Kansas City

An Independent Licensee of the Blue Cross and Blue Shield Association

Jivi (antihemophilic factor [recombinant] PEGylated-aucl lyophilized powder)

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Policy

Blue Cross and Blue Shield of Kansas City (Blue KC) will provide coverage for **Jivi (antihemophilic factor [recombinant] PEGylated-aucl lyophilized powder)**when it is determined to be medically necessary because the criteria shown below are met.

When Policy Topic is covered

Jivi may be considered **medically necessary** when all of the following criteria are met:

FDA-Approved Indication

1. **Hemophilia A.** Approve for 1 year if the patient meets the following criteria (A and B):

A) The agent is prescribed by or in consultation with a hemophilia specialist; AND

B) The patient is ≥ 12 years of age.

Jivi is indicated in a variety of clinical scenarios in previously-treated adults and adolescents 12 years of age and older with hemophilia A.[1] Many other recombinant, third-generation, Factor VIII products (Advate, Novoeight, Nuwiq, Kovaltry, Afstyla), including extended half-life Factor VIII products (Eloctate, Adynovate), are indicated for use in young children in similar clinical scenarios in which Jivi is FDA-approved.[3] In studies involving previously-treated children with hemophilia A, adverse reactions due to immune response to PEG were noted in children < 6 years of age.[1]

Drug must be sourced from an approved specialty infusion provider.

When Policy Topic is not covered

Jivi is considered **not medically necessary** when the above criteria is not met and **investigational** for all other uses.

Considerations

Jivi requires prior authorization through the Clinical Pharmacy Department.

This Blue Cross and Blue Shield of Kansas City policy statement was developed using available resources such as, but not limited to: Food and Drug Administration (FDA) approvals, Facts and Comparisons, National specialty guidelines, local medical policies of other health plans, Medicare (CMS), local providers.

Description of Procedure or Service

Jivi, a DNA-derived antihemophilic factor (recombinant) PEGylated agent, is a Factor VIII product indicated for use in previously-treated adults and adolescents ≥ 12 years of age with hemophilia A in the following clinical scenarios: 1) on-demand treatment and control of bleeding episodes; 2) perioperative management of bleeding; and 3) routine prophylaxis to reduce the frequency of bleeding episodes.[1] Limitations of use include that Jivi is not indicated for use in children < 12 years of age due to a greater risk for hypersensitivity reactions. Also, Jivi is not indicated for use in previously-untreated patients, or for the treatment of von Willebrand disease. Dosing for the control of bleeding episodes and perioperative management is based on a unit per kg basis depending on the desired factor VIII increase. For routine prophylaxis, the initial regimen is 30 to 40 IU/kg intravenously (IV) twice weekly. Based on the bleeding episodes, the dose may be adjusted to 45 to 60 IU/kg IV once every 5 days. The regimen may be further individually adjusted to less or more frequent dosing.

Clinical Efficacy

The efficacy of Jivi for the clinical scenarios in which it is indicated was assessed in one international clinical trial involving male patients ≥ 12 years of age (n = 134).[1,2] Patients had severe hemophilia A (Factor VIII activity < 1%) and no history of Factor VIII inhibitors. Approximately 90% of bleeds were successfully treated with one or two infusions of Jivi. In patients using Jivi for prophylaxis, the annualized bleeding rate was reduced by approximately 88% when given once every 5 days in comparison to on-demand therapy ($P < 0.0001$). Use of Jivi for perioperative management led to either 'good' or 'excellent' hemostatic control in all of the 20 major surgeries performed.

Safety

The most common adverse events reported with Jivi in patients ≥ 12 years of age were headache (14%), cough (7%), pyrexia (5%), and nausea (5%).[1] In complete clinical studies involving pediatric previously-treated patients < 12 years of age (44 patients < 6 years of age and 29 patients 6 to < 12 years of age), adverse reactions due to immune response to PEG were noted in children < 6 years of age. In 23% of patients in this younger age group, loss of drug effect to

neutralizing anti-PEG IgM antibodies were noted during the first 4 exposure days. In 7% of patients < 6 years of age, loss of drug effect was noted, along with hypersensitivity reactions.

Rationale

Prior authorization is required to ensure the safe, clinically appropriate and cost effective use of Jivi while maintaining optimal therapeutic outcomes.

References

1. Jivi® lyophilized power for solution for intravenous use [prescribing information]. Whippany, NJ: Bayer HealthCare; August 2018.
2. Reding MT, Hg HJ, Poulsen H, et al. Safety and efficacy of BAY 94-9027, a prolonged-half-life factor VIII. *J Thromb Haemost*. 2017;15(3):411-419.
3. 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 23, 2018. Available at: https://www.hemophilia.org/node/3675. Accessed on August 30, 2018.

Billing Coding/Physician Documentation Information

J7199	Hemophilia clotting factor, not otherwise classified

Additional Policy Key Words

Factor VIII (recombinant), long-acting

Policy Implementation/Update Information

02/2019	New policy titled Jivi (antihemophilic factor [recombinant] PEGylated-aucl lyophilized powder)