Clinical Policy: Rilonacept (Arcalyst)

Reference Number: CP.PHAR.266
Effective Date: 07/16
Last Review Date: 07/17

Coding Implications
Revision Log

See Important Reminder at the end of this policy for important regulatory and legal information.

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 health plans affiliated with Centene Corporation® 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 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 Centene benefit or member has previously met all initial approval criteria;

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).

Approval duration: 12 months

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

1. Currently receiving medication via health plan benefit and documentation supports positive response to therapy.

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

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

III. Diagnoses/Indications for which coverage is NOT authorized:

A. Non-FDA approved indications, which are not addressed in this policy, unless there is sufficient documentation of efficacy and safety according to the off label use policy – CP. PHAR.57 or evidence of coverage documents

IV. 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.

V. 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

VI. Product Availability

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

VII. 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 June 27, 2017.

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
Policy split from CP.PHAR.47 Cryopyrin-Associated Periodic Syndromes (CAPS) Treatments; Updated contraindications per PI.	5/16	07/16
Converted to new template. Section II: added examples of CAPS related symptoms to assess on continued authorization. Removed safety restrictions that are not a black box warning.	07/17	07/17

Important Reminder

This clinical policy has been developed by appropriately experienced and licensed health care professionals based on a review and consideration of currently available generally accepted standards of medical practice; peer-reviewed medical literature; government agency/program approval status; evidence-based guidelines and positions of leading national health professional organizations; views of physicians practicing in relevant clinical areas affected by this clinical policy; and other available clinical information. The Health Plan makes no representations and accepts no liability with respect to the content of any external information used or relied upon in developing this clinical policy. This clinical policy is consistent with standards of medical practice current at the time that this clinical policy was approved. "Health Plan" means a health plan that has adopted this clinical policy and that is operated or administered, in whole or in part, by Centene Management Company, LLC, or any of such health plan's affiliates, as applicable.

The purpose of this clinical policy is to provide a guide to medical necessity, which is a component of the guidelines used to assist in making coverage decisions and administering benefits. It does not constitute a contract or guarantee regarding payment or results. Coverage decisions and the administration of benefits are subject to all terms, conditions, exclusions and limitations of the coverage documents (e.g., evidence of coverage, certificate of coverage, policy,

contract of insurance, etc.), as well as to state and federal requirements and applicable Health Plan-level administrative policies and procedures.

This clinical policy is effective as of the date determined by the Health Plan. The date of posting may not be the effective date of this clinical policy. This clinical policy may be subject to applicable legal and regulatory requirements relating to provider notification. If there is a discrepancy between the effective date of this clinical policy and any applicable legal or regulatory requirement, the requirements of law and regulation shall govern. The Health Plan retains the right to change, amend or withdraw this clinical policy, and additional clinical policies may be developed and adopted as needed, at any time.

This clinical policy does not constitute medical advice, medical treatment or medical care. It is not intended to dictate to providers how to practice medicine. Providers are expected to exercise professional medical judgment in providing the most appropriate care, and are solely responsible for the medical advice and treatment of members. This clinical policy is not intended to recommend treatment for members. Members should consult with their treating physician in connection with diagnosis and treatment decisions.

Providers referred to in this clinical policy are independent contractors who exercise independent judgment and over whom the Health Plan has no control or right of control. Providers are not agents or employees of the Health Plan.

This clinical policy is the property of the Health Plan. Unauthorized copying, use, and distribution of this clinical policy or any information contained herein are strictly prohibited. Providers, members and their representatives are bound to the terms and conditions expressed herein through the terms of their contracts. Where no such contract exists, providers, members and their representatives agree to be bound by such terms and conditions by providing services to members and/or submitting claims for payment for such services.

Note: For Medicaid members, when state Medicaid coverage provisions conflict with the coverage provisions in this clinical policy, state Medicaid coverage provisions take precedence. Please refer to the state Medicaid manual for any coverage provisions pertaining to this clinical policy.