Reference number(s)
1800-A

Specialty Guideline Management Arcalyst

Products Referenced by this Document

Drugs that are listed in the following table include both brand and generic and all dosage forms and strengths unless otherwise stated. Over-the-counter (OTC) products are not included unless otherwise stated.

Brand Name	Generic Name
Arcalyst	rilonacept

Indications

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications[1]

- Treatment of Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Autoinflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS) in adults and pediatric patients 12 years of age and older.
- Maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA) in adults and pediatric patients weighing at least 10 kilograms (kg).
- Treatment of recurrent pericarditis (RP) and reduction in risk of recurrence in adults and pediatric patients 12 years and older.

All other indications are considered experimental/investigational and not medically necessary.

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Documentation

Submission of the following information is necessary to initiate the prior authorization review:

Deficiency of Interleukin-1 Receptor Antagonist (DIRA)

Initial requests

IL1RN gene variant status

Recurrent Pericarditis (RP)

Initial requests

- Chart notes, medical record documentation, or test results supporting the diagnosis of pericarditis (e.g., electrocardiogram [ECG], cardiovascular magnetic resonance [CMR] findings).
- Chart notes, medical record documentation, or claims history supporting previous medications tried, including response to therapy.

Continuation requests

Chart notes or medical record documentation supporting positive clinical response.

Prescriber Specialties

This medication must be prescribed by or in consultation with one of the following:

- Cryopyrin-associated periodic syndromes (CAPS) and deficiency of interleukin-1 receptor antagonist (DIRA): rheumatologist or immunologist
- Recurrent pericarditis (RP): cardiologist, rheumatologist, or immunologist

Coverage Criteria

Cryopyrin-Associated Periodic Syndromes (CAPS)[1-3]

Authorization of 12 months may be granted for members 12 years of age or older for treatment of CAPS when both of the following criteria are met:

- Member has a diagnosis of familial cold autoinflammatory syndrome (FCAS) with classic signs and symptoms (i.e., recurrent, intermittent fever and rash that were often exacerbated by exposure to generalized cool ambient temperature) or Muckle-Wells syndrome (MWS) with

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classic signs and symptoms (i.e., chronic fever and rash of waxing and waning intensity, sometimes exacerbated by exposure to generalized cool ambient temperature).
- Member has functional impairment limiting the activities of daily living.

Deficiency of Interleukin-1 Receptor Antagonist (DIRA)[1,4]

Authorization of 12 months may be granted for members weighing at least 10 kg for treatment of DIRA when both of the following criteria are met:

- Member has IL1RN gene variants.
- Arcalyst will be used for maintenance of remission following treatment with Kineret (anakinra).

Recurrent Pericarditis (RP)[1,5-8]

Authorization of 12 months may be granted for members 12 years of age or older for treatment of recurrent pericarditis when both of the following criteria are met:

- Member has had at least two episodes of pericarditis with pericarditis being defined as both of the following:
    - Clinical presentation (e.g. chest pain, infarct-like symptoms, arrhythmias, heart failure, aborted sudden cardiac death)
    - At least one of the following:
        - Pericardial rubs
        - PR depression, widespread ST-segment elevation
        - C-reactive protein (CRP) elevation
        - New or worsening pericardial effusion
        - Pericardial edema and/or late gadolinium enhancement (LGE) from CMR findings
- Member has failed at least two agents of standard therapy (e.g., colchicine, non-steroidal anti-inflammatory drugs [NSAIDs], corticosteroids).

Continuation of Therapy

Cryopyrin-Associated Periodic Syndromes (CAPS)[3]

Authorization of 12 months may be granted for all members 12 years of age or older (including new members) who are using the requested medication for CAPS and who achieve or maintain a positive clinical response as evidenced by low disease activity or improvement in signs and symptoms of the condition.

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Deficiency of Interleukin-1 Receptor Antagonist (DIRA)

Authorization of 12 months may be granted for all members weighing at least 10 kg (including new members) who are using the requested medication for DIRA and who achieve or maintain a positive clinical response as evidenced by low disease activity or improvement in signs and symptoms of the condition.

Recurrent Pericarditis (RP)[1,5-8]

Authorization of 12 months may be granted for all members 12 years of age or older (including new members) who are using the requested medication for recurrent pericarditis and who achieve or maintain a positive clinical response as evidenced by decreased recurrence of pericarditis or improvement in signs and symptoms of the condition when there is improvement in any of the following:

- Pericarditic or pleuritic chest pain
- Pericardial or pleural rubs
- ECG
- Pericardial effusion
- CRP
- CMR

Other[1]

For all indications: Member has been evaluated for tuberculosis (TB) infection prior to initiation of treatment with a biologic or targeted synthetic drug associated with an increased risk of TB.

For all indications: Member cannot use the requested medication concomitantly with any other biologic drug or targeted synthetic drug.

References

1. Arcalyst [package insert]. London, UK: Kiniksa Pharmaceuticals (UK), Ltd.; October 2025.
2. Hoffman HM, Throne ML, Amar NJ, et al. Efficacy and safety of rilonacept (interleukin-1 trap) in patients with cryopyrin-associated periodic syndromes. Results from two sequential placebo-controlled studies. Arthritis Rheum. 2008;58(8):2443-52.
3. Lachmann HJ, Kone-Paut I, Kuemmerle-Deschner JB, et al. Canakinumab in CAPS Study Group. Use of canakinumab in the cryopyrin-associated periodic syndrome. N Engl J Med. 2009;360(23):2416-2425.
4. Garg M, de Jesus A, Chapelle D, et al. Rilonacept maintains long-term inflammatory remission in patients with deficiency of the IL-1 receptor antagonist. JCI Insight. 2017;2(16):e94838.

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5. Schulz-Menger J, Collini V, Groschel J, et al. 2025 ESC Guidelines for the management of myocarditis and pericarditis. Eur Heart J. 2025; 46: 3952-4041.
6. Chiabrando JG, Bonaventura A, Vecchié A, et al. Management of acute and recurrent pericarditis: JACC State-of-the-art review. J Am Coll Cardiol. 2020;75(1):76-92.
7. 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.
8. Kumar S, Furqan M, Kafil T, et al. (2023). The paradigm shift in the management of recurrent pericarditis. American College of Cardiology. https://www.acc.org/Latest-in-Cardiology/Articles/2022/12/19/14/52/The-Paradigm-Shift-in-the-Management-of-Recurrent-Pericarditis.

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