| Reference number(s) |
| --- |
| 2056- A |

# Specialty Guideline 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 Indication[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 are considered experimental/investigational and not medically necessary.

## Documentation

Submission of the following information is necessary to initiate the prior authorization review:

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

- Continuation requests: chart notes documenting a clinically positive response to therapy, which shall include improvement, stabilization, or slowing of disease progression.

# Prescriber Specialties

This medication must be prescribed by or in consultation with a physician who specializes in the treatment of metabolic disease and/or lysosomal storage disorders.

# 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

Authorization of 12 months may be granted for continued treatment in members requesting reauthorization for an indication listed in the Coverage Criteria section who have a clinically positive response to therapy, which shall include improvement, stabilization, or slowing of disease progression.

# 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).