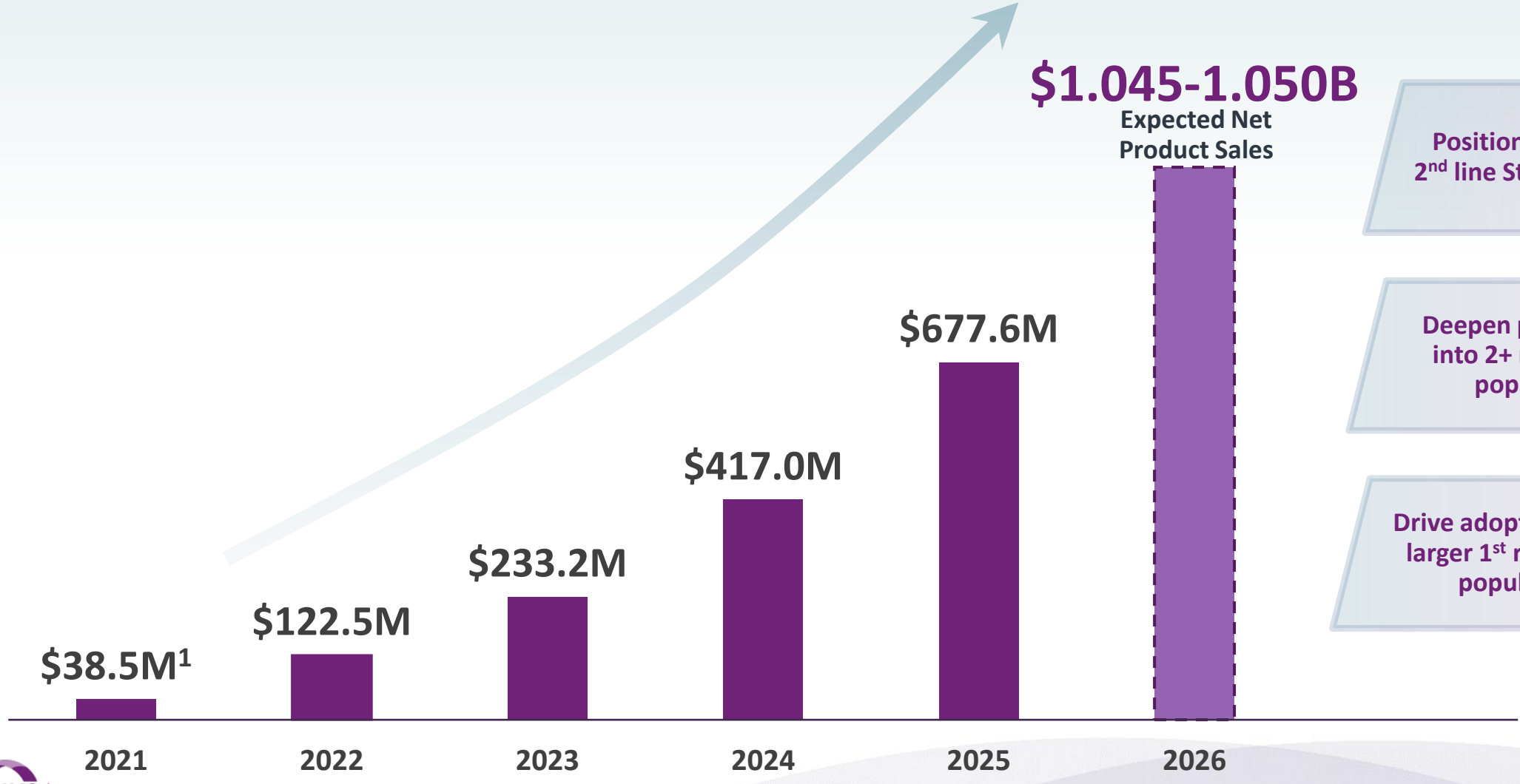
Distribution

- 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

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

2026 ARCALYST Net Product Sales Guidance

Revenue guidance raised to $1.045-$1.050B from $980-$995M



- Position ARCALYST as 2nd line Standard of Care
- Deepen penetration into 2+ recurrence population
- Drive adoption among larger 1st recurrence population

1) 2021 = 9 months of availability (Q2-Q4).

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.

*Kiniksa exclusively licensed rights for the development and commercialization of ARCALYST in APAC (ex-Japan) to Huadong Medicine

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 = total cost of goods sold - amortization of Regeneron milestone payment.

CAPS = Cryopyrin-Associated Periodic Syndromes; DIRA = Deficiency of the Interleukin-1 Receptor Antagonist; MENA =Middle East and North Africa; APAC = Asia Pacific Region

KPL-387

MONOCLONAL ANTIBODY IL-1 RECEPTOR ANTAGONIST INHIBITING IL-1α AND IL-1β SIGNALING

DISEASE AREA: Recurrent pericarditis; painful and debilitating autoinflammatory cardiovascular disease

SCIENTIFIC RATIONALE: Inhibition of IL-1α and IL-1β signaling well-established for the treatment for recurrent pericarditis[1-3]

REGULATORY: U.S. Orphan Drug Designation for the treatment of pericarditis

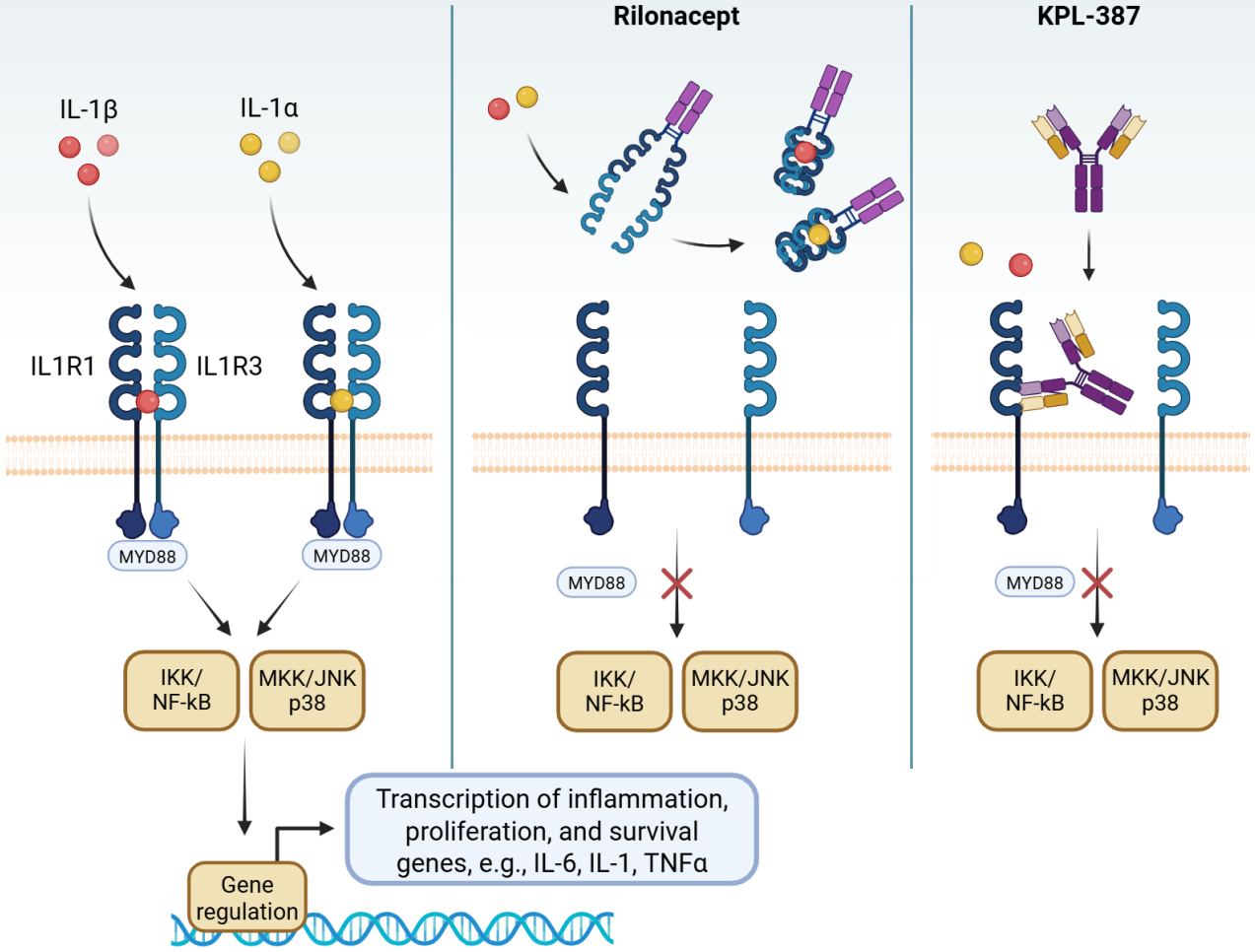
STATUS: PASTORALE, pivotal Phase 3 portion of the Phase 2/3 clinical trial, enrolling and dosing patients; pivotal (Phase 3) data expected in 2H 2027

ECONOMICS: Independently developed and wholly-owned

RIGHTS: All indications worldwide

1) Klein AL, Imazio M, Cremer P, et al. Phase 3 trial of interleukin-1 trap rilonacept in recurrent pericarditis. N Engl J Med. 2021;384(1):31-41; ARCALYST (rilonacept) prescribing information 2021; 2) Arnold, D. D., Yalamanoglu, A., & Boyman, O. (2022). Systematic review of safety and efficacy of IL-1-targeted biologics in treating immune-mediated disorders. Frontiers in immunology, 13, 888392; 3) Dinarello CA, Simon A, van der Meer JWM. Treating inflammation by blocking interleukin-1 in a broad spectrum of diseases. Nat rev drug Discov, 2012;11(8);633-652.

KPL-387: Independently Developed IL-1 Receptor Antagonist for the Treatment of Recurrent Pericarditis



Created in BioRender. Lawton, M. (2025) https://BioRender.com/hvx08bv

KPL-387

- Fully human IgG2 **monoclonal antibody**
- **Binds to IL-1R1**, inhibiting both IL-1α & IL-1β cytokine-mediated signaling
- **IL-1α and IL-1β inhibition is well-established** and **well-tolerated**
- **Monthly** dosing potential with **single subcutaneous self-injection** in **liquid formulation**

IL-1R1 = interleukin-1 receptor 1; IL-1R3 = interleukin-1 receptor 3; IL-1α = interleukin-1 alpha; IL-1β = interleukin-1 beta; IgG2 = immunoglobulin G2; MYD88 = myeloid differentiation primary response 88; IKK = IkappaB kinase; NF-κB = nuclear factor-kappa B; MKK = mitogen-activated protein kinase kinase; JNK = jun N-terminal kinase; p38 = p38 mitogen-activated protein kinase; IL-6 = interleukin 6; TNFα = tumor necrosis factor-alpha

KPL-387 Program Aims to Address Key Patient Needs and Expand Market

The **vast majority** of surveyed HCPs report that an efficacious IL-1α & IL-1β inhibitor with the **target profile of KPL-387** would be **best positioned** to **address unmet needs** of patients living with recurrent pericarditis and is likely to **expand the market**

Key Needs Filled

- **Less frequent dosing**
- **Streamlined preparation**
- **Patient-friendly administration**

Patient Preferences

~75% Of all RP patients prefer the KPL-387 target profile over available **commercial and investigational therapies**

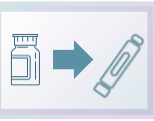
~70% Of all RP patients would be willing to stay on a monthly autoinjector for **longer, with fewer missed doses**, compared to a weekly subcutaneous dosing presentation

~75% Of ARCALYST-naïve patients would be more willing to take an injectable therapy **if presented in an autoinjector**

*"It [would be] **easy to use** because there is no need to mix and wait for it to work. The autoinjector [would] make it simple. It [would] also be **more convenient to take it once a month.**" – ARCALYST patient*

HCP Preferences

~92% Report high likelihood of prescribing KPL-387 for **new patients**, in the context of available commercial and investigational therapies



Current ARCALYST patients demonstrate high compliance and adherence, but HCPs receptive to switching **upon patient request**



HCPs indicate a sizable **increase in proportion of patients likely to use an IL-1α & β inhibitor** if KPL-387 comes to market

*"The dosing regimen of once monthly versus once weekly is a **game changer** for patients." – Physician*

Source: Kiniksa data on file.

Registrational Program Includes Pivotal and Supplemental Components

KPL-387 Development Program

	Phase	Study Design & Type	Patient Population	Treatment Duration
Pivotal Study	***Phase 3***	Event-Driven, Double-Blind, Placebo-Controlled, Randomized-Withdrawal Study[1]	Qualifying Pericarditis Episode	Event-Driven
Supplemental Studies	***Phase 1***	SAD/MAD Study	Healthy Participants	Single Dose & 12 Weeks (MAD)
	Phase 2	Dose-Focusing Study[1]	Qualifying Pericarditis Episode	24 Weeks
		Transition to KPL-387 Monotherapy Dosing & Administration Study[2]	Well-Controlled Recurrent Pericarditis[3]	16 Weeks
	LTEs	Eligible Patients Completing Phase 2 Dose-Focusing Study[1]		Up to 24 Months Additional Treatment[4]
		Eligible Patients Completing Phase 2 Transition to KPL-387 Monotherapy Dosing & Administration Study		Up to 24 months Additional Treatment[4]
		Eligible Patients Completing Phase 3 Pivotal Study[1]		Up to 24 Months Additional Treatment[4]

1) NCT07010159; 2) Supplemental study evaluating the efficacy/safety of dosing regimens used to transition patients with well-controlled RP to KPL-387 monotherapy from stable prior treatment with standard therapies; 3) No recurrence within 3 months prior to baseline; CRP < 0.5 mg/dL within 14 days of Baseline and NRS ≤ 3 at Baseline; no clinical worsening or suspicion of impending recurrence; 4) Up to 24 months or the time KPL-387 is approved for commercial use in that region to treat recurrent pericarditis.

LTE = long-term extension; SAD = single ascending dose; MAD = multiple ascending dose

KPL-387 Development Program Builds on Successful RHAPSODY Design

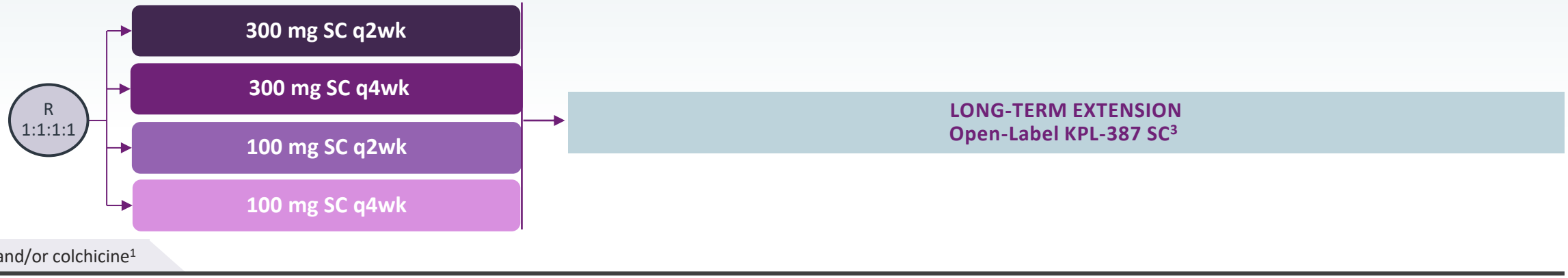
Phase 2/3 recurrent pericarditis clinical trial

Dose-Focusing

Population:
- Up to ~80 patients diagnosed with RP presenting at screening with a qualifying pericarditis episode despite treatment with conventional oral therapies.

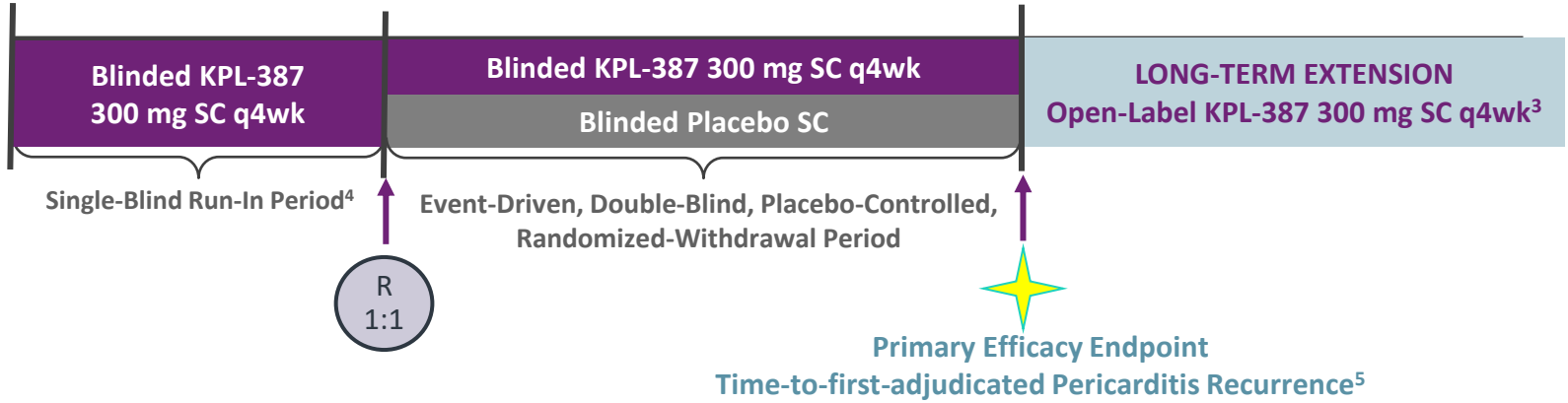
Primary Efficacy Endpoint
- Time to Treatment Response[2]

Key Secondary Endpoints
- Time to Pain Response
- Time to CRP normalization (CRP ≤ 0.5 mg/dL)



Pivotal

Population:
- Up to ~85 patients diagnosed with RP presenting at screening with a qualifying pericarditis episode despite treatment with conventional oral therapies.



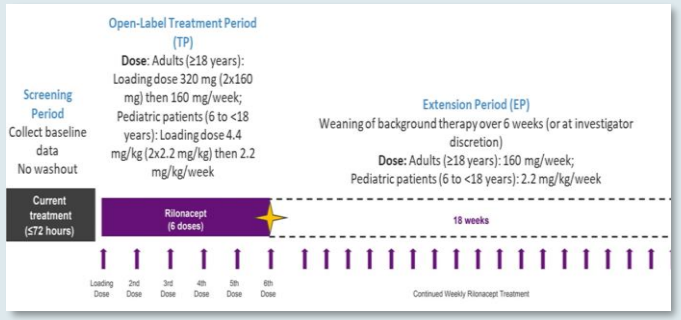
1) KPL-387 will be administered in addition to conventional oral pericarditis medications (NSAIDs and/or colchicine) from baseline to Week 1, and then oral medications will be weaned off to achieve KPL-387 monotherapy by Week 2. Participants previously treated with glucocorticoids must have discontinued their use at least 72 hours prior to first study drug administration; 2) Treatment Response is defined as Pain Response (NRS score ≤ 2 on the 11-point daily pericarditis pain NRS) and at least one CRP level ≤ 0.5 mg/dL within 7 days before or after the Pain Response; 3) Up to 24 months or the time KPL-387 is approved for commercial use in that region to treat recurrent pericarditis; 4) Duration of the run-in period undisclosed in order to maintain study subjects blinded to the start of the randomized-withdrawal period; 5) Time to Pericarditis Recurrence is defined as the time from randomization in the Randomized Withdrawal (RW) Period to the date of the first Pericarditis Recurrence for each participant. Only CEC-confirmed Pericarditis Recurrences will be considered as events for the primary efficacy analysis in the pivotal portion.
NSAID = non-steroidal anti-inflammatory drug; RP = recurrent pericarditis; CRP = C-reactive protein; NRS = numerical rating scale (for chest pain); R = randomization; SC = subcutaneous

Dose-Focusing Portion Builds on Precedent of Rilonacept Phase 2 Trial

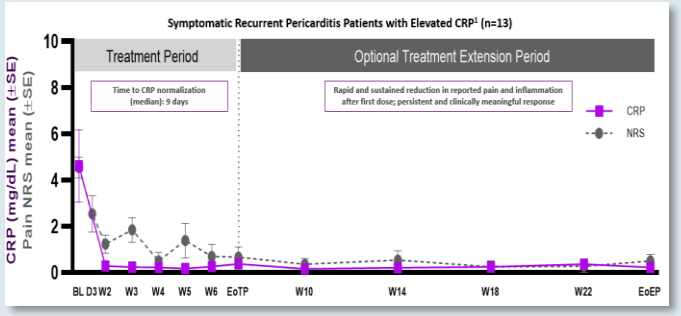
Cadence & magnitude of initial response and duration of action in Phase 2 inform Phase 3 efficacy, recurrence risk reduction

Open-Label Phase 2 Clinical Trial of Rilonacept in Pericarditis Populations[1]

(n=13)



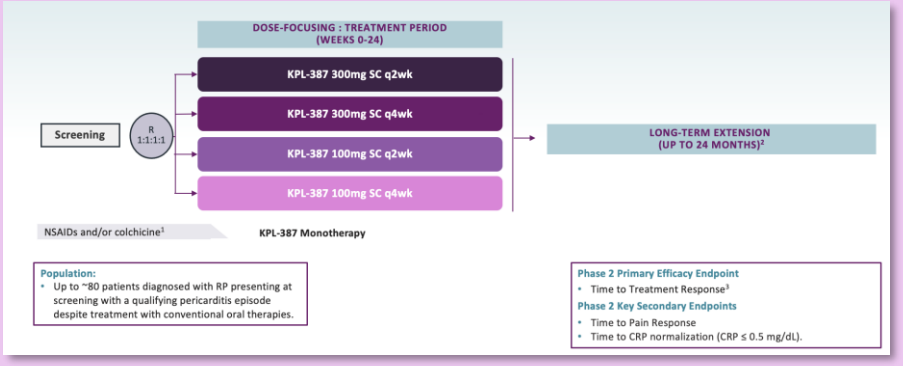
Test of treatment response and durability: Addition of rilonacept in patients failing standard oral therapies



IL-1 pathway inhibition with rilonacept resulted in rapid & sustained reduction in reported pain & inflammation after first dose; persistent & clinically meaningful response[2]

Phase 2 Dose-Focusing Portion of KPL-387 Phase 2/3 Clinical Trial[3]

(n= up to ~20/arm)



Anchoring on prior rilonacept clinical trial experience, the dose-focusing portion assesses 4 dose levels for cadence/magnitude of initial response and durability, informing PK/PD relationship and Phase 3 dose confirmation

1) NCT03980522; 2) Klein AL, Lin D, Cremer PC, Nasir S, Luis SA, Abbate A, Ertel A, LeWinter M, Beutler A, Fang F, Paolini JF. Efficacy and safety of rilonacept for recurrent pericarditis: results from a phase II clinical trial. *Heart*. 2020 Nov 23;107(6):488–96; 3) NCT07010159.

PK = pharmacokinetic; PD = pharmacodynamic

Phase 2 Data Supported Selection of KPL-387 300 mg SC q4wk Dose Level for Pivotal Phase 3 Study

Primary and Secondary Endpoints*	KPL-387 300 mg SC q4wk
Time to Treatment Response[1]	4.0 (3.0, 6.0) days
Time to Pain Response	4.0 (3.0, 6.0) days
Time to CRP Normalization[2]	8.0 (7.0, 9.0) days

*Reported as median (95% CI)

Participants in the 300 mg SC q4wk dose group experienced rapid and sustained Treatment Response[1], with reductions in NRS pain and inflammation[2] as soon as after the first dose

KPL-387 efficacy was durable throughout the monthly dosing interval

Treatment effects were consistent with those seen in prior studies of rilonacept in recurrent pericarditis**

KPL-387 was generally well-tolerated, consistent with the well-known safety profile IL-1 pathway inhibition

Phase 2 data presented are based on an interval analysis of the ongoing Phase 2 study and are subject to change following study completion, final data cleaning, and completion of analyses. Final results may differ from those presented herein.

**Cross-trial comparisons may not be reliable due to differences in study protocol, patient populations, and other factors.

1) Treatment Response is defined as Pain Response (NRS score ≤ 2 on the 11-point daily pericarditis NRS pain scale) and at least one CRP level ≤ 0.5 mg/dL within 7 days before or after the day of Pain Response; 2) CRP normalization defined as CRP ≤ 0.5 mg/dL.

SC = subcutaneous; q4wk = every four weeks; NRS = Numeric Rating Scale; CRP = C-reactive protein

Financials
Third Quarter 2026

Third Quarter 2026 Financial Results

Income Statement	Three Months Ended September 30,	
	2026	2025
Product Revenue	$286.0M	$180.9M
License and Collaboration Revenue	$0.0M	$0.0M
Total Revenue	**$286.0M**	**$180.9M**
Cost of Goods Sold	$27.8M	$20.3M
Collaboration Expenses[1]	$105.8M	$63.3M
Research and Development	$32.8M	$24.2M
Selling, General and Administrative	$65.4M	$49.1M
Total Operating Expenses	**$231.7M**	**$156.8M**
Operating Income	**$54.2M**	**$24.0M**
Other Income, Net	$4.8M	$3.1M
Income Tax Provision	($11.7M)	($8.7M)
Net Income	**$47.3M**	**$18.4M**

Collaboration Expenses[1]	Three Months Ended September 30,	
	2026	2025
ARCALYST Net Sales	**$286.0M**	**$180.9M**
Profit Split-Eligible Cost of Goods Sold	($26.2M)	($20.0M)
Commercial, Marketing, Regulatory and Other Expenses	($48.2M)	($34.3M)
ARCALYST Collaboration Operating Profit	**$211.6M**	**$126.6M**
ARCALYST Collaboration Expense	$105.8M	$63.3M
ARCALYST Out-Licensing[2]	$0.0M	$0.0M
Other Collaboration Expenses	$0.0M	$0.0M
Total Collaboration Expenses	**$105.8M**	**$63.3M**

Balance Sheet	September 30, 2026	December 31, 2025
Cash, Cash Equivalents and Short-term Investments	**$568.8M**	**$414.1M**

Full Year 2026 Financial Guidance

ARCALYST Net Revenue Guidance: $1.045-$1.050B
R&D Expense Guidance: $160-$170M
SG&A Expense Guidance: $270-$280M

Figures represented to the nearest tenth; totals may not sum exactly due to rounding.
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) Revenue associated with ARCALYST Out-Licensing is included in License and Collaboration Revenue.

KINIKSA

Appendix Out-Licensing Agreements

Out-Licensing Agreements

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

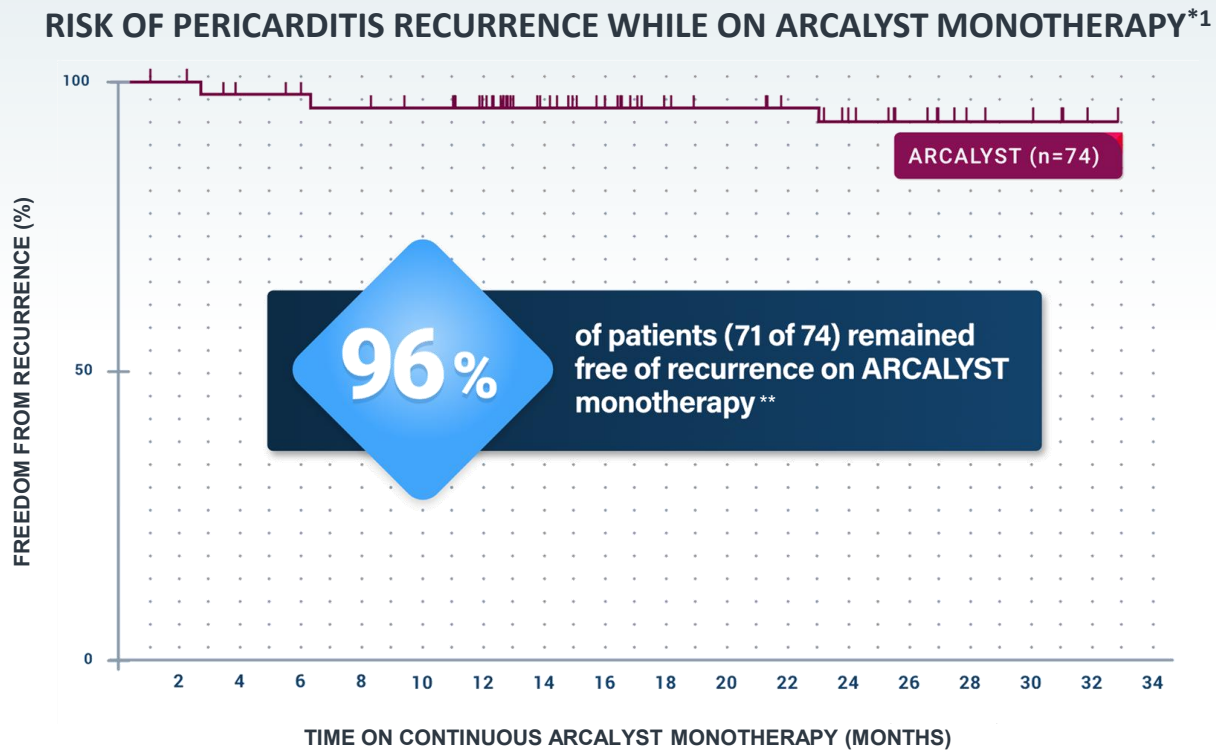
- In February 2022, Kiniksa announced a strategic collaboration with Huadong to develop and commercialize ARCALYST in Greater China and multiple other countries in the Asia Pacific region, excluding Japan
- In January 2025, Kiniksa received a $20M milestone payment, $10M of which flows through to Regeneron, for the approval of ARCALYST in mainland China
- Kiniksa remains eligible to receive specified sales-based milestones along with tiered royalty payments

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 $570 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

Appendix
ARCALYST (rilonacept)

ARCALYST Monotherapy Provided Long-Term Prevention of Recurrence Over Three-Year Duration of RHAPSODY



A new integrated analysis of RHAPSODY supports evidence-based use of ARCALYST as a monotherapy in long-term recurrent pericarditis management, minimizing need for adjunctive treatments and reducing polypharmacy

*Post hoc analysis. Freedom from recurrence was quantified from randomized withdrawal start (if randomized to ARCALYST) or from initiation of bailout ARCALYST (if randomized to placebo) until the end of treatment;
**Three observed adjudicated recurrences (3 of 74) were each associated with temporary interruptions of 1 to 3 doses of therapy.

1) Lotan, D, Imazio, M, Klein, AL, Abbate, A, Arad, M, Cremer, PC, Luis, SA, Wang, S, Curtis, A, Clair, J, Paolini, JF. Rilonacept as Monotherapy for Long-Term Recurrent pericarditis Management: Sustained Disease Control Over Three Years. Poster presented at 2025 European Society of Cardiology Congress. Madrid, Spain.

RW = randomized withdrawal; LTE = long-term extension

Real-World Outcomes Data Affirm Benefit of Sustained ARCALYST Treatment

Rilonacept provides superior clinical outcomes and reduced hospitalizations/ED visits versus 2nd-line glucocorticoids

Clinical Outcomes and Healthcare Resource Utilization in Patients With RP

				Annualized Recurrence Rate (per PY)			Hospitalizations (per 100 PY)			ED visits (per 100 PY)		
Therapy (n)		Median [SD] duration on 1L NSAIDs ± Colchicine, years	Median [SD] duration on 2L regimen, years	While on 1L NSAIDs ± Colchicine[a]	While on 2L regimen	Relative risk difference [P-value]	While on 1L NSAIDs ± Colchicine	While on 2L regimen	Relative risk difference [P-value]	While on 1L NSAIDs ± Colchicine	While on 2L regimen	Relative risk difference [P-value]
1st Line	**NSAIDs ± Colchicine*** (n=85)	1.5 [0.6, 2.8]	N/A	0.74[b]	N/A	N/A	11.2	N/A	N/A	39.2	N/A	N/A
2nd Line	**IL-1 Pathway Inhibition** (n=104)	2.1 [0.4, 4.5]	1.8 [0.8, 2.6]	1.41[c]	0.043[d]	– 97% [<0.001]	11.9	0	– 100% [0.02]	34.1	0.009	– 99.9% [0.01]
	Rilonacept (n=82)	1.4 [0.3, 3.8]	1.9 [0.9, 2.7]	1.52[e]	0.032[f]	– 97.9% [<0.001]	11.8	0	– 100% [0.018]	42.2	0	– 100% [0.008]
	Anakinra (n=22)	2.8 [0.1, 3.9]	1.7 [1.1, 2.4]	1.12[g]	0.18[h]	– 83.9% [<0.001]	12.2	0	– 100% [0.02]	21.4	0.021	– 99.9% [0.01]
	Glucocorticoids (n=87)	0.1 [0.08, 0.8]	0.6 [0.12, 0.9]	2.13[i]	3.3[j]	+ 54.9% [0.034]	16.4	22.7	+ 38.4% [<0.001]	65.5	153.4	+ 134.2% [0.006]
	csDMARD** (n=2)	0.5 [0.4, 0.5]	0.5 [0.4, 0.6]	3.33[k]	2[l]	– 39.9% [0.8]	0	0	N/A	0	0	N/A

*Patients remaining on NSAIDs ± Colchicine who did not escalate therapy to 2nd-line treatment; **2L csDMARDs included patients on azathioprine (n=1) and methotrexate (n=1); [a]Incident episode was managed with NSAIDs and/or colchicine and/or glucocorticoids; [b]11% of events confirmed by chest pain + RHAPSODY adjudication criteria; 7% did not meet RHAPSODY adjudication criteria; 81% had no CRP value ≤ 30 days of each event; [c]19% of events confirmed by chest pain + RHAPSODY adjudication criteria;12% did not meet RHAPSODY adjudication criteria; 70% had no CRP value ≤ 30 days of each event; [d]20% of events confirmed by chest pain + RHAPSODY adjudication criteria; 30% of events did not meet RHAPSODY adjudication criteria; 50% of events had no CRP value ≤ 30 days of each event; [e]21% of events confirmed based on chest pain + RHAPSODY adjudication criteria; 12% of events did not meet RHAPSODY adjudication criteria; 67% of events had no CRP value ≤ 30 days of each event; [f]17% of events confirmed by chest pain + RHAPSODY adjudication criteria; 50% of events did not meet RHAPSODY adjudication criteria; 33% of events had no CRP value ≤ 30 days of each event. 67% of events on rilonacept were associated with dose interval elongation; 33% of events were breakthrough but had no CRP values that met RHAPSODY adjudication criteria ≤ 30 days of chest pain; [g]15% of events confirmed by chest pain + RHAPSODY adjudication criteria; 11% of events did not meet RHAPSODY adjudication criteria; 74% of events did not have a CRP value ≤ 30 days of each event; [h]22% of events confirmed by chest pain + RHAPSODY adjudication criteria; 22% of events did not meet RHAPSODY adjudication criteria; 56% of events did not have a CRP value ≤ 30 days of each event. 56% of events were associated with dose interval elongation; 44% of events were breakthrough,75% of breakthrough events had CRP values that met RHAPSODY adjudication criteria ≤ 30 days of chest pain; [i]29% of events confirmed based on chest pain + RHAPSODY adjudication criteria; 27% of events did not meet RHAPSODY adjudication criteria; 44% of events had no CRP value ≤ 30 days of each event; [j]62% of events confirmed by chest pain + RHAPSODY adjudication criteria; 7% of events did not meet RHAPSODY adjudication criteria; 31% of events had no CRP value ≤ 30 days of each event. All 86% of events were associated with dose tapering; 14% of events were breakthrough, all breakthrough events had CRP values that met RHAPSODY adjudication criteria ≤ 30 days of chest pain; [k]33% of events confirmed by chest pain + RHAPSODY adjudication criteria;67% of events had no CRP value ≤ 30 days of each event; [l]100% of events confirmed by chest pain + RHAPSODY adjudication criteria. All events were breakthrough but had no CRP values that met RHAPSODY adjudication criteria ≤ 30 days of chest pain.

1) Cremer, PC, Garshick, M, Luis, SA, Raisinghani, A, Weber, B, Al-Kazaz, M, Hagerty, T, Ryan, JJ, Salik, JR, Corey, T, Parameswaran, V, Klein, AL, Paolini, JF. Rilonacept Reduces Pericarditis Recurrence Risk, Hospitalization, and ED Visits: Outcomes From the RESONANCE Patient Registry. Poster presented at 2026 American College of Cardiology Congress. New Orleans, LA USA.

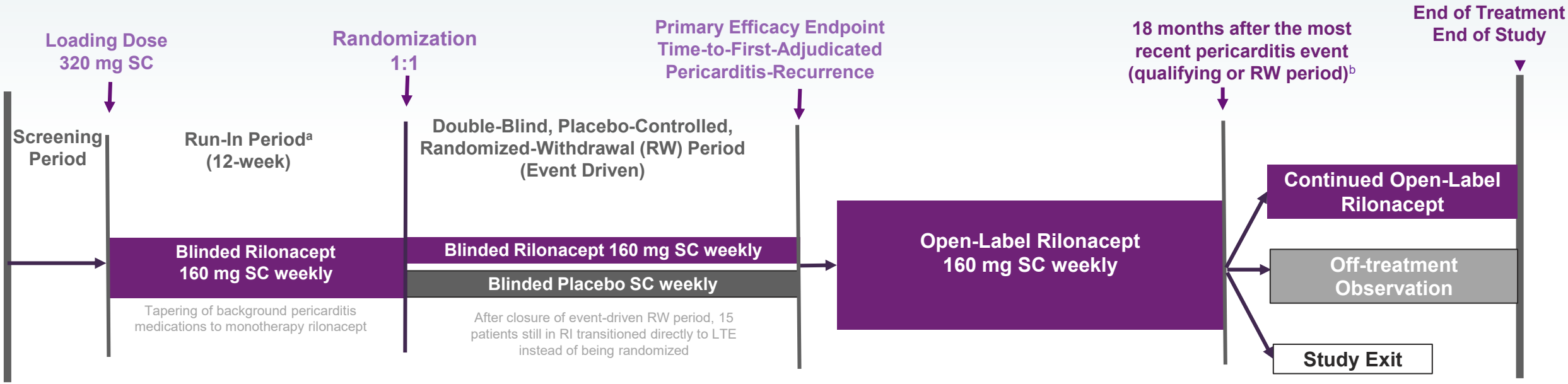
PY = patient-years; ED = emergency department; SD = standard deviation; NSAIDs = non-steroidal anti-inflammatory drugs; IL-1 = interleukin-1

RHAPSODY Design

Event-Driven Pivotal Study

Median rilonacept treatment duration prior to the LTE (RI+RW) was 9 months (range, 3-14)

Long-Term Extension (LTE) (up to 24 months)



[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)

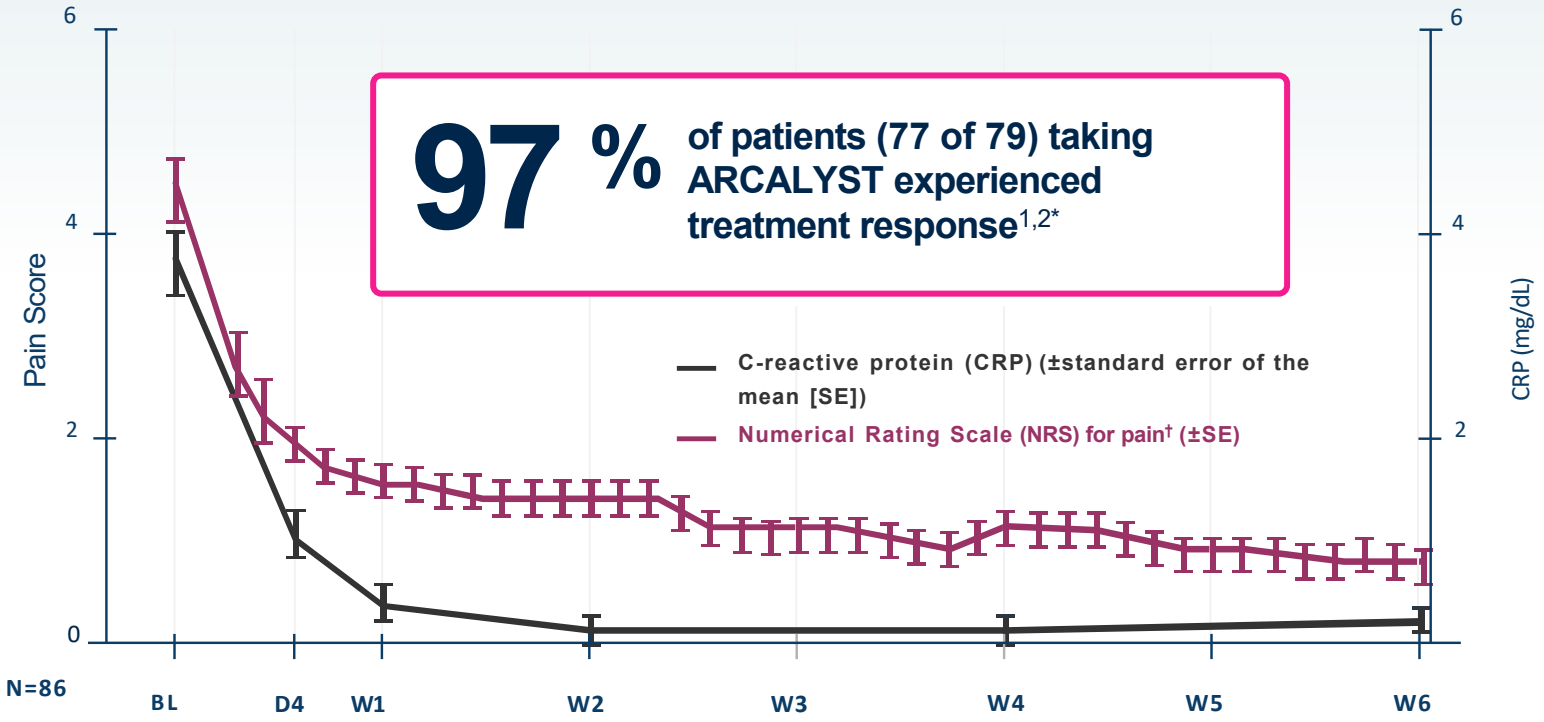
OR

- Discontinue the LTE completely (no further observation)

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)

Rilonacept Initiation Resulted in Rapid Resolution of Acute Pericarditis Episodes

RHAPSODY run-in period



Most patients experienced treatment response*, pain relief, and inflammation resolution as early as after the first dose[2]

Median time to **treatment response***

5.0 days

(95% CI: 4.0, 7.0)

Median time to **pain response**

5.0 days

(95% CI: 4.0, 6.0)

Median time to **CRP normalization**

7.0 days

(95% CI: 5.0, 8.0)

*Time to treatment response was defined as the time from the first dose to the first day when pericardial pain was NRS ≤2 (0-10) and CRP ≤0.5 mg/dL (measured within 7 days before or after the pain response).
†Ranging from 0 to 10, with higher scores indicating greater pain severity.

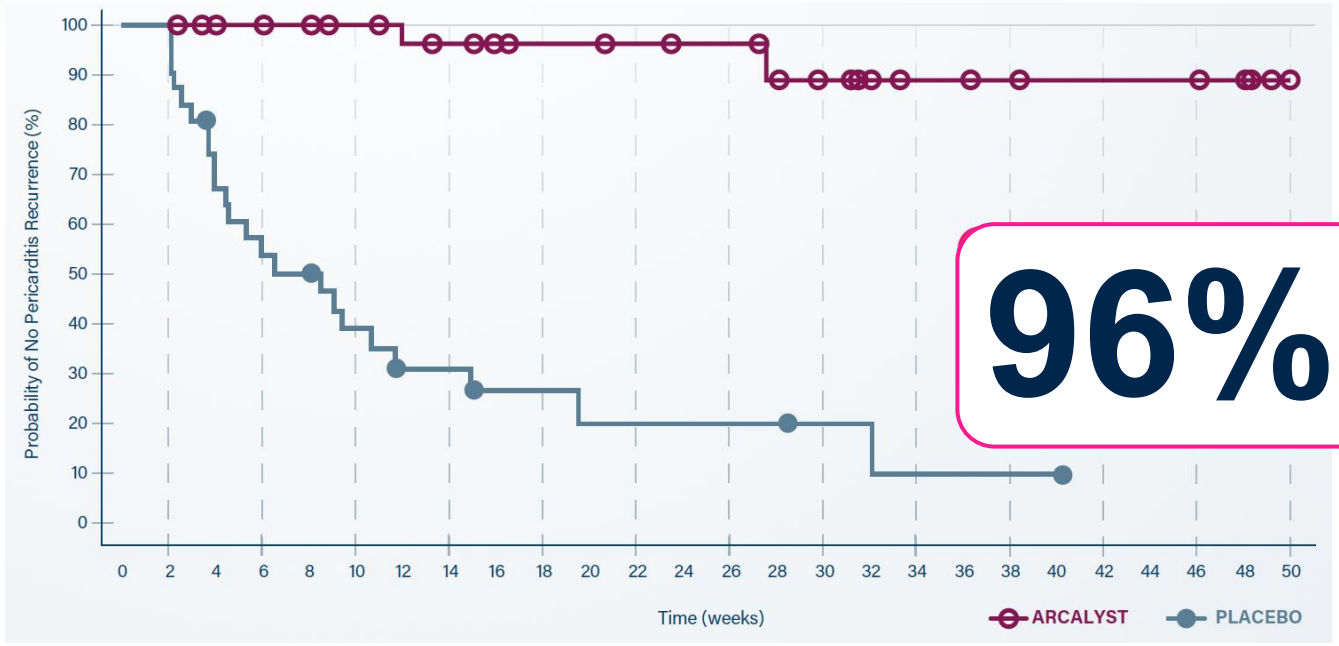
1) ARCALYST Package insert. Kiniksa Pharmaceuticals; 2021; 2) Klein AL, Imazio M, Cremer P, et al. Phase 3 trial of interleukin-1 trap rilonacept in recurrent pericarditis. N Engl J Med. 2021;384(1):31-41.

96% Reduction in Risk of Pericarditis Recurrence

Pivotal Phase 3 RHAPSODY Data

ARCALYST reduced the risk of pericarditis recurrence

The primary efficacy endpoint was time to first adjudicated pericarditis recurrence in the randomized withdrawal period.



The median time to recurrence on ARCALYST could not be estimated due to the low number of recurrences

- 2 of 30 of patients treated with ARCALYST had a recurrence
- The 2 pericarditis recurrences with ARCALYST occurred during temporary interruptions of 1 to 3 doses of ARCALYST

96%

reduction in the risk of recurrent pericarditis
(hazard ratio: 0.04; *p*<0.0001)

The median time to recurrence on placebo was 8.6 weeks (95% CI: 4.0, 11.7)

- 74% (23 of 31) of patients treated with placebo experienced a recurrence at the time that the event-driven portion of the trial was closed
- Consistent with the expected washout pharmacokinetics of once-weekly ARCALYST at steady state

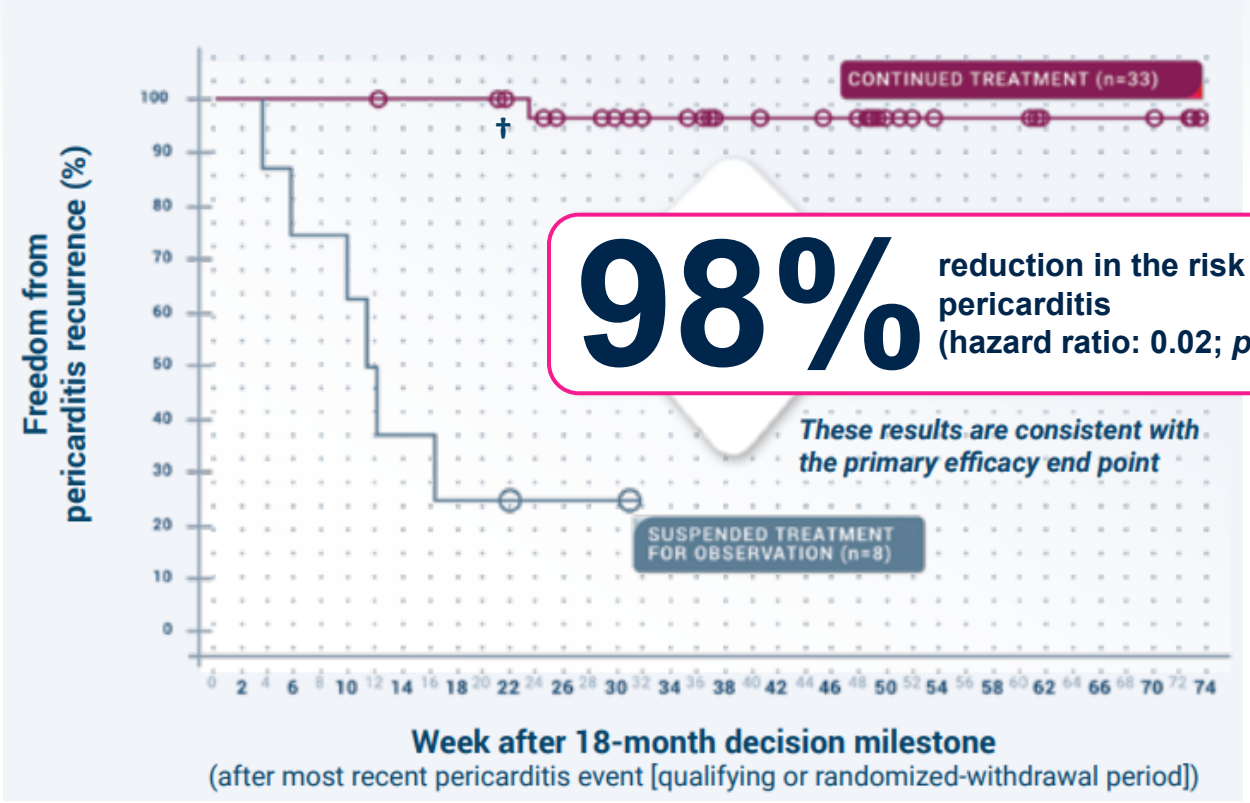
KINIKSA

Sources: Klein AL, Imazio M, Cremer P, et al. Phase 3 trial of interleukin-1 trap rilonacept in recurrent pericarditis. N Engl J Med. 2021;384(1):31-41; ARCALYST (rilonacept) prescribing information 2021

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

ARCALYST reduced the risk of pericarditis recurrence

Recurrence-free rate was 97% (32 of 33) for ARCALYST and 25% (2 of 8) for those who suspended therapy



98% **reduction in the risk of recurrent pericarditis (hazard ratio: 0.02; *p*<0.0001)**

These results are consistent with the primary efficacy end point

3% (1 of 33) of patients who continued ARCALYST treatment experienced a recurrence (during a treatment interruption of 4 weekly doses).[1]

- The median time to recurrence could not be estimated due to low number

75% (6 of 8) of patients who suspended treatment for observation experienced a recurrence.[1]

- The median time to recurrence after suspension of ARCALYST treatment was **11.8 weeks**

Control of recurrent pericarditis requires continued blockade of IL-1 signaling for the duration of disease.[2,3]

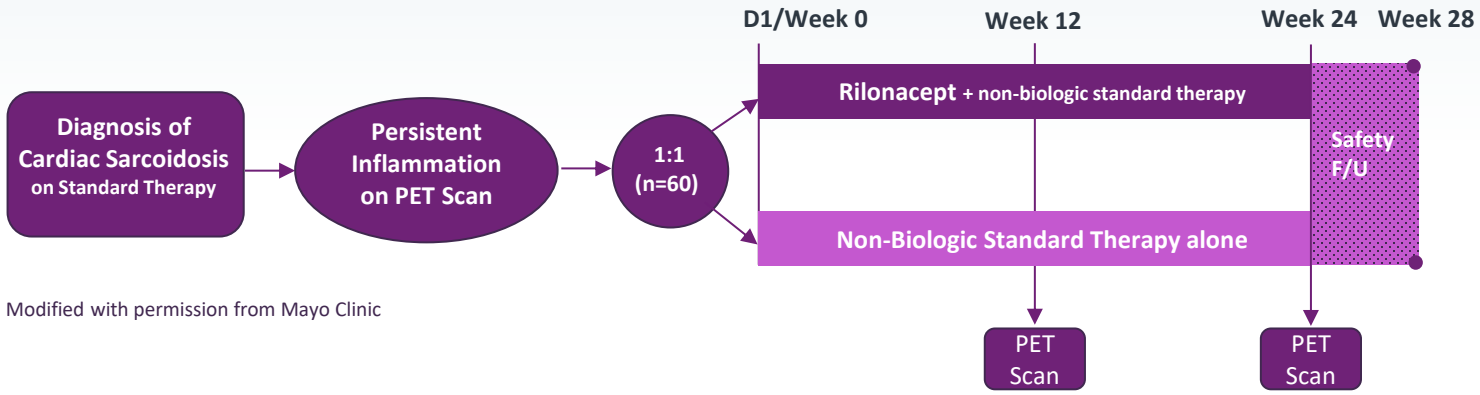
†The patient with a recurrence at 23.4 weeks had interrupted rilonacept treatment ~4 weeks prior.

1) Imazio M, Klein AL, Brucato A, et al. Sustained pericarditis recurrence risk reduction with long-term rilonacept. *J Am Heart Assoc.* 2024:e032516; 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; 3) Imazio M, Mardigyan V, Andreis A, Franchin L, De Biasio M, Collini V. New developments in the management of recurrent pericarditis. Can J Cardiol. 2023;39(8):1103-1110

Randomized Phase 2 Trial of Rilonacept in Cardiac Sarcoidosis

Collaborative study agreement with Mayo Clinic and The Johns Hopkins University

PROBE-design study to evaluate efficacy and safety of rilonacept over 24 weeks of treatment in participants with cardiac sarcoidosis[1]



Modified with permission from Mayo Clinic

Primary Efficacy Endpoint

- Change from baseline in number of segments with FDG uptake on cardiac FDG-PET scan at Week 24

Key Secondary Endpoints

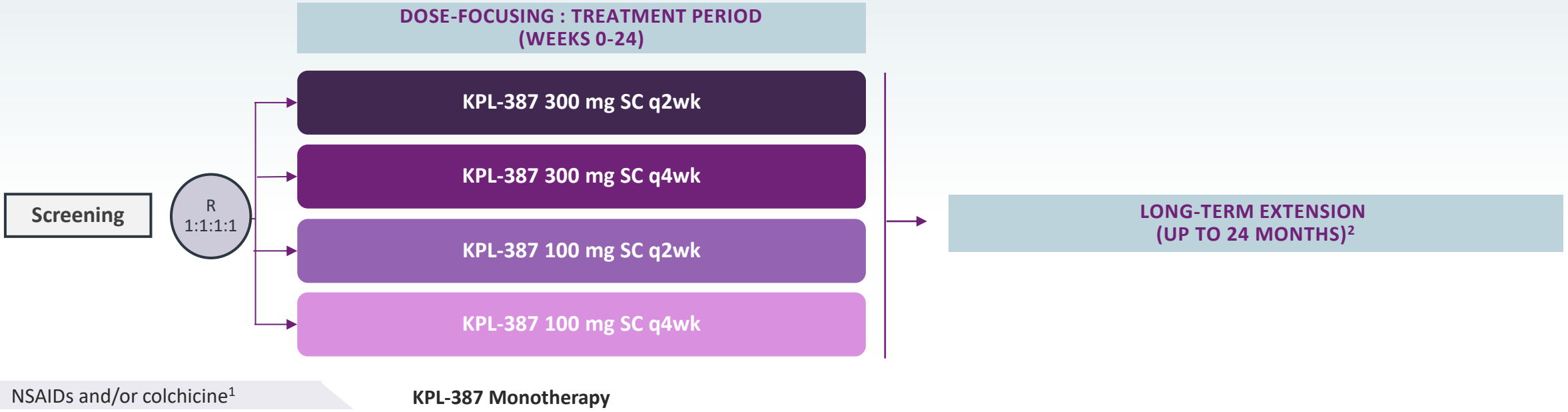
- Change from baseline in number of segments with FDG uptake at Week 12
- Change from baseline in SUV_{Max} at Week 12
- Change from baseline in SUV_{Max} at Week 24
- Change from baseline in SPRS on FDG-PET scan at Week 24

1.) Mayo Clinic is the IND-holder (IND: 172350); Clinicaltrials.gov NCT06660732.
PROBE = prospective, randomized, open label, blinded endpoint; CS = cardiac sarcoidosis; SC = subcutaneous; FDG = flurodeoxyglucose; PET = positron emission tomography; SUV_{Max} = maximum standardized uptake value; SPRS = summed perfusion rest score; qwk = every week; F/U = follow-up.

Appendix
KPL-387

KPL-387 Phase 2/3 Recurrent Pericarditis Clinical Trial

Phase 2: Dose-focusing study



Population:

- Up to ~80 patients diagnosed with RP presenting at screening with a qualifying pericarditis episode despite treatment with conventional oral therapies.

Phase 2 Primary Efficacy Endpoint

- Time to Treatment Response[3]

Phase 2 Key Secondary Endpoints

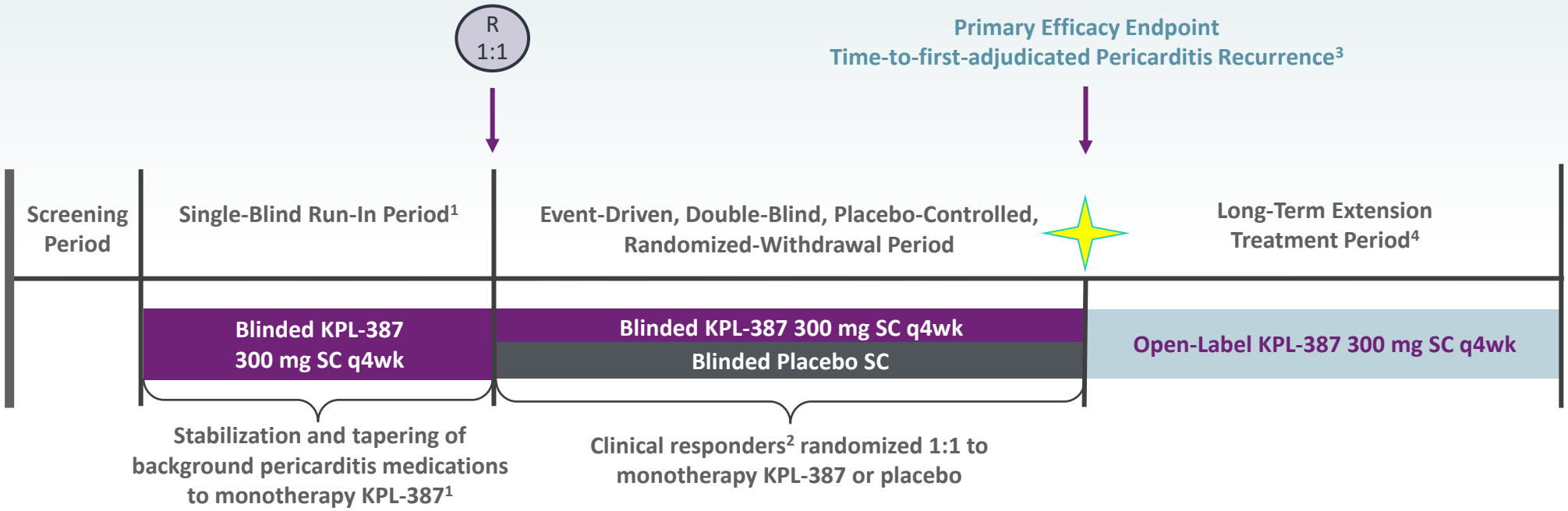
- Time to Pain Response
- Time to CRP normalization (CRP ≤ 0.5 mg/dL).

1) KPL-387 will be administered in addition to conventional oral pericarditis medications (NSAIDs and/or colchicine) from baseline to Week 1 and then weaned off pericarditis medications to achieve KPL-387 monotherapy by Week 2. Participants previously treated with glucocorticoids must have discontinued their use at least 72 hours prior to first study drug administration; 2) Up to 24 months or the time KPL-387 is approved for commercial use in that region to treat recurrent pericarditis; 3) Treatment Response is defined as Pain Response (NRS score ≤ 2 on the 11-point daily pericarditis pain NRS) and at least one CRP level ≤ 0.5 mg/dL within 7 days before or after the Pain Response.

NSAID = non-steroidal anti-inflammatory drug; RP = recurrent pericarditis; CRP = C-reactive protein; NRS = numerical rating scale (for chest pain); R = randomization; SC = subcutaneous

KPL-387 Phase 2/3 Recurrent Pericarditis Clinical Trial

Phase 3: Pivotal study



Population:

- Up to ~85 patients diagnosed with RP presenting at screening with a qualifying pericarditis episode despite treatment with conventional oral therapies.

Primary Efficacy Endpoint

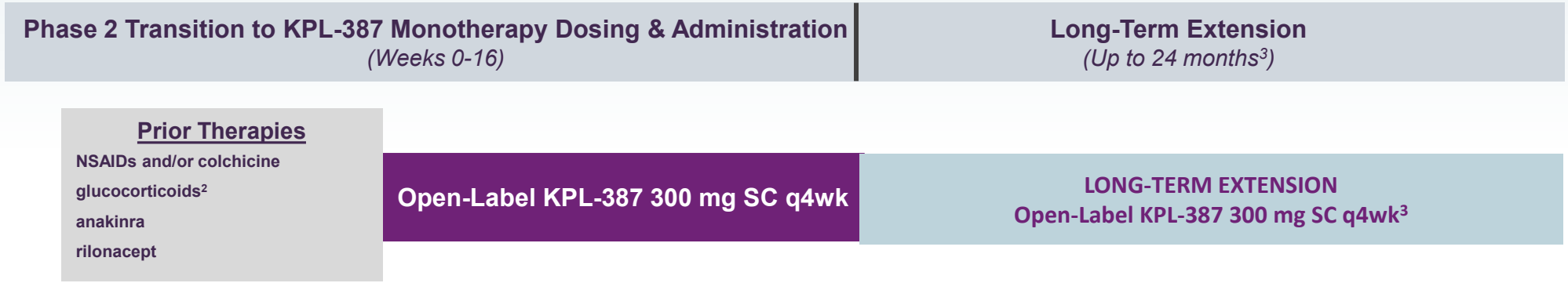
- Time-to-first-adjudicated Pericarditis Recurrence[3]

1) Duration of the run-in period undisclosed in order to maintain study subjects blinded to the start of the randomized-withdrawal period; 2) Clinical response defined as the weekly average of daily pericarditis pain ≤ 2.0 on the 11-point NRS and a CRP level ≤ 0.5 mg/dL, while receiving monotherapy KPL-387 without a recurrence; 3) Time to Pericarditis Recurrence is defined as the time from randomization in the Randomized Withdrawal (RW) Period to the date of the first Pericarditis Recurrence for each participant. Only CEC-confirmed Pericarditis Recurrences will be considered as events for the primary efficacy analysis in the Phase 3 pivotal study; 4) Up to 24 months or the time KPL-387 is approved for commercial use in that region to treat recurrent pericarditis.

CEC = Clinical Endpoint Committee; CRP = C-reactive protein; NRS = numerical rating scale (for chest pain); R = randomization; SC = subcutaneous

Transition to KPL-387 Monotherapy Dosing & Administration Study

Supplemental Phase 2 study evaluating the transition of patients from standard therapies to KPL-387 monotherapy



Population

- Up to ~80 participants with well-controlled RP[1] receiving standard therapies: NSAIDs and/or colchicine, glucocorticoids[2], and/or IL-1α and IL-1β inhibition (anakinra or rilonacept)

Study Objective

- To evaluate the efficacy/safety of dosing regimens used to transition patients with well-controlled RP to KPL-387 monotherapy from stable prior treatment with standard therapies

1) No recurrence within 3 months prior to baseline; CRP < 0.5 mg/dL within 14 days of Baseline and NRS ≤ 3 at Baseline; no clinical worsening or suspicion of impending recurrence; 2) Glucocorticoids or IL-1 pathway inhibitors may be used alone or in combination with NSAIDs and/or colchicine; 3) Up to 24 months or the time KPL-387 is approved for commercial use in that region to treat recurrent pericarditis.
NSAID = non-steroidal anti-inflammatory drug; RP = recurrent pericarditis; SC = subcutaneous

KPL-387 Phase 1 SAD/MAD Study

First-in-human study evaluating safety, tolerability, pharmacokinetics, and immunogenicity

Part A: Single Ascending Dose Cohorts (1-11) | **Part B: Multiple Ascending Dose Cohorts (12-14)**

SC Administration

- 7 600 mg (6 active/2 placebo)
- 6 300 mg (6 active/2 placebo)
- 5 100 mg (6 active/2 placebo)
- 4 30 mg (6 active/2 placebo)
- 3 10 mg (6 active/2 placebo)
- 2 3 mg (4 active/2 placebo)
- 1 1 mg (4 active/2 placebo)

IV Administration

- 11 1000 mg (6 active/2 placebo)
- 10 300 mg (6 active/2 placebo)
- 9 100 mg (6 active/2 placebo)
- 8 30 mg (6 active/2 placebo)

SC Administration

- 14 300 mg, q2wk, 12 wks (6 active/2 placebo)
- 13 300 mg, q4wk, 12 wks (6 active/2 placebo)
- 12 100 mg, q4wk, 12 wks (6 active/2 placebo)

Enrollment:
- Approximately 112 Subjects

Population:
- Healthy Volunteers

Primary Endpoints:
- Safety
- Tolerability

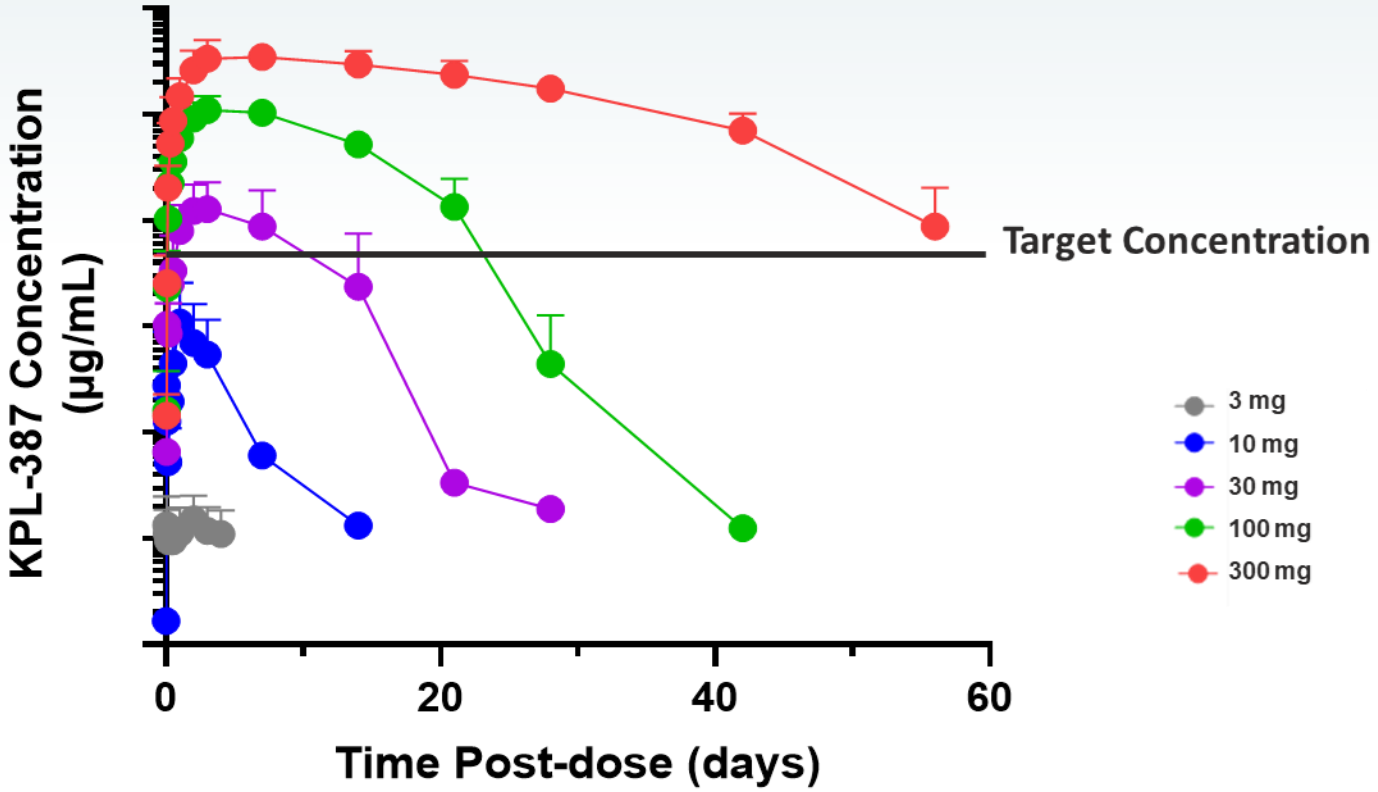
Key Secondary Endpoints:
- Pharmacokinetics
- ADA

SAD = single ascending dose; MAD = multiple ascending dose; SC = subcutaneous; IV = intravenous; q2wk = every 2 weeks; q4wk = every 4 weeks; ADA = anti-drug antibodies

The Single-Dose PK of KPL-387 Support the Monthly Dose Paradigm

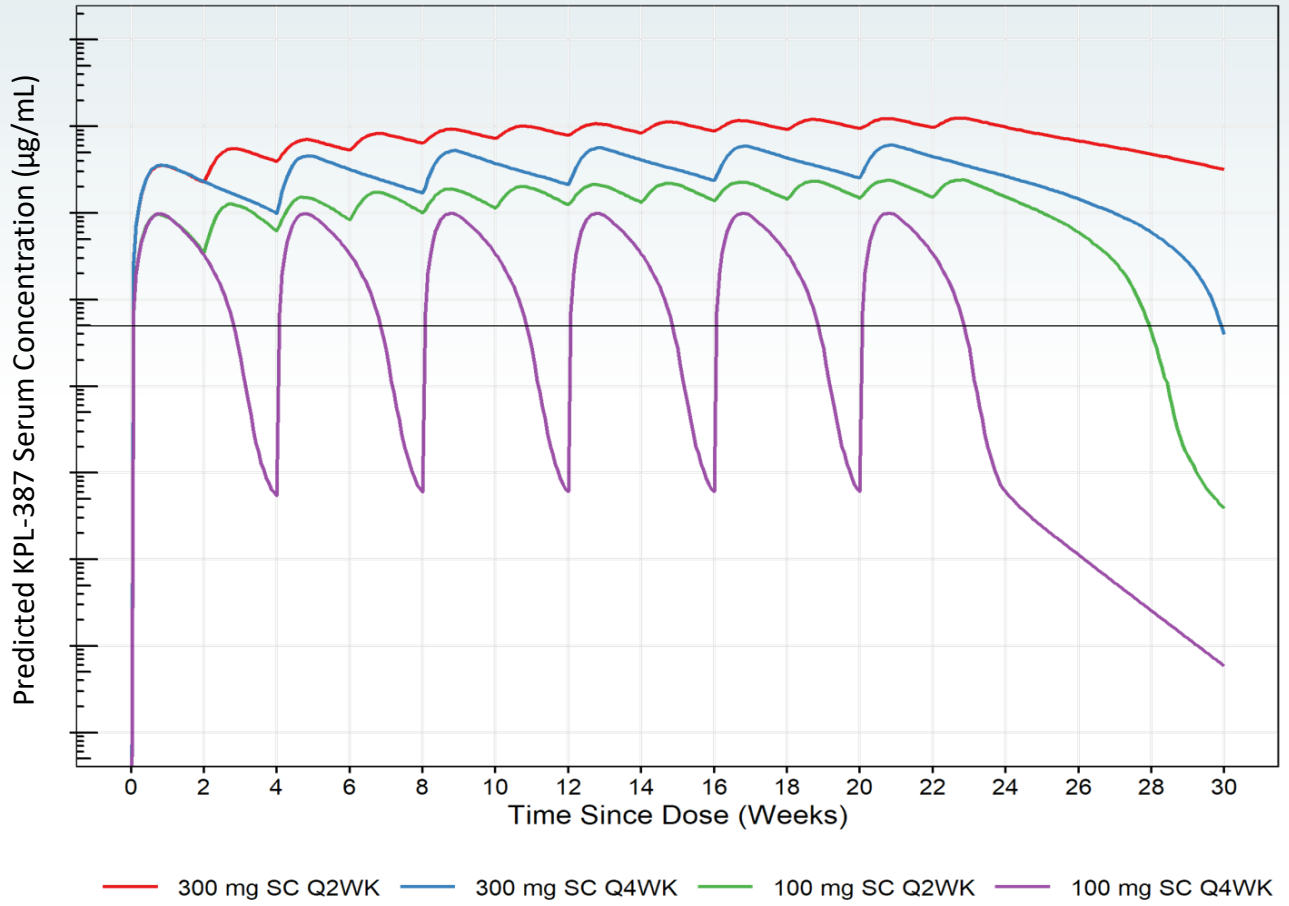
Topline data from KPL-387 Phase 1 single ascending dose study

KPL-387 Pharmacokinetics (Subcutaneous Administration) in Healthy Volunteers



PK = Pharmacokinetics

PK-Modeling and Dose Simulations for KPL-387 Phase 2/3 RP Study



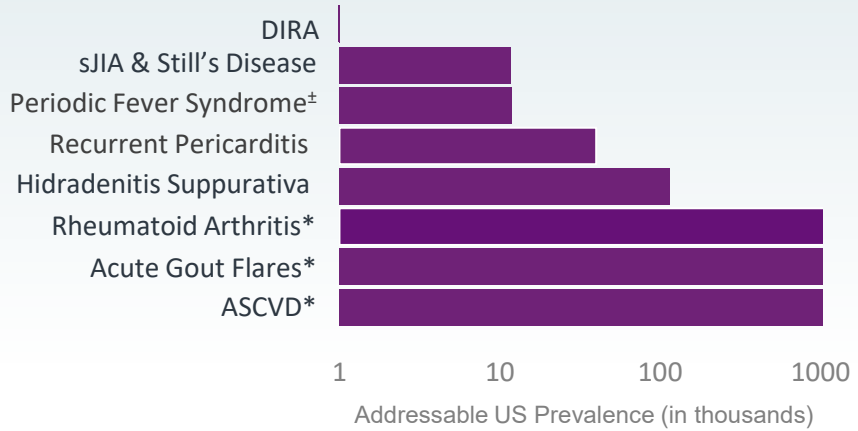
Target Concentration

Pharmacokinetic modeling of KPL-387 at the anticipated therapeutic dose supports the monthly dose paradigm

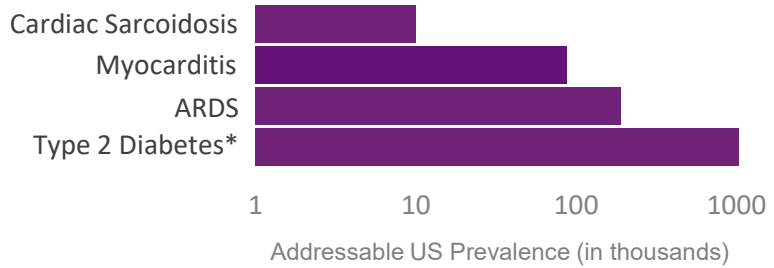
Appendix KPL-1161

IL-1α and IL-1β Inhibition is Well Established in Cardiovascular and Inflammatory Diseases

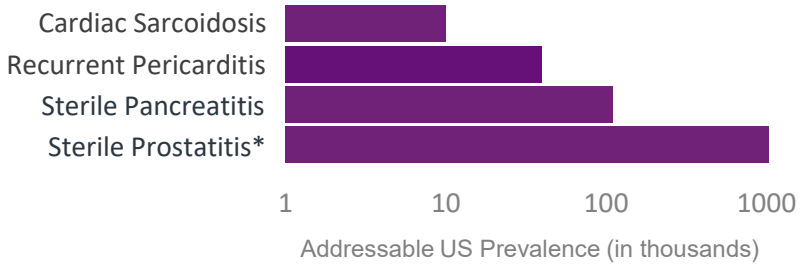
Indications with Regulatory Approval or Positive RCT Trials



Indications with Positive Initial Clinical Data



Indications with Pending Data & Ongoing Trials



INDICATION SELECTION CRITERIA

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

±Includes HIDS/MKD, TRAPS, FMF; *Indications with an addressable US prevalence of >800,000

RCT = Randomized Controlled Trials; DIRA = Deficiency of Interleukin-1 Receptor Antagonist; sJIA = Systemic Juvenile Idiopathic Arthritis; CAPS = Cryopyrin-Associated Periodic Syndromes; TRAPS = Tumor Necrosis Factor Receptor-Associated Periodic Syndrome; HIDS = Hyperimmunoglobulin D Syndrome; FMF = Familial Mediterranean Fever; ASCVD = Atherosclerotic Cardiovascular Disease; ARDS = Acute Respiratory Distress Syndrome

Sources: Aksentijevich I. N Engl J Med. 2009; Gerfaud-Valentin M. Autoimmun Rev. 2014; Georgin-Lavialle. Rev Med Interne. 2023; Klein A. J Am Heart Assoc. 2021; Garg A. JAMA Dermatol. 2017; Hunter TM. Rheumatol Int. 2017; Keenan RT. Am J Med. 2011; Zhang X. Am J Prev Cardiol. 2024; Santulli G. Am J Cardiol. 2023; Wang YW. BMC Public Health. 2023; Diamond M. StatPearls. 2025; Bullard KM. CDC MMWR. 2018; Hoque R. Pancreas. 2012; Pendegast HJ. StatPerls 2024; 2024 ClearView Analysis.

KINIKSA

Corporate Presentation

OCTOBER 2026