

## VPRIV® (velaglucerase alfa) (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) [Pharmacy Benefit]:

- Vpriv 400 unit powder for injection: 18 vials per 14 days

#### B. Max Units (per dose and over time) [Medical Benefit]:

- 72 billable units every 14 days

### III. Initial Approval Criteria

#### Type 1 Gaucher's Disease †

- Patient age at least 4 years or older; **AND**
- Patient has a documented diagnosis of Type 1 Gaucher Disease as confirmed by reduced glucocerebrosidase activity in peripheral leukocytes; **AND**
- **Adults only criteria (patient at least 18 years or older):** Patient's disease results in one or more of the following:
  - Anemia [hemoglobin less than or equal to 11 g/dL (women) or 12 g/dL (men) ]; **OR**
  - Moderate to severe hepatomegaly (liver size 1.25 or more times normal) or splenomegaly (spleen size 5 or more times normal); **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/mm<sup>3</sup>); **AND**
- Must be used as a single agent

† FDA Approved Indication(s)

#### IV. Renewal Criteria

- Patient continues to meet the criteria 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
  - Improvement in platelet counts; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include severe hypersensitivity reactions, etc.

#### V. Dosage/Administration

Indication	Dose
Type 1 Gaucher Disease	Up to 60 units/kg every other week as a 60-minute intravenous infusion

#### VI. Billing Code/Availability Information

Jcode:

J3385 – Injection, velaglycerase alfa, 100 units: 100 units = 1 billable unit

NDC:

Vpriv 400 unit powder for injection: 54092-0701-xx

#### VII. References

1. VPRIV [package insert]. Cambridge, MA; Shire Human Genetic Therapies; April 2015. Accessed June 2018.
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. Balwani M, Burrow TA, Charrow J, et al. Recommendations for the use of eligustat in the treatment of adults with Gaucher disease Type 1 in the United States. Mol Genet Metab. 2016 Feb; 117(2):95-103. Doi: `0.1016/j.ymgme.2015.09.002. Epub 2015 Sep 7

## 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) and Local Coverage Determinations (LCDs) 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): 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 Government Benefit Administrators, 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