CareSource Rxinnovations

PHARMACY POLICY STATEMENT
Arkansas PASSE

DRUG NAME	Beqvez (fidanacogene elaparvovec-dzkt)
BENEFIT TYPE	Medical
STATUS	Prior Authorization Required

Beqvez, approved by the FDA in 2024, is an adeno-associated virus vector-based gene therapy indicated for the treatment of adults with moderate to severe hemophilia B (congenital factor IX deficiency) who: Currently use factor IX prophylaxis therapy, or, have current or historical life-threatening hemorrhage, or, have repeated, serious spontaneous bleeding episodes, and, do not have neutralizing antibodies to adeno-associated virus serotype Rh74var (AAVRh74var) capsid as detected by an FDA-approved test.
Hemophilia is a disease that interferes with the normal coagulation process. Hemophilia B is less common than hemophilia A. It is caused by a faulty gene leading to a lack of clotting factor IX. Standard treatment is by replacement of the missing factor. Severe patients may require infusions 3 times per week. Successful gene therapy would obviate the need for prophylactic factor product use.
Beqvez is the second gene therapy approved for hemophilia B, following Hemgenix. It is designed to introduce in the transduced cells a functional copy of the factor IX gene encoding a high-activity FIX variant (FIX-R338L, hFIX Padua) to increase circulating factor IX activity.

Beqvez (fidanacogene elaparvovec-dzkt) will be considered for coverage when the following criteria are met:

Hemophilia B

For **initial** authorization:

1. Member is a male at least 18 years of age; AND
2. Medication must be prescribed by or in consultation with a hematologist; AND
3. Member has a documented diagnosis of hemophilia B; AND
4. Member has severe or moderately severe disease as indicated by 2% or less of normal circulating factor IX (2 IU/dL or less); AND
5. Member meets one of the following:
   a) Currently using Factor IX prophylaxis
   b) Has current or historical life-threatening hemorrhage
   c) Has repeated, serious spontaneous bleeding episodes; AND
6. Member's current weight is provided for dose calculation; AND
7. Baseline liver function tests (LFTs) have been or will be completed; AND
8. Member has tested negative for neutralizing antibodies to adeno-associated virus serotype Rh74var (AAVRh74var) capsid; AND
9. Factor IX (FIX) inhibitor testing is NOT positive (≥0.6 Bethesda Units [BU]) AND member does NOT have a prior history of factor IX inhibitor; AND
10. Member does NOT have any of the following:
    a) Significant liver disease such as portal hypertension, splenomegaly, hepatic encephalopathy
    b) Active hepatitis B or C
    c) CD4+ cell count <200 mm^3 or viral load ≥20 copies/mL if serological evidence of HIV-1 or HIV-2 infection
    d) Prior gene therapy.
11. **Dosage allowed/Quantity limit:** One-time IV infusion of 5×10^{11} vector genomes per kg (vg/kg).

If all the above requirements are met, the medication will be approved for 3 months.
For **reauthorization**: 1. Beqvez is a one-time single infusion and will not be reauthorized.

CareSource considers Beqvez (fidanacogene elaparvovec-dzkt) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
05/06/2024	New policy for Beqvez created.

References:

1. Beqvez [prescribing information]. Pfizer Inc.; 2024.
2. George LA, Sullivan SK, Giermasz A, et al. Hemophilia B Gene Therapy with a High-Specific-Activity Factor IX Variant. *N Engl J Med*. 2017;377(23):2215-2227. doi:10.1056/NEJMoa1708538
3. A Study to Evaluate the Efficacy and Safety of Factor IX Gene Therapy With PF-06838435 in Adult Males With Moderately Severe to Severe Hemophilia B (BENEGENE-2). ClinicalTrials.gov identifier: NCT03861273. Updated May 3, 2024. Accessed May 6, 2024. https://classic.clinicaltrials.gov/ct2/show/NCT03861273
4. Srivastava A, Santagostino E, Dougall A, et al. WFH Guidelines for the Management of Hemophilia, 3rd edition [published correction appears in Haemophilia. 2021 Jul;27(4):699]. *Haemophilia*. 2020;26 Suppl 6:1-158. doi:10.1111/hae.14046

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