SmartPA Criteria Proposal

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**Drug/Drug Class:** Cryopyrin-Associated Periodic Syndrome (CAPS) Agents PDL

**First Implementation Date:** May 26, 2010

**Revised Date:** October 14, 2021

**Prepared For:** MO HealthNet

**Prepared By:** MO HealthNet/Conduent

**Criteria Status:** ☒ Revision of Existing Criteria

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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), 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.

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**Program-Specific Information:**

<table>
<thead>
<tr>
<th>Preferred Agents</th>
<th>Non-Preferred Agents</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ilaris®</td>
<td>Arcalyst®</td>
</tr>
<tr>
<td>Kineret®</td>
<td></td>
</tr>
</tbody>
</table>

**Type of Criteria:**
- ☐ Increased risk of ADE
- ☒ Preferred Drug List
- ☐ Appropriate Indications
- ☐ Clinical Edit
- ☐ 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: [ ]  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

9. USPDI, Micromedex; 2021.
10. Facts and Comparisons eAnswers (online); 2021 Clinical Drug Information, LLC.