

<b>Clinical Policy Title:</b>	riloncept
<b>Policy Number:</b>	RxA.016
<b>Drug(s) Applied:</b>	Arcalyst®
<b>Original Policy Date:</b>	02/07/2020
<b>Last Review Date:</b>	12/11/2025
<b>Line of Business Policy Applies to:</b>	All lines of business (except Medicare)

## Criteria

### I. Initial Approval Criteria

#### A. Cryopyrin-Associated Periodic Syndromes (CAPS) (must meet all):

1. Diagnosis of familial cold autoinflammatory syndrome (FCAS) or Muckle-Wells syndrome (MWS);
2. Documentation of one of the following (a or b):
  - a. For FCAS, classic signs and symptoms (e.g., recurrent, intermittent fever and rash often exacerbated by exposure to generalized cool ambient temperature) AND functional impairment limiting activities of daily living;
  - b. For MWS, classic signs and symptoms (e.g., chronic fever and rash of waxing and waning intensity, sometimes exacerbated with exposure to generalized cool ambient temperature) AND functional impairment limiting activities of daily living.

**Approval Duration:**

**All Lines of Business (except Medicare):** 6 months

#### B. Deficiency of interleukin-1 (IL-1) receptor antagonist (DIRA) (must meet all):

1. Diagnosis of deficiency of interleukin-1 receptor antagonist confirmed by the presence of loss-of-function ILRN mutations;
2. Member is in remission and has been stable for ≥ 6 months.

**Approval Duration:**

**All Lines of Business (except Medicare):** 6 months

#### C. Recurrent Pericarditis (must meet all):

1. Diagnosis of recurrent pericarditis as evidenced by at least 2 episodes that occur a minimum of 4-6 weeks apart;
2. Trial and failure of at least one of the following: non-steroidal anti-inflammatory drugs (e.g., ibuprofen, naproxen), colchicine, or corticosteroids (e.g., prednisone), unless contraindicated or clinically significant adverse effects are experienced.

**Approval Duration:**

**All Lines of Business (except Medicare):** 6 months

### II. Continued Therapy Approval

#### A. All Indications in Section I (must meet all):

1. Auto-approval based on lookback functionality within the past 120 days as a proxy for member responding positively to therapy.

This clinical policy has been developed to authorize, modify, or determine coverage for individuals with similar conditions. Specific care and treatment may vary depending on individual need and benefits covered by the plan. This policy is not intended to dictate to providers how to practice medicine, nor does it constitute a contract or guarantee regarding payment or results. This document may contain prescription brand name drugs that are trademarks of pharmaceutical manufacturers that are not affiliated with RxAdvance.

**Approval Duration**

**All Lines of Business (except Medicare): 12 months**

**References**

- Hoffman HM, Throne ML, Amar NJ, et al. Efficacy and safety of riloncept (interleukin-1 trap) in patients with cryopyrin-associated periodic syndromes: results from two sequential placebo-controlled studies. Arthritis and Rheumatism. 2008;58(8): 2443-2452. Available at: <https://pubmed.ncbi.nlm.nih.gov/18668535/>. Accessed August 28, 2024.

Review/Revision History	Review/Revision Date	P&T Approval Date
Policy established.	01/2020	02/07/2020
Policy updated – added clarifying information regarding dosing and administration.	05/2020	05/20/2020
<p>Policy was reviewed:</p> <ol style="list-style-type: none"> <li>Policy title table was updated: Clinical Policy Title was updated to ‘riloncept’, Drug(s) Applied was updated to ‘Arcalyst®’, Line of business policy applies was updated to All lines of business.</li> <li>Initial and continued approval criteria was added for DIRA.</li> <li>Continued therapy approval criteria II.A.1 and II.B.1 was rephrased to “Currently receiving medication that has been authorized by RxAdvance...”.</li> <li>Commercial approval durations were updated to 6 months from 6 months or to the member’s renewal date, whichever is longer. Approval duration for HIM was removed.</li> <li>References were updated.</li> </ol>	01/19/2021	03/09/2021
<p>Policy was reviewed:</p> <ol style="list-style-type: none"> <li>Initial Approval Criteria: Updated               <ol style="list-style-type: none"> <li>I.B.3 and I.B.4: Age criteria rephrased to be clear on age requirements</li> <li>I.C: Updated to include approval criteria for indication, recurrent pericarditis.</li> </ol> </li> <li>Continued Therapy Approval Criteria: Updated               <ol style="list-style-type: none"> <li>II.A.1, and II.B.1 were rephrased to "Member is currently receiving medication that has been authorized by RxAdvance...".</li> </ol> </li> </ol>	11/18/2021	01/17/2022

<p>b. II.C: Updated to include approval criteria for indication, recurrent pericarditis.</p> <p>3. References were reviewed and updated.</p>		
<p>Policy was reviewed:</p> <ol style="list-style-type: none"> <li>1. Initial Approval Criteria, I.A.2: Updated prescriber criteria from Prescribed by or in consultation with a rheumatologist to Prescribed by or in consultation with a rheumatologist, immunologist, allergist, dermatologist, neurologist or specialist with expertise in management of CAPS.</li> <li>2. Initial Approval Criteria, I.A.4: Updated to include new documentation criteria Documentation of one of the following (a or b):             <ol style="list-style-type: none"> <li>a. For FCAS, classic signs and symptoms e.g., recurrent, intermittent fever and rash often exacerbated by exposure to generalized cool ambient temperature) AND functional impairment limiting activities of daily living;</li> <li>b. For MWS, classic signs and symptoms (e.g., chronic fever and rash of waxing and waning intensity, sometimes exacerbated with exposure to generalized cool ambient temperature) AND functional impairment limiting activities of daily living;</li> </ol> </li> <li>3. Initial Approval Criteria, I.B.1: Updated diagnostic criteria from Diagnosis of deficiency of interleukin-1 receptor antagonist to Diagnosis of deficiency of interleukin-1 receptor antagonist confirmed by the presence of loss-of-function ILRN mutations.</li> <li>4. Initial Approval Criteria, I.B.3: Updated age criteria from Age ≥ 18 years, or if age &lt; 18 years, member’s body weight is ≥ 10 kg to Weight is ≥ 10 kg.</li> <li>5. Initial Approval Criteria, I.B.4: "If switching from another IL-1 blocker, the previous medication has been discontinued and Arcalyst® is started at the time of the next dose" replaced with "Member is in remission and has been stable for ≥ 6 months".</li> </ol>	<p>07/05/2022</p>	<p>10/19/2022</p>

<ul style="list-style-type: none"> <li>6. Initial Approval Criteria, I.C.1: Updated diagnostic criteria from Diagnosis of recurrent pericarditis to Diagnosis of recurrent pericarditis as evidenced by at least 2 episodes that occur a minimum of 4-6 weeks apart.</li> <li>7. Initial Approval Criteria, I.C.4: Updated to include new trial and failure criteria Trial and failure of at least one of the following: non-steroidal anti-inflammatory drugs (e.g., ibuprofen, naproxen), colchicine, or corticosteroids (e.g., prednisone), unless contraindicated or clinically significant adverse effects are experienced.</li> <li>8. Continued Therapy Approval Criteria II.A.3: Updated response to therapy criteria from Member is responding positively to therapy to "Member has experienced disease stability or improvement in clinical symptoms while on therapy as evidenced by (a, b, c, d, e)..."</li> <li>9. References were reviewed and updated.</li> </ul>		
<p>Policy was reviewed.</p>	<p>10/19/2023</p>	<p>10/19/2023</p>
<p>Policy was reviewed:</p> <ul style="list-style-type: none"> <li>1. Removed age restrictions.</li> <li>2. Removed prescriber restrictions.</li> <li>3. Removed dose restrictions.</li> <li>4. Updated diagnosis descriptions.</li> <li>5. Updated Continued therapy approval with auto-approval based on lookback functionality within the past 120 days.</li> <li>6. Removed reauthorization requirement for positive response to therapy.</li> <li>7. Updated approval duration verbiage.</li> <li>8. Reauthorization criteria for all the diagnosis merged under "All Indications in Section I".</li> <li>9. References were reviewed and updated.</li> </ul>	<p>8/28/2024</p>	<p>9/13/2024</p>
<p>Policy reviewed</p>	<p>12/11/2025</p>	<p>12/11/2025</p>

