Definition

Velaglucerase alfa for injection is indicated for long-term enzyme replacement therapy (ERT) in adults and pediatric patients 4 years and older with type 1 Gaucher disease. Clinical studies have demonstrated the efficacy of velaglucerase alfa in non-neuropathic Gaucher disease as initial treatment among patients with no prior ERT exposure and among patients switching from imiglucerase to velaglucerase alfa.

Related Medical Guideline

Off-Label Use of FDA-Approved Drugs and Biologicals

LENGTH OF AUTHORIZATION

Coverage will be for 12 months and may be renewed.

DOSING LIMITS

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

- 72 billable units every 14 days

Guideline

I. INITIAL APPROVAL CRITERIA

Type 1 Gaucher’s Disease †

- Patient age at least 4 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) or splenomegaly (spleen size 5 or more times normal); 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 distention, diminished quality of life, etc.); OR
  - Thrombocytopenia (platelet count less than or equal to 120,000/mm3); AND

• **Must be used as a single agent**

† FDA Approved Indication(s)

II. **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 severe hypersensitivity reactions, etc.

**Limitations/Exclusions**

Vpriv is not considered medically necessary for indications other than those listed above due to insufficient evidence of therapeutic value.

**Applicable Procedure Codes**

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<thead>
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<th>Description</th>
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<tbody>
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<td>J3385</td>
<td>Injection, velaglucerase alfa, up to 60 units</td>
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**Applicable NDCs**

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<td>54092-0701-XX</td>
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**Applicable Diagnosis Codes**

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<td>E75.22</td>
<td>Gaucher disease</td>
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Revision History

N/A

References