CVS Caremark®

|  |
| --- |
| 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 | alpha1-proteinase inhibitor [human] |
| Glassia | alpha1-proteinase inhibitor [human] |
| Prolastin-C | alpha1-proteinase inhibitor [human] |
| Zemaira | alpha1-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 Indications1-5

#### Aralast NP

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

#### Zemaira

Chronic augmentation and maintenance therapy in adults with alpha1-proteinase inhibitor deficiency and clinical evidence of emphysema.

### Compendial Uses9

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:

Alpha1-proteinase Inhibitor (alpha1-antitrypsin) Deficiency:

* Pretreatment serum alpha1-antitrypsin (AAT) level
* Pretreatment post-bronchodilation forced expiratory volume in 1 second (FEV1)
* AAT protein phenotype or genotype

## Coverage Criteria

### Alpha1-proteinase Inhibitor (alpha1-antitrypsin) Deficiency6-8

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:

* 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

### Alpha1-proteinase Inhibitor (alpha1-antitrypsin) Deficiency

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.

### 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.