

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

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### 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]:

<b><u>Loading Dose:</u></b>
– 345 mg weekly x 4 doses
<b><u>Maintenance Dose:</u></b>
– 1.5 mg/kg weekly dosing = 180 mg weekly
– 3 mg/kg every 2 week dosing = 345 mg every 2 weeks
– 6 mg/kg every 4 week dosing = 690 mg every 4 weeks

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

<b><u>Loading Dose:</u></b>
– 690 billable units (BU) weekly x 4 doses
<b><u>Maintenance Dose:</u></b>
– 1.5 mg/kg weekly dosing = 360 BU weekly
– 3 mg/kg every 2 week dosing = 690 BU every 2 weeks
– 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 30 mg, 60 mg, 105 mg, 150 mg, and 300 mg.*

### III. Initial Approval Criteria <sup>1-3,8,10,11</sup>

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 confirmed inhibitors to Factor VIII; **AND**
- Must be 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 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)

### **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:
  - Patient must have severe hemophilia A (factor VIII level of <1%); **OR**
  - Patient has had 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 <sup>1-3,8</sup>**

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 and thrombotic events, 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)

## Dosage/Administration <sup>1-3</sup>

Indication	Dose
Routine Prophylaxis in Congenital Hemophilia A with or without inhibitors	<p><u>Loading Dose:</u></p> <p>Administer 3 mg/kg by subcutaneous injection once weekly for the first 4 weeks</p> <p><u>Maintenance Dose:</u></p> <ul style="list-style-type: none"> <li>Administer 1.5 mg/kg once weekly; <b>OR</b></li> <li>Administer 3 mg/kg every two weeks; <b>OR</b></li> <li>Administer 6 mg/kg every four weeks</li> </ul>

## V. 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	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

## VI. References

- Hemlibra [package insert]. South San Francisco, CA; Genentech, Inc. March 2023. Accessed May 2023.
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- Guidelines for the Management of Hemophilia. 3<sup>rd</sup> Edition. World Federation of Hemophilia 2020. Available at: <https://www1.wfh.org/publications/files/pdf-1863.pdf>. Accessed May 2023.
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8. MASAC RECOMMENDATION CONCERNING PROPHYLAXIS. 2016 National Hemophilia Foundation. MASAC Document #241; February 2016. Available at: <https://www.hemophilia.org/>. Accessed May 2023.
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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. First Coast Service Options, Inc. Local Coverage Article: Billing and Coding: Hemophilia Clotting Factors (A56482). Centers for Medicare & Medicaid Services Inc. Updated on 10/28/2022 with effective date 10/01/2022. Accessed May 2023.
13. 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 2023.
14. Novitas Solutions, Inc. Local Coverage Article: Billing and Coding: Hemophilia Factor Products (A56433). Centers for Medicare & Medicaid Services Inc. Updated on 10/14/2022 with effective date 10/01/2022. Accessed May 2023.

## Appendix 1 – Covered Diagnosis Codes

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):

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