Medical Policy

Utilization Management and Clinical Medical Policy

Policy Name:	Policy Number:	Scope:	Origination Date:	Effective Date:
Elosulfase alfa (Vimizim®)	**MP-RX-FP-102-23**	☒ **MMM MA** ☒ **MMM MultiHealth**	**11/30/2023** **Last Review Date:** **10/21/2026**	**10/21/2026** **Frequently Revision:** **Annual**

Service Category:

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

Service Description:

This document addresses the use of elosulfase alfa (Vimizim®) , a hydrolytic lyposomal glycosaminoglycan (GAG)- specific enzyme approved by the Food and Drug Administration (FDA) for the treatment of Mucopolysaccharidosis type IVA (MPS IVA; Morquio A syndrome).

Background Information:

This document addresses Vimizim (elosulfase alfa), an enzyme replacement therapy approved by the Food and Drug Administration (FDA) to treat individuals with Mucopolysaccharidosis type IVA (MPS IVA; Morquio A syndrome).

The mucopolysaccharidoses are a group of inherited metabolic diseases caused by the deficiency of lysosomal enzymes needed to breakdown mucopolysaccharides, also known as glycosaminoglycans (GAGs). The progressive accumulation of GAGs in lysosomes leads to respiratory, cardiac, skeletal and connective tissue, 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. For MPS IVA, diagnosis is established by identification of low N-acetylgalactosamine-6-sulfatase (GALNS) enzyme activity or biallelic pathogenic variants in GALNS on molecular genetic testing.

Vimizim has a black box warning for hypersensitivity reactions including anaphylaxis and risk of acute respiratory complications. Life-threatening anaphylactic reactions have occurred during the early course of enzyme replacement therapy and after extended duration of therapy. Appropriate medical support should be available during Vimizim administration, including access to cardiopulmonary resuscitation equipment. If a severe hypersensitivity reaction occurs, Vimizim should be discontinued and appropriate medical treatment, including epinephrine, should be initiated. Individuals should be educated on the signs and symptoms of anaphylaxis and to seek immediate medical care should they occur. Individuals with acute febrile or respiratory illness may be at risk of serious acute exacerbation of their respiratory disease and require additional monitoring; careful consideration should be given to the patient's clinical status prior to administration, and delaying the infusion should be considered. Patients with MPS IVA should also be monitored for signs and symptoms of spinal or cervical cord compression, a known serious complication of MPS IVA.

Approved Indications

A. Mucopolysaccharidosis type IVA (MPS IVA; Morquio A syndrome).

Other Uses

A. None.

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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.

elosulfase alfa (Vimizim®)

A. Criteria For Initial Approval (*Provider must submit documentation [such as office chart notes, lab results, pathology reports, imaging studies, and any other pertinent clinical information] supporting the patient's diagnosis for the drug and confirming that the patient has met **all** approval criteria.)*

Initial requests for Vimizim (elosulfase alfa) may be approved if the following criteria are met:

i. Individual has a diagnosis of mucopolysaccharidosis IVA (Morquio A syndrome); **AND**

ii. Documentation is provided that diagnosis is confirmed by one of the following (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 biallelic pathogenic or likely pathogenic variants in the GALNS gene; **AND**

iii. Individual demonstrates clinical signs and symptoms of Morquio A syndrome including, but not limited to, skeletal abnormalities, short stature, gait impairment, reduced endurance, knee deformity, corneal opacity, pectus carinatum, or respiratory involvement (Hendriksz 2015, Wood 2013).

B. Criteria For Continuation of Therapy

Continuation requests for Vimizim may be approved if the following criteria are met:

i. Individual has no evidence of unacceptable toxicity or severe hypersensitivity reaction that would preclude continued treatment; **AND**

ii. Documentation is provided to show clinically significant improvement or stabilization in clinical signs and symptoms of disease compared to baseline or to the expected natural history trajectory of disease, including but not limited to improvement or stabilization in endurance or walking distance, pulmonary function, activities of daily living, fine or gross motor function, mobility, respiratory status, or reduction in urinary keratan sulfate/GAG excretion.

C. Authorization Duration

i. Initial Approval Duration: 1 year

ii. Reauthorization Approval Duration: 1 year

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D. Conditions Not Covered

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

Requests for Vimizim (elosulfase alfa) may not be approved for the following:

i. Individual is using Vimizim 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. Therapeutic Alternatives

The list below includes preferred alternative therapies recommended in the approval criteria and may be subject to prior authorization.

i. N/A

B. 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	Recommended Dosage/Limit
Vimizim (elosulfase alfa) injection 5 mg/5mL vial	• 2 mg/kg IV per body weight, based on infusion volume, once a week
Exceptions	
N/A	

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Codes Information:

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.

ICD-10 Diagnostic Codes:

Codes	Description
E76.210	Morquio A mucopolysaccharidoses

HCPCS Codes:

Codes	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

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Reference Information:

1. Akyol MU, Alden TD, Amartino H, et al. Recommendations for the management of MPS IVA: systematic evidence and consensus based guidance. Orphanet J Rare Dis. 2019;14(1):137. doi: 10.1186/s13023-019-1074-9
2. BioMarin Pharmaceutical Inc. Vimizim® (elosulfase alfa) injection, for intravenous use: Prescribing Information. Revised October 2025. Accessed July 6, 2026. https://www.vimizim.com/en-us/wp-content/uploads/sites/2/2025/12/prescribing-information-2025.pdf?v=0.51
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. Regier DS, Oetgen M, Tanpaiboon P. Mucopolysaccharidosis Type IVA. GeneReviews®. University of Washington, Seattle; updated August 12, 2021. Accessed July 6, 2026. https://www.ncbi.nlm.nih.gov/books/NBK148668/
8. Wood TC, Harvey K, Beck M, et al. Diagnosing mucopolysaccharidosis IVA. J Inherit Metab Dis. 2013;36:293–307

Federal and state laws or requirements, contract language, and Plan utilization management programs or polices may take precedence over the application of this clinical criteria.

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Policy History:

Type of Review	Summary of Changes	P&T Approval Date	UM/CMPC Approval Date
Annual Review	Updated the Service Description and Background Information to align with the current FDA Prescribing Information, including terminology clarifications and updated boxed warning language for hypersensitivity reactions, anaphylaxis, and acute respiratory complications. Updated initial approval criteria to clarify diagnostic confirmation through reduced GALNS enzyme activity or biallelic pathogenic/likely pathogenic GALNS variants. Refined continuation of therapy criteria to include absence of unacceptable toxicity or severe hypersensitivity reactions and documentation of clinical improvement or stabilization. Updated quantity limitations to reflect current PI dosing. Coding Reviewed: no changes. Updated reference information and incorporated new policy template.	10/15/2026	10/21/2026
Annual Review	Minimal changes; word formatting. Coding reviewed: No changes.	9/5/2025	9/16/2025
Annual Review	Clarify diagnosis confirmation criteria. Wording and formatting changes. Coding Reviewed: No changes.	3/14/2025	4/2/2025
Policy Inception	Elevance Health's Medical Policy adoption.	N/A	11/30/2023