Reference number
1801-A

SPECIALTY GUIDELINE MANAGEMENT

ILARIS (canakinumab)

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 Adult-Onset Still's Disease (AOSD) and Systemic Juvenile Idiopathic Arthritis (SJIA) in patients aged 2 years and older.

B. Compendial Use
Gout and pseudogout

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

II. CRITERIA FOR INITIAL APPROVAL

A. Periodic Fever Syndromes
1. Authorization of 12 months may be granted for treatment of CAPS when all of the following criteria are met:
   a. Member has a diagnosis of familial cold auto-inflammatory 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 all 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 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 all of the following criteria are met:
   a. Member has had active flares within the last 6 months.
   b. Physician's Global Assessment 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. Active Systemic Juvenile Idiopathic Arthritis (sJIA)

1. Authorization of 12 months may be granted for members who have received a biologic indicated for active systemic juvenile idiopathic arthritis.

2. Authorization of 12 months may be granted for the treatment of active sJIA when any of the following criteria is met:
   a. Member has had an inadequate response to at least a 1-month trial of nonsteroidal anti-inflammatory drugs (NSAIDs).
   b. Member has had an inadequate response to at least a 2-week trial of corticosteroids.
   c. Member has had an inadequate response to at least a 3-month trial of methotrexate or leflunomide.

C. Adult-Onset Still's Disease (AOSD)

1. Authorization of 12 months may be granted for members who have received a biologic indicated for active adult-onset Still's disease.

2. Authorization of 12 months may be granted for the treatment of active adult-onset Still's disease when any of the following criteria is met:
   a. Member has had an inadequate response to at least a 1-month trial of nonsteroidal anti-inflammatory drugs (NSAIDs).
   b. Member has had an inadequate response to at least a 1-month trial of corticosteroids.
   c. Member has had an inadequate response to at least a 3-month trial of a conventional DMARD (e.g., methotrexate).

D. Management of gout and pseudogout flares

Authorization of 6 months may be granted for the management of flares for gout or pseudogout (also known as calcium pyrophosphate deposition disease) when any of the following criteria is met:

1. Member has had an inadequate response or intolerance to maximum tolerated doses of non-steroidal anti-inflammatory drugs (NSAIDs), colchicine and oral and injectable corticosteroid.
2. Member has a contraindication to NSAIDs and colchicine, and has a clinical reason to avoid repeated courses of corticosteroids.

III. CONTINUATION OF THERAPY

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 II and who achieve or maintain positive clinical response as evidenced by low disease activity or improvement in signs and symptoms of the condition.

IV. OTHER

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

* If the screening testing for TB is positive, there must be further testing to confirm there is no active disease. 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.

** Risk factors for TB include: Persons with close contact to people with infectious TB disease; persons who have recently immigrated from areas of the world with high rates of TB (e.g., Africa, Asia, Eastern Europe, Latin America, Russia); children less than 5 years of age who have a positive TB test; groups with high rates of TB transmission (e.g., homeless persons, injection drug users, persons with HIV infection); persons who work or reside with people who are at an increased risk for active TB (e.g., hospitals, long-term care facilities, correctional facilities, homeless shelters).

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

V. REFERENCES

1. Ilaris [package insert]. East Hanover, NJ: Novartis Pharmaceuticals Corporation; September 2020.
2. Galeotti C, Meinzer U, Quartier P, et al. Efficacy of interleukin-1-targeting drugs in mevalonate kinase deficiency. *Rheumatology* (Oxford). 2012;51(10):1855-1859.
3. 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.
4. 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.
5. DRUGDEX® System (electronic version). Truven Health Analytics, Ann Arbor, MI. Available at http://www.micromedexsolutions.com [available with subscription]. Accessed November 15, 2020.
6. Schlesinger N, Alten RE, Bardin T, et al: Canakinumab for acute gouty arthritis in patients with limited treatment options: results from two randomized, multicentre, active-controlled, double-blind trials and their initial extensions. *Ann Rheum Dis*. 2012; 71(11):1839-1848.
7. 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.
8. Zhang W, Doherty M, Pascual E, et al. EULAR recommendations for calcium pyrophosphate deposition. Part II: Management. *Ann Rheum Dis*. 2011;70:571–575.
9. Centers for Disease Control and Prevention. Tuberculosis (TB). TB risk factors. Available at: https://www.cdc.gov/tb/topic/basics/risk.htm. Accessed: November 13, 2020.
10. 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.