DESCRIPTION:

Prolastin-C®, Aralast NP®, Zemaira®, and Glassia® are all FDA approved agents for use as replacement therapy in congenital alpha-1 antitrypsin (AAT) deficiency with clinical emphysema. AAT deficiency is an autosomal recessive genetic disorder that results in decreased levels of the protease inhibitor alpha-1 antitrypsin. When circulating levels of AAT drop below a minimal protective level the alveolar walls are damaged from excess protease. Patients with this disorder develop early onset panacinar emphysema (i.e. affecting all parts of the lobules). They are also at risk for developing chronic liver disease (hepatitis, cirrhosis), panniculitis (an inflammation of the layer of fat beneath the skin) and vasculitis. AAT deficiency represents about 3% of all emphysema cases reported in the US.

POLICY:

Based upon our criteria and review of the peer-reviewed literature, treatment with Prolastin-C®, Aralast NP®, Zemaira®, and Glassia® administered in accordance with FDA guidelines, has been medically proven to be effective and therefore, appropriate if all of the following criteria are met:

1. Patient must be followed by and have a prescription written by a pulmonologist AND
2. Patient must currently be a non-smoker documented by a negative cotinine urine test.  
   a. If using nicotine replacement products but no longer smoking, then urine anabasine measurements should also be ordered and must be negative AND
3. Patient must be a PiZZ, PiSZ, PiZ(null) or Pi(null)(null), as they are at the greatest risk for developing panacinar emphysema AND
4. Treatment should only be initiated when patient’s alpha 1-antitrypsin (AAT) levels are less than 11 mmol/L OR 80mg/dL AND have documented evidence of emphysema as FEV1< 65% of predicted value. Augmentation therapy is NOT recommended for patients without symptomatic emphysema AND
5. Patients should demonstrate 1 or more of the following: signs of significant lung disease such as chronic productive cough or unusual frequency of lower respiratory infection, airflow obstruction, accelerated decline of FEV1 or chest radiograph or CT scan evidence of emphysema, especially in the absence of a recognized risk factor (smoking, occupational dust exposure, etc)

POLICY GUIDELINES:

1. Patients MUST have clinically demonstrable panacinar emphysema
2. Patients with emphysema due to AAT deficiency should be maintained on regimens similar to those patients with emphysema not associated with AAT deficiency, including: maximum doses of beta-adrenergic bronchodilators, anticholinergics and antibiotics, when appropriate. Patients should also have vaccinations against influenza and pneumococcus and supplemental oxygen therapy when indicated.
SUBJECT: Alpha-1 Antitrypsin Therapy (AAT); Alpha-1 Proteinase Inhibitors (human): Prolastin-C®, Zemaira®, Aralast NP® and Glassia®

POLICY NUMBER: Pharmacy-02
EFFECTIVE DATE: 6/05
REVIEW DATE: 01/15, 08/13, 05/13, 03/13, 11/11, 7/10, 7/09, 5/08, 9/07, 10/06, 9/05, 8/06
Last Updated: 1/2/2015

If the member’s subscriber contract excludes coverage for a specific service or prescription drug, it is not covered under that contract. In such cases, medical or drug policy criteria are not applied. Medical or drug policies apply to commercial, SafetyNet, and Health Care Reform products only when a contract benefit for the specific service exists.

3. Treatment will only be covered when administered as an IV infusion.
4. Safety and effectiveness in children has not been established

Exclusion Criteria:
The use of alpha-1 Antitrypsin therapy will not be covered in any of the following situations:
1. Active smokers
2. Treatment of cystic fibrosis
3. Current non-smokers who start smoking after initial approval can be denied further treatment
4. Augmentation therapy will not be granted for PiMZ heterozygotes or other AAT deficiencies (i.e. PiMM)

RATIONALE:

CODES:
Number Description
Eligibility for reimbursement is based upon the benefits set forth in the member’s subscriber contract. Codes may not be covered under all circumstances. Please read the policy and guidelines statements carefully. Codes may not be all inclusive as the AMA and CMS code updates may occur more frequently than policy updates. Code Key: Experimental/Investigational = (E/I), Not medically necessary/ appropriate = (NMN).

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HCPCS:
J0256 Aralast NP, Prolastin-C, Zemaira, Glassia

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