Elaprase® (idursulfase)
(Intravenous)

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I. Length of Authorization

- Initial: Prior authorization validity will be provided initially for 12 months (365 days).
- Renewal: Prior authorization validity may be renewed every 12 months (365 days) thereafter.

II. Dosing Limits

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

- 240 billable units every 28 days

III. Initial Approval Criteria [1,4,5,7,9,10]

Prior authorization validity is provided in the following conditions:

- Member is at least 16 months of age; **AND**
- Documented baseline age-appropriate values for one or more of the following have been obtained:
  - Members 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 measure (CHAQ/HAQ/MPS HAQ), and/or urinary glycosaminoglycan (uGAG); **OR**
  - Members 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 members in which FVC or 6-MWT are not suitable for measuring, requests will be reviewed on a case-by-case basis.*

Universal Criteria [1,5,11-13]

- Therapy is being used to treat non-central nervous system manifestations of the disease and member does not have severe, irreversible cognitive impairment; **AND**

NOTE: *Requests for continued therapy in members with severe, irreversible cognitive impairment will be reviewed on a case-by-case basis.*

Hunter syndrome (Mucopolysaccharidosis II; MPS II) † Φ [1,5]

- Member 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 (or likely pathogenic) variants 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]

Prior authorization validity may be renewed based on the following criteria:

- Member 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 members with severe genetic mutations, acute respiratory complications, acute cardiorespiratory failure, etc.; **AND**
- Member has demonstrated a beneficial response to therapy compared to pretreatment age-appropriate baseline values in one or more of the following:
  - Members 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**
  - Members 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 billable unit = 1 mg

NDC:

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

VII. References

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Appendix A – Non-Quantitative Treatment Limitations (NQTL) Factor Checklist

Non-quantitative treatment limitations (NQTLs) refer to the methods, guidelines, standards of evidence, or other conditions that can restrict how long or to what extent benefits are provided under a health plan. These may include things like utilization review or prior authorization. The utilization management NQTL applies comparably, and not more stringently, to mental health/substance use disorder (MH/SUD) Medical Benefit Prescription Drugs and medical/surgical (M/S) Medical Benefit Prescription Drugs. The table below lists the factors that were considered in designing and applying prior authorization to this drug/drug group, and a summary of the conclusions that Prime's assessment led to for each.

Factor	Conclusion
Indication	Yes: Consider for PA
Safety and efficacy	Yes: Consider for PA
Potential for misuse/abuse	No: PA not a priority
Cost of drug	Yes: Consider for PA

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

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
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
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA
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