Authorization

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

Dosing Limits

A. Max Units (per dose and over time) [Medical Benefit]:
   • 115 billable units every 14 days

Dosing and Administration

Guideline

Initial Approval Criteria

Coverage is provided in the following conditions:

Fabry Disease (alpha-galactosidase A deficiency) †
   • Patient is 8 years of age or older; AND
   • Documented diagnosis of Fabry disease with biochemical/genetic confirmation by one of the following:
α-galactosidase A (α-Gal A) activity in plasma, isolated leukocytes, and/or cultured cells (males only); OR

- Plasma or urinary globotriaosylceramide(Gb3/GL-3) or globotriaosylsphingosine (lyso-Gb3); OR

- Detection of pathogenic mutations in the GALA/GLA gene by molecular genetic testing; AND

  - Baseline value for plasma GL-3 and/or GL-3 inclusions; AND
  - Must not be used in combination with migalastat

† FDA approved indication(s)

Renewal Criteria

Authorizations can be renewed based on the following criteria:

- Patient continues to meet the criteria identified in section III; AND

- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include the following: severe hypersensitivity reactions, severe infusion site reactions, compromised cardiac function, etc.; AND

- Disease response with treatment as defined by a reduction in plasma GL-3 and/or GL-3 inclusions compared to pre-treatment baseline

Limitations/Exclusions

Fabrazyme is considered investigational when used for any indication not listed above.

Applicable Procedure Codes

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<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>J0180</td>
<td>Injection, agalsidase beta, 1 mg; 1 billable unit = 1 mg</td>
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</table>

Applicable Procedure Codes

<table>
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<tr>
<th>Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>54868-0041-xx</td>
<td>Fabrazyme 5 mg single-use vial for injection</td>
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<tr>
<td>54868-0040-xx</td>
<td>Fabrazyme 35 mg single-use vial for injection</td>
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Applicable Diagnosis Codes

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<th>Description</th>
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<tr>
<td>E75.21</td>
<td>Fabry (-Anderson) disease</td>
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References


