Alpha1-Proteinase Inhibitors

NOTICE

This policy contains information which is clinical in nature. The policy is not medical advice. The information in this policy is used by Wellmark to make determinations whether medical treatment is covered under the terms of a Wellmark member's health benefit plan. Physicians and other health care providers are responsible for medical advice and treatment. If you have specific health care needs, you should consult an appropriate health care professional. If you would like to request an accessible version of this document, please contact customer service at 800-524-9242.

BENEFIT APPLICATION

Benefit determinations are based on the applicable contract language in effect at the time the services were rendered. Exclusions, limitations or exceptions may apply. Benefits may vary based on contract, and individual member benefits must be verified. Wellmark determines medical necessity only if the benefit exists and no contract exclusions are applicable. This policy may not apply to FEP. Benefits are determined by the Federal Employee Program.

DESCRIPTION

The intent of the Alpha1-Proteinase Inhibitors drug policy is to ensure appropriate selection of patients for therapy based on product labeling, clinical guidelines and clinical studies while steering utilization to the most cost-effective medication within the therapeutic class. For this program, Prolastin-C is the preferred product. The criteria will require the use of the health plan's preferred product before the use of targeted products (Aralast NP, Glassia, Zemaira), unless there are clinical circumstances that exclude the use of the preferred products and may be based on previous use of a product. The coverage review process will ascertain situations where a clinical exception can be made. This program applies to all members requesting treatment with a non-preferred product.

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

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

3. 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)

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

Table. Alpha1-Proteinase Inhibitor Products

Medication	Generic Name
Preferred Products:	
Prolastin-C	alpha1-proteinase inhibitor [human]
Targeted Products:	
Aralast NP	alpha1-proteinase inhibitor [human]
Glassia	alpha1-proteinase inhibitor [human]
Zemaira	alpha1-proteinase inhibitor [human]

POLICY

Documentation

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

- Pretreatment serum alpha$_1$-antitrypsin (AAT) level
- Pretreatment post-bronchodilation forced expiratory volume in 1 second (FEV_1)
- AAT protein phenotype

Criteria for Initial Approval

I. 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 predicted.
   - The member has a documented PiZZ, PiZ (null), or Pi (null, null) phenotype (homozygous) AAT deficiency or other phenotype 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 phenotype AAT deficiency.
   - The member has experienced an intolerable adverse event with the preferred product, Prolastin-C, if requesting Aralast-NP, Glassia, or Zemaira

Continuation of Therapy

I. 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.

Prior approval is required. Submit a prior approval/treatment request now.

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.

Dosing and Administration

Approvals may be subject to dosing limits in accordance with FDA-approved labeling, accepted compendia, and/or evidence-based practice guidelines.

PROCEDURES AND BILLING CODES

To report provider services, use appropriate CPT* codes, Alpha Numeric (HCPCS level 2) codes, Revenue codes, and/or ICD diagnostic codes.

- J0256 – Inj, alpha 1 proteinase inhibitor (human), not otherwise specified, 10 mg (Prolastin-C, Aralast NP, Zemaira)
- J0257 – Inj, alpha 1 proteinase inhibitor (human), 10 mg (Glassia)

REFERENCES

- Aralast NP [package insert]. Westlake Village, CA: Baxalta US Inc.; September 2015.
- Glassia [package insert]. Westlake Village, CA: Baxalta US Inc.; June 2017.
- Prolastin-C [package insert]. Research Triangle Park, NC: Grifols Therapeutics Inc.; September 2017.
- Zemaira [package insert]. Kankakee, IL: CSL Behring LLC; September 2015.
- 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.
- 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.
- 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.

POLICY HISTORY

Policy #: 05.02.39
Policy Creation: January 2018
Reviewed: April 2020
Revised: January 2020
Current Effective Date: January 1, 2020