Definition

Mepsevii® (Vestronidase Alfa-vjbk) (Intravenous)

Last Review Date: January 1, 2019
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

<table>
<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

- J397 Injection, vestronidase alfa-vjbk, 1mg

Applicable NDCs

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

Applicable Diagnosis Codes

- ICD-10 E76.29 Other mucopolysaccharidoses

References