AETNA BETTER HEALTH®
Coverage Policy/Guideline

Name:	Arcalyst	Page:	1 of 4
Effective Date:	10/25/2023	Last Review Date:	10/2023

Applies to:	☐Illinois	☐Florida	☒Florida Kids
	☐New Jersey	☒Maryland	☐Michigan
	☒Pennsylvania Kids	☐Virginia	☒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:

FDA-Approved Indications

A. Treatment of Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Autoinflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS) in adults and children 12 years of age and older.
B. Maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA) in adults and pediatric patients weighing at least 10 kg.
C. 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:

Non-preferred: Arcalyst

Policy/Guideline:

Documentation for all indications:
The patient is unable to take ONE preferred anti-TNF (Enbrel or preferred adalimumab product) AND Kevzara, 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:

A. Deficiency of interleukin-1 receptor antagonist (DIRA) initial requests: *IL1RN* mutation status

B. Recurrent pericarditis:
1. Initial requests: Chart notes, medical record documentation, or claims history supporting previous medications tried, including response to therapy.
2. Continuation requests: Chart notes or medical record documentation supporting positive clinical response.

Prescriber Specialty:
This medication must be prescribed by or in consultation with one of the following:
A. Cryopyrin associated periodic syndromes (CAPS) and deficiency of interleukin-1 receptor antagonist (DIRA): rheumatologist or immunologist
B. Recurrent pericarditis: cardiologist, rheumatologist, or immunologist

Criteria for Initial Approval:

A. Cryopyrin-associated periodic syndromes (CAPS)
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:
1. 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).
2. Member has functional impairment limiting the activities of daily living.

B. Deficiency of interleukin-1 receptor antagonist (DIRA)
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:
1. Member has *IL1RN* mutations.
2. Arcalyst will be used for maintenance of remission following treatment with Kineret (anakinra).

C. Recurrent pericarditis
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:
1. Member has had at least two episodes of pericarditis.
2. Member has failed at least 2 agents of standard therapy (e.g., colchicine, non-steroidal anti-inflammatory drugs [NSAIDs], corticosteroids).

Continuation of Therapy:

A. Cryopyrin-associated periodic syndromes (CAPS)
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 who achieve or maintain positive clinical response as evidenced by low disease activity or improvement in signs and symptoms of the condition.

B. 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 positive clinical response as evidenced by low disease activity or improvement in signs and symptoms of the condition.

C. Recurrent pericarditis
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:

1. Pericarditic chest pain
2. Pericardial rubs
3. Electrocardiogram (ECG)
4. Pericardial effusion
5. C-reactive protein (CRP)

Other Criteria:
Member has had a documented negative tuberculosis (TB) test (which can include a tuberculosis skin test [PPD], an interferon-release assay [IGRA], or a chest x-ray)* within 6 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. 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.

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 10, 2023 from: https://www.cdc.gov/tb/topic/testing/tbtesttypes.htm.
4. 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.
5. Chiabrando JG, Bonaventura A, Vecchié, et al. Management of acute and recurrent pericarditis: *JACC* State-of-the-art review. *J Am Coll Cardiol.* 2020;75(1):76-92.
6. 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.
7. 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.
8. 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. https://doi.org/10.1172/jci.insight.94838.