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Section:	Prescription Drugs	**Effective Date:**	April 1, 2024
Subsection:	Endocrine and Metabolic Drugs	**Original Policy Date:**	March 22, 2006
Subject:	Naglazyme	**Page:**	1 of 3

Last Review Date: March 8, 2024

Naglazyme

Description

Naglazyme (galsulfase)

Background
Naglazyme is indicated for Maroteaux-Lamy Syndrome (MPS VI), which is an inherited lysosomal storage disorder caused by the deficiency of N-acetylgalactosamine 4-sulfatase. N-acetylgalactosamine 4-sulfatase is an enzyme required for the breakdown of certain complex carbohydrates. The deficiency of this enzyme results in the accumulation of the glycosaminoglycan (GAG) substrate throughout the body. Naglazyme provides the needed enzyme that will be taken up into the lysosomes and increase the catabolism of GAG. Naglazyme has been shown to help people with MPS VI walk farther and climb more stairs (1).

Regulatory Status
FDA-approved indication: Naglazyme is a hydrolytic lysosomal glycosaminoglycan (GAG)-specific enzyme indicated for patients with Mucopolysaccharidosis VI (MPS VI; Maroteaux-Lamy syndrome). Naglazyme has been shown to improve walking and stair-climbing capacity (1).

Physicians should monitor patients for the development of immune complex mediated reactions while receiving the infusion. Appropriate medical monitoring and support measures should be available during infusion for the possible risk of acute cardio respiratory failure and respiratory support for acute respiratory complications. Pretreatment with antihistamines with or without antipyretics is recommended prior to the start of the infusion to reduce risk of reactions (1).

Related policies
Aldurazyme, Elaprase, Mepsevii, Vimizim

Policy

This policy statement applies to clinical review performed for pre-service (Prior Approval, Precertification, Advanced Benefit Determination, etc.) and/or post-service claims.

Naglazyme may be considered **medically necessary** if the conditions indicated below are met.

Naglazyme may be considered **investigational** for all other indications.

Prior-Approval Requirements

Diagnosis

Patient must have the following:

Maroteaux-Lamy Syndrome

Prior – Approval *Renewal* Requirements

Same as above

Policy Guidelines

Pre - PA Allowance

None

Prior - Approval Limits

Duration 2 years

Prior – Approval *Renewal* Limits

Same as above

Rationale

Summary
Naglazyme is a hydrolytic lysosomal glycosaminoglycan (GAG)-specific enzyme indicated for patients with Mucopolysaccharidosis VI (MPS VI; Maroteaux-Lamy syndrome). Naglazyme has

been shown to improve walking and stair-climbing capacity. Observing of life-threatening anaphylactic reactions should be done during infusions. Pretreatment with antihistamines with or without antipyretics is recommended prior to the start of infusion to reduce the risk of infusion-reactions (1).

Prior authorization is required to ensure the safe, clinically appropriate, and cost-effective use of Naglazyme while maintaining optimal therapeutic outcomes.

References

1. Naglazyme [package insert]. Novato, CA: BioMarin Pharmaceutical Inc; December 2019.

Policy History

Date	Action
March 2010	Criteria reviewed and the approval time limit was changed to lifetime to be consistent with the other enzyme agents and ICD-10 codes added.
September 2011	Annual Review
September 2012	Annual editorial review and reference update
June 2013	Annual editorial review and reference update
September 2014	Annual editorial review and reference update
September 2015	Annual editorial review and reference update
September 2016	Annual editorial review Policy number changed from 5.08.17 to 5.30.01
December 2017	Annual review
June 2018	Annual editorial review
December 2019	Annual editorial review. Changed approval duration from lifetime to 2 years
December 2020	Annual review and reference update
September 2021	Annual review
September 2022	Annual review
March 2023	Annual review
March 2024	Annual review

Keywords

This policy was approved by the FEP® Pharmacy and Medical Policy Committee on March 8, 2024 and is effective on April 1, 2024.