

When requesting Naglazyme® (galsulfase), the individual requiring treatment must be diagnosed with the following FDA-approved indication and meet the specific coverage guidelines and applicable safety criteria for the covered indication.

FDA-approved indication¹

Naglazyme® (galsulfase) is indicated for the treatment of Mucopolysaccharidosis VI (MPS VI, Maroteaux-Lamy syndrome).

Approved Off-label Compendial use²

- No off-label uses meet evidence standards

Coverage Guidelines

Mucopolysaccharidosis VI (Maroteaux-Lamy syndrome)

Has a diagnosis of Mucopolysaccharidosis VI (MPS VI, Maroteaux-Lamy syndrome) confirmed by enzyme assay demonstrating a deficiency of N-acetylgalactosamine-4-sulfatase enzyme activity or by genetic testing^{1,3}

Approval duration: 12 months

Additional Information for Prescribers¹

Our prescribers are alerted to the following safety concerns as educational points/reminders:

- Life-threatening anaphylactic reactions have been observed in some Individuals during Naglazyme® (galsulfase) infusions and up to 24 hours after infusion.
- If anaphylaxis or severe allergic reactions occur, immediately discontinue infusion and initiate appropriate treatment, which may include resuscitation, epinephrine, administering additional antihistamines, antipyretics or corticosteroids.
- Immune-mediated reactions can occur with Naglazyme® (galsulfase). Monitor Individuals for the development of immune complex-mediated reactions while receiving Naglazyme® (galsulfase).
- Pretreatment with antihistamines with or without antipyretics is recommended prior to the start of infusion to reduce the risk of infusion reactions. If infusion reactions occur, decreasing the infusion rate, temporarily stopping the infusion, or administering additional antihistamines and/or antipyretics is recommended.
- Sleep apnea is common in MPS VI individuals and antihistamine pretreatment may increase the risk of apneic episodes.
- An infusion-associated reaction (IAR) may not occur until multiple infusions have been given. First IARs occurred as late as 146 weeks in the clinical studies of Naglazyme® (galsulfase).

- There have been post-marketing reports of individuals treated with Naglazyme® (galsulfase) who experienced the onset or worsening of spinal or cervical cord compression requiring decompression surgery. Individuals should be monitored for signs and symptoms of spinal/cervical cord compression and given appropriate clinical care.

References

1. Naglazyme® prescribing information. Novato, CA: BioMarin Pharmaceutical Inc.: Revised March 2013.
2. Klasco RK (Ed): DRUGDEX® system. Thomson Micromedex, Greenwood Village, CO. Accessed February 14, 2017. <https://www.micromedexsolutions.com>
3. Lehman T, Miller N, Norquist B, et al. Diagnosis of the mucopolysaccharidoses. *Rheumatology*. 2011;50:v41-v48.