



Elaprase® (idursulfase) (Intravenous)

Document Number: SHP-0034

Last Review Date: 02/02/2023

Date of Origin: 01/01/2012

Dates Reviewed: 12/2011, 02/2013, 02/2014, 12/2014, 10/2015, 10/2016, 10/2017, 10/2018, 02/2019, 02/2020, 02/2021, 02/2022, 04/2022, 02/2023

I. Length of Authorization

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

II. Dosing Limits

A. Quantity Limit (max daily dose) [NDC Unit]:

- Elaprase 6 mg/3 mL vial: 10 vials per 7 days

B. Max Units (per dose and over time) [HCPCS Unit]:

- 60 billable units every 7 days

III. Initial Approval Criteria ^{1,4,5,7,9,10}

Submission of medical records (chart notes) related to the medical necessity criteria is REQUIRED on all requests for authorizations. Records will be reviewed at the time of submission. Please provide documentation related to diagnosis, step therapy, and clinical markers (i.e. genetic and mutational testing) supporting initiation when applicable. Medical records may be submitted via direct upload through the PA web portal or by fax.

Coverage is provided in the following conditions:

- Patient is at least 16 months of age; **AND**
- Documented baseline age-appropriate values for one or more of the following have been obtained:
 - Patients 5 years of age or greater: 6-minute walk test (6MWT), percent predicted forced vital capacity (FVC), joint range of motion, left ventricular hypertrophy, growth, quality of life (CHAQ/HAQ/MPS HAQ), and/or urinary glycosaminoglycan (uGAG); **OR**
 - Patients 16 months to less than 5 years of age: spleen volume, liver volume, FVC, 6-MWT, and/or urinary glycosaminoglycan (uGAG); **AND**

****NOTE:** For very young patients in which FVC or 6-MWT are not suitable for measuring, requests will be reviewed on a case-by case basis.

Universal Criteria ¹

- Patient does not have severe cognitive impairment; **AND**

Hunter syndrome (Mucopolysaccharidosis II; MPS II) † ‡ **Φ** ^{1,5}

- Patient has a definitive diagnosis of MPS II as confirmed by one of the following:
 - Deficient or absent 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

† FDA Approved Indication(s); ‡ Compendia recommended indication(s); **Φ** Orphan Drug

IV. Renewal Criteria ^{1,4,5,7,9,10}

Coverage may be renewed based on the following criteria:

- Patient continues to meet the universal and other indication-specific relevant criteria such as concomitant therapy requirements (not including prerequisite therapy), performance status, etc. identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: severe hypersensitivity reactions including anaphylaxis, antibody development and serious adverse reactions in Hunter Syndrome patients with severe genetic mutations, acute respiratory complications, acute cardiorespiratory failure, etc.; **AND**
- Patient has demonstrated a beneficial response to therapy compared to pretreatment age-appropriate baseline values in one or more of the following:
 - Patients 5 years of age or greater: stabilization or improvement in percent predicted FVC and/or 6-MWT, increased joint range of motion, decreased left ventricular hypertrophy, improved growth, improved quality of life (clinically meaningful change in the CHAQ/HAQ/MPS HAQ disability index), and/or uGAG levels; **OR**
 - Patients 16 months to less than 5 years of age: reductions in spleen volume and/or liver volume or stabilization/improvement in FVC and/or 6-MWT, and/or uGAG levels

V. Dosage/Administration ¹

Indication	Dose
Hunter Syndrome; MPS II	0.5 mg/kg of body weight administered once weekly as an intravenous infusion

VI. Billing Code/Availability Information

HCP Code:

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

NDC:

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

VII. References

1. Elaprase [package insert]. Lexington, MA; Shire Human Genetic Therapies, Inc; September 2021. Accessed December 2022.
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3. Scarpa M, Almássy 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 Nov 7;6:72. doi: 10.1186/1750-1172-6-72.
4. Muenzer J, Bodamer O, Burton B, et al. The role of enzyme replacement therapy in severe Hunter syndrome-an expert panel consensus. *Eur J Pediatr*. 2012 Jan;171(1):181-8.
5. Scarpa M. Mucopolysaccharidosis Type II. GeneReviews®. www.ncbi.nlm.nih.gov/books/NBK1274/. Initial Posting: November 6, 2007; Last Update: October 4, 2018. Accessed on December 19, 2022.
6. Burrow T, Leslie ND. Review of the use of idursulfase in the treatment of mucopolysaccharidosis II. *Biologics*. 2008 Jun; 2(2): 311–320.
7. Giugliani R, Villareal MLS, Valdez CAA, et al. Guidelines for diagnosis and treatment of Hunter Syndrome for clinicians in Latin America. *Genet Mol Biol*. 2014 Jun; 37(2): 315–329.
8. Burton BK, Giugliani R. Diagnosing Hunter syndrome in pediatric practice: practical considerations and common pitfalls. *Eur J Pediatr* 2012; 171:631.
9. Muenzer J, Wraith J, Beck M, *et al*. A phase II/III clinical study of enzyme replacement therapy with idursulfase in mucopolysaccharidosis II (Hunter syndrome). *Genet Med* 8, 465–473 (2006) doi:10.1097/01.gim.0000232477.37660.fb
10. Muenzer J, Beck M, Eng CM, et al. Long-term, open-labeled extension study of idursulfase in the treatment of Hunter syndrome. *Genet Med*. 2011 Feb;13(2):95-101. doi: 10.1097/GIM.0b013e3181fea459.

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
E76.1	Mucopolysaccharidosis, type II

ELAPRASE® (idursulfase) Prior Auth Criteria

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Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD), Local Coverage Determinations (LCDs), and Local Coverage Articles (LCAs) may exist and compliance with these policies is required where applicable. They can be found at:

<https://www.cms.gov/medicare-coverage-database/search.aspx>. Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD/LCA): N/A

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	Applicable State/US Territory	Contractor
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC