



## Prior Authorization Approval Criteria

### *Alpha-1 proteinase inhibitor*

<b>Generic name:</b>	Alpha-1 proteinase inhibitor
<b>Brand names:</b>	Aralast, Prolastin, Zemaira
<b>Medication class:</b>	Alpha-1 proteinase inhibitor
<b>FDA-approved uses:</b>	<p>Prolastin: Augmentation therapy in congenital alpha<sub>1</sub>-antitrypsin deficiency with clinical panacinar emphysema</p> <p>Aralast, Zemaira: Chronic augmentation and maintenance therapy in individuals with alpha-1 proteinase inhibitor (A1PI) deficiency and clinical evidence of emphysema</p>
<b>Available dosage forms:</b>	400 mg, 500 mg, 1000 mg lyophilized powder for reconstitution for IV administration
<b>Usual dose:</b>	60 mg/kg IV once weekly. Use within 3 hours after reconstituted product is warmed to room temperature.
<b>Duration of therapy:</b>	<p>Chronic. Maintain AAT above protective threshold of 80 mg/dL (11 mmol/L).</p> <p><i>Note:</i> Zemaira has only been studied for 24 weeks. Clinical data demonstrating the long-term effects of chronic augmentation therapy with Zemaira are not available.</p>

**Cost:**

Aralast:	400 mg = \$204	Cost/month = \$8,568	<b>Annual cost = \$111,384 (3/1/05)</b>
Prolastin:	500 mg = \$175	Cost/month = \$5,880	<b>Annual cost = \$76,440 (3/1/05)</b>
Zemaira:	1000 mg = \$430	Cost/month = \$8,600	<b>Annual cost = \$111,800 (5/1/07)</b>

*Note:* As of 5/1/07, all 3 of these products are billed under the same HCPCS code.

**Criteria for use** (*bullet points below are all inclusive unless otherwise noted*):

- Patient has clinically documented alpha-1 antitrypsin (AAT) deficiency
- Must also have clinical evidence of emphysema

**Criteria for continuation of therapy:** Clinical evidence of efficacy (i.e., elevation of AAT levels)

**Caution:**

- Use caution in patients at risk for fluid overload
- Use Prolastin only in patients with PiZZ, PiZ(null) or Pi(null)(null) alpha-1 antitrypsin deficiency phenotypes
- May carry the risk of transmission of infectious agents

**Monitoring:** A1PI serum levels; vital signs during infusion.

**Contraindications:**

- Hypersensitivity to alpha-1 proteinase inhibitor
- Patients with selective IgA deficiencies (IgA less than 15 mg/dL) who have known antibody against IgA (anti-IgA antibody)

**Not approved if:**

- Above criteria are not met.
- Emphysema is due to environmental triggers.
- Emphysema is caused by tobacco use.

**Special considerations:**

All studies to date have included a small number of patients for a period not exceeding 6 months. AAT augmentation therapy has been shown to increase AAT levels, but the ability to alter/halt progression to emphysema has not been demonstrated. Long-term studies are needed to determine the long term effects of AAT therapy.

FCHP Pharmacy and Therapeutics Committee approval: \_\_\_\_\_

Date: \_\_\_\_\_

Adopted: 04/13/05  
First revision: 12/12/07