Mepsevii® (vestronidase alfa-vjbk) (Intravenous)

Last Review Date: January 1, 2020
Number: MG.MM.PH.122

Medical Guideline Disclaimer

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Definition

Mepsevii is a recombinant human lysosomal beta glucuronidase indicated in pediatric and adult patients for the treatment of Mucopolysaccharidosis VII.

Length of Authorization

Coverage will be provided for 6 months and may be renewed.

Dosing Limits

Max Units (per dose and over time) [Medical Benefit]:
- 460 mg every 14 days

Guideline

I. INITIAL APPROVAL CRITERIA

Mepsevii may be considered medically necessary if one of the below conditions are met AND use is consistent with the medical necessity criteria that follows:

Mucopolysaccharidosis VII†
- Patient has a definitive diagnosis of MPS VII confirmed by BOTH of the following:
  - Beta-glucuronidase enzyme deficiency in peripheral blood leukocytes; AND
  - Detection of pathogenic mutations in the GUSB gene by molecular genetic testing; AND
- Patient age is 5 months or older; AND
Documented baseline value for one or more of the following: six minute walk test (6MWT), motor function [i.e., Bruininks-Oseretsky Test of Motor Proficiency (BOT-2)], liver and/or spleen volume, urinary excretion of glycosaminoglycans (GAGs) such as chondroitin sulfate and dermatan sulfate, skeletal involvement, pulmonary function tests, etc.

† FDA-labeled indication(s)

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

II. RENEWAL CRITERIA

- Patient continues to meet the Initial Approval Criteria; AND
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include the following: anaphylaxis and severe allergic reactions, etc.; AND
- Patient has responded to therapy compared to pretreatment baseline in one or more of the following:
  - Stability or improvement in 6MWT and/or motor function
  - Reduction in liver and/or spleen volume
  - Reduction in urinary excretion of GAGs
  - Stability of skeletal disease
  - Stability or improvement in pulmonary function tests

Dosage/Administration

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<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
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<tr>
<td>Mucopolysaccharidosis VII</td>
<td>4 mg/kg of body weight administered as an intravenous infusion over approximately 4 hours once every 2 weeks</td>
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Applicable Procedure Codes

| J3397 | Injection, vestronidase alfa-vjbk, 1mg |

Applicable NDCs

| 69794-0001-XX | Mepsevii 10 mg/5 mL single-dose vial |

Applicable Diagnosis Codes

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<thead>
<tr>
<th>ICD-10</th>
<th>ICD-10 Description</th>
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<tr>
<td>E76.29</td>
<td>Other mucopolysaccharidoses</td>
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References