

Reference number
2052-A

# SPECIALTY GUIDELINE MANAGEMENT

## ELAPRASE (idursulfase)

### POLICY

#### I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

##### FDA-Approved Indication

Elaprase is indicated for patients with Hunter syndrome (Mucopolysaccharidosis II, MPS II). Elaprase has been shown to improve walking capacity in patients 5 years and older. 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 with Elaprase has reduced spleen volume similarly to that of adults and children 5 years of age and older. The safety and efficacy of Elaprase have not been established in pediatric patients less than 16 months of age.

All other indications are considered experimental/investigational and not medically necessary.

#### II. DOCUMENTATION

Submission of the following information is necessary to initiate the prior authorization review:

- A. Initial requests: iduronate 2-sulfatase enzyme assay or genetic testing results supporting diagnosis.
- B. Continuation requests: chart notes documenting a clinically positive response to therapy, which shall include improvement, stabilization, or slowing of disease progression.

#### III. CRITERIA FOR INITIAL APPROVAL

##### **Mucopolysaccharidosis II (MPS II)**

Authorization of 12 months may be granted for treatment of MPS II when the diagnosis of MPS II was confirmed by enzyme assay demonstrating a deficiency of iduronate 2-sulfatase enzyme activity or by genetic testing.

#### IV. CONTINUATION OF THERAPY

Authorization of 12 months may be granted for continued treatment in members requesting reauthorization for mucopolysaccharidosis II (MPS II) who have a clinically positive response to therapy, which shall include improvement, stabilization, or slowing of disease progression.

#### V. REFERENCES

1. Elaprase [package insert]. Lexington, MA: Shire Human Genetic Therapies, Inc.; November 2018.
2. Muenzer J, Beck M, Eng CM, et al. Multidisciplinary management of Hunter syndrome. *Pediatrics*. 2009;124(6):e1228-e1239.