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, 08/14/2024, 09/17/2025, 07/21/2026

Pharmacy Scope: Medicaid

Medical Scope: Medicaid, Commercial, Medicare

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]:

Loading Dose: • 345mg weekly x 4 doses
Maintenance Dose: • 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]:

Loading Dose: • 690 billable units (BU) weekly x 4 doses
Maintenance Dose: • 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 12mg, 30 mg, 60 mg, 105 mg, 150 mg, and 300mg.

III. Summary of Evidence

Hemlibra is a factor IXa and X indicated for the routine prophylaxis to prevent or reduce the frequency of bleeding episodes in patients with hemophilia A. Efficacy in patients with hemophilia A without FVIII inhibitors was established in two clinical trials, Haven 3 and Haven 4. Haven 3 was a randomized, multicenter, open-label, clinical trial in 152 patients with hemophilia A who received on-demand or prophylactic treatment. Patients received Hemlibra 3mg/kg once weekly for the first 4 weeks followed by either 1.5mg/kg once weekly or 3mg/kg once every two weeks or no prophylaxis. Hemlibra prophylaxis resulted in a statistically significant ($p<0.0001$) 68% reduction in bleed rate for treated bleeds compared to previous FVIII prophylaxis as well as significant reductions observed in all bleeds, spontaneous bleeds, joint bleeds and target joint bleeds. Haven 4 was a single-arm, multicenter, open-label clinical trial that included 41 patients with hemophilia A, with or without FVIII inhibitors. Patients received 3mg/kg weekly for the first 4 weeks followed by 6 mg/kg once every four weeks thereafter. Results show an annualized bleed rate of 1.2 for treated bleeds with four patients experiencing zero treated bleeds, demonstrating efficacy in reducing bleed rate. Adverse effects include injection site reactions, headache and arthralgia.

IV. Initial Approval Criteria

- Will NOT be used in combination with another agent used as prophylactic therapy for Hemophilia A *(Note: The prophylactic use of factor VIII (FVIII) products may be continued during the first week of Hemlibra prophylaxis)*** See chart below*

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

- Diagnosis of congenital factor VIII deficiency has been confirmed by blood coagulation testing; **AND**
- Member 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 members with severe factor VIII deficiency (factor VIII level of <1%); **OR**
    - Used as secondary prophylaxis in members with at least TWO documented episodes of spontaneous bleeding into joints; **AND**
- Not used in combination with Immune Tolerance Induction (ITI); **AND**
    - Member has at least two documented episodes of spontaneous bleeding into joints; **OR**
    - Member has documented trial and failure of Immune Tolerance Induction (ITI); **OR**
    - Member has documented trial and failure of or is currently on routine prophylaxis with a bypassing agent (i.e., NovoSeven, FEIBA).

***Note:** Members 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

- Diagnosis of congenital factor VIII deficiency has been confirmed by blood coagulation testing; **AND**
- 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 members with severe factor VIII deficiency (factor VIII level of <1%); **OR**
  - Secondary prophylaxis in members with at least TWO documented episodes of spontaneous bleeding into joints; **AND**
- Member 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

******Drugs to treat Hemophilia A or B***

Hemophilia A & B Drug Chart	
Factor VIIa (Hemophilia A or B)	
Novoseven RT	J7189
Sevenfact	J7212
Anti-Inhibitor Coagulant Complex (Hemophilia A or B)	
Feiba	J7198
Factor VIII (Hemophilia A)	
Advate	J7192
Kogenate FS	J7192
Helixate FS	J7192
Recombinate	J7192
Kovaltry	J7211
Eloctate	J7205
Koate / Koate-DVI	J7190

Hemofil M	J7190
Novoeight	J7182
Nuwiq	J7209
Obizur	J7188
Xyntha / Xyntha Solofuse	J7185
Afstyla	J7210
Adynovate	J7207
Jivi	J7208
Esperoct	J7204
Altuviiio	J7214
Humate-P	J7187
Alphanate	J7186
Wilate	J7183
Hemlibra	J7170
Factor IX (Hemophilia B)	
AlphaNine SD	J7193
Mononine	J7193
Alprolix	J7201
Profilnine	J7194
BeneFIX	J7194
Ixinity	J7213
Rixubis	J7200
Idelvion	J7202

Rebinyn	J7203
Tissue Factor Pathway Inhibitor (Hemophilia A or B)	
Alhemo	J7173
Hympavzi	J7172
Antithrombin-Directed Double-Stranded Small Interfering Ribonucleic acid (siRNA) (Hemophilia A or B)	
Qfitlia	J7174

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

- Prescriptions cannot be filled without an expressed need from the member, 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 member 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:

- Member continues to meet criteria identified in section IV; **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**
- 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 member is fully adherent to therapy, etc.); **AND**
- Member 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 member 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
Hemlibra	12mg/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

VIII. References

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Appendix 1 – Covered Diagnosis Codes

Hemlibra

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