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5.70.04

Section:	Prescription Drugs	**Effective Date:**	April 1, 2021
Subsection:	Analgesics and Anesthetics	**Original Policy Date:**	September 8, 2011
Subject:	Arcalyst	**Page:**	1 of 5

Last Review Date: March 12, 2021

Arcalyst

Description

Arcalyst (rilonacept)

Background
Arcalyst is used in the treatment Cryopyrin-Associated Periodic Syndromes (CAPS) disorders, including Familial Cold Auto-Inflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS) in adults and children 12 and under; and for maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA). 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 DIRA and CAPS sufferers with Familial Cold Auto-Inflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS) (1).

Regulatory Status
FDA-approved indication: 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 at least 10 kg

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 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 patients under 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** for the treatment of familial cold auto-inflammatory syndrome form of CAPS or the Muckle-Wells syndrome form of CAPS and in maintenance remission of DIRA 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. Familial Cold Auto-inflammatory Syndrome (FCAS) form of CAPS (Cryopyrin-Associated Periodic Syndromes)

2. Muckle-Wells Syndrome (MWS) form of CAPS (Cryopyrin-Associated Periodic Syndromes), also known as Cold-Induced Auto-inflammatory Syndrome-1

AND ALL of the following:
1. **NO** concurrent use with a Tumor Necrosis Factor (TNF) antagonist
2. **NO** concurrent use with an interleukin-1 receptor antagonist
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
3. **NO** concurrent use with a Tumor Necrosis Factor (TNF) antagonist
4. **NO** concurrent use with an interleukin-1 receptor antagonist
5. **NO** evidence of active or chronic infections
6. **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; and for the maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA). 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]. Tarrytown, NY: Regeneron Pharmaceuticals Inc; December 2020.

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

Keywords

This policy was approved by the FEP® Pharmacy and Medical Policy Committee on March 12, 2021 and is effective on April 1, 2021.