



Ilaris® (canakinumab)

When requesting Ilaris® (canakinumab), the individual requiring treatment must be diagnosed with an FDA-approved indication and meet the specific coverage guidelines and applicable safety criteria for the covered indication.

FDA-Approved Indications

Ilaris is indicated for the treatment of:

- Cryopyrin-Associated Periodic Syndromes (CAPS) including Familial Cold Autoinflammatory Syndrome, Muckle-Wells Syndrome, and Neonatal Onset Multisystem Inflammatory Disease (NOMID) or Chronic Infantile Neurological Cutaneous and Articular (CINCA) Syndrome
- Tumor Necrosis Factor (TNF) Receptor Associated Periodic Syndrome (TRAPS)
- Hyperimmunoglobulin D (Hyper-IgD) Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD)
- Familial Mediterranean Fever (FMF)
- Active Systemic Juvenile Idiopathic Arthritis (sJIA) in patients 2 years of age and older
- Adult onset Still's Disease

Coverage Guidelines

The individual must meet the following criteria for initial authorization of select indications:

Cryopyrin-Associated Periodic Syndromes (CAPS)

- An individual is 4 years of age or older; **AND**
- Ilaris is prescribed by or in consultation with a rheumatologist, geneticist, allergist/immunologist, or dermatologist.

Familial Mediterranean Fever (FMF)

- Ilaris is prescribed by or in consultation with a rheumatologist, nephrologist, geneticist, gastroenterologist, oncologist, or hematologist.

Hyperimmunoglobulin D syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD) OR Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS)

- Ilaris is prescribed by or in consultation with a rheumatologist, nephrologist, geneticist, oncologist, or hematologist.

Systemic Juvenile Idiopathic Arthritis (sJIA)

- An individual is 2 years of age or older; **AND**
- Ilaris is prescribed by or in consultation with a rheumatologist; **AND**

- An individual meets one of the following conditions:
 - Has tried at least 2 other biologic agents (e.g., tocilizumab, Kineret, abatacept, etanercept, adalimumab, infliximab); **OR**
 - Has features of poor prognosis (e.g., arthritis of the hip, radiographic damage, active systemic disease such as fever, elevated inflammatory markers, or requirement for treatment with systemic glucocorticoids) and has tried Actemra or Kineret; **OR**
 - Has active systemic features with concerns of progression to macrophage activation syndrome (MAS) and has tried Kineret.

Still's Disease

- The individual is 18 years of age or older; **AND**
 - **Note:** If the patient is under 18 years of age, refer to criteria for systemic juvenile idiopathic arthritis
- Ilaris is prescribed by or in consultation with a rheumatologist; **AND**
- An individual meets one of the following conditions:
 - Has tried at least 2 other biologic agents (e.g., tocilizumab, Kineret, abatacept, etanercept, adalimumab, infliximab); **OR**
 - Has features of poor prognosis (e.g., arthritis of the hip, radiographic damage, active systemic disease such as fever, elevated inflammatory markers, or requirement for treatment with systemic glucocorticoids) and has tried Actemra or Kineret; **OR**
 - Has active systemic features with concerns of progression to macrophage activation syndrome (MAS) and has tried Kineret.

The individual must meet the following criteria for reauthorization (all indications):

- An individual is responding to Ilaris therapy.

Approval duration (initial): 3 months (*applies to CAPS, sJIA, and Still's Disease*)

Approval duration (initial): 4 months (*applies to FMF, TRAPS, and HIDS/MKD*)

Approval duration (renewal): 12 months

Dosing Recommendations

Cryopyrin-Associated Periodic Syndromes (CAPS):

The recommended dose is 150 mg for patients with body weight greater than 40 kg and 3 mg/kg for patients with body weight 15 to 40 kg administered subcutaneously every 8 weeks.

Tumor Necrosis Factor (TNF) receptor Associated Periodic Syndrome (TRAPS), Hyperimmunoglobulin D (Hyper-IgD) Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD), Familial Mediterranean Fever (FMF)

For body weight less than or equal to 40 kg: The recommended starting dose is 2 mg/kg administered subcutaneously every 4 weeks. The dose may be increased to 4 mg/kg every 4 weeks if the clinical response is inadequate.

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For body weight greater than 40 kg: The recommended starting dose is 150 mg every 4 weeks. The dose can be increased to 300 mg every 4 weeks if the clinical response is inadequate.

Systemic Juvenile Idiopathic Arthritis and Still's Disease:

The recommended dose is 4 mg/kg, for patients 2 years of age or older with body weight 7.5 kg or greater, administered subcutaneously every 4 weeks. The maximum dosage is 300mg per dose.

References

1. Ilaris® for subcutaneous injection [prescribing information]. East Hanover, NJ: Novartis Pharmaceuticals Corporation; June 2020.
2. Shinkai K, McCalmont TH, Leslie KS. Cryopyrin-associated periodic syndromes and autoinflammation. *Clin Exp Dermatol*. 2008;33:1-9.
3. Ozen S, Hoffman HM, Frenkel J, et al. Familial Mediterranean Fever (FMF) and beyond: a new horizon. Fourth International Congress on the Systemic Autoinflammatory Diseases held in Bethesda, USA; 6-10 November 2005. *Ann Rheum Dis*. 2006;65(7):961-964.
4. Genetics Home Reference. US National Library of Medicine. Available at: <https://ghr.nlm.nih.gov/>. Accessed on March 30, 2020. Search terms: TRAPS, familial Mediterranean fever, MKD.
5. ter Haar NM, Oswald M, Jeyaratnam J, et al. Recommendations for the management of autoinflammatory diseases. *Ann Rheum Dis*. 2015;74(9):1636-1644.
6. Ozen S, Demirkaya E, Erer B, et al. EULAR recommendations for the management of familial Mediterranean fever. *Ann Rheum Dis*. 2016;75(4):644-651.
7. Kimura Y, Morgan DeWitt E, Beukelman T, et al. Adding Canakinumab to the Childhood Arthritis and Rheumatology Research Alliance Consensus Treatment Plans for Systemic Juvenile Idiopathic Arthritis: comment on the article by DeWitt et al. *Arthritis Care Res (Hoboken)*. 2014;66(9):1430-1431.
8. DeWitt EM, Kimura Y, Beukelman T, et al. Consensus treatment plans for new-onset systemic juvenile idiopathic arthritis. *Arthritis Care Res (Hoboken)*. 2012;64(7):1001-1010.
9. Ringold S, Weiss PF, Beukelman T, et al. 2013 update of the 2011 American College of Rheumatology recommendations for the treatment of juvenile idiopathic arthritis: recommendations for the medical therapy of children with systemic juvenile idiopathic arthritis and tuberculosis screening among children receiving biologic medications. *Arthritis Rheum*. 2013;65(10):2499-2512.
10. Novartis. Efficacy, safety and tolerability of ACZ885 in patients with active rheumatoid arthritis. In: ClinicalTrials.gov [Internet]. Bethesda (MD): National Library of Medicine (US). Accessed on March 30, 2020. Available from: <http://www.clinicaltrials.gov/ct2/show/NCT00424346?term=acz885&rank=10>. NLM Identifier: NCT00424346
11. Alten R, Gomez-Reino J, Durez P, et al. Efficacy and safety of the human anti-IL-1 β monoclonal antibody canakinumab in rheumatoid arthritis: results of a 12-week, Phase II, dose-finding study. *BMC Musculoskelet Disord*. 2011;12:153.