

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
- Gout Flares

Coverage Guidelines

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

Cryopyrin-Associated Periodic Syndromes (CAPS)

- Is 4 years of age or older;
- Ilaris is prescribed by or in consultation with a rheumatologist, geneticist, allergist, immunologist, or dermatologist.

Familial Mediterranean Fever (FMF)

- Is 2 years of age or older;
- Has tried colchicine, unless contraindicated;
- Will be taking Ilaris in combination with colchicine, unless colchicine is contraindicated or not tolerated;
- Prior to starting Ilaris, C-reactive protein level is ≥ 10 mg/L OR elevated at least two times the upper limit of normal;

- Has a history of at least one flare per month despite use of colchicine OR has been hospitalized for a severe flare;
- Ilaris is prescribed by or in consultation with a rheumatologist, nephrologist, geneticist, gastroenterologist, oncologist, or hematologist.

Hyperimmunoglobulin D syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD)

- Is 2 years of age or older;
- Prior to starting Ilaris, C-reactive protein level is ≥ 10 mg/L OR elevated at least two times the upper limit of normal;
- Has a history of at least three febrile acute flares within the previous 6-month period OR has been hospitalized for a severe flare;
- Ilaris is prescribed by or in consultation with a rheumatologist, nephrologist, geneticist, oncologist, or hematologist.

Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS)

- Is 2 years of age or older;
- Prior to starting Ilaris, C-reactive protein level is ≥ 10 mg/L OR elevated at least two times the upper limit of normal;
- Has a history of at least six flares per year OR has been hospitalized for a severe flare;
- Ilaris is prescribed by or in consultation with a rheumatologist, nephrologist, geneticist, oncologist, or hematologist.

Systemic Juvenile Idiopathic Arthritis (sJIA)

- Is 2 years of age or older;
- Ilaris is prescribed by or in consultation with a rheumatologist;
- Meets one of the following conditions:
 - o Has tried at least 2 other biologic agents (e.g., tocilizumab, Kineret, abatacept, etanercept, adalimumab, infliximab); OR
 - o 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
 - o Has active systemic features with concerns of progression to macrophage activation syndrome (MAS) and has tried Kineret.

Still's disease, Adult onset

- Is 18 years of age or older;
- Ilaris is prescribed by or in consultation with a rheumatologist;
- Meets one of the following conditions:
 - o Has tried at least 2 other biologic agents (e.g., tocilizumab, Kineret, abatacept, etanercept, adalimumab, infliximab); OR
 - o 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

- o Has active systemic features with concerns of progression to macrophage activation syndrome (MAS) and has tried Kineret.

Note: If the patient is <18 years of age, refer to criteria for systemic juvenile idiopathic arthritis (sJIA)

Gout Flares

- Is 18 years of age or older;
- Meets one of the following:
 - o Has an intolerance, contraindication, or inadequate response to nonsteroidal anti-inflammatory drugs (NSAIDs) and colchicine; OR
 - o Has been previously treated with corticosteroids (oral or injectable) and is unable to be retreated with a repeat course;
- Is currently receiving or will be treated with a concomitant urate lowering agent for the prevention of gout unless contraindicated (e.g., allopurinol, febuxostat, probenecid);
- Ilaris is prescribed by or in consultation with a rheumatologist.

The individual must meet **all** the following criteria for reauthorization (all indications except gout flares):

- Has been established on Ilaris therapy for at least 6 months;
Note: A patient who has received < 6 months of therapy, or who is restarting therapy with Ilaris, is reviewed under initial authorization criteria.
- Meets one of the following;
 - o Has experienced an improvement in at least one objective measure compared with baseline (e.g., resolution of fever, improvement in rash or skin manifestations, improvement of serum markers [e.g., C-reactive protein, amyloid A], reduction in proteinuria, and/or stabilization of serum creatinine); OR
 - o Has experienced an improvement in at least one symptom compared with baseline (e.g., less joint pain/tenderness, stiffness, or swelling, improved function or activities of daily living).

Approval duration (initial): 6 months

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, administered subcutaneously every 8 weeks.

The recommended dose is 2 mg/kg, for patients with body weight 15 to 40 kg, administered subcutaneously every 8 weeks. For children 15 to 40 kg with an inadequate response, the dose can be increased to 3 mg/kg.

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

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For body weight less than or equal to 40 kg: The recommended dose is 2 mg/kg administered subcutaneously every 4 weeks. The dose can be increased to 4 mg/kg every 4 weeks if the clinical response is not adequate.

For body weight greater than 40 kg: The recommended dose is 150 mg administered subcutaneously every 4 weeks. The dose can be increased to 300 mg every 4 weeks if the clinical response is not adequate.

Systemic Juvenile Idiopathic Arthritis, Still's disease

The recommended dose with a body weight greater than or equal to 7.5 kg is 4 mg/kg (with a maximum of 300 mg) administered subcutaneously every 4 weeks.

Gout Flares

The recommended dose is 150 mg administered subcutaneously. May retreat at an interval of at least 12 weeks.

References

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12. FitzGerald JD, Dalbeth N, Mikuls T, et al. 2020 American College of Rheumatology guideline for the management of gout. *Arthritis Care & Research*. 2020;72(6): 744-760.