



## Alpha-1-Proteinase Inhibitors Infusion Therapy

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### Related Medical Guideline

[Off-Label Use of FDA-Approved Drugs and Biologicals](#)

### Dosage/Administration

Indication	Dose
All Indications	60 mg/kg by intravenous (IV) infusion administered once every 7 days

### Length of Authorization

Coverage will be provided for 12 months and may be renewed.

### Applicable Products

[Aralast NP®](#)

[Glassia®](#)

[Prolastin®](#)

[Prolastin®-C](#)

[Zemaira®](#)

## I. Initial Approval Criteria

Alpha-1-Proteinase Inhibitors are considered medically necessary for emphysema secondary to alpha-1-antitrypsin (ATT) deficiency when the following criteria are met; all:

1. Presence AAT deficiency with PiZZ, PiZ (null) or Pi (null,null) phenotypes
2. Presence of AAT deficiency and clinical evidence of panacinar emphysema
3. Low serum AAT concentration ( $\leq 11$  uM/L [35% of normal] or  $\leq 80$  mg/dL [measured by radial immunodiffusion] or  $\leq 0.8$  g/L [measured by nephelometry])
4. Member is a non-smoker

## II. Renewal Criteria

Coverage will be given for 12 months and is eligible for renewal when the following criteria is met:

1. Patient continues to meet INITIAL APPROVAL CRITERIA.
2. Positive response to treatment (defined by elevation of AAT levels above baseline and/or substantial reduction in deterioration-rate of lung function, as measured by percent-predicted FEV1)
3. Absence of unacceptable drug-toxicity (i.e., hypersensitivity reactions)

## Limitations/Exclusions

1. Alpha-1-Proteinase Inhibitors are considered investigational and not medically necessary for any indications other than those listed above due to insufficient evidence of therapeutic value.

## Revision History

4/1/18	Added dose and administration table, Added length of authorization, Extended length of renewal approval from 6 months to 12 months
1/12/18	added prerequisite that member must be a non-smoker

## Applicable Procedure Codes

J0256	Injection, alpha 1-proteinase inhibitor (human), not otherwise specified, 10 mg
J0257	Injection, alpha 1 proteinase inhibitor (human), (GLASSIA), 10 mg
S9346	Home infusion therapy, alpha-1-proteinase inhibitor (e.g., Prolastin); administrative services, professional pharmacy services, care coordination, and all necessary supplies and equipment (drugs and nursing visits coded separately), per diem

## Applicable Diagnosis Codes

E88.01	Alpha-1-antitrypsin deficiency
J43.0	Unilateral pulmonary emphysema [MacLeod's syndrome]
J43.1	Panlobular emphysema
J43.2	Centrilobular emphysema
J43.8	Other emphysema
J43.9	Emphysema, unspecified

## References

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3. Aralast NP [package insert]. Westlake Village, CA; Baxter Healthcare; March 2014. Accessed May 2015.
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6. American Thoracic Society/European Respiratory Society Statement: Standards for the Diagnosis and Management of Individuals with Alpha-1 Antitrypsin Deficiency approved by the ATS Board of Directors, December 2002, and by the ERS Executive Committee, February 2003.  
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