

Cerezyme® (imiglucerase) (Intravenous)

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I. Length of Authorization

Coverage will be for 12 months and may be renewed.

II. Dosing Limits

A. Quantity Limit (max daily dose) [NDC unit]:

- Cerezyme 200 unit injection: 70 vials per 28 days
- Cerezyme 400 unit injection: 36 vials per 28 days

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

- 700 billable units every 14 days

III. Initial Approval Criteria ^{1,9,10,11,12}

Universal Criteria

- Must be used as a single agent; **AND**

Type 1 Gaucher Disease †Φ

- Patient age at least 2 years or older; **AND**
- Patient has a documented diagnosis of Type 1 Gaucher Disease as confirmed by a beta-glucosidase leukocyte (BGL) test with significantly reduced or absent glucocerebrosidase enzyme activity; **AND**
- Adults only (i.e., patients at least 18 years or older): Patient's disease results in one or more of the following:
 - Anemia [i.e., *hemoglobin less than or equal to 11 g/dL (women) or 12 g/dL (men)*] not attributed to iron, folic acid or vitamin B12 deficiency; **OR**
 - Moderate to severe hepatomegaly (liver size 1.25 or more times normal volume) or splenomegaly (spleen size 5 or more times normal volume); **OR**

- Skeletal disease (e.g. lesions, remodeling defects and/or deformity of long bones, osteopenia/osteoporosis, etc.); **OR**
- Symptomatic disease (e.g. bone pain, fatigue, dyspnea, angina, abdominal distension, diminished quality of life, etc.); **OR**
- Thrombocytopenia (platelet count less than or equal to 120,000/mm³).

† FDA Approved Indication(s); Ⓢ Orphan Drug

IV. Renewal Criteria ^{1,9,10,11,12}

- Patient continues to meet the universal and other indication-specific relevant criteria identified in section III; **AND**
- Disease response as indicated by one or more of the following (compared to pre-treatment baseline):
 - Improvement in symptoms (e.g. bone pain, fatigue, dyspnea, angina, abdominal distension, diminished quality of life, etc.)
 - Reduction in size of liver or spleen
 - Improvement in hemoglobin/anemia
 - Improvement in skeletal disease (e.g. increase in lumbar spine and/or femoral neck BMD, no bone crises or bone fractures, etc.)
 - Improvement in platelet counts; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include hypersensitivity reactions.

V. Dosage/Administration

Indication	Dose
Type 1 Gaucher Disease	<ul style="list-style-type: none"> ▪ Initial dosages range from 2.5 U/kg of body weight 3 times a week to 60 U/kg once every 2 weeks based on disease severity. Cerezyme is administered by intravenous infusion over 1–2 hours. ▪ Dosage adjustments should be made on an individual basis and may increase or decrease, based on achievement of therapeutic goals as assessed by routine comprehensive evaluations of the patient’s clinical manifestations.

VI. Billing Code/Availability Information

HCPCS code:

- J1786 – Injection, imiglucerase, 10 units: 1 billable unit = 10 units

NDC:

- Cerezyme 200 unit injection; SDV: 58468-1983-xx
- Cerezyme 400 unit injection; SDV: 58468-4663-xx

VII. References

1. Cerezyme [package insert]. Cambridge, MA; Genzyme Corporation.; April 2018. Accessed May 2020.
2. Anderson HC, et al. Consensus Statement by the International Collaborative Gaucher Group (ICGG) U.S. Coordinators on Individualization of ERT for Type-1 Gaucher Disease. September, 2000.
3. Charrow, et al. Gaucher Disease: Recommendations on Diagnosis, Evaluation and Monitoring (Special Article). Archives of Internal Medicine 1998;158: 1754-1760.
4. Pastores GM, Weinreb NJ, Aerts H, et al. Therapeutic goals in the treatment of Gaucher disease. Semin Hematol 2004; 41:4.
5. Baldellou A, Andria G, Campbell PE, et al. Paediatric non-neuronopathic Gaucher disease: recommendations for treatment and monitoring. Eur J Pediatr 2004; 163:67.
6. Charrow J, Andersson HC, Kaplan P, et al. The Gaucher Registry: Demographics and disease characteristics of 1698 patients with Gaucher disease. Arch Intern Med 2000; 160:2835.
7. Martins AM, Valadares ER, Porta G, et al. Recommendations on diagnosis, treatment, and monitoring for Gaucher disease. J Pediatr. 2009 Oct;155(4 Suppl):S10-8.
8. Kaplan P, Baris H, De Meirleir L, et al. Revised recommendations for the management of Gaucher disease in children. Eur J Pediatr. 2013 Apr;172(4):447-58. doi: 10.1007/s00431-012-1771-z. Epub 2012 Jul 8.
9. Pastores GM, Hughes DA. Gaucher Disease. GeneReviews. www.ncbi.nlm.nih.gov/books/NBK1269/ (Accessed on May 28, 2020).
10. Grabowski GA, Barton NW, Pastores G, et al. Enzyme Therapy in Type 1 Gaucher Disease: Comparative Efficacy of Mannose-Terminated Glucocerebrosidase From Natural and Recombinant Sources. Ann Intern Med 1995 Jan 1;122(1):33-9. doi: 10.7326/0003-4819-122-1-199501010-00005.
11. Biegstraaten M, Cox TM, Belmatoug N, et al. Management goals for type 1 Gaucher disease: An expert consensus document from the European working group on Gaucher disease. Blood Cells, Molecules and Diseases 68 (2018) 203-208.
12. DuaPuri R, Kapoor S, Kishnani PS, et al. Diagnosis and Management of Gaucher Disease in India – Consensus Guidelines of the Gaucher Disease Task Force of the Society for Indian Academy of Medical Genetics and the Indian Academy of Pediatrics. Indian Pediatrics. 5;2018: 143-153.

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
E75.22	Gaucher disease

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

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 Determination (NCD) Local Coverage Determinations (LCDs), and Local Coverage Article may exist and compliance with these policies is required where applicable. They can be found at: <http://www.cms.gov/medicare-coverage-database/search/advanced-search.aspx>. Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD/LCA): 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, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
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