RX.PA.004.MPC Alpha$_1$-Proteinase Inhibitors (Human): Aralast NP™, Glassia™, Prolastin-C®, Zemaira®

The purpose of this policy is to define the prior authorization process for Alpha$_1$ Proteinase Inhibitors.

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 alpha$_1$-proteinase inhibitor (alpha$_1$-PI).

Prolastin-C® is indicated for chronic augmentation and maintenance therapy in adults with emphysema due to deficiency of alpha$_1$-proteinase inhibitor (alpha$_1$-antitrypsin deficiency).

Zemaira® is indicated for chronic augmentation and maintenance therapy in individuals with alpha$_1$-PI deficiency and clinical evidence of emphysema.

The recommendations are based upon the American Thoracic Society/European Respiratory Society Statement.

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.

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

PROCEDURE

A. Initial Authorization Criteria:

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 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 50mg/dL as determined by nephelometry
  - Less than 80mg/dL as determined by radial immunodiffusion
- Must be a non-smoker or has quit smoking for at least 6 months
- Prescriber attests that conventional therapy for emphysema will continue to be used concurrently
- Must not have selective IgA deficiencies with known antibodies against IgA (anti-IgA antibodies)

B. Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc) listed in the FDA approved labeling.

C. Alpha 1-Proteinase Inhibitors will be considered investigational or experimental for any other use and will not be covered.

D. <u>Reauthorization Criteria:</u>

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 all the following:

- MPC Renewal:
  - Documentation from the provider that the member remains a candidate for treatment with an Alpha 1-Proteinase Inhibitor based upon the prescriber's assessment while on therapy
  - Chart documentation showing medication prescribed by or in consultation with a pulmonologist
  - Chart documentation showing member is experiencing positive response to therapy
- Non- MPC Renewal:
  - Members who have previously been taking an Alpha 1-Proteinase Inhibitor and are requesting a non-MPC renewal should be considered under criterion A (Initial Authorization Criteria).
  - Member has not been receiving medication samples for an Alpha 1-Proteinase Inhibitor; AND
  - Provider has a documented clinical response showing member is experiencing positive response to therapy

Limitations:

Length of Authorization (if above criteria met)	
Initial Authorization	6 months
Reauthorization	12 months

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.

HCPCS Code(s):

Code	Description
J0256	Injection, alpha 1-proteinase inhibitor (human), not otherwise specified, 10 mg
J0257	Injection, alpha 1 proteinase inhibitor (human), (GLASSIA), 10 mg

REFERENCES

1. Silverman EK, Sandhaus RA. Clinical practice. Alpha$_1$-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 (alpha$_1$-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.

REVIEW HISTORY

DESCRIPTION OF REVIEW / REVISION	DATE APPROVED
Update to reauthorization section with MPC vs Non-MPC renewal criteria and change to authorization/reauthorization timeframes (6 months and 12 months)	*09/2022*
Annual review	*02/2022*

Addition of dosing requirements and off-label restrictions	*12/2021*
P&T Review	*11/2020*