Mepsevii® (Vestronidase Alfa-vjbk) (Intravenous)

Last Review Date: January 1, 2019	Number: MG.MM.PH.122

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

Indication	Dose
Mucopolysaccharidosis VII	4 mg/kg of body weight administered as an intravenous infusion over approximately 4 hours once every 2 weeks

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

ICD-10	ICD-10 Description
E76.29	Other mucopolysaccharidoses

References

1. Mepsevii [package insert]. Novato, CA; Ultragenyx Pharmaceutical Inc.; November 2017.
2. Montaño AM, Lock-Hock N, Steiner RD, et al. Clinical course of sly syndrome (mucopolysaccharidosis type VII). J Med Genet. 2016 Jun;53(6):403-18.