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

Cryopyrin-Associated Periodic Syndromes, Familial Cold Autoinflammatory Syndrome and Muckle-Wells Syndrome
Arcalyst is indicated for the 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 and older.

Deficiency of IL-1 Receptor Antagonist
Arcalyst is indicated for the maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA) in adults and pediatric patients weighing at least 10 kg.

Recurrent Pericarditis
Arcalyst is indicated for the treatment of recurrent pericarditis (RP) and reduction in risk of recurrence in adults and pediatric patients 12 years and older.

Coverage Guidelines

Cryopyrin-Associated Periodic Syndromes including Familial Cold Autoinflammatory Syndrome, Muckle-Wells Syndrome, and Neonatal Onset Multisystem Inflammatory Disease, or Chronic Infantile Neurological Cutaneous and Articular Syndrome

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

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

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

- Has been receiving Arcalyst therapy for at least 6 months *(Note: A patient who has received < 6 months of therapy, or who is restarting therapy with Arcalyst, is reviewed under initial authorization criteria);*
- Meets one of the following;
    - Has experienced an improvement in at least one objective measure compared to baseline (e.g., resolution of fever, improvement in rash or skin manifestations, improvement of serum markers [e.g., C-reactive protein,

© 2024 eviCore healthcare. All rights reserved.

amyloid A], reduction in proteinuria, and/or stabilization of serum creatinine); OR
    - Has experienced an improvement in at least one symptom compared to baseline (e.g., less joint pain/tenderness, decreased fatigue improved function or activities of daily living).

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

The individual must meet **all** of the following criteria for re-authorization:
- Has been receiving Arcalyst therapy for at least 6 *months (Note: A patient who has received < 6 months of therapy, or who is restarting therapy with Arcalyst, is reviewed under initial authorization criteria);*
- 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], reduction in proteinuria, and/or stabilization of serum creatinine); OR
    - Has experienced an improvement in at least one symptom compared with baseline (e.g., less joint pain/tenderness, stiffness, or swelling, improvement of skin or bone symptoms).

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;
- Has recurrent pericarditis;
- Prior to starting treatment with Arcalyst, had a history of at least three episodes of pericarditis;
- Arcalyst is prescribed by or in consultation with a cardiologist or rheumatologist;
- Meets one of the following:

 © 2024 eviCore healthcare. All rights reserved.

- Is receiving standard treatment for the current episode (e.g., nonsteroidal anti-inflammatory drugs [NSAIDS], colchicine, and/or systemic corticosteroids); OR
- Has a contraindication to standard treatment (e.g., NSAIDS, colchicine, and/or systemic corticosteroids).

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

- Has been receiving Arcalyst therapy for at least 3 months *(Note: A patient who has received < 3 months of therapy, or who is restarting therapy with Arcalyst, is reviewed under initial authorization criteria);*
- 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).

Approval duration (initial): 3 months
Approval duration (renewal): 12 months

Dosing Recommendations

For adult patients 18 years and older:
For CAPS, FCAS, MWS and RP: 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.

For pediatric patients aged 12 to 17 years:
For CAPS, FCAS, MWS, and RP: 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: Pediatric patients weighing ≥ 10 kg: The recommend dose is 4.4 mg/kg, up to a maximum of 320 mg, administered via subcutaneous injection once weekly.

References

1. Arcalyst® for injection [prescribing information]. Tarrytown, NY: Regeneron Pharmaceuticals; May 2021.
2. Chiabrando JG, Bonaventura A, Vecchie A, et al. Management of acute and recurrent pericarditis. *J Am Coll Cardiol*. 2020;75(1):76-92.
3. 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.

 © 2024 eviCore healthcare. All rights reserved.