

SPECIALTY GUIDELINE 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 are considered experimental/investigational and not medically necessary.

II. DOCUMENTATION

Submission of the following information is necessary to initiate the prior authorization review:
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 Hemlibra 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).

IV. CONTINUATION OF THERAPY

Authorization of 12 months may be granted for continued treatment in members requesting reauthorization for an indication listed in Section III when the member is experiencing benefit from therapy (e.g., reduced frequency or severity of bleeds).

V. DOSAGE AND ADMINISTRATION

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

Hemlibra 2417-A, 3536-A SGM P2021a

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- 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

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

VII. REFERENCES

- Hemlibra [package insert]. South San Francisco, CA: Genentech, Inc.; October 2018.
- 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.
- National Hemophilia Foundation. MASAC recommendations concerning products licensed for the treatment of hemophilia and other bleeding disorders. Revised August 2020. MASAC Document #263. https://www.hemophilia.org/sites/default/files/document/files/263_treatment.pdf. Accessed November 20, 2020.
- 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 November 25, 2020.
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- Leissinger C, Carcao M, Gill JC, et al. Desmopressin (DDAVP) in the management of patients with congenital bleeding disorders. *Haemophilia*. 2014;20:158-167.