Draft Revised Policy: Do Not Implement

Alpha-1 Proteinase Inhibitor (Aralast NP®, Glassia®, Prolastin-C®, Zemaira®)

IMPORTANT REMINDER

We develop Medical Policies to provide guidance to Members and Providers. This Medical Policy relates only to the services or supplies described in it. The existence of a Medical Policy is not an authorization, certification, explanation of benefits or a contract for the service (or supply) that is referenced in the Medical Policy. For a determination of the benefits that a Member is entitled to receive under his or her health plan, the Member's health plan must be reviewed. If there is a conflict between the medical policy and a health plan or government program (e.g., TennCare), the express terms of the health plan or government program will govern.

The proposal is to add text/statements in red and to delete text/statements with strikethrough:

POLICY

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

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

COVERAGE CRITERIA ~~FOR INITIAL APPROVAL~~

Alpha1-proteinase Inhibitor (alpha1-antitrypsin) Deficiency:

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 (FEV_1) 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)

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 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.

MEDICATION QUANTITY LIMITS

Drug Name	Diagnosis	Maximum Dosing Regimen
Aralast NP (Alpha1-Proteinase Inhibitor (Human); Glassia (Alpha1-Proteinase Inhibitor (Human); Prolastin-C(Alpha1-Proteinase Inhibitor (Human); Zemaira (Alpha1-Proteinase Inhibitor (Human))	Alpha1-Antitrypsin Deficiency (Alpha-1 Proteinase Inhibitor Deficiency)	Route of Administration: Intravenous ≥18 year(s) 60mg/kg every week

APPLICABLE TENNESSEE STATE MANDATE REQUIREMENTS

BlueCross BlueShield of Tennessee's Medical Policy complies with Tennessee Code Annotated Section 56-7-2352 regarding coverage of off-label indications of Food and Drug Administration (FDA) approved drugs when the off-label use is recognized in one of the statutorily recognized standard reference compendia or in the published peer-reviewed medical literature.

ADDITIONAL INFORMATION

For appropriate chemotherapy regimens, dosage information, contraindications, precautions, warnings, and monitoring information, please refer to one of the standard reference compendia (e.g., the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) published by the National Comprehensive Cancer Network®, Drugdex Evaluations of Micromedex Solutions at Truven Health, or The American Hospital Formulary Service Drug Information).

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.

EFFECTIVE DATE

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