**Medical Guideline Disclaimer**

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**Definition**

Vimizim is a purified human enzyme produced by recombinant DNA technology that provides exogenous N-acetylgalactosamine-6-sulfatase. The mannose-6-phosphate-terminated oligosaccharide chains of elosulfase alfa bind to mannose-6-phosphate receptors of lysosomal cells resulting in cellular uptake of elosulfase alfa and increased catabolism of keratan sulfate (KS) and chondroitin-6-sulfate (C6S).

**Length of Authorization**

Coverage will be provided for 12 months and may be renewed.

**Dosing Limits**

- **Max Units (per dose and over time) [Medical Benefit]**:
  - 230 billable units (230 mg) per 7 days

**I. INITIAL APPROVAL CRITERIA**

**Vimizim may be considered medically necessary for the following:**

1. **Mucopolysaccharidosis IV-A (MPS-IV-A, Morquio A Syndrome)**
   a. Patient is 5 years of age or older; **AND**
   b. Documented diagnosis of Mucopolysaccharidosis, MPS-IV-A with biochemical/genetic confirmation by one of the following:
      i. Absence or marked reduction in N-acetylgalactosamine 6-sulfatase (GALNS) enzyme activity; **OR**
      ii. Sequence analysis and/or deletion/duplication analysis of the GALNS gene for biallelic mutation; **AND**
   c. Documented baseline six minute walk test (6-MWT), a current (within 30 days) test will be needed for renewal.
II. RENEWAL CRITERIA

- Disease response with treatment as defined by an improvement in 6-MWT from baseline; **AND**
- Absence of unacceptable toxicity from the drug including hypersensitivity reactions.

Dosage/Administration

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
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<tbody>
<tr>
<td>Mucopolysaccharidosis</td>
<td>– 2 mg/kg Intravenously administered once weekly over a minimum of 3.5 to 4.5 hours.</td>
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Applicable Procedure Codes

- **J1322** Vimizim 5 mg injection; 1 billable unit = 1 mg

Applicable NDCs

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

Applicable Diagnosis Codes

<table>
<thead>
<tr>
<th>ICD-10</th>
<th>ICD-10 Description</th>
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<tbody>
<tr>
<td>E76.210</td>
<td>Morquio A mucopolysaccharidoses</td>
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References