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Ascension

Galsulfase

(Naglazyme®) J1458 (1 mg)

Covered with prior authorization

Requests for Naglazyme® (galsulfase) may be approved if the following criteria are met:

- Documentation is provided that individual has a diagnosis of Mucopolysaccharidosis VI (Maroteaux-Lamy syndrome) confirmed by (Akyol 2019, Wood 2012):
    - 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**
    - Documented N-acetylgalactosamine-4-sulfatase (arylsulfatase B) gene mutation.

Requests for Naglazyme® (galsulfase) may **not** be approved if the above criteria are not met and for all other indications not included above.

Initial and renewal authorizations are for up to 12 months.

Annual reauthorizations will require medical chart documentation that the patient has been seen within the past 12 months and that markers of disease are improved by therapy. Documentation provided should 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.

When coverage is available and medically necessary, the dosage, frequency, duration of therapy, and site of care should be reasonable, clinically appropriate, and supported by evidence-based literature and adjusted based upon severity, alternative available treatments, and previous response to therapy.

Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement.

Naglazyme® (galsulfase) How Supplied and Usual Dosing:

- Naglazyme® (galsulfase) 5 mg vial - 1 mg/kg once a week

Exclusion criteria:

- Naglazyme® (galsulfase) may not be approved when the above criteria are not met and for all other indications.
- Doses, durations, or dosing intervals that exceed FDA maximum limits for any FDA-approved indication or are not supported by industry-accepted practice guidelines or peer-reviewed literature for the relevant off-label use;
- Individuals with significant known risk factors unless the record provides an assessment of clinical benefit that outweighs the risk.

U.S. Food and Drug Administration:
This section is to be used for informational purposes. FDA approval alone is not a basis for coverage. Naglazyme® (galsulfase) is an enzyme replacement therapy approved by the Food and Drug Administration (FDA) to treat individuals with Mucopolysaccharidosis VI (Maroteaux-Lamy syndrome). The mucopolysaccharidoses are a group of inherited metabolic diseases caused by the deficiency of lysosomal enzymes needed to break down mucopolysaccharides or glycosaminoglycan (GAG). The progressive accumulation of GAG in lysosomes leads to respiratory, cardiac, skeletal and connectivity, neurologic and ophthalmologic complications. There are seven distinct types of mucopolysaccharidosis (I, II, III, IV, VI, VII, and IX). Accurate diagnosis is important to provide disease-specific enzyme replacement therapy. Diagnosis is confirmed through urinary GAG concentration measurement, enzymatic activity measurement or genetic testing.

Key References Accessed 8/2022:

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.
3. 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.
4. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2021; Updated periodically.
5. Valayannopoulos V, Nicely H, Harmatz P, Turbeville S. Mucopolysaccharidosis VI. Orphanet J Rare Dis. 2010; 5:5.
6. 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.
7. 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.

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Date	Summary of Changes
August 2022	Criteria for use summary developed by the Ascension Medical Specialty Prior Authorization Team.
September 2022	Criteria for use summary approved by the Ascension Ambulatory Care Expert Review Panel.
October 2022	Criteria for use summary approved by the Ascension Therapeutic Affinity Group.

If you have questions, call 833-980-2352 to speak to a member of the Ascension Rx prior authorization team, or email your questions to smarthealthspecialty@ascension.org.