Protocol for Cryopyrin-Associated Periodic Syndromes (CAPS) Products

Approved July 2020

Arcalyst® (rilonacept)
Ilaris® (canakinumab)
Kineret® (anakinra)

Background:
Three clinically overlapping, interleukin (IL) 1-associated, autoinflammatory disorders are known collectively as the cryopyrin-associated periodic syndromes (CAPS) or cryopyrinopathies: familial cold autoinflammatory syndrome (FCAS), Muckle-Wells syndrome (MWS), and neonatal-onset multisystem inflammatory disorder (NOMID, also known as chronic infantile neurologic cutaneous and articular [CINCA] syndrome).

***Arcalyst® (rilonacept)** is an interleukin-1blocker indicated for the treatment of CAPS, including FCAS and MWS in children 12 and older including:*

- *Familial Cold Autoinflammatory Syndrome (FCAS)*
- *Muscle-Wells Syndrome (MWS)*

***Ilaris® (canakinumab)** is an interleukin-1β blocker indicated for the treatment of:*

- *Cryopyrin-associated periodic syndromes (CAPS) in adults and children 4 years of age and older including:*
  - *Familial Cold Autoinflammatory Syndrome (FCAS)*
  - *Muckle-Wells Syndrome (MWS)*
- *Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS) in adult and pediatric patients*
- *Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD) in adults and pediatric patients*
- *Familial Mediterranean Fever (FMF) in adult and pediatric patients*
- *Active Systemic Juvenile Idiopathic Arthritis (SJIA)*

***Kineret® (anakinra)** is an interleukin-1 receptor antagonist indicated for the treatment of:*

- *Rheumatoid Arthritis (RA)*
- *Cryopyrin-associated periodic syndromes*
- *Patient has Schnitzler syndrome*
- *Patient has moderate to severe Hidradenitis Suppurativa (HS)*

Criteria for approval:

1. Medication is prescribed by or in consultation with a rheumatologist or physician experienced in the treatment of genetic disorders. For diagnosis of Hidradenitis Suppurativa the medication is prescribed by or in consultation with a dermatologist. For diagnosis of Schnitzler syndrome, the medication is prescribed by or in consultation with a rheumatologist, dermatologist, or immunologist **AND**

2. Medication will not be used in combination with any other biologic DMARD or Targeted Immune Modulator for the same diagnosis **AND**

3. Weight must be received for drugs that have weight-based dosing **AND**

4. Medication is prescribed in accordance with Food and Drug Administration (FDA) established indication and dosing regimens or in accordance with medically appropriate off-label indication and dosing according to American Hospital Formulary Service, Micromedex, Clinical Pharmacology, Lexi-Drugs, national guidelines, or other peer-reviewed evidence

For Arcalyst:

A. Patient is ≥ 12 years old **AND**
B. Patient has a confirmed diagnosis of Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Auto-inflammatory syndrome (FCAS) and Muckle-Wells Syndrome (MWS)

For Ilaris: The member meets at least one of the following:

A. Patient has a confirmed diagnosis of Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Auto-inflammatory syndrome (FCAS) and Muckle-Wells Syndrome (MWS) and meets the following criteria:
    1. Patient is ≥ 4 years old; **OR**

B. Patient has a confirmed diagnosis of Familial Mediterranean Fever (FMF) and meets the following criteria:
    1. For children 4 years or older, the patient has inadequate response to or is intolerant to colchicine; **OR**

C. Patient has a confirmed diagnosis of Hyperimmunoglobulin D syndrome (HIDS)/mevalonate kinase deficiency (MKD); **OR**

D. Patient has a confirmed diagnosis of Tumor necrosis factor receptor associated periodic syndrome (TRAPS); **OR**

E. Patient has confirmed diagnosis of active systemic juvenile idiopathic arthritis (SJIA) and meets the following criteria:
    1. Patient is ≥ 2 years with SJIA **AND**
    2. For patients with active systemic features:

i. Unless contraindicated to all, patient has inadequate response to or is intolerant to systemic corticosteroids or non-steroidal anti-inflammatory drugs (NSAIDS) or methotrexate or leflunomide or anakinra (Kineret®) or tocilizumab (Actemra®) **OR**

3. For patients without active systemic features
    i. Unless contraindicated to all, patient has had an inadequate response to or is intolerant to one of the following:
        1. DMARD (i.e., methotrexate or leflunomide) plus anakinra
        2. DMARD (i.e., methotrexate or leflunomide) plus tocilizumab
        3. DMARD (i.e., methotrexate or leflunomide) plus TNF-α inhibitor (e.g., adalimumab, etanercept, infliximab)
        4. abatacept

For Kineret:

A. Member does not have known hypersensitivity to E coli-derived proteins **AND**

B. Patient has one of the following diagnoses:
    1. Patient has a diagnosis of Rheumatoid Arthritis (RA) and meets the following criteria:
        i. Patient is ≥ 18 years
        ii. Patient has moderately to severely active disease
        iii. Unless contraindicated to all, patient has had intolerance or an inadequate response to at least a 3-months trial of one of the following disease modifying antirheumatic drugs (DMARDs): Hydroxychloroquine, Leflunomide, Methotrexate, or Sulfasalazine; **OR**

    2. Patient has confirmed diagnosis of Cryopyrin-Associated Periodic Syndromes (CAPS):
        i. Neonatal-Onset Multisystem Inflammatory Disease (NOMID) also known as chronic infantile neurologic cutaneous articular syndrome (CINCA); **OR**

    3. Patient has a diagnosis of Systemic Juvenile Idiopathic Arthritis (SJIA) defined as <u>one</u> of the following:
        i. Patients has active systemic features
            a. Unless contraindicated to all, patient has had an inadequate response to or is intolerant to systemic corticosteroids or non-steroidal anti-inflammatory drugs (NSAIDS) **OR**
        ii. Patients without active systemic features
            a. Unless contraindicated to all, patient has had an inadequate response to or is intolerant to methotrexate, leflunomide, non-steroidal anti-inflammatory drugs (NSAIDS), or intra-articular glucocorticosteroids **OR**

iii. Patient has macrophage activation syndrome (MAS); **OR**

4. Patient has Schnitzler syndrome
    i. Member is ≥ 18 years old; **OR**

5. Patient has moderate to severe Hidradenitis Suppurativa (HS)

Continuation of therapy:

A. Documentation of positive clinical response to medication from baseline

B. For dose increase requests, weight must be received for drugs that have weight-based dosing.

C. Medication is prescribed in accordance with Food and Drug Administration (FDA) established indication and dosing regimens or in accordance with medically appropriate off-label indication and dosing according to American Hospital Formulary Service, Micromedex, Clinical Pharmacology, Lexi-Drugs, national guidelines, or other peer-reviewed evidence

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