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 for the treatment of Maroteaux-Lamy syndrome.

Naglazyme may be considered investigational in patients without a diagnosis of Maroteaux-Lamy syndrome.

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