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
III. CRITERIA FOR INITIAL APPROVAL

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) 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 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