Premier Health Plan

POLICY AND PROCEDURE

POLICY TITLE: ***Ilaris (canakinumab)***
DEPARTMENT: **Clinical Pharmacy Services- Utilization Management**
ORIGINAL DATE: ***October 2009 (as adopted from UPMC Health Plan)***

Last P & T Committee Approval Date: *February 2018*

Product Applicability: *mark all applicable products below:*

COMMERCIAL	[ ] HMO [ ] PPO *Products*: [ ] Small [ ] Indiv. [ ] Large *Exchange*: [ ] Shop [ ] Indiv. [x] All
OTHER	[x] Self-funded/ASO

PURPOSE

The purpose of this policy is to define the prior authorization process for Ilaris (canakinumab).

Ilaris (canakinumab) is an interleukin-1B receptor blocker indicated for:

- Treatment of Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Autoinflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS), in adults and children 4 years of age and older.
- Treatment of active Systemic Juvenile Idiopathic Arthritis (SJIA) in patients aged 2 years and older

DEFINITIONS

Evanescent rash- a macular, salmon-colored rash on the trunk and extremities that is transient

Familial Cold Autoinflammatory Syndrome (FCAS) - An autosomal dominant inherited inflammatory disease characterized by episodes of rash, fever, and joint pain, following exposure to cold with attacks occurring 1-2 hours after exposure to cold and lasting less than 24 hours.

Familial Mediterranean Fever (FMF) - an autosomal recessive inherited disorder with a childhood onset and > 90% of patients having the first attack of periodic fever by the age of 20 years. The attacks may be triggered by certain events (cold exposure, infections, stress, menstrual cycle, exercise, fat-rich meals) and usually last about three days with fever and severe pain. Symptoms can include: peritonitis, arthritis, pleuritis, arthritis, and erysipelas like erythema.

Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD – an autosomal recessive disorder with a childhood onset, and characterized by recurring episodes of systemic inflammation and high spiking fevers. The attack can be spontaneous or may be triggered by certain events (vaccination, minor trauma, stress, surgery). The attacks last 3-7 days and the patient can experience prodromal symptoms. Symptoms can include: painful lymph nodes, abdominal pain, vomiting, diarrhea, skin-rash, join pain headache, hepatosplenomegaly.

Muckle-Wells Syndrome (MWS) - An autosomal dominant inherited inflammatory disease characterized by chronic recurrent urticaria, periodic arthritis, sensorineural deafness, general inflammation, and secondary amyloidosis.

NLRP3 gene - Nucleotide-binding domain, leucine-rich family, pyrin domain containing 3 gene. This gene encodes cryopyrin, a protein contained in intracellular inflammasomes. Cryopyrin regulates protease caspase-1 and controls interleukin-1 beta.

Papilledema - bilateral optic disc swelling due to raised intracranial pressure

Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS) - an autosomal dominant disorder with a childhood onset, and characterized by periodic episodes of high spiking fevers. The attacks may be triggered by certain events (local injury, minor infection, exercise, stress, hormonal change) and last 1-3 weeks. The patient can experience myalgia as a prodromal symptom. Symptoms can include: rash, muscle cramps, headache abdominal pain, pleuritis, pericarditis, painful lymph nodes, conjunctivitis, periorbital edema.

POLICY

It is the policy of the Health Plan to maintain a prior authorization process that promotes appropriate utilization of specific drugs with potential for misuse or limited indications. This process involves a review using Food and Drug Administration (FDA) criteria to make a determination of Medical Necessity, and approval by the Pharmacy & Therapeutics Committee of the criteria for prior authorization, as described in RX.002 Pharmacy and Therapeutics Committee and RX.003-Prior Authorization Process.

The drug, Ilaris (canakinumab), is subject to the prior authorization process.

PROCEDURE

Initial Authorization Criteria:

Must meet all of the criteria listed under the respective diagnosis:

1. **For all diagnoses:**
   - Must have a negative tuberculosis skin test [such as Tuberculin PPD (purified protein derivative) test] or Interferon-Gamma Release Assay (IGRA) whole-blood test [such as QuantiFERON®-TB Gold In-Tube test (QFT-GIT) or T-SPOT®.TB test (T-Spot)]
   - Must currently not be using a Tumor Necrosis Factor (TNF) blocking agent or other biologic agent
   - Must have no evidence of infection

2. **Muckle-Wells Syndrome:**
   - Must be prescribed by a rheumatologist, dermatologist, immunologist, or genetic specialist
   - Must be age 4 years or older
   - Must have a diagnosis of Muckle-Wells syndrome (MWS). Chart documentation of a clinical work-up to rule out other diagnoses and clinical rationale for the diagnosis and exclusion of other diagnoses must be provided. The diagnosis must either be confirmed by genetic testing or a clinical diagnosis defined as one of the following scenarios:
     - Must have a mutation in the NLRP3 (formerly CIAS1) gene. Documentation of lab result confirming mutation is required.
     - Must meet 3 of the following diagnostic criteria (chart documentation required):
       - History of intermittent episodes of fever for at least 24 hours duration
       - Autosomal dominant pattern of disease inheritance
       - Presence of severe fatigue
       - Presence of musculoskeletal symptoms (e.g. arthralgia, arthritis, myalgia)
       - Presence of ocular symptoms (e.g. conjunctivitis, anterior uveitis, papilledema)
       - Presence of erythematous rash

        - Presence of amyloidosis
        - Presence of hearing loss
- Must have an adequate trial of at least 3 months of Kineret (anakinra) with an inadequate response or significant side effects/toxicity or have a contraindication to this therapy

3. Familial Cold Autoinflammatory Syndrome (FCAS):

- Must be prescribed by a rheumatologist, dermatologist, immunologist, or genetic specialist
- Must be age 4 years or older
- Must have a diagnosis of Familial Cold Autoinflammatory Syndrome (FCAS). Chart documentation of a clinical work-up to rule out other diagnoses and clinical rationale for the diagnosis and exclusion of other diagnoses must be provided. The diagnosis must either be confirmed by genetic testing or a clinical diagnosis defined as one of the following scenarios:
    - Must have a mutation in the NLRP3 (formerly CIAS1) gene. Documentation of lab result confirming mutation is required.
    - Must meet 3 of the following diagnostic criteria (chart documentation required):
        - History of intermittent episodes of fever and rash that primarily follow natural, experimental, or both types of generalized cold exposures for less than 24 hours duration
        - Autosomal dominant pattern of disease inheritance
        - Age of onset < 6 months of age
        - Presence of conjunctivitis associated with attacks
        - Absence of deafness, periorbital edema, lymphadenopathy, and serositis
- Must have an adequate tiral of at least 3 months of Kineret (anakinra) wth an inadequate response or significant side effects/toxicity or have a contraindication to this therapy

4. Familial Mediterranean Fever (FMF)

- Must be prescribed by a rheumatologist, dermatologist, immunologist, or genetic specialist
- Must have a diagnosis of Familial Mediterranean Fever (FMF). Chart documentation of a clinical work-up to rule out other diagnoses and clinical rationale for the diagnosis and exclusion of other diagnoses must be provided.

The diagnosis must either be confirmed by genetic testing or a clinical diagnosis defined as one of the following scenarios:
    - Must have a mutation of the MEFV gene. Documentation of lab result confirming mutation is required.
    - Must meet 3 of the following diagnostic criteria (chart documentation required):
        - History of intermittent episodes of fever and pain for at least 3 days duration
        - Autosomal recessive pattern of disease inheritance
        - Presence of peritonitis
        - Presence of arthritis
        - Presence of pleuritis
        - Presence of erysipelas-like erythema
- Must have an adequate trial of a least 3 months of colchicine with an inadequate response or significant side effects/toxicity or have a contraindication to this therapy

- Must have an adequate trial of at least 3 months of Kineret (anakinra) with an inadequate response or significant side effects/toxicity or have a contraindication to this therapy

5. Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD)
- Must be prescribed by a rheumatologist, dermatologist, immunologist, or genetic specialist
- Must be age 2 years or older
- Must have a diagnosis of Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD). Chart documentation of a clinical work-up to rule out other diagnoses and clinical rationale for the diagnosis and exclusion of other diagnoses must be provided. The diagnosis must either be confirmed by genetic testing or a clinical diagnosis defined as one of the following scenarios:
    - Must have a mutation in the MVK gene. Documentation of lab result confirming mutation is required.
    - Must meet 3 of the following diagnostic criteria (chart documentation required):
        - History of intermittent episodes of fever and inflammation for at least 3 days duration
        - Autosomal recessive pattern of disease inheritance
        - Presence of painful lymph nodes
        - Presence of joint pain

        - Presence of headache
        - Presence of hepatosplenomegaly.
        - Presence of abdominal pain, vomiting, diarrhea
        - Presence of a skin rash

- Must have an adequate trial of at least 3 months of Kineret (anakinra) with an inadequate response or significant side effects/toxicity or have a contraindication to this therapy

6. Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS)

- Must be prescribed by a rheumatologist, dermatologist, immunologist, or genetic specialist
- Must have a diagnosis of Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS). Chart documentation of a clinical work-up to rule out other diagnoses and clinical rationale for the diagnosis and exclusion of other diagnoses must be provided. The diagnosis must either be confirmed by genetic testing or a clinical diagnosis defined as one of the following scenarios:
    - Must have a mutation in the TNFRSF1A gene. Documentation of lab result confirming mutation is required.
    - Must meet 3 of the following diagnostic criteria (chart documentation required):
        - History of intermittent episodes of fever for at least 1 week
        - duration
        - Autosomal dominant pattern of disease inheritance
        - Presence of painful lymph nodes
        - Presence of headache
        - Presence of a skin rash
        - Presence of muscle cramps
        - Presence of abdominal pain
        - Presence of pleuritis
        - Presence of pericarditis
        - Presence of conjunctivitis, periorbital edema

- Must have an adequate trial of a least 3 months of Enbrel (etanercept) with an inadequate response or significant side effects/toxicity or have a contraindication to this therapy

7. **Juvenile Idiopathic Arthritis with systemic symptoms includes systemic juvenile idiopathic arthritis (SJIA):**
    - Must be prescribed by a pediatric rheumatologist
    - Must be age 2 years or older
    - Must have a diagnosis of active juvenile idiopathic arthritis. Chart documentation of a clinical work-up to rule out other diagnoses and clinical rationale for the diagnosis and exclusion of other diagnoses must be provided. Chart documentation must demonstrate all of the following:
        - History of fever for at least 2 week duration
        - History of at least 1 of the following:
            - Evanscent rash
            - History of arthritis in 1 or more joints
            - Generalized lymph node enlargement
            - Hepatomegaly or splenomegaly
            - Pericarditis, pleuritis, or peritonitis
    - Must have an adequate trial of at least 3 months of Kineret (anakinra) wth an inadequate response or significant side effects/toxicity or have a contraindication to this therapy

Reauthorization Criteria:

All prior authorization renewals are reviewed to determination the Medical Necessity for the continuation of treatment. Authorization is extended as specified below:

1. **For Periodic Fever Syndromes (MWS, FCAS, FMF, HIDS/MKD, TRAPS)**
    - For an additional 3 months based upon:
        - Chart documentation from the prescriber that the member's condition has improved based upon the prescriber's assessment while on therapy.
        - Documentation that there is no evidence of infection.
2. **For SJIA**
    - For an additional year based upon:
        - Chart documentation from the prescriber that the member's condition has improved based upon the prescriber's assessment while on therapy.
        - Documentation that there is no evidence of infection.

Limitations:

Length of Authorization (if above criteria met)	
Initial Authorization	Up to 3 months
Reauthorization	• SJIA: Up to 1 year • Periodic Fever Syndromes (MWS, FCAS, FMF, HIDS/MKD, TRAPS): Up to 3 months

If the established criteria are not met, the request is referred to a Medical Director for review.

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RECORD RETENTION

Records Retention for Evolent Health documents, regardless of medium, are provided within the Evolent Health records retention policy and as indicated in CORP.028.E Records Retention Policy and Procedure.

REVIEW HISTORY

DESCRIPTION OF REVIEW / REVISION	DATE APPROVED
Annual Review	*02/16, 02/17, 02/18*
Criteria Update	*12/16, 04/17*