Policy Title:	Mepsevii (vestronidase alfa-vjbk) (Intravenous)		
		Department:	PHA
Effective Date:	01/01/2020		
Review Date:	01/22/2020, 5/27/2021, 02/17/2022		

Purpose: To support safe, effective and appropriate use of Mepsevii (vestronidase alfa-vjbk).

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

Policy Statement:

Mepsevii 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 Mepsevii will be reviewed prospectively via the prior authorization process based on criteria below.

Initial Criteria:

Authorization may be granted for treatment of Mucopolysaccharidosis VII (MPS VII or Sly syndrome) when all of the following criteria are met:

- 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 aged 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, shoulder flexion, visual acuity, etc.;
- MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements

Continuation of Therapy Criteria:

Authorizations may be renewed when all of the following criteria are met:

- Patient continues to meet all initial criteria; AND

- Patient is tolerating treatment with 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, shoulder flexion, visual acuity, and/or other motor functions
    - Reduction in liver and/or spleen volume
    - Reduction in urinary excretion of GAGs
    - Stability of skeletal disease
    - Stability or improvement in pulmonary function tests

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:

Indication	**Dose**	**Maximum dose (1 billable unit = 1 mg)**
Mucopolysaccharidosis VII (Sly Syndrome)	4 mg/kg administered every two weeks as an intravenous infusion	460 billable units every 14 days

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:

HCPCS/CPT Code	Description
J3397	Injection, vestronidase alfa-vjbk, 1 mg

References:

1. Mepsevii [package insert]. Novato, CA; Ultragenyx Pharmaceutical Inc.; December 2020. Accessed February 2022.