

Market Applicability														
Market	DC	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	NA

\*FHK- Florida Healthy Kids

## Alpha-1 Proteinase Inhibitors

Override(s)	Approval Duration
Prior Authorization	1 Year

Medications
Aralast NP (alpha-1 proteinase inhibitor)
Glassia (alpha-1 proteinase inhibitor)
Prolastin-C (alpha-1 proteinase inhibitor)
Zemaira (alpha-1 proteinase inhibitor)

### APPROVAL CRITERIA

Requests for alpha-1 proteinase inhibitor therapy may be approved if the following criteria are met:

- I. Individual has a diagnosis of congenital alpha-1 antitrypsin deficiency (alpha-1 proteinase inhibitor deficiency); **AND**
- II. Individual has a confirmed alpha-1 antitrypsin level is less than or equal to 11 µmol/L ((approximately equivalent to 80 mg/dL measured by radial immunodiffusion or 57 mg/dL measured by nephelometry) (ATS/ERS, 2003; Stoller, 2017); **AND**
- III. Individual is currently a non-smoker (ATS/ERS, 2003; CTS, 2013); **AND**
- IV. Individual has clinically evident emphysema; **AND**
- V. One of the following:
  - A. Individual has moderate airflow obstruction evidenced by a forced expiratory volume (FEV<sub>1</sub>) of 30-65% of predicted value prior to initiation of therapy (ATS/ERS, 2003); **OR**
  - B. Individual has a rapid decline in lung function as measured by a change in FEV<sub>1</sub> greater than 120 ml/year (ATS/ERS, 2003).

Alpha-1 proteinase inhibitor therapy may **not** be approved for the following:

- I. Individuals with IgA antibodies.

This policy does not apply to health plans or member categories that do not have pharmacy benefits, nor does it apply to Medicare. Note that market specific restrictions or transition-of-care benefit limitations may apply.

Market Applicability														
Market	DC	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	NA

\*FHK- Florida Healthy Kids

State Specific Mandates		
State name	Date effective	Mandate details (including specific bill if applicable)
N/A	N/A	N/A

**Key References:**

1. 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(7):818-900.
2. Clinical Pharmacology [database online]. Tampa, FL: Gold Standard, Inc.: 2018. URL: <http://www.clinicalpharmacology.com>. Updated periodically.
3. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. <http://dailymed.nlm.nih.gov/dailymed/about.cfm>. Accessed: September 24, 2018.
4. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
5. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2018; Updated periodically.
6. Marciniuk DD, Hernandez P, Balter M, et al. Alpha-1 antitrypsin deficiency targeted testing and augmentation therapy: a Canadian Thoracic Society clinical practice guideline. Can Respir J. 2012; 19(2):109-116.
7. Stoller JK. Clinical manifestations, diagnosis, and natural history of alpha-1 antitrypsin deficiency. Updated: May 25, 2018. In: UpToDate, Post TW (Ed), UpToDate, Waltham, MA. Accessed: September 26, 2018.
8. Stoller JK. Treatment of alpha-1 antitrypsin deficiency. Updated: April 20, 2018. In: UpToDate, Post TW (Ed), UpToDate, Waltham, MA. Accessed: September 26, 2018.
9. Stoller JK, Lacbawan FL, Aboussouan LS. Alpha-1 antitrypsin deficiency. GeneReviews (online). University of Washington, Seattle. Updated: January 19, 2017. Available at: <http://www.ncbi.nlm.nih.gov/books/NBK1519/?report=classic>. Accessed: September 25, 2018.

This policy does not apply to health plans or member categories that do not have pharmacy benefits, nor does it apply to Medicare. Note that market specific restrictions or transition-of-care benefit limitations may apply.