SmartPA Criteria Proposal

Drug/Drug Class:	Cryopyrin-Associated Periodic Syndrome (CAPS) Agents PDL Edit
First Implementation Date:	May 26, 2010
Proposed Date:	June 18, 2020
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), neonatal-onset multisystem inflammatory disease (NOMID) (aka chronic infantile neurological cutaneous articular (CINCA) syndrome. CAPS is caused by mutations 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. Diagnosis is based on symptoms, but a proper diagnosis should include all autoinflammatory disorders, but specifically CIAS1. 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®	• Arcalyst® • Kineret®

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 aged 2 years and older

Approval Criteria

- Documented compliance on current therapy regimen **OR**
- For Ilaris:
    - Documented diagnosis of juvenile idiopathic arthritis in the past 2 years
        - Participants aged 2 years or older **AND**
        - **Adequate therapeutic trial of methotrexate in the past 720 days OR**
        - **Contraindication to methotrexate therapy AND**
        - **Adequate therapeutic 6 month trial of tumor necrosis factor (TNF) inhibitors 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
        - Participants aged 4 years or older **OR**
    - Documented diagnosis of periodic fever syndromes in the past year **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 cryopyrin-associated periodic syndrome in the past year
    - Participant aged 12 years or older **OR**
- For Kineret:
    - Documented diagnosis of neonatal-onset multisystem inflammatory disease the past 2 years **OR**
    - Documented diagnosis of rheumatoid arthritis in the past 2 years
        - Adequate therapeutic trial of methotrexate in the past 720 days OR
        - Contraindication to methotrexate therapy AND
        - Adequate therapeutic 6 month trial of tumor necrosis factor (TNF) inhibitors defined as:
            - Combination therapy of 2 TNF inhibitors OR
            - Monotherapy of 1 TNF inhibitor

Denial Criteria

- For Ilaris and Arcalyst: concurrent therapy with a tumor necrosis factor inhibitor in the past 45 days
- Lack of adequate trial on required preferred agents
- Therapy will be denied if no approval criteria are 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

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: "Targeted Immune Modulators – Therapeutic Class Review", Conduent Business Services, L.L.C., Richmond, VA; May 2019.
3. Evidence-Based Medicine Analysis: "Cryopyrin-Associated Periodic Syndrome (CAPS) treatment", UMKC-DIC; March 2020.
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; 2018.
7. Ilaris [package insert]. East Hanover, NJ: Novartis Pharmaceuticals; 2016.
8. Arcalyst [package insert]. Tarrytown, NY: Regeneron Pharmaceuticals; 2020.
9. Lippincott, Williams, Wilkins. PDR Electronic Library, Montvale NJ; 2020.
10. USPDI, Micromedex; 2020.
11. Facts and Comparisons eAnswers (online); 2020 Clinical Drug Information, LLC.