Alpha₁-Proteinase Inhibitors

Description

Aralast NP, Glassia, Prolastin-C, Zemaira

Background

Aralast NP, Glassia, Prolastin-C, and Zemaira are intravenous infusions indicated for individuals with clinically evident emphysema due to severe deficiency of Alpha₁-PI, also known as alpha₁-antitrypsin (AAT) deficiency. These medications increase antigenic and functional (anti-neutrophil elastase capacity, ANEC) serum levels and antigenic lung epithelial lining fluid levels of Alpha₁-PI. Intravenous administration of purified preparations of pooled donor-derived human AAT has been shown to augment levels of AAT and the AAT-related anti-elastase capacity of serum and lung epithelial lining fluid. The current U.S. Food and Drug Administration (FDA)-approved intravenous augmentation therapy dose for chronic administration is 60 mg/kg body weight, administered weekly (1-6).

Regulatory Status

FDA-approved indications: Aralast NP, Glassia, Prolastin-C, and Zemaira are indicated for chronic augmentation therapy in individuals with clinically evident emphysema due to severe congenital deficiency of alpha₁-PI (1-4).

The safety of Alpha₁-Proteinase Inhibitors in patients with severe renal impairment (creatinine clearance (CrCl) less than 30 mL/min) or end-stage renal disease has not been studied. The safety of Alpha₁-Proteinase Inhibitors in patients with moderate to severe hepatic impairment has not been studied (1-4).
Intravenous augmentation therapy is recommended for individuals with AATD and an FEV1 in the range of 30%-65% predicted (strong recommendation, high quality evidence) (6).

High value is placed on the potential to prolong survival in this group, the finding that intravenous augmentation therapy is associated with lower levels of elastin degradation products in individuals with AATD, and lower rates of loss of CT lung density in individuals with AATD-COPD receiving augmentation therapy. Low value is placed on the cost of this therapy (6).

The safety and effectiveness of Alpha\textsubscript{1}-Proteinase Inhibitors in pediatric patients have not been established (1-4).

Related policies

Policy

This policy statement applies to clinical review performed for pre-service (Prior Approval, Precertification, Advanced Benefit Determination, etc.) and/or post-service claims.

Aralast NP, Glassia, Prolastin-C, and Zemaira may be considered medically necessary in patients age 18 years or age or older in individuals with emphysema and if the conditions indicated below are met.

Aralast NP, Glassia, Prolastin-C, and Zemaira are considered investigational in patients less than 18 years of age and for all other indications.

Prior-Approval Requirements

Age

18 years of age and older

Diagnosis

The patient must have the following:

1. Emphysema
   a. Clinically documented alpha\textsubscript{1} antitrypsin (AAT) deficiency with a pretreatment serum AAT level less than 11 \( \mu \)M/L (80 mg/dl by radial immunodiffusion or 50 mg/dl by nephelometry)
   b. Patient must NOT be a current smoker
5.45.09

Section: Prescription Drugs  Effective Date: April 1, 2019
Subsection: Respiratory Agents  Original Policy Date: July 21, 2017
Subject: Alpha1-Proteinase Inhibitors  Page: 3 of 4

...c. Documented progressive emphysema with ONE of the following:
   i. Moderate airflow obstruction is evidenced by forced expiratory volume (FEV₁) of 30-65% of predicted value, prior to initiation of therapy
   ii. Individual has a rapid decline in lung function as measured by a change in FEV₁ greater than 120 ml/year

Prior – Approval Renewal Requirements

Age  18 years of age and older

Diagnosis

The patient must have the following:

1. Emphysema
   a. Patient must NOT be a current smoker
   b. Clinical evidence of efficacy with ONE of the following:
      i. Elevation of AAT levels (above protective threshold)
      ii. Reduction in rate of deterioration of lung function with a reduction in FEV₁ rate of decline

Policy Guidelines

Pre - PA Allowance
None

Prior - Approval Limits
Duration  3 months

Prior – Approval Renewal Limits
Duration  12 months

Rationale

Summary
Aralast NP, Glassia, Prolastin-C, and Zemaira are intravenous infusions indicated for individuals with clinically evident emphysema due to severe deficiency of Alpha₁-PI, also known as alpha₁-antitrypsin (AAT) deficiency. The safety of Alpha₁-Proteinase Inhibitors in patients with severe
renal impairment (creatinine clearance less than 30 mL/min), end-stage renal disease or moderate to severe hepatic impairment has not been studied. The safety and effectiveness of Alpha1-Proteinase Inhibitors in pediatric patients have not been established (1-4).

Prior authorization is required to ensure the safe, clinically appropriate and cost-effective use of Aralast NP, Glassia, Prolastin-C, and Zemaira while maintaining optimal therapeutic outcomes.

References

Policy History

<table>
<thead>
<tr>
<th>Date</th>
<th>Action</th>
</tr>
</thead>
<tbody>
<tr>
<td>July 2017</td>
<td>Addition to PA</td>
</tr>
<tr>
<td>September 2017</td>
<td>Annual review and reference update</td>
</tr>
<tr>
<td>March 2018</td>
<td>Annual review and reference update</td>
</tr>
<tr>
<td>March 2019</td>
<td>Annual review and reference update</td>
</tr>
</tbody>
</table>

Keywords

This policy was approved by the FEP® Pharmacy and Medical Policy Committee on March 15, 2019 and is effective on April 1, 2019.