PHARMACY POLICY STATEMENT
Ohio Medicaid

<table>
<thead>
<tr>
<th>DRUG NAME</th>
<th>Arcalyst (Rilonacept)</th>
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<tbody>
<tr>
<td>BILLING CODE</td>
<td>J2793</td>
</tr>
<tr>
<td>BENEFIT TYPE</td>
<td>Medical</td>
</tr>
<tr>
<td>STATUS</td>
<td>Prior Authorization Required</td>
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</table>

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 (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 **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 12 years of age or older; 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 Nonsteroidal Anti-inflammatory Drugs (NSAIDs) and colchicine (or corticosteroids if both are contraindicated); 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 (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. 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.

<table>
<thead>
<tr>
<th>DATE</th>
<th>ACTION/DESCRIPTION</th>
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<tbody>
<tr>
<td>6/11/21</td>
<td>New policy for Arcalyst created.</td>
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<tr>
<td>10/06/2022</td>
<td>Updated benefit to medical due to OH single PBM.</td>
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</table>

References:


Effective date: 10/01/2022
Revised date: 10/06/2022

OH-MED-P-366685