

Mepsevii™ (vestronidase alfa-vjbk) (Intravenous)

Document Number: IC-0346

Last Review Date: 02/04/2019

Date of Origin: 12/12/2017

Dates Reviewed: 12/2017, 10/2018, 02/2019

I. Length of Authorization

Coverage will be provided for twelve months and may be renewed.

II. Dosing Limits

A. Quantity Limit (max daily dose) [Pharmacy Benefit]:

- Mepsevii 10 mg vial: 46 vials per 14 days

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

- 460 billable units (460 mg) every 14 days

III. Initial Approval Criteria

Coverage is provided in the following conditions:

Mucopolysaccharidosis VII (MPS VII; Sly syndrome) †

- Patient has a definitive diagnosis of MPS VII confirmed by BOTH of the following:
 - Beta-glucuronidase enzyme deficiency in peripheral blood leukocytes; **AND**
 - Detection of pathogenic mutations in the *GUSB* gene by molecular genetic testing; **AND**
- Patient aged 5 months or older; **AND**
- Documented baseline value for one or more of the following: six minute walk test (6MWT), motor function [i.e., Bruininks-Oseretsky Test of Motor Proficiency (BOT-2)], liver and/or spleen volume, urinary excretion of glycosaminoglycans (GAGs) such as chondroitin sulfate and dermatan sulfate, skeletal involvement, pulmonary function tests, shoulder flexion, visual acuity, etc.

† FDA-approved indication(s)

IV. Renewal Criteria

Authorizations can be renewed based on the following criteria:

- Patient continues to meet the criteria identified in section III; **AND**

- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include the following: anaphylaxis and severe allergic reactions, etc.; **AND**
- Patient has responded to therapy compared to pretreatment baseline in one or more of the following:
 - Stability or improvement in 6MWT, shoulder flexion, visual acuity, and/or other motor functions
 - Reduction in liver and/or spleen volume
 - Reduction in urinary excretion of GAGs
 - Stability of skeletal disease
 - Stability or improvement in pulmonary function tests

V. Dosage/Administration

Indication	Dose
Mucopolysaccharidosis VII (Sly syndrome)	4 mg/kg of body weight administered as an intravenous infusion over approximately 4 hours once every 2 weeks

VI. Billing Code/Availability Information

Jcode:

- J3397 – Injection, vestronidase alfa-vj bk, 1 mg: 1 billable unit = 1 mg

NDC:

- Mepsevii 10 mg/5 mL single-dose vial: 69794-0001-xx

VII. References

1. Mepsevii [package insert]. Novato, CA; Ultragenyx Pharmaceutical Inc.; November 2017. Accessed January 2019.
2. Montañó AM, Lock-Hock N, Steiner RD, et al. Clinical course of sly syndrome (mucopolysaccharidosis type VII). *J Med Genet.* 2016 Jun;53(6):403-18.

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
E76.29	Other mucopolysaccharidoses

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 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