### Medical Necessity Guidelines:

**Cerezyme® (imiglucerase), Elelyso® (taliglucerase alfa), and VPRIV® (velaglucerase alfa)**

*Effective: January 1, 2023*

#### Prior Authorization Required

If **REQUIRED**, submit supporting clinical documentation pertinent to service request.

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<thead>
<tr>
<th>Applies to:</th>
<th>Yes ☒ No ☐</th>
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<tr>
<td><strong>Commercial Products</strong></td>
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<tr>
<td>☒ Harvard Pilgrim Health Care Commercial products; Fax 617-673-0988</td>
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<tr>
<td>☒ Tufts Health Plan Commercial products; Fax 617-673-0988</td>
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<tr>
<td>☒ CareLink® – Refer to CareLink Procedures, Services and Items Requiring Prior Authorization</td>
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<td><strong>Public Plans Products</strong></td>
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<tr>
<td>☒ Tufts Health Direct – A Massachusetts Qualified Health Plan (QHP) (a commercial product); Fax 617-673-0988</td>
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<td>☐ Tufts Health Together – MassHealth MCO Plan and Accountable Care Partnership Plans; Fax 617-673-0988</td>
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<tr>
<td>☐ Tufts Health RITogether – A Rhode Island Medicaid Plan; Fax 617-673-0988</td>
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<tr>
<td>☐ Tufts Health Unify® – OneCare Plan (a dual-eligible product); Fax 617-673-0956</td>
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<td><em>The MNG applies to Tufts Health Unify members unless a less restrictive LCD or NCD exists.</em></td>
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<td><strong>Senior Products</strong></td>
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<td>☐ Harvard Pilgrim Health Care Stride Medicare Advantage; Fax 617-673-0956</td>
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<tr>
<td>☐ Tufts Health Plan Senior Care Options (SCO), (a dual-eligible product); Fax 617-673-0956</td>
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<td>☐ Tufts Medicare Preferred HMO, (a Medicare Advantage product); Fax 617-673-0956</td>
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<td>☐ Tufts Medicare Preferred PPO, (a Medicare Advantage product); Fax 617-673-0956</td>
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**Note:** While you may not be the provider responsible for obtaining prior authorization, as a condition of payment you will need to ensure that prior authorization has been obtained.

### Overview

Gaucher disease (GD) is a rare and debilitating genetic disorder in which patients lack the enzyme b-glucocerebrosidase, which is essential for the proper lipid metabolism. As a result of this missing enzyme, there is a build-up of the glycolipid glucocerebroside, which can cause a host of problems, most importantly, hepatomegaly (enlarged liver), splenomegaly (enlarged spleen), bone disease and severe anemia (low blood counts). The mainstay of treatment for this disease focuses on replacing the missing enzyme, which provides some relief, but is not a cure.

Type 1 GD (GD1) is the most prevalent type in the United States, Europe, and Israel and occurs with greater frequency in the Ashkenazi Jewish population. In the United States, Europe, and Israel, approximately 90 percent of patients have GD1. GD1 is characterized by variability in signs, symptoms, severity, and progression, even among siblings with the same genotype and monozygotic twins. Symptomatic patients have visceral involvement, bone disease, and bleeding. Fatigue is common, and pubertal delay with associated delay in growth may occur. Bone disease is common in all patients, especially those who have undergone splenectomy. Variability is described in individuals homozygous for the c.1226A>G allele (p.N409S or N370S variant), ranging from clinically significant anemia, thrombocytopenia, hepatosplenomegaly, marrow infiltration, bony abnormalities, and osteopenia/osteoporosis, to essentially asymptomatic with no or mild hematologic and skeletal findings on examination.

### Food and Drug Administration (FDA) Approved Indications:
- **Cerezyme** (imiglucerase for injection) is indicated for long-term enzyme replacement therapy for pediatric and adult patients with a confirmed diagnosis of Type 1 Gaucher disease that results in one or more of the following conditions: anemia, thrombocytopenia, bone disease, or hepatomegaly or splenomegaly.
- **Elelyso** (taliglucerase alfa) for injection is a hydrolytic lysosomal glucocerebroside-specific enzyme indicated for the treatment of patients 4 years and older with a confirmed diagnosis of Type 1 Gaucher disease.
- **VPRIV** (velaglucerase alfa) is a hydrolytic lysosomal glucocerebroside-specific enzyme indicated for long-term enzyme replacement therapy for patients with type 1 Gaucher disease.

### Clinical Guideline Coverage Criteria

The Plan may cover Cerezyme (imiglucerase), Elelyso (taliglucerase alfa) or VPRIV (velaglucerase alfa) when all the following clinical criteria are met:

1. Documented diagnosis of Type 1 Gaucher disease
   AND
2. Documentation of one of the following:
   a. For Cerezyme, the Member is at least 2 years of age
   b. For Elelyso, the Member is at least 4 years of age
   c. For VPRIV, the Member is at least 4 years of age
   AND
3. Documentation of at least one of the following conditions:
   a. Anemia
   b. Bone disease
   c. Hepatomegaly or splenomegaly
   d. Thrombocytopenia
   AND
4. For Cerezyme requests only, documentation the Member has demonstrated an inadequate response to Elelyso or VPRIV or the provider has indicated clinical inappropriateness of treatment with Elelyso or VPRIV therapy or the member is age 2 or 3.

### Limitations

- The Plan does not cover enzyme replacement therapy for Type 2 or Type 3 Gaucher Disease.
- Coverage will be limited to FDA-approved dosing regimens.

### Codes

The following code(s) require prior authorization:

### Table 1: HCPCS Codes

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<tr>
<th>HCPCS Codes</th>
<th>Description</th>
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<tr>
<td>J1786</td>
<td>Injection, imiglucerase, 10 units</td>
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<tr>
<td>J3060</td>
<td>Injection, taliglucerase alfa, 10 units</td>
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<tr>
<td>J3358</td>
<td>Injection, velaglucerase alfa, 100 units</td>
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### References:


Approval And Revision History
- September 13, 2022: Reviewed by Pharmacy and Therapeutics Committee (P&T)
- September 21, 2022: Reviewed by the Medical Policy Approval Committee (MPAC)

Background, Product and Disclaimer Information

Medical Necessity Guidelines are developed to determine coverage for benefits and are published to provide a better understanding of the basis upon which coverage decisions are made. We make coverage decisions using these guidelines, along with the Member’s benefit document, and in coordination with the Member’s physician(s) on a case-by-case basis considering the individual Member’s health care needs.

Medical Necessity Guidelines are developed for selected therapeutic or diagnostic services found to be safe and proven effective in a limited, defined population of patients or clinical circumstances. They include concise clinical coverage criteria based on current literature review, consultation with practicing physicians in our service area who are medical experts in the particular field, FDA and other government agency policies, and standards adopted by national accreditation organizations. We revise and update Medical Necessity Guidelines annually, or more frequently if new evidence becomes available that suggests needed revisions.

For self-insured plans, coverage may vary depending on the terms of the benefit document. If a discrepancy exists between a Medical Necessity Guideline and a self-insured Member’s benefit document, the provisions of the benefit document will govern. For Tufts Health Together (Medicaid), coverage may be available beyond these guidelines for pediatric members under age 21 under the Early and Periodic Screening, Diagnostic and Treatment (EPSDT) benefits of the plan in accordance with 130 CMR 450.140 and 130 CMR 447.000, and with prior authorization.

Treating providers are solely responsible for the medical advice and treatment of Members. The use of this guideline is not a guarantee of payment or a final prediction of how specific claim(s) will be adjudicated. Claims payment is subject to eligibility and benefits on the date of service, coordination of benefits, referral/authorization, utilization management guidelines when applicable, and adherence to plan policies, plan procedures, and claims editing logic.