Naglazyme (galsulfase)

Override(s)	Approval Duration
Prior Authorization	1 year

Medications	Dosing Limit
Naglazyme (galsulfase) 5 mg vial	1 mg/kg once per week

APPROVAL CRITERIA

Initial requests for Naglazyme (galsulfase) may be approved if the following criteria are met:

I. Individual has a diagnosis of Mucopolysaccharidosis VI (Maroteaux-Lamy syndrome) confirmed by (Akyol 2019, Wood 2012)**:**
   A. Documentation is provided showing an increase in dermatan sulfate in the urine and a decrease in the activity of N-acetylgalactosamine-4-sulfatase (arylsulfatase B) enzyme as measured in fibroblasts or leukocytes combined with normal enzyme activity level of another sulfatase; **OR**
   B. Documentation is provided showing an N-acetlygalactosamine-4-sulfatase (arylsulfatase B) gene mutation.

Continuation requests for Naglazyme (galsulfase) may be approved if the following criterion is met:

I. Documentation is provided to show clinically significant improvement or stabilization in clinical signs and symptoms of disease (including but not limited to reduction in urinary GAG excretion, reduction in hepatosplenomegaly, improvement in pulmonary function, improvement in walking distance and/or improvement in fine or gross motor function) compared to the predicted natural history trajectory of disease.

Naglazyme (galsulfase) may not be approved when the above criteria are not met and for all other indications.

Key References:

1. Akyol MU, Alden TD, Amartino H, et al. Recommendations for the management of MPS VI: systematic evidence and consensus-based guidance. *Orphanet J Rare Dis*. 2019;14(1):118. doi: 10.1186/s13023-019-1080-y.
2. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: September 9, 2023.
3. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
4. Lehman TJ, Miller N, Norquist B, Underhill L, Keutzer J. Diagnosis of the mucopolysaccharidoses. *Rheumatology (Oxford)*. 2011;50 Suppl 5:v41-v48. doi:10.1093/rheumatology/ker390.
5. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc. Updated periodically.
6. Valayannopoulos V, Nicely H, Harmatz P, Turbeville S. Mucopolysaccharidosis VI. *Orphanet J Rare Dis*. 2010; 5:5.

7. Wang RY, Bodamer OA, Watson MS, Wilcox WR. American College of Medical Genetics (ACMG) Work Group on Diagnostic Confirmation of Lysosomal Storage Diseases. Lysosomal storage diseases: diagnostic confirmation and management of presymptomatic individuals. *Genet Med*. 2011; 13(5):457-484.
8. Wood T, Bodamer OA, Burin MG, et al. Expert recommendations for the laboratory diagnosis of MPS VI. *Mol Genet Metab*. 2012; 106(1):73-82.

Federal and state laws or requirements, contract language, and Plan utilization management programs or polices may take precedence over the application of this clinical criteria.