



Drug Coverage Policy

Effective Date.....5/1/2024
Coverage Policy Number.....IP0355
Policy Title.....Hemophilia – Eptacog
Products - Sevenfact

Hemophilia – Eptacog Products - Sevenfact

- **Sevenfact® (Factor VIIa [recombinant]-jncw intravenous infusion – LFB S.A./Hema Biologics)**

INSTRUCTIONS FOR USE

The following Coverage Policy applies to health benefit plans administered by Cigna Companies. Certain Cigna Companies and/or lines of business only provide utilization review services to clients and do not make coverage determinations. References to standard benefit plan language and coverage determinations do not apply to those clients. Coverage Policies are intended to provide guidance in interpreting certain standard benefit plans administered by Cigna Companies. Please note, the terms of a customer’s particular benefit plan document [Group Service Agreement, Evidence of Coverage, Certificate of Coverage, Summary Plan Description (SPD) or similar plan document] may differ significantly from the standard benefit plans upon which these Coverage Policies are based. For example, a customer’s benefit plan document may contain a specific exclusion related to a topic addressed in a Coverage Policy. In the event of a conflict, a customer’s benefit plan document always supersedes the information in the Coverage Policies. In the absence of a controlling federal or state coverage mandate, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of 1) the terms of the applicable benefit plan document in effect on the date of service; 2) any applicable laws/regulations; 3) any relevant collateral source materials including Coverage Policies and; 4) the specific facts of the particular situation. Each coverage request should be reviewed on its own merits. Medical directors are expected to exercise clinical judgment and have discretion in making individual coverage determinations. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. In certain markets, delegated vendor guidelines may be used to support medical necessity and other coverage determinations.

Medical Necessity Criteria

Sevenfact is considered medically necessary when ONE the following is met:

1. **Hemophilia A with Inhibitors.** Individual meets **ALL** of the following criteria:
 - A. Age 12 years or older
 - B. **ONE** of the following:
 - i. Positive inhibitor titer at least 5 Bethesda Units or greater

- ii. History of an inhibitor with anamnestic response to increased Factor VIII replacement therapy, which according to the prescriber, precludes the use of Factor VIII replacement to treat bleeding episodes
- iii. History of an inhibitor with refractory hemostatic response to increased Factor VIII replacement therapy, which according to the prescriber, precludes the use of Factor VIII replacement to treat bleeding episodes
- C. Medication is prescribed by, or in consultation with, a hematologist

Dosing. Up to 2,700 mcg/kg intravenously per 28 days

2. **Hemophilia B with Inhibitors.** Individual meets **ALL** of the following criteria:
- A. Age 12 years or older
 - B. **ONE** of the following:
 - i. Positive inhibitor titer at least 5 Bethesda Units or greater
 - ii. History of an inhibitor with anamnestic response to increased Factor IX replacement therapy, which according to the prescriber, precludes the use of Factor IX replacement to treat bleeding episodes
 - iii. History of an inhibitor with refractory hemostatic response to increased Factor IX replacement therapy, which according to the prescriber, precludes the use of Factor IX replacement to treat bleeding episodes
 - C. Medication is prescribed by, or in consultation with, a hematologist

Dosing. Up to 2,700 mcg/kg intravenously per 28 days

When coverage is available and medically necessary, the dosage, frequency, duration of therapy, and site of care should be reasonable, clinically appropriate, and supported by evidence-based literature and adjusted based upon severity, alternative available treatments, and previous response to therapy.

Receipt of sample product does not satisfy any criteria requirements for coverage.

Reauthorization Criteria

Continuation of Sevenfact is considered medically necessary for **ALL** covered diagnoses when the above medical necessity criteria are met AND there is documentation of beneficial response.

Authorization Duration

Initial approval duration: up to 12 months
 Reauthorization approval duration: up to 12 months

Conditions Not Covered

Any other use is considered experimental, investigational, or unproven.

Coding Information

- 1) This list of codes may not be all-inclusive.
- 2) Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement.

Considered Medically Necessary when criteria in the applicable policy statements listed above are met:

HCPCS Codes	Description
J7212	Factor VIIa (antihemophilic factor, recombinant)-jncw (Sevenfact), 1 mcg

Background

OVERVIEW

Sevenfact, a recombinant Factor VIIa product, is indicated for the treatment and control of bleeding episodes occurring in adults and adolescents (≥ 12 years of age) with **hemophilia A or B with inhibitors**.¹ As a limitation of use, Sevenfact is not indicated for the treatment of patients with congenital Factor VII deficiency.

Disease Overview

In hemophilia A and B, antibodies to exogenous clotting factor, known as “inhibitors”, may develop. Approximately 30% of patients with severe hemophilia A and up to 5% of patients with severe hemophilia B develop inhibitors to Factor VIII or Factor IX during their lifetime.² A high-responding inhibitor (≥ 5 Bethesda Units [BU]) tends to persist, whereas low-responding inhibitors of < 5 BU may wane without changes to the treatment regimen. Presence of inhibitors is associated with higher disease burden, increased risk of musculoskeletal complications, pain, physical limitations, and treatment challenges.^{2,3}

Dosing Information

Sevenfact is only indicated in the acute treatment setting for treatment of bleeding events. In the prescribing information, it is noted that maximum tolerated doses have not been determined for Sevenfact, and cumulative daily doses greater than 900 mcg/kg, which may be associated with greater risk of thromboembolic complications, have not been studied.¹ The National Hemophilia Foundation Medical and Scientific Advisory Council (MASAC) provides recommendations regarding doses of clotting factor concentrate in the home (2016).⁴ Per MASAC guidance, patients on prophylaxis should also have a minimum of one major dose and two minor doses on hand for breakthrough episodes. The guidance also notes that an adequate supply of clotting factor concentrate is needed to accommodate weekends and holidays. Therefore, maximum doses in this policy allow for three days of acute bleeding per 28 days. Doses exceeding this quantity will be reviewed on a case-by-case basis by a clinician.

Guidelines

National Bleeding Disorders Foundation MASAC guidelines (revised August 2023) recognize both Sevenfact and NovoSeven RT® (coagulation Factor VIIa [recombinant] intravenous infusion) as treatments for **hemophilia A or B with inhibitors**.⁵ No preference is stated for one agent over the other. It is noted that choice of product depends on multiple factors, including type of inhibitor (low- or high-responding), current titer, location of bleed, and previous response. Of note, NovoSeven RT, but not Sevenfact, is recognized as a treatment option in other settings, such as acquired hemophilia A and congenital Factor VII deficiency.

World Federation of Hemophilia (WFH) guidelines (2020) support recombinant Factor VIIa for patients with high-titer inhibitors who require acute treatment or around surgery/invasive procedures.³ For low-titer inhibitors, Factor VIII or IX replacement may be used. These products may also be used for patients with a history of a high-titer inhibitor whose titer has fallen to low or undetectable levels. However, once an anamnestic response occurs, further treatment with Factor

replacement is typically no longer effective, and bypass agent therapy (e.g., recombinant Factor VIIa) is needed.

References

1. Sevenfact® intravenous infusion [prescribing information]. Les Ulis, France/Louisville, KY: LFB S.A./Hema Biologics; November 2022.
2. Meeks SL, Leissing CA. The evolution of factor VIIa in the treatment of bleeding in haemophilia with inhibitors. *Haemophilia*. 2019;25(6):911-918.
3. Srivastava A, Santagostino E, Dougall A, et al; WFH Guidelines for the Management of Hemophilia panelists and co-authors. WFH Guidelines for the Management of Hemophilia, 3rd edition. *Haemophilia*. 2020 Aug;26 Suppl 6:1-158.
4. MASAC (Medical and Scientific Advisory Council) recommendations regarding doses of clotting factor concentrate in the home. MASAC Document #242. Adopted on June 7, 2016. Available at: <https://www.hemophilia.org/sites/default/files/document/files/242.pdf>. Accessed on November 5, 2023.
5. National Bleeding Disorders Foundation. MASAC (Medical and Scientific Advisory Council) recommendations concerning products licensed for the treatment of hemophilia and selected disorders of the coagulation system (August 2023). MASAC Document #280. Available at: <https://www.hemophilia.org/sites/default/files/document/files/MASAC-Products-Licensed.pdf>. Accessed on November 5, 2023.

Revision Details

Type of Revision	Summary of Changes	Date
Annual Revision	<ul style="list-style-type: none">• Updated coverage policy title.	5/1/2024

The policy effective date is in force until updated or retired.

“Cigna Companies” refers to operating subsidiaries of The Cigna Group. All products and services are provided exclusively by or through such operating subsidiaries, including Cigna Health and Life Insurance Company, Connecticut General Life Insurance Company, Evernorth Behavioral Health, Inc., Cigna Health Management, Inc., and HMO or service company subsidiaries of The Cigna Group. © 2024 The Cigna Group.