AETNA BETTER HEALTH® Coverage Policy/Guideline				
Name:	Arcalyst		Page:	1 of 5
Effective Date:	10/15/2025		Last Review Date:	9/2025
Applies to:	☒Illinois ☐New Jersey ☐Pennsylvania Kids	☐Florida ☐Maryland ☐Virginia	☐Florida Kids ☐Michigan ☐Kentucky PRMD	

Intent:

The intent of this policy/guideline is to provide information to the prescribing practitioner outlining the coverage criteria for Arcalyst under the patient's prescription drug benefit.

Description:

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.

Applicable Drug List:

Arcalyst

Policy/Guideline:

Documentation for all indications:
The patient is unable to take THREE preferred products, where indicated, for the given diagnosis due to a trial and inadequate treatment response or intolerance, or a contraindication. Documentation is required for approval.

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 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)[1,2,4]

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 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)[1,5]

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)[1,6-9]

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.
- 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)[4]

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.

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)[1,6-9]

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
- Electrocardiogram (ECG)
- Pericardial effusion
- C-reactive protein (CRP)

Other[1,3]

For all indications: Member has had a documented negative tuberculosis (TB) test (which can include a tuberculosis skin test [TST] or an interferon-release assay [IGRA]) within 12 months of initiating therapy for persons who are naïve to biologic drugs or targeted synthetic drugs associated with an increased risk of TB.

If the screening testing for TB is positive, there must be further testing to confirm there is no active disease (e.g., chest x-ray). Do not administer the requested medication to members with active TB infection. If there is latent disease, TB treatment must be started before initiation of the requested medication.

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

Approval Duration and Quantity Restrictions:

Approval:
Initial Approval: 12 months
Renewal Approval: 12 months

Quantity Level Limit:

- Arcalyst (rilonacept) injection 220 mg vial: 8 vials per 28 days

References:

1. Arcalyst [package insert]. London, UK: Kiniksa Pharmaceuticals (UK), Ltd.; May 2021.
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. Testing for TB Infection. Centers for Disease Control and Prevention. Retrieved on April 18, 2025 from: https://www.cdc.gov/tb/index.html.
4. 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.
5. 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.
6. Adler Y, Charron P, Imazio M, et al. 2015 ESC Guidelines for the diagnosis and management of pericardial diseases: The Task Force for the Diagnosis and Management of Pericardial Diseases of the European Society of Cardiology (ESC) Endorsed by: The European Association for Cardio-Thoracic Surgery (EACTS). Eur Heart J. 2015;36(42):2921-64.
7. 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.

8. 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.
9. 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.