Draft Revised Policy: Do Not Implement

Canakinumab (Ilaris®)

IMPORTANT REMINDER

We develop Medical Policies to provide guidance to Members and Providers. This Medical Policy relates only to the services or supplies described in it. The existence of a Medical Policy is not an authorization, certification, explanation of benefits or a contract for the service (or supply) that is referenced in the Medical Policy. For a determination of the benefits that a Member is entitled to receive under his or her health plan, the Member's health plan must be reviewed. If there is a conflict between the medical policy and a health plan or government program (e.g., TennCare), the express terms of the health plan or government program will govern.

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
   1. Periodic Fever Syndromes:
      a. Cryopyrin-Associated Periodic Syndromes (CAPS)
         Ilaris is indicated for the treatment of Cryopyrin-Associated Periodic Syndromes (CAPS), in adults and children 4 years of age and older including Familial Cold Autoinflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS).
      b. Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS)
         Ilaris is indicated for the treatment of TRAPS in adult and pediatric patients.
      c. Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD)
         Ilaris is indicated for the treatment of HIDS and MKD in adult and pediatric patients.
      d. Familial Mediterranean Fever (FMF)
         Ilaris is indicated for the treatment of FMF in adult and pediatric patients.
   2. Still's disease (Adult-onset Still's Disease [AOSD] and systemic Juvenile Idiopathic Arthritis [sJIA]):
      Ilaris is indicated for the treatment of active Still's disease, including AOSD and sJIA in patients aged 2 years and older.
   3. Gout flares:
      Ilaris is indicated 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.

B. Compendial Use
   Pseudogout

All other indications are considered experimental/investigational and not medically necessary.

II. DOCUMENTATION

Submission of the following information is necessary to initiate the prior authorization review:

A. Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS) and Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD): For initial requests: Chart notes, medical record documentation, or laboratory result (if applicable) indicating number of active flares within the last 6 months and Physician's Global Assessment (PGA) score or C-reactive protein (CRP) level.

B. Familial Mediterranean Fever (FMF): (initial requests only)
   1. Chart notes or medical record documentation indicating number of active flares within the last 6 months.
   2. Laboratory results, chart notes, or medical record documentation of CRP level.
   3. Chart notes, medical record documentation, or claims history supporting previous medications tried (if applicable), including response to therapy. If therapy is not advisable, documentation of clinical reason to avoid therapy.

C. Systemic Juvenile Idiopathic Arthritis (sJIA) and Adult-onset Still's disease (AOSD)
   1. Initial requests: Chart notes, medical record documentation, or claims history supporting previous medications tried, including response to therapy (if applicable).
   2. Continuation requests: Chart notes or medical record documentation supporting positive clinical response.

D. Gout and pseudogout flares: (initial requests only) Chart notes, medical record documentation, or claims history supporting previous medications tried (if applicable), including response to therapy. If therapy is not advisable, documentation of clinical reason to avoid therapy.

III. PRESCRIBER SPECIALTIES

This medication must be prescribed by or in consultation with one of the following:
A. Cryopyrin-associated periodic syndromes (CAPS), TRAPS, HIDS/MKD, and FMF: rheumatologist or immunologist
B. Systemic juvenile idiopathic arthritis (sJIA), AOSD, gout, and pseudogout: rheumatologist

IV. CRITERIA FOR INITIAL APPROVAL

A. Periodic fever syndromes

1. Authorization of 12 months may be granted for members 4 years of age or older for treatment of CAPS when both of the following criteria are met:
   a. Member has a diagnosis of familial cold autoinflammatory syndrome (FCAS) with classic signs and symptoms (i.e., recurrent, intermittent fever and rash that were often exacerbated by exposure to generalized cool ambient temperature) or Muckle-Wells syndrome (MWS) with classic signs and symptoms (i.e., chronic fever and rash of waxing and waning intensity, sometimes exacerbated by exposure to generalized cool ambient temperature).
   b. Member has functional impairment limiting the activities of daily living.

2. Authorization of 12 months may be granted for treatment of TRAPS when both of the following criteria are met:
   a. Member has chronic or recurrent disease activity with active flares within the last 6 months.
   b. Physician's Global Assessment (PGA) score greater than or equal to 2 or C-reactive protein (CRP) greater than 10 mg/L.

3. Authorization of 12 months may be granted for treatment of HIDS/MKD when both of the following criteria are met:
   a. Member has had active flares within the last 6 months.

b. Physician's Global Assessment (PGA) score greater than or equal to 2 or C-reactive protein (CRP) greater than 10 mg/L.

4. Authorization of 12 months may be granted for treatment of FMF when all of the following criteria are met:
   a. Member has active disease with flares within the last 6 months.
   b. C-reactive protein (CRP) greater than 10 mg/L.
   c. Member has had an inadequate response or intolerance to or has a contraindication to colchicine.

B. Systemic juvenile idiopathic arthritis (sJIA)

1. Authorization of 12 months may be granted for members 2 years of age or older who have previously received a biologic indicated for active sJIA.

2. Authorization of 12 months may be granted for members 2 years of age or older for treatment of active sJIA when both of the following criteria are met:
   a. Member has active systemic features (e.g., fever, evanescent rash, lymphadenopathy, hepatomegaly, splenomegaly, serositis).
   b. Member has had an inadequate response to non-steroidal anti-inflammatory drugs (NSAIDs) or systemic glucocorticoids.

C. Adult-onset Still's disease (AOSD)

1. Authorization of 12 months may be granted for adult members who have previously received a biologic indicated for active AOSD.

2. Authorization of 12 months may be granted for adult members for treatment of active AOSD when both of the following criteria are met:
   a. Member has active systemic features (e.g., fever, arthralgia/arthritis, evanescent rash, lymphadenopathy, hepatomegaly, splenomegaly, sore throat).
   b. Member meets any of the following:
      i. Member has had an inadequate response to a trial of nonsteroidal anti-inflammatory drugs (NSAIDs).
      ii. Member has had an inadequate response to a trial of corticosteroids.
      iii. Member has had an inadequate response to a trial of a conventional synthetic drug (e.g., methotrexate).

D. Gout and pseudogout flares

Authorization of 12 months may be granted for adult members for the treatment of flares for gout and pseudogout (also known as calcium pyrophosphate deposition disease) when both of the following criteria are met:

1. Member has experienced at least three flares in the last 12 months.
2. Member has had an inadequate response, intolerance or contraindication to non-steroidal anti-inflammatory drugs (NSAIDs), colchicine and corticosteroids.

V. CONTINUATION OF THERAPY

A. Systemic juvenile idiopathic arthritis (sJIA)

Authorization of 12 months may be granted for all members 2 years of age or older (including new members) who are using the requested medication for sJIA and who achieve or maintain a positive clinical response as evidenced by low disease activity or improvement in signs and symptoms of the condition when there is improvement in any of the following from baseline:

1. Number of joints with active arthritis (e.g., swelling, pain, limitation of motion)
2. Number of joints with limitation of movement
3. Functional ability
4. Systemic features (e.g., fever, evanescent rash, lymphadenopathy, hepatomegaly, splenomegaly, serositis)

B. Adult-onset Still's disease (AOSD)
Authorization of 12 months may be granted for all adult members (including new members) who are using the requested medication for AOSD and who achieve or maintain a positive clinical response as evidenced by low disease activity or improvement in signs and symptoms of the condition when there is improvement in any of the following from baseline:

1. Number of joints with active arthritis (e.g., swelling, pain, limitation of motion)
2. Number of joints with limitation of movement
3. Functional ability
4. Systemic features (e.g., fever, evanescent rash, lymphadenopathy, hepatomegaly, splenomegaly, serositis)

C. Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Autoinflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS)
Authorization of 12 months may be granted for all members 4 years of age or older (including new members) who are using the requested medication for CAPS, including FCAS and MWS, and who achieve or maintain a positive clinical response as evidenced by low disease activity or improvement in signs and symptoms of the condition.

D. All other diagnoses
Authorization of 12 months may be granted for all members (including new members) who are using the requested medication for an indication outlined in Section IV and who achieve or maintain a positive clinical response as evidenced by low disease activity or improvement in signs and symptoms of the condition.

VI. OTHER

For all indications: Member has had a documented negative tuberculosis (TB) test (which can include a tuberculosis skin test [TST ~~PPD~~] or an interferon-release assay [IGRA]~~, or a chest x-ray~~)* within 6 months of initiating therapy for persons who are naïve to biologic drugs or targeted synthetic drugs associated with an increased risk of TB.

*If the screening testing for TB is positive, there must be further testing to confirm there is no active disease (e.g., chest x-ray). Do not administer the requested medication to members with active TB infection. If there is latent disease, TB treatment must be started before initiation of the requested medication.

For all indications: Member cannot use the requested medication concomitantly with any other biologic drug or targeted synthetic drug.

APPLICABLE TENNESSEE STATE MANDATE REQUIREMENTS

BlueCross BlueShield of Tennessee's Medical Policy complies with Tennessee Code Annotated Section 56-7-2352 regarding coverage of off-label indications of Food and Drug Administration (FDA) approved drugs when the off-label use is recognized in one of the statutorily recognized standard reference compendia or in the published peer-reviewed medical literature.

ADDITIONAL INFORMATION

For appropriate chemotherapy regimens, dosage information, contraindications, precautions, warnings, and monitoring information, please refer to one of the standard reference compendia (e.g., the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) published by the National Comprehensive Cancer Network®, Drugdex Evaluations of Micromedex Solutions at Truven Health, or The American Hospital Formulary Service Drug Information).

REFERENCES

1. Ilaris [package insert]. East Hanover, NJ: Novartis Pharmaceuticals Corporation; August 2023.
2. De Benedetti F, Gattorno M, Anton J, et al; Canakinumab for the treatment of autoinflammatory recurrent fever syndromes. N Engl J Med. 2018;378:1908-19.Lachmann HJ, Kone-Paut I, Kuemmerle-Deschner JB, et al; Canakinumab in CAPS Study Group. Use of canakinumab in the cryopyrin-associated periodic syndrome. *N Engl J Med*. 2009;360(23):2416-2425.
3. 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: recommendations for the medical therapy of children with systemic juvenile idiopathic arthritis and tuberculosis screening among children receiving biologic medications. *Arthritis Care Res*. 2013;65(10):1551-63.
4. DRUGDEX® System (electronic version). Truven Health Analytics, Ann Arbor, MI. Available at http://www.micromedexsolutions.com [available with subscription]. Accessed August 30, 2023.
5. Richette P, Doherty M, Pascual E, et al. 2016 updated EULAR evidence-based recommendations for the management of gout. *Ann Rheum Dis*. 2017;76:29–42.
6. Zhang W, Doherty M, Pascual E, et al. EULAR recommendations for calcium pyrophosphate deposition. Part II: Management. *Ann Rheum Dis*. 2011;70:571–575.
7. Testing for TB Infection. Centers for Disease Control and Prevention. Retrieved on November 17, 2023 from: https://www.cdc.gov/tb/topic/testing/tbtesttypes.htm.
8. FitzGerald JD, Dalbeth N, Mikuls T, et al. 2020 American College of Rheumatology Guideline for the Management of Gout [published correction appears in *Arthritis Care Res (Hoboken)*. 2020 Aug;72(8):1187]. *Arthritis Care Res (Hoboken)*. 2020;72(6):744-760.
9. Onel KB, Horton DB, Lovell DJ, et al. 2021 American College of Rheumatology guideline for the treatment of juvenile idiopathic arthritis: therapeutic approaches for oligoarthritis, temporomandibular joint arthritis, and systemic juvenile idiopathic arthritis. *Arthritis Rheumatol*. 2022;74(4):553-569.
10. Efthimiou P, Kontzias A, Hur P, et al. Adult-onset Still's disease in focus: Clinical manifestations, diagnosis, treatment, and unmet needs in the era of targeted therapies. *Semin Arthritis Rheum*. 2021;51(4):858-874.

EFFECTIVE DATE

ID_CHS