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DRUG POLICY

Kineret (anakinra)

NOTICE

This policy contains information which is clinical in nature. The policy is not medical advice. The information in this policy is used by Wellmark to make determinations whether medical treatment is covered under the terms of a Wellmark member's health benefit plan. Physicians and other health care providers are responsible for medical advice and treatment. If you have specific health care needs, you should consult an appropriate health care professional. If you would like to request an accessible version of this document, please contact customer service at 800-524-9242.

BENEFIT APPLICATION

Benefit determinations are based on the applicable contract language in effect at the time the services were rendered. Exclusions, limitations or exceptions may apply. Benefits may vary based on contract, and individual member benefits must be verified. Wellmark determines medical necessity only if the benefit exists and no contract exclusions are applicable. This medical policy may not apply to FEP. Benefits are determined by the Federal Employee Program.

DESCRIPTION

The intent of the Kineret drug policy is to ensure appropriate selection of patients for therapy based on product labeling, clinical guidelines and clinical studies while steering utilization to the most cost-effective medication within the therapeutic class. For this program, adalimumab-aacf, Enbrel, Cosentyx, Otezla, Rinvoq, Simponi, Skyrizi, Otulfi (ustekinumab-aaaz), Tremfya, and Xeljanz/Xeljanz XR are the preferred products and will apply to members requesting treatment for an indication that is FDA-approved for the preferred product. The criteria will require the use of two of the health plan's preferred products before the use of non-preferred products unless there are clinical circumstances that exclude the use of all the preferred products, the patient is currently receiving treatment with the non-preferred drug and experience a positive therapeutic outcome, or there is only one preferred product for an indication.

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) in patients 18 years of age or older who have failed 1 or more disease modifying antirheumatic drugs (DMARDs)
2. Cryopyrin-Associated Periodic Syndromes (CAPS), including Neonatal-Onset Multisystem Inflammatory Disease (NOMID)
3. Deficiency of Interleukin-1 Receptor Antagonist (DIRA)

Compendial Uses

1. Systemic juvenile idiopathic arthritis (sJIA)
2. Adult-onset Still's disease (AOSD)

3. Multicentric Castleman's disease
4. Recurrent pericarditis (RP)
5. Hyperimmunoglobulin D syndrome (HIDS) (Mevalonate Kinase Deficiency [MKD])
6. Schnitzler's syndrome
7. Gout and pseudogout (calcium pyrophosphate deposition)
8. CAR T-Cell Related Toxicities – Cytokine release syndrome (CRS)
9. Erdheim-Chester Disease
10. Immune checkpoint inhibitor-related toxicity—immunotherapy-related hemophagocytic lymphohistiocytosis (HLH)-like syndrome

POLICY

Required Documentation:

Submission of the following information is necessary to initiate prior authorization review:

Rheumatoid arthritis (RA)

1. For initial requests:
 - A. Chart notes, medical record documentation, or claims history supporting previous medications tried (if applicable), including response to therapy. If therapy is not advisable, documentation of clinical reason to avoid therapy.
 - B. Laboratory results, chart notes, or medical record documentation of biomarker testing (i.e., rheumatoid factor [RF], anti-cyclic citrullinated peptide [anti-CCP], and C-reactive protein [CRP] and/or erythrocyte sedimentation rate [ESR]) (if applicable).
2. For continuation requests: Chart notes or medical record documentation supporting positive clinical response.

Adult-onset Still's disease (AOSD) and systemic juvenile idiopathic arthritis (sJIA)

1. Initial requests: Chart notes, medical record documentation, or claims history supporting previous medications tried (if applicable), including response to therapy. If therapy is not advisable, documentation of clinical reason to avoid therapy.
2. Continuation requests: Chart notes or medical record documentation supporting positive clinical response.

Neonatal-onset multisystem inflammatory disease (NOMID): For continuation requests

1. Chart notes, medical record documentation, or laboratory results supporting positive clinical response.

Deficiency of interleukin-1 receptor antagonist (DIRA)

2. Initial requests: *IL1RN* mutation status.

Recurrent pericarditis

1. Initial requests: Chart notes, medical record documentation, or claims history supporting previous medications tried, including response to therapy.
2. Continuation requests: Chart notes or medical record documentation supporting positive clinical response.

Hyperimmunoglobulin D syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD)

1. Initial requests: Chart notes, medical record documentation, or laboratory result (if applicable) indicating number of active flares within the last 6 months and Physician's Global Assessment score or C-reactive protein (CRP) level.

Gout and pseudogout flares, CAR T related toxicities, and Immune Checkpoint Inhibitor-Related Toxicity:For initial requests

1. Chart notes, medical record documentation, or claims history supporting previous medications tried (if applicable), including response to therapy. If therapy is not advisable, documentation of clinical reason to avoid therapy.

Must meet BOTH the Preferred Drug Plan Design and Criteria for Initial Approval/Continuation of Therapy when both are applicable.

Preferred Drug Plan Design

Rheumatoid Arthritis

1. Criteria for initial approval for rheumatoid arthritis will only apply when at least ONE of the following criteria are met:
 - A. Member has had an inadequate response to treatment or intolerable adverse event with at least TWO of the preferred products (Enbrel, adalimumab-aacf, Rinvoq, Simponi, and Xeljanz/Xeljanz XR)
 - B. Member has a clinical reason to avoid TNF-inhibitors (Enbrel, adalimumab-aacf, and Simponi) (See Appendix A) AND has had an inadequate response to treatment or intolerable adverse event with the preferred products, Rinvoq AND Xeljanz or Xeljanz XR
 - C. Member is currently receiving treatment with the requested product through insurance (excludes obtainment as samples or via manufacturer's patient assistance programs) and experiencing a positive therapeutic outcome

Note: Submission of chart notes detailing the outcomes of treatment, intolerable adverse event(s) experienced, contraindication(s), or exclusion(s) to treatment with preferred product(s) is required (where applicable).

Prescriber Specialties (initial approvals only)

This medication must be prescribed by or in consultation with one of the following:

1. Rheumatoid arthritis, adult-onset Still's disease, and systemic juvenile idiopathic arthritis, gout, and pseudogout: rheumatologist
2. Cryopyrin-associated periodic syndromes (CAPS), including neonatal-onset multisystem inflammatory disease (NOMID), deficiency of interleukin-1 receptor antagonist (DIRA), and hyperimmunoglobulin D syndrome (HIDS)/mevalonate kinase deficiency (MKD): rheumatologist or immunologist
3. Recurrent pericarditis: cardiologist, rheumatologist, or immunologist
4. Schnitzler's syndrome: rheumatologist, dermatologist, or immunologist
5. Multicentric Castleman's disease, CAR T-cell related toxicities, Erdheim-Chester disease, and immune checkpoint inhibitor-related toxicity: oncologist or hematologist

Criteria for Initial Approval

Moderately to Severely Active Rheumatoid Arthritis (RA)

1. Authorization of 12 months may be granted for adult members who have previously received a biologic or targeted synthetic drug (e.g., Rinvoq, Xeljanz) indicated for moderately to severely active rheumatoid arthritis.
2. Authorization of 12 months may be granted for adult members for treatment of moderately to severely active RA when all of the following criteria are met:
 - A. Member meets either of the following criteria:
 - 1) Member has been tested for either of the following biomarkers and the test was positive:
 - a. Rheumatoid factor (RF)
 - b. Anti-cyclic citrullinated peptide (anti-CCP)
 - 2) Member has been tested for ALL of the following biomarkers:
 - a. RF

- b. Anti-CCP
 - c. C-reactive protein (CRP) and/or erythrocyte sedimentation rate (ESR)
- B. Member has experienced an inadequate response to at least a 3-month trial of methotrexate despite adequate dosing (i.e., titrated to at least 15 mg/week), or the member has an intolerance or contraindication to methotrexate (see Appendix B).
- C. Member has experienced an inadequate response to at least a 3-month trial of a biologic or a targeted synthetic drug (e.g., Rinvoq, Xeljanz) or has an intolerance to a biologic or targeted synthetic drug.

Adult-onset Still's Disease (AOSD)

1. Authorization of 12 months may be granted for members who have received a biologic indicated for active adult-onset Still's disease.
2. Authorization of 12 months may be granted for treatment of adult-onset Still's disease when both of the following criteria is met:
 - A. Member has active systemic features (e.g., fever, arthralgia/arthritis, evanescent rash, lymphadenopathy, hepatomegaly, splenomegaly, sore throat).
 - B. Member meets any of the following:
 - 1) Member has had an inadequate response to a trial of nonsteroidal anti-inflammatory drugs (NSAIDs).
 - 2) Member has had an inadequate response to a trial of corticosteroids.
 - 3) Member has had an inadequate response to a trial of a conventional synthetic drug (e.g., methotrexate).

Systemic Juvenile Idiopathic Arthritis (sJIA)

1. Authorization of 12 months may be granted for treatment of active sJIA for members who have previously received a biologic indicated for active sJIA.
2. Authorization of 12 months may be granted for the treatment of active sJIA when both of the following criteria are met:
 - A. Member has active systemic features (e.g., fever, evanescent rash, lymphadenopathy, hepatomegaly, splenomegaly, or serositis).
 - B. Member has had an inadequate response to non-steroidal anti-inflammatory drugs (NSAIDs) or systemic glucocorticoids.

Neonatal-Onset Multisystem Inflammatory Disease (NOMID)

1. Authorization of 12 months may be granted for treatment of cryopyrin-associated periodic syndromes (CAPS), including NOMID (also known as chronic infantile neurologic cutaneous and articular syndrome [CINCA]).

Deficiency of interleukin-1 receptor antagonist (DIRA)

1. Authorization of 12 months may be granted for the treatment of genetically confirmed deficiency of interleukin-1 receptor antagonist (DIRA) due to *IL1RN* mutations.

Recurrent Pericarditis

1. Authorization of 12 months may be granted for the treatment of recurrent pericarditis when both of the following criteria are met:
 - A. Member has had at least two episodes of pericarditis.
 - B. Member has failed at least 2 agents of standard therapy (e.g., colchicine, non-steroidal anti-inflammatory drugs [NSAIDs], corticosteroids).

Multicentric Castleman's Disease

1. Authorization of 12 months may be granted for the treatment of multicentric Castleman's disease when both of the following criteria are met:

- A. The requested medication will be used as a single-agent.
- B. The disease has progressed following treatment of relapsed/refractory or progressive disease.

Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD)

1. Authorization of 12 months may be granted for the treatment of HIDS/MKD when all of the following criteria are met:
 - A. Member has had active flares within the last 6 months
 - B. Physician's Global Assessment greater than or equal to 2 or C-reactive protein (CRP) greater than 10 mg/L

Schnitzler's syndrome

1. Authorization of 12 months may be granted for treatment of Schnitzler's syndrome when all of the following criteria are met:
 - A. Member has an urticarial rash, monoclonal IgM (or IgG) gammopathy and at least two of the following signs and symptoms: fever, joint pain or inflammation, bone pain, palpable lymph nodes, enlargement of the liver or spleen, elevated numbers of white blood cells (leukocytosis), elevated red blood cell (erythrocyte) sedimentation rate or abnormalities on bone morphological study (e.g., increased bone density)
 - B. Other possible causes of the signs and symptoms have been ruled out, including but not limited to: hyperimmunoglobulin D syndrome, adult-onset Still disease, urticarial hypocomplementemic vasculitis, acquired C1 inhibitor deficiency and cryoglobulinemia.

Gout and pseudogout flares

1. Authorization of 12 months may be granted for adult members for the treatment of flares for gout and pseudogout (also known as calcium pyrophosphate deposition disease) when both of the following criteria are met:
 - A. Member has experienced at least three gout flares in the last 12 months
 - B. Member has had an inadequate response, intolerance, or contraindication to non-steroidal anti-inflammatory (NSAIDs), colchicine, and corticosteroids

CAR T-cell related toxicities and Cytokine release syndrome (CRS)

1. Authorization of 1 month may be granted for the management of chimeric antigen receptor (CAR) T cell induced cytokine release syndrome when the following criteria is met:
 - A. Cytokine release syndrome is refractory to high dose corticosteroids and anti-IL-6 therapy
 - B. Kineret will be used as replacement for the second dose of tocilizumab when supplies are limited or unavailable
2. Authorization of 3 months may be granted for the prophylaxis of CAR T-cell-induced toxicity in members at high risk of developing high-grade immune effector cell-associated neurotoxicity syndrome (ICANS).

Erdheim-Chester Disease

1. Authorization of 12 months may be granted for the treatment of Erdheim-Chester disease.

Immune Checkpoint Inhibitor-Related Toxicity

1. Authorization of 6 months may be granted for the treatment of immune checkpoint inhibitor-related toxicity when the member has immunotherapy-related hemophagocytic lymphohistiocytosis (HLH)-like syndrome and either of the following criteria is met:
 - A. Member has had an inadequate response to systemic corticosteroids.
 - B. Member has an intolerance or contraindication to corticosteroids.

Continuation of Therapy

Moderately to severely active rheumatoid arthritis (RA)

1. Authorization of 12 months may be granted for all members (including new members) who are using the requested medication for moderately to severely active rheumatoid arthritis and who achieve or maintain a positive clinical response as evidenced by disease activity improvement of at least 20% from baseline in tender joint count, swollen joint count, pain, or disability.

Adult-onset Still's disease (AOSD) and systemic juvenile idiopathic arthritis (sJIA)

1. Authorization of 12 months may be granted for all members (including new members) who are using the requested medication for adult-onset Still's disease or systemic juvenile idiopathic arthritis and who achieve or maintain a positive clinical response as evidenced by low disease activity or improvement in signs and symptoms of the condition when there is improvement in any of the following from baseline:
 - A. Number of joints with active arthritis (e.g., swelling, pain, limitation of motion)
 - B. Number of joints with limitation of movement
 - C. Functional ability
 - D. Systemic features (e.g., fever, evanescent skin rash, lymphadenopathy, hepatomegaly, splenomegaly, or serositis)

Neonatal-onset multisystem inflammatory disease (NOMID)

1. Authorization of 12 months may be granted for all members (including new members) who are using the requested medication for CAPS, including NOMID (also known as CINCA), and who achieve or maintain a positive clinical response as evidenced by low disease activity or improvement in signs and symptoms of the condition when there is improvement in any of the following from baseline:
 - A. Fever
 - B. Skin rash
 - C. Joint pain and/or inflammation
 - D. Central nervous system (CNS) symptoms (e.g., meningitis, headache, cerebral atrophy, uveitis, hearing loss)
 - E. Inflammatory markers (e.g., serum amyloid A [SAA], C-reactive protein [CRP], erythrocyte sedimentation rate [ESR])

Recurrent pericarditis

1. Authorization of 12 months may be granted for all members (including new members) who are using the requested medication for recurrent pericarditis and who achieve or maintain a positive clinical response as evidenced by low disease activity or improvement in signs and symptoms of the condition when there is improvement in any of the following:
 - A. Pericarditic chest pain
 - B. Pericardial rubs
 - C. Electrocardiogram (ECG)
 - D. Pericardial effusion
 - E. C-reactive protein (CRP)

Multicentric Castleman's disease

1. Authorization of 12 months may be granted for continued treatment of multicentric Castleman's disease in members requesting reauthorization who have not experienced disease progression or an unacceptable toxicity.

CAR T-Cell-Related Toxicities and Immune Checkpoint Inhibitor-Related Toxicity

1. All members (including new members) requesting authorization for continuation of therapy must meet all requirements in the coverage criteria.

All other indications

1. Authorization of 12 months may be granted for all members (including new members) who are using the requested medication for an indication outlined in the Criteria for Initial Approval and who achieve or maintain positive clinical response as evidenced by low disease activity or improvement in signs and symptoms of the condition.

Other

For all indications: Member has had a documented negative TB test (which can include a tuberculosis skin test [TST] or an interferon-release assay [IGRA])* within 12 months of initiating therapy for persons who are naïve to biologic drugs or targeted synthetic drugs associated with an increased risk of TB.

* If the screening testing for TB is positive, there must be further testing to confirm there is no active disease (e.g., a chest x-ray). Do not administer the requested medication to members with active TB infection. If there is latent disease, TB treatment must be started before initiation of treatment.

For all indications: Member cannot use the requested medication concomitantly with any other biologic drug or targeted synthetic drug.

Dosage and Administration

Approvals may be subject to dosing limits in accordance with FDA-approved labeling, accepted compendia, and/or evidence-based practice guidelines.

Kineret is considered **not medically necessary** for members who do not meet the criteria set forth above.

Quantity Limits

Trade Name	Generic Name	Quantity Limit
Kineret®	anakinra	<p>RA: 28 syringes per 28 days</p> <p>NOMID and DIRA: Initial dosing of 1mg/kg, maintenance dosing of 3-4mg/kg and maximum dosing of 8mg/kg daily</p> <p>For all other indications: 112 syringes per 28 days</p>

Appendices

Appendix A: Clinical reasons to avoid TNF-inhibitors

1. History of demyelinating disorder
2. History of congestive heart failure
3. History of hepatitis B infection
4. Autoantibody formation/lupus-like syndrome
5. Risk of lymphoma

Appendix B: Examples of Contraindications to Methotrexate

1. Clinical diagnosis of alcohol use disorder, alcoholic liver disease or other chronic liver disease
2. Breastfeeding
3. Blood dyscrasias (e.g., thrombocytopenia, leukopenia, significant anemia)
4. Elevated liver transaminases
5. History of intolerance or adverse event
6. Hypersensitivity
7. Interstitial pneumonitis or clinically significant pulmonary fibrosis
8. Myelodysplasia
9. Pregnancy or currently planning pregnancy
10. Renal impairment

11. Significant drug interaction

PROCEDURES AND BILLING CODES

To report provider services, use appropriate CPT* codes, Alpha Numeric (HCPCS level 2) codes, Revenue codes, and/or ICD diagnostic codes.

- N/A

REFERENCES

- Kineret [package insert]. Stockholm, Sweden: Swedish Orphan Biovitrum AB (publ); December 2020.
- DRUGDEX® System (electronic version). Truven Health Analytics, Ann Arbor, MI. Available at <http://www.micromedexsolutions.com> [available with subscription]. Accessed November 15, 2020.
- 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.
- Laskari K, Tzioufas AG, Moutsopoulos HM. Efficacy and long-term follow-up of IL-1R inhibitor anakinra in adults with Still's disease: a case-series study. *Arthritis Res Ther*. 2011;13(3):R91.
- Lequerre T, Quartier P, Rosellini D, et al. Interleukin-1 receptor antagonist (anakinra) treatment in patients with systemic-onset juvenile idiopathic arthritis or adult onset Still disease: preliminary experience in France. *Ann Rheum Dis*. 2008;67:302-308.
- The NCCN Drugs & Biologics Compendium™. National Comprehensive Cancer Network, Inc. <http://www.nccn.org>. Accessed November 15, 2020.
- Smolen JS, Landewé R, Billsma J, et al. EULAR recommendations for the management of rheumatoid arthritis with synthetic and biological disease-modifying antirheumatic drugs: 2019 update. *Ann Rheum Dis*. 2020;79(6):685-699.
- Singh JA, Furst DE, Bharat A, et al. 2012 Update of the 2008 American College of Rheumatology recommendations for the use of disease-modifying antirheumatic drugs and biologic agents in the treatment of rheumatoid arthritis. *Arthritis Care Res*. 2012;64(5):625-639.
- Saag KG, Teng GG, Patkar NM, et al. American College of Rheumatology 2008 recommendations for the use of nonbiologic and biologic disease-modifying antirheumatic drugs in rheumatoid arthritis. *Arthritis Rheum*. 2008;59(6):762-784.
- Aletaha D, Neogi T, Silman AJ, et al. 2010 Rheumatoid Arthritis Classification Criteria. An American College of Rheumatology/European League Against Rheumatism Collaborative Initiative. *Arthritis Rheum*. 2010;62:2569-2581.
- Anderson J, Caplan L, Yazdany J, et al. Rheumatoid Arthritis Disease Activity Measures: American College of Rheumatology Recommendations for Use in Clinical Practice. *Arthritis Rheum*. 2010;64:640-647.
- Beukelman T, Patkar NM, Saag KG, et al. 2011 American College of Rheumatology recommendations for the treatment of juvenile idiopathic arthritis: initiation and safety monitoring of therapeutic agents for the treatment of arthritis and systemic features. *Arthritis Care Res*. 2011;63(4):465-482.
- Quartier P, Allantaz F, Cimaz R, et al. A multicentre, randomized, double-blind, placebo-controlled trial with the interleukin-1 receptor antagonist anakinra in patients with systemic-onset juvenile idiopathic arthritis (ANAJIS trial). *Ann Rheum Dis*. 2011;70:747-754.
- Efthimiou P, Paik P K, Bielory L. Diagnosis and Management of Adult Onset Still's Disease. *Ann Rheum Dis*. 2006 May;65(5):564-72. Epub 2005 Oct 11.
- National Organization for Rare Disorders. Adult Onset Still's Disease. URL: <https://www.rarediseases.org/rare-disease-information/rare-diseases/byID/1210/viewFullReport>. Accessed November 15, 2020.

- 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.
- Kostjukovits S, Kalliokoski L, Antila K, Korppi M. Treatment of hyperimmunoglobulinemia D Syndrome with biologics in children: review of the literature and Finnish experience. *Eur J Pediatr*. 2015 Jun;174(6):707-14.
- National Organization for Rare Disorders. Mevalonate Kinase Deficiency. <http://rarediseases.org/rare-diseases/hyper-igd-syndrome>. Accessed November 15, 2020.
- American College of Rheumatology. Hyperimmunoglobulin D Syndrome. <http://www.rheumatology.org/I-AM-A/Patient-Caregiver/Diseases-Conditions/Hyperimmunoglobulin-D-Syndrome-Juvenile>. Accessed November 15, 2020.
- Simon A, Asli B, Braun-Falco M, et al. Schnitzler's syndrome: diagnosis, treatment, and follow-up. *Allergy*. 2013;68:562–568.
- Lipsker: The Schnitzler syndrome. *Orphanet Journal of Rare Diseases*. 2010;5:38.
- Centers for Disease Control and Prevention. Tuberculosis (TB). TB risk factors. Available at: <https://www.cdc.gov/tb/topic/basics/risk.htm>. Accessed: November 15, 2021.
- Lyseng-Williamson KA. Anakinra in Still's disease: A profile of its use. *Drugs Ther Perspect*. 2018;34(12):543-553.
- Yoo DH. Biologics for the treatment of adult-onset still's disease. *Expert Opin Biol Ther*. 2019 Aug 14:1-18 [Epub ahead of print].
- Richette P, Doherty M, Pascual E, et al. 2016 updated EULAR evidence-based recommendations for the management of gout. *Ann Rheum Dis*. 2017;76:29–42.
- Zhang W, Doherty M, Pascual E, et al. EULAR recommendations for calcium pyrophosphate deposition. Part II: Management. *Ann Rheum Dis*. 2011;70:571–575.
- Ottaviani S., Brunier L, Sibilia J, et al. Efficacy of anakinra in calcium pyrophosphate crystal-induced arthritis: A report of 16 cases and review of the literature. *Joint Bone Spine*. 2013;80:178-182.
- FitzGerald JD, Dalbeth N, Mikuls T, et al. 2020 American College of Rheumatology Guideline for the Management of Gout [published correction appears in *Arthritis Care Res (Hoboken)*. 2020 Aug;72(8):1187]. *Arthritis Care Res (Hoboken)*. 2020;72(6):744-760.
- Aksentijevich I, Masters SL, Ferguson PJ, et al. An autoinflammatory disease with deficiency of the interleukin-1-receptor antagonist. *N Engl J Med*. 2009;360(23):2426-37.
- Bonilla FA, Khan DA, Ballas ZK, et al. Practice parameter for the diagnosis and management of primary immunodeficiency. *J Allergy Clin Immunol*. 2015;136(5):1186-205.
- Smolen JS, Aletaha D. Assessment of rheumatoid arthritis activity in clinical trials and clinical practice. In: UpToDate, Post TW (Ed), UpToDate, Waltham, MA. Available with subscription. URL: www.uptodate.com. Accessed March 19, 2021.
- Fraenkel L, Bathon JM, England BR, et al. 2021 American College of Rheumatology guideline for the treatment of rheumatoid arthritis. *Arthritis Care Res*. 2021;0:1-16.
- Chiabrando JG, Bonaventura A, Vecchié, et al. Management of acute and recurrent pericarditis: JACC State-of-the-art review. *J Am Coll Cardiol*. 2020;75(1):76-92.
- Efthimiou P, Kontzias A, Hur P, et al. Adult-onset Still's disease in focus: Clinical manifestations, diagnosis, treatment, and unmet needs in the era of targeted therapies. *Semin Arthritis Rheum*. 2021;51(4):858-874.
- Onel KB, Horton DB, Lovell DJ, et al. 2021 American College of Rheumatology guideline for the treatment of juvenile idiopathic arthritis: therapeutic approaches for oligoarthritis, temporomandibular joint arthritis, and systemic juvenile idiopathic arthritis. *Arthritis Rheumatol*. 2022;74(4):553-569.

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POLICY HISTORY

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