# 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]:**

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

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

| |
|---|
| **Loading Dose:**<br>– 690 billable units (BU) weekly x 4 doses |
| **Maintenance Dose:**<br>– 1.5 mg/kg weekly dosing = 360 BU weekly<br>– 3 mg/kg every 2 week dosing = 690 BU every 2 weeks<br>– 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 [1-3,8,10-13]

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 [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 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 [1]

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

## 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 | 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 |

## VI. References


1. Hemlibra [package insert]. South San Francisco, CA; Genentech, Inc. January 2024. Accessed February 2024.
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4. Annual Review of Factor Replacement Products. Oklahoma Health Care Authority Review Board. Updated April 2016. Accessed April 2022.
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: https://www.hemophilia.org/. Accessed May 2023.
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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. 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 February 2024.
15. 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 February 2024.
16. 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 February 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, LLC |
| 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, 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 |