MEDICAL MUTUAL®

Drug Policy

Policy:	**201805 MRx**	**Initial Effective Date: 02/18/2018**
Code(s):	**HCPCS J7170**	**Annual Review Date: 03/21/2024**
SUBJECT:	**Hemlibra (emicizumab-kxwh injection)**	**Last Revised Date: 03/21/2024**

☒Subject to Site of Care

Prior approval is required for some or all procedure codes listed in this Corporate Medical Policy.

Initial and renewal requests for the medication(s) listed in this policy are subject to site of care management. When billed under the medical benefit, administration of the medication will be restricted to a non-hospital facility-based location (i.e., home infusion provider, provider's office, free-standing ambulatory infusion center) unless the member meets the site of care exception criteria. To view the exception criteria and a list of medications subject to site of care management please click here.

OVERVIEW

Hemlibra is a bispecific factor IXa- and factor X-directed antibody indicated for routine prophylaxis to prevent or reduce the frequency of bleeding episodes in adult and pediatric patients ages newborn and older with hemophilia A (congenital factor VIII deficiency).

POLICY STATEMENT

This policy involves the use of Hemlibra. Prior authorization is recommended for medical benefit coverage of Hemlibra. Approval is recommended for those who meet the conditions of coverage in the **Initial Approval and Renewal Criteria, Preferred Drug (when applicable), Dosing/Administration, Length of Authorization, and Site of Care (when applicable)** for the diagnosis provided. The requirement that the patient meet the Criteria and Preferred Drug for coverage of the requested medication applies to the initial authorization only. All approvals for initial therapy are provided for the initial approval duration noted below; if reauthorization is allowed, a response to therapy is required for continuation of therapy.

Documentation Required: When documentation is required, the prescriber must provide written documentation supporting the trials of these other agents, noted in the criteria as [documentation required]. Documentation should include chart notes, prescription claims records, and/or prescription receipts.

I. Length of Authorization

Coverage will be provided for 3 months and may be renewed every 12 months thereafter.

II. Dosing Limits

A. Quantity Limit (max daily dose) [NDC Unit]:

Loading Dose: • 345 mg weekly x 4 doses
Maintenance Dose: • 6 mg/kg every 4 week dosing = 690 mg every 4 weeks

B. Max Units (per dose and over time) [HCPCS Unit]:

Loading Dose: • 690 billable units (BU) weekly x 4 doses
Maintenance Dose: • 6 mg/kg every 4 week dosing = 1380 BU every 4 weeks

Note: Patient must be dosed at a frequency that will produce the least wastage per dose based on available vial sizes of 12 mg, 30 mg, 60 mg, 105 mg, 150 mg, and 300 mg.

III. Initial Approval Criteria [1-3,8,10-14]

Coverage is provided in the following conditions:

- Diagnosis of congenital factor VIII deficiency has been confirmed by blood coagulation testing; **AND**

Hemophilia A (congenital factor VIII deficiency) with inhibitors † Φ

- Patient has inhibitors to Factor VIII with a current or historical titer of ≥ 5 Bethesda Units (BU)******; **AND**
- Must be used as routine prophylaxis to prevent or reduce the frequency of bleeding episodes; **AND**
  - Used as primary prophylaxis in patients with severe factor VIII deficiency (factor VIII level of <1%); **OR**
  - Used as secondary prophylaxis in patients with at least TWO documented episodes of spontaneous bleeding into joints; **AND**
- Not used in combination with Immune Tolerance Induction (ITI); **AND**
  - Patient has had at least two documented episodes of spontaneous bleeding into joints; **OR**
  - Patient had a documented trial and failure of Immune Tolerance Induction (ITI); **OR**
  - Patient had a documented trial and failure of, or is currently on, routine prophylaxis with a bypassing agent (i.e., NovoSeven, Feiba)

***Note:** Patients with inhibitor titer levels >0.6 BU to <5 BU who are not responding to or are not a candidate for standard factor replacement, will be evaluated on a case-by-case basis.*

Hemophilia A (congenital factor VIII deficiency) without inhibitors † Φ

- Must be used for routine prophylaxis to prevent or reduce the frequency of bleeding episodes; **AND**
- Used as treatment in one of the following:
  - Primary prophylaxis in patients with severe factor VIII deficiency (factor VIII level of <1%); **OR**
  - Secondary prophylaxis in patients with at least TWO documented episodes of spontaneous bleeding into joints; **AND**
- Patient is not a suitable candidate for treatment with shorter half-life Factor VIII (recombinant) products at a total weekly dose of 100 IU/kg or less (as attested by the prescribing physician with appropriate clinical rationale)

† FDA Approved Indication(s); ‡ Compendia Recommended Indication(s); **Φ** Orphan Drug

IV. Renewal Criteria [1-3,8]

Coverage can be renewed based upon the following criteria:

- Patient continues to meet the indication-specific relevant criteria such as concomitant therapy requirements (not including prerequisite therapy), performance status, etc. identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: thrombotic microangiopathy, thromboembolic events (thromboembolism, pulmonary embolism), development of neutralizing antibodies (inhibitors), etc.; **AND**
- Patient has demonstrated a beneficial response to therapy (i.e., the frequency of bleeding episodes has decreased from pre-treatment baseline)

V. Dosage/Administration [1]

Indication	Dose
Routine Prophylaxis in Congenital Hemophilia A with or without inhibitors	Loading Dose: Administer 3 mg/kg by subcutaneous injection once weekly for the first 4 weeks Maintenance Dose: • Administer 1.5 mg/kg once weekly; **OR** • Administer 3 mg/kg every two weeks; **OR** • Administer 6 mg/kg every four weeks

VI. Billing Code/Availability Information

HCPCS Code:

- J7170 - Injection, emicizumab-kxwh, 0.5 mg; 1 billable unit = 0.5 mg

NDC:

Drug	Strength	Form	NDC
Hemlibra	12 mg/0.4 mL	SDV	50242-0927-xx
	30 mg/mL	SDV	50242-0920-xx
	60 mg/0.4 mL	SDV	50242-0921-xx
	105 mg/0.7 mL	SDV	50242-0922-xx
	150 mg/mL	SDV	50242-0923-xx
	300 mg/2 mL	SDV	50242-0930-xx

VII. References

1. Hemlibra [package insert]. South San Francisco, CA; Genentech, Inc. January 2024. Accessed May 2024.
2. MASAC Recommendations Concerning Products Licensed for the Treatment of Hemophilia and Selected Disorders of the Coagulation System. Revised April 11, 2024. National Hemophilia Foundation. MASAC Document #284; April 2024. Available at: https://www.bleeding.org. Accessed May 2024.
3. Guidelines for the Management of Hemophilia. 3rd Edition. World Federation of Hemophilia 2020. Available at: https://www1.wfh.org/publications/files/pdf-1863.pdf. Accessed May 2024.
4. Annual Review of Factor Replacement Products. Oklahoma Health Care Authority Review Board. Updated Dec 2020. Accessed May 2024.
5. Graham A1, Jaworski K. Pharmacokinetic analysis of anti-hemophilic factor in the obese patient. Haemophilia. 2014 Mar;20(2):226-9.
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7. Mingot-Castellano, et al. Application of Pharmacokinetics Programs in Optimization of Haemostatic Treatment in Severe Hemophilia a Patients: Changes in Consumption, Clinical Outcomes and Quality of Life. Blood. 2014 December; 124 (21).
8. MASAC Recommendation Concerning Prophylaxis for Hemophilia A and B with and without Inhibitors. Revised April 27, 2022. National Hemophilia Foundation. MASAC Document #267; April 2022. Available at: https://www.bleeding.org. Accessed May 2024.
9. UKHCDO protocol for first line immune tolerance induction for children with severe haemophilia A: A protocol from the UKHCDO Inhibitor and Paediatric Working Parties. 2017. Available at: http://www.ukhcdo.org/guidelines. Accessed May 2024.

10. Oldenburg J, Mahlangu JN, Kim B, et al. Emicizumab Prophylaxis in Hemophilia A with Inhibitors. N Engl J Med. 2017 Aug 31;377(9):809-818. doi: 10.1056/NEJMoa1703068. Epub 2017 Jul 10.
11. Pipe SW, Shima M, Lehle M, et al. Efficacy, safety, and pharmacokinetics of emicizumab prophylaxis given every 4 weeks in people with haemophilia A (HAVEN 4): a multicentre, open-label, non-randomised phase 3 study. Lancet Haematol. 2019 Jun;6(6):e295-e305. doi: 10.1016/S2352-3026(19)30054-7. Epub 2019 Apr 16.
12. Young G, Liesner R, Chang T, et al. A multicenter, open-label phase 3 study of emicizumab prophylaxis in children with hemophilia A with inhibitors. Blood. 2019 Dec 12;134(24):2127-2138. doi: 10.1182/blood.2019001869. PMID: 31697801; PMCID: PMC6908828.
13. Mahlangu J, Oldenburg J, Paz-Priel I, et al. Emicizumab Prophylaxis in Patients Who Have Hemophilia A without Inhibitors. N Engl J Med. 2018 Aug 30;379(9):811-822. doi: 10.1056/NEJMoa1803550. PMID: 30157389.
14. Hoots, WK. (2024). Hemophilia A and B: Routine management including prophylaxis. In Leung LLK, Tirnauer JS (Eds.), *UptoDate*. Last updated: April 16, 2024. Accessed May 13, 2024. Available from https://www.uptodate.com/contents/hemophilia-a-and-b-routine-management-including-prophylaxis?search=hemophilia%20a&source=search_result&selectedTitle=2~150&usage_type=default&display_rank=2#H978189854
15. First Coast Service Options, Inc. Local Coverage Article: Billing and Coding: Hemophilia Clotting Factors (A56482). Centers for Medicare & Medicaid Services Inc. Updated on 09/29/2023 with effective date 10/01/2023. Accessed May 2024.
16. Palmetto GBA. Local Coverage Article: Billing and Coding: Guidance for Anti-Inhibitor Coagulant Complex (AICC) National Coverage Determination (NCD) 110.3 (A56065). Centers for Medicare & Medicaid Services Inc. Updated on 11/14/2022 with effective date 11/24/2022. Accessed May 2024.
17. Novitas Solutions, Inc. Local Coverage Article: Billing and Coding: Hemophilia Factor Products (A56433). Centers for Medicare & Medicaid Services Inc. Updated on 09/29/2023 with effective date 10/01/2023. Accessed May 2024.

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
D66	Hereditary factor VIII deficiency

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

The preceding information is intended for non-Medicare coverage determinations. Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determinations (NCDs) and/or Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. Local Coverage Articles (LCAs) may also exist for claims payment purposes or to clarify benefit eligibility under Part B for drugs which may be self-administered. The following link may be used to search for NCD, LCD, or LCA documents: https://www.cms.gov/medicare-coverage-database/search.aspx. Additional indications, including any preceding information, may be applied at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes		
Jurisdiction	**NCD/LCA/LCD Document (s)**	**Contractor**
H,L	A56433	Novitas Solutions, Inc.
J,M	A56065	Palmetto GBA
N	A56482	First Coast Service Options, Inc.

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT,	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC

Documentation Requirements:

The Company reserves the right to request additional documentation as part of its coverage determination process. The Company may deny reimbursement when it has determined that the services performed were not medically necessary, investigational or experimental, not within the scope of benefits afforded to the member and/or a pattern of billing or other practice has been found to be either inappropriate or excessive. Additional documentation supporting medical necessity for the services provided must be made available upon request to the Company. Documentation requested may include patient records, test results and/or credentials of the provider ordering or performing a service. The Company also reserves the right to modify, revise, change, apply and interpret this policy at its sole discretion, and the exercise of this discretion shall be final and binding.

FOR MEDICAL BENEFIT COVERAGE REQUESTS:

Prior approval is required for HCPCS Codes J7170