

Reference number(s)
1877-A

## **SPECIALTY GUIDELINE MANAGEMENT**

### **Alpha<sub>1</sub>-Proteinase Inhibitors**

**ARALAST NP (alpha<sub>1</sub>-proteinase inhibitor [human])**  
**GLASSIA (alpha<sub>1</sub>-proteinase inhibitor [human])**  
**PROLASTIN-C (alpha<sub>1</sub>-proteinase inhibitor [human])**  
**ZEMAIRA (alpha<sub>1</sub>-proteinase inhibitor [human])**

#### **POLICY**

##### **I. INDICATIONS**

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

##### FDA-Approved Indications

1. Aralast NP  
Chronic augmentation therapy in adults with clinically evident emphysema due to severe congenital deficiency of alpha<sub>1</sub>-proteinase inhibitor (alpha<sub>1</sub>-antitrypsin deficiency)
2. Glassia  
Chronic augmentation and maintenance therapy in adults with clinically evident emphysema due to severe hereditary deficiency of alpha<sub>1</sub>-proteinase inhibitor (alpha<sub>1</sub>-antitrypsin deficiency)
3. Prolastin-C  
Chronic augmentation and maintenance therapy in adults with clinical evidence of emphysema due to severe hereditary deficiency of alpha<sub>1</sub>-proteinase inhibitor (alpha<sub>1</sub>-antitrypsin deficiency)
4. Zemaira  
Chronic augmentation and maintenance therapy in adults with alpha<sub>1</sub>-proteinase inhibitor deficiency and clinical evidence of emphysema

All other indications are considered experimental/investigational and not medically necessary.

##### **II. DOCUMENTATION**

Submission of the following information is necessary to initiate the prior authorization review:

1. Pretreatment serum alpha<sub>1</sub>-antitrypsin (AAT) level
2. Pretreatment post-bronchodilation forced expiratory volume in 1 second (FEV<sub>1</sub>)
3. AAT protein phenotype

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### III. CRITERIA FOR INITIAL APPROVAL

Authorization of 12 months may be granted for treatment of emphysema due to alpha<sub>1</sub>-antitrypsin (AAT) deficiency when all of the following criteria are met:

1. The member's pretreatment serum AAT level is less than 11 micromol/L (80 mg/dL by radial immunodiffusion or 50 mg/dL by nephelometry).<sup>5,6</sup>
2. The member's pretreatment post-bronchodilation forced expiratory volume in 1 second (FEV<sub>1</sub>) is greater than or equal to 25% and less than or equal to 80% of the predicted value.<sup>6</sup>
3. The member has a documented PiZZ, PiZ (null), or Pi (null, null) phenotype (homozygous) AAT deficiency or other phenotype associated with serum AAT concentrations of less than 11 micromol/L (80 mg/dL by radial immunodiffusion or 50 mg/dL by nephelometry).<sup>6</sup>
4. The member does not have the PiMZ or PiMS phenotype AAT deficiency.<sup>7</sup>

### IV. CONTINUATION OF THERAPY

Authorization of 12 months may be granted for continued treatment of emphysema due to alpha<sub>1</sub>-antitrypsin (AAT) deficiency when the member is experiencing beneficial clinical response from therapy.

### V. OTHER

Note: If the member is a current smoker, they should be counseled on the harmful effects of smoking on pulmonary conditions and available smoking cessation options.

### VI. REFERENCES

1. Aralast NP [package insert]. Westlake Village, CA: Baxalta US Inc.; December 2018.
2. Glassia [package insert]. Westlake Village, CA: Baxalta US Inc.; June 2017.
3. Prolastin-C [package insert]. Research Triangle Park, NC: Grifols Therapeutics Inc.; August 2018.
4. Zemaira [package insert]. Kankakee, IL: CSL Behring LLC; April 2019.
5. American Thoracic Society/European Respiratory Society statement: standards for the diagnosis and management of individuals with alpha-1 antitrypsin deficiency. *Am J Respir Crit Care Med.* 2003;168:818-900.
6. Marciniuk DD, Hernandez P, Balter M, et al. Alpha-1 antitrypsin deficiency targeted testing and augmentation therapy: a Canadian Thoracic Society clinical practice guideline. *Can Respir J.* 2012;19:109-116.
7. Sandhaus RA, Turino G, Brantly ML, et al. The diagnosis and management of alpha-1 antitrypsin deficiency in the adult. *Chronic Obstr Pulm Dis.* 2016;3(3):668-82.