# Mepsevii (vestronidase alfa)

| Override(s) | Approval Duration |
|---|---|
| Prior Authorization | Initial therapy: 6 months<br>Continuation therapy: 6 months |

| Medications | Dosing Limit |
|---|---|
| Mepsevii (vestronidase alfa-vjbk) 10 mg vial | 4 mg/kg once every 2 weeks |

## APPROVAL CRITERIA

Initial requests for Mepsevii (vestronidase alfa-vjbk) may be approved if the following criteria are met:

I. Individual has a diagnosis of Mucopolysaccharidosis type VII (Sly syndrome); **AND**
II. Documentation is provided that diagnosis is demonstrated by (NCT 02230566; Lehman 2011):
    A. Deficiency in beta-glucuronidase enzyme activity as measured in fibroblasts or leukocytes; **OR**
    B. GUSB gene mutation; **AND**
III. Documentation is provided that elevated urine glycosaminoglycans excretion is at a minimum of 3-fold over the mean normal for age at screening (NCT 02230566).

Continuation requests for Mepsevii (vestronidase alfa-vjbk) may be approved if the following criteria are met:

I. Documentation is provided to show clinically significant improvement or stabilization in clinical signs and symptoms of disease (including but not limited to reduction in urinary GAG excretion, reduction in hepatosplenomegaly, improvement in pulmonary function, improvement in walking distance and/or improvement in fine or gross motor function) compared to the predicted natural history trajectory of disease (NCT 02230566).

Mepsevii (vestronidase alfa) may not be approved when the above criteria are not met and for all other indications.

**Note:**
Mepsevii has a black box warning for anaphylaxis. Life-threatening anaphylactic reactions have occurred during Mepsevii infusions so appropriate medical support should be available during Mepsevii administration. The Mepsevii infusion should be discontinued if the individual experiences anaphylaxis, and individuals should be observed for 60 minutes after Mepsevii infusion.

**Key References**:


1. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: September 9, 2023.
2. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
3. Lehman TJ, Miller N, Norquist B, Underhill L, Keutzer J. Diagnosis of the mucopolysaccharidoses. *Rheumatology (Oxford)*. 2011;50 Suppl 5:v41-v48. doi:10.1093/rheumatology/ker390.
4. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc. Updated periodically.
5. Ultragenyx Pharmaceutical Inc. A phase 3 study of UX003 rhGUS enzyme replacement therapy in patients with MPS 7. NLM Identifier: NCT 02230566. Last updated: July 30, 2020. Available at: https://clinicaltrials.gov/ct2/show/NCT02230566?term=Recombinant+Human+Beta-glucuronidase&rank=3. Accessed: September 9, 2023.


Federal and state laws or requirements, contract language, and Plan utilization management programs or polices may take precedence over the application of this clinical criteria.