

## PHARMACY POLICY STATEMENT Ohio Medicaid

<b>DRUG NAME</b>	<b>Ilaris (canakinumab)</b>
BILLING CODE	Must use valid NDC
BENEFIT TYPE	Pharmacy
SITE OF SERVICE ALLOWED	Home/Office/Outpatient
STATUS	Prior Authorization Required

Ilaris is an interleukin-1 $\beta$  blocker that was initially approved by the FDA in 2009. It is indicated for the treatment of certain autoinflammatory Periodic Fever Syndromes, and for Still's Disease (Adult-Onset Still's Disease [AOSD] and Systemic Juvenile Idiopathic Arthritis [SJIA]). Ilaris binds to IL-1 $\beta$  and neutralizes its activity by blocking its interaction with IL-1 receptors, but it does not bind IL-1 $\alpha$  or IL-1 receptor antagonist (IL-1ra).

CAPS refer to rare genetic syndromes generally caused by mutations in the NLRP-3 gene (also known as CIAS1). The NLRP-3 gene encodes the protein cryopyrin, an important component of the inflammasome. Cryopyrin controls the activation of IL-1 $\beta$ . Mutations in NLRP-3 result in an overactive inflammasome resulting in excessive release of activated IL-1 $\beta$  that drives inflammation. Still's disease is a severe autoinflammatory disease, driven by innate immunity by means of proinflammatory cytokines such as IL-1 $\beta$ . AOSD and SJIA are thought to represent a continuum of the same disease entity.

Ilaris (canakinumab) will be considered for coverage when the following criteria are met:

### Adult-Onset Still's Disease (AOSD)

For **initial** authorization:

1. Member must have a confirmed diagnosis of active Adult-Onset Still's Disease supported by chart notes; AND
2. Medication must be prescribed by or in consultation with a rheumatologist; AND
3. Must have a negative tuberculosis test within the past 12 months; AND
4. Member has tried and failed, or unable to tolerate **both** of the following (taken together or separately):
  - a) A trial of a corticosteroid (prednisone or methylprednisolone)
  - b) A 2-month trial of a conventional DMARD (e.g., methotrexate, cyclosporine, leflunomide, etc.).
5. **Dosage allowed/Quantity limit:** 4 mg/kg (up to max dose 300 mg) subcutaneously every 4 weeks.

***If all the above requirements are met, the medication will be approved for 6 months.***

For **reauthorization**:

1. Chart notes have been provided that show the member has shown improvement of signs and symptoms of disease.

***If all the above requirements are met, the medication will be approved for an additional 12 months.***

### Cryopyrin-Associated Periodic Syndrome (CAPS)

For **initial** authorization:

1. Member must be 4 years of age or older; AND
2. Medication must be prescribed by or in consultation with a rheumatologist or other specialist familiar with periodic fever syndromes; AND
3. Member must be diagnosed with Familial Cold Autoinflammatory Syndrome (FCAS) or Muckle-Wells Syndrome (MWS); AND
4. Member has elevated inflammatory markers (e.g., serum levels of amyloid A, C-reactive protein, erythrocyte sedimentation rate); AND
5. Member displays symptoms of CAPS (e.g., skin rash, musculoskeletal pain, central nervous system manifestations, hearing loss, conjunctivitis, cold/stress-triggered flares); AND
6. Must have a negative tuberculosis test within the past 12 months.
7. **Dosage allowed/Quantity limit:** 150 mg for body weight > 40 kg; 2 mg/kg for body weight between 15 kg and 40 kg. For children 15 kg to 40 kg with an inadequate response, the dose can be increased to 3 mg/kg. Administer subQ every 8 weeks.

***If all the above requirements are met, the medication will be approved for 6 months.***

For **reauthorization**:

1. Chart notes demonstrate positive clinical response including decreased inflammatory marker values and symptom improvement.

***If all the above requirements are met, the medication will be approved for an additional 12 months.***

## Familial Mediterranean Fever (FMF)

For **initial** authorization:

1. Medication is prescribed by or in consultation with a rheumatologist or other physician experienced with periodic fever syndromes; AND
2. Member has a diagnosis of familial Mediterranean fever; AND
3. Member has had a compliant trial and failure of colchicine at maximal appropriate dose for at least 6 months unless contraindicated or intolerable; AND
4. Must have a negative tuberculosis test within the past 12 months.
5. **Dosage allowed/Quantity limit:** Body weight ≤ 40 kg: starting dose is 2 mg/kg every 4 weeks. The dose can be increased to 4 mg/kg every 4 weeks if the clinical response is not adequate. Body weight > 40 kg: starting dose is 150 mg every 4 weeks. The dose can be increased to 300 mg every 4 weeks if the clinical response is not adequate.

***If all the above requirements are met, the medication will be approved for 6 months.***

For **reauthorization**:

1. Chart notes have been provided showing response to therapy such as reduced severity and/or frequency of flares.

***If all the above requirements are met, the medication will be approved for an additional 12 months.***

## Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD)

For **initial** authorization:

1. Medication is prescribed by or in consultation with a rheumatologist or other physician experienced with periodic fever syndromes; AND
2. Member has a diagnosis of HIDS/MKD; AND
3. Must have a negative tuberculosis test within the past 12 months.
4. **Dosage allowed/Quantity limit:** Body weight  $\leq$  40 kg: starting dose is 2 mg/kg every 4 weeks. The dose can be increased to 4 mg/kg every 4 weeks if the clinical response is not adequate. Body weight  $>$  40 kg: starting dose is 150 mg every 4 weeks. The dose can be increased to 300 mg every 4 weeks if the clinical response is not adequate.

***If all the above requirements are met, the medication will be approved for 6 months.***

For **reauthorization**:

1. Chart notes have been provided showing response to therapy such as reduced severity and/or frequency of flares.

***If all the above requirements are met, the medication will be approved for an additional 12 months.***

## Systemic Juvenile Idiopathic Arthritis (SJIA)

For **initial** authorization:

1. Member must be 2 years of age or older; AND
2. Member must have a documented negative TB test (i.e., tuberculosis skin test (PPD), an interferon-release assay (IGRA)) within 12 months prior to starting therapy; AND
3. Medication must be prescribed by a rheumatologist; AND
4. Member must have active systemic juvenile idiopathic arthritis, as indicated by arthritis involving two or more joints AND **one** or more of the following:
  - a) Evanescent erythematous rash;
  - b) Fever for at least two weeks
  - c) Generalized lymphadenopathy;
  - d) Hepatomegaly or splenomegaly;
  - e) Pericarditis, pleuritis, or peritonitis; AND
5. Member must have inadequate response to ALL of the following:
  - a) Glucocorticoid injection;
  - b) Methotrexate;
  - c) NSAIDs after a 12-week trial.
6. **Dosage allowed/Quantity limit:** 4 mg/kg (with a maximum of 300 mg) for members with a body weight  $\geq$  7.5 kg. Administer subcutaneously every 4 weeks.

***If all the above requirements are met, the medication will be approved for 12 months.***

For **reauthorization**:

1. Must have been retested for TB with a negative result within the past 12 months; AND
2. Member must be in compliance with all other initial criteria; AND
3. Chart notes have been provided that show the member has shown improvement of signs and symptoms of disease.

***If all the above requirements are met, the medication will be approved for an additional 12 months.***

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

For **initial** authorization:

1. Medication is prescribed by or in consultation with a rheumatologist or other physician experienced with periodic fever syndromes; AND
2. Member has a diagnosis of TRAPS; AND
3. Must have a negative tuberculosis test within the past 12 months.
4. **Dosage allowed/Quantity limit:** Body weight  $\leq$  40 kg: starting dose is 2 mg/kg every 4 weeks. The dose can be increased to 4 mg/kg every 4 weeks if the clinical response is not adequate. Body weight  $>$  40 kg: starting dose is 150 mg every 4 weeks. The dose can be increased to 300 mg every 4 weeks if the clinical response is not adequate.

***If all the above requirements are met, the medication will be approved for 6 months.***

For **reauthorization**:

1. Chart notes have been provided showing response to therapy such as reduced severity and/or frequency of flares.

***If all the above requirements are met, the medication will be approved for an additional 12 months.***

**CareSource considers Ilaris (canakinumab) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.**

DATE	ACTION/DESCRIPTION
05/09/2017	New policy for Ilaris created. Policy SRx-0042 archived. For CAPS diagnosis: laboratory evidence requirement of a genetic mutation added. Diagnoses of TRAPS, HIDS/MKD and FMF were added. List of diagnoses considered not medically necessary added.
07/14/2017	Documentation of negative TB test was added to all diagnosis.
03/20/2019	TB test allowed to be done within 12 months prior to initiation of therapy; chest x-ray option removed.
09/29/2020	New diagnosis of Adult Onset Still's Disease added. Status corrected.
06/15/2021	At end of policy, replaced specific list of excluded diseases with general statement. CAPS: Updated references. Removed genetic test requirement (mutation not present in many patients), added biomarker and symptoms instead. Reduced initial approval duration from 12 months to 6 months, should see response much sooner. Specified renewal criteria. FMF: Updated references. Added specialist. Added diagnosis. Removed baseline PGA score. Removed CRP level. Removed minimum number of flares. Added trial of colchicine per guidelines. Specified renewal criteria. HIDS/MKD: Updated references. Added specialist. Added diagnosis. Removed baseline PGA score. Removed CRP level. Removed minimum number of flares. Reduced initial approval duration. Specified renewal criteria. TRAPS: Updated references. Added specialist. Added diagnosis. Removed baseline PGA score. Removed CRP level. Removed minimum number of flares. Reduced initial approval duration. Specified renewal criteria.
02/18/2022	Transferred to new template. AOSD: Added new reference. Changed wording of TB test requirement. Removed meet initial criteria from reauth section.

References:

OH-MED-P-366685

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