EVH Clinical Guideline 5003.CC for Alpha$_1$ Proteinase Inhibitors

Guideline Number: EVH_CG_5003.CC	Applicable Codes	
"Evolent" refers to Evolent Health LLC and Evolent Specialty Services, Inc. *© 2024 - 2026 Evolent. All rights Reserved.*		
Original Date: January 2024	**Last Revised Date:** January 2026	**Implementation Date:** March 2026

TABLE OF CONTENTS

STATEMENT

General Information

- *It is the policy of the Health Plan to maintain a prior authorization process that promotes appropriate utilization of specific drugs with potential for misuse or limited indications. This process involves a review using Food and Drug Administration (FDA) criteria to make a determination of Medical Necessity, and approval by the Medical Policy Committee.*
- *If the established criteria are not met, the request is referred to a Medical Director for review, if required for the plan and level of request.*

Purpose

The purpose of this guideline is to define the prior authorization process for the following alpha$_1$ proteinase inhibitors.

Scope

This guideline applies to all practitioners who are involved in providing the requested drug. This guideline is specific to the Health Plan's medical benefit.

INITIAL REVIEW CRITERIA

The request must meet all of the criteria listed below.

- Must be prescribed by, or in consultation with, a pulmonologist
- Must be age 18 years or older
- Must have a confirmed diagnosis of congenital alpha1-antitrypsin deficiency with clinically evident emphysema or airflow obstruction
- Must have an alpha$_1$-antitrypsin phenotype of PI*ZZ, PI*ZNull or PI*NullNull
- Must have a baseline (pretreatment) serum alpha1-antitrypsin concentration of less than 11µmol/L as documented by either of the following:
  - Less than 57mg/dL as determined by nephelometry
  - Less than 80mg/dL as determined by radial immunodiffusion
- Must have a forced expiratory volume (FEV_1) of <65% of predicted value OR a decline of FEV_1 by ≥100 mL/year
- Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc.) listed in the FDA approved labeling
- Must have documentation or attestation from the provider that:
  - The member is currently a non-smoker

- The member does not have selective immunoglobulin (IgA) deficiencies with known antibodies against IgA (anti-IgA antibodies)

REAUTHORIZATION CRITERIA

All prior authorization renewals are reviewed on an annual basis to determine the Medical Necessity for continuation of therapy. The request must meet all of the criteria listed below.

- Must have a positive clinical response (e.g., decreased frequency of exacerbations, slowed rate of FEV1 decline)
- Must remain a non-smoker
- Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc.) listed in the FDA approved labeling

APPROVAL DURATIONS

Initial Authorization	Up to 1 year
Reauthorization	Same as initial

CODING AND STANDARDS

Codes

Code	Brand	Description
J0256	Aralast NP, Prolastin-C, Zemaira	Injection, alpha 1-proteinase inhibitor (human), not otherwise specified, 10mg
J0257	Glassia	Injection, alpha 1 proteinase inhibitor (human), (GLASSIA), 10 mg

Applicable Lines of Business

☐	CHIP (Children's Health Insurance Program)
☐	Commercial
☐	Exchange/Marketplace
☒	Medicaid
☐	Medicare Advantage

BACKGROUND

Aralast NP is indicated for chronic augmentation therapy in patients having congenital deficiency of alpha1-proteinase inhibitor (alpha1-PI) with clinically evident emphysema.

Glassia is indicated for chronic augmentation and maintenance therapy in adults with emphysema due to congenital deficiency of alpha1-proteinase inhibitor (alpha1-PI).

Prolastin-C is indicated for chronic replacement therapy of individuals having congenital deficiency of alpha1-PI (alpha1-antitrypsin deficiency) with clinically demonstrable panacinar emphysema.

Zemaira is indicated for chronic augmentation and maintenance therapy in individuals with alpha1-PI deficiency and clinical evidence of emphysema.

Definitions

Alpha$_1$- Antitrypsin Deficiency – a rare genetic condition characterized by low levels of serum alpha$_1$-antitrypsin (AAT). AAT is a serine protease inhibitor that inhibits neutrophil elastase (NE). Neutrophil elastase degrades elastin and other extracellular matrix components. The imbalance between the AAT and NE increases the risk of emphysema. AAT deficiency also increases the risk of liver disease and several other conditions

POLICY HISTORY

Date	Summary
January 2026	• Converted to new guideline format; no major changes
April 2024	• Added FEV_1 Thresholds for Initial Criteria; Updated Reauthorization criteria
January 2024	• New Guideline

LEGAL AND COMPLIANCE

Guideline Approval

Committee

Reviewed / Approved by Evolent Administrative Services Medical Policy Committee

Disclaimer

Evolent Clinical Guidelines do not constitute medical advice. Treating health care professionals are solely responsible for diagnosis, treatment, and medical advice. Evolent uses Clinical Guidelines in accordance with its contractual obligations to provide utilization management. Coverage for services varies for individual members according to the terms of their health care coverage or government program. Individual members' health care coverage may not utilize some Evolent Clinical Guidelines. Evolent clinical guidelines contain guidance that requires prior authorization and service limitations. A list of procedure codes, services or drugs may not be all inclusive and does not imply that a service or drug is a covered or non-covered service or drug. Evolent reserves the right to review and update this Clinical Guideline in its sole discretion. Notice of any changes shall be provided as required by applicable provider agreements and laws or regulations. Members should contact their Plan customer service representative for specific coverage information.

REFERENCES

1. Silverman EK, Sandhaus RA. Clinical practice. Alpha1-antitrypsin deficiency. N Engl J Med. 2009 Jun 25;360(26):2749-57.
2. 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.
3. Fairbanks KD, Tavill AS. Liver disease in alpha 1-antitrypsin deficiency: a review. Am J Gastroenterol. 2008 Aug;103(8):2136-41
4. Tirado-Conde G, Lara B, Miravitlles M. Augmentation therapy for emphysema due to alpha-1-antitrypsin deficiency. Ther Adv Respir Dis. 2008 Feb;2(1):13-21.
5. Aralast NP (alpha1-proteinase inhibitor [human]) package insert. Cambridge, MA: Takeda Pharmaceuticals USA Inc; May 2025.
6. Zemaira (alpha1-proteinase inhibitor, human) package insert. Kankakee, IL: CSL Behring LLC, January 2024.
7. Prolastin – C (alpha1-proteinase inhibitor [human]) package insert. Research Triangle Park, NC: Grifols Therapeutics Inc: January 2022.
8. Prolastin-C Liquid (alpha1-proteinase inhibitor [human]) package insert. Research Triangle Park, NC: Grifols Therapeutics LLC; May 2020.
9. Glassia [package insert]. Cambridge, MA: Takeda Pharmaceuticals USA Inc; February 2025.
10. Global Initiative for Chronic Obstructive Lung Disease (GOLD). Global Strategy for the Diagnosis, Management and Prevention of Chronic Obstructive Pulmonary Disease: 2024 Report. www.goldcopd.org
11. Alpha-1 antitrypsin deficiency targeted testing and augmentation therapy: a Canadian Thoracic Society clinical practice guideline. Marciniuk DD, Hernandez P, Balter M, Bourbeau J, Chapman KR, Ford GT, Lauzon JL, Maltais F, O'Donnell DE, Goodridge D, Strange C, Cave AJ, Curren K, Muthuri S, Canadian Thoracic Society COPD Clinical Assembly Alpha-1 Antitrypsin Deficiency Expert Working Group Can Respir J. 2012 Mar-Apr;19(2):109-16.