Policy Title:	Ilaris (canakinumab) (Subcutaneous)		
		Department:	PHA
Effective Date:	01/01/2020		
Review Date:	09/25/2019, 12/18/19, 1/29/20		
Revision Date:	09/25/2019, 1/29/20		

Purpose: To support safe, effective and appropriate use of Ilaris (canakinumab).

Scope: Medicaid, Exchange, Medicare-Medicaid Plan (MMP)

Policy Statement:

Ilaris (canakinumab) is covered under the Medical Benefit when used within the following guidelines. Use outside of these guidelines may result in non-payment unless approved under an exception process.

Procedure:

Coverage of Ilaris (canakinumab) will be reviewed prospectively via the prior authorization process based on criteria below.

Initial Criteria

- Patient has been evaluated and screened for the presence of latent TB infection prior to initiating treatment; AND
- Patient does not have an active infection, including clinically important localized infections; AND
- Must not be administered concurrently with live vaccines; AND
- Patient is not on concurrent therapy with other IL-1 blocking agents (e.g., anakinra, rilonacept, etc.): AND
- Patient is not on concurrent treatment with another TNF inhibitor, biologic response modifier or other non-biologic immunomodulating agent (i.e., apremilast, tofacitinib, baricitinib);
- MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements

Cryopyrin-Associated Periodic Syndromes (CAPS)

- Patient is over the age of 4; AND
- Must be used as a single agent; AND
- Patient has documented baseline serum levels of inflammatory proteins (C-Reactive Protein [CRP] and/or Serum Amyloid A [SAA]; AND

- Patient has documented laboratory evidence of a genetic mutation in the Cold-Induced Auto-inflammatory Syndrome 1 (CIAS1), also known as NLRP3; AND
    - Diagnosis of Familial Cold Autoinflammatory Syndrome (FCAS); OR
    - Diagnosis of Muckle-Wells Syndrome (MWS); AND
- Patient has two or more of any of the CAPS-typical symptoms:
    - urticaria-like rash
    - cold-triggered episodes
    - sensorineural hearing loss
    - musculoskeletal symptoms
    - chronic aseptic meningitis
    - skeletal abnormalities

Systemic Juvenile Idiopathic Arthritis

- Patient is over the age of 2; AND
- Patient has active Systemic Juvenile Idiopathic Arthritis (sJIA); AND
- Patient has had at least a 1-month trial and failure (unless contraindicated or intolerant) of previous therapy with either oral non-steroidal anti-inflammatory drugs (NSAIDs) OR a systemic glucocorticoid (prednisone, methylprednisolone, etc.)

Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS)

- Patient is over the age of 2; AND
- Patient has chronic or recurrent disease (defined as 6 or more flares per year); AND
- Patient has documented baseline serum levels of C-Reactive Protein (CRP)

Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD)

- Patient is over the age of 2; AND
- Patient has a confirmed diagnosis based on genetic/enzymatic laboratory findings; AND
- Patient has a documented prior history of greater than or equal to 3 febrile acute flares within a 6 month period

Familial Mediterranean Fever (FMF)

- Patient is over the age of 2; AND
- Patient has failed on colchicine therapy or has a documented allergy or intolerance; AND
- Patient has active disease defined as at least one flare per month

Continuation of Therapy Criteria:

- Patient continues to meet initial criteria; AND
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include the following: severe hypersensitivity reactions, serious infections (including but not limited to tuberculosis), and macrophage activation syndrome (MAS); AND
- Patient is receiving ongoing monitoring for presence of TB or other active infections; AND

Cryopyrin-Associated Periodic Syndromes

- Disease response as indicated by improvement in patient's symptoms from baseline AND improvement in serum levels of inflammatory proteins (e.g. CRP and/or SAA, etc) from baseline

Systemic Juvenile Idiopathic Arthritis

- Disease response as indicated by improvement in signs and compared to baseline such as the number of tender and swollen joint counts.

Tumor Necrosis Factor Receptor Associated Periodic Syndrome; Hyperimmunoglobulin D Syndrome/Mevalonate Kinase Deficiency; Familial Mediterranean Fever

- Disease response as indicated by improvement in patient's symptoms from baseline AND improvement of serum levels of CRP.

Coverage durations:

- Initial coverage: 6 months
- Continuation of therapy coverage: 6 months

*** Requests will also be reviewed to National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) if applicable. ***

Dosage/Administration:

Indication	Dose
Cryopyrin-Associated Periodic Syndromes	Weight greater than 40 kg • 150 mg subcutaneously every 8 weeks Weight equal to 15-40 kg • 2 mg/kg subcutaneously every 8 weeks. May be increased to 3 mg/kg if inadequate response.
Systemic Juvenile Idiopathic Arthritis	Weight is greater than or equal to 7.5 kg • 4 mg/kg (with a maximum of 300mg) subcutaneously every 4 weeks
All other indications	Weight greater than 40 kg • 150 mg subcutaneously every 4 weeks. May increase dose to 300mg if inadequate response. Weight less than or equal to 40 kg • 2 mg/kg subcutaneously every 4 weeks. May be increased to 4 mg/kg if inadequate response

Dosing Limits:

Indication	**Maximum dose (1 billable unit = 1 mg)**
Cryopyrin-Associated Periodic Syndromes	150 billable units every 8 weeks (56 days)
Systemic Juvenile Idiopathic Arthritis	300 billable units every 4 weeks (28 days)
Tumor Necrosis Factor Receptor Associated Periodic Syndrome	300 billable units every 4 weeks (28 days)
Hyperimmunoglobulin D Syndrome/Mevalonate Kinase Deficiency	300 billable units every 4 weeks (28 days)
Familial Mediterranean Fever	300 billable units every 4 weeks (28 days)

Investigational use: All therapies are considered investigational when used at a dose or for a condition other than those that are recognized as medically accepted indications as defined in any one of the following standard reference compendia: American Hospital Formulary Service Drug information (AHFS-DI), Thomson Micromedex DrugDex, Clinical Pharmacology, Wolters Kluwer Lexi-Drugs, or Peer-reviewed published medical literature indicating that sufficient evidence exists to support use. Neighborhood does not provide coverage for drugs when used for investigational purposes.

Applicable Codes:
Below is a list of billing codes applicable for covered treatment options. The below tables are provided for reference purposes and may not be all-inclusive. Requests received with codes from tables below do not guarantee coverage. Requests must meet all criteria provided in the procedure section.

The following HCPCS/CPT code is:

HCPCS/CPT Code	**Description**
J0638	Injection, canakinumab, 1mg

References:

1. Ilaris [package insert]. East Hanover, NJ; Novartis Pharmaceuticals Corporation; December 2016. Accessed July 2018.
2. Lachmann, HJ, Kone-Paut, I, Kuemmerle-Deschner, JB, et al. Use of canakinumab in the cryopyrin-associated periodic syndrome. N Engl J Med. 2009 Jun 4; 360(23):2416-25.

3. Ruperto N, Brunner H, Quartier P, et al. Two Randomized Trials of Canakinumab in Systemic Juvenile Idiopathic Arthritis. N Engl J Med 2012; 367:2396-2406.
4. Ringold, S., Weiss, P. F., Beukelman, T., DeWitt, E. M., Ilowite, N. T., Kimura, Y., Laxer, R. M., Lovell, D. J., Nigrovic, P. A., Robinson, A. B. and Vehe, R. K. (2013), 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 & Rheumatism, 65: 2499–2512. doi: 10.1002/art.38092
5. DeWitt EM, Kimura Y, Beukelman T, et al. Consensus treatment plans for new-onset systemic juvenile idiopathic arthritis. Arthritis Care Res (Hoboken). 2012 Jul;64(7):1001- 10.
6. Kuemmerle-Deschner JB, Ozen S, Tyrrell PN, et al. Diagnostic criteria for cryopyrinassociated periodic syndrome (CAPS). Ann Rheum Dis. 2017 Jun;76(6):942-947. doi: 10.1136/annrheumdis-2016-209686.
7. Terreri MT, Bernardo WM, Len CA, et al. Guidelines for the management and treatment of periodic fever syndromes: Cryopyrin-associated periodic syndromes (cryopyrinopathies –CAPS). Rev Bras Reumatol Engl Ed. 2016 Jan-Feb;56(1):44-51. doi: 10.1016/j.rbre.2015.08.020.