Clinical Policy: Galsulfase (Naglazyme)
Reference Number: CP.PHAR.161
Effective Date: 02.16
Last Review Date: 05.19
Line of Business: Commercial, HIM-Medical Benefit, Medicaid

See Important Reminder at the end of this policy for important regulatory and legal information.

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 health plans affiliated with Centene Corporation® that Naglazyme is medically necessary when the following criteria are met:

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 per week.
   Approval duration:
   Medicaid/HIM – 6 months
   Commercial – 6 months or to the member’s renewal date, whichever is longer

B. Other diagnoses/indications
   1. Refer to the off-label use policy for the relevant line of business if diagnosis is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized): CP.CPA.09 for commercial and CP.PMN.53 for Medicaid and HIM-Medical Benefit.
II. Continued Therapy
   A. Mucopolysaccharidosis VI: Maroteaux-Lamy Syndrome (must meet all):
      1. Currently receiving medication via Centene benefit or member has previously met
         initial approval criteria;
      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 per week.
   Approval duration:
   Medicaid/HIM – 12 months
   Commercial – 6 months or to the member’s renewal date, whichever is longer
   B. Other diagnoses/indications (must meet 1 or 2):
      1. Currently receiving medication via Centene benefit and documentation supports
         positive response to therapy.
         Approval duration: Duration of request or 6 months (whichever is less);
      2. Refer to the off-label use policy for the relevant line of business if diagnosis is NOT
         specifically listed under section III (Diagnoses/Indications for which coverage is
         NOT authorized): CP.CPA.09 for commercial and CP.PMN.53 for Medicaid and
         HIM-Medical Benefit.

III. Diagnoses/Indications for which coverage is NOT authorized:
   A. Non-FDA approved indications, which are not addressed in this policy, unless there is
      sufficient documentation of efficacy and safety according to the off label use policy –
      CP.CPA.09 for commercial and CP.PMN.53 for Medicaid and HIM-Medical Benefit
      or evidence of coverage documents.

IV. 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;
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- Vision problems;
- Respiratory infections;
- Breathing problems, sleep apnea;
- High blood pressure;
- Joint stiffness;
- Hepatomegaly, splenomegaly.

V. Dosage and Administration

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dosing Regimen</th>
<th>Maximum Dose</th>
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<tbody>
<tr>
<td>MPS VI</td>
<td>1 mg/kg IV once weekly</td>
<td>1 mg/kg/week</td>
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</table>

VI. Product Availability
Vial: 5 mg/5 mL

VII. References

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.

<table>
<thead>
<tr>
<th>HCPCS Codes</th>
<th>Description</th>
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<tbody>
<tr>
<td>J1458</td>
<td>Injection, galsulfase, 1 mg</td>
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Reviews, Revisions, and Approvals

<table>
<thead>
<tr>
<th>Description</th>
<th>Date</th>
<th>P&amp;T Approval Date</th>
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<tbody>
<tr>
<td>Policy split from USS.SPMN.33 Lysosomal Storage Disorders and converted to new template. Added age restriction per PI. Modified approval duration to 6 months for initial and 12 months for re-auth.</td>
<td>08.16</td>
<td>09.16</td>
</tr>
<tr>
<td>Modified age restriction to 3 months per PI. Added prescriber requirement. Added max dose criteria. Added requirement for positive response to therapy.</td>
<td>06.17</td>
<td>08.17</td>
</tr>
<tr>
<td>Policy converted to newer template. Age restriction added. Added appendix B.</td>
<td>09.05.17</td>
<td>11.17</td>
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<tr>
<td>2Q 2018 annual review: no significant changes from previously approved corporate policy; policies combined for Commercial and Medicaid lines of business; HIM added; Commercial: added diagnosis confirmation testing requirement; added age limit; added specific</td>
<td>02.26.18</td>
<td>05.18</td>
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## Important Reminder

This clinical policy has been developed by appropriately experienced and licensed health care professionals based on a review and consideration of currently available generally accepted standards of medical practice; peer-reviewed medical literature; government agency/program approval status; evidence-based guidelines and positions of leading national health professional organizations; views of physicians practicing in relevant clinical areas affected by this clinical policy; and other available clinical information. The Health Plan makes no representations and accepts no liability with respect to the content of any external information used or relied upon in developing this clinical policy. This clinical policy is consistent with standards of medical practice current at the time that this clinical policy was approved. “Health Plan” means a health plan that has adopted this clinical policy and that is operated or administered, in whole or in part, by Centene Management Company, LLC, or any of such health plan’s affiliates, as applicable.

The purpose of this clinical policy is to provide a guide to medical necessity, which is a component of the guidelines used to assist in making coverage decisions and administering benefits. It does not constitute a contract or guarantee regarding payment or results. Coverage decisions and the administration of benefits are subject to all terms, conditions, exclusions and limitations of the coverage documents (e.g., evidence of coverage, certificate of coverage, policy, contract of insurance, etc.), as well as to state and federal requirements and applicable Health Plan-level administrative policies and procedures.

This clinical policy is effective as of the date determined by the Health Plan. The date of posting may not be the effective date of this clinical policy. This clinical policy may be subject to applicable legal and regulatory requirements relating to provider notification. If there is a discrepancy between the effective date of this clinical policy and any applicable legal or regulatory requirement, the requirements of law and regulation shall govern. The Health Plan retains the right to change, amend or withdraw this clinical policy, and additional clinical policies may be developed and adopted as needed, at any time.

This clinical policy does not constitute medical advice, medical treatment or medical care. It is not intended to dictate to providers how to practice medicine. Providers are expected to exercise professional medical judgment in providing the most appropriate care, and are solely responsible for the medical advice and treatment of members. This clinical policy is not intended to recommend treatment for members. Members should consult with their treating physician in connection with diagnosis and treatment decisions.

### Reviews, Revisions, and Approvals

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<tr>
<td>examples of positive response to therapy for reauthorization; changed approval durations from length of benefit to 6/12 months; references reviewed and updated.</td>
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<tr>
<td>2Q 2019 annual review: no significant changes; references reviewed and updated.</td>
<td>02.28.19</td>
<td>05.19</td>
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Note: For Medicaid members, when state Medicaid coverage provisions conflict with the coverage provisions in this clinical policy, state Medicaid coverage provisions take precedence. Please refer to the state Medicaid manual for any coverage provisions pertaining to this clinical policy.

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