Clinical Policy Title:	galsulfase
Policy Number:	RxA.233
Drug(s) Applied:	Naglazyme®
Original Policy Date:	02/07/2020
Last Review Date:	06/10/2021
Line of Business Policy Applies to:	All lines of business

Background

Naglazyme® is a hydrolytic lysosomal glycosaminoglycan-specific enzyme indicated for patients with mucopolysaccharidosis VI (MPS VI; Maroteaux-Lamy syndrome). Naglazyme® has been shown to improve walking and stair-climbing capacity.

Dosing Information

Drug Name	Indication	Dosing Regimen	Maximum Dose
galsulfase (Naglazyme®)	MPS VI	1 mg/kg of body weight IV once weekly	1 mg/kg of body weight /week

Dosage Forms

- Single dose vial: 5 mg/5 mL (1 mg/ml).

Clinical Policy

Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria. The provision of provider samples does not guarantee coverage under the provisions of the pharmacy benefit administer by RxAdvance. All criteria for initial approval must be met in order to obtain coverage.

I. Initial Approval Criteria

A. Mucopolysaccharidosis VI: Maroteaux-Lamy Syndrome (must meet all)**:**

1. Diagnosis of MPS VI (Maroteaux-Lamy syndrome) confirmed by one of the following (a or b):
   a. Enzyme assay demonstrating a deficiency in N-acetylgalactosamine 4-sulfatase (arylsulfatase B) activity;
   b. DNA testing;
2. Age ≥ 3 months;
3. Dose does not exceed 1 mg per kg of body weight per week.

Approval Duration
Commercial: 6 months
Medicaid: 6 months

II. Continued Therapy Approval

A. Mucopolysaccharidosis VI: Maroteaux-Lamy Syndrome (must meet all)**:**

1. Member is currently receiving medication that has been authorized by RxAdvance or the member has

met initial approval criteria listed in this policy;
2. Member is responding positively to therapy as evidenced by improvement in the individual member's MPS VI (Maroteaux-Lamy syndrome) manifestation profile (see Appendix D for examples);
3. If request is for a dose increase, new dose does not exceed 1 mg per kg of body weight per week.

Approval Duration
Commercial: 6 months
Medicaid: 12 months

III. Appendices

APPENDIX A: Abbreviation/Acronym Key
FDA: Food and Drug Administration
MPS VI: Mucopolysaccharidosis VI

APPENDIX B: Therapeutic Alternatives
- Not applicable

APPENDIX C: Contraindications/Boxed Warnings
- Contraindication(s):
  - None reported
- Boxed Warning(s):
  - None reported

APPENDIX D: General Information
The presenting symptoms and clinical course of MPS VI can vary from one individual to another. Some examples, however, of improvement in MPS VI disease as a result of Naglazyme® therapy may include improvement in:
- 12-minute walking test distance;
- 3-minute stair climb rate;
- Poor endurance;
- Vision problems;
- Respiratory infections;
- Breathing problems, sleep apnea;
- High blood pressure;
- Joint stiffness;
- Hepatomegaly, splenomegaly.

References
1. Naglazyme® Prescribing Information. Novato, CA: BioMarin Pharmaceutical, Inc.; December, 2019. Available at http://www.naglazyme.com. Accessed February 22, 2021.
2. Muenzer J. The mucopolysaccharidoses: a heterogeneous group of disorders with variable pediatric presentations. J Pediatr. 2004; 144(5 Suppl): S27-S34.

Review/Revision History	Review/Revised Date	P&T Approval Date
Policy established.	01/2020	02/07/2020
Policy was reviewed: 1. Clinical Policy Title was updated.	07/28/2020	09/14/2020

2. Drug(s) Applied was updated. 3. Line of Business Policy Applies to was update to all lines of business. 4. Continued Therapy criteria II.A.1 was rephrased to "Currently receiving medication that has been authorized by RxAdvance..." 5. Initial Approval criteria: Commercial approval duration was updated from member's renewal date to 6 months. 6. 6. Continued Approval criteria: Commercial approval duration was updated from member's renewal date to 6 months. 7. References were updated.		
Policy was reviewed: 1. Last review date was updated. 2. Clinical policy verbiage added " The provision of provider samples does not guarantee…". 3. References were reviewed and updated. 4. Dosing regimen, maximum dose, initial and continued criteria updated to specify: 1 mg/kg of body weight IV once weekly.	02/22/2021	06/10/2021