

Vimizim® (elosulfase alfa) (Intravenous)

Document Number: IC-0190

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Date of Origin: 04/29/2014

Dates Reviewed: 02/2015, 01/2016, 01/2017, 01/2018, 02/2019

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]:

- Vimizim 5mg/5ml: 184 vials every 28 days

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

- 230 billable units (230 mg) every 7 days

III. Initial Approval Criteria

Mucopolysaccharidosis IVA (MPS IVA, Morquio A Syndrome)

- Patient is 5 years of age or older; **AND**
- Documented diagnosis of Mucopolysaccharidosis IVA with biochemical/genetic confirmation by one of the following:
 - Absence or marked reduction in N-acetylgalactosamine 6-sulfatase (GALNS) enzyme activity; **OR**
 - Sequence analysis and/or deletion/duplication analysis of the *GALNS* gene for biallelic mutation; **AND**
- Documented baseline value for one or more of the following: endurance tests (e.g., six minute walk test (6-MWT) or timed 25-foot walk test (T25FW)), pulmonary function tests (e.g., FVC), etc.

IV. Renewal Criteria

- Patient continues to meet the criteria identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: anaphylaxis/hypersensitivity reactions, acute respiratory complications, spinal/cervical cord compression, etc.; **AND**
- Patient has shown a response to therapy as evidenced by one or more of the following markers when compared to pretreatment baseline values:
 - Stability or improvement on endurance tests
 - Stability or improvement in pulmonary function tests

V. Dosage/Administration

Indication	Dose
Mucopolysaccharidosis IVA	2 mg per kg body weight administered once every week as an intravenous infusion over a minimum of 3.5 to 4.5 hours, based on infusion volume

VI. Billing Code/Availability Information

Jcode:

- J1322 – Injection, elosulfase alfa, 1 mg : 1 billable unit = 1 mg

NDC:

- Vimizim 5mg/5ml injection: 68135-0100-xx

VII. References

1. Vimizim [package insert]. Novato, CA; Biomarin Pharmaceutical Inc.; February 2014. Accessed January 2019.
2. Regier DS, Oetgen M, Tanpaiboon P. Mucopolysaccharidosis Type IVA. In: Pagon RA, Adam MP, Bird TD, Dolan CR, Fong CT, Smith RJH, Stephens K, editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2014. 2013 Jul 11 [updated 2014 Mar 13].
3. Hendriksz CJ, Berger KI, Giugliani R, et al. International Guidelines for the Management and Treatment of Morquio A Syndrome. *Am J Med Genet A*. 2015 Jan; 167(1): 11–25. Published online 2014 Oct 24. doi: 10.1002/ajmg.a.36833
4. Regier DS, Oetgen M, Tanpaiboon P. Mucopolysaccharidosis Type IVA. 2013 Jul 11 [Updated 2016 Mar 24]. In: Adam MP, Ardinger HH, Pagon RA, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2019. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK148668/>

Appendix 1 – Covered Diagnosis Codes

ICD-10	Description
E76.210	Morquio A 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