Mass General Brigham
Health Plan

Kineret (anakinra)
Effective 02/01/2022

Plan	☐ MassHealth UPPL ☒Commercial/Exchange	**Program Type**	☒ Prior Authorization ☐ Quantity Limit ☐ Step Therapy
Benefit	☒ Pharmacy Benefit ☐ Medical Benefit (NLX)		
Specialty Limitations	This medication has been designated specialty and must be filled at a contracted specialty pharmacy.		
Contact Information	**Specialty Medications**		
	All Plans	Phone: 866-814-5506	Fax: 866-249-6155
	Non-Specialty Medications		
	MassHealth	Phone: 877-433-7643	Fax: 866-255-7569
	Commercial	Phone: 800-294-5979	Fax: 888-836-0730
	Exchange	Phone: 855-582-2022	Fax: 855-245-2134
	Medical Specialty Medications (NLX)		
	All Plans	Phone: 844-345-2803	Fax: 844-851-0882
Exceptions	N/A		

Overview
The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met, and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications
1. Moderately to severely active rheumatoid arthritis (RA)
2. Cryopyrin-Associated Periodic Syndromes (CAPS)
3. Neonatal-Onset Multisystem Inflammatory Disease (NOMID)
4. Deficiency of interleukin one receptor antagonist (DIRA)

Compendial Uses
1. Systemic juvenile idiopathic arthritis (sJIA)
2. Adult-onset Still's disease
3. Non-Hodgkin's lymphoma – Castleman's disease
4. Recurrent pericarditis
5. Hyperimmunoglobulin D syndrome [Mevalonate Kinase Deficiency (MKD)]

All other indications are considered experimental/investigational and are not a covered benefit.

Coverage Guidelines

Moderately to Severely Active Rheumatoid Arthritis (RA)

1. Authorization may be granted for members new to the plan who are currently receiving treatment with Kineret, excluding when the product is obtained as samples or via manufacturer's patient assistance programs.

OR

2. Authorization may be granted for treatment of moderately to severely active RA for members who meet one of the following criteria:
   a. Member has experienced an inadequate response or intolerance to ALL preferred products (Enbrel, Humira and Rinvoq).
   b. Member has a contraindication to Enbrel, Humira AND Rinvoq and meets one of the following:
      - Member has experienced an inadequate response to at least a 3-month trial of methotrexate despite adequate dosing (i.e., titrated to 20 mg/week).
      - Member has an intolerance or contraindication to methotrexate (see Appendix).

Adult-Onset Still's Disease

Authorization may be granted for members who meet ANY of the following criteria:

1. Member has experienced an inadequate response to at least a 3-month trial of methotrexate
2. Member has intolerance or contraindication to methotrexate (See Appendix)
3. Member has a febrile disease

Active Systemic Juvenile Idiopathic Arthritis (sJIA)

1. Authorization may be granted for the treatment of sJIA for members who have received Actemra or Ilaris in a paid claim through a pharmacy or medical benefit within the previous 120 days.

OR

2. Authorization may be granted for the treatment of active sJIA for members who have had an inadequate response to a trial of NSAIDS and one of the following: corticosteroids, methotrexate, or leflunomide.

Neonatal-Onset Multisystem Inflammatory Disease (NOMID)

Authorization of 24 months may be granted for the treatment of Cryopyrin-associated periodic syndromes (CAPS), including NOMID (also known as Chronic Infantile Neurological Cutaneous and Articular syndrome (CINCA).

Recurrent Pericarditis

Authorization of 12 months may be granted for the treatment of recurrent pericarditis for members who have failed a first-line therapy agent (i.e., colchicine).

Non-Hodgkin's Lymphoma – Multicentric Castleman's Disease

Authorization of 12 months may be granted for the treatment of multicentric Castleman's disease.

Hyperimmunoglobulin D Syndrome [Mevalonate Kinase Deficiency (MKD)]

Authorization of 24 months may be granted for the treatment of hyperimmunoglobulin D syndrome.

Deficiency of Interleukin One Receptor Antagonist (DIRA)

Authorization may be granted for members who meet all of the following criteria:

1. The diagnosis has been confirmed by genetic testing documenting mutations involving the IL1RN
2. The diagnosis of primary immunodeficiency has been ruled out
3. The member has experienced at least one of the following conditions:

- Infantile pustulosis (neonatal onset pustulosis
- Infantile pustular psoriasis
- SAPHO syndrome (synovitis, acne, pustulosis, hyperostosis and osteitis

4. The member has failed high-dose corticosteroids

Continuation of Therapy

Adult-Onset Still's Disease, Rheumatoid Arthritis and Juvenile Idiopathic Arthritis
Reauthorization may be granted for all members (including new members) who have achieved or maintained a positive clinical response after at least 3 months of therapy with Kineret as evidenced by low disease activity or improvement in signs and symptoms of the condition.

Neonatal-Onset Multisystem Inflammatory Disease (NOMID), Multicentric Castleman's disease, and Hyperimmunoglobulin D Syndrome
Reauthorization of may be granted for all members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria and documentation has been submitted of positive clinical response as evidenced by low disease activity or improvement in signs and symptoms of the condition.

Recurrent Pericarditis
Authorization of 6-month intervals may be granted for all members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria and documentation has been submitted of positive clinical response as evidenced by low disease activity or improvement in signs and symptoms of the condition.

Deficiency of Interleukin One Receptor Antagonist (DIRA)
Authorization may be granted for all members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria and documentation has been submitted of positive clinical response as evidenced by low disease activity or improvement in signs and symptoms of the condition.

Limitations

1. Initial approvals will be based on diagnosis:
    a. Moderately to Severely Active Rheumatoid Arthritis (RA) – 24 months
    b. Adult-Onset Still's Disease – 24 months
    c. Active Systemic Juvenile Idiopathic Arthritis (sJIA) – 24 months
    d. Neonatal-Onset Multisystem Inflammatory Disease (NOMID) – 24 months
    e. Recurrent Pericarditis – 12 months
    f. Non-Hodgkin's Lymphoma – Multicentric Castleman's Disease – 12 months
    g. Hyperimmunoglobulin D Syndrome [Mevalonate Kinase Deficiency (MKD)] – 24 months
    h. Deficiency of Interleukin One Receptor Antagonist (DIRA) – 12 months

2. Reauthorizations will be based on diagnosis:
    a. Adult-Onset Still's Disease, Rheumatoid Arthritis and Juvenile Idiopathic Arthritis – 24 months
    b. Neonatal-Onset Multisystem Inflammatory Disease (NOMID), Multicentric Castleman's disease, Hyperimmunoglobulin D Syndrome and DIRA- 12 months
    c. Recurrent Pericarditis – 6 months

Appendix

Examples of Contraindications to Methotrexate

1. History of intolerance or adverse event
2. Alcoholic liver disease or other chronic liver disease
3. Elevated liver transaminases
4. Interstitial pneumonitis or clinically significant pulmonary fibrosis
5. Renal impairment
6. Current pregnancy or planning pregnancy
7. Breastfeeding
8. Blood dyscrasias (e.g., thrombocytopenia, leukopenia, significant anemia)
9. Myelodysplasia
10. Hypersensitivity
11. Significant drug interaction

References

1. Kineret [package insert]. Stockholm, Sweden: Swedish Orphan Biovitrum AB (publ); December 2020.
2. National Organization for Rare Disorders. URL: https://www.rarediseases.org/rare-disease-information/rare-diseases/byID/1210/viewFullReport. Accessed April 18, 2017.
3. Adler Y, Charron P, Imazio M, et al. 2015 ESC Guidelines for the diagnosis and management of pericardial diseases: The Task Force for the Diagnosis and Management of Pericardial Diseases of the European Society of Cardiology (ESC) Endorsed by: The European Association for Cardio-Thoracic Surgery (EACTS). Eur Heart J. 2015 Nov 7; 36 (42): 2921-64.
4. Kostjukovits S, Kalliokoski L, Antila K, Korppi M. Treatment of hyperummunoglobulinemia D Syndrome with biologics in children: review of the literature and Finnish experience. Eur J Pediatr. 2015 Jun; 174 (6): 707-14
5. National Organization for Rare Disorders. Hyperimmunoglobulin D Syndrome. http://rarediseases.org/rare-diseases/hyper-igd-syndrome. Accessed April 18, 2017.
6. American College of Rheumatology. Hyperimmunoglobulin D Syndrome. http://www.rheumatology.org/I-AM-A/Patient-Caregiver/Diseases-Conditions/Hyperimmunoglobulin-D-Syndrome-Juvenile. Accessed April 18, 2017.
7. Ortiz-Sanjuán F, Blanco R, Riancho-Zarrabeitia L, et al. Efficacy of Anakinra in Refractory Adult-Onset Still's Disease: Multicenter Study of 41 Patients and Literature Review. Medicine (Baltimore) 2015; 94: e1554.
8. Lazaros G, Imazio M, Brucato A, et al. Anakinra: an emerging option for refractory idiopathic recurrent pericarditis: a systematic review of published evidence. J Cardiovasc Med (Hagerstown) 2016; 17:256.
9. Brucato A, Imazio M, Gattorno M, et al. Effect of anakinra on recurrent pericarditis among patients with colchicine resistance and corticosteroid dependence: the AIRTRIP randomized clinical trial. *JAMA*. 2016;316(18):1906-1912.[PubMed 27825009]
10. Ringold S, Weiss PF, Beukelman T, et al. 2013 Update of the 2011 American College of Rheumatology Recommendations for the Treatment of Juvenile Idiopathic Arthritis: Recommendations for the Medical Therapy of Children With Systemic Juvenile Idiopathic Arthritis and Tuberculosis Screening Among Children Receiving Biologic Medications. *Arthritis & Rheumatism.* 2013; 65:2499-2512.
11. The NCCN Drugs & Biologics Compendium™. National Comprehensive Cancer Network, Inc. http://www.nccn.org. Accessed April 24, 2017.
12. Cowen EW, Goldbach-Mansky R. DIRA, DITRA, and new insights into pathways of skin inflammation: what's in a name? Arch Dermatol 2012; 148:381

13. Nakagawa K, Gonzalez-Roca E, Souto A, et al. Somatic NLRP3 mosaicism in Muckle-Wells syndrome. A genetic mechanism shared by different phenotypes of cryopyrin-associated periodic syndromes. Ann Rheum Dis 2015; 74:603

Review History

03/21/05 – Reviewed
05/15/05 – Implemented
02/27/06 – Reviewed
02/25/08 – Reviewed
02/23/09 – Reviewed
02/22/10 – Reviewed
02/28/11 – Reviewed
02/27/12 – Reviewed
02/25/13 – Reviewed
02/24/14 – Reviewed
02/23/15 – Reviewed
02/22/16 – Reviewed
02/27/17 – Adopted SGM & PDS
02/26/18 – Updated
02/20/19 – Updated
11/20/19 - Added Rinvoq as required preferred trial for RA
09/22/2021 – Reviewed and Updated Sept P&T; Added new indication of DIRA with criteria and limitations; References updated. Effective 02/01/2022.
09/21/2022 - Reviewed at Sept P&T; no clinical changes.