CareSource Rxinnovations

PHARMACY POLICY STATEMENT
Georgia Medicaid

DRUG NAME	Arcalyst (rilonacept)
BENEFIT TYPE	Pharmacy
STATUS	Prior Authorization Required

Arcalyst is an interleukin 1 (IL-1) antagonist indicated for Cryopyrin-Associated Periodic Syndromes (CAPS), Deficiency of IL-1 Receptor Antagonist (DIRA), and recurrent pericarditis.
CAPS refer to rare genetic syndromes generally caused by mutations in the NLRP-3 [Nucleotide-binding domain, leucine rich family (NLR), pyrin domain containing 3] gene (also known as Cold-Induced Auto-inflammatory Syndrome-1 [CIAS1]). Mutations in NLRP-3 result in an overactive inflammasome leading to an excessive release of activated IL-1β that drives inflammation.
DIRA is an auto-inflammatory, autosomal recessive disorder caused by loss of function mutations in the IL1RN gene, which encodes IL-1 receptor antagonist (IL-1ra), resulting in unopposed signaling of the proinflammatory cytokines IL-1α and IL-1β through the IL-1 receptor.
Interleukin-1 (IL-1) is a key cytokine that mediates the pathophysiology of many inflammatory processes, and it has also been implicated as a causative factor in pericarditis.

Arcalyst (rilonacept) will be considered for coverage when the following criteria are met:

Cryopyrin-Associated Periodic Syndromes (CAPS)

For **initial** authorization:

1. Member is at least 12 years of age; AND
2. Medication must be prescribed by or in consultation with a rheumatologist or other specialist familiar with CAPS; AND
3. Member has a diagnosis of Familial Cold Auto-Inflammatory Syndrome (FCAS) or Muckle-Wells Syndrome (MWS); AND
4. Member has elevated inflammatory markers (e.g. serum levels of amyloid A, C-reactive protein, erythrocyte sedimentation rate); AND
5. Member displays symptoms of CAPS (e.g. skin rash, musculoskeletal pain, central nervous system manifestations, hearing loss, conjunctivitis, cold/stress-triggered flares); AND
6. Member has had a negative tuberculosis test within the past 12 months.
7. **Dosage allowed/Quantity limit:**
   Adults: loading dose, 320 mg SUBQ (160 mg at 2 different sites); then 160 mg SUBQ once weekly.
   Pediatric: (12 to 17 years of age) loading dose, 4.4 mg/kg SUBQ (MAX of 320 mg) as 1 or 2 injections with a MAX volume of 2 mL; then 2.2 mg/kg (MAX 160 mg) SUBQ once weekly.
   Quantity limit (maintenance): 4 vials per 28 days (4 doses). Note: Each vial is 220 mg.

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:
1. Chart notes demonstrate positive clinical response including decreased inflammatory marker values and symptom improvement.

If all the above requirements are met, the medication will be approved for an additional 12 months.

Deficiency of IL-1 Receptor Antagonist (DIRA)

For **initial** authorization:
1. Medication must be prescribed by or in consultation with a rheumatologist, dermatologist, or geneticist; AND
2. Member has a diagnosis of DIRA confirmed by ALL of the following:
   a) Genetic testing shows IL1RN mutation,
   b) Member has baseline symptoms of skin and/or bone inflammation,
   c) Inflammatory markers (erythrocyte sedimentation rate [ESR], C-reactive protein [CRP]) are elevated at baseline; AND
3. Member has had a negative tuberculosis test within the past 12 months.
4. **Dosage allowed/Quantity limit:**
   Adults: 320 mg (160 mg at 2 different sites on the same day) subQ once weekly
   Pediatric patients weighing 10 kg or more: 4.4 mg/kg subQ once weekly in 1 or 2 injections (if 2 injections, administer at 2 different sites on the same day); MAX dosage, 320 mg
   Quantity limit: 8 vials per 28 days (4 doses). Note: Each vial is 220 mg.

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:
1. Must demonstrate sustained positive clinical response to therapy such as inflammatory remission, resolution of skin and/or bone symptoms, normalization of ESR and/or CRP.

If all the above requirements are met, the medication will be approved for an additional 12 months.

Recurrent Pericarditis

For **initial** authorization:
1. Member is at least 12 years of age; AND
2. Drug is prescribed by or in consultation with a cardiologist; AND
3. Member has a diagnosis of recurrent pericarditis, presenting with at least the 3rd episode of acute pericarditis; AND
4. Member's C-reactive protein (CRP) level is equal to or greater than 1 mg/dL; AND
5. Member has tried and failed ALL of the following (in combination):
   a) Aspirin or Nonsteroidal Anti-inflammatory Drug (NSAID; e.g., ibuprofen, indomethacin)
   b) Colchicine
   c) Low-dose corticosteroid; AND
6. Member has had a negative tuberculosis test within the past 12 months.
7. **Dosage allowed/Quantity limit:**
   Adults: Loading dose, 320 mg SUBQ (160 mg at 2 different sites); then 160 mg SUBQ once weekly
   Pediatrics: (12 to 17 years) Loading dose, 4.4 mg/kg SUBQ (MAX of 320 mg) as 1 or 2 injections with a MAX volume of 2 mL; then 2.2 mg/kg (MAX 160 mg) SUBQ once weekly.
   Quantity limit (maintenance): 4 vials per 28 days (4 doses). Note: Each vial is 220 mg.

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:
1. Member has a documented clinical response to treatment such as decreased recurrence of pericarditis, significantly improved chest pain, and normalized inflammatory markers (e.g., CRP).

If all the above requirements are met, the medication will be approved for an additional 12 months.

CareSource considers Arcalyst (rilonacept) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
06/11/2021	New policy for Arcalyst created.
02/18/2022	Annual review; no updates.
08/09/2024	CAPS, DIRA: Updated refs. CAPS, Pericarditis: Corrected QL from 8 vials/28 days to 4 vials/28 days (maintenance). Pericarditis: Added aspirin as option vs NSAID; changed steroid from an option to a requirement to be used along with previous steps and specified low dose (Adler 2015); added decreased recurrence to reauth.

References:

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3. 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):2443-2452. doi:10.1002/art.23687
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7. 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):2921-2964. doi:10.1093/eurheartj/ehv318
8. 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. doi:10.1016/j.jacc.2019.11.021

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