# SmartPA Criteria Proposal

<table>
<thead>
<tr>
<th>Drug/Drug Class:</th>
<th>Cryopyrin-Associated Periodic Syndrome (CAPS) Agents PDL Edit</th>
</tr>
</thead>
<tbody>
<tr>
<td>First Implementation Date:</td>
<td>May 26, 2010</td>
</tr>
<tr>
<td>Revised Date:</td>
<td>October 1, 2020</td>
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<tr>
<td>Prepared For:</td>
<td>MO HealthNet</td>
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<td>Prepared By:</td>
<td>MO HealthNet/Conduent</td>
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<tr>
<td>Criteria Status:</td>
<td>☐ Existing Criteria ☒ Revision of Existing Criteria ☐ New Criteria</td>
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## 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.

<table>
<thead>
<tr>
<th>Program-Specific Information:</th>
<th>Preferred Agents</th>
<th>Non-Preferred Agents</th>
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<tbody>
<tr>
<td>• Ilaris®</td>
<td>• Arcalyst®</td>
<td>• Kineret®</td>
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<table>
<thead>
<tr>
<th>Type of Criteria:</th>
<th>☐ Increased risk of ADE</th>
<th>☒ Preferred Drug List</th>
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<tbody>
<tr>
<td>Data Sources:</td>
<td>☐ Only Administrative Databases</td>
<td>☒ Databases + Prescriber-Supplied</td>
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</tbody>
</table>

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*Other company trademarks are also acknowledged.*
**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 adult-onset Still’s disease: Clinical Consultant Review OR
  - Documented diagnosis of juvenile idiopathic arthritis
    - Participants aged 2 years or older 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 OR
  - Documented diagnosis of rheumatoid arthritis
    - Participants 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) 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 with diagnosis of cryopyrin-associated periodic syndrome or periodic fever syndromes
- Lack of adequate trial on required preferred agents
- Therapy will be denied if no approval criteria are met

**Required Documentation**

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

**Disposition of Edit**

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

1 year

References

10. USPDI, Micromedex; 2020.
11. Facts and Comparisons eAnswers (online); 2020 Clinical Drug Information, LLC.