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Section:	Prescription Drugs	**Effective Date:**	April 1, 2024
Subsection:	Analgesics and Anesthetics	**Original Policy Date:**	September 8, 2011
Subject:	Arcalyst	**Page:**	1 of 5

Last Review Date: March 8, 2024

Arcalyst

Description

Arcalyst (rilonacept)

Background
Arcalyst (rilonacept) is indicated for the treatment of Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Auto-Inflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS); for maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA); and for recurrent pericarditis (RP). Arcalyst blocks interleukin-1 which is a signaling protein secreted by certain immune-related cells in the body. Interleukin-1 acts as a messenger to regulate inflammatory responses, but in excess it can be harmful and has been shown to be key in the inflammation seen in CAPS, DIRA and pericarditis (1).

Regulatory Status
FDA-approved indications: Arcalyst (rilonacept) is an interleukin-1 blocker indicated for: (1)

1. Treatment of Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Auto-inflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS) in adults and children 12 and older
2. Maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA) in adults and pediatric patients weighing 10 kg or more
3. Treatment of recurrent pericarditis (RP) and reduction in risk of recurrence in adults and children 12 years and older

Interleukin-1 blockade may interfere with immune response to infections. Serious, life threatening infections have been reported in patients taking Arcalyst. In addition, taking Arcalyst

with tumor necrosis factor (TNF) inhibitors or other interleukin-1 blockers may further increase the risk of serious infections and an increased risk of neutropenia. The concomitant administration of Arcalyst with TNF-blocking agents is not recommended. Discontinue treatment with Arcalyst if patient develops a serious infection. Arcalyst should not be initiated in patients with active or chronic infections (1).

Live vaccines should not be given concurrently with Arcalyst. Prior to initiation of therapy with Arcalyst, patients should receive all recommended vaccinations (1).

The safety and effectiveness of Arcalyst in CAPS/FCAS/MWS and recurrent pericarditis in patients less than 12 years of age and in DIRA pediatric patients weighing less than 10 kg have not been established (1).

Related policies
Ilaris, Kineret

Policy

This policy statement applies to clinical review performed for pre-service (Prior Approval, Precertification, Advanced Benefit Determination, etc.) and/or post-service claims.

Arcalyst may be considered **medically necessary** if the conditions indicated below are met.

Arcalyst may be considered **investigational** for all other indications.

Prior-Approval Requirements

Age 12 years of age or older

Diagnoses

Patient must have **ONE** of the following:

1. Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Auto-inflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS)
2. Recurrent pericarditis (RP)

AND ALL of the following:

1. **NO** concurrent use with a tumor necrosis factor (TNF) inhibitor (e.g., Cimzia, Enbrel, Humira, Remicade, Simponi)
2. **NO** concurrent use with another interleukin-1 receptor antagonist (e.g., Ilaris, Kineret)
3. **NO** evidence of active or chronic infections
4. **NOT** given concurrently with live vaccines

Age No age restriction

Diagnosis

Patient must have the following:

1. Deficiency of Interleukin-1 Receptor Antagonist (DIRA)

AND ALL of the following:

1. Will be used as maintenance of remission
2. Pediatric patients must weigh at least 10 kg
2. **NO** concurrent use with a tumor necrosis factor (TNF) inhibitor (e.g., Cimzia, Enbrel, Humira, Remicade, Simponi)
3. **NO** concurrent use with another interleukin-1 receptor antagonist (e.g., Ilaris, Kineret)
4. **NO** evidence of active or chronic infections
5. **NOT** given concurrently with live vaccines

Prior – Approval *Renewal* Requirements

Same as above

Policy Guidelines

Pre - PA Allowance

None

Prior - Approval Limits

Duration 2 years

Prior – Approval *Renewal* Limits

Same as above

Rationale

Summary
Arcalyst (rilonacept) is an interleukin-1 blocker indicated for the treatment of Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Auto-inflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS) in adults and children 12 and older; for the maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA); and for recurrent pericarditis (RP). The concomitant administration of Arcalyst with TNF-blocking agents or other interleukin-1 blockers is not recommended due to the increased risk of serious infection or neutropenia. Arcalyst should not be initiated in patients with active or chronic infections. Live vaccines should not be given concurrently with Arcalyst. Lifetime treatment is required to maintain the patient in remission (1).

Prior authorization is required to ensure the safe, clinically appropriate, and cost-effective use of Arcalyst while maintaining optimal therapeutic outcomes.

References

1. Arcalyst [package insert]. London, UK: Kiniksa Pharmaceuticals, Ltd.; May 2021.

Policy History

Date	Action
September 2012	Annual editorial and reference update
June 2013	Annual editorial review and reference update
June 2014	Annual editorial review and reference update
June 2015	Annual editorial review and reference update
March 2016	Annual editorial review and reference update Removal of the Tumor Necrosis Factor (TNF) antagonist examples and interleukin-1 receptor antagonist examples Policy number changed from 5.02.04 to 5.70.04
March 2017	Annual review and reference update
March 2018	Annual editorial review
March 2019	Annual review
March 2020	Annual editorial review. Changed approval duration from lifetime to 2 years
January 2021	Addition of indication: maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA)
March 2021	Annual review

April 2021	Addition of indication: recurrent pericarditis. Revised CAPS indication to match package insert. Added examples of TNF antagonists and IL-1 antagonists
June 2021	Annual review
March 2022	Annual review and reference update
March 2023	Annual review. Changed policy number to 5.70.004
March 2024	Annual review

Keywords

This policy was approved by the FEP® Pharmacy and Medical Policy Committee on March 8, 2024 and is effective on April 1, 2024.