Hemgenix® (etranacogene dezaparvovec-drlb)
(Intravenous)

Document Number: IC-0688

Last Review Date: 01/06/2025
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Dates Reviewed: 01/2023, 06/2024, 01/2025

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 [1-13]

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) † Φ

- Patient is at least 18 years of age; **AND**
- Patient has a diagnosis of moderately severe or severe congenital factor IX deficiency bleeding phenotype (i.e., ≤2% of normal circulating factor IX), as attested by the managing physician 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., fidanacogene elaparvovec); **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 (if test result is positive, re-test within approximately 2 weeks. If re-test is also positive, Hemgenix 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 Hemgenix occurs);* **AND**
- Patient will discontinue Factor IX prophylaxis therapy upon achieving FIX levels of 5% from etranacogene dezaparvovec treatment; **AND**
- Patient's baseline anti-AAV5 antibody titer is used as part of the evaluation process by the managing physician; **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

Notes:

- It may take several weeks before improved hemostatic control becomes apparent after etranacogene dezaparvovec infusion; therefore, continued hemostatic support with exogenous human Factor IX may be needed during the first weeks after etranacogene dezaparvovec infusion.
- Use of exogenous Factor IX concentrates before and after etranacogene dezaparvovec administration may impede assessment of endogenous, etranacogene dezaparvovec-derived Factor IX activity.

† 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)

V. Dosage/Administration [1]

Indication	Dose
Hemophilia B (Congenital Factor IX Deficiency)	The recommended dose of Hemgenix is 2 x 10^{13} genome copies (gc) per kilogram (kg) of body weight (or 2 mL/kg body weight) administered as an intravenous infusion. Calculate the dose as follows: – Hemgenix dose (in mL) = patient body weight (in kilogram) x 2

	The multiplication factor 2 represents the per kilogram dose (2 × 10^{13} gc/kg) divided by the amount of genome copies per mL of the Hemgenix solution (1 × 10^{13} gc/mL). – Number of Hemgenix vials needed = Hemgenix dose (in mL) divided by 10 (round up to next whole number of vials). *The division factor 10 represents the extractable volume of Hemgenix from each vial (10 mL).*
• Prepare Hemgenix using sterile technique under aseptic conditions, proper engineering controls (e.g., biological safety cabinet or isolator) and according to institutional policies. • Do not expose Hemgenix to the light of an ultraviolet radiation disinfection lamp. • Confirm that the patient's identity matches with the patient-specific identifier number on the outer carton. • Verify the required dose of Hemgenix based on the patient's body weight. • Confirm that the carton contains sufficient number of vials to prepare the diluted Hemgenix patient-specific infusion bag. • Parenteral drug products should be inspected visually for particulate matter and discoloration prior to administration, whenever solution and container permit. • For single-dose intravenous infusion only. • DO NOT administer Hemgenix as an intravenous push or bolus. • DO NOT infuse the diluted Hemgenix solution in the same intravenous line with any other products. • DO NOT use a central line or port.	

VI. Billing Code/Availability Information

HCPCS code:

- J1411 – Injection, etranacogene dezaparvovec-drlb, per therapeutic dose; 1 billable unit = 1 kit (based on weight chart below)

NDC(s):

Hemgenix kit sizes:

Total number of vials	Patient Weight (kg)	Total Volume (mL)	NDC
10	46-50	100	00053-0100-10
11	51-55	110	00053-0110-11
12	56-60	120	00053-0120-12
13	61-65	130	00053-0130-13
14	66-70	140	00053-0140-14
15	71-75	150	00053-0150-15
16	76-80	160	00053-0160-16
17	81-85	170	00053-0170-17
18	86-90	180	00053-0180-18
19	91-95	190	00053-0190-19
20	96-100	200	00053-0200-20
21	101-105	210	00053-0210-21
22	106-110	220	00053-0220-22
23	111-115	230	00053-0230-23
24	116-120	240	00053-0240-24
25	121-125	250	00053-0250-25
26	126-130	260	00053-0260-26
27	131-135	270	00053-0270-27
28	136-140	280	00053-0280-28

29	141-145	290	00053-0290-29
30	146-150	300	00053-0300-30
31	151-155	310	00053-0310-31
32	156-160	320	00053-0320-32
33	161-165	330	00053-0330-33
34	166-170	340	00053-0340-34
35	171-175	350	00053-0350-35
36	176-180	360	00053-0360-36
37	181-185	370	00053-0370-37
38	186-190	380	00053-0380-38
39	191-195	390	00053-0390-39
40	196-200	400	00053-0400-40
41	201-205	410	00053-0410-41
42	206-210	420	00053-0420-42
43	211-215	430	00053-0430-43
44	216-220	440	00053-0440-44
45	221-225	450	00053-0450-45
46	226-230	460	00053-0460-46
47	231-235	470	00053-0470-47
48	236-240	480	00053-0480-48

VII. References

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Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
D67	Hereditary factor IX deficiency

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

The preceding information is intended for non-Medicare coverage determinations. Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determinations (NCDs) and/or Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. Local Coverage Articles (LCAs) may also exist for claims payment purposes or to clarify benefit eligibility under Part B for drugs which may be self-administered. The following link may be used to search for NCD, LCD, or LCA documents: https://www.cms.gov/medicare-coverage-database/search.aspx. Additional indications, including any preceding information, may be applied at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCA/LCD): N/A

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC