



# SmartPA Criteria Proposal

<b>Drug/Drug Class:</b>	Cryopyrin-Associated Periodic Syndrome (CAPS) Agents PDL Edit
<b>First Implementation Date:</b>	May 26, 2010
<b>Revised Date:</b>	October 14, 2021
<b>Prepared For:</b>	MO HealthNet
<b>Prepared By:</b>	MO HealthNet/Conduent
<b>Criteria Status:</b>	<input type="checkbox"/> Existing Criteria <input checked="" type="checkbox"/> Revision of Existing Criteria <input type="checkbox"/> 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 3 gene 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
	<ul style="list-style-type: none"> <li>• Ilaris®</li> <li>• Kineret®</li> </ul>	<ul style="list-style-type: none"> <li>• Arcalyst®</li> </ul>

**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 **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**
    - Participant aged 2 years or older
- 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 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

## 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:   
MedWatch Form:

Progress Notes:   
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.