

MEPSEVII™ (vestronidase alfa-vjbk) Injection

When requesting MEPSEVII™ (vestronidase alfa-vjbk), the individual requiring treatment must be diagnosed with an FDA-approved indication and meet the specific coverage guidelines and applicable safety criteria for the covered indication.

FDA-approved indication

MEPSEVII is indicated in pediatric and adult patients for the treatment of Mucopolysaccharidosis VII (MPS VII, Sly syndrome).

Approved Off-label Compendial use(s)

None

Coverage Guidelines

The individual must meet all of the following criteria for approval:

- Must have a diagnosis of Mucopolysaccharidosis VII (MPS VII, Sly syndrome) confirmed by enzyme assay demonstrating a deficiency of β -glucuronidase enzyme activity or by genetic testing
- MEPSEVII will not be used primarily to treat the central nervous system manifestation of MPS VII
- In the event of anaphylaxis, the appropriate medical support will be readily available when MEPSEVII is administered.

Dosing Recommendation

The recommended dosage is 4 mg/kg via intravenous infusion every 2 weeks.

Approval duration (initial and renewal): 12 months

References

1. MEPSEVII prescribing information. Novato, CA. Ultragenyx Pharmaceutical Inc. November 2017.
2. Montano AM, Lock-Hock N, Steiner RD, et al. Clinical course of sly syndrome (Mucopolysaccharidosis type VII). *J Med Genet.* 2016;53(6):403-418. Available at: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4893087/pdf/jmedgenet-2015-103322.pdf>. Accessed on January 18, 2018.
3. National Organization for Rare Disorders: Mucopolysaccharidosis Type VII. Available at: <https://rarediseases.org/rare-diseases/sly-syndrome/>. Accessed on January 18, 2018.
4. National MPS Society: MPS VII (Sly syndrome). Available at: <https://mpssociety.org/learn/diseases/mps-vii/>. Accessed on January 18, 2018.