Clinical Policy: Rilonacept (Arcalyst)

Reference Number: PA.CP.PHAR.266
Effective Date: 01/18
Last Review Date: 04/18

Coding Implications
Revision Log

Description
Rilonacept (Arcalyst®) is a dimeric fusion protein consisting of the ligand-binding domains of the extracellular portions of the human interleukin-1 receptor component (IL-1RI) and IL-1 receptor accessory protein (IL-1RAcP) linked in-line to the Fc portion of human IgG1. Rilonacept blocks IL-1β signaling by acting as a soluble decoy receptor that binds IL-1β and prevents its interaction with cell surface receptors. Rilonacept also binds IL-1α and IL-1 receptor antagonist (IL-1ra) with reduced affinity.

FDA approved indication
Arcalyst (rilonacept) is an interleukin-1 blocker 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 and older.

Policy/Criteria
Provider must submit documentation (which may include office chart notes and lab results) supporting that member has met all approval criteria

It is the policy of PA Health and Wellness® that Arcalyst is **medically necessary** when one of the following criteria are met:

I. Initial Approval Criteria

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

1. Diagnosis of Cryopyrin Associated Periodic Syndrome (CAPS) and (a or b):
   a. Familial Cold Auto-inflammatory Syndrome (FCAS);
   b. Muckle-Wells syndrome (MWS);
2. Prescribed by or in consultation with a rheumatologist;
3. Age ≥ 12 years;
4. Dose does not exceed a loading dose of 320mg (as two injections) and once weekly dosing of 160mg (as a single injection).

Approval duration: 6 months

B. Other diagnoses/indications: Refer to PA.CP.PHAR.57 if diagnosis is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized).

II. Continued Approval

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

1. Currently receiving medication via PA Health and Wellness benefit or member has previously met all initial approval criteria or Continuity of Care policy applies;
2. Member is responding positively to therapy (e.g., normalization of CRP improvement in joint pain, rash, feelings of fever/chills, eye redness/pain and fatigue);
3. If request is for a dose increase, new dose does not exceed once weekly dosing of 160 mg (as a single injection).

CLINICAL POLICY
Rilonacept

Approval duration: 12 months

B. Other diagnoses/indications (must meet 1 or 2)**:**

C. Currently receiving medication via Pennsylvania Health and Wellness benefit or the Continuity of Care policy (PA.LTSS.PHAR.01) applies and documentation supports positive response to therapy; or

Approval duration: Duration of request or 6 months (whichever is less); or

1. Refer to PA.CP.PHAR.57 if diagnosis is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized).

III. Appendices/General Information

Appendix A: Abbreviation/Acronym Key

CAPS: Cryopyrin-Associated Periodic Syndromes
CINCA: chronic infantile neurologic cutaneous articular syndrome
CRP: C-reactive protein
FCAS: Familial Cold Autoinflammatory Syndrome
IL-1RI: Human interleukin-1 receptor component
IL-1RAcP: IL-1 receptor accessory protein
IL-1ra: IL-1α and IL-1 receptor antagonist
MWS: Muckle-Wells Syndrome
NOMID: neonatal-onset multisystem inflammatory disease
TNF: tumor necrosis factor

Appendix B: General Information

- Three related conditions make up the broader disease known as CAPS: FCASMWS, 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 TNF inhibitors (such as Enbrel, Humira, or Remicade) and IL-1 blocking agents (such as Kineret) is not recommended because this may increase the risk of serious infections.

IV. Dosage and Administration

Indication	Dosing Regimen	Maximum Dose
CAPS (FCAS, MWS)	18 yrs and older: 320 mg SC loading dose followed by 160 mg SC once weekly 12 to 17 yrs: 4.4 mg/kg SC loading dose (maximum 320 mg) followed by 2.2 mg/kg (maximum of 160 mg) SC once weekly	320 mg loading dose; 160 mg weekly maintenance dose

V. Product Availability

Drug	Availability
Rilonacept (Arcalyst)	Single-use vial: 220 mg

VI. References

1. Arcalyst Prescribing Information. Tarrytown, NY: Regeneron Pharmaceuticals, Inc.; September 2016. Available at https://www.regeneron.com/sites/default/files/Arcalyst_FPI.pdf. Accessed February 1, 2018.
2. Hoffman, HM, Throne ML, Amar NJ, et al. Efficacy and safety of rilonacept (interleukin-1 trap) in patients with cryopyrin-associated periodic syndromes. Arthritis and Rheumatism. 2008;58(8): 2443-2452.

Coding Implications
Codes referenced in this clinical policy are for informational purposes only. Inclusion or exclusion of any codes does not guarantee coverage. Providers should reference the most up-to-date sources of professional coding guidance prior to the submission of claims for reimbursement of covered services.

HCPCS Codes	Description
J2793	Injection, rilonacept, 1 mg

Reviews, Revisions, and Approvals	Date	Approval Date
2Q 2018 annual review: references reviewed and updated.		