Elosulfase alfa (Vimizim®)	Place of Service Office Administration Outpatient Facility Infusion Administration Infusion Center Administration Home Infusion HCPCS: J1322 per 1 mg

Condition listed in policy (*see criteria for details*)

- Mucopolysaccharidosis type IVA (MPS IVA / Morquio A syndrome)

AHFS therapeutic class: Enzymes

Mechanism of action: Exogenous enzyme N-acetylgalactosamine-6-sulfatase

(1) Special Instructions and pertinent Information

Covered under the Medical Benefit, please submit clinical information for prior authorization review.

****CRITERIA FOR HOSPITAL OUTPATIENT FACILITY ADMINISTRATION ****

AAAAI Guidelines 2011, MCG™ Care Guidelines, 19th edition, 2015

Members with the following plans: **PPO, Direct Contract HMO, and when applicable, Medi-Cal, ASO/Shared Advantage/HMO (non-direct contract)** may be required to have their medication administered at a preferred site of service, including the home, a physician's office, or an independent infusion center not associated with a hospital.

For members that cannot receive infusions in the preferred home or ambulatory setting AND meet one of the following criteria points, drug administration may be performed at a hospital outpatient facility infusion center.

ADMINISTRATION OF VIMIZIM IN THE HOSPITAL OUTPATIENT FACILITY SITE OF CARE REQUIRES ONE OF THE FOLLOWING: (*Supporting Documentation must be submitted)*

1. **Patient is initiating therapy (allowed for the first 4 infusions) with Vimizim or is being re-initiated on Vimizim after at least 6 months off therapy.** *Subsequent doses will require medical necessity for continued use in the hospital outpatient facility site of care.*

Or

Additional clinical monitoring is required during administration as evidenced by one of the following:

2. **Patient has experienced a previous severe adverse event on Vimizim based on documentation submitted.**

3. **Patient continues to experience moderate to severe adverse events on Vimizim based on documentation submitted, despite receiving premedication such as acetaminophen, steroids, diphenhydramine, fluids, etc.**

4. **Patient is clinically unstable based on documentation submitted.**

5. **Patient is physically or cognitively unstable based on documentation submitted.**

(2) Prior Authorization/Medical Review is required for the following condition(s)

All requests for elosulfase alfa (Vimizim®) must be sent for clinical review and receive authorization prior to drug administration or claim payment.

Mucopolysaccharidosis type IVA (MPS IVA; Morquio A syndrome)

1. Patients with documented clinical diagnosis of Mucopolysaccharidosis type IVA (MPS IVA; Morquio A syndrome) based on clinical signs and symptoms, **AND**
2. Either of the following:
    a. Documented reduced GALNS enzyme activity, OR
    b. Genetic testing confirming diagnosis of MPS IVA

Covered dose
Up to 2 mg/kg IV weekly

Coverage period
Indefinite

ICD-10:
E76.210

(3) The following condition(s) DO NOT require Prior Authorization/Preservice

All requests for elosulfase alfa (Vimizim®) must be sent for clinical review and receive authorization prior to drug administration or claim payment.

(4) This Medication is NOT medically necessary for the following condition(s)

Coverage for a Non-FDA approved indication, requires that criteria outlined in Health and Safety Code § 1367.21, including objective evidence of efficacy and safety are met for the proposed indication.

Please refer to the Provider Manual and User Guide for more information.

(5) Additional Information

How supplied:
5 mg/5 mL (single-use vials)

MPS IV, also known as Morquio syndrome, is an autosomal recessive disorder that occurs in 2 forms with similar clinical features. MPS IVA caused by mutations in the gene encoding galactosamine-6-sulfatase (GALNS) and MPS IVB is due to mutations in the gene encoding beta-galactosidase (GLB1).

The primary manifestations of Morquio syndrome are skeletal and include short stature, pectus carinatum, kyphoscoliosis, genu valgum, coxa valga, dysostosis multiples, spondyloepiphyseal dysplasia, and playspondyly.

(6) References

- AHFS®. Available by subscription at http://www.lexi.com
- DrugDex®. Available by subscription at http://www.micromedexsolutions.com/home/dispatch
- 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;167A(1):11-25. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4309407/pdf/ajmg0167-0011.pdf
- Vimizim® (elosulfase alfa) [Prescribing Information]. Novato, CA: BioMarin Pharmaceuticals. 12/2019.

(7) Policy Update

Date of last review: 3Q2023
Date of next review: 3Q2024
Changes from previous policy version:

- No clinical change to policy following routine annual review.

BSC Drug Coverage Criteria to Determine Medical Necessity
Reviewed by P&T Committee