Definition
Aldurazyme is a recombinant form of human alpha-L-iduronidase indicated for enzyme replacement therapy in patients with Mucopolysaccharidosis I.

Length of Authorization
Coverage will be provided for 12 months and may be renewed

Dosing Limits

Max Units (per dose and over time) [Medical Benefit]:
- 667 billable units every 7 days

Guideline

I. INITIAL APPROVAL CRITERIA
Coverage is provided in the following conditions:

Mucopolysaccharidosis I (MPS I) †
- Patient has a definitive diagnosis of MPS I confirmed by one of the following:
Detection of pathogenic mutations in the *IDUA* gene by molecular genetic testing; **OR**
- Detection of deficient activity of the lysosomal enzyme α-L-iduronidase; **AND**
- Diagnosis of Hurler (severe) or Hurler-Scheie (attenuated) forms of disease **OR**
- Diagnosis of Scheie (attenuated) form of disease with moderate to severe symptoms; **AND**
- Patient is 6 months of age or older; **AND**
- Patient has absence of severe cognitive impairment; **AND**
- Documented baseline value for urinary glycosaminoglycan (uGAG); **AND**
- Documented baseline values for one or more of the following:
  - **Patients 6 years or greater:** percent predicted forced vital capacity (FVC) and/or 6-minute walk test; **OR**
  - **Patients 6 months to less than 6 years:** cardiac status, upper airway obstruction during sleep, growth velocity, mental development, FVC, and/or 6-minute walk test

† FDA approved indication(s)

**II. RENEWAL CRITERIA**

Authorizations can be renewed based on the following criteria:
- Patient continues to meet the criteria identified above; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include the following: severe hypersensitivity reactions, acute respiratory complications, acute cardiorespiratory failure, severe infusion reactions, 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 6 years or greater:** stability or improvement in percent predicted FVC and/or 6-minute walk test; **OR**
  - **Patients 6 months to less than 6 years:** stability or improvement in cardiac status, upper airway obstruction during sleep, growth velocity, mental development, FVC and/or 6-minute walk test

**Dosing/Administration**

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<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
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<tr>
<td>Mucopolysaccharidosis I (MPS I)</td>
<td>0.58 mg/kg of body weight administered once weekly, as an intravenous infusion, over 3-4 hours.</td>
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**Applicable Procedure Codes**

| J1931 | Injection, laronidase, 0.1 mg; 1 billable unit = 0.1 mg |
Applicable NDC’s

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<thead>
<tr>
<th>NDC Code</th>
<th>Description</th>
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<tr>
<td>58468-0070-xx</td>
<td>Aldurazyme 2.9 mg/5 mL single-dose vial</td>
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ICD-10  | ICD-10 Description  
E76.01 | Hurler’s syndrome  
E76.02 | Hurler-Scheie syndrome  
E76.03 | Scheie's syndrome  

Revision History

References