Elaprase® (idursulfase)
(Intravenous)

Document Number: IC-0034

Last Review Date: 10/02/2018
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Dates Reviewed: 12/2011, 02/2013, 02/2014, 12/2014, 10/2015, 10/2016, 10/2017, 10/2018

I. Length of Authorization

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

II. Dosing Limits

A. Quantity Limit (max daily dose) [Pharmacy Benefit]:

– Elaprase 6 mg vial: 10 vials per 7 days

B. Max Units (per dose and over time) [Medical Benefit]:

- 60 billable units every 7 days

III. Initial Approval Criteria

Coverage is provided in the following conditions:

Hunter syndrome (Mucopolysaccharidosis II; MPS II) †

- Patient is at least 16 months old; **AND**
- Patient has absence of severe cognitive impairment; **AND**
- Diagnosis has been confirmed by one of the following:
  - Deficient 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; **AND**
- Documented baseline value for urinary glycosaminoglycan (uGAG)
- Documented baseline values for one or more of the following:
  - Patients 5 years or greater: 6-minute walk test (6-MWT) and/or percent predicted forced vital capacity (FVC); **OR**
  - Patients < 5 years: spleen volume, liver volume, FVC, and/or 6-minute walk test

† FDA Approved Indication(s)

IV. Renewal Criteria

Authorizations can be renewed based on the following criteria:

- Patient continues to meet the criteria in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include the following: severe hypersensitivity including anaphylactic and anaphylactoid reactions, antibody development and serious adverse reactions, acute respiratory complications, acute cardiorespiratory failure, etc.; **AND**
- Patient does not have progressive/irreversible severe cognitive impairment; **AND**
- Patient has a documented reduction in uGAG levels; **AND**
- Patient has demonstrated a beneficial response to therapy compared to pretreatment baseline in one or more of the following:
    - Patients 5 years or greater: stabilization or improvement in 6-MWT and/or FVC; **OR**
    - Patients < 5 years: spleen volume, and/or liver volume or stabilization/improvement in FVC and/or 6-MWT

V. Dosage/Administration

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

Jcode:

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; June 2013. Accessed August 2018.
2. 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):267-77. Epub 2007 Nov 23.
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/ (Accessed on September 8, 2017).
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.

Appendix 1 – Covered Diagnosis Codes

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

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) and Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. They can be found at: http://www.cms.gov/medicare-coverage-database/search/advanced-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):

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 Government Benefit Administrators, 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)

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
15	KY, OH	CGS Administrators, LLC