SmartPA Criteria Proposal

Drug/Drug Class:	Cryopyrin-Associated Periodic Syndrome (CAPS) Agents PDL Edit
First Implementation Date:	May 26, 2010
Proposed Date:	June 17, 2021
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**
☐ **Appropriate Indications**
☒ **Preferred Drug List**
☐ **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 aged ~~2 years and older~~

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
  - Participant aged 12 years or older **AND**
  - Documented diagnosis of cryopyrin-associated periodic syndrome ~~in the past year~~ **OR**
  - **Documented diagnosis of recurrent pericarditis**
- For Ilaris:
  - **Documented diagnosis of juvenile idiopathic arthritis or adult-onset Still's disease~~: Clinical Consultant Review OR~~**
    - Participant aged 2 years or older **AND**
    - **Adequate therapeutic 6 month trial of tumor necrosis factor (TNF) inhibitor defined as:**
      - **Combination therapy of 2 TNF inhibitors OR**
      - **Monotherapy of 1 TNF inhibitor OR**
  - Documented diagnosis of cryopyrin-associated periodic syndrome ~~in the past year~~
    - Participant aged 4 years or older **OR**
  - Documented diagnosis of periodic fever syndromes ~~in the past year~~ **OR**
- **For Kineret:**
  - **Documented diagnosis of neonatal-onset multisystem inflammatory disease OR**
  - **Documented diagnosis of rheumatoid arthritis:**
    - **Participant aged 18 years or older AND**
    - **Adequate therapeutic trial of methotrexate OR**
    - **Contraindication to methotrexate therapy AND**
    - **Adequate therapeutic 6 month trial of tumor necrosis factor (TNF) inhibitor defined as:**
      - **Combination therapy of 2 TNF inhibitors OR**
      - **Monotherapy of 1 TNF inhibitor**
- **Documented diagnosis of deficiency of interleukin-1 receptor antagonist (DIRA) and claim is for Arcalyst or Kineret**

Denial Criteria

- Therapy will be denied if all approval criteria are not met
- For Ilaris and Arcalyst: concurrent therapy with a tumor necrosis factor inhibitor in the past 45 days **with diagnosis of cryopyrin-associated periodic syndrome or periodic fever syndromes**
- Lack of adequate trial on required preferred agents

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

1. Drug Effectiveness Review Project – Drug Class Review “Targeted Immune Modulators”. Center for Evidence-Based Policy, Oregon Health & Science University, March 2012; updated June 2016.
2. Evidence-Based Medicine and Fiscal Analysis: “Systemic Immunomodulators, CAPS Agents – Therapeutic Class Review”, Conduent Business Services, L.L.C., Richmond, VA; June 2021.
3. Evidence-Based Medicine Analysis: “Cryopyrin-Associated Periodic Syndrome (CAPS) treatment”, UMKC-DIC; May 2021.
4. 2015 American College of Rheumatology Guideline for the Treatment of Rheumatoid Arthritis, Singh et al. Arthritis Care & Research – DOI 10.1002/acr.22783
5. Nigrovic, P., (2019). Cryopyrin-associated periodic syndromes and related disorders. In E. TePas (Ed.), *UpToDate*.
6. Kineret [package insert]. Stockholm, Sweden: Swedish Orphan Biovitrum AB; December 2020.
7. Ilaris [package insert]. East Hanover, NJ: Novartis Pharmaceuticals; September 2020.
8. Arcalyst [package insert]. London, UK: Kiniksa Pharmaceuticals (UK), Ltd.; March 2021.
9. USPDI, Micromedex; 2021.
10. Facts and Comparisons eAnswers (online); 2021 Clinical Drug Information, LLC.