

STANDARD MEDICARE PART B MANAGEMENT

HEMLIBRA (emicizumab-kxwh)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications

Hemlibra is 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) with or without factor VIII inhibitors.

All other indications will be assessed on an individual basis. Submissions for indications other than those enumerated in this policy should be accompanied by supporting evidence from Medicare approved compendia.

II. DOCUMENTATION

The following documentation must be available, upon request, for all submissions:

For continuation requests: Chart notes documenting benefit from therapy (e.g., reduced frequency or severity of bleeds).

III. CRITERIA FOR INITIAL APPROVAL

Hemophilia A (congenital factor VIII deficiency)

Authorization of 12 months may be granted for treatment of hemophilia A (congenital factor VIII deficiency) when all of the following criteria is met:

- A. Member must be using the requested medication for routine prophylaxis to prevent or reduce the frequency of bleeding episodes.
- B. Member meets one of the following criteria:
 1. Member has mild disease (See Appendix A) and has had an insufficient response to desmopressin or a documented clinical reason for not using desmopressin (See Appendix B).
 2. Member has moderate or severe disease (See Appendix A).
- C. Prophylactic use of factor VIII products (e.g., Advate, Adynovate, Eloctate) will be discontinued after the first week of starting therapy with the requested medication.

IV. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must be currently receiving therapy with the requested agent.

Authorization for 12 months may be granted when all of the following criteria are met:

- A. The member is currently receiving therapy with the requested medication
- B. The requested medication is being used to treat an indication enumerated in Section III
- C. The member is receiving benefit from therapy (e.g., reduced frequency or severity of bleeds)
- D. The member is not using the requested medication in combination with factor VIII products (e.g., Advate, Adynovate, Eloctate, etc.) for prophylactic use.

V. DOSAGE AND ADMINISTRATION

For initial and continuation requests, dosing does not exceed the following:

- A. Induction: 3mg/kg subcutaneously once weekly for the first 4 weeks.
- B. Maintenance: 1.5mg/kg once weekly, or 3mg/kg once every 2 weeks, or 6mg/kg once every 4 weeks.

VI. APPENDICES

Appendix A: Classification of Hemophilia by Clotting Factor Level (% Activity) and Bleeding Episodes

Severity	Clotting Factor Level % activity*	Bleeding Episodes
Severe	<1%	Spontaneous bleeding episodes, predominantly into joints and muscles Severe bleeding with trauma, injury or surgery
Moderate	1% to 5%	Occasional spontaneous bleeding episodes Severe bleeding with trauma, injury or surgery
Mild	6% to 40%	Severe bleeding with serious injury, trauma or surgery

*Factor assay levels are required to determine the diagnosis and are of value in monitoring treatment response.

Appendix B: Clinical Reasons For Not Utilizing Desmopressin in Patients with Hemophilia A

- a. Age < 2 years
- b. Pregnancy
- c. Fluid/electrolyte imbalance
- d. High risk for cardiovascular or cerebrovascular disease (especially the elderly)
- e. Predisposition to thrombus formation
- f. Trauma requiring surgery
- g. Life-threatening bleed
- h. Contraindication or intolerance to desmopressin
- i. Stimate Nasal Spray is unavailable due to backorder/shortage issues (where applicable)

VII. REFERENCES

1. Hemlibra [package insert]. South San Francisco, CA: Genentech, Inc.; June 2022.
2. Srivastava A, Santagostino E, Dougall A, et al. WFH Guidelines for the Management of Hemophilia, 3rd edition. *Haemophilia*. 2020;26 Suppl 6:1-158. doi:10.1111/hae.14046.
3. National Hemophilia Foundation. MASAC recommendations concerning products licensed for the treatment of hemophilia and other bleeding disorders. Revised March 2022. MASAC Document #272. https://www.hemophilia.org/sites/default/files/document/files/272_Treatment.pdf. Accessed December 3, 2022.

Reference number(s)
4798-A

4. National Hemophilia Foundation. Hemophilia A (Factor VIII Deficiency). Available at: <http://www.hemophilia.org/NHFWeb/MainPgs/MainNHF.aspx?menuid=180&contentid=45&rptname=bleeding>. Accessed December 3, 2022.
5. AHFS DI (Adult and Pediatric) [database online]. Hudson, OH: Lexi-Comp, Inc.; http://online.lexi.com/lco/action/index/dataset/complete_ashp [available with subscription]. Accessed December 3, 2022.
6. Leissinger C, Carcao M, Gill JC, et al. Desmopressin (DDAVP) in the management of patients with congenital bleeding disorders. *Haemophilia*. 2014;20:158-167.