Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	X

Mepsevii (vestronidase alfa)

Override(s)	Approval Duration
Prior Authorization	Initial therapy: 6 months 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 of diagnosis based on leukocyte or fibroblast glucuronidase enzyme assay OR genetic testing (NCT 02230566, Lehman 2011); **AND**
III. Elevated urine glycosaminoglycans excretion 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. There is documentation of 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.

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	X

State Specific Mandates		
State name N/A	Date effective N/A	Mandate details (including specific bill if applicable) N/A

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: August 31, 2019.
2. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
3. Lehman TJ, Miller N, Norquist B, et al. Diagnosis of the mucopolysaccharidoses. Rheumatology. 2011; 50:V41-V46.
4. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2019; 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: January 4, 2019. Available at: https://clinicaltrials.gov/ct2/show/NCT02230566?term=Recombinant+Human+Beta-glucuronidase&rank=3. Accessed: August 31, 2019.

This policy does not apply to health plans or member categories that do not have pharmacy benefits, nor does it apply to Medicare. Note that market specific restrictions or transition-of-care benefit limitations may apply.