Clinical Policy: Canakinumab (Ilaris)

Reference Number: PA.CP.PHAR.246
Effective Date: 01/18
Last Review Date: 04/18

Coding Implications
Revision Log

Description
The intent of the criteria is to ensure that patients follow selection elements established Pennsylvania Health and Wellness® clinical policy for Canakinumab (Ilaris®).

FDA Approved Indication(s)
Ilaris is indicated for the treatment of:

- Periodic fever syndromes:
  - Cryopyrin-Associated Periodic Syndromes (CAPS) in adults and children 4 years of age and older including:
    - Familial Cold Autoinflammatory Syndrome (FCAS)
    - Muckle-Wells Syndrome (MWS)
  - Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS) in adult and pediatric patients
  - Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD) in adult and pediatric patients
  - Familial Mediterranean Fever (FMF) in adult and pediatric patients
- Active Systemic Juvenile Idiopathic Arthritis (SJIA) in patients aged 2 years and older

Policy/Criteria
It is the policy of Pennsylvania Health and Wellness® that Ilaris is **medically necessary** when the following criteria are met:

I. Initial Approval Criteria

A. Periodic Fever Syndromes (must meet all)**:**

1. Diagnosis of FCAS, MWS, TRAPS, HIDS/MKD, or FMF;
2. Prescribed by or in consultation with a rheumatologist;
3. Member meets one of the following (a or b):
   a. FCAS or MWS: age ≥ 4 years;
   b. TRAPS, HIDS/MKD, or FMF: age ≥ 2 years;
4. For FMF, member meets one of the following (a or b):
   a. Age < 4 years;
   b. Failure of a ≥ 6 month trial of colchicine at up to maximally indicated doses, unless contraindicated or clinically significant adverse effects are experienced;
5. Dose does not exceed one of the following (a or b):
   a. FCAS or MWS: 150 mg every 8 weeks;
   b. TRAPS, HIDS/MKD, or FMF: 300 mg every 4 weeks.

Approval duration: 3 months for FCAS or MWS; 6 months for all other indications

B. Systemic Juvenile Idiopathic Arthritis (must meet all):
1. Prescribed by or in consultation with a dermatologist, rheumatologist; or gastrointestinal (GI) specialist;
2. Diagnosis of systemic juvenile idiopathic arthritis (SJIA);
3. Age ≥ to 2 years;
4. Member meets one of the following (a or b):
   a. Failure of a ≥ 3 consecutive month trial of methotrexate (MTX) or leflunomide at up to maximally indicated doses unless contraindicated or clinically significant adverse effects are experienced;
   b. Failure of a ≥ 2 week trial of a systemic corticosteroid at up to maximally indicated doses unless contraindicated or clinically significant adverse effects are experienced;
5. Prescribed dose of Ilaris does not exceed 300 mg every 4 weeks;

C. Other diagnoses/indications: Refer to PA.CP.PHAR.57 - Global Biopharm Policy.

II. Continued Approval

A. All Indications (must meet all)**:**
1. Currently receiving medication via PA Health and Wellness benefit or member has previously met all initial approval criteria or Continuity of Care policy applies;
2. Member is responding positively to therapy:
3. If request is for a dose increase, new dose does not exceed one of the following (a or b):
   a. FCAS or MWS: 150 mg every 8 weeks;
   b. TRAPS, HIDS/MKD, FMF, or SJIA: 300 mg every 4 weeks.

Approval duration: 12 months

B. Other diagnoses/indications (must meet 1 or 2)**:**
1. Currently receiving medication via Pennsylvania Health and Wellness benefit or the Continuity of Care policy (PA.LTSS.PHAR.01) applies and documentation supports positive response to therapy; or
2. Refer to PA.CP.PHAR.57 - Global Biopharm Policy.

Background

Description/Mechanism of Action:

Canakinumab is a human monoclonal anti-human IL-1β antibody of the IgG1/κ isotype. Ilaris binds to human IL- 1β and neutralizes its activity by blocking its interaction with IL-1 receptors, but it does not bind IL-1α or IL-1 receptor antagonist (IL-1ra).

Cryopyrin-associated periodic syndromes (CAPS) refer to rare genetic syndromes generally caused by mutations in the NLRP-3 [nucleotide-binding domain, leucine rich family (NLR), pyrin domain containing 3] gene (also known as Cold-Induced Auto-inflammatory Syndrome-1 [CIAS1]). CAPS disorders are inherited in an autosomal dominant pattern with male and female offspring equally affected. Features common to all disorders include fever, urticaria-like rash,

arthralgia, myalgia, fatigue, and conjunctivitis. The NLRP-3 gene encodes the protein cryopyrin, an important component of the inflammasome. Cryopyrin regulates the protease caspase-1 and controls the activation of interleukin-1 beta (IL-1β). Mutations in NLRP-3 result in an overactive inflammasome resulting in excessive release of activated IL-1β that drives inflammation. SJIA is a severe autoinflammatory disease, driven by innate immunity by means of pro-inflammatory cytokines such as interleukin 1β (IL-1β).

Formulations:
Ilaris is supplied in a sterile, single-use, colorless, 6 mL glass vial containing 150 of canakinumab as a white, preservative-free, lyophilized powder. Reconstitution with 1 mL of preservative-free sterile water for injection is required prior to administration.

Appendices

Appendix A: Abbreviation Key

CAPS: cryopyrin-associated periodic syndromes
CHAQ: Childhood Health Assessment Questionnaire
CRP: C-reactive protein
FCAS: familial cold autoinflammatory syndrome
FMF: familial Mediterranean fever
HIDS: hyperimmunoglobulin D syndrome
MKD: mevalonate kinase deficiency
MWS: Muckle-Wells syndrome
NSAID: non-steroidal anti-inflammatory drug
SAA: serum amyloid A
SJIA: active systemic juvenile idiopathic arthritis
TB: tuberculosis
TRAPS: tumor necrosis factor receptor associated periodic syndrome

Coding Implications

Codes referenced in this clinical policy are for informational purposes only. Inclusion or exclusion of any codes does not guarantee coverage. Providers should reference the most up-to-date sources of professional coding guidance prior to the submission of claims for reimbursement of covered services.

HCPCS Codes	Description
J0638	Injection, canakinumab, 1 mg

Reviews, Revisions, and Approvals	Date	Approval Date
2Q 2018 annual review: condensed all periodic fever syndromes into one criteria set; duration of initial approval for periodic fever syndromes modified to 6 months; added dermatologist and gastrointestinal specialist requirement to SJIA; removed requirement for TB testing from all criteria; references reviewed and updated.	2.27.18	

References

1. Ilaris Prescribing Information. East Hanover, NJ; Novartis Pharmaceuticals Corporation; September 2016. Available at www.ilaris.com. Accessed February 27, 2018.
2. Ringold S, Weiss PF, et al. 2013 update of the 2011 American College of Rheumatology recommendations for the treatment of juvenile idiopathic arthritis. Arthritis Care Res. 2013; 65(10): 2499-2512.
3. Beukelman T, et al. 2011 American College of Rheumatology recommendations for the treatment of juvenile idiopathic arthritis: initiation and safety monitoring of therapeutic agents for the treatment of arthritis and systemic features. Arthritis Care & Research, 2011; 63(4): 465-482.
4. Kimura Y. Systemic juvenile idiopathic arthritis: Treatment. In: UpToDate, Waltham, MA: Walters Kluwer Health; 2016. Available at: www.UpToDate.com. Accessed February 27, 2018..
5. Ozen S, Demirkaya E, Erer B, et al. EULAR recommendations for the management of familial Mediterranean fever. Ann Rheum Dis. 2016; 75(4): 644-651.