

Reference number
1800-A

SPECIALTY GUIDELINE MANAGEMENT

ARCALYST (rilonacept)

POLICY

I. 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

Arcalyst is indicated for:

- A. Treatment of Cryopyrin Associated Periodic Syndromes (CAPS), including Familial Cold Auto-inflammatory 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.

II. DOCUMENTATION

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

- A. Deficiency of interleukin-1 receptor antagonist (DIRA): *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.

III. CRITERIA FOR INITIAL APPROVAL

A. Cryopyrin-associated periodic syndrome (CAPS)

Authorization of 12 months may be granted for treatment of CAPS when all of the following criteria are met:

1. Member has a diagnosis of familial cold auto-inflammatory 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.

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B. Deficiency of interleukin-1 receptor antagonist (DIRA)

Authorization of 12 months may be granted for treatment of DIRA when all of the following criteria are met:

1. Member has loss-of-function *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 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 therapy with colchicine and non-steroidal anti-inflammatory drugs (NSAIDs).

IV. CONTINUATION OF THERAPY

A. Recurrent pericarditis

Authorization of 12 months may be granted for all members (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)

B. All other indications

Authorization of 12 months may be granted for all members (including new members) who are using the requested medication for an indication outlined in Section III and who achieve or maintain positive clinical response as evidenced by low disease activity or improvement in signs and symptoms of the condition.

V. OTHER

For all indications: Member has had a documented negative 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 DMARDs or targeted synthetic DMARDs associated with an increased risk of TB, and repeated yearly for members with risk factors** for TB that are continuing therapy with biologics.

* 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.

** Risk factors for TB include: Persons with close contact to people with infectious TB disease; persons who have recently immigrated from areas of the world with high rates of TB (e.g., Africa, Asia, Eastern Europe, Latin America, Russia); children less than 5 years of age who have a positive TB test; groups with high rates of TB transmission (e.g., homeless persons, injection drug users, persons with HIV infection); persons who work or reside with people who are at an increased risk for active TB (e.g., hospitals, long-term care facilities, correctional facilities, homeless shelters).

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

VI. REFERENCES

1. Arcalyst [package insert]. Tarrytown, NY: Regeneron Pharmaceuticals, Inc.; March 2021.
2. Hoffman HM, Throne ML, Amar NJ, et al. Efficacy and safety of riloncept (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. Centers for Disease Control and Prevention. Tuberculosis (TB). TB risk factors. Available at: <https://www.cdc.gov/tb/topic/basics/risk.htm>. Accessed: November 13, 2020.
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 Nov 7;36(42):2921-64.