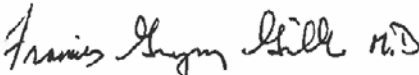


**Prior Authorization Review Panel**

**Prior Authorization Review Panel**

**CHC-MCO Policy Submission**

A separate copy of this form must accompany each policy submitted for review.  
Policies submitted without this form will not be considered for review.

<b>Plan: PA Health &amp; Wellness</b>	<b>Submission Date: 05/01/2020</b>
<b>Policy Number: PA.CP.PHAR.161</b>	<b>Effective Date: 01/2018</b> <b>Revision Date: 04/15/2020</b>
<b>Policy Name: Galsulfase (Naglazyme)</b>	
<p><b>Type of Submission – <u>Check all that apply:</u></b></p> <p> <input type="checkbox"/> <b>New Policy</b>  <input checked="" type="checkbox"/> <b>Revised Policy*</b>  <input type="checkbox"/> <b>Annual Review - No Revisions</b>  <input type="checkbox"/> <b>Statewide PDL</b> - <i>Select this box when submitting policies for Statewide PDL implementation and when submitting policies for drug classes included on the Statewide PDL.</i> </p>	
<p><b>*All revisions to the policy <u>must</u> be highlighted using track changes throughout the document.</b></p> <p><b>Please provide any changes or clarifying information for the policy below:</b></p> <p>2Q 2020 annual review: references reviewed and updated.</p>	
<p><b>Name of Authorized Individual (Please type or print):</b></p> <p><b>Francis G. Grillo, MD</b></p>	<p><b>Signature of Authorized Individual:</b></p> 

## Clinical Policy: Galsulfase (Naglazyme)

Reference Number: PA.CP.PHAR.161

Effective Date: 01/18

Last Review Date: 04/2020

[Coding Implications](#)

[Revision Log](#)

### Description

Galsulfase (Naglazyme®) is a hydrolytic lysosomal glycosaminoglycan-specific enzyme.

### FDA approved indication

Naglazyme is indicated for the treatment of patients with mucopolysaccharidosis VI (MPS VI; Maroteaux-Lamy syndrome). Naglazyme has been shown to improve walking and stair-climbing capacity.

### Policy/Criteria

*Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.*

It is the policy of Pennsylvania Health and Wellness that Naglazyme is **medically necessary** when the following criteria are met:

#### I. Initial Approval Criteria

##### A. Maroteaux-Lamy Syndrome (Mucopolysaccharidosis VI [MPS VI]): (must meet all):

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

**Approval duration: 6 months**

##### B. Other diagnoses/indications: Refer to PA.CP.PMN.53

#### II. Continued Approval

##### A. Maroteaux-Lamy Syndrome (MPS VI): (must meet all):

1. Currently receiving medication via Pennsylvania Health and Wellness benefit or member has previously met all initial approval criteria; or the Continuity of Care policy (PA.LTSS.PHAR.01) applies;
1. 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*);
2. If request is for a dose increase, new dose does not exceed 1 mg/kg/week.

**Approval duration: 12 months**

##### B. Other diagnoses/indications (must meet 1 or 2):

1. Currently receiving medication via Pennsylvania Health and Wellness benefit and documentation supports positive response to therapy; or the Continuity of Care policy (PA.LTSS.PHAR.01) applies; or
2. Refer to PA.CP.PMN.53

### **III. Appendices/General Information**

*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.

### **IV. Dosage and Administration**

<b>Indication</b>	<b>Dosing Regimen</b>	<b>Maximum Dose</b>
MPS VI	1 mg/kg IV once weekly	1 mg/kg/week

### **V. Product Availability**

Vial: 5 mg/5 mL

### **VI. References**

1. Naglazyme Prescribing Information. Novato, CA: BioMarin Pharmaceutical, Inc.; December 2019. Available at <http://www.naglazyme.com>. Accessed February 20,2020.
2. Muenzer J. The mucopolysaccharidoses: a heterogeneous group of disorders with variable pediatric presentations. J Pediatr. 2004; 144(5 Suppl): S27-S34.

3. Akyol MU, Alden TD, Amartino H, et al. Recommendations for the management of MPS VI: systematic evidence- and consensus-based guidance. Orphanet J of Rare Dis, 2019;12(118)1-21.

**Coding Implications**

Codes referenced in this clinical policy are for informational purposes only. Inclusion or exclusion of any codes does not guarantee coverage. Providers should reference the most up-to-date sources of professional coding guidance prior to the submission of claims for reimbursement of covered services.

HCPCS Codes	Description
J1458	Injection, galsulfase, 1 mg

Reviews, Revisions, and Approvals	Date	Approval Date
2Q 2018 annual review: Modified age restriction to 3 months per PI. Added prescriber requirement. Added max dose criteria. Added requirement for positive response to therapy. R eferences reviewed and updated.	02.13.18	04.18
2Q 2019 annual review: references reviewed and updated.	04/2019	
2Q 2020 annual review: references reviewed and updated.	04/2020	