Reference number(s)
1877-A

SPECIALTY GUIDELINE MANAGEMENT
Alpha$_1$-Proteinase Inhibitors

ARALAST NP (alpha$_1$-proteinase inhibitor [human])
GLASSIA (alpha$_1$-proteinase inhibitor [human])
PROLASTIN-C (alpha$_1$-proteinase inhibitor [human])
ZEMAIRA (alpha$_1$-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$_1$-proteinase inhibitor (alpha$_1$-antitrypsin deficiency)

2. Glassia
   Chronic augmentation and maintenance therapy in adults with clinically evident emphysema due to severe hereditary deficiency of alpha$_1$-proteinase inhibitor (alpha$_1$-antitrypsin deficiency)

3. Prolastin-C
   Chronic augmentation and maintenance therapy in adults with clinical evidence of emphysema due to severe hereditary deficiency of alpha$_1$-proteinase inhibitor (alpha$_1$-antitrypsin deficiency)

4. Zemaira
   Chronic augmentation and maintenance therapy in adults with alpha$_1$-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$_1$-antitrypsin (AAT) level
2. Pretreatment post-bronchodilation forced expiratory volume in 1 second (FEV$_1$)
3. AAT protein phenotype

III. CRITERIA FOR INITIAL APPROVAL

Authorization of 12 months may be granted for treatment of emphysema due to alpha$_1$-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 (FEV_1) 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) 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).
4. The member does not have the PiMZ or PiMS phenotype AAT deficiency.

IV. CONTINUATION OF THERAPY

Authorization of 12 months may be granted for continued treatment of emphysema due to alpha$_1$-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.