Medical Policy:

Vimizim® (elosulfase alfa) intravenous infusion

POLICY NUMBER	LAST REVIEW	ORIGIN DATE
MG.MM.PH.310	February 6, 2025	March 2019

The treating physician or primary care provider must submit to EmblemHealth, or ConnectiCare, as applicable (hereinafter jointly referred to as "EmblemHealth"), the clinical evidence that the member meets the criteria for the treatment or surgical procedure. Without this documentation and information, EmblemHealth will not be able to properly review the request preauthorization or post-payment review. The clinical review criteria expressed below reflects how EmblemHealth determines whether certain services or supplies are medically necessary. This clinical policy is not intended to pre-empt the judgment of the reviewing medical director or dictate to health care providers how to practice medicine. Health care providers are expected to exercise their medical judgment in rendering appropriate care. Health care providers are expected to exercise their medical judgment in rendering appropriate care.

EmblemHealth established the clinical review criteria based upon a review of currently available clinical information (including clinical outcome studies in the peer reviewed published medical literature, regulatory status of the technology, evidence-based guidelines of public health and health research agencies, evidence-based guidelines and positions of leading national health professional organizations, views of physicians practicing in relevant clinical areas, and other relevant factors). EmblemHealth expressly reserves the right to revise these conclusions as clinical information changes and welcomes further relevant information. Each benefit program defines which services are covered. The conclusion that a particular service or supply is medically necessary does not constitute a representation or warranty that this service or supply is covered and/or paid for by EmblemHealth, as some programs exclude coverage for services or supplies that EmblemHealth considers medically necessary.

If there is a discrepancy between this guideline and a member's benefits program, the benefits program will govern. Identification of selected brand names of devices, tests and procedures in a medical coverage policy is for reference only and is not an endorsement of any one device, test or procedure over another. In addition, coverage may be mandated by applicable legal requirements of a state, the Federal Government or the Centers for Medicare & Medicaid Services (CMS) for Medicare and Medicaid members. All coding and web site links are accurate at time of publication.

EmblemHealth may also use tools developed by third parties, such as the MCG™ Care Guidelines, to assist us in administering health benefits. The MCG™ Care Guidelines are intended to be used in connection with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice. EmblemHealth Services Company, LLC, has adopted this policy in providing management, administrative and other services to EmblemHealth Plan, Inc., EmblemHealth Insurance Company, EmblemHealth Services Company, LLC, and Health Insurance Plan of Greater New York (HIP) related to health benefit plans offered by these entities. ConnectiCare, an EmblemHealth company, has also adopted this policy. All of the aforementioned entities are affiliated companies under common control of EmblemHealth Inc.

Definitions

Vimizim, a human N-acetylgalactosamine-6-sulfatase, is indicated for patients with Mucopolysaccharidosis type IVA (Morquio A syndrome [MPS IVA]). It is produced in Chinese hamster ovary cells via recombinant DNA technology. Vimizim is a hydrolytic lysosomal enzyme which is taken up by lysosomes and hydrolyzes sulfate from the non-reduced ends of the glycosaminoglycans keratan sulfate and chondroitin-6-sulfate.

Length of Authorization

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

Dosing Limits [Medical Benefit]

Approve up to 2 mg/kg administered intravenously no more frequently than once a week; 230 billable units (230 mg) every 7 days

Guideline

I. INITIAL CRITERIA

1. **Mucopolysaccharidosis Type IVA (Morquio A Syndrome):**

A. Patient is at least 5 years of age; **AND**
B. The diagnosis is established by **ONE** of the following (i or ii):
    i. Patient has a laboratory test demonstrating deficient *N*-acetylgalactosamine-6-sulfatase activity in leukocytes or fibroblasts; **OR**
    ii. Patient has a molecular genetic test demonstrating *N*-acetylgalactosamine-6-sulfatase gene mutation; **AND**
C. Documented baseline for **ONE** or more of the following:
    i. Endurance tests (e.g., six-minute walk test [6-MWT], timed 25-foot walk test [T25FW],
    ii. three-minute stair climb test [3-MSCT])
    iii. Pulmonary function tests (e.g., maximum voluntary ventilation [MVV], percent predicted
    iv. forced vital capacity [FVC], etc.)
    v. Urine keratan sulfate (KS) or urine glycosaminoglycan (GAG) levels
D. Vimizim is prescribed by or in consultation with a geneticist, endocrinologist, a metabolic disorder sub-specialist, or a physician who specializes in the treatment of lysosomal storage disorders.

II. RENEWAL CRITERIA

1. Patient continues to meet indication-specific criteria such as concomitant therapy identified in INITIAL CRITERIA; **AND**
2. Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: anaphylaxis and hypersensitivity reactions, acute respiratory complications, spinal/cervical cord compression, etc.; **AND**
3. Patient has shown a response to therapy as evidenced by the following when compared to pretreatment baseline values:
    a. Stability or improvement in endurance tests (e.g., six-minute walk test [6-MWT], timed 25-foot walk test [T25FW], three-minute stair climb test [3-MSCT]); **OR**
    b. Stability or improvement in pulmonary function tests (e.g., FVC, etc.); **OR**
    c. Stability or reduction in urine keratan sulfate (KS) or urine glycosaminoglycan (GAG) levels

Applicable Procedure Codes

Code	Description
J1322	Injection, elosulfase alfa, 1 mg

Applicable NDCs

Code	Description
68135-0100-01	Vimizim 5MG/5ML Solution

ICD-10 Diagnoses

Code	Description
E76.210	Morquio A mucopolysaccharidoses

Revision History

Company(ies)	DATE	REVISION

EmblemHealth & ConnectiCare	2/6/2025	Annual Review: Initial criteria: added: "Documented baseline for ONE or more of the following: Endurance tests (e.g., six-minute walk test [6-MWT], timed 25-foot walk test [T25FW], three-minute stair climb test [3-MSCT]), Pulmonary function tests (e.g., maximum voluntary ventilation [MVV], percent predicted , forced vital capacity [FVC], etc.), Urine keratan sulfate (KS) or urine glycosaminoglycan (GAG) levels" Renewal Criteria: Added: "Stability or reduction in urine keratan sulfate (KS) or urine glycosaminoglycan (GAG) levels"
EmblemHealth & ConnectiCare	1/2/2024	Annual Review: Updated dosing limits, added age restriction, added renewal criteria
EmblemHealth & ConnectiCare	4/07/2023	Transfer from CCUM template to Co-Branded Medical template Retired MG.MM.PH.178
EmblemHealth & ConnectiCare	4/06/2022	Annual Revision: no criteria changes
EmblemHealth & ConnectiCare	4/07/2021	Annual Revision: no criteria changes

References

1. Vimizim® intravenous infusion [prescribing information]. Novato, CA: BioMarin; January 2021.