Idursulfase (Elaprase®)	Place of Service Infusion Center Administration Home Infusion Administration Office Administration Outpatient Facility Infusion Administration* [*Prior authorization required – see section (1)] HCPCS: **J1743** per 1 mg

Conditions listed in policy (*see criteria for details*)

- Mucopolysaccharidosis II (MPSII), also known as Hunter's Syndrome

AHFS therapeutic class: Enzymes

Mechanism of action: Idursulfase is a biosynthetic, (recombinant DNA origin) human enzyme replacement therapy for the treatment of Hunter syndrome, also known as Mucopolysaccharidosis II (MPS II).

(1) Special Instructions and pertinent Information

Covered under the medical benefit, please submit clinical information for prior authorization review.

****CRITERIA FOR HOSPITAL OUTPATIENT FACILITY ADMINISTRATION ****
AAAAI Guidelines 2011, MCG™ Care Guidelines, 19th edition, 2015

Members with the following plans: **PPO, Direct Contract HMO, and when applicable, ASO/Shared Advantage/HMO (non-direct contract)** may be required to have their medication administered at a preferred site of service, including the home, a physician's office, or an independent infusion center not associated with a hospital.
For members that cannot receive infusions in the preferred home or ambulatory setting AND meet one of the following criteria points, drug administration may be performed at a hospital outpatient facility infusion center.

ADMINISTRATION OF ELAPRASE IN THE HOSPITAL OUTPATIENT FACILITY SITE OF CARE REQUIRES ONE OF THE FOLLOWING: (*Supporting Documentation must be submitted)*

1. **Patient is initiating therapy (allowed for the first 4 infusions) with Elaprase or is being re-initiated on Elaprase after at least 6 months off therapy.** *Subsequent doses will require medical necessity for continued use in the hospital outpatient facility site of care.*

Or

Additional clinical monitoring is required during administration as evidenced by one of the following:

2. **Patient has experienced a previous severe adverse event on Elaprase based on documentation submitted.**

3. **Patient continues to experience moderate to severe adverse events on Elaprase based on documentation submitted, despite receiving premedication such as acetaminophen, steroids, diphenhydramine, fluids, etc.**

4. **Patient is clinically unstable based on documentation submitted.**

5. **Patient is physically or cognitively unstable based on documentation submitted.**

(2) Prior Authorization/Medical Review is required for the following condition(s)

All requests for Elaprase® (idursulfase) must be sent for clinical review and receive authorization prior to drug administration or claim payment.

Mucopolysaccharidosis II (MPS II) or Hunter's Syndrome

1. Either of the following:
    a. Documented reduced enzyme activity of iduronate-2-sulfatase (12S), **OR**
    b. Genetic testing confirming diagnosis of MPS II

Covered Doses
Up to 0.5 mg/kg administered intravenously once weekly

Coverage Period
indefinite

ICD-10:
E76.1

(3) The following condition(s) DO NOT require Prior Authorization/Preservice

All requests for Elaprase® (idursulfase) must be sent for clinical review and receive authorization prior to drug administration or claim payment.

(4) This Medication is NOT medically necessary for the following condition(s)

Coverage for a Non-FDA approved indication, requires that criteria outlined in Health and Safety Code § 1367.21, including objective evidence of efficacy and safety are met for the proposed indication.

Please refer to the Provider Manual and User Guide for more information.

(5) Additional information

How supplied:
6mg/3ml (single use vials)

Due to potential for hypersensitivity reactions, medical support should be readily available when Elaprase® is administered.

(6) References

- AHFS®. Available by subscription at http://www.lexi.com
- DrugDex®. Available by subscription at http://www.thomsonhc.com
- Elaprase® (idursufase) [Prescribing information]. Lexington, MA: Shire Human Genetic Therapies, Inc.; 9/2021.
- Wang RY, Bodamer OA, et al. American College of Medical Genetics (ACMG) Work Group on Diagnostic Confirmation of Lysosomal Storage Diseases. Lysosomal storage diseases: diagnostic confirmation and management of presymptomatic individuals. Genet Med. 2011; 13(5):457-484.
- Wraith JE, Scarpa M, Beck M, et al. Mucopolysaccharidosis type II (Hunter syndrome): a clinical review and recommendations for treatment in the era of enzyme replacement therapy. Eur J Pediatr. 2008 Mar; 167(3): 267–277.

(7) Policy Update

Date of last review: 3Q2023

Date of next review: 3Q2024

Changes from previous policy version:

- No clinical change to policy following routine annual review.

BSC Drug Coverage Criteria to Determine Medical Necessity
Reviewed by P&T Committee