Pharmacy Medical Necessity Guidelines: Hemlibra® (emicizumab-kxwh)

Effective: January 14, 2020

Prior Authorization Required	√	Type of Review – Care Management	
Not Covered		Type of Review – Clinical Review	√
Pharmacy (RX) or Medical (MED) Benefit	Rx	Department to Review	RXUM

These pharmacy medical necessity guidelines apply to the following:

Commercial Products

☒ Tufts Health Plan Commercial products – large group plans
☒ Tufts Health Plan Commercial products – small group and individual plans
☒ Tufts Health Freedom Plan products – large group plans
☒ Tufts Health Freedom Plan products – small group plans
- CareLink℠ – Refer to CareLink Procedures, Services and Items Requiring Prior Authorization

Tufts Health Public Plans Products

☒ Tufts Health Direct – A Massachusetts Qualified Health Plan (QHP) (a commercial product)
☒ Tufts Health Together – MassHealth MCO Plan and Accountable Care Partnership Plans
☒ Tufts Health RITogether – A Rhode Island Medicaid Plan

Fax Numbers:

RXUM: 617.673.0988

Note: This guideline does not apply to Medicare Members (includes dual eligible Members).

OVERVIEW

FOOD AND DRUG ADMINISTRATION (FDA)-APPROVED INDICATIONS

Hemlibra (emicizumab-kxwh) is a bispecific factor IXa- and factor X-directed antibody indicated for routine prophylaxis to prevent or reduce the frequency of bleeding episodes in adults and pediatric patients with hemophilia A (congenital factor VIII deficiency) with or without factor VIII inhibitors.

Hemophilia is one of the most common congenital bleeding disorders known to be due to defects in distinct and unrelated genes. Hemophilia is a clinically heterogeneous disorder resulting in deficiency of plasma factor VIII (FVIII) or factor IX (FIX) coagulant activity. There are two main types of hemophilia: hemophilia A (also known as antihemophilic factor [AHF] deficiency, FVIII deficiency, or classic hemophilia) and hemophilia B (also known as FIX deficiency or Christmas disease). Both types of hemophilia are X-linked bleeding disorders almost solely affecting males. Both FVIII and FIX deficiencies increase the risk of bleeding by reducing the amount of activated factor X (FX) and thrombin available to make a stable fibrin clot. Depending on the severity of the disease, a hemorrhage can occur spontaneously or can be precipitated by trauma.

The severity of bleeding in hemophilia is directly related to the degree of factor deficiency. Severity of hemophilia A and B factor deficiency is classified as severe, moderate, or mild, depending on the degree of factor levels present and relating directly to the expected frequency of bleeding. Normal factor levels are 50-150%. Severe hemophilia A or B is defined as a factor level of less than 1%; moderate hemophilia A or B is defined as a factor level of 1-5%; and mild hemophilia is defined as a factor level of 5-40%. Patients with severe hemophilia usually bleed frequently (one to two times per week) into muscles or joints and it often happens spontaneously. In moderate to mild disease, spontaneous bleeding is rare, and may never occur in mild disease. Bleeding in moderate to mild disease may occur after surgery, injury, or dental work.

Inhibitors can develop when a person with hemophilia has an immune response to treatment with clotting factor concentrates (factor products). Inhibitors, also called antibodies, form in the blood to fight against foreign proteins in the factor concentrates and they prevent the factor concentrate from being able to stop the bleeding. Overall disease management becomes even more complicated in patients who have developed inhibitors. Historically, management strategies in patients with inhibitors include high-dose clotting factor concentrates, immune tolerance induction therapy, and use of bypassing agents (e.g., FEIBA, NovoSevenRT).

COVERAGE GUIDELINES

The plan may authorize coverage of Hemlibra (emicizumab-kxwh) for Members when all of the following criteria are met:

1. Documented diagnosis of hemophilia A

AND

2. Documentation for use as prophylaxis to prevent or reduce the frequency of bleeding episodes

AND

3. Documentation of severe disease as evidence by <1% (<0.01 IU/mL) of endogenous factor VIII

LIMITATIONS

None

CODES

None

REFERENCES

1. Centers for Disease Control and Prevention. Hemophilia Inhibitors. 2017 Feb. URL: cdc.gov/ncbddd/hemophilia/inhibitors.html. Available from the Internet. Accessed 2019 January 23.
2. Hemlibra (emicizumab-kxwh) [package insert]. San Francisco, CA; Genentech, Inc.: 2018 October.
3. National Hemophilia Foundation. MASAC Recommendation on Administration of Inhibitor Bypassing Agents in the Home for Patients with Hemophilia and Inhibitors. 2015 June. URL: hemophilia.org/Researchers-Healthcare-Providers/Medical-and-Scientific-Advisory-Council-MASAC/MASAC-Recommendations/MASAC-Recommendation-on-Administration-of-Inhibitor-Bypassing-Agents-in-the-Home-for-Patients-with-Hemophilia-and-Inhibitors. Available on the Internet. Accessed 2019 January 23.
4. World Federation of Hemophilia. About Bleeding Disorders. 2012 May. URL: wfh.org/en/page.aspx?pid=1282. Available on the Internet. Accessed 2019 January 23.
5. World Federation of Hemophilia. Protocols for the treatment of hemophilia and von Willebrand disease. February 2018. URL: hog.org/publications/page/protocols-for-the-treatment-of-hemophilia-and-von-willebrand-disease-2. Available from Internet. Accessed 2019 January 23.

APPROVAL HISTORY

June 12, 2018: Reviewed by Pharmacy & Therapeutics Committee.

Subsequent endorsement date(s) and changes made:

1. February 12, 2019: Removed the following criterion due to updated approved indication in patients with or without inhibitors: "Documentation of an inhibitor titer of ≥5 BU/mL (high titer level)."
2. January 14, 2020: No changes.

BACKGROUND, PRODUCT AND DISCLAIMER INFORMATION

Pharmacy Medical Necessity Guidelines have been developed for determining coverage for plan benefits and are published to provide a better understanding of the basis upon which coverage decisions are made. The plan makes coverage decisions on a case-by-case basis considering the individual member's health care needs. Pharmacy Medical Necessity Guidelines are developed for selected therapeutic classes or drugs found to be safe, but proven to be effective in a limited, defined population of patients or clinical circumstances. They include concise clinical coverage criteria based on current literature review, consultation with practicing physicians in the service area who are medical experts in the particular field, FDA and other government agency policies, and standards adopted by national accreditation organizations. The plan revises and updates Pharmacy Medical Necessity Guidelines annually, or more frequently if new evidence becomes available that suggests needed revisions.

For self-insured plans, coverage may vary depending on the terms of the benefit document. If a discrepancy exists between a Pharmacy Medical Necessity Guideline and a self-insured Member's benefit document, the provisions of the benefit document will govern.

Treating providers are solely responsible for the medical advice and treatment of members. The use of this policy is not a guarantee of payment or a final prediction of how specific claim(s) will be adjudicated. Claims payment is subject to member eligibility and benefits on the date of service, coordination of benefits, referral/authorization and utilization management guidelines when applicable, and adherence to plan policies and procedures and claims editing logic.

Provider Services