

## 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)

- An individual is 2 years of age or older; AND
- Has tried colchicine, unless contraindicated; AND
- Will be taking colchicine in combination with Ilaris, unless colchicine is contraindicated or not tolerated; AND
- Prior to starting Ilaris, the individual meets both of the following:
  - C-reactive protein level is  $\geq 10$  mg/L OR elevated at least two times the upper limit of normal; AND
  - Has a history of at least one flare per month despite use of colchicine OR was hospitalized for a severe flare; AND

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

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

- An individual is at least 2 years of age or older; AND
- Prior to starting Ilaris, the individual meets both of the following:
  - C-reactive protein level is  $\geq 10$  mg/L OR elevated at least two times the upper limit of normal; AND
  - Has a history of at least three febrile acute flares within the previous 6-month period OR was hospitalized for a severe flare; AND
- Ilaris is prescribed by or in consultation with a rheumatologist, nephrologist, geneticist, oncologist, or hematologist.

### **Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS)**

- An individual is at least 2 years of age or older; AND
- Prior to starting Ilaris, the individual meets both of the following:
  - C-reactive protein level is  $\geq 10$  mg/L OR elevated at least two times the upper limit of normal; AND
  - Has a history of at least six flares per year OR was hospitalized for a severe flare; AND
- 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

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

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

- Has been established on Ilaris therapy for at least 6 months; AND
- Meets one of the following;
  - Has experienced an improvement in at least one objective measure compared with baseline (e.g., resolution of fever, improvement of serum markers [e.g., C-reactive protein, amyloid A]); OR
  - Has experienced an improvement in at least one symptom compared with baseline (e.g., decreased pain, improved function or activities of daily living).
- Note: A patient who has received < 6 months of therapy, or who is restarting therapy with Ilaris, is reviewed under initial authorization criteria.

**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 no more frequently than every 8 weeks.

The recommended dose is 3 mg/kg, for patients with body weight 15 to 40 kg, administered subcutaneously no more frequently than 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: Administer 4 mg/kg per dose administered subcutaneously no more frequently than every 4 weeks.

For body weight greater than 40 kg: Administer 300 mg per dose administered subcutaneously no more frequently than once every 4 weeks.

### Systemic Juvenile Idiopathic Arthritis, Still's disease

Administer 4 mg/kg to a maximum of 300 mg per dose administered subcutaneously no more frequently than once every 4 weeks.

## References

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