Elaprase (idursulfase)

Effective Date: January 1, 2021	Number: MG.MM.PH.308

Definitions

Elaprase is human iduronate-2-sulfatase (idursulfase), produced in a human cell line using recombinant DNA technology. Idursulfase hydrolyzes the 2-sulfate esters of terminal iduronate sulfate residues from dermatan and heparin sulfate in lysosomes of various cell types. Elaprase is indicated for patients with Hunter syndrome (Mucopolysaccharidosis type II [MPS II]). Elaprase has been shown to improve walking capacity in patients ≥ 5 years of age. In patients 16 months to 5 years of age, no data are available to demonstrate improvement in disease-related symptoms or long-term clinical outcome; however, treatment has reduced spleen volume similar to that of patients ≥ 5 years of age.

Dosing

Each dose must not exceed 0.5 mg/kg administered intravenously no more frequently than once a week.

Length of Coverage

- Approvals will be granted for 12 months

Guideline

Mucopolysaccharidosis Type II (Hunter Syndrome)

- The diagnosis is established by **one** of the following:

- Patient has a laboratory test demonstrating deficient iduronate-2-sulfatase activity in leukocytes, fibroblasts, serum, or plasma; **OR**
- Patient has a molecular genetic test demonstrating iduronate-2-sulfatase gene mutation; **AND**

- Elaprase is prescribed by or in consultation with a geneticist, endocrinologist, a metabolic disorder sub-specialist, or a physician who specializes in the treatment of lysosomal storage disorders.

Limitations/Exclusions

- Coverage is not recommended for circumstances not listed in the Guideline. Criteria will be updated as new published data are available.

Applicable Procedure Codes

J1743	Injection, idursulfase, 1 mg; 1 mg = 1 billable unit

Applicable NDC Codes

54092-0700-xx	Elaprase 6 mg/3 mL single-use vial for injection

Applicable Diagnosis Codes

E76.1	Mucopolysaccharidosis, type II

Revisions

1/1/2021	Criteria apply to Commercial, Medicare, and Medicaid members.

REFERENCES

1. Elaprase® injection for intravenous use [prescribing information]. Lexington, MA: Shire Human Genetic Therapies, Inc.; November, 2018.
2. Scarpa M, Almassy Z, Beck M, et al. Mucopolysaccharidosis type II: European recommendations for the diagnosis and multidisciplinary management of a rare disease. *Orphanet J Rare Dis*. 2011;6:72.
3. Muenzer J, Beck M, Eng CM, et al. Multidisciplinary management of Hunter syndrome. *Pediatrics*. 2009;124:e1228-e1239.
4. Giugliani R, Federhen A, Munoz Rojas MV, et al. Mucopolysaccharidosis I, II, and VI: Brief review and guidelines for treatment. *Genet Mol Biol*. 2010;33:589-604.
5. D'Avanzo F, Rigon L, Zanetti A, Tomanin R. Mucopolysaccharidosis type II: One hundred years of research, diagnosis, and treatment. *Int J Mol Sci*. 2020;21:E1258.