The following policy is for medication that falls under the Medicare Part D benefit only.

Prior Authorization Group: Alpha1-Antitrypsin Replacement Therapy Policy

Drug(s): ARALAST (proteinase inhibitor), PROLASTIN (proteinase inhibitor), ZEMAIRA (proteinase inhibitor)

Covered Uses: Emphysema due to alpha1–antitrypsin (AAT) deficiency. All criteria in "other criteria" section must be met for coverage.

Required Medical Information: AAT serum level, pulmonary function tests including FEV1, smoking status, previous and current therapies. Medical history.

Age Restrictions: N/A

Prescriber Restrictions: Must be ordered or followed by a pulmonologist

Other Criteria: Coverage is provided when documentation submitted identifies emphysema due to AAT deficiency and all of the following criteria are met:

1. AAT serum level less than 11 µM or less than 80mg/dL
2. Progressive clinically evident emphysema with a documented rate of decline in forced expiratory volume in 1 second (FEV1) post bronchodilation between 30 and 65% predicted except when:
    a. Nearly normal pulmonary function if they experience a rapid decline in lung function (FEV1 greater than 120 ml/yr) **OR**
    b. Poor lung function and currently receiving standard treatment
3. Non-smoker
4. Phenotype is identified as PiZZ, PiZ(null) or Pi(null)(null)
5. Failed optimal alternate pharmacologic and non-pharmacologic therapies (e.g. bronchodilators, inhaled steroids if appropriate, oxygen, pulmonary rehabilitation, etc.).

Continued therapy will be considered up to a maximum of 12 month intervals based on demonstrated response in slowing progression of lung function decline AND member compliance with all pharmacologic therapies.

Exclusion Criteria: Not covered if any of the following situations are true:
1. PiMZ or PiMS phenotypes
2. Members identified with selective IgA deficiencies (IgA level less than 15mg/dl) that have known antibodies against IgA, since they may experience severe reactions
3. Dosing exceeding package labeling
4. Frequency exceeding once weekly infusions
5. Member not compliant with therapy. Coverage is not provided for doses exceeding package labeling.

Coverage Duration: 3 months initial approval. Twelve months for extension of therapy.