

**Ilaris (canakinumab)**  
**Effective 07/01/2023**

<b>Plan</b>	<input checked="" type="checkbox"/> MassHealth UPPL <input type="checkbox"/> Commercial/Exchange	<b>Program Type</b>	<input checked="" type="checkbox"/> Prior Authorization
<b>Benefit</b>	<input checked="" type="checkbox"/> Pharmacy Benefit <input checked="" type="checkbox"/> Medical Benefit (NLX)		<input type="checkbox"/> Quantity Limit <input type="checkbox"/> Step Therapy
<b>Specialty Limitations</b>	This medication has been designated specialty and must be filled at a contracted specialty pharmacy.		
<b>Contact Information</b>	<b>Specialty Medications</b>		
	All Plans	Phone: 866-814-5506	Fax: 866-249-6155
	<b>Non-Specialty Medications</b>		
	MassHealth	Phone: 877-433-7643	Fax: 866-255-7569
	Commercial	Phone: 800-294-5979	Fax: 888-836-0730
	Exchange	Phone: 855-582-2022	Fax: 855-245-2134
	<b>Medical Specialty Medications (NLX)</b>		
	All Plans	Phone: 844-345-2803	Fax: 844-851-0882
<b>Exceptions</b>	N/A		

### Overview

Ilaris® (canakinumab) is a human monoclonal IL-1 $\beta$  antibody that is indicated for Active Still's disease (including Adult-Onset Still's Disease [AOSD] and Systemic Juvenile Idiopathic Arthritis [SJIA]), Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Auto-inflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS), Familial Mediterranean fever (FMF), Mevalonate kinase deficiency (MKD); also known as Hyperimmunoglobulin D syndrome (HIDS) and Tumor necrosis factor receptor associated periodic syndrome (TRAPS).

### Coverage Guidelines

Authorization may be reviewed on a case by case basis for members who are new to the plan currently receiving treatment with requested medication excluding when the product is obtained as samples or via manufacturer's patient assistance programs.

#### OR

Authorization may be granted for members when all the following criteria are met, and documentation is provided:

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

ALL of the following:

1. Diagnosis of adult-onset Still's disease
2. Member is  $\geq$  2 years of age
3. Paid claims or physician attestation of inadequate response or adverse reaction to ONE or contraindication to ALL corticosteroids

4. Paid claims or physician attestation of inadequate response, adverse reaction, or contraindication to Kineret® (anakinra)
5. Appropriate dosing

**Cryopyrin-Associated Periodic Syndromes, including: Familial cold autoinflammatory syndrome (FCAS) and Muckle-Wells syndrome (MWS)**

**ALL** of the following:

1. Diagnosis of one of the following:
  - a. Familial cold autoinflammatory syndrome (FCAS)
  - b. Muckle-Wells syndrome (MWS)
2. Member is  $\geq 4$  years of age
3. **ONE** of the following:
  - a. Evidence of symptoms indicative of the disease
  - b. Confirmation of diagnosis through genetic testing
4. Appropriate dosing

**Familial Mediterranean fever (FMF)**

**ALL** of the following:

1. Diagnosis Familial Mediterranean Fever
2. Paid claims or physician attestation of inadequate response, adverse reaction, or contraindication to colchicine
3. **ONE** of the following:
  - a. Evidence of symptoms indicative of the disease (see Appendix for examples)
  - b. Confirmation of diagnosis through genetic testing
4. Appropriate dosing

**Hyperimmunoglobulin D syndrome (HIDS)/Mevalonate kinase deficiency (MKD)**

**ALL** of the following:

1. Diagnosis of one of the following:
  - a. Hyperimmunoglobulin D syndrome
  - b. Mevalonate kinase deficiency
2. **ONE** of the following:
  - a. Evidence of symptoms indicative of the disease (see Appendix for examples)
  - b. Confirmation of diagnosis through genetic testing
3. Appropriate dosing

**Systemic juvenile idiopathic arthritis (SJIA)**

**ALL** of the following:

1. Diagnosis of systemic juvenile idiopathic arthritis
2. Member is  $\geq 2$  years of age
3. Paid claims or physician attestation of inadequate response or adverse reaction to **ONE** or a contraindication to **ALL** corticosteroids
4. Paid claims or physician attestation of inadequate response, adverse reaction, or contraindication to Kineret® (anakinra)
5. Appropriate dosing

**Tumor necrosis factor receptor associated periodic syndrome (TRAPS)**

**ALL** of the following:



1. Diagnosis of tumor necrosis factor receptor associated periodic syndrome
2. ONE of the following:
  - a. Evidence of symptoms indicative of the disease (see Appendix for examples)
  - b. Confirmation of diagnosis through genetic testing
3. Appropriate dosing

**Continuation of Therapy**

Reauthorization by prescriber will infer a positive response to therapy and dosing is appropriate.

**Limitations**

1. Initial approvals will be granted for 6 months
2. Reauthorizations will be granted for 12 months
3. Dosing information:

Ilaris® (canakinumab)  Single-use 1 mL vial containing 150 mg of solution QTY 1 mL=150 mg	<b>Pediatric Dosing</b>  <b>CAPS (includes FCAS, MWD) (≥4 years of age):</b> <i>Members weighing &gt;40 kg:</i> SQ injection: 150 mg every eight weeks <i>Members weighing &gt;15 kg and ≤40 kg:</i> SQ injection: 2 mg/kg every eight weeks; dose can be increased to 3 mg/kg every eight weeks if response is inadequate	<b>Adult Dosing</b>  <b>CAPS (includes FCAS, MWD):</b> <i>Members weighing &gt;40 kg:</i> SQ injection: 150 mg every eight weeks <i>Members weighing &gt;15 kg and ≤40 kg:</i> SQ injection: 2 mg/kg every eight weeks; dose can be increased to 3 mg/kg every eight weeks if response is inadequate
	<b>FMF, HIDS/MKD and TRAPS:</b> <i>Members weighing &gt;40 kg:</i> SQ injection: 150 mg every four weeks; dose can be increased to 4 mg/kg every four weeks if response is inadequate <i>Members weighing ≤40 kg:</i> SQ injection: 2 mg/kg every four weeks; dose can be increased to 4 mg/kg every four weeks if response is inadequate	<b>FMF, HIDS/MKD and TRAPS:</b> <i>Members weighing &gt;40 kg:</i> SQ injection: 150 mg every four weeks; dose can be increased to 4 mg/kg every four weeks if response is inadequate <i>Members weighing ≤40 kg:</i> SQ injection: 2 mg/kg every four weeks; dose can be increased to 4 mg/kg every four weeks if response is inadequate
<b>AOSD and SJIA (≥2 years of age):</b> <i>Members weighing ≥7.5 kg:</i> 4 mg/kg (maximum: 300 mg) every four weeks via SQ injection	<b>AOSD and SJIA:</b> 4 mg/kg (maximum: 300 mg) every four weeks via SQ injection	

**Appendix**

**Disease Symptoms**

**Tumor necrosis factor receptor associated periodic syndrome (TRAPS):** characterized by recurrent fevers over months or years every five to six weeks, focal myalgias, conjunctivitis, periorbital edema,



abdominal pain, monoarticular arthritis, and rash.

**Recurrent pericarditis:** The predominant feature of recurrent pericarditis is usually pleuritic chest pain (often sharp, worse when lying flat, and alleviated when leaning forward), which may follow exertion. Some members may also report dyspnea or malaise.

**Familial cold autoinflammatory syndrome (FCAS):** mildest phenotype; characterized by intermittent cold-induced rash with fever and arthralgia.

**Muckle-Wells syndrome (MWS):** characterized by recurrent episodes of fever and urticaria associated with joint and ocular manifestations, deafness and reactive amyloid A amyloidosis.

**Neonatal onset multisystem inflammatory disease (NOMID):** most severe spectrum of the disease; characterized by erythematous rash resembling urticaria, fever, impaired growth, chronic meningitis, hearing loss, uveitis, lymphadenopathy and hepatosplenomegaly. Limb and joint pain is common.

**Hyperimmunoglobulin D syndrome (HIDS)/Mevalonate kinase deficiency (MKD):** characterized by episodic attacks of fever lasting three to seven days are accompanied, in most cases, by chills, cervical lymphadenopathy, abdominal pain, vomiting, and/or diarrhea. Other symptoms include headache, arthralgias/arthritis, aphthous ulceration, a pleomorphic rash, and splenomegaly. Elevated levels of immunoglobulin D (IgD) are often present.

#### References

1. Ilaris® [package insert]. Hanover (NJ): Novartis Pharmaceuticals Corp. 2020 Sep

#### Review History

06/14/2023 - Created for P&T in order to match MH UPPL. Effective 7/1/23.

