Policy Title: Cerezyme (imiglucerase), Elelyso (taliglucerase alfa), VPRIV (velaglucerase alfa) Intravenous

Department: PHA

Effective Date: 01/01/2020

Review Date: 04/19/2019, 9/18/2019, 12/18/19, 1/29/20, 2/04/2021

Revision Date: 04/19/2019, 9/18/2019, 1/29/20, 2/04/2021

Purpose: To support safe, effective and appropriate use of Cerezyme (imiglucerase), Elelyso (taliglucerase alfa), and VPRIV (velaglucerase alfa) to treat Gaucher’s disease.

Scope: Medicaid, Commercial, Medicare-Medicaid Plan (MMP)

Policy Statement:
Medications to treat Gaucher’s disease are covered under the Medical Benefit when used within the following guidelines. Use outside of these guidelines may result in non-payment unless approved under an exception process.

Procedure:
Coverage of Cerezyme (imiglucerase), Elelyso (taliglucerase alfa), and VPRIV (velaglucerase alfa) will be reviewed prospectively via the prior authorization process based on criteria below.

Coverage Criteria:
- Patient must have a confirmed diagnosis of type 1 Gaucher disease (GD1) when the diagnosis of Gaucher disease was confirmed by enzyme assay demonstrating a deficiency of beta-glucocerebrosidase (glucosidase) enzyme activity or by genetic testing; AND
- Requests for Elelyso (taliglucerase alfa) or VPRIV (velaglucerase alfa) must have a documented failure, intolerance or contraindication to Cerezyme (imiglucerase); OR
- Patients that are currently on treatment with Elelyso (taliglucerase alfa) or VPRIV (velaglucerase alfa) can remain on treatment; OR
- MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements

Continuation of Therapy Criteria:
- The patient meets all initial criteria; AND
- Patient is tolerating and responding to medication (improvement in symptoms compared to pre-treatment baseline, such as e.g. bone pain, fatigue, dyspnea, angina, abdominal distension, diminished quality of life, etc.) and there continues to be a medical need for the medication.
Coverage duration: 6 months

*** Requests will also be reviewed to National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) if applicable. ***

Dosage/Administration:

Cerezyme:

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
<th>Maximum dose (1 billable unit = 10 units)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type 1 Gaucher Disease</td>
<td>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.</td>
<td>700 billable units every 14 days</td>
</tr>
</tbody>
</table>

Elelyso:

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
<th>Maximum dose (1 billable unit = 10 units)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type 1 Gaucher Disease</td>
<td>Up to 60 units/kg every other week as a 60-120-minute intravenous infusion</td>
<td>700 billable units every 14 days</td>
</tr>
</tbody>
</table>

VPRIV:

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
<th>Maximum dose (1 billable unit = 10 units)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type 1 Gaucher Disease</td>
<td>Up to 60 units/kg every other week as a 60-minute intravenous infusion</td>
<td>72 billable units every 14 days</td>
</tr>
</tbody>
</table>

Investigational use: All therapies are considered investigational when used at a dose or for a condition other than those that are recognized as medically accepted indications as defined in any one of the following standard reference compendia: American Hospital Formulary Service Drug information (AHFS-DI), Thomson Micromedex DrugDex, Clinical Pharmacology, Wolters Kluwer Lexi-Drugs, or Peer-reviewed published medical literature indicating that sufficient evidence exists to support use. Neighborhood does not provide coverage for drugs when used for investigational purposes.

Applicable Codes:
Below is a list of billing codes applicable for covered treatment options. The below tables are provided for reference purposes and may not be all-inclusive. Requests received with codes from tables below do not guarantee coverage. Requests must meet all criteria provided in the procedure section.

The following HCPCS/CPT codes are:

<table>
<thead>
<tr>
<th>HCPCS/CPT Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>J1786</td>
<td>Injection, imiglucerase, 10 units</td>
</tr>
<tr>
<td>J3060</td>
<td>Injection, taliglucerase alfa, 10 units</td>
</tr>
<tr>
<td>J3385</td>
<td>Injection, velaglucerase alfa, 100 units</td>
</tr>
</tbody>
</table>

References: