Coverage of drugs is first determined by the member’s pharmacy or medical benefit. Please consult with or refer to the Evidence of Coverage document.

I. **FDA Approved Indications:**

- Aralast: Chronic augmentation therapy in adults with clinically evident emphysema due to severe congenital deficiency of alpha1-proteinase inhibitor (Alpha1-PI), also known as alpha1-antitrypsin deficiency.
- Glassia, Prolastin, and Zemaira: Chronic augmentation and maintenance therapy in adults with clinically evident emphysema due to severe congenital deficiency of alpha1-proteinase inhibitor (Alpha1-PI), also known as alpha1-antitrypsin deficiency.

II. **Health Net Approved Indications and Usage Guidelines:**

- Diagnosis of emphysema due to congenital deficiency of alpha 1-proteinase inhibitor deficiency (alpha1-antitrypsin deficiency)
  
  AND

- Patient is 18 years of age or older
  
  AND

- Patient has never smoked or has quit smoking for at least the prior 6 months
  
  AND

- Has clinical signs and symptoms consistent with emphysema
  
  AND

- Impedance forced expiratory volume per one second (iFEV\(_1\)) <80% of predicted volume or residual volume > 120% of predicted volume, or diffusion capacity < 50% of predicted value or forced expiratory volume in one second/forced vital capacity (FEV\(_1\)/FVC) < 70%
  
  AND

- Alpha-1antitrypsin (AAT) level \(\leq 11\) umol/L (corresponds to 80 mg/dl measured by radial immunodiffusion or 0.8 g/L/dl if measured by nephelometry)
  
  OR

- If the patient has an AAT level >11 umol/L, then the patient must have one of the high-risk phenotypes (i.e. PiZZ, PiZnull, Pi(null, null), or one of a few rare phenotypes [e.g. Pi(Malton, Malton)].
III. **Coverage is Not Authorized For:**
- Immunoglobulin A (IgA) deficient patients (IgA level less than 15mg/dL) with known antibody against IgA
- Non-FDA approved indications, which are not listed in the Health Net Approved Indications and usage guidelines section unless there is sufficient documentation of efficacy and safety in the published literature.

IV. **General Information:**
- Augmentation therapy by boosting AAT levels is not a cure, will not reverse lung damage that has already occurred, and has not yet been proven to retard the progression of emphysema.
- Clinical data demonstrating the long-term effects of chronic augmentation or replacement therapy of individuals with alpha-1 proteinase inhibitor are not available.
- IgA may be present in alpha 1-proteinase inhibitors and patients may experience severe reactions, including anaphylaxis.
- The American Thoracic Society (ATS) and the European Respiratory Society (ERS) state that alpha-1-proteinase inhibitor therapy does not confer benefit in, and is not recommended for, patients who have alpha-1-proteinase-associated liver disease.
- Aralast NP, Glassia, Prolastin-C, Zemaira: Safety and effectiveness in the pediatric population have not been established
- Aralast, Glassia, Prolastin, and Zemaira increase antigenic and functional (anti-neutrophil elastase capacity [ANEC]) serum levels and antigenic lung epithelial lining fluid levels of alpha 1-proteinase inhibitor.
- The effect of augmentation therapy with any alpha 1-proteinase inhibitor on pulmonary exacerbations and on the progression of emphysema in alpha1-antitrypsin deficiency has not been conclusively demonstrated in randomized, controlled clinical trials
- Clinical data demonstrating the long-term effects of chronic augmentation and maintenance therapy with Aralast, Glassia, Prolastin and Zemaira are not available

V. **Therapeutic Alternatives:**
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VI. **Recommended Dosing Regimen and Authorization Limit:**

<table>
<thead>
<tr>
<th>Drug</th>
<th>Dosing Regimen</th>
<th>Authorization Limit</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aralast NP</td>
<td>60 mg/kg IV QWK</td>
<td>Length of benefit</td>
</tr>
<tr>
<td></td>
<td>Maximum Infusion Rate:</td>
<td></td>
</tr>
<tr>
<td></td>
<td>0.2 mL/kg/min</td>
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</tr>
<tr>
<td>Zemaira</td>
<td>60 mg/kg IV QWK over 15 minutes</td>
<td>Length of benefit</td>
</tr>
<tr>
<td></td>
<td>Approximate Infusion Rate:</td>
<td></td>
</tr>
<tr>
<td></td>
<td>0.08 mL/kg/min</td>
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</tbody>
</table>
### Prior Authorization Protocol

**ARALAST NP®, GLASSIA®, PROLASTIN-C®, ZEMAIRA® (alpha 1-proteinase inhibitors)**

**NATL**

<table>
<thead>
<tr>
<th>Drug</th>
<th>Dosing Regimen</th>
<th>Authorization Limit</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prolastin-C</td>
<td>60 mg/kg IV QWK over 15 minutes Approximate Infusion Rate: 0.08 mL/kg/min</td>
<td>Length of benefit</td>
</tr>
<tr>
<td>Glassia</td>
<td>60 mg/kg IV QWK over 15 minutes Maximum Infusion Rate: 0.04 mL/kg/min</td>
<td>Length of benefit</td>
</tr>
</tbody>
</table>

#### VII. Product Availability:

- Aralast NP Vial: 0.5 gm lyophilized powder for injection with diluent; 1 gm lyophilized powder for injection with diluent
- Zemaira Vial: 1000 mg lyophilized powder for injection with diluent
- Prolastin-C Vial: 1000 mg lyophilized powder for injection with diluent
- Glassia Vial: 1 gm in 50 mL of solution

#### VIII. References:

Prior Authorization Protocol

ARALAST NP™, GLASSIA™, PROLASTIN-C®, ZEMAIRA® (alpha 1-proteinase inhibitors)

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The materials provided to you are guidelines used by this health plan to authorize, modify, or determine coverage for persons with similar illnesses or conditions. Specific care and treatment may vary depending on individual needs and the benefits covered under your contract.