Medical Policy

BlueCross BlueShield Kansas
An Independent licensee of the
Blue Cross Blue Shield Association

Title: Beqvez

Professional / Institutional
Original Effective Date: January 8, 2026
Latest Review Date:
Current Effective Date: January 8, 2026

State and Federal mandates and health plan member contract language, including specific provisions/exclusions, take precedence over Medical Policy and must be considered first in determining eligibility for coverage. To verify a member's benefits, contact Blue Cross and Blue Shield of Kansas Customer Service.

The BCBSKS Medical Policies contained herein are for informational purposes and apply only to members who have health insurance through BCBSKS or who are covered by a self-insured group plan administered by BCBSKS. Medical Policy for FEP members is subject to FEP medical policy which may differ from BCBSKS Medical Policy.

The medical policies do not constitute medical advice or medical care. Treating health care providers are independent contractors and are neither employees nor agents of Blue Cross and Blue Shield of Kansas and are solely responsible for diagnosis, treatment and medical advice.

If your patient is covered under a different Blue Cross and Blue Shield plan, please refer to the Medical Policies of that plan.

cPOLICY AGENT SUMMARY – MEDICAL PRIOR AUTHORIZATION

<table>
<tr><th>Indication</th><th>Dose</th></tr>
<tr><td>Hemophilia B (Congenital Factor IX Deficiency)</td><td>The recommended dose of Beqvez is a single-dose intravenous infusion of 5 × 10^{11} vector genomes per kg (vg/kg) of body weight.

<u>Calculate patient's dose weight</u>:

– Dosing is based on the patient's body mass index (BMI) in kg/m^2
<table>
<tr><th>• Patient's BMI</th><th>• Patient's Dose Weight</th></tr>
<tr><td>≤30 kg/m^2</td><td>Dose Weight=Actual body weight</td></tr>
<tr><td>>30 kg/m^2</td><td>Determine using the following calculation:</td></tr>
</table>
</td></tr>
</table>

	Dose Weight (kg) = 30 kg/m^2 x [Height (m)]^2

Calculation of patient's dose volume in mL:

- Dose weight in kilograms (kg) divided by 20 = dose in mL
  (The division factor 20 represents the amount of vector genomes per mL of the Beqvez suspension (1×10^{13} vg/mL) divided by the per kilogram dose (5×10^{11} vg/kg))

- *Beqvez contains genetically modified vectors. Personal protective equipment (including gloves, safety goggles, laboratory coat and sleeves) should be worn while preparing or administering.*
- *Confirm that the patient's identity matches the patient-specific identifier number on the outer carton.*
- *Store in the original package to avoid direct sunlight and ultraviolet light exposure.*
- *Thaw Beqvez vials for 1 hour at room temperature 15 °C to 30 °C (59 °F to 86 °F) in the upright orientation in the inner carton. Vials may be gently swirled but not shaken or inverted.*
- *DO NOT administer as an intravenous push or bolus.*
- *DO NOT infuse the diluted suspension in the same intravenous line with any other products.*
- *DO NOT use a central line or port.*

PRIOR AUTHORIZATION CLINICAL CRITERIA FOR APPROVAL

I. Length of Authorization

Coverage will be provided for one dose and may not be renewed.

II. Dosing Limits

Max Units (per dose and over time) [HCPCS Unit]:

- 1 billable unit for one dose

III. Initial Approval Criteria [1-12]

Submission of medical records (chart notes) related to the medical necessity criteria is REQUIRED on all requests for authorizations. Records will be reviewed at the time of submission. Please provide documentation related to diagnosis, step therapy, and clinical markers (i.e. genetic and mutational testing) supporting initiation when applicable. Please provide documentation via direct upload through the PA web portal or by fax.

Coverage is provided in the following conditions:

Hemophilia B (Congenital Factor IX Deficiency) †

- Patient is at least 18 years of age; AND
- Patient has a diagnosis of moderate to severe congenital factor IX deficiency (i.e., ≤2% of normal circulating factor IX), as confirmed by blood coagulation testing, for which the subject is on continuous routine factor IX prophylaxis, unless there is a

contraindication or intolerance *(Note: Continuous routine prophylaxis is defined as the intent of treating with an a priori defined frequency of infusions (e.g., twice weekly, once every two weeks, etc.) as documented in the medical records)*; **AND**

- Patient has not received prior hemophilia AAV-vector–based gene therapy (e.g., etranacogene dezaparvovec); **AND**
- Patient has one or more of the following:
    - Currently use Factor IX prophylaxis therapy (e.g., AlphaNine SD, Alprolix, BeneFIX, Idelvion, Ixinity, Mononine, Profilnine, Rebinyn, Rixubis, etc.); **OR**
    - Have current or historical life-threatening hemorrhage; **OR**
    - Have repeated, serious spontaneous bleeding episodes, (e.g., intramuscular hematomas requiring hospitalization, hemarthrosis, central nervous system (CNS) bleeding (including intracranial hemorrhage), pulmonary hemorrhage, life-threatening gastrointestinal (GI) hemorrhage and umbilical cord bleeding); **AND**
- Patient has been tested and found negative for Factor IX inhibitor titers (i.e., <0.6 Bethesda Units) and does not have a prior history of inhibitors *(Note: if test result is positive, re-test within approximately 2 weeks. If re-test is also positive, fidanacogene elaparvovec should not be given)*; **AND**
- Patient Factor IX activity will be monitored periodically (e.g., weekly for 3 months) as well as presence of inhibitors if bleeding is not controlled *(Note: patients will continue to require exogenous Factor IX until response to fidanacogene elaparvovec occurs)*; **AND**
- Patient will discontinue Factor IX prophylaxis therapy upon achieving FIX levels of 5% from fidanacogene elaparvovec treatment; **AND**
- Patient is adeno-associated virus serotype Rh74var capsid (AAVRh74var) neutralizing antibody negative as determined by an FDA-approved or CLIA-compliant test❖; **AND**
- Patient will have baseline liver function assessed prior to and after therapy according to the monitoring schedule outlined in the product labeling with corticosteroids administered in response to elevations; **AND**
- Patients with preexisting risk factors for hepatocellular carcinoma (e.g., patients with cirrhosis, advanced hepatic fibrosis, hepatitis C or B, non-alcoholic fatty liver disease (NAFLD), chronic alcohol consumption, non-alcoholic steatohepatitis (NASH), and advanced age) will have abdominal ultrasound screenings and be monitored regularly (e.g., annually) for alpha-fetoprotein (AFP) elevations following administration; **AND**
- Patient does not have current liver-related coagulopathy, hypoalbuminemia, persistent jaundice, or cirrhosis), portal hypertension, splenomegaly, hepatic encephalopathy, hepatic fibrosis, or active viral hepatitis; **AND**

- Patient has been tested for HIV and does not have an active infection (i.e., either CD4+ cell count <200 mm^3 or viral load ≥20 copies/mL in cases of serological evidence of HIV-1 or HIV-2 infection); AND
- Patient has been counseled on avoidance of potentially hepatoxic substances (e.g., alcohol) which may reduce the efficacy of fidanacogene elaparvovec

Notes:

- Monitor Factor IX activity levels as outlined in the prescribing information to confirm adequate endogenous Factor IX activity levels to support discontinuation of pre-infusion Factor IX prophylaxis therapy.
- Exogenous Factor IX or other hemostatic products may also be required in case of surgery, invasive procedures, trauma, or bleeds in the event that fidanacogene elaparvovec-derived Factor IX activity is deemed insufficient for adequate hemostasis in such situations.
- Use of exogenous Factor IX concentrates before and after fidanacogene elaparvovec administration may impede assessment of endogenous, fidanacogene elaparvovec-derived Factor IX activity.

❖ If confirmed using an immunotherapy assay-http://www.fda.gov/companiondiagnostics

† FDA Approved Indication(s); ‡ Compendia Recommended Indication(s); Φ Orphan Drug

IV. Renewal Criteria

- Duration of authorization has not been exceeded (refer to Section I)

Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage of these services as it applies to an individual member.

CLINICAL RATIONALE

See package insert for FDA preshttps://dailymed.nlm.nih.gov/dailymed/index.cfm

CODING

The following codes for treatment and procedures applicable to this policy are included below for informational purposes. This may not be a comprehensive list of procedure codes applicable to this policy.

Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage of these services as it applies to an individual member.

The code(s) listed below are medically necessary ONLY if the procedure is performed according to the "Policy" section of this document.

HCPCS code(s):

- J1414 – Injection, fidanacogene elaparvovec-dzkt, per therapeutic dose; 1 billable unit = 1 kit (based on weight chart below) *(Effective 01/01/2025)*
- J3590 – Unclassified biologics *(Discontinue use on 01/01/2025)*
- C9172 – Injection, fidanacogene elaparvovec-dzkt, per therapeutic dose; 1 billable unit = 1 kit (based on weight chart below) *(Discontinue use on 01/01/2025)*

NDC(s):

Beqvez Multi-Vial kit sizes:

Patient Dose Weight (kg)	Total number of vials per Kit	NDC
≤75	4	00069-2004-04
>75 to ≤95	5	00069-2005-05
>95 to ≤115	6	00069-2006-06
>115 to ≤135	7	00069-2007-07

REVISIONS	
Posted: Effective:	New medical policy (Hemophilia B Gene Therapy Medical Drug Criteria Program) added to the bcbsks.com web site. Hemgenix is the only drug in policy. Policy is maintained by Prime Therapeutics LLC.
10-02-2023	Policy reviewed by Prime Therapeutics with no revisions
Posted 07-01-2024 Effective 08-01-2024	Beqvez added as target to Hemophilia B Gene Therapy Medical Drug Criteria Program
10-08-2024	Policy reviewed by Prime Therapeutics with non-clinical edits
Posted: 12-09-2025 Effective: 01-08-2026	New medical added to the bcbsks.com web site . Policy maintained by Prime Therapeutics LLC.

REFERENCES

1. Beqvez [package insert]. New York, NY; Pfizer, Inc., April 2024. Accessed November 2024.
2. MASAC Recommendations Concerning Products Licensed for the Treatment of Hemophilia and Selected Disorders of the Coagulation System. National Hemophilia Foundation. MASAC Document #290; October 2024. Available at:https://www.bleeding.org/healthcare-professionals/guidelines-on-care/masac-documents/masac-document-290-masac-recommendations-concerning-products-licensed-for-the-treatment-of-hemophilia-and-selected-disorders-of-the-coagulation-system. Accessed November 2024.

3. Guidelines for the Management of Hemophilia. 3rd Edition. World Federation of Hemophilia 2020. Available at: https://www1.wfh.org/publications/files/pdf-1863.pdf. Accessed November 2024.
4. Graham A1, Jaworski K. Pharmacokinetic analysis of anti-hemophilic factor in the obese patient. Haemophilia. 2014 Mar;20(2):226-9.
5. Croteau SE1, Neufeld EJ. Transition considerations for extended half-life factor products. Haemophilia. 2015 May;21(3):285-8.
6. Mingot-Castellano, et al. Application of Pharmacokinetics Programs in Optimization of Haemostatic Treatment in Severe Hemophilia a Patients: Changes in Consumption, Clinical Outcomes and Quality of Life. Blood. 2014 December; 124 (21).
7. MASAC Recommendation Concerning Prophylaxis For Hemophilia A And B With And Without Inhibitors. National Hemophilia Foundation. MASAC Document #267 (Replaces Document #241); April 2022. Available at: https://www.bleeding.org/sites/default/files/document/files/267_Prophylaxis.pdf. Accessed November 2024.
8. Rayment R, Chalmers E, Forsyth K, et al. Guidelines on the use of prophylactic factor replacement for children and adults with Haemophilia A and B. B J Haem:190;5, Sep 2020. https://doi.org/10.1111/bjh.16704.
9. Peyvandi F, Palla R, Menegatti M, et al. Coagulation factor activity and clinical bleeding severity in rare bleeding disorders: results from the European Network of Rare Bleeding Disorders. *J Thromb Haemost*. 2012;10:615-621.
10. MASAC Recommendations on Hemophilia Treatment Center Preparedness for Delivering Gene Therapy for Hemophilia. National Hemophilia Foundation. MASAC Document #282. October 2023. Available at: https://www.bleeding.org/healthcare-professionals/guidelines-on-care/masac-documents/masac-document-282-masac-recommendations-on-hemophilia-treatment-center-preparedness-for-delivering-gene-therapy-for-hemophilia. Accessed November 2024.
11. Thornburg, C.D., Simmons, D.H., von Drygalski, A. Evaluating gene therapy as a potential paradigm shift in treating severe hemophilia. BioDrugs. 2023. DOI: 10.1007/s40259-023-00615-4.
12. Klamroth R, Cuker A, Alzahrani H, et al. Efficacy and Safety of Fidanacogene Elaparvovec in Adults with Moderately Severe or Severe Hemophilia B: Results from the Phase 3 BENEGENE-2 Gene Therapy Trial. Hamostaseologie 2024; 44(S 01): S81-S82. DOI: 10.1055/s-0044-1779185.