Corporate Medical Policy: Canakinumab (Ilaris®) **"Notification"** **POLICY EFFECTIVE JULY 1, 2026**

Restricted Product(s):

- canakinumab (Ilaris®) subcutaneous injection for administration by a healthcare professional

FDA Approved Use:

- For the treatment of autoinflammatory Periodic Fever Syndromes:
  - Cryopyrin-Associated Periodic Syndromes (CAPS), in adults and pediatric patients 4 years of age and older, including:
    - Familial Cold Auto-inflammatory Syndrome (FCAS)
    - Muckle-Wells Syndrome (MWS)
  - Tumor Necrosis Factor (TNF) Receptor Associated Periodic Syndrome (TRAPS) in adult and pediatric patients
  - Hyperimmunoglobulin D (Hyper-IgD) Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD) in adult and pediatric patients
  - Familial Mediterranean Fever (FMF) in adult and pediatric patients
- For the treatment of active Still's disease, including Adult-Onset Still's Disease (AOSD) and Systemic Juvenile Idiopathic Arthritis (SJIA) in patients 2 years of age and older
- For the symptomatic treatment of adult patients with gout flares in whom non-steroidal anti-inflammatory drugs (NSAIDs) and colchicine are contraindicated, are not tolerated, or do not provide an adequate response, and in whom repeated courses of corticosteroids are not appropriate

Criteria for Medical Necessity:

The restricted product(s) may be considered medically necessary when the following criteria are met:

Initial Criteria for Approval:

1. The patient has a diagnosis of **Cryopyrin-Associated Periodic Syndrome (CAPS)** (including Familial Cold Auto-Inflammatory Syndrome [FCAS] or Muckle-Wells Syndrome [MWS]); **AND**
   a. The patient is 4 years of age or older; **AND**
   b. The diagnosis has been confirmed by ONE of the following:
      i. Genetic testing demonstrating presence of a mutation in the Cold-Induced Auto-Inflammatory Syndrome 1 (*CIAS1*) gene, also known as *NLRP3* **[medical record documentation required]**; **OR**
      ii. The patient has BOTH of the following **[medical record documentation required]**:

1. Elevated baseline serum inflammatory markers (i.e., C-reactive protein [CRP] and/or serum amyloid A [SAA]); **AND**
2. At least TWO of the following signs or symptoms for CAPS:
    a. Urticaria-like rash; **OR**
    b. Cold/stress-triggered flares; **OR**
    c. Chronic aseptic meningitis; **OR**
    d. Sensorineural hearing loss; **OR**
    e. Musculoskeletal symptoms (e.g., arthralgia, arthritis, myalgia); **OR**
    f. Skeletal abnormalities (e.g., epiphysial overgrowth/frontal bossing); **OR**

2. The patient has a diagnosis of **Tumor Necrosis Factor (TNF) Receptor Associated Periodic Syndrome (TRAPS)**; **AND**
    a. The patient is 2 years of age or older; **AND**
    b. The diagnosis has been confirmed by genetic testing demonstrating presence of a mutation in the Tumor Necrosis Factor Receptor-1 (TNFR1) gene (*TNFRSF1A*) **[medical record documentation required]**; **AND**
    c. The patient has chronic or recurrent disease, defined as greater than 6 flares per year; **AND**
    d. The patient has documented baseline serum C-reactive protein (CRP) levels **[medical record documentation required]**; **OR**

3. The patient has a diagnosis of **Hyperimmunoglobulin D (Hyper-IgD) Syndrome (HIDS) / Mevalonate Kinase Deficiency (MKD)**; **AND**
    a. The patient is 2 years of age or older; **AND**
    b. The diagnosis has been confirmed by ONE of the following:
        i. Genetic testing demonstrating presence of a mutation in the *MVK* gene **[medical record documentation required]**; **OR**
        ii. The patient has significantly elevated serum IgD levels **[medical record documentation required]**; **AND**
    c. The patient has a history of at least 3 febrile episodes within a 6-month period **[medical record documentation required]**; **AND**
    d. The patient has documented baseline serum C-reactive protein (CRP) levels **[medical record documentation required]**; **OR**

4. The patient has a diagnosis of **Familial Mediterranean Fever (FMF)**; **AND**
    a. The patient is 2 years of age or older; **AND**
    b. The diagnosis has been confirmed by genetic testing demonstrating at least one known *MEFV* exon 10 mutation **[medical record documentation required]**; **AND**
    c. The patient has active disease, defined as at least one febrile episode per month; **AND**
    d. ONE of the following:

i. The patient has tried and had an inadequate response with colchicine for at least a 6-month trial **[medical record documentation required]**; **OR**
ii. The patient has an intolerance, FDA labeled contraindication, or hypersensitivity to colchicine **[medical record documentation required]**; **AND**

e. The patient has documented baseline serum C-reactive protein (CRP) levels **[medical record documentation required]**; **OR**

5. The patient has a diagnosis of **active systemic juvenile idiopathic arthritis (SJIA)**; **AND**
   a. The patient is 2 years of age or older; **AND**
   b. The patient has documented active systemic features (e.g., ongoing fever for at least 2 weeks, evanescent erythematous rash, generalized lymphadenopathy, ≥1 joint with active arthritis, hepatomegaly, splenomegaly, serositis); **AND**
   c. ONE of the following:
      i. The patient has tried and had an inadequate response to TWO of the following drug classes used in the treatment of SJIA **[medical record documentation required]**:
         1. DMARDs (i.e., methotrexate, leflunomide) for at least a 3-month trial; **OR**
         2. Systemic corticosteroids (oral or intravenous) for at least a 1-month trial; **OR**
         3. NSAIDs (e.g., ibuprofen, celecoxib) for at least a 1-month trial; **OR**
      ii. The patient has an intolerance, FDA labeled contraindication, or hypersensitivity to ALL of the drug classes listed above (i.e., DMARDs, systemic corticosteroids, NSAIDs) **[medical record documentation required]**; **OR**
      iii. The patient is currently established on a biologic or systemic immunomodulator agent that is FDA approved for the treatment of SJIA (excluding sample use) **[medical record documentation required]**; **AND**
         1. The patient has had positive clinical benefit (e.g., improvement in signs and symptoms, reduction in disease severity, etc.) from use of the biologic or systemic immunomodulator agent **[medical record documentation required]**; **OR**

6. The patient has a diagnosis of **active adult-onset Still's disease (AOSD)**; **AND**
   a. The patient is 18 years of age or older; **AND**
   b. ONE of the following:
      i. The patient has tried and had an inadequate response to at least ONE corticosteroid (e.g., prednisone, methylprednisolone, etc.) for at least a 1-month trial **[medical record documentation required]**; **OR**
      ii. The patient has an intolerance, FDA labeled contraindication, or hypersensitivity to ALL corticosteroids **[medical record documentation required]**; **AND**
   c. ONE of the following:

i. The patient has tried and had an inadequate response to methotrexate for at least a 3-month trial **[medical record documentation required]**; **OR**
ii. The patient has an intolerance, FDA labeled contraindication, or hypersensitivity to methotrexate **[medical record documentation required]**; **OR**

d. The patient is currently established on a biologic or systemic immunomodulator agent that is FDA approved for the treatment of AOSD (excluding sample use) **[medical record documentation required]**; **AND**
   i. The patient has had positive clinical benefit (e.g., improvement in signs and symptoms, reduction in disease severity, etc.) from use of the biologic or systemic immunomodulator agent **[medical record documentation required]**; **OR**

7. The patient has a diagnosis of **gout flares**; **AND**
   a. The patient is 18 years of age or older; **AND**
   b. The patient has experienced greater than or equal to 3 flares in the past 12 months **[medical record documentation required]**; **AND**
   c. ONE of the following **[medical record documentation required]**:
      i. The patient has tried and had an inadequate response to ONE non-steroidal anti-inflammatory drug (NSAID); **OR**
      ii. The patient has an intolerance or hypersensitivity to ONE non-steroidal anti-inflammatory drug (NSAID); **OR**
      iii. The patient has an FDA labeled contraindication to ALL non-steroidal anti-inflammatory drugs (NSAIDs); **AND**
   d. ONE of the following **[medical record documentation required]**:
      i. The patient has tried and had an inadequate response to colchicine; **OR**
      ii. The patient has an intolerance or hypersensitivity to colchicine; **OR**
      iii. The patient has an FDA labeled contraindication to colchicine; **AND**
   e. Repeated courses of corticosteroids are not appropriate for the patient **[medical record documentation required]**; **AND**

8. The patient will NOT be receiving canakinumab (Ilaris®) in combination with another biologic immunomodulator agent; **AND**
9. The prescriber is a specialist in the area of the patient's diagnosis (e.g., allergist, immunologist, pediatrician, rheumatologist) or has consulted with a specialist in the area of the patient's diagnosis; **AND**
10. The requested quantity does NOT exceed the maximum units allowed for the duration of approval (see table below); **AND**
11. For requests for injection or infusion administration of the requested medication in an **inpatient or outpatient hospital setting,** Site of Care Criteria applies (outlined below)*

Duration of Approval:

Gout flares: 84 days (12 weeks)

All other indications: 365 days (1 year)

Continuation Criteria for Approval:

1. The patient was approved through Blue Cross NC initial criteria for approval; **OR**
2. The patient would have met initial criteria for approval at the time they started therapy; **AND**
3. The patient has had clinical benefit while on canakinumab (Ilaris®) therapy (e.g., reduction of inflammatory markers [i.e., CRP, ESR, SAA], improvement in signs and symptoms, reduction in disease severity, etc.); **AND**
4. For patients requiring re-treatment of gout flares, the patient has had at least a 12-week interval since the last dose of canakinumab (Ilaris®) was administered; **AND**
5. The patient will NOT be receiving canakinumab (Ilaris®) in combination with another biologic immunomodulator agent; **AND**
6. The prescriber is a specialist in the area of the patient's diagnosis (e.g., allergist, immunologist, pediatrician, rheumatologist) or has consulted with a specialist in the area of the patient's diagnosis; **AND**
7. The requested quantity does NOT exceed the maximum units allowed for the duration of approval (see table below); **AND**
8. For requests for injection or infusion administration of the requested medication in an **inpatient or outpatient hospital setting,** Site of Care Criteria applies (outlined below)*

Duration of Approval:

Gout flares: 84 days (12 weeks)
All other indications: 365 days (1 year)

FDA Label Reference				
Medication	**Indication**	**Dosing**	**HCPCS**	**Maximum Units***
canakinumab (Ilaris®) subcutaneous (SC) injection	CAPS	Body weight >40 kg: 150 mg SC every 8 weeks Body weight ≥15 kg and ≤40 kg: 2 mg/kg SC every 8 weeks	J0638	1,050

FDA Label Reference				
Medication	**Indication**	**Dosing**	**HCPCS**	**Maximum Units***
		Dosage may be increased to 3 mg/kg SC every 8 weeks in pediatric patients 15 kg to 40 kg with an inadequate response		
	TRAPS, HIDS/MKD, and FMF	Body weight >40 kg: 150 mg SC every 4 weeks; may be increased to 300 mg every 4 weeks if the clinical response is not adequate Body weight ≤40 kg: 2 mg/kg SC every 4 weeks; may be increased to 4 mg/kg every 4 weeks if the clinical response is not adequate		3,900
	Still's disease (AOSD and SJIA)	4 mg/kg (maximum 300 mg) for patients with a body weight ≥7.5 kg SC every 4 weeks		3,900
	Gout flares in adults	150 mg SC. Patients requiring re-treatment should have an interval of at least 12 weeks before a new dose is administered.		150

***Maximum units allowed for duration of approval**

***Site of Care Medical Necessity Criteria**

1. For requests for injection or infusion administration in an **inpatient setting**, the injection or infusion may be given if the above medical necessity criteria are met AND the inpatient admission is NOT for the sole purpose of administering the injection or infusion; **OR**

2. For requests for injection or infusion administration in an **outpatient hospital setting**, the injection or infusion may be given if the above medical necessity criteria are met AND ONE of the following must be met:
    a. History of a severe adverse event following the injection or infusion of the requested medication (i.e., anaphylaxis, seizure, thromboembolism, myocardial infarction, renal failure); **OR**
    b. Conditions that cause an increased risk for severe adverse event (i.e., unstable renal function, cardiopulmonary conditions, unstable vascular access); **OR**
    c. History of mild adverse events that have not been successfully managed through mild pre-medication (e.g., diphenhydramine, acetaminophen, steroids, fluids, etc.); **OR**
    d. Inability to physically and cognitively adhere to the treatment schedule and regimen complexity; **OR**
    e. New to therapy, defined as initial injection or infusion OR less than 3 months since initial injection or infusion; **OR**
    f. Re-initiation of therapy, defined as ONE of the following:
        i. First injection or infusion after 6 months of no injections or infusions for drugs with an approved dosing interval less than 6 months duration; **OR**
        ii. First injection or infusion after at least a 1-month gap in therapy outside of the approved dosing interval for drugs requiring every 6 months dosing duration; **OR**
    g. Requirement of a change in the requested restricted product formulation; **AND**
3. If the Site of Care Medical Necessity Criteria in #1 or #2 above are not met, the injection or infusion will be administered in a **home-based infusion** or physician office setting with or without supervision by a certified healthcare professional.

References: all information referenced is from FDA package insert unless otherwise noted below.

1. 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. American College of Rheumatology. October 2013. *Arthritis & Rheumatism*. 65(10):2499-2512.
2. Romano M, Arici ZS, Piskin D, et al. The 2021 EULAR/American College of Rheumatology Points to Consider for Diagnosis, Management and Monitoring of the Interleukin-1 Mediated Autoinflammatory Diseases: Cryopyrin-Associated Periodic Syndromes, Tumour Necrosis Factor Receptor-Associated Periodic Syndrome, Mevalonate Kinase Deficiency, and Deficiency of the Interleukin-1 Receptor Antagonist. *Arthritis Rheumatol*. 2022 Jul;74(7):1102-1121.

Policy Implementation/Update Information: Criteria and treatment protocols are reviewed annually by the Blue Cross NC P&T Committee, regardless of change. This policy is reviewed in Q1 annually.

July 2026: Criteria change: For CAPS: Added required diagnostic confirmation by either genetic testing or presence of elevated inflammatory markers plus at least two CAP-typical symptoms. For TRAPS: Added required diagnostic confirmation by genetic testing; Added required

presence of chronic or recurrent disease (>6 flares per year) and submission of baseline CRP levels. For HIDS and MKD: Combined criteria into one section; Added required diagnostic confirmation by either genetic testing or presence of significantly elevated serum IgD levels; Added required history of at least 3 febrile episodes within a 6-month period and submission of baseline CRP levels. For FMF: Added required diagnostic confirmation by genetic testing; Added required presence of active disease (1 febrile episode per month) and submission of baseline CRP levels. For SJIA and AOSD: Added allowance for patients currently established on a biologic or systemic immunomodulator agent that is FDA approved for treatment of the requested indication for those who have had positive clinical benefit from use of the biologic or systemic immunomodulator agent; Added defined trial duration for conventional agents (corticosteroids and methotrexate) for AOSD to align with SJIA requirements. Added age requirement for all indications where not already present. Added examples of clinical benefit within continuation criteria. Other minor formatting changes made throughout policy for clarity with no change to intent. **Policy notification given 5/1/2026 for effective date 7/1/2026.**
November 2025: Criteria change: Updated Site of Care medical necessity criteria to add additional bypass for patients with a history of severe adverse events or conditions that cause an increased risk for severe adverse event to align with the Place of Service for Medical Infusions policy for clarity of intent.
September 2025: Criteria change: Adjusted maximum units according to FDA label for clarity. Other minor updates made throughout policy for clarity with no change to policy intent.
March 2024: Criteria change: Add new indication for gout flares with corresponding criteria and dosing table updates.
October 2021: Criteria change: Added Site of Care medical necessity criteria. **Policy notification given 8/2/2021 for effective date 10/1/2021**.
June 2021: Criteria change: Medical record documentation required for trial and failure of conventional agents; removed criterion point regarding medication history indicating use of another biologic immunomodulator agent for the treatment of SJIA; minor updates to language and formatting for clarity.
June 2021: Criteria change: Added maximum units; medical policy formatting change. **Policy notification given 4/16/2021 for effective date 6/16/2021**.
*Further historical criteria changes and updates available upon request from Medical Policy and/or Corporate Pharmacy.