Fabrazyme® (agalsidase beta)
(Intravenous)

Last Review Date: January 1, 2020
Number: MG.MM.PH.82

Medical Guideline Disclaimer
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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

Fabrazyme Package Insert

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:
  o $\alpha$-galactosidase A ($\alpha$-Gal A) activity in plasma, isolated leukocytes, and/or cultured cells (males only); OR
  o Plasma or urinary globotriaosylceramide (Gb$_3$/GL-3) or globotriaosylsphingosine (lyso-Gb$_3$); OR
  o 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 above; 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>
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<tbody>
<tr>
<td>J0180</td>
<td>Injection, agalsidase beta, 1 mg; 1 billable unit = 1 mg</td>
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<table>
<thead>
<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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<tr>
<th>Code</th>
<th>Description</th>
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<tr>
<td>E75.21</td>
<td>Fabry (-Anderson) disease</td>
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References


