Clinical Policy Title:	rilonacept
Policy Number:	RxA.16
Drug(s) Applied:	Arcalyst®
Original Policy Date:	02/07/2020
Last Review Date:	03/09/2021
Line of Business Policy Applies to:	All lines of business

Background

Rilonacept (Arcalyst®) is an interleukin-1 inhibitor.

Arcalyst® is indicated for:

- The treatment of cryopyrin-associated periodic syndromes (CAPS), including familial cold autoinflammatory syndrome (FCAS) and Muckle-Wells syndrome (MWS) in adults and children 12 years and older.
- Maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA) in adults and pediatric patients weighing at least 10 kg.

Dosing Information

Drug Name	Indication	Dosing Regimen	Maximum Dose
rilonacept (Arcalyst®)	CAPS (FCAS, MWS)	Age ≥ 18 years: 320 mg SC loading dose followed by 160 mg SC once weekly Age 12 to 17 years: 4.4 mg/kg SC loading dose followed by 2.2 mg/kg SC once weekly	Loading dose: 320 mg/ week; Maintenance dose: 160 mg/ week
	DIRA	4.4 mg/kg once weekly	320 mg/ week

Dosage Forms

- Single-use vial for reconstitution: 220 mg (each reconstituted 2 ml vial delivers 160 mg)

Clinical Policy

Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.

I. Initial Approval Criteria

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

1. Diagnosis of FCAS or MWS;

2. Prescribed by or in consultation with a rheumatologist;
3. Age 12 years or more;
4. Dose does not exceed a loading dose of 320 mg (as two injections on same day at 2 different sites) and once weekly dosing of 160 mg (as a single injection).

Approval Duration:
Commercial: 6 months
Medicaid: 6 months

B. DIRA (must meet all)**:**

1. Diagnosis of deficiency of interleukin-1 receptor antagonist;
2. Prescribed by or in consultation with a rheumatologist or orthopaedist;
3. Age is 18 years or older;
4. For age less than 18 years, member's body weight is 10 kg or more;
5. If switching from another IL-1 blocker, the previous medication has been discontinued and Arcalyst is begun at the time of the next dose;
6. Dose does not exceed 320 mg once weekly (as two injections on same day at two different sites).

Approval Duration:
Commercial: 6 months
Medicaid: 6 months

II. Continued Therapy Approval

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

1. Currently receiving medication that has been authorized by RxAdvance or member has previously met initial approval criteria listed in this policy;
2. Member is responding positively to therapy;
3. If request is for a dose increase, new dose does not exceed once weekly dosing of 160 mg (as a single injection).

Approval Duration
Commercial: 12 months
Medicaid: 12 months

B. DIRA (must meet all)**:**

1. Currently receiving medication that has been authorized by RxAdvance or member has previously met initial approval criteria listed in this policy;
2. Member is responding positively to therapy;
3. If request is for a dose increase, new dose does not exceed once weekly dosing of 320 mg.

Approval Duration
Commercial: 12 months
Medicaid: 12 months

III. Appendices

APPENDIX A: Abbreviation/Acronym Key
CAPS: Cryopyrin-Associated Periodic Syndromes
DIRA: Deficiency of Interleukin-1 Receptor Antagonist
FCAS: Familial Cold Autoinflammatory Syndrome
MWS: Muckle-Wells Syndrome
SC: subcutaneous/subcutaneously

APPENDIX B: Therapeutic Alternatives

Below are suggested therapeutic alternatives based on clinical guidance. Please check drug formulary for preferred agents and utilization management requirements.

Drug Name	Dosing Regimen	Dose Limit/ Maximum Dose
Kineret® (anakinra)	1 to 2 mg/kg SC once daily, initially. If needed, increase by 0.5 to 1 mg/kg increments.	100 mg/day SC

Therapeutic alternatives are listed as Brand name® (generic) when the drug is available by brand name only and generic (Brand name®) when the drug is available by both brand and generic.

APPENDIX C: Contraindications/Boxed Warnings

- Contraindication(s):
  - None reported

- Boxed Warning(s):
  - None reported

APPENDIX D: General Information

- Three related conditions make up the broader disease known as CAPS: FCAS, MWS, and neonatal-onset multisystem inflammatory disease (NOMID), also known as chronic infantile neurologic cutaneous articular syndrome (CINCA). Arcalyst® is not FDA approved for use in patients with NOMID/CINCA.
- Concomitant administration of Arcalyst® with tumor necrosis factor (TNF) inhibitors (e.g., Enbrel, Humira, or Remicade) and interleukin-1 blocking agents (e.g., Kineret) is not recommended because this may increase the risk of serious infections.
- Examples of positive response to therapy include reduction/normalization of: C-reactive protein levels, serum amyloid A levels, flare frequency, or severity and duration of symptoms (e.g., joint pain, rash, fever/chills, eye pain, fatigue).
- Do not initiate treatment with Arcalyst® in patients with active or chronic infections.
- Live vaccines should not be given concurrently with Arcalyst®. All recommended vaccinations should be completed prior to initiation of therapy with Arcalyst®.

References

1. Arcalyst® Prescribing Information. Tarrytown, NY: Regeneron Pharmaceuticals, Inc.; December 2020. Available at: https://www.Arcalyst®.com/. Accessed January 19, 2021.
2. 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 and Rheumatism. 2008;58(8): 2443-2452. Accessed January 19, 2021.
3. Clinical Pharmacology [database online] powered by ClinicalKey. Tampa, FL: Elsevier, 2020. Accessed with subscription at: http://www.clinicalkey.com. Updated January 14, 2020. Accessed January 19, 2021.
4. Rilonacept, Lexi-Drug. Lexicomp. Wolters Kluwer Health, Inc. Riverwoods, IL. Accessed with subscription at: http://online.lexi.com. Accessed January 19, 2021.

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
Policy was reviewed: 1. Policy title table was updated: Clinical Policy Title was updated to 'rilonacept', Drug(s) Applied was updated to 'Arcalyst®', Line of business policy applies was updated to All lines of business. 2. Background was updated: Indication 'Maintenance of remission of DIRA...' was added. 3. Dosing information was added for DIRA. 4. Initial and continued approval criteria was added for DIRA. 5. Continued therapy approval criteria II.A.1 and II.B.1 was rephrased to "Currently receiving medication that has been authorized by RxAdvance…". 6. Commercial approval durations were updated to 6 months from6 months or to the member's renewal date, whichever is longer. Approval duration for HIM was removed. 7. Appendix A was updated for DIRA. 8. Appendix B: Therapeutic Alternatives was added. 9. Appendix D was updated. "Live vaccines.." 10. References were updated.	01/19/2021	03/09/2021