Premier Health Plan

POLICY AND PROCEDURE

POLICY TITLE: Arcalyst (**Rilonacept)**
DEPARTMENT: **Clinical Pharmacy Services- Utilization Management**
ORIGINAL DATE: ***July 2008 (as adopted from UPMC Health Plan)***

Last P & T Committee Approval Date: *February 2018*

Product Applicability: *mark all applicable products below:*

COMMERCIAL	[ ] HMO [ ] PPO *Products*: [ ] Small [ ] Indiv. [ ] Large *Exchange*: [ ] Shop [ ] Indiv. [x] All
OTHER	[x] Self-funded/ASO

PURPOSE

The purpose of this policy is to define the prior authorization process for Arcalyst (rilonacept).

Arcalyst (Rilonacept) is an interleukin-1 inhibitor indicated for the treatment for the cryopyrin-associated periodic syndromes (CAPS), including familial cold autoinflammatory syndrome (FCAS) and Muckle-Wells syndrome (MWS) in patients 12 years of age or older.

DEFINITIONS

Familial Cold Autoinflammatory Syndrome (FCAS): an autosomal dominant inherited inflammatory disease characterized by episodes of rash, fever, and joint pain, following exposure to cold with attacks occurring 1-2 hours after exposure to cold and lasting less than 24 hours.

Muckle-Wells Syndrome (MWS): an autosomal dominant inherited inflammatory disease characterized by chronic recurrent urticaria, periodic arthritis, sensorineural deafness, general inflammation, and secondary amyloidosis.

NLRP3 gene: nucleotide-binding domain, leucine-rich family, pyrin domain containing 3 gene. This gene encodes cryopyrin, a protein contained in intracellular inflammasomes. Cryopyrin regulates protease caspase-1 and controls interleukin-1 beta.

POLICY

It is the policy of the Health Plan to maintain a prior authorization process that promotes appropriate utilization of specific drugs with potential for misuse or limited indications. This process involves a review

using Food and Drug Administration (FDA) criteria to make a determination of Medical Necessity, as defined in CRM.015-Medical Necessity, and approval by the Pharmacy & Therapeutics Committee of the criteria for prior authorization, as described in RX.003-Prior Authorization Process.

The drug, Arcalyst (Rilonacept), is subject to the prior authorization process.

PROCEDURE

Initial Authorization Criteria:

Must meet all of the criteria listed under the respective diagnosis:

1. For Muckle-Wells Syndrome (MWS)

- Must have a negative tuberculosis skin test [such as Tuberculin PPD (purified protein derivative) test] or Interferon-Gamma Release Assay (IGRA) whole-blood test [such as QuantiFERON®-TB Gold In-Tube test (QFT-GIT) or T-SPOT®.TB test (T-Spot)]
- Must be prescribed by a rheumatologist, dermatologist, immunologist, or genetic specialist
- Must be age 12 years and older
- Must have a diagnosis of Muckle-Wells syndrome (MWS). Chart documentation of a clinical work-up to rule out other diagnoses and clinical rationale for the diagnosis and exclusion of other diagnoses must be provided. The diagnosis must either be confirmed by genetic testing or a clinical diagnosis defined as one of the following scenarios:
    - Must have a mutation in the NLRP3 (formerly CIAS1) gene. Documentation of lab result confirming mutation is required.
    - Must meet 3 of the following diagnostic criteria (chart documentation required):
        - Autosomal dominant pattern of disease inheritance
        - Presence of severe fatigue
        - Presence of musculoskeletal symptoms (e.g. arthralgia, arthritis, myalgia)
        - Presence of ocular symptoms (e.g. conjunctivitis, anterior uveitis, papilledema)
        - Presence of erythematous rash
        - Duration of most febrile episodes lasting greater than 24 hours
        - Presence of amyloidosis
        - Presence of hearing loss
- Must have baseline lipid panel assessment
- Must currently not be using a Tumor Necrosis Factor (TNF) blocking agent or other biologic agent
- Must have no evidence of infection

2. For Familial Cold Autoinflammatory Syndrome (FCAS)

- Must have a negative tuberculosis skin test or IGRA whole-blood test
- Must be prescribed by a rheumatologist, dermatologist, immunologist, or genetic specialist
- Must be age 12 years and older
- Must have a diagnosis of Familial Cold Autoinflammatory Syndrome (FCAS). Chart documentation of a clinical work-up to rule out other diagnoses and clinical rationale for the diagnosis and exclusion of other diagnoses must be provided. The diagnosis must either be confirmed by genetic testing or a clinical diagnosis defined as one of the following scenarios:
    - Must have a mutation in the NLRP3 (formerly CIAS1) gene. Documentation of lab result confirming mutation is required.
    - Must meet 4 of the following diagnostic criteria (chart documentation required):
        - Recurrent intermittent episodes of fever and rash that primarily follow natural, experimental, or both types of generalized cold exposures

  - Autosomal dominant pattern of disease inheritance
  - Age of onset less than 6 months of age
  - Duration of most attacks less than 24 hours
  - Presence of conjunctivitis associated with attacks
  - Absence of deafness, periorbital edema, lymphadenopathy, and serositis
- Must have baseline lipid panel assessment
- Must currently not be using a Tumor Necrosis Factor (TNF) blocking agent or other biologic agent
- Must have no evidence of infection

Reauthorization Criteria:
All prior authorization renewals are reviewed on an annual basis to determine the medical necessity for continuation of therapy. Authorizations may be extended at one-year intervals based upon:

- Chart documentation from the prescriber that the member's disease has improved based upon the prescriber's assessment while on therapy
- Documentation that there is no evidence of infection
- Documentation that a lipid panel has been assessed within 3 months of initiation of therapy (for initial re-authorization) and at regular intervals thereafter (for annual reauthorizations)

Limitations:

Length of Authorization (if above criteria met)	
Initial Authorization	Up to 3 months
Reauthorization	Up to 1 year
Quantity Level Limit	
Arcalyst	4 vials per month

If the established criteria are not met, the request is referred to a Medical Director for review.

REFERENCES

1. Arcalyst [package insert]. Regeneron Pharmaceuticals: Tarrytown, NY; April 2010.
2. Hoffman HM, Throne ML, Amar J, et al. Efficacy and safety of rilonacept (interleukin-1 trap) in patients with cryopyrin-associated periodic syndromes. *Arthritis Rheum* 2008;58(8):2443-2452
3. Goldbach-Mansky R, Shroff SD, Wilson M, et al. A pilot study to evaluate the safety and efficacy of the long-acting interleukin-1 inhibitor rilonacept (interleukin-1 trap) in patients with familial cold autoinflammatory syndrome. *Arthritis Rheum* 2008;58(8):2432-2442
4. Hoffman HM, Wanderer AA, Broide DH. Familial cold autoinflammatory syndrome: phenotype and genotype of an autosomal dominant periodic fever. J Allergy Clin Immunol 2001;108:615-20
5. Aksentijevich I, Putnam CD, Remmers EF, et al. The clinical continuum of crypyrinopathies: novel CIAS1 mutation in north American patients and a new cryopyrin model. Arthritis Rheum 2007;56(4):1273-1285.
6. Kuemmerle-Deschner JB, Lohse P, Koetter I, et al. NLRP3 E311K mutation in a large family with Muckle-Wells syndrome – description of a heterogenous phenotype and response to treatment. Arthritis Research & Therapy 2011;13:R196
7. Kummerle-Deschner JB, Tyrrell PN, Reess F, et al. Risk factors for severe Muckle-Wells syndrome. Arthritis Rheum 2010;62(10):3783-3791

8. Kuemmerle-Deschner JB, Tyrrell P, Roetter I, et al. Efficacy and safety of anakinra therapy in pediatric and adult patients with the autoinflammatory Muckle-Wells Syndrome. Arthritis Rheum 2011;63(3):840-849
9. Yu JR, Kieron LS. Cryopyrin-associated periodic syndrome: an update on diagnosis and treatment response. Curr Allergy Asthma Rep 2011;11:12-20
10. Hashkes PJ. Autoinflammatory Syndromes. Pediatr Clin N Am 2012;59:447-470Hoffman HM. Familial cold autoinflammatory syndrome. Orphanet Encyclopedia. February 2005: http://orpha.net/data/patho/GB/uk-FCAS.pdf. Accessed March 9, 2013.
11. Grateu G. Muckle-wells syndrome. Orphanet Encyclopedia. June 2003. http://www.orpha.net/data/patho/GB/uk-MWS.pdf

RECORD RETENTION

Records Retention for Evolent Health documents, regardless of medium, are provided within the Evolent Health records retention policy and as indicated in CORP.028.E Records Retention Policy and Procedure.

REVIEW HISTORY

DESCRIPTION OF REVIEW / REVISION	DATE APPROVED
Annual Review	*02/17, 02/18*