

Vpriv® (velaglucerase alfa)

When requesting Vpriv® (velaglucerase alfa), the individual requiring treatment must be diagnosed with the following FDA-approved indication and meet the specific coverage guidelines and applicable safety criteria for the covered indications.

FDA-approved indication

- Vpriv is indicated for the treatment of individuals with Type 1 Gaucher disease.

Coverage Guidelines

Type 1 Gaucher Disease

The individual must meet **all** of the following criteria for approval:

- The diagnosis is established by one of the following:
 - Demonstration of deficient β - glucocerebrosidase activity in leukocytes or fibroblasts; OR
 - Molecular genetic testing documenting glucocerebrosidase gene mutation;
- AND
- This is being prescribed by or in consultation with a geneticist, endocrinologist, a metabolic disorder sub-specialist, or a physician who specializes in the treatment of lysosomal storage disorders.

Approval duration: 1 year

Dosing Guidelines

- 60 U/kg administered intravenously no more frequently than once every 2 weeks

References

1. Vpriv® for injection [prescribing information]. Lexington, MA: Shire Human Genetic Therapies; December 2020.
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3. Cox T. Gaucher disease: clinical profile and therapeutic development. *Biologics.* 2010;4:299-313.
4. Jmoudiak, M. and Futerman, AH. Gaucher disease: Pathological mechanisms and modern management. *Br J Haematol.* 2005;129(2):178–188.
5. Grabowski GA. Lysosomal storage disease 1- phenotype, diagnosis, and treatment of Gaucher's disease. *Lancet.* 2008;372:1263-1271.
6. Zimran A. How I treat Gaucher disease. *Blood.* 2011;118:1463-1471.
7. Stirnemann J, Belmatoug N, Camou F, et al. A review of Gaucher disease pathophysiology, clinical presentation and treatments. *Int J Mol Sci.* 2017;18:441.
8. Baris HN, Cohen IJ, Mistry PK. Gaucher disease: The metabolic defect, pathophysiology, phenotypes and natural history. *Pediatr Endocrinol Rev.* 2014;12:72-81.