**Alpha-1-Proteinase Inhibitors Infusion Therapy**

Last Review Date: September 9, 2016  
Number: MG.MM.PH.27

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**Related Medical Guideline**

[Off-Label Use of FDA-Approved Drugs and Biologicals](#)

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**Dosage and Administration**

Refer to drug-specific labeling

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**Applicable Products**

- Aralast NP®
- Glassia®
- Prolastin®
- Prolastin®-C
- Zemaira®

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**Guideline**

Alpha-1-Proteinase Inhibitors are considered medically necessary for emphysema secondary to alpha-1-antitrypsin (ATT) deficiency when the following criteria are met; all:

1. Presence AAT deficiency with PiZZ, PiZ (null) or Pi (null,null) phenotypes
2. Presence of AAT deficiency and clinical evidence of panacinar emphysema
3. Low serum AAT concentration ($\leq$ 11 uM/L [35% of normal]) or $\leq$ 80 mg/dL [measured by radial immunodiffusion] or $\leq$ 0.8 g/L [measured by nephelometry]
Renewal Criteria

Coverage will be given for six months and is eligible for renewal when both of the following criteria are met:

1. Positive response to treatment (defined by elevation of AAT levels above baseline and/or substantial reduction in deterioration-rate of lung function, as measured by percent-predicted FEV1)

2. Absence of unacceptable drug-toxicity (i.e., hypersensitivity reactions)

Limitations/Exclusions

Alpha-1-Proteinase Inhibitors are considered investigational and not medically necessary for any indications other than those listed above due to insufficient evidence of therapeutic value.

References


Applicable Diagnosis Codes

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<thead>
<tr>
<th>Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>E88.01</td>
<td>Alpha-1-antitrypsin deficiency</td>
</tr>
<tr>
<td>J43.0</td>
<td>Unilateral pulmonary emphysema [MacLeod's syndrome]</td>
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<tr>
<td>J43.1</td>
<td>Panlobular emphysema</td>
</tr>
<tr>
<td>J43.2</td>
<td>Centrilobular emphysema</td>
</tr>
<tr>
<td>J43.8</td>
<td>Other emphysema</td>
</tr>
<tr>
<td>J43.9</td>
<td>Emphysema, unspecified</td>
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</tbody>
</table>

Applicable Procedure Codes

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>J0256</td>
<td>Injection, alpha 1-proteinase inhibitor (human), not otherwise specified, 10 mg</td>
</tr>
<tr>
<td>J0257</td>
<td>Injection, alpha 1 proteinase inhibitor (human), (GLASSIA), 10 mg</td>
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<tr>
<td>S9346</td>
<td>Home infusion therapy, alpha-1-proteinase inhibitor (e.g., Prolastin); administrative services, professional pharmacy services, care coordination, and all necessary supplies and equipment (drugs and nursing visits coded separately), per diem</td>
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