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

Initial 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. Documentation is provided that individual's 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 has clinically evident emphysema (or chronic obstructive pulmonary disease [COPD]); **AND**
IV. Individual is currently a non-smoker (ATS/ERS, 2003; CTS, 2013); **AND**
V. One of the following:
    A. Documentation is provided that 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. Documentation is provided that individual has a rapid decline in lung function as measured by a change in FEV_1 greater than 120 ml/year (ATS/ERS, 2003).

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

I. Documentation is provided that there is clinically significant improvement or stabilization in clinical signs and symptoms of disease (including but not limited to decreased frequency of exacerbations, slowed rate of FEV_1 decline, preservation of CT scan lung density or improvement in symptom burden); **AND**
II. Individual remains a non-smoker.

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

I. Individuals with IgA antibodies; **OR**

II. May not be approved when the above criteria are not met and for all other indications.

Key References:

1. American Thoracic Society(ATS)/European Respiratory Society (ERS) 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 30, 2022.
3. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
4. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2022; 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: September 13, 2022. In: UpToDate, Post TW (Ed), UpToDate, Waltham, MA. Accessed: October 1, 2022.
7. Stoller JK. Treatment of alpha-1 antitrypsin deficiency. Updated: November 4, 2021. In: UpToDate, Post TW (Ed), UpToDate, Waltham, MA. Accessed: October 1, 2022.
8. Stoller JK, Hupertz V, Aboussouan LS. Alpha-1 Antitrypsin Deficiency. GeneReviews (online). University of Washington, Seattle. Updated: May 21, 2020. Available at: http://www.ncbi.nlm.nih.gov/books/NBK1519/?report=classic. Accessed: October 4, 2022.