Elaprase® (idursulfase)  
(Intravenous)

Effective Date: 03/01/2020  
Review Date: 01/28/2021, 02/11/2021  
Revision date: 01/28/2021, 2/11/2021

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 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\textsuperscript{1,4,5,7,9,10}

Coverage is provided in the following conditions:

MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements.

- Patient is at least 16 months of age: \textbf{AND}
- Patient has absence of severe cognitive impairment: \textbf{AND}
- Documented baseline value for urinary glycosaminoglycan (uGAG) has been obtained: \textbf{AND}
- Documented baseline values for one or more of the following have been obtained:
  - Patients 5 years or greater: 6-minute walk test (6-MWT), percent predicted forced vital capacity (FVC), joint range of motion, left ventricular hypertrophy, growth, quality of life (CHAQ/HAQ/MPS HAQ): \textbf{OR}
  - Patients < 5 years: spleen volume, liver volume, FVC, and/or 6-minute walk test: \textbf{AND}

\textbf{Hunter syndrome (Mucopolysaccharidosis II; MPS II) † Φ}

- Diagnosis has been 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); Ø Orphan Drug

### IV. Renewal Criteria¹,⁴,⁵,⁷,⁹,¹⁰

Coverage can be renewed based on the following criteria:

- Patient continues to meet indication-specific relevant criteria 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 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 percent predicted FVC and/or 6-minute walk test, 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): **OR**
  - Patients < 5 years: reductions in spleen volume and/or liver volume or stabilization/improvement in FVC and/or 6-MWT

### V. Dosage/Administration¹,⁹,¹⁰

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
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<tbody>
<tr>
<td>Hunter Syndrome: MPS II</td>
<td>0.5 mg/kg of body weight administered once weekly as an intravenous infusion</td>
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</table>

### 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


Appendix 1 – Covered Diagnosis Codes

<table>
<thead>
<tr>
<th>ICD-10</th>
<th>ICD-10 Description</th>
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<tbody>
<tr>
<td>E76.1</td>
<td>Mucopolysaccharidosis, type II</td>
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</table>
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: [http://www.cms.gov/medicare-coverage-database/search/advanced-search.aspx](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/LCA): N/A

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<td>Noridian Healthcare Solutions, LLC</td>
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<tr>
<td>F (2 &amp; 3)</td>
<td>AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ</td>
<td>Noridian Healthcare Solutions, LLC</td>
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<tr>
<td>5</td>
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<td>MN, WI, IL</td>
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