Geisinger Health Plan

POLICIES AND PROCEDURE MANUAL

Policy: MBP 43.0

Section: Medical Benefit Pharmaceutical Policy

Subject: Alpha 1-Antitrypsin Inhibitor Therapy (Prolastin-C, Aralast, Zemaira, Glassia)

I. Policy:
Alpha 1-Antitrypsin Inhibitor Therapy (Prolastin-C, Aralast, Zemaira, Glassia)

II. Purpose/Objective:
To provide a policy of coverage regarding Alpha 1-Antitrypsin Inhibitor Therapy (Prolastin-C, Aralast, Zemaira, Glassia)

III. Responsibility:
A. Medical Directors
B. Medical Management
C. Pharmacy Department

IV. Required Definitions
1. Attachment – a supporting document that is developed and maintained by the policy writer or department requiring/authoring the policy.
2. Exhibit – a supporting document developed and maintained in a department other than
3. the department requiring/authoring the policy.
4. Devised – the date the policy was implemented.
5. Revised – the date of every revision to the policy, including typographical and grammatical changes.
6. Reviewed – the date documenting the annual review if the policy has no revisions necessary.

V. Additional Definitions
Medical Necessity or Medically Necessary means Covered Services rendered by a Health Care Provider that the Plan determines are:

a. appropriate for the symptoms and diagnosis or treatment of the Member's condition, illness, disease or injury;
b. provided for the diagnosis and the direct care and treatment of the Member's condition, illness disease or injury;
c. in accordance with current standards good medical treatment practiced by the general medical community;
d. not primarily for the convenience of the Member, or the Member's Health Care Provider; and
e. the most appropriate source or level of service that can safely be provided to the Member. When applied to hospitalization, this further means that the Member requires acute care as an inpatient due to the nature of the services rendered or the Member's condition, and the Member cannot receive safe or adequate care as an outpatient

Medicaid Business Segment
Medical Necessity shall mean a service or benefit that is compensable under the Medical Assistance Program and if it meets any one of the following standards:

(i) the service or benefit will, or is reasonably expected to, prevent the onset of an illness, condition or disability.

(ii) the service or benefit will, or is reasonably expected to, reduce or ameliorate the physical, mental or development effects of an illness, condition, injury or disability.

(iii) the service or benefit will assist the Member to achieve or maintain maximum functional capacity in performing daily activities, taking into account both the functional capacity of the Member and those functional capacities that are appropriate for members of the same age

DESCRIPTION:
Alpha 1-antitrypsin (AAT) deficiency is an autosomal co-dominant genetic condition that may predispose people with the condition to emphysema in adult life, and liver disease in adults and children. In rare cases, alpha-1 antitrypsin deficiency also causes a skin condition characterized by hardened skin with painful lumps or patches.

CRITERIA FOR USE: Requires Prior Authorization by Medical Director or Designee

Alpha 1-antitrypsin inhibitor therapy will be considered medically necessary when all of the following criteria are met:

Diagnosis of emphysema secondary to alpha 1-antitrypsin deficiency when **all** of the following criteria are met:

- The insured individual has a diagnosis of panacinar emphysema and a documented decline in forced expiratory volume in 1 second (FEV_1) in spite of optimal medical therapy (e.g., bronchodilators, inhaled or oral corticosteroids, oxygen, if indicated); **AND**

- The insured individual has a phenotype* associated with causing serum alpha 1-antitrypsin of less than 80 mg/dL [PiZZ, PiZ(*null*),or Pi(*null)(null*)]; **AND**

- The insured individual has an alpha 1-antitrypsin serum level below the value of 35% of normal (less than 80 mg/dL), which is considered to be the threshold thought to protect against emphysema; **AND**

- The insured individual is a non-smoker or ex-smoker **(This criterion is not applicable to Medicare business segment)**

***Note**:

Phenotype		Risk	Commercial standard plasma level, mg/dL
MM	The most common allele. PI*MM homozygotes have no increased risk for developing AAT deficiency-associated lung disease	No increased risk	150-350
MZ	"Carrier" status	Possible mild increased risk	90-210
SS	The homozygous form is associated with plasma levels about 60% of normal.	No increased risk	100-200
SZ	SZ phenotypes are at increased risk of developing AAT deficiency-associated diseases	Mild increased risk (20-50%)	75-120
ZZ	The most frequent deficient AAT allele. ZZ homozygotes have plasma levels of AAT that are about 15% of the normal plasma concentration and are at the greatest risk for developing AAT deficiency-associated lung disease	High risk (80-100%)	20-45
null	(null)(null) or Z(null) homozygotes are associated with the most severe deficiency, producing no active AAT, or less than 1% of the normal amount of plasma AAT	High risk (100% by age 30 yrs)	0

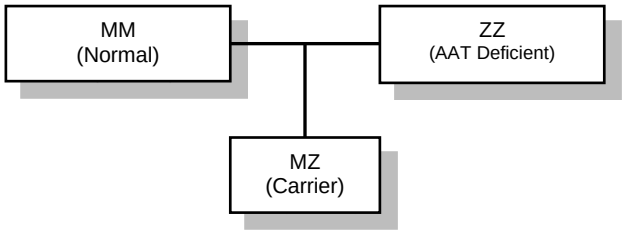
Adapted from: American Thoracic Society. Am Rev Respir Dis 1989;140:1494

AUTHORIZATION DURATION: Initial approval will be for **12 months** or less if the reviewing provider feels it is medically appropriate. Subsequent approvals will be for an additional **12 months** or less if the reviewing provider feels it is medically appropriate and will require medical record documentation of continued disease improvement or lack of disease progression. The medication will no longer be covered if patient experiences toxicity or worsening of disease.

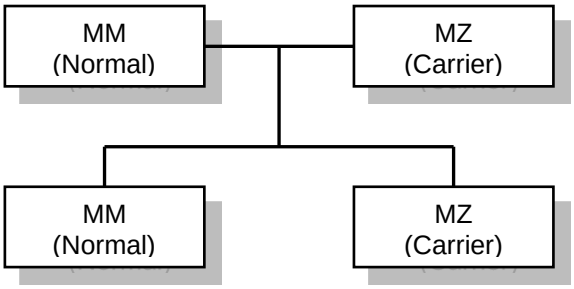
LIMITATIONS: There is insufficient evidence in the published, peer-reviewed medical literature of any proven value in initiating alpha 1-antitrypsin inhibitor replacement therapy in affected individuals who lack clinical evidence of emphysema. The use of alpha 1-antitrypsin augmentation therapy in this population is considered experimental, investigational or unproven, and is **NOT COVERED.**

Additional information:
The greatest risk of emphysema occurs in those individuals with the rare homozygous null phenotype who completely lack Alpha 1-antitrypsin [Pi(null) (null)] or those with other rare phenotypes who produce only 1 to 2% of normal levels [PiZ(null)].

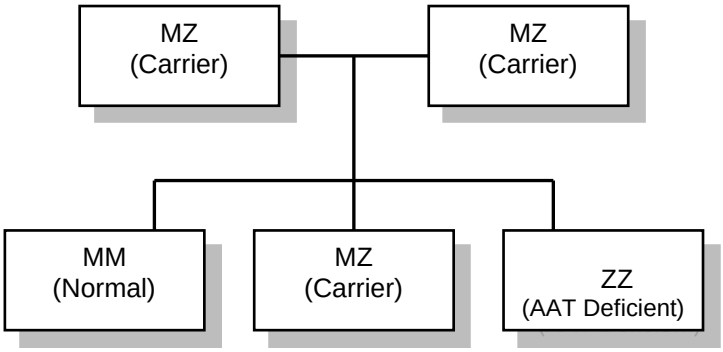
A person who inherits a normal gene, *phenotype* [PiMM], from one parent and an AAT deficient gene, (PiZZ *phenotype)* from the other parent becomes a *carrier* of the AAT-deficient gene. Few if any symptoms will appear in a person said to be a *carrier* of the gene.



If one parent is a carrier phenotype, and the other is normal phenotype, they have a 50/50 chance of having a carrier child.



If both parents are *carriers*, they can pass down normal, carrier or AAT-deficient genes to their children.



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LINE OF BUSINESS:
Eligibility and contract specific benefit limitations and/or exclusions will apply. Coverage statements found in the line of business specific benefit document will supersede this policy.

This policy will be revised as necessary and reviewed no less than annually**.**

Devised: 10/2006

Revised: 08/14, 03/24/15

Reviewed: 02/08, 3/09, 5/10, 6/11, 08/14, 3/31/16, 1/31/17, 10/31/17, 9/28/18, 8/29/19, 8/26/20, 8/19/21