Medical Policy

Healthcare Services Department

Policy Name	Policy Number	Scope
Vimizim (elosulfase alfa)	MP-RX-FP-102-23	☒ MMM MA ☒ MMM Multihealth

Service Category

☐ Anesthesia
☐ Surgery
☐ Radiology Procedures
☐ Pathology and Laboratory Procedures
☐ Medicine Services and Procedures
☐ Evaluation and Management Services
☐ DME/Prosthetics or Supplies
☒ Part B DRUG

Service Description

This document addresses the use of **Vimizim** (elosulfase alfa) , a drug approved by the Food and Drug Administration (FDA) for the treatment of Mucopolysaccharidosis IVA (Morquio A syndrome).

Background Information

The mucopolysaccharidoses are a group of inherited metabolic diseases caused by the deficiency of lysosomal enzymes needed to breakdown mucopolysaccharides or glycosaminoglycan (GAGs). The progressive accumulation of GAGs in lysosomes leads to respiratory, cardiac, skeletal and connectivity, neurologic and ophthalmologic complications. There are seven distinct types of mucopolysaccharidosis (I, II, III, IV, VI, VII and IX). Accurate diagnosis is important to provide disease-specific enzyme replacement therapy. Diagnosis is confirmed through urinary GAG concentration measurement, enzymatic activity measurement or genetic testing.
Vimizim has a black box warning for anaphylaxis. Life-threatening anaphylactic reactions have occurred during Vimizim infusions. Appropriate medical support should be available during Vimizim administration. Individuals should be educated on the signs and symptoms of anaphylaxis and to seek immediate medical care should they occur. Individuals with acute respiratory illness may be at risk of serious acute exacerbation of their respiratory disease and require additional monitoring

Approved Indications

A. For patients with Mucopolysaccharidosis type IVA (MPS IVA; Morquio A syndrome).

Other Uses

A. N/A

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Applicable Codes

The following list(s) of procedure and/or diagnosis codes is provided for reference purposes only and may not be all inclusive. Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement policy. Benefit coverage for health services is determined by the member specific benefit plan document and applicable laws that may require coverage for a specific service. The inclusion of a code does not imply any right to reimbursement or guarantee claim payment. Other Policies and Guidelines may apply.

HCPCS	Description
J1322	Injection, elosulfase alfa, 1 mg [Vimizim
S9357	Home infusion therapy, enzyme replacement intravenous therapy, (e.g., Imiglucerase); administrative services, professional pharmacy services, care coordination, and all necessary supplies and equipment (drugs and nursing visits coded separately), per diem

ICD-10	Description
E76.210	Morquio A mucopolysaccharidoses

Medical Necessity Guidelines

When a drug is being reviewed for coverage under a member's medical benefit plan or is otherwise subject to clinical review (including prior authorization), the following criteria will be used to determine whether the drug meets any applicable medical necessity requirements for the intended/prescribed purpose.

Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.

Vimizim *(elosulfase alfa)*

A. Prescriber Specialties

i. n/a

B. Criteria For Initial Approval

i. Individual has a diagnosis of mucopolysaccharidosis IVA (Morquio A syndrome); AND
ii. Documentation is provided that diagnosis is confirmed by (Akyol 2019, Wood 2013):
    a. Documented reduced fibroblast or leukocyte N-acetylgalactosamine-6-sulfatase (GALNS) enzyme activity combined with normal enzyme activity level of another sulfatase; OR
    b. Documented genetic testing; AND
iii. Confirmed clinical signs and symptoms of Morquio A syndrome (for example, knee deformity, corneal opacity or pectus carinatum) (Hendriksz 2015, Wood 2013).

C. Criteria For Continuation of Therapy

i. Documentation is provided to show clinically significant improvement or stabilization in clinical signs and symptoms of disease (including but not limited to reduction in urinary GAG excretion, reduction in hepatosplenomegaly, improvement in pulmonary function, improvement in walking distance and/or improvement in fine or gross motor function) compared to the predicted natural history trajectory of disease

D. Authorization Duration

i. Mucopolysaccharidosis type IVA (MPS IVA; Morquio A syndrome
    a. Initial Approval Duration: 1 year
    b. Reauthorization Approval Duration: 1 year

E. **Conditions Not Covered**

Any other use is considered experimental, investigational, or unproven, including the following (this list may not be all inclusive):

i. Individual is using to treat mucopolysaccharidosis IVB (Morquio B syndrome); OR
ii. May not be approved when the above criteria are not met and for all other indications

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Limits or Restrictions

A. Quantity Limitations

Approvals may be subject to dosing limits in accordance with FDA-approved labeling, accepted compendia, and/or evidence-based practice guidelines. The chart below includes dosing recommendations as per the FDA-approved prescribing information.

Drug	Limit
Vimizim (elosulfase alfa) 5 mg vial	2 mg/kg once a week
Exceptions	
N/A	

Reference Information

1. Akyol MU, Alden TD, Amartino H, et al. Recommendations for the management of MPS IVA: systematic evidence and consensusbased guidance. Orphanet J Rare Dis. 2019;14(1):137. doi: 10.1186/s13023-019-1074-9
2. based guidance. Orphanet J Rare Dis. 2019;14(1):137. doi: 10.1186/s13023-019-1074-9 2. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: September 9, 2022.
3. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically
4. Hendriksz CJ, Berger KI, Giugliani R, at al. International guidelines for the management and treatment of Morquio A syndrome. Am J Med Genet A. 2015; 167A(1):11-25
5. Lehman TJ, Miller N, Norquist B, Underhill L, Keutzer J. Diagnosis of the mucopolysaccharidoses. Rheumatology (Oxford). 2011;50 Suppl 5:v41-v48. doi:10.1093/rheumatology/ker390.
6. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2022; Updated periodically.
7. Wood TC, Harvey K, Beck M, et al. Diagnosing mucopolysaccharidosis IVA. J Inherit Metab Dis. 2013;36:293–307

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Policy History

Revision Type	Summary of Changes	P&T Approval Date	MPCC Approval Date
Policy Inception	Elevance Health's Medical Policy adoption.	N/A	11/30/2023

Revised: 9/27/23