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of the Blue Cross and Blue Shield Association

Medical benefit drug policies are a source for BCBSM and BCN medical policy information only. These documents are not to be used to determine benefits or reimbursement. Please reference the appropriate certificate or contract for benefit information. This policy may be updated and therefore subject to change.

Effective Date: 02/14/2019

Hemlibra® (emicizumab-kxwh)

FDA approval: 11/16/2017

HCPCS: J7170

Benefit: Both – Medical and Pharmacy

Policy:

Requests must be supported by submission of chart notes and patient specific documentation.

- A. Coverage of the requested drug is provided when all the following are met:
 - a. For prophylaxis of bleeding episodes in patients diagnosed with hemophilia A with inhibitors
 - i. Prescribed by or in consultation with a specialist that works in a hemophilia treatment center OR a hematologist with hemophilia experience
 - ii. Documentation of a high titer for factor VIII inhibitors measuring ≥ 5 Bethesda Units per milliliter (BU/mL)
 - iii. Will not be used in combination with Immune Tolerance Induction (ITI)
 - iv. Documentation of the number of bleeds experienced in the past 12 months
 - v. Prescribing appropriate FDA approved weight-based dosing
 - b. For prophylaxis of bleeding episodes in patients diagnosed with hemophilia A without inhibitors
 - i. Must be treatment experienced with factor VIII product
 - ii. Must not be well controlled on a stable dose of a factor VIII product
 - iii. Prescribed by or in consultation with a specialist that works in a hemophilia treatment center OR a hematologist with hemophilia experience
 - iv. Documentation of severe hemophilia A with factor VIII level $<1\%$
 - v. Documentation of the number of bleeds experienced within the past 12 months
 - vi. Trial and failure of the preferred products as specified in the BCBSM/BCN utilization management medical drug list
 - vii. Prescribing appropriate FDA approved weight-based dosing
- B. Quantity Limitations, Authorization Period and Renewal Criteria
 - a. Quantity Limit: FDA approved dosing
 - b. Initial Authorization Period: 6 months
 - c. Renewal Criteria:
 - i. Continuation of coverage will be provided when treatment has been proven successful

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- through a decrease in the number of bleeds
- ii. No development of anti-drug antibodies

***Note: Coverage may differ for Medicare Part B members based on any applicable criteria outlined in Local Coverage Determinations (LCD) or National Coverage Determinations (NCD) as determined by Center for Medicare and Medicaid Services (CMS). See the CMS website at <http://www.cms.hhs.gov/>. Determination of coverage of Part B drugs is based on medically accepted indications which have supported citations included or approved for inclusion determined by CMS approved compendia.

Therapeutic considerations:

A. FDA approved indication / Diagnosis

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.

**Please refer to most recent prescribing information.*
<https://www.hemlibra.com>

B. Background Information

- a. Hemophilia A accounts for 80% of patient with hemophilia with numbers totaling close to 16,000 patients in the US. It is estimated that 1 in 3 people with severe hemophilia A (those with factor VIII levels less than 1% in serum) will develop inhibitors to factor VIII, or antibodies to factor VIII replacement therapies. Presence of inhibitors to factor VIII can result in an increased risk for life threatening bleeds or repeated bleeds. The standard treatment for patients with hemophilia A are factor VIII concentrates. When there is a failure of a patient to respond to factor VIII concentrates, there is cause to suspect the presence of inhibitors. Patients with low amounts of inhibitors to factor VIII can be treated with higher doses of factor VIII to neutralize the inhibitors and stop the bleed. Patients with a history of high amounts of inhibitors to factor VIII but currently have low amounts of factor VIII inhibitors may be treated similarly to those with low levels of factor VIII inhibitors, however an anamnestic response can occur within three to five days precluding further treatment with factor VIII replacement therapies. Another product commercially available and used as prophylaxis of bleeding episodes in hemophilia A in those with inhibitors to factor VIII is FEIBA, a bypassing agent which bypasses factor VIII and allows continuation down the clotting cascade without factor VIII involvement.

C. Efficacy

**Please refer to most recent prescribing information.*

D. Medication Safety Considerations

Black Box Warning: Yes

**Please refer to most recent prescribing information.*

E. Dosing and administration

- a. Dosing:
 - i. Induction:
 - 1. 3 mg/kg once weekly for the first 4 weeks
 - ii. Maintenance:
 - 1. 1.5 mg/kg once every week

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2. 3 mg/kg once every 2 weeks
3. 6 mg/kg once every 4 weeks

**Please refer to most recent prescribing information.*

F. How supplied

- a. Single dose vials
 - i. 30 mg/mL
 - ii. 60 mg/0.4mL
 - iii. 105 mg/0.7 mL
 - iv. 150 mg/mL

References:

1. Hemlibra [prescribing information]. South San Francisco, CA: Genentech, Inc.; October 2018.
2. Srivastava A, Brewer AK, Mauser-Bunschoten EP, et al. World Federation of Hemophilia. Guidelines for the management of hemophilia. *Haemophilia*. Available at: <https://www1.wfh.org/publication/files/pdf-1472.pdf> Accessed on: November 19, 2018.
3. Oldenburg J, Mahlangu JN, Kim B, et al. Efficacy of emicizumab prophylaxis in hemophilia A with inhibitors. *NEMJ*. 2017; 377: 809 – 18.
4. Mahlangu JN, Oldenburg J, Paz-Priel I, et al. Efficacy of emicizumab prophylaxis in patients who have hemophilia A without inhibitors. *NEJM*. August 2018; 319 (9): 811 – 22.
5. Clinicaltrials.gov. A multicenter, open-label, phase III clinical trial to evaluate the efficacy, safety, and pharmacokinetics of subcutaneous administration of emicizumab in hemophilia A pediatric patients with inhibitors. (NCT02795767) Available at: <https://clinicaltrials.gov/ct2/show/NCT02795767>. Accessed on: November 19, 2018.
6. Clinicaltrials.gov. A randomized, multicenter, open-label, phase III clinical trial to evaluate the efficacy, safety, and pharmacokinetics of prophylactic emicizumab versus no prophylaxis in hemophilia A patients without inhibitors. (NCT02847637) Available at: <https://clinicaltrials.gov/ct2/show/NCT02847637>. Accessed on: November 19, 2018.
7. Hemophilia A. National Organization for Rare Diseases. Danbury, CT. <https://rarediseases.org/rare-diseases/hemophilia-a/>. Accessed on: December 4, 2018.
8. Kempton CL, White II GC. How we treat a hemophilia A patient with a factor VIII inhibitor. *Blood*. 2009; 133: 11-17. <http://www.bloodjournal.org/content/113/1/11>.
9. Franchini M, Lippi G. Acquired factor VIII inhibitors. *Blood*. 2008; 112: 250-255. <http://www.bloodjournal.org/content/112/2/250?sso-checked=true>.

Policy History		
#	Date	Change Description
1.2	Effective Date: 03/01/2019	PA added to BCBS and BCN
1.1	Effective Date: 02/14/2019	Criteria Update
1.0	Effective Date: 12/06/2018	New Policy

* The prescribing information for a drug is subject to change. To ensure you are reading the most current information it is advised that you reference the most updated prescribing information by visiting the drug or manufacturer website or <http://dailymed.nlm.nih.gov/dailymed/index.cfm>.

Authorization Request Form
Hemlibra® (emicizumab-kxwh injection)
HCPs CODE: J7170



This form is to be used by participating physicians to obtain coverage for Hemlibra®. For commercial members only, please complete this form and submit via fax to 1-877-325-5979. If you have any questions regarding this process, please contact BCBSM Provider Relations and Servicing or the Medical Drug Helpdesk at 1-800-437-3803 for assistance.

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PATIENT INFORMATION		PHYSICIAN INFORMATION	
Name		Name	
ID Number		Specialty	
D.O.B. ☐ Male ☐ Female		Address	
Diagnosis		City /State/Zip	
Drug Name ☐ Hemlibra®		Phone/Fax: P: () - F: () -	
Dose and Quantity Weight (kg)		NPI	
Directions		Contact Person	
Date of Service(s)		Contact Person Phone / Ext.	

STEP 1: DISEASE STATE INFORMATION

- Is this request for: ☐ Initiation ☐ Continuation Date patient started therapy: _____
- How is this medication being administered? ☐ Self-administered (Please fax this completed form to BCBSM at (866) 601-4425 if you have pharmacy coverage with BCBSM)
☐ Health Care Professional administered (**Continue to #3**)_
- Site of administration? ☐ Provider office/Home infusion ☐ Other: _____
☐ Hospital outpatient facility (go to #4) Reason for hospital outpatient administration: _____
- Please specify location of administration if hospital outpatient infusion? _____
- Please provide the NPI number for the place of administration: _____
- Initiation AND Continuation of therapy:**
 - Will Hemlibra be used in combination with Immune Tolerance Induction (ITI)? ☐ Yes ☐ No
 - Is the patient diagnosed with hemophilia A WITH factor VIII inhibitors? ☐ Yes ☐ No
 - Is the patient diagnosed with hemophilia A WITHOUT factor VIII inhibitors? ☐ Yes ☐ No
 - If yes, please provide a list of all hemophilia agents the patient tried and failed, length of therapy, and the reason for failure: _____
 - Please indicate how the patient's hemophilia is classified:
 - ☐ Mild hemophilia (factor VIII level of 6% - 40%)
 - ☐ Moderate hemophilia (factor VIII level of 1% - 5%)
 - ☐ Severe hemophilia (factor VIII level < 1%)
 - ☐ Unknown
 - Provide the number of bleeds the patient has experienced in the past 12 months prior to starting Hemlibra: _____
- Is Hemlibra being dispensed by a treatment center associated with hemophilia that provides high quality hemophilia care with outcome-based results (ie: hemophilia treatment center)? ☐ Yes ☐ No
- Continuation of therapy:**
 - Has the number of bleeding episodes decreased since starting Hemlibra? ☐ Yes ☐ No
 - Number of bleeding episodes yearly after starting Hemlibra: _____
 - Has the patient developed anti-drug antibodies since initiating treatment with Hemlibra? ☐ Yes ☐ No
- Please add any other supporting medical information necessary for our review

Coverage will not be provided if the prescribing physician's signature and date are not reflected on this document.

☐ Request for expedited review: I certify that applying the standard review time frame may seriously jeopardize the life or health of the member or the member's ability to regain maximum function

Physician's Name	Physician Signature	Date
Step 2: Checklist ☐ Form Completely Filled Out ☐ Attached Chart Notes	☐ List of medications patient tried and failed ☐ Blood coagulation test	
Step 3: Submit	By Fax: BCBSM Specialty Pharmacy Mailbox 1-877-325-5979	By Mail: BCBSM Specialty Pharmacy Program P.O. Box 312320, Detroit, MI 48231-2320

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