First Quarter 2026 Financial Results and Recent Portfolio Execution

APRIL 28, 2026

Agenda

- **Introduction** | *Sanj K. Patel, Chief Executive Officer*
- **ARCALYST® Commercial Execution** | *Ross Moat, Chief Operating Officer*
- **KPL-387 Program Review** | *John F. Paolini, Chief Medical Officer*
- **First Quarter 2026 Financial Results** | *Mark Ragosa, Chief Financial Officer*
- **Closing Remarks** | *Sanj K. Patel, Chief Executive Officer*
- **Q&A Session**

KINIKSA

Forward Looking Statements

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Introduction

Sanj K. Patel
Chief Executive Officer

Q1 2026 Business Highlights

Driving ARCALYST Revenue

- **Q1 2026 ARCALYST revenue of $214.3M**
- **Full year 2026 net revenue guidance raised** to between **$930-$945 million** from previous guidance of $900-$920 million

Advancing Clinical Portfolio

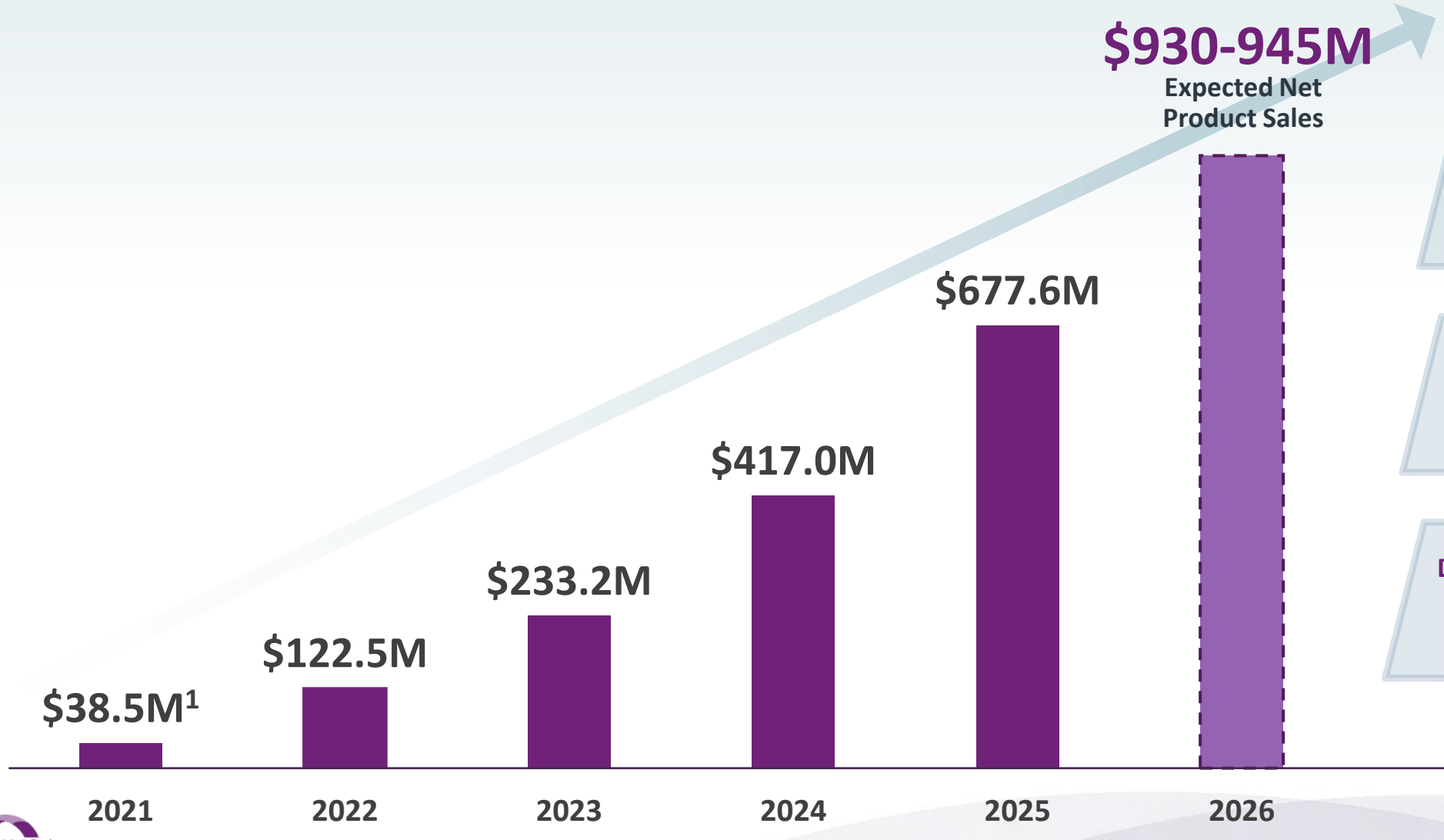
- **KPL-387 Phase 2 data** from Phase 2/3 recurrent pericarditis trial **on track for 2H 2026**
- **Phase 3 pivotal trial** expected to initiate by the **end of 2026**
- **KPL-1161 Phase 1** first-in-human trial to initiate by the **end of 2026**

Maintaining Financial Strength

- Strong financial position with **~$468M in cash**
- Current operating plan expected to remain **cash flow positive** on an annual basis
- Financial strength provides capacity to **continue investing in value creating opportunities**

2026 ARCALYST Net Product Sales Guidance

Revenue guidance raised to $930-$945M from $900-$920M



Positioning ARCALYST as 2nd line Standard of Care

Deepening penetration into 2+ recurrence population

Driving adoption among larger 1st recurrence population

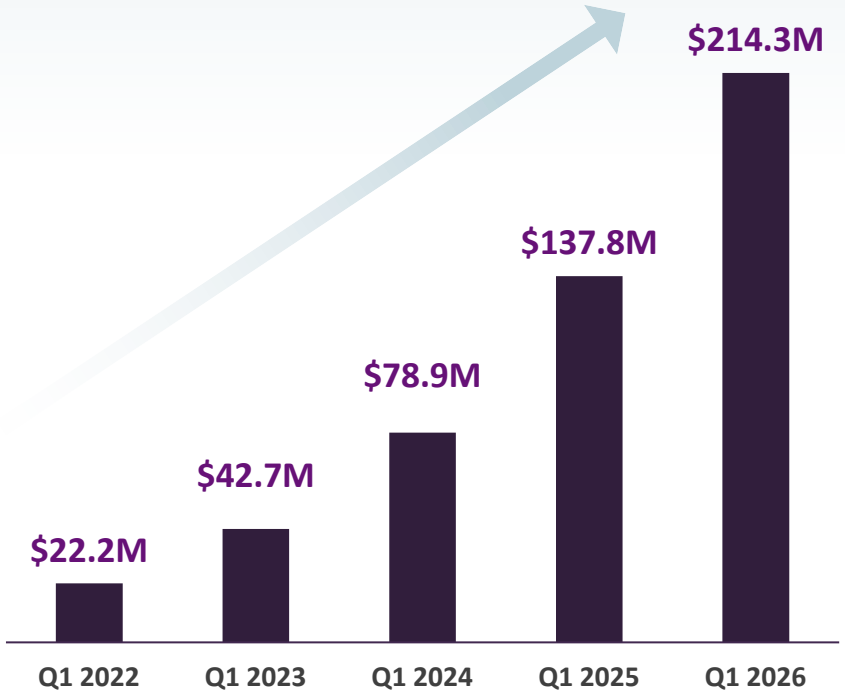
1) 2021 = 9 months of availability (Q2-Q4).

ARCALYST Commercial Execution

Ross Moat
Chief Operating Officer

Expanding Adoption of IL-1α & IL-1β Inhibition Driving ARCALYST Sales

Year-Over-Year Net Revenue Growth



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

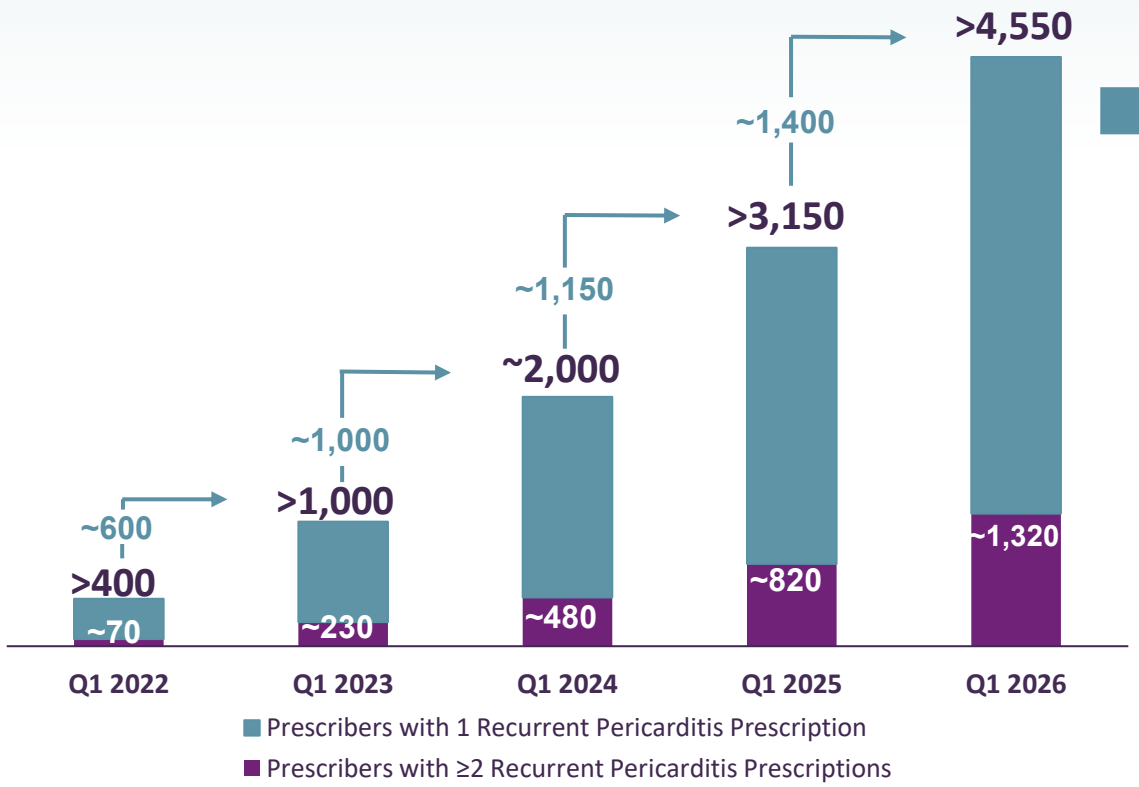
Initial Target Population

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

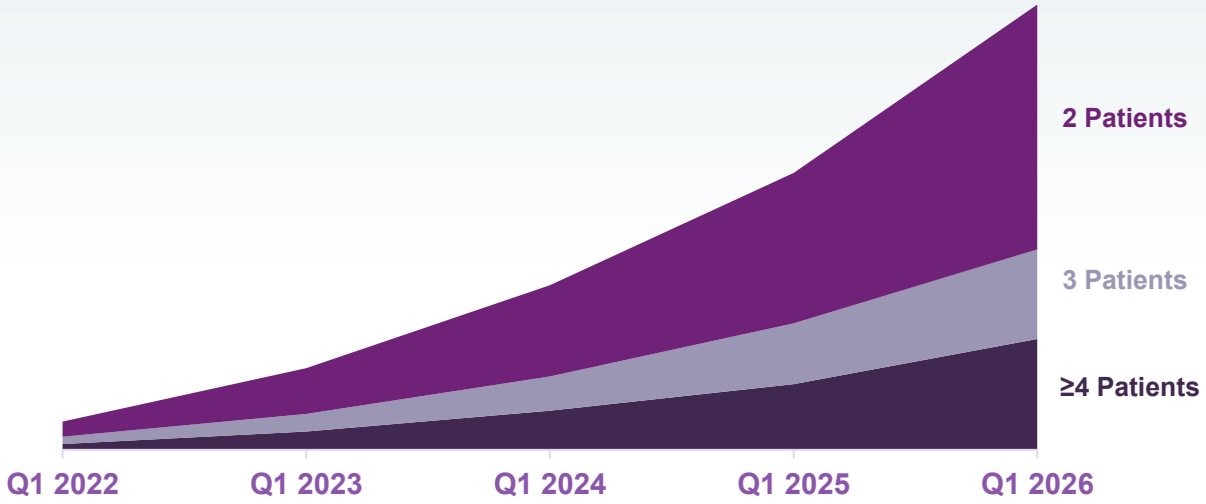
1) HCP market research 2026; Kiniksa data on file.

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 Q1 2026 came from repeat prescribers**

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Highly Targeted DTC Campaign Promotes Education and Self-Advocacy

Empowering recurrent pericarditis patients to ask about ARCALYST with *Heart's Home™* connected TV ad

Efficiently Expanding Our Reach to Additional Patients

- Campaign adapts proven direct-to-consumer tactics for **cost-effective use in rare disease**
- Utilizes advanced analytics to direct content to an **enriched population of likely recurrent pericarditis patients**
- Leverages connected TV to reach individual patients; **significantly more cost-effective than traditional TV**
- Potential to provide **additional upside beyond current revenue guidance**

ARCALYST is prescribed ~80% of the time when requested by a patient[1]

1) HCP market research 2026; Kiniksa data on file.

KPL-387 Program Review

John F. Paolini
Chief Medical Officer

KPL-387 Development Program Includes Pivotal & Supplemental Studies

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]

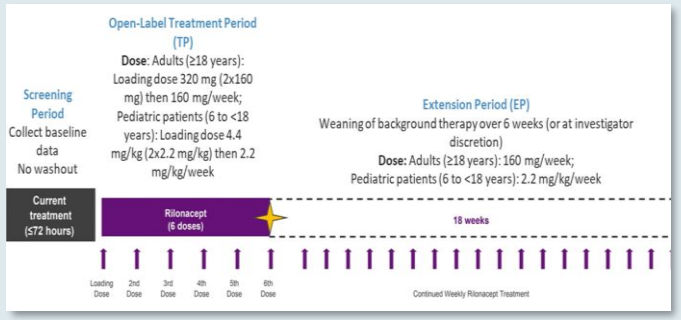
1) NCT07010159; 2) Supplemental study evaluating the efficacy/safety of dosing regimens used to transition patients with well-controlled RP to KPL-387 monotherapy from stable prior treatment with standard therapies; 3) No recurrence within 3 months prior to baseline; CRP < 0.5 mg/dL within 14 days of 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

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

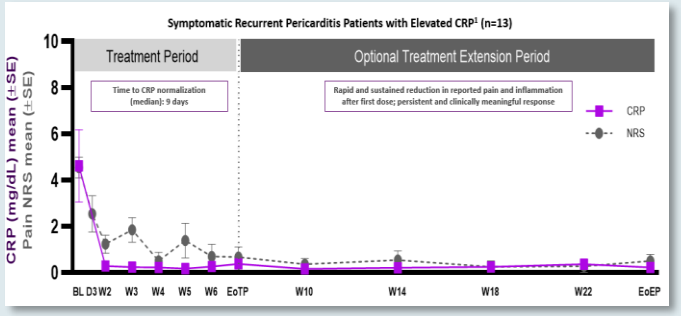
Cadence & magnitude of initial response and duration of action in Phase 2 inform Phase 3 efficacy, recurrence risk reduction

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

(n=13)



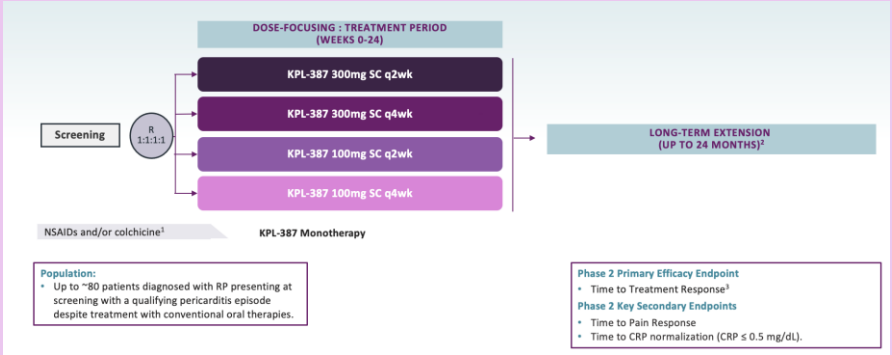
Test of treatment response and durability: Addition of rilonacept in patients failing standard oral therapies



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

First Quarter 2026 Financials

Mark Ragosa
Chief Financial Officer

First Quarter 2026 Financial Results

Income Statement	Three Months Ended March 31,	
	2026	2025
Product Revenue	$214.3M	$137.8M
License and Collaboration Revenue	$0.0M	$0.0M
Total Revenue	**$214.3M**	**$137.8M**
Cost of Goods Sold	$20.8M	$17.9M
Collaboration Expenses[1,2]	$75.6M	$43.8M
Research and Development	$27.5M	$19.3M
Selling, General and Administrative	$61.2M	$43.5M
Total Operating Expenses	**$185.0M**	**$124.5M**
Operating Income (Loss)	**$29.3M**	**$13.3M**
Other Income, Net	$3.4M	$2.3M
Income Tax Provision	($10.1M)	($7.0M)
Net Income	**$22.6M**	**$8.5M**

Collaboration Expenses[1]	Three Months Ended March 31,	
	2026	2025
ARCALYST Net Sales	**$214.3M**	**$137.8M**
Profit Split-Eligible Cost of Goods Sold[2]	($20.3M)	($17.6M)
Commercial, Marketing, Regulatory and Other Expenses	($42.8M)	($32.6M)
ARCALYST Collaboration Operating Profit	**$151.2M**	**$87.6M**
ARCALYST Collaboration Expense	$75.6M	$43.8M
ARCALYST Out-Licensing[3]	$0.0M	$0.0M
Other Collaboration Expenses	$0.0M	$0.0M
Total Collaboration Expenses	**$75.6M**	**$43.8M**

Balance Sheet	March 31, 2026	December 31, 2025
Cash, Cash Equivalents and Short-term Investments	**$468.1M**	**$414.1M**

Operating Plan Expected to Remain Cash Flow Positive on an Annual Basis

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 good sold – amortization of Regeneron milestone payment; 3) Revenue associated with ARCALYST Out-Licensing is included in Licensing and Collaboration Revenue.

Closing Remarks

Sanj K. Patel

Chief Executive Officer

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