

Hemophilia Products – Anti-Inhibitor Antibody: Hemlibra (emicizumab-kxwh) (Subcutaneous)

Effective date: 01/01/2020

Review date: 12/18/19, 1/22/20, 2/25/2021, 06/24/2021, 6/16/2022, 6/22/2023, 12/07/2023, 01/04/2024, 05/15/2024

Scope: Medicaid Commercial, Medicare-Medicaid Plan (MMP)

I. Length of Authorization

Unless otherwise specified*, the initial authorization will be provided for 3 months and may be renewed every 12 months thereafter.

Note: The cumulative amount of medication the patient has on-hand will be taken into account for authorizations.

** Initial and renewal authorization periods may vary by specific covered indication*

II. Dosing Limits

A. Quantity Limit (max daily dose) [Pharmacy Benefit]:

<p><u>Loading Dose:</u></p> <ul style="list-style-type: none"> • 345mg weekly x 4 doses
<p><u>Maintenance Dose:</u></p> <ul style="list-style-type: none"> • 1.5mg/kg weekly = 180mg weekly • 3mg/kg every 2 weeks = 345mg every 2 weeks • 6mg/kg every 4 weeks = 690mg every 4 weeks

B. Max Units (per dose and over time) [Medical Benefit]:

<p><u>Loading Dose:</u></p> <ul style="list-style-type: none"> • 690 billable units (BU) weekly x 4 doses
<p><u>Maintenance Dose:</u></p> <ul style="list-style-type: none"> • 1.5mg/kg weekly = 360 BU weekly • 3mg/kg every 2 weeks = 690 BU every 2 weeks • 6mg/kg every 4 weeks = 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 30 mg, 60 mg, 105 mg, 150 mg, and 300mg.

III. Summary of Evidence

Clinical trials evaluating the efficacy and safety of Hemlibra in patients with hemophilia A with factor VIII inhibitors have demonstrated significant reductions in the frequency of bleeding episodes compared to standard prophylactic treatment regimens. Hemlibra has been shown to effectively mimic the function of factor VIII by bridging activated factor IX and factor X, thereby restoring the coagulation cascade, and reducing the risk of bleeding events. The most common adverse reactions include injection site reaction, headache, and arthralgia.

IV. Initial Approval Criteria

- MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements.

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

- Diagnosis of congenital factor VIII deficiency has been confirmed by blood coagulation testing; **AND**
- Confirmation the patient has inhibitors to Factor VIII ; **AND**
- Used as routine prophylaxis to prevent or reduce the frequency of bleeding episodes; **AND**
- Not used in combination with Immune Tolerance Induction (ITI); **AND**
 - Patient has at least two documented episodes of spontaneous bleeding into joints; **OR**
 - Patient has documented trial and failure of Immune Tolerance Induction (ITI); **OR**
 - Patient has documented trial and failure of or is currently on routine prophylaxis with a bypassing agent (i.e., NovoSeven, FEIBA).

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

- Diagnosis of congenital factor VIII deficiency has been confirmed by blood coagulation testing; **AND**
- Used for routine prophylaxis: **AND**
 - Patient must have severe hemophilia A (factor VIII level of <1%); **OR**
 - Patient has 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

V. Dispensing Requirements for Rendering Providers (Hemophilia Management Program)

- Prescriptions cannot be filled without an expressed need from the patient, caregiver or prescribing practitioner. Auto-filling is not allowed.
- Monthly, rendering provider must submit for authorization of dispensing quantity before delivering factor product.
- The cumulative amount of medication(s) the patient has on-hand should be taken into account when dispensing factor product.
- Dispensing requirements for renderings providers are a part of the hemophilia management program. This information is not meant to replace clinical decision making when initiating or modifying medication therapy and should only be used as a guide.

VI. Renewal Criteria

Coverage can be renewed based upon the following criteria:

- Patient continues to meet criteria identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: thrombotic microangiopathy and thrombotic events, thromboembolic events (thromboembolism, pulmonary embolism), development of neutralizing antibodies (inhibitors), etc.; **AND**
- Any increases in dose must be supported by an acceptable clinical rationale (i.e. weight gain, half-life study results, increase in breakthrough bleeding when patient is fully adherent to therapy, etc.); **AND**
- Patient has demonstrated a beneficial response to therapy (i.e., the frequency of bleeding episodes has decreased from pre-treatment baseline); **AND**
- The cumulative amount of medication(s) the patient has on-hand will be taken into account when authorizing.

Routine prophylaxis to prevent or reduce the frequency of bleeding episode

- Renewals will be approved for a 12 month authorization period

Dosage/Administration

Indication	Dose
Routine Prophylaxis in Congenital Hemophilia A with or without inhibitors	3 mg/kg by subcutaneous injection once weekly for the first 4 weeks, followed by 1.5 mg/kg once weekly, 3 mg/kg every two weeks, or 6 mg/kg every four weeks

VII. Billing Code/Availability Information

HCPCS Code:

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

NDC:

Drug	Strength	Form	NDC
	12mg/0.4 mL	SDV	50242-0927-xx
Hemlibra	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

VIII. References

1. Hemlibra [package insert]. South San Francisco, CA; Genentech, Inc. July 2023. Accessed November 2023.
2. MASAC RECOMMENDATIONS CONCERNING PRODUCTS LICENSED FOR THE TREATMENT OF HEMOPHILIA AND OTHER BLEEDING DISORDERS. 2016 National Hemophilia Foundation. MASAC Document #249; October 2016. Available at: <http://www.hemophilia.org>. Accessed January 2018.
3. Guidelines for the Management of Hemophilia. 2nd Edition. World Federation of Hemophilia. 2013. Available at: <https://www1.wfh.org/publication/files/pdf-1472.pdf>. Accessed January 2019.
4. Annual Review of Factor Replacement Products. Oklahoma Health Care Authority Review Board. Updated April 2016. Access June 2016.
5. Graham A1, Jaworski K. Pharmacokinetic analysis of anti-hemophilic factor in the obese patient. Haemophilia. 2014 Mar;20(2):226-9.
6. Croteau SE1, Neufeld EJ. Transition considerations for extended half-life factor products. Haemophilia. 2015 May;21(3):285-8.
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. 2016 National Hemophilia Foundation. MASAC Document #241; February 2016. Available at: <http://www.hemophilia.org>. Accessed January 2019.
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 January 2019.

Appendix 1 – Covered Diagnosis Codes

Hemlibra

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

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

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 Determination (NCD), Local Coverage Determinations (LCDs), and Local Coverage Articles (LCAs) may exist and compliance with these policies is required where applicable. They can be found at: <https://www.cms.gov/medicare-coverage-database/search.aspx>. Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD/LCA):

Jurisdiction(s): N	NCD/LCD Document (s): A56482
https://www.cms.gov/medicare-coverage-database/new-search/search-results.aspx?keyword=a56482&areaId=all&docType=NCA%2CCAL%2CNCD%2CMEDCAC%2CTA%2CMCD%2C6%2C3%2C5%2C1%2CF%2CP	
Jurisdiction(s): J,M	NCD/LCD Document (s): A56065
https://www.cms.gov/medicare-coverage-database/new-search/search-results.aspx?keyword=a56065&areaId=all&docType=NCA%2CCAL%2CNCD%2CMEDCAC%2CTA%2CMCD%2C6%2C3%2C5%2C1%2CF%2CP	
Jurisdiction(s): H,L	NCD/LCD Document (s): A56433
https://www.cms.gov/medicare-coverage-database/new-search/search-results.aspx?keyword=a56433&areaId=all&docType=NCA%2CCAL%2CNCD%2CMEDCAC%2CTA%2CMCD%2C6%2C3%2C5%2C1%2CF%2CP	

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD): N/A

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, AZ	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.

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	Applicable State/US Territory	Contractor
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, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC

Policy Rationale: Hemlibra was reviewed by the Neighborhood Health Plan of Rhode Island Pharmacy & Therapeutics (P&T) Committee. Neighborhood adopted the following clinical coverage criteria to ensure that its members use Hemlibra according to Food and Drug Administration (FDA) approved labeling and/or relevant clinical literature. Neighborhood worked with network prescribers and pharmacists to draft these criteria. These criteria will help ensure its members are using this drug for a medically accepted indication, while minimizing the risk for adverse effects and ensuring more cost-effective options are used first, if applicable and appropriate. For INTEGRITY (Medicare-Medicaid Plan) members, these coverage criteria will only apply in the absence of National Coverage Determination (NCD) or Local Coverage Determination (LCD) criteria. Neighborhood will give individual consideration to each request it reviews based on the information submitted by the prescriber and other information available to the plan.