

Arcalyst® (rilonacept)

When requesting Arcalyst® (rilonacept), the individual requiring treatment must be diagnosed with an FDA-approved indication and meet the specific coverage guidelines and applicable safety criteria for the covered indication.

FDA-approved Indications

- Arcalyst is an interleukin-1 blocker indicated for the treatment of cryopyrin-associated periodic syndromes (CAPS), including familial cold autoinflammatory syndrome (FCAS), Muckle-Wells syndrome (MWS), and Neonatal Onset Multisystem Inflammatory Disease or chronic infantile neurological cutaneous and articular syndrome in adults and children 12 years of age and older as well as the maintenance of remission in patients weighing at least 10 kg with interleukin-1 receptor antagonist (DIRA) deficiency.
- Arcalyst is indicated for the treatment of recurrent disease and reduction of risk of recurrence in patients greater than or equal to 12 years of age with pericarditis.

Coverage Guidelines

Cryopyrin-Associated Periodic Syndromes (CAPS)

The individual must meet **all** of the following criteria for initial authorization:

- Is 12 years of age or older; AND
- Arcalyst is prescribed by or in consultation with a rheumatologist, geneticist, allergist, immunologist, or dermatologist.

For re-authorization, the individual must meet **all** of the following criteria:

- Has been established on Arcalyst therapy for at least 6 months; AND
- Meets one of the following;
 - Has experienced an improvement in at least one objective measure compared with baseline (e.g., resolution of fever, improvement of serum markers [e.g., C-reactive protein, amyloid A]); OR
 - Has experienced an improvement in at least one symptom compared with baseline (e.g., decreased joint pain, improved function or activities of daily living).
- Note: A patient who has received < 6 months of therapy, or who is restarting therapy with Arcalyst, is reviewed under initial authorization criteria.

Approval duration (initial): 6 months

Approval duration (renewal): 12 months

Deficiency of Interleukin-1 Receptor Antagonist (DIRA)

The individual must meet **all** of the following criteria for initial authorization:

- The patient weighs greater than or equal to 10 kg (22 pounds); AND
- Genetic testing has confirmed a mutation in the *IL1RN* gene; AND
- The patient has demonstrated a clinical benefit with Kineret (e.g., normalized acute phase reactants, resolution of fever, skin rash, and bone pain, and reduced dosage of corticosteroids); AND
- Arcalyst is prescribed by or in consultation with a rheumatologist, geneticist, dermatologist, or a physician specializing in the treatment of autoinflammatory disorders.

For re-authorization, the individual must meet **all** of the following criteria:

- Has been established on Arcalyst therapy for at least 6 months; AND
- Meets one of the following:
 - Has experienced an improvement in at least one objective measure compared with baseline (e.g., improvement in rash or skin manifestations, improvement of serum markers [e.g., C-reactive protein, erythrocyte sedimentation rate]); OR
 - Has experienced an improvement in at least one symptom compared with baseline (e.g., less joint pain, stiffness, or swelling).
- Note: A patient who has received < 6 months of therapy, or who is restarting therapy with Arcalyst, is reviewed under initial authorization criteria.

Approval duration (initial): 6 months

Approval duration (renewal): 12 months

Pericarditis

The individual must meet **all** of the following criteria for initial authorization:

- Is 12 years of age or older; AND
- Arcalyst is prescribed by or in consultation with a cardiologist or rheumatologist; AND
- The patient has recurrent pericarditis; AND
- Prior to starting treatment with Arcalyst, the patient has a history of at least three episodes of pericarditis; AND
- The patient meets one of the following:
 - For the current episode, the patient is receiving standard treatment; OR
 - Standard treatment (e.g., nonsteroidal anti-inflammatory drugs [NSAIDs], colchicine, and/or systemic corticosteroids) is contraindicated.

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For re-authorization, the individual must meet **all** of the following criteria:

- Has been established on Arcalyst therapy for at least 3 months; AND
- Meets one of the following:
 - Has experienced an improvement in at least one objective measure compared with baseline (e.g., continued resolution of fever, normalization of inflammatory markers [e.g., C-reactive protein, erythrocyte sedimentation rate]): OR
 - Has experienced an improvement in at least one symptom compared with baseline (e.g., resolution of chest pain or pericarditis pain).
- Note: A patient who has received < 90 days of therapy, or who is restarting therapy with Arcalyst, is reviewed under initial authorization criteria.

Approval duration (initial): 3 months

Approval duration (renewal): 12 months

Dosing Recommendations

For adult patients 18 years and older:

For CAPS: The recommended dose is a loading dose of 320 mg administered via subcutaneous injection followed by 160 mg once weekly.

For DIRA: The recommended dose is 320 mg administered via subcutaneous injection once weekly given as 2 separate injections (160 mg [2mL] per injection) on the same day at 2 different sites. *Switching from another interleukin-1 blocker:* Discontinue the interleukin-1 blocker and begin Arcalyst at the same time of the next scheduled dose.

For pediatric patients aged 12 to 17 years:

For CAPS: The recommended dose is a loading dose of 4.4 mg/kg, up to a maximum of 320 mg, administered via subcutaneous injection followed by 2.2 mg/kg, up to a maximum of 160 mg, administered via subcutaneous injection once weekly.

For DIRA: Children weighing ≥ 10 kg: The recommended dose is 4.4 mg/kg, up to a maximum of 320 mg, administered via subcutaneous injection once weekly as 1 or 2 separate injections on the same day at different sites.

References

1. Arcalyst® for injection [prescribing information]. Tarrytown, NY: Regeneron Pharmaceuticals; May 2021.
2. Hansmann S, Lainka E, Horneff G, et al. Consensus protocols for the diagnosis and management of the hereditary autoinflammatory syndromes CAPS, TRAPS and MKD/HIDS: a German PRO-KIND initiative. *Pediatr Rheumatol Online J*. 2020;18(1):17.
3. Chiabrando JG, Bonaventura A, Vecchie A, et al. Management of acute and recurrent pericarditis. *J Am Coll Cardiol*. 2020;75(1):76-92.
4. 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.