Medical Guideline Disclaimer

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I. Length of Authorization

Coverage will be for 12 months and may be renewed.

II. Dosing Limits

A. Max Units (per dose and over time) [Medical Benefit]:

- 700 billable units every 14 days

III. Initial Approval Criteria

Type 1 Gaucher Disease†

- Patient age at least 2 years or older; AND
- Patient has a documented diagnosis of Type 1 Gaucher Disease as confirmed by reduced glucocerebrosidase activity in peripheral leukocytes; AND
- Adults only criteria (patient at least 18 years or older): Patient’s disease results in one or more of the following:
  - Anemia [hemoglobin less than or equal to 11 g/dL (women) or 12 g/dL (men)]; OR
  - Moderate to severe hepatomegaly (liver size 1.25 or more times normal volume) or splenomegaly (spleen size 5 or more times normal volume); OR
- Skeletal disease (e.g. lesions, remodeling defects and/or deformity of long bones, osteopenia/osteoporosis, etc.); OR
- Symptomatic disease (e.g. bone pain, fatigue, dyspnea, angina, abdominal distension, diminished quality of life, etc.); OR
- Thrombocytopenia (platelet count less than or equal to 120,000/mm³).

- Must be used as a single agent

† FDA Approved Indication(s)

IV. Renewal Criteria

- Patient continues to meet the criteria in Section III; AND
- Disease response as indicated by one or more of the following (compared to pre-treatment baseline):
  - Improvement in symptoms (e.g. bone pain, fatigue, dyspnea, angina, abdominal distension, diminished quality of life, etc.)
  - Reduction in size of liver or spleen
  - Improvement in hemoglobin/anemia
  - Improvement in skeletal disease
  - Improvement in platelet counts; AND
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include hypersensitivity reactions.

V. Dosage/Administration

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
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| Type 1 Gaucher Disease      | - Initial dosages range from 2.5 U/kg of body weight 3 times a week to 60 U/kg once every 2 weeks based on disease severity. Cerezyme is administered by intravenous infusion over 1–2 hours.  
- Dosage adjustments should be made on an individual basis and may increase or decrease, based on achievement of therapeutic goals as assessed by routine comprehensive evaluations of the patient’s clinical manifestations. |

Limitations/Exclusions

Cerezyme® (imiglucerase) is not considered medically necessary for indications other than those listed above due to insufficient evidence of therapeutic value.
Applicable Procedure Codes

<table>
<thead>
<tr>
<th>Procedure Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>J1786</td>
<td>Injection, imiglucerase, 10 units</td>
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Applicable NDCs

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<tr>
<th>NDC Details</th>
<th>Description</th>
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<tr>
<td>58468-1983-xx</td>
<td>Cerezyme 200 unit injection</td>
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<tr>
<td>58468-4663-xx</td>
<td>Cerezyme 400 unit injection</td>
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Applicable Diagnosis Codes

<table>
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<tr>
<th>Diagnosis Code</th>
<th>Description</th>
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<tr>
<td>E75.22</td>
<td>Gaucher disease</td>
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Revision History

VI. References