	Must use valid NDC
BENEFIT TYPE	Pharmacy
SITE OF SERVICE ALLOWED	Home
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-1ƅ 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-1Ů and IL-1ƅ 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]TJ 12.0 Ms orage is AND least

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, flor92 (t)-
   manifestations, hearing lo
6. Member has had a negati
7. **Dosage allowed/Quantit**

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 (if administered as 2 injections, then administer at 2 different sites); then 2.2 mg/kg (MAX 160 mg) SUBQ once weekly.
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. 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 **reauthorization**:

1. Member has a documented clinical response to treatment such as 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
6/11/21	New policy for Arcalyst created.

References:

1. Arcalyst (Rilonacept) [package insert]. London, UK; Kiniksa Pharmaceuticals (UK), Ltd.; Revised 03/2021.
2. Aksentijevich I, Masters SL, Ferguson PJ, et al. An autoinflammatory disease with deficiency of the interleukin-1-