Elaprase® (idursulfase)
(Intravenous)

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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]

Site of care specialty infusion program requirements are met (refer to EOCCO Site of Care Policy).

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 [1,11-13]

- Therapy is being used to treat non-central nervous system manifestations of the disease and patient does not have severe, irreversible 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 reduction in uGAG levels; **OR**
    - Patients 16 months to less than 5 years of age: reductions in spleen and/or liver volume, stabilization/improvement in FVC and/or 6-MWT, and/or reduction in 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

HCPCS 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; Takeda Pharmaceuticals U.S.A., Inc,; September 2021. Accessed December 2023.
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12. Zhou J, Lin J, Leung WT, Wang L. A basic understanding of mucopolysaccharidosis: Incidence, clinical features, diagnosis, and management. Intractable Rare Dis Res. 2020 Feb;9(1):1-9. doi: 10.5582/irdr.2020.01011.

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Appendix 1 – Covered Diagnosis Codes

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

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

The preceding information is intended for non-Medicare coverage determinations. 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 Determinations (NCDs) and/or Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. Local Coverage Articles (LCAs) may also exist for claims payment purposes or to clarify benefit eligibility under Part B for drugs which may be self-administered. The following link may be used to search for NCD, LCD, or LCA documents: https://www.cms.gov/medicare-coverage-database/search.aspx. Additional indications, including any preceding information, may be applied 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