Policy Title: Kanuma (sebelipase alfa) (Intravenous)

Policy Number:  
Department: PHA

Effective Date: 01/01/2020

Review Date: 09/25/2019, 1/15/2020, 4/15/2021

Revision Date: 09/25/2019, 1/15/2020, 4/15/2021

Purpose: To support safe, effective and appropriate use of Kanuma (sebelipase alfa).

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

Policy Statement:

Kanuma (sebelipase alfa) is 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 Kanuma (sebelipase alfa) will be reviewed prospectively via the prior authorization process based on criteria below.

Initial Criteria:

Lysosomal Acid Lipase (LAL) deficiency

- Diagnosis has been confirmed by enzyme assay demonstrating a deficiency of lysosomal acid lipase enzyme activity or genetic testing revealing mutation in the lipase A, lysosomal acid type (LIPA) gene; AND
- Patient is at least 1 month old; AND
- Prescribing physician is a specialist in genetics and metabolism; AND
- Weight, baseline liver function and baseline lipid panel is provided;
- MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements.

Continuation of Therapy Criteria:

- Patient continues to meet initial criteria; AND
- Patient is tolerating treatment; AND
- Recent weight, liver function and lipid panel after treatment initiation is provided; AND
• Documented clinical benefit with therapy (e.g. reduction in LDL, triglycerides, AST or ALT, increase in HDL, improvement in weight-for-age z-scores, reduction in liver fat content)

Coverage durations:
• Initial coverage: 6 months
• Continuation of therapy coverage: 6 months

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

Dosage/Administration:

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
<th>Maximum dose (1 billable unit = 1 mg)</th>
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</thead>
<tbody>
<tr>
<td>LAL deficiency</td>
<td>Pediatric &amp; Adult patients:</td>
<td>340 billable units once weekly</td>
</tr>
<tr>
<td></td>
<td>• 1 mg/kg administered once every other week as an IV infusion</td>
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<td></td>
<td>Rapidly progressive disease presenting within the first 6 months of life:</td>
<td></td>
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<tr>
<td></td>
<td>• 1 mg/kg administered once weekly as an IV infusion</td>
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<tr>
<td></td>
<td>• May increase to 3 mg/kg once weekly for patients who do not achieve an optimal clinical response</td>
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</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 code is:

<table>
<thead>
<tr>
<th>HCPCS/CPT Code</th>
<th>Description</th>
</tr>
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<tbody>
<tr>
<td>J2840</td>
<td>Injection, sebelipase alfa, 1mg</td>
</tr>
</tbody>
</table>

References: