

POLICY: Hematology – Corifact® (Factor XIII Concentrate [human] injection for intravenous use – CSL Behring)

APPROVAL DATE: 09/11/2019

OVERVIEW

Corifact, a Factor XIII concentrate, is indicated for routine prophylactic treatment and perioperative management of surgical bleeding in adult and pediatric patients with congenital Factor XIII deficiency.¹

Disease Overview

Congenital Factor XIII deficiency is caused by defects in both Factor XIII A and Factor XIII B genes.²⁻⁵ However, most cases are due to genetic alterations on the Factor XIII A gene. The estimated prevalence of Factor XIII A 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 XIII A-Subunit [recombinant] injection for intravenous use).

Guidelines

The National Hemophilia Foundation Medical and Scientific Advisory Council has guidelines for the treatment of hemophilia and other bleeding disorders (revised April 2018).⁶ 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).⁷ 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.

POLICY STATEMENT

Prior authorization is recommended for medical benefit coverage Corifact. Approval is recommended for those who meet the Criteria and Dosing for the listed indication(s). Extended approvals are allowed if the patient continues to meet the criteria and dosing. Requests for doses outside of the established dosing documented in this policy will be considered on a case-by-case basis by a clinician (i.e., Medical Director or Pharmacist). Because of the specialized skills required for evaluation and diagnosis of patients treated with Corifact, as well as the monitoring required for adverse events and long-term efficacy, the agent is required to be prescribed by or in consultation with a physician who specializes in the condition being treated.

RECOMMENDED AUTHORIZATION CRITERIA

Coverage of Corifact is recommended for patients who meet criteria:

FDA-Approved Indication

1. **Congenital Factor XIII Deficiency.** Approve for 1 year if the agent is prescribed by or in consultation with a hematologist.

Dosing. Approve up to 160 IU/kg by intravenous infusion no more frequently than once every 28 days.

CONDITIONS NOT RECOMMENDED FOR APPROVAL

1. **Other Indications.** Coverage is not recommended for circumstances not listed in the Authorization Criteria. Criteria will be updated as new published data are available.

REFERENCES

1. Corifact® injection for intravenous use [prescribing information]. Kankakee, IL: CSL Behring; September 2017.
2. Menegatti M, Peyvandi F. Treatment of rare factor deficiencies other than hemophilia. *Blood*. 2019;133(5):415-424.
3. Ashley C, Change E, Davis J, et al. Efficacy and safety of prophylactic treatment with plasma-derived factor XIII concentrate (human) in patients with congenital factor XIII deficiency. *Haemophilia*. 2015;21(1):102-108.
4. Janbain M, Nugent DJ, Powell JS, et al. Use of Factor XIII (FXIII) concentrate in patients with congenital FXIII deficiency undergoing surgical procedures. *Transfusion*. 2015;55(1):45-50.
5. Palla R, Peyvandi F, Shapiro AD. Rare bleeding disorders: diagnosis and treatment. *Blood*. 2015;125(13):2052-2061.
6. MASAC (Medical and Scientific Advisory Council) recommendations concerning products licensed for the treatment of hemophilia and other bleeding disorders (Revised April 2018). MASAC Document #253. Adopted on April 19, 2018. Available at: <https://www.hemophilia.org/node/3675>. Accessed on June 7, 2019.
7. 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 July 22, 2019.

HISTORY

Type of Revision	Summary of Changes	Approval Date
New policy	--	09/11/2019