Clinical Policy: Elosulfase alfa (Vimizim)
Reference Number: ERX.SPMN.147
Effective Date: 10/16
Last Review Date: 09/16

See Important Reminder at the end of this policy for important regulatory and legal information.

Policy/Criteria
It is the policy of health plans affiliated with Envolve Pharmacy Solutions® that elosulfase alfa (Vimizim®) is medically necessary when one of the following criteria set is met:

I. Initial Approval Criteria
   A. MPS Type IVA: Morquio A Syndrome (must meet all):
      1. Age ≥ 5 years;
      2. Diagnosis of MPS type IVA (Morquio A syndrome) confirmed by one of the following:
         a. Enzyme assay demonstrating a deficiency of N-acetylgalactosamine-6-sulfatase enzyme activity;
         b. DNA testing.

   Approval duration: 6 months

   B. Other diagnoses/indications: Refer to ERX.SPMN.16 - Global Biopharm Policy.

II. Continued Approval
   A. MPS Type IVA: Morquio A Syndrome (must meet all):
      1. Currently receiving medication via health plan benefit or member has previously met all initial approval criteria.

   Approval duration: 12 months

   B. Other diagnoses/indications (must meet 1 or 2):
      1. Currently receiving medication via health plan benefit and documentation supports positive response to therapy; or
      2. Refer to ERX.SPMN.16 - Global Biopharm Policy.
Background

Description/Mechanism of Action:
Elosulfase alfa is a purified human enzyme produced by recombinant DNA technology in a Chinese hamster ovary cell line. Human N-acetylgalactosamine-6-sulfatase (EC 3.1.6.4) is a hydrolytic lysosomal glycosaminoglycan-specific enzyme that hydrolyzes sulfate from either galactose-6-sulfate or N-acetyl-galactosamine-6-sulfate on the non-reducing ends of the glycosaminoglycans keratan sulfate (KS) and chondroitin-6-sulfate (C6S).

Vimizim is intended to provide the exogenous enzyme N-acetylgalactosamine-6-sulfatase that will be taken up into the lysosomes and increase the catabolism of the GAGs KS and C6S. Elosulfase alfa uptake by cells into lysosomes is mediated by the binding of mannose-6-phosphate-terminated oligosaccharide chains of elosulfase alfa to mannose-6-phosphate receptors.

FDA Approved Indication(s):
Vimizim is an enzyme/intravenous injectable solution indicated for:
- Mucopolysaccharidosis type IVA (MPS IVA; Morquio A syndrome).

Appendices

Appendix A: Abbreviation Key
GAG: glycosaminoglycans
MPS: mucopolysaccharidosis

Coding Implications
Codes referenced in this clinical policy are for informational purposes only. Inclusion or exclusion of any codes does not guarantee coverage. Providers should reference the most up-to-date sources of professional coding guidance prior to the submission of claims for reimbursement of covered services.

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<thead>
<tr>
<th>HCPCS Codes</th>
<th>Description</th>
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<tr>
<td>J1322</td>
<td>Injection, elosulfase alfa, 1 mg</td>
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Reviews, Revisions, and Approvals

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<tr>
<th>Date</th>
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References
**Important Reminder**

This clinical policy has been developed by appropriately experienced and licensed health care professionals based on a review and consideration of currently available generally accepted standards of medical practice; peer-reviewed medical literature; government agency/program approval status; evidence-based guidelines and positions of leading national health professional organizations; views of physicians practicing in relevant clinical areas affected by this clinical policy; and other available clinical information.

This Clinical Policy is not intended to dictate to providers how to practice medicine, nor does it constitute a contract or guarantee regarding payment or results. Providers are expected to exercise professional medical judgment in providing the most appropriate care, and are solely responsible for the medical advice and treatment of members.

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