Reference number(s)
1877-A

Specialty Guideline Management Alpha1-Proteinase Inhibitors

Products Referenced by this Document

Drugs that are listed in the following table include both brand and generic and all dosage forms and strengths unless otherwise stated. Over-the-counter (OTC) products are not included unless otherwise stated.

Brand Name	Generic Name
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]

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[1-5]

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).

Glassia

Chronic augmentation and maintenance therapy in adults with clinically evident emphysema due to severe hereditary deficiency of alpha1-proteinase inhibitor (alpha1-antitrypsin deficiency).

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).

Zemaira

Chronic augmentation and maintenance therapy in adults with alpha$_1$-proteinase inhibitor deficiency and clinical evidence of emphysema.

Compendial Uses[9]

Acute graft-versus-host disease (GVHD)

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

Documentation

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

Alpha$_1$-proteinase Inhibitor (alpha$_1$-antitrypsin) Deficiency:

- Pretreatment serum alpha$_1$-antitrypsin (AAT) level
- Pretreatment post-bronchodilation forced expiratory volume in 1 second (FEV$_1$)
- AAT protein phenotype or genotype

Coverage Criteria

Alpha$_1$-proteinase Inhibitor (alpha$_1$-antitrypsin) Deficiency[6-8]

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:

- 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).
- 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.
- 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).
- The member does not have the PiMZ or PiMS AAT deficiency.

Acute Graft-Versus-Host Disease (GVHD)[9]

Authorization of 12 months may be granted for the treatment of steroid-refractory acute graft-versus-host disease (GVHD) following hematopoietic stem cell transplantation.

Continuation of Therapy

Alpha$_1$-proteinase Inhibitor (alpha$_1$-antitrypsin) Deficiency

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.

Acute Graft-Versus-Host Disease (GVHD)

All members requesting authorization for continuation of therapy must meet all requirements in the coverage criteria.

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.

References

1. Aralast NP [package insert]. Lexington, MA: Baxalta US Inc.; March 2023.
2. Glassia [package insert]. Lexington, MA: Takeda Pharmaceuticals US Inc.; September 2023.
3. Prolastin-C Liquid [package insert]. Research Triangle Park, NC: Grifols Therapeutics LLC.; May 2020.
4. Prolastin-C [package insert]. Research Triangle Park, NC: Grifols Therapeutics LLC.; January 2022.
5. Zemaira [package insert]. Kankakee, IL: CSL Behring LLC; January 2024.
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.

9. The NCCN Drugs & Biologics Compendium 2024 National Comprehensive Cancer Network, Inc. http://www.nccn.org. Accessed December 16, 2024.