Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

Alpha-1 Proteinase Inhibitors

Override(s)	Approval Duration
Prior Authorization	1 Year

Medications	Dosing Limit
Aralast NP (alpha-1 proteinase inhibitor)	60 mg/kg once per week
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 confirmedalpha-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_1) 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_1 greater than 120 ml/year (ATS/ERS, 2003).

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

I. All other indications not included above; **OR**
II. Individuals with IgA antibodies.

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

State Specific Mandates		
State name N/A	Date effective N/A	Mandate details (including specific bill if applicable) 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. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: September 20, 2019.
3. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
4. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2019; Updated periodically.
5. 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.
6. 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 22, 2019.
7. Stoller JK. Treatment of alpha-1 antitrypsin deficiency. Updated: February 5, 2019. In: UpToDate, Post TW (Ed), UpToDate, Waltham, MA. Accessed: September 22, 2019.
8. 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 22, 2019.