Medical Policy:

Naglazyme® (galsulfase) intravenous infusion

POLICY NUMBER	LAST REVIEW	ORIGIN DATE
MG.MM.PH.305	March 17, 2025	December 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

Naglazyme, a human N-acetylgalactosamine 4-sulfatase, is indicated for patients with Mucopolysaccharidosis type VI (Maroteaux – Lamy syndrome [MPS VI]). It is produced in a Chinese hamster ovary cell line via recombinant DNA technology. The enzyme catalyzes the hydrolysis of the sulfate ester from the glycosaminoglycans, chondroitin 4-sulfate and dermatan sulfate. Naglazyme has been shown to improve walking and stair climbing capacity.

Length of Authorization

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

Dosing Limits [Medical Benefit]

Each dose must not exceed 1 mg/kg administered intravenously no more frequently than once weekly.

Max Units (per dose and over time) [HCPCS Unit]:

115 billable units every 7 days

Guideline

I. INITIAL CRITERIA

1. **Mucopolysaccharidosis Type VI (Maroteaux – Lamy Syndrome).** Approve if the patient meets the following criteria:
   A. The diagnosis is established by **ONE** of the following (i or ii):
      i. Patient has a laboratory test demonstrating deficient *N*-acetylgalactosamine 4-sulfatase (arylsulfatase B) activity in leukocytes or fibroblasts; **OR**
      ii. Patient has a molecular genetic test demonstrating biallelic pathogenic or likely pathogenic arylsulfatase B (*ARSB*) gene variants; **AND**
   B. Patient is at least 5 years of age; **AND**
   C. Documented baseline 12-minute walk test (12-MWT), 3-minute stair climb test (3-MSCT), and/or pulmonary function tests (e.g., FEV1, etc.); **AND**
   D. Documented baseline value for urinary glycosaminoglycan (uGAG); **AND**
   E. Therapy is being used to treat non-central nervous system manifestations of the disease and patient does not have severe, irreversible cognitive impairment; **AND**
   F. Naglazyme 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

Coverage can be renewed based on the following criteria:

1. Patient continues to meet Initial Criteria; **AND**
2. Absence of unacceptable toxicity from the drug. *Examples of unacceptable toxicity include: anaphylaxis and hypersensitivity reactions, immune-mediated reactions, acute respiratory complications associated with administration, acute cardiorespiratory failure, severe infusion reactions, spinal or cervical cord compression, etc*.; **AND**
3. Patient has a documented reduction in uGAG levels compared to pretreatment baseline; **AND**
4. Patient has demonstrated a beneficial disease response to therapy compared to pre-treatment baseline values as defined in **ONE** or more of the following:
   A. Improvement in or stability of 12-minute walk test (12-MWT); **OR**
   B. Improvement in or stability of 3-minute stair climb test (3-MSCT); **OR**
   C. Improvement in or stability of pulmonary function testing (e.g., FEV1, etc.)

Applicable Procedure Codes

Code	Description
J1458	Injection, galsulfase, 1 mg

Applicable NDCs

Code	Description
68135-0020-01	Naglazyme 1mg/mL Solution

ICD-10 Diagnoses

Code	Description
E76.29	Other mucopolysaccharidoses

Revision History

Company(ies)	DATE	REVISION
EmblemHealth & ConnectiCare	3/17/2025	Annual Review: Initial Criteria: Removed" arylsulfatase B gene mutation; AND" from the following statement: "Patient has a molecular genetic test demonstrating arylsulfatase B gene mutation; AND" and replaced with: "biallelic pathogenic or likely pathogenic arylsulfatase B (*ARSB*) gene variants; AND" Added: "Patient is at least 5 years of age; AND Documented baseline 12-minute walk test (12-MWT), 3-minute stair climb test (3-MSCT), and/or pulmonary function tests (e.g., FEV1, etc.); AND Documented baseline value for urinary glycosaminoglycan (uGAG); AND Therapy is being used to treat non-central nervous system manifestations of the disease and patient does not have severe, irreversible cognitive impairment; AND" Added "RENEWAL CRITERIA Coverage can be renewed based on the following criteria: Patient continues to meet Initial Criteria; AND Absence of unacceptable toxicity from the drug. *Examples of unacceptable toxicity include: anaphylaxis and hypersensitivity reactions, immune-mediated reactions, acute respiratory complications associated with administration, acute cardiorespiratory failure, severe infusion reactions, spinal or cervical cord compression, etc*.; AND Patient has a documented reduction in uGAG levels compared to pretreatment baseline; AND Patient has demonstrated a beneficial disease response to therapy compared to pre-treatment baseline values as defined in ONE or more of the following: Improvement in or stability of 12-minute walk test (12-MWT); OR Improvement in or stability of 3-minute stair climb test (3-MSCT); OR Improvement in or stability of pulmonary function testing (e.g., FEV1, etc.)"
EmblemHealth & ConnectiCare	2/1/2024	Annual Review: no criteria changes
EmblemHealth & ConnectiCare	04/07/2023	Transfer from CCUM template to Co-Branded Medical template Retired MG.MM.PH.93
EmblemHealth & ConnectiCare	04/06/2022	Annual Revision: no criteria changes
EmblemHealth & ConnectiCare	04/07/2021	Annual Revision: no criteria changes

References

1. Naglazyme® intravenous infusion [prescribing information]. Novato, CA: BioMarin; April 2020.