

Reference number(s)
1877-A

SPECIALTY GUIDELINE MANAGEMENT Alpha₁-Proteinase Inhibitors

ARALAST NP (alpha₁-proteinase inhibitor [human])
GLASSIA (alpha₁-proteinase inhibitor [human])
PROLASTIN-C (alpha₁-proteinase inhibitor [human])
ZEMAIRA (alpha₁-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 alpha₁-proteinase inhibitor (alpha₁-antitrypsin deficiency)
2. Glassia
Chronic augmentation and maintenance therapy in adults with clinically evident emphysema due to severe hereditary deficiency of alpha₁-proteinase inhibitor (alpha₁-antitrypsin deficiency)
3. Prolastin-C
Chronic augmentation and maintenance therapy in adults with clinical evidence of emphysema due to severe hereditary deficiency of alpha₁-proteinase inhibitor (alpha₁-antitrypsin deficiency)
4. Zemaira
Chronic augmentation and maintenance therapy in adults with alpha₁-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 alpha₁-antitrypsin (AAT) level
2. Pretreatment post-bronchodilation forced expiratory volume in 1 second (FEV₁)
3. AAT protein phenotype or genotype

III. CRITERIA FOR INITIAL APPROVAL

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Authorization of 12 months may be granted for treatment of emphysema due to alpha₁-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).
2. The member's pretreatment post-bronchodilation forced expiratory volume in 1 second (FEV₁) is greater than or equal to 25% and less than or equal to 80% of the predicted value.
3. The member has a documented PiZZ, PiZ (null), or Pi (null, null) (homozygous) AAT deficiency or other phenotype or genotype associated with serum AAT concentrations of less than 11 micromol/L (80 mg/dL by radial immunodiffusion or 50 mg/dL by nephelometry).
4. The member does not have the PiMZ or PiMS AAT deficiency.

IV. CONTINUATION OF THERAPY

Authorization of 12 months may be granted for continued treatment of emphysema due to alpha₁-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]. Lexington, MA: Baxalta US Inc.; December 2018.
2. Glassia [package insert]. Lexington, MA: Takeda Pharmaceuticals US Inc.; March 2022.
3. Prolastin-C Liquid [package insert]. Research Triangle Park, NC: Grifols Therapeutics Inc.; May 2020.
4. Prolastin-C [package insert]. Research Triangle Park, NC: Grifols Therapeutics Inc.; January 2022.
5. Zemaira [package insert]. Kankakee, IL: CSL Behring LLC; September 2022.
6. 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.
7. 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.
8. 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.