Authorization

- Coverage will be provided for 12 months and may be renewed.

Dosing Limits

A. Max Units (per dose and over time) [Medical Benefit]:

- 60 billable units every 7 days

Dosing and Administration

Elaprase Package Insert

Guideline

Initial Criteria

Coverage is provided in the following conditions:

**Hunter syndrome (Mucopolysaccharidosis II; MPS II) †**

- Patient is at least 5 years old; **AND**
- Patient has absence of severe cognitive impairment; **AND**
- Diagnosis has been confirmed by one of the following:
Deficient iduronate 2-sulfatase (I2S) enzyme activity in white cells, fibroblasts, or plasma in the presence of normal activity of at least one other sulfatase; OR
- Detection of pathogenic mutations in the IDS gene by molecular genetic testing; AND
- Documented baseline value for urinary glycosaminoglycan (uGAG)
- Documented baseline values for one or more of the following:
  - Patients 5 years or greater: 6-minute walk test (6-MWT) and/or percent predicted forced vital capacity (FVC); OR
  - Patients < 5 years: spleen volume, liver volume, FVC, and/or 6-minute walk test

† FDA Approved Indication(s)

Renewal Criteria

Authorizations can be renewed based on the following criteria:

- Patient continues to meet the criteria above; AND
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include the following: severe hypersensitivity including anaphylactic and anaphylactoid reactions, antibody development and serious adverse reactions, acute respiratory complications, acute cardiorespiratory failure, etc.; AND
- Patient does not have progressive/irreversible severe cognitive impairment; AND
- Patient has a documented reduction in uGAG levels; AND
- Patient has demonstrated a beneficial response to therapy compared to pretreatment baseline in one or more of the following:
  - Patients 5 years or greater: stabilization or improvement in 6-MWT and/or FVC; OR
  - Patients < 5 years: spleen volume, and/or liver volume or stabilization/improvement in FVC and/or 6-MWT

Limitations/Exclusions

Elaprase is considered investigational when used for any indication not listed above.

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
References


