# Clinical Policy: Emicizumab-kxwh (Hemlibra)

Reference Number: PA. CP.PHAR.370
Effective Date: 01.19
Last Review Date: 01.19

Coding Implications
Revision Log

**Description**
Emicizumab-kxwh (Hemlibra®) is a bispecific factor IXa- and factor X-directed antibody.

**FDA Approved Indication(s)**
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.

**Policy/Criteria**
*Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.*

It is the policy of health plans affiliated with PA Health & Wellness that Hemlibra is **medically necessary** when the following criteria are met:

**I. Initial Approval Criteria**

**A. Congenital Hemophilia A With or Without Inhibitors** (must meet all):

1. Prescribed for routine prophylaxis of bleeding episodes in patients with congenital hemophilia A (factor VIII deficiency);
2. Prescribed by or in consultation with a hematologist;
3. Dose does not exceed 3 mg/kg per week during the first four weeks of therapy, followed by either 1.5 mg/kg per week, 3 mg/kg once every two weeks, or 6 mg/kg once every four weeks thereafter.

**Approval duration: 6 months**

**B. Other diagnoses/indications**

1. Refer to the off-label use policy for the relevant line of business if diagnosis is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized): PA.CP.PMN.53.

**II. Continued Therapy**

**A. Congenital Hemophilia A With or Without Inhibitors** (must meet all)**:**

1. Currently receiving medication via Pennsylvania Health and Wellness benefit or member has previously met all initial approval criteria or the Continuity of Care policy (PA.LTSS.PHAR.01) applies;
2. Member is responding positively to therapy (*see Appendix D*);
3. If request is for a dose increase, new dose does not exceed 3 mg/kg per week during the first four weeks of therapy, followed by either 1.5 mg/kg per week, 3 mg/kg once every two weeks or 6 mg/kg once every four weeks thereafter.

**Approval duration: 6 months**

**B. Other diagnoses/indications** (must meet 1 or 2)**:**
1. Currently receiving medication via Pennsylvania Health and Wellness benefit and documentation supports positive response to therapy or the Continuity of Care policy (PA.LTSS.PHAR.01) applies.
   **Approval duration: Duration of request or 6 months (whichever is less)**; or
2. Refer to the off-label use policy for the relevant line of business if diagnosis is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized): PA.CP.PMN.53.

## III. Diagnoses/Indications for which coverage is NOT authorized:

**A.** Non-FDA approved indications, which are not addressed in this policy, unless there is sufficient documentation of efficacy and safety according to the off label use policy – PA.CP.PMN.53 or evidence of coverage documents.

## IV. Appendices/General Information

*Appendix A: Abbreviation/Acronym Key*
FDA: Food and Drug Administration

*Appendix B: Therapeutic Alternatives*
Not applicable

*Appendix C: Contraindications*
- Contraindication(s): none reported
- Black box warning(s): Thrombotic microangiopathy and thromboembolism: Cases of thrombotic microangiopathy and thrombotic events were reported when on average a cumulative amount of >100 U/kg/24 hours of activated prothrombin complex concentrate (aPCC) was administered for 24 hours or more to patients receiving Hemlibra prophylaxis. Monitoring is recommended for the development of thrombotic microangiopathy and thrombotic events if aPCC is administered. Discontinuation of aPCC and suspended dosing of Hemlibra is also recommended if symptoms occur.

*Appendix D: General Information*
- The elimination half-life of Hemlibra is 27.8 ± 8.1 days. Therefore, the “on-demand” use of Hemlibra for the treatment of acute bleeding episodes is inappropriate.
- There is insufficient data to support the use of Hemlibra for the treatment of hemophilia B either with or without inhibitors.
- There is potential for thrombotic microangiopathy and thrombotic events when used concurrently with Feiba > 100 U/kg/day for 24 hours or more. Additional monitoring is recommended with concomitant use of the two agents. Discontinuation of Feiba and suspended dosing of Hemlibra is recommended if symptoms occur.
- The World Federation of Hemophilia recommends starting primary prophylaxis before the second clinically evident large joint bleed, and before 3 years of age, to prevent future bleeding episodes and the resulting complications

- Examples of member responding positively to therapy may include: reduction in number of all bleeds over time, reduction in number of joint bleeds over time, or reduction in number of target joint bleeds over time.

## V. Dosage and Administration

| Indication | Dosing Regimen | Maximum Dose |
|---|---|---|
| Routine prophylaxis of bleeding episodes | 3 mg/kg SC weekly for four weeks, followed by 1.5 mg/kg SC weekly or 3 mg/kg once every two weeks or 6 mg/kg once every four weeks thereafter | 3 mg/kg/week for the first 4 weeks, followed by 1.5 mg/kg/week thereafter |

## VI. Product Availability

Single-dose vials for injection: 30 mg/mL, 60 mg/0.4 mL, 105 mg/0.7 mL, 150 mg/mL

## VII. References

1. Hemlibra Prescribing Information. South San Francisco, CA: Genentech, Inc.; October 2018. Available at: https://www.gene.com/download/pdf/hemlibra_prescribing.pdf. Accessed October 16, 2018.

## Coding Implications

Codes referenced in this clinical policy are for informational purposes only. Inclusion or exclusion of any codes does not guarantee coverage. Providers should reference the most up-to-date sources of professional coding guidance prior to the submission of claims for reimbursement of covered services.

| HCPCS Codes | Description |
|---|---|
| J7170 | Injection, emicizumab-kxwh, 0.5 mg |

| Reviews, Revisions, and Approvals | Date | P&T Approval Date |
|---|---|---|
| Policy created | 01.19 | |
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