SmartPA Criteria Proposal

Drug/Drug Class:	Cryopyrin-Associated Periodic Syndrome (CAPS) Agents PDL Edit
First Implementation Date:	May 26, 2010
Revised Date:	October 5, 2023
Prepared For:	MO HealthNet
Prepared By:	MO HealthNet/Conduent
Criteria Status:	☐ Existing Criteria ☒ Revision of Existing Criteria ☐ New Criteria

Executive Summary

Purpose: The MO HealthNet Pharmacy Program will implement a state-specific preferred drug list.

Why Issue Selected: Cryopyrin-associated periodic syndrome (CAPS) is a group of rare autosomal-dominant, interleukin (IL) 1-associated, auto-inflammatory disorders. The group includes familial cold autoinflammatory (urticaria) syndromes (FCAS/FCU), Muckle-Wells syndrome (MWS), and neonatal-onset multisystem inflammatory disease (NOMID) (aka chronic infantile neurological cutaneous articular (CINCA) syndrome. CAPS is caused by pathogenic variants in the nucleotide-binding domain, leucine rich family, pyrin domain containing 3gene or the cold-induced auto-inflammatory syndrome-1 (CIAS1) gene. Cryopyrin, a protein encoded by this gene, regulates IL-1beta activation and a deficiency in cryopyrin causes excessive inflammation. Symptoms include rash, headaches, periodic fevers, general malaise, joint pain, and conjunctivitis. Prevalence of the disease is estimated to be 1 in 1 million people worldwide. Symptoms are typically triggered by cold or cooling temperatures in participants with FCAS/FCU or MWS and can last for 1-3 days. MWS can also be triggered by stress or other unknown factors and may be characterized by a progressive, hearing loss starting in adolescence. NOMID/CINCA is characterized by continuous inflammation in multiple organs starting during infancy. Most participants suffer from chronic inflammation of the central nervous system resulting in chronic aseptic meningitis, severe headaches, elevated brain pressures, and progressive hearing loss, along with cognitive and mental deficits.

Total program savings for the PDL classes will be regularly reviewed.

Program-Specific Information:

Preferred Agents	Non-Preferred Agents
• Ilaris® • Kineret®	• Arcalyst®

Type of Criteria:

☐ **Increased risk of ADE**
☒ **Preferred Drug List**
☐ **Appropriate Indications**
☐ **Clinical Edit**

Data Sources:

☐ **Only Administrative Databases**
☒ **Databases + Prescriber-Supplied**

Setting & Population

- Drug class for review: Cryopyrin-Associated Periodic Syndrome (CAPS) Agents
- Age range: All appropriate MO HealthNet participants

Approval Criteria

- Documented compliance on current therapy regimen **OR**
- Failure to achieve desired therapeutic outcomes with trial on 1 or more preferred agents
  - Documented trial period of preferred agents **OR**
  - Documented ADE/ADR to preferred agents **AND**
- For Arcalyst:
  - Documented diagnosis of deficiency of interleukin-1 receptor antagonist (DIRA) **OR**
  - Participant aged 12 years or older **AND**
  - Documented diagnosis of cryopyrin-associated periodic syndrome **OR**
  - Documented diagnosis of recurrent pericarditis
- For Ilaris:
  - Documented diagnosis of juvenile idiopathic arthritis
    - Participant aged 2 years or older **AND**
    - Adequate therapeutic 6 month trial of tumor necrosis factor (TNF) inhibitor (trial defined as duration of therapy with class not agent)
  - Documented diagnosis of cryopyrin-associated periodic syndrome
    - Participant aged 4 years or older **OR**
  - Documented diagnosis of periodic fever syndromes **OR adult-onset Still's disease**
    - Participant aged 2 years or older
  - **Documented diagnosis of gout**
    - **Documented contraindication, intolerance, or inadequate response to NSAIDs and colchicine AND**
    - **Documented reason repeated courses of corticosteroids are not appropriate**
- For Kineret:
  - Documented diagnosis of deficiency of interleukin-1 receptor antagonist (DIRA) **OR**
  - Documented diagnosis of neonatal-onset multisystem inflammatory disease **OR**
  - Documented diagnosis of rheumatoid arthritis:
    - Participant aged 18 years or older **AND**
    - Adequate therapeutic 6 month trial of tumor necrosis factor (TNF) inhibitor (trial defined as duration of therapy with class not agent)

Denial Criteria

- Concurrent therapy with a tumor necrosis factor inhibitor in the past 45 days with diagnosis of cryopyrin-associated periodic syndrome, deficiency of interleukin-1 receptor antagonist, or periodic fever syndromes
- Lack of adequate trial on required preferred agents
- Therapy will be denied if all approval criteria are not met

Required Documentation

Laboratory Results:		Progress Notes:	X
MedWatch Form:		Other:	

Disposition of Edit

Denial: Exception Code “0160” (Preferred Drug List)
Rule Type: PDL

Default Approval Period

1 year

References

- Evidence-Based Medicine and Fiscal Analysis: “Therapeutic Class Review: IMMUNOLOGIC AGENTS Systemic Immunomodulators, Cryopyrin-Associated Periodic Syndrome (CAPS) Agents”, Gainwell Technologies; Last updated May 3, 2023.
- Evidence-Based Medicine Analysis: “Cryopyrin-Associated Periodic Syndrome (CAPS) Treatment”. UMKC-DIC; February 2023.
- Nigrovic, P., (2023). Cryopyrin-associated periodic syndromes and related disorders. In E. TePas (Ed.), *UpToDate.*
- Kineret [package insert]. Stockholm, Sweden: Swedish Orphan Biovitrum AB; December 2020.
- Ilaris [package insert]. East Hanover, NJ: Novartis Pharmaceuticals; September 2020.
- Arcalyst [package insert]. London, UK: Kiniksa Pharmaceuticals (UK), Ltd.; March 2021.
- USPDI, Micromedex; 2023.
- Clinical Pharmacology [online]. Tampa (FL): Elsevier. 2023.