Effective Date **5/1/2023**
Next Review Date… **5/1/2024**
Coverage Policy Number **IP0552**

Factor XIII (Plasma-Derived)

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Related Coverage Resources

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. 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.

Overview

This policy supports medical necessity review for Factor XIII Concentrate (human) intravenous infusion (**Corifact**®).

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

Initial Approval Criteria

Factor XIII Concentrate (human) intravenous infusion (Corifact) is considered medically necessary for the treatment of congenital Factor XIII deficiency when the individual meets ALL of the following criteria:

1. **ONE** of the following conditions is met:
    a. Peri-operative management of bleeding
    b. Routine prophylaxis to reduce the frequency of bleeding episodes

c. Treatment of bleeding episodes

2. Medication is prescribed by or in consultation with a hematologist

Dosing. Up to 160 IU/kg by intravenous infusion 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.

Continuation of Therapy Criteria

Continuation of Factor XIII Concentrate (human) intravenous infusion (Corifact) is considered medically necessary for the treatment of congenital Factor XIII deficiency when initial criteria are met AND beneficial response is demonstrated.

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
J7180	Injection, factor XIII (antihemophilic factor, human), 1 IU

Background

OVERVIEW
Corifact, a Factor XIII concentrate, is indicated for adult and pediatric patients with congenital Factor XIII deficiency for:[1]

- **Peri-operative management** of surgical bleeding.
- **Routine prophylactic** treatment.

Disease Overview
Congenital Factor XIII deficiency is caused by defects in both Factor XIIIA and Factor XIIIB genes.[2,3] However, most cases are due to genetic alterations on the Factor XIIIA gene. The estimated prevalence of Factor XIIIA deficiency is one case in 2 million patients. Clinical symptoms include delayed wound healing, bleeding of soft and subcutaneous tissue, recurrent spontaneous miscarriage, and central nervous system (CNS) bleeding, which may be life-threatening. If patients have severe Factor XIII deficiency, early manifestations include bleeding from the umbilical cord or CNS. Prospective data showed that a level of 30% Factor XIII clotting activity is an adequate therapeutic target for most patients. Treatment of Factor XIII deficiency involves use of fresh

frozen plasma, cryoprecipitate, Corifact, or Tretten® (coagulation Factor XIIIA-Subunit [recombinant] intravenous infusion).

Guidelines
The National Hemophilia Foundation Medical and Scientific Advisory Council has guidelines for the treatment of hemophilia and other bleeding disorders (revised March 2022).[4] Corifact is recommended in patients who have Factor XIII deficiency.

Dosing Considerations
Dosing of clotting factor concentrates is highly individualized. MASAC provides recommendations regarding doses of clotting factor concentrate in the home (2016).[5] The number of required doses varies greatly and is dependent on the severity of the disorder and the prescribed regimen. Per MASAC guidance, patients on prophylaxis should also have a minimum of one major dose and two minor doses on hand for breakthrough bleeding in addition to the prophylactic doses used monthly. 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 prophylactic dosing plus three days of acute bleeding or perioperative management per 28 days. Doses exceeding this quantity will be reviewed on a case-by-case basis by a clinician.

References

1. Corifact® intravenous infusion [prescribing information]. Kankakee, IL: CSL Behring; December 2019.
2. Menegatti M, Peyvandi F. Treatment of rare factor deficiencies other than hemophilia. *Blood*. 2019;133(5):415-424.
3. Pelcovits A, Schiffman F, Niroula R. Factor XIII deficiency: a review of clinical presentation and management. *Hematol Oncol Clin North Am*. 2021;35(6):1171-1180.
4. MASAC (Medical and Scientific Advisory Council) recommendations concerning products licensed for the treatment of hemophilia and other bleeding disorders (Revised March 2022). MASAC Document #272. Adopted on April 27, 2022. Available at: https://www.hemophilia.org/sites/default/files/document/files/272_Treatment.pdf. Accessed on October 13, 2022.
5. National Hemophilia Foundation. MASAC (Medical and Scientific Advisory Council) recommendations regarding doses of clotting factor concentrate in the home (Revised June 7, 2016). MASAC Document #242. Adopted on June 7, 2016. Available at: https://www.hemophilia.org/sites/default/files/document/files/242.pdf. Accessed on October 13, 2022.