

## PHARMACY POLICY STATEMENT

### Marketplace

DRUG NAME	Arcalyst (Rilonacept)
BILLING CODE	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-1β. *W K D W G U L Y H V L Q I O D P P D W L R Q.*

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-1β. *D Q G , / - W K U R X J K W K H , / - 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

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.
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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