RX.PA.004.CCH ALPHA$_1$ PROTEINASE INHIBITORS (HUMAN)

The purpose of this policy is to define the prior authorization process for Aralast NP, Glassia, Prolastin-C, and Zemaira.

Aralast NP is indicated for chronic augmentation therapy in patients having congenital deficiency of alpha$_1$-proteinase inhibitor (alpha$_1$-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 (alpha$_1$-PI).

Prolastin-C is indicated for chronic replacement therapy of individuals having congenital deficiency of alpha$_1$-PI (alpha$_1$-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 alpha1-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

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.

The drugs, Aralast NP, Glassia, Prolastin-C, and Zemaira, are subject to the prior authorization process.

PROCEDURE

Initial Authorization Criteria:

Must meet all 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 alpha$_1$-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 alpha$_1$-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. Authorization may be extended at 1-year intervals based upon chart documentation of the following:

- Must have a positive clinical response (e.g., decreased frequency of exacerbations, slowed rate of FEV_1 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

Limitations:

Length of Authorization (if above criteria met)	
Initial Authorization	Up to 1 year
Reauthorization	Same as initial

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.

Codes:

CPT Codes / HCPCS Codes / ICD-10 Codes		
Code	Brand	Description
J0256	Aralast NP, Prolastin-C, Zemaira	INJECTION, ALPHA 1-PROTEINASE INHIBITOR (HUMAN), NOT OTHERWISE SPECIFIED, 10 MG
J0257	Glassia	Injection, alpha 1 proteinase inhibitor (human), (GLASSIA), 10 mg

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. Westlake Village, CA: Baxter Healthcare Corporation. May 2007.
6. Zemaira (alpha1-proteinase inhibitor, human) package insert. Kankakee, IL: CSL Behring LLC, January 2007.
7. Prolastin (alpha1-proteinase inhibitor [human]) package insert. Research Triangle Park, NC: Talecris Biotherapeutics, Inc. June 2008.
8. Prolastin-C (alpha1-proteinase inhibitor [human]) package insert. Research Triangle Park, NC: Talecris Biotherapeutics, Inc. October 2009.
9. Glassia [package insert]. Westlake Village, CA: Baxter Healthcare; August 2010.
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.

Revision History

DESCRIPTION OF REVIEW / REVISION	DATE APPROVED
New Policy	04/2024
Added FEV1 Thresholds for Initial Criteria; Updated Reauthorization Criteria	XX/XX

Record Retention

Records Retention for Evolent Health documents, regardless of medium, are provided within the Evolent Health records retention policy and as indicated in CORP.028.E Records Retention Policy and Procedure.

Disclaimer

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CountyCare reserves the right to review and update the medical payment and prior authorization guidelines in its sole discretion. Notice of such changes, if necessary, shall be provided in accordance with the terms and conditions of provider agreements and any applicable laws or regulations.

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