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Section:	Prescription Drugs	**Effective Date:**	July 1, 2022
Subsection:	Analgesics and Anesthetics	**Original Policy Date:**	September 1, 2011
Subject:	Ilaris	**Page:**	1 of 6

Last Review Date: June 16, 2022

Ilaris

Description

Ilaris (canakinumab)

Background
Ilaris (canakinumab) is a recombinant human monoclonal anti-human interleukin-1β (IL-1β) antibody designed to bind selectively to and neutralize the activity of IL-1β, a proinflammatory cytokine. Ilaris is indicated for the treatment of Cryopyrin-Associated Periodic Syndrome (CAPS), Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS), Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD), Familial Mediterranean Fever (FMF), and active Still's disease. Ilaris is given as a subcutaneous injection by a healthcare provider (1).

Regulatory Status
FDA-approved indications: Ilaris is an interleukin-1β blocker indicated for the treatment of: (1)

1. Periodic Fever Syndromes:
    a. Cryopyrin-Associated Periodic Syndromes (CAPS), in adults and children 4 years of age and older including:
    b. Familial Cold Auto-Inflammatory Syndrome (FCAS)
    c. Muckle-Wells Syndrome (MWS)
    d. Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS) in adult and pediatric patients
    e. Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD) in adult and pediatric patients
    f. Familial Mediterranean fever (FMF) in adult and pediatric patients
2. Active Still's disease, including Adult-Onset Still's Disease (AOSD) and Systemic Juvenile Idiopathic Arthritis (SJIA) in patients aged 2 years and older.

Ilaris has been associated with an increased risk of serious infections. Physicians should exercise caution when administering Ilaris to patients with infections, a history of recurring infections or underlying conditions which may predispose them to infections. Discontinue treatment with Ilaris if a patient develops a serious infection. Do not administer Ilaris to patients during an active infection requiring medical intervention (1).

In clinical trials, Ilaris has not been administered concomitantly with tumor necrosis factor (TNF) inhibitors. An increased incidence of serious infections and an increased risk of neutropenia have been associated with administration of another interleukin-1 (IL-1) blocker in combination with TNF inhibitors in another patient population. Use of Ilaris with TNF inhibitors may also result in similar toxicities and is not recommended because this may increase the risk of serious infections. Drugs that affect the immune system by blocking TNF have been associated with an increased risk of new tuberculosis and reactivation of latent tuberculosis (TB). It is possible that use of IL-1 inhibitors such as Ilaris increases the risk of reactivation of tuberculosis or of opportunistic infections. (1).

Live vaccines should not be given concurrently with Ilaris. Prior to initiation of therapy with Ilaris, patients should receive all recommended vaccinations as IL-1 blockade may interfere with immune response to infections (1).

The safety and effectiveness of Ilaris in AOSD/SJIA, TRAPS, HIDS/MKD, and FMF patients under 2 years of age and in CAPS patients under 4 years of age have not been established (1).

Related policies
Arcalyst, 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.

Ilaris may be considered **medically necessary** in patients 4 years of age or older for the treatment of Cryopyrin Associated Periodic Syndromes (CAPS); in patients 2 years of age or older with active Still's disease, Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS), Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD), or Familial Mediterranean Fever (FMF); and if the conditions indicated below are met.

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

Prior-Approval Requirements

Diagnoses

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

Age 2 years of age or older

1. Active Still's disease, including Adult-Onset Still's disease (AOSD) and Systemic Juvenile Idiopathic Arthritis (SJIA)
2. Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS)
3. Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD)
4. Familial Mediterranean Fever (FMF)

Age 4 years of age or older

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

AND NONE of the following:

a. Concurrently using a tumor necrosis factor (TNF) antagonist (e.g., Cimzia, Enbrel, Humira, Remicade, Simponi)
b. Concurrently using another interleukin-1 receptor antagonist (e.g., Arcalyst, Kineret)
c. Evidence of an active infection requiring medical intervention

Prior – Approval *Renewal* Requirements

Diagnoses

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

Age 2 years of age or older

1. Still's disease, including Adult-Onset Still's disease (AOSD) and Systemic Juvenile Idiopathic Arthritis (SJIA)
    a. Condition has improved or stabilized while on therapy
2. Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS)
3. Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD)

4. Familial Mediterranean Fever (FMF)

Age 4 years of age or older

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

AND NONE of the following:

a. Concurrently using a tumor necrosis factor (TNF) antagonist (e.g., Cimzia, Enbrel, Humira, Remicade, Simponi)
b. Concurrently using another interleukin-1 receptor antagonist (e.g., Arcalyst, Kineret)
c. Evidence of an active infection requiring medical intervention

Policy Guidelines

Pre - PA Allowance

None

Prior - Approval Limits

Duration 12 months

Prior – Approval *Renewal* Limits

Duration 18 months

Rationale

Summary

Ilaris (canakinumab) is an interleukin-1β blocker indicated for the treatment of active Still's disease, Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS), Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD), Familial Mediterranean Fever (FMF), and Cryopyrin-Associated Periodic Syndromes (CAPS) including Familial Cold Autoinflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS). Ilaris has been associated with an increased risk of serious infections. Do not administer Ilaris to patients during an active infection requiring medical intervention. Ilaris is given as a subcutaneous injection by a healthcare provider. The safety and effectiveness of Ilaris in

AOSD/SJIA, TRAPS, HIDS/MKD, and FMF patients under 2 years of age and in CAPS patients under 4 years of age have not been established (1).

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

References

1. Ilaris [package insert]. East Hanover, NJ: Novartis Pharmaceuticals Corp; September 2020.

Policy History

Date	Action
September 2011	Annual editorial and reference update
September 2012	Annual editorial and reference update
June 2013	Annual editorial and reference update A new FDA indication was approved for Active Systemic Juvenile Idiopathic Arthritis (SJIA) and added to criteria.
June 2014	Annual editorial and reference update
March 2016	Annual editorial and reference update Removal of the Tumor Necrosis Factor (TNF) antagonist examples and interleukin-1 receptor antagonist examples Policy code changed from 5.02.09 to 5.70.09
October 2016	Addition of new FDA indications for TRAPS, HIDS/MKD, and FMF
December 2016	Annual review
March 2017	Annual editorial review
March 2018	Annual editorial review and reference update
August 2018	Addition of renewal requirements and changed approval lengths
November 2018	Annual review
March 2019	Annual review
March 2020	Annual review
July 2020	Addition of indication: Active Still's disease, including Adult-Onset Still's disease (AOSD)
September 2020	Annual review
April 2021	Revised CAPS indication to match package insert. Added examples of TNF antagonists and IL-1 antagonists. Added Kineret to Related Policies
June 2021	Annual review and reference update
June 2022	Annual review

Keywords

This policy was approved by the FEP® Pharmacy and Medical Policy Committee on June 16, 2022 and is effective on July 1, 2022.