

# 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>Proposed Date:</b>	June 18, 2020
<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), 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 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. 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
	<ul style="list-style-type: none"> <li>• Ilaris®</li> </ul>	<ul style="list-style-type: none"> <li>• Arcalyst®</li> <li>• Kineret®</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 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:

MedWatch Form:

Other:

## Disposition of Edit

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

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## Default Approval Period

1 year

## References

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6. Kineret [package insert]. Stockholm, Sweden: Swedish Orphan Biovitrum AB; 2018.
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8. Arcalyst [package insert]. Tarrytown, NY: Regeneron Pharmaceuticals; 2020.
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11. Facts and Comparisons eAnswers (online); 2020 Clinical Drug Information, LLC.

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