Genglycos™ (pariglasgene brecaparvovec-opnr)
(Intravenous)

Document Number: IC-0880

Last Review Date: 10/01/2026
Date of Origin: 10/01/2026
Dates Reviewed: 10/2026

I. Length of Authorization

- Initial: Prior authorization validity will be provided initially for one dose.
- Renewal: Prior authorization validity may NOT be renewed.

II. Dosing Limits

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

- 1 kit of 15 vials for a total of 30 mL (each vial contains 3.0 x 10^{13} genome copies-gc/mL – 2 mL)

III. Initial Approval Criteria

Submission of supporting clinical documentation (including but not limited to medical records, chart notes, lab results, and confirmatory diagnostics) related to the medical necessity criteria is REQUIRED on all requests for authorizations. Records will be reviewed at the time of submission as part of the evaluation of this request. 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. Failure to submit the medical records may result in the denial of the request due to inability to establish medical necessity in accordance with policy guidelines.

Prior authorization validity is provided in the following conditions:

Glycogen Storage Disease type Ia (GSDIa) † Φ [1-5]

- Member is at least 8 years of age; **AND**
- Member does NOT have any FDA labeled contraindications to the requested agent (e.g., severe hepatic fibrosis or cirrhosis); **AND**
- Member has a diagnosis of glycogen storage disease type Ia (also known as von Gierke disease) as established by the following:
  - Member has biallelic pathogenic variants in the *G6PC1* gene identified by molecular genetic testing; **OR**
  - Member has deficient hepatic enzyme activity (i.e., glucose-6-phosphatase catalytic activity) as evidenced from a liver biopsy specimen; **AND**

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- Member is currently receiving a therapeutic regimen of cornstarch (or equivalent), following international guidelines/recommendations with stable nutrition, glycemic, and clinical status and will use therapy to reduce daily cornstarch intake as an adjunct to nutritional management; **AND**
- Member does not have detectable anti-adeno-associated virus, serotype 8 (anti-AAV8) antibody titers at baseline; **AND**
- Member will have adrenal function assessed including performing a clinical examination and laboratory testing (i.e., morning serum cortisol and ACTH) prior to start of therapy and as clinically indicated; **AND**
- Member will have liver function assessed including performing a clinical examination and laboratory testing (i.e., alanine aminotransferase (ALT), aspartate aminotransferase (AST), and total bilirubin) prior to start of therapy and then every 2 to 4 weeks until month 6, and as clinically indicated**; AND**
- Member will use immunosuppressive corticosteroid therapy, following Genglycos, and will be monitored for signs and symptoms of hepatotoxicity and/or adrenal insufficiency and adrenal crisis; **AND**
- Member does not have pre-existing hepatic impairment or an acute hepatic viral infection; **AND**
- Member has not received prior adeno-associated viral (AAV) vector-based gene therapy; **AND**
- Member has not received a liver transplant

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

IV. Renewal Criteria [1]

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

V. Dosage/Administration [1]

<table>
<tr><th>Indication</th><th>Dose</th></tr>
<tr><td>GSDIa</td><td>– The recommended dose of Genglycos is 1.0 × 10^{13} gc/kg body weight, administered as a single intravenous infusion.
<table>
<tr><th>Member body weight range (kg)</th><th>Total Dose Volume (mL)</th><th>Number of vials</th></tr>
<tr><td>15 to <21</td><td>6</td><td>3</td></tr>
<tr><td>21 to <27</td><td>8</td><td>4</td></tr>
<tr><td>27 to <33</td><td>10</td><td>5</td></tr>
<tr><td>33 to <39</td><td>12</td><td>6</td></tr>
<tr><td>39 to <45</td><td>14</td><td>7</td></tr>
<tr><td>45 to <51</td><td>16</td><td>8</td></tr>
<tr><td>51 to <57</td><td>18</td><td>9</td></tr>
<tr><td>57 to <63</td><td>20</td><td>10</td></tr>
<tr><td>63 to <69</td><td>22</td><td>11</td></tr>
<tr><td>69 to <75</td><td>24</td><td>12</td></tr>
<tr><td>75 to <81</td><td>26</td><td>13</td></tr>
<tr><td>81 to <87</td><td>28</td><td>14</td></tr>
<tr><td>≥87</td><td>30</td><td>15</td></tr>
<tr><td colspan="3">a. Dose volume is rounded up or down to the nearest 2 mL.
b. Members weighing >87 kg should be dosed at the 87 kg weight.</td></tr>
</table></td></tr>
</table>

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- Verify the required dose of Genglycos based on member's body weight, and confirm the kit contains enough vials to prepare the Genglycos infusion for the member.
- Vials should not be thawed or punctured until immediately prior to use. If the member is not expected to be ready for same-day administration, store Genglycos in a manner consistent with how it is outlined in the prescribing information.
- Pre-medicate with acetaminophen and non-sedating antihistamines 30 to 60 minutes prior to infusion of Genglycos.
- Administer Genglycos as a single-dose intravenous infusion through a peripheral venous catheter.
- Do not administer Genglycos as an intravenous push or bolus.

VI. Billing Code/Availability Information

HCPCS code(s):

- J3590 – Unclassified biologics
- C9399 – Unclassified drugs or biologicals *(Hospital Outpatient Use Only)*

NDC:

- Genglycos 3.0 × 10^{13} genome copies (gc) per mL – 2 mL single dose vial

Member body weight range (kg)	Total Dose Volume (mL)	Number of vials	NDC
15 to <21	6	3	69794-0333-03
21 to <27	8	4	69794-0334-04
27 to <33	10	5	69794-0335-05
33 to <39	12	6	69794-0336-06
39 to <45	14	7	69794-0337-07
45 to <51	16	8	69794-0338-08
51 to <57	18	9	69794-0339-09
57 to <63	20	10	69794-0340-10
63 to <69	22	11	69794-0341-11
69 to <75	24	12	69794-0342-12
75 to <81	26	13	69794-0343-13
81 to <87	28	14	69794-0344-14
≥87	30	15	69794-0345-15

VII. References

1. Genglycos [package insert]. Novato, CA; Ultragenyx Pharm., Inc.; August 2026. Accessed September 2026.
2. ClinicalTrials.gov. NCT01395641. A Phase 3, Randomized, Double-Blind, Placebo-Controlled Study of Adeno-Associated Virus Serotype 8-Mediated Gene Transfer of Glucose-6-Phosphatase in Patients With Glycogen Storage Disease Type Ia. Search ClinicalTrials.gov for: NCT05139316 | Card Results | ClinicalTrials.gov.
3. Bali DS, El-Gharbawy A, Austin S, et al. Glycogen Storage Disease Type I. 2006 Apr 19 [Updated 2021 Oct 14]. In: Adam MP, Bick S, Mirzaa GM, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2026. Available from: https://www.ncbi.nlm.nih.gov/books/NBK1312/.
4. Glycogen storage disease type 1A. National Institutes of Health: Genetic and Rare Diseases Information Center Website. https://rarediseases.info.nih.gov/diseases/7864/glycogen-storage-disease-type-1a. Updated March 1, 2019. Accessed September 3, 2026.
5. Mitchell J, Abdenur J, de Boer F, et al. Efficacy and safety results from a pivotal phase 3 trial of DTX401, an AAV8-mediated liver-directed gene therapy, in individuals with glycogen storage

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disease type Ia (GSDIa). *Endocrine Abstracts* (2025) **110** RC3.6.
DOI: 10.1530/endoabs.110.RC3.6

Appendix A – Non-Quantitative Treatment Limitations (NQTL) Factor Checklist

Non-quantitative treatment limitations (NQTLs) refer to the methods, guidelines, standards of evidence, or other conditions that can restrict how long or to what extent benefits are provided under a health plan. These may include things like utilization review or prior authorization. The utilization management NQTL applies comparably, and not more stringently, to mental health/substance use disorder (MH/SUD) Medical Benefit Prescription Drugs and medical/surgical (M/S) Medical Benefit Prescription Drugs. The table below lists the factors that were considered in designing and applying prior authorization to this drug/drug group, and a summary of the conclusions that Prime's assessment led to for each.

Factor	Conclusion
Indication	Yes: Consider for PA
Safety and efficacy	No: PA not a priority
Potential for misuse/abuse	No: PA not a priority
Cost of drug	Yes: Consider for PA

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
E74.01	von Gierke disease

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	Wellpoint Federal

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Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
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	Wellpoint Federal
15	KY, OH	CGS Administrators, LLC

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