

Policy: MBP 175.0

Section: Medical Benefit Pharmaceutical Policy

Subject: Mepsevii (vestronidase alfa-vjkb)

I. Policy:

Mepsevii (vestronidase alfa-vjkb)

II. Purpose/Objective:

To provide a policy of coverage regarding Mepsevii (vestronidase alfa-vjkb)

III. Responsibility:

- A. Medical Directors
- B. Medical Management
- C. Pharmacy Department

IV. Required Definitions

1. Attachment – a supporting document that is developed and maintained by the policy writer or department requiring/authoring the policy.
2. Exhibit – a supporting document developed and maintained in a department other than
3. the department requiring/authoring the policy.
4. Devised – the date the policy was implemented.
5. Revised – the date of every revision to the policy, including typographical and grammatical changes.
6. Reviewed – the date documenting the annual review if the policy has no revisions necessary.

V. Additional Definitions

Medical Necessity or Medically Necessary means Covered Services rendered by a Health Care Provider that the Plan determines are:

- a. appropriate for the symptoms and diagnosis or treatment of the Member's condition, illness, disease or injury;
- b. provided for the diagnosis and the direct care and treatment of the Member's condition, illness disease or injury;
- c. in accordance with current standards good medical treatment practiced by the general medical community;
- d. not primarily for the convenience of the Member, or the Member's Health Care Provider; and
- e. the most appropriate source or level of service that can safely be provided to the Member. When applied to hospitalization, this further means that the Member requires acute care as an inpatient due to the nature of the services rendered or the Member's condition, and the Member cannot receive safe or adequate care as an outpatient

Medicaid Business Segment

Medically Necessary — A service, item, procedure, or level of care compensable under the Medical Assistance program that is necessary for the proper treatment or management of an illness, injury, or disability is one that:

- i. Will, or is reasonably expected to, prevent the onset of an illness, condition, injury or disability.
- ii. Will, or is reasonably expected to, reduce or ameliorate the physical, mental or developmental effects of an illness, condition, injury or disability.
- iii. Will assist the Member to achieve or maintain maximum functional capacity in performing daily activities, taking into account both the functional capacity of the Member and those functional capacities that are appropriate for Members of the same age.

DESCRIPTION:

Mepsevii (vestronidase alfa-vjvk) is a recombinant human beta-glucuronidase (GUS), which provides exogenous GUS enzyme for uptake into cellular lysosomes. Mannose-6-phosphate (M6P) residues on the oligosaccharide chains allow binding of the enzyme to cell surface receptors, leading to cellular uptake of the enzyme, targeting to lysosomes and subsequent catabolism of accumulated glycosaminoglycans (GAGs) in affected tissues.

CRITERIA FOR USE: Requires Prior Authorization by Medical Director or Designee

Mepsevii (vestronidase alfa-vjvk) will be considered medically for all lines of business necessary when ALL of the following criteria are met:

- Prescribed by or in consultation with a specialist in genetic disorders OR metabolic disorders **AND**
 - Medical record documentation of a diagnosis of Mucopolysaccharidosis VII (MPS VII, Sly syndrome) confirmed by ALL of the following:
 - Elevated urinary glycosaminoglycans (GAGs) at least three times the upper limit of normal (3xULN) **AND**
 - Enzyme activity assay (beta-glucuronidase deficiency) OR genetic testing (mutation of chromosome 7q21.11) **AND**
 - At least one of the following clinical signs or symptoms: enlarged liver and spleen, joint limitations, airway obstructions or pulmonary dysfunction
- AND**
- Medical record documentation of a baseline evaluation, including a standardized assessment of motor function (e.g. 6-minute walk test (6MWT)), urinary GAGs level, and pulmonary function test (PFT)

Note: Some patients may only have elevated GAGs two times the upper limit of normal (2xULN). Elevated GAGs and two mutations consistent with MPS VII are appropriate to diagnosis patients with MPS VII when diagnosed through newborn screening or sibling screening.

AUTHORIZATION DURATION: If determined to be medically necessary, Mepsevii should be approved for an initial authorization duration of **6 months**. Subsequent authorizations of Mepsevii should be approved for an authorization duration of **12 months** when the following criteria are met.

- Medical record documentation of improvement or maintenance of motor function, urinary GAGs level, pulmonary function, or other clinical signs/symptoms (i.e. decreased liver/spleen size, improvement in joint function, etc.)

Note: For Medicaid (GHP Family), any requests for services that do not meet criteria set in the PARP will be evaluated on a case-by-case basis.

LINE OF BUSINESS:

Eligibility and contract specific benefit limitations and/or exclusions will apply. Coverage statements found in the line of business specific benefit document will supersede this policy.

This policy will be revised as necessary and reviewed no less than annually.

Devised: 5/15/18

Revised: 12/17/22 (LOB carve out, Medicaid PARP statement), 12/5/23 (Medicaid business segment)

Reviewed: 4/22/19, 1/1/20, 1/1/21, 12/17/21

MA UM Committee approval: 12/31/23