Vimizim® (elosulfase alfa)  
(Intravenous)

Last Review Date: 02/04/2020  
Date of Origin: 04/29/2014  
Dates Reviewed: 02/2015, 01/2016, 01/2017, 01/2018, 02/2019, 02/2020

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

B. Max Units (per dose and over time) [HCPCS Unit]:
   - 230 billable units (230 mg) every 7 days

III. Initial Approval Criteria\textsuperscript{1,4,5,6}

Coverage is provided in the following conditions:

Universal Criteria

- Patient is 5 years of age or older: AND

Mucopolysaccharidosis IVA (MPS IVA, Morquio A Syndrome)

- 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
  - Detection of biallelic pathogenic mutations in the GALNS gene by genetic molecular testing (i.e., sequence analysis and/or deletion/duplication analysis): 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), three minute stair climb test (3-MSCT)), and/or pulmonary function tests (e.g., FVC), etc.
IV. Renewal Criteria\textsuperscript{1,4,5,6}

- Patient continues to meet universal and other indication-specific relevant criteria such as concomitant therapy requirements (not including prerequisite therapy), performance status, etc. identified in section III: \textbf{AND}
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: anaphylaxis and hypersensitivity reactions, acute respiratory complications, spinal/cervical cord compression, etc.: \textbf{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 in six minute walk test (6-MWT), three minute stair climb test (3-MSCT)
  - Stability or improvement in pulmonary function tests

V. Dosage/Administration\textsuperscript{1,4,5,6}

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
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<tr>
<td>Mucopolysaccharidosis IVA</td>
<td>2 mg/kg administered once every week as an intravenous (IV) infusion</td>
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VI. Billing Code/Availability Information

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

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

VII. References


Appendix 1 – Covered Diagnosis Codes

<table>
<thead>
<tr>
<th>ICD-10</th>
<th>Description</th>
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<tr>
<td>E76.210</td>
<td>Morquio A mucopolysaccharidoses</td>
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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 Articles 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/Articles): N/A