Reference number(s)
2691-A

Standard Medicare Part B Management Naglazyme

Products Referenced by this Document

Drugs that are listed in the following table include both brand and generic and all dosage forms and strengths unless otherwise stated. Over the counter (OTC) products are not included unless otherwise stated.

Brand Name	Generic Name
Naglazyme	galsulfase

Indications

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-approved Indications[1]

Naglazyme is indicated for patients with Mucopolysaccharidosis VI (MPS VI, Maroteaux-Lamy syndrome). Naglazyme has been shown to improve walking and stair-climbing capacity.

All other indications will be assessed on an individual basis. Submissions for indications other than those listed in this criteria should be accompanied by supporting evidence from Medicare approved compendia.

Documentation

The following documentation must be available, upon request, for all submissions:

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Initial Requests:

- N-acetylgalactosamine-4-sulfatase (arylsulfatase B) enzyme assay or genetic testing results supporting diagnosis.

Continuation Requests:

- Chart notes or medical records documenting a clinically positive response to therapy, which shall include improvement, stabilization, or slowing of disease progression (e.g., improvement, stabilization, or slowing of disease progression for growth rate, 12-minute walk test, 3-minute stair climb test, urinary glycosaminoglycan (GAG) levels).

Coverage Criteria

Mucopolysaccharidosis VI (MPS VI, Maroteaux-Lamy syndrome)[1,2]

Authorization of 12 months may be granted for treatment of MPS VI (Maroteaux-Lamy syndrome) when the diagnosis of MPS VI was confirmed by enzyme assay demonstrating a deficiency of N-acetylgalactosamine-4-sulfatase (arylsulfatase B) enzyme activity or by genetic testing.

Continuation of Therapy

All members (including new members) requesting authorization for continuation of therapy must be currently receiving therapy with the requested agent.

Authorization of 12 months may be granted when all of the following criteria are met:

- The member is currently receiving therapy with the requested medication.
- The requested medication is being used to treat an indication listed in the Coverage Criteria section.
- The member is receiving benefit from therapy. Benefit is defined as a clinically positive response to therapy, which shall include improvement, stabilization, or slowing of disease progression (e.g., improvement, stabilization, or slowing of disease progression for growth rate, 12-minute walk test, 3-minute stair climb test, urinary glycosaminoglycan (GAG) levels).

Summary of Evidence

The contents of this policy were created after examining the following resources:

- The prescribing information for Naglazyme.
- The available compendium

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- National Comprehensive Cancer Network (NCCN) Drugs and Biologics Compendium
- Micromedex DrugDex
- American Hospital Formulary Service- Drug Information (AHFS-DI)
- Lexi-Drugs
- Clinical Pharmacology

After reviewing the information in the above resources, the FDA-approved indications listed in the prescribing information for Naglazyme are covered.

Explanation of Rationale

Support for FDA-approved indications can be found in the manufacturer's prescribing information.

Support for using enzyme assays or genetic testing prior to starting Naglazyme to treat MPS VI can be found in a guideline by Akyol et al. The diagnosis of MPS VI can be confirmed by one of two ways: confirmation of ASB enzyme activity in cultured fibroblasts or isolated leukocytes of less than 10% of the lower limit of normal or demonstration of two disease-causing variants.

References

1. Naglazyme [package insert]. Novato, CA: BioMarin Pharmaceutical Inc.; September 2024.
2. Akyol, M.U., Alden, T.D., Amartino, H. et al. Recommendations for the management of MPS VI: systematic evidence- and consensus-based guidance. Orphanet J Rare Dis 14, 118 (2019).
3. Decker C, Yu ZF, Giugliani R, et al. Enzyme replacement therapy for mucopolysaccharidosis VI: Growth and pubertal development in patients treated with recombinant human N-acetylgalactosamine 4-sulfatase. *J Pediatr Rehabil Med*. 2010;3(2):89-100. doi:10.3233/PRM-2010-0113

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