

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

CG-DRUG-92

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

Augmentation therapy with intravenous alpha-1 proteinase inhibitors (Aralast NP, Glassia, Prolastin-C, and Zemaira) may be approved for adults with congenital alpha-1 antitrypsin deficiency when all of the following criteria are met:

- I. Confirmation that alpha-1 antitrypsin level is less than or equal to 11  $\mu\text{mol/L}$ \*; **AND**
- II. Individual is currently a non-smoker; **AND**
- III. Individual has clinically evident emphysema; **AND**
- IV. One of the following:
  - A. Moderate airflow obstruction is evidenced by forced expiratory volume ( $\text{FEV}_1$ ) of 30-65% of predicted value, prior to initiation of therapy; **OR**
  - B. Individual has a rapid decline in lung function as measured by a change in  $\text{FEV}_1$  greater than 120 ml/year.

\*Note: Serum levels of alpha-1 antitrypsin can be assessed by immunodiffusion, immune turbidimetry, rocket immunoelectrophoresis, or nephelometry. Given the variations in testing modalities, it is essential to know the range of normal values for the test used in a given individual. For example, 11  $\mu\text{mol/L}$  is approximately equivalent to a commercial standard level of 80 mg/dL by immunodiffusion or 57 mg/dL by nephelometry.

Use of alpha-1 proteinase inhibitors (Aralast NP, Glassia, Prolastin-C, and Zemaira) may **not** be approved for 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.

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Use of alpha-1 proteinase inhibitors (Aralast NP, Glassia, Prolastin-C, and Zemaira) is considered investigational and may **not** be approved when the criteria above are not met and for all other indications including, but not limited to:

- I. Bronchopulmonary dysplasia;
- II. Cystic fibrosis;
- III. Diabetes mellitus;
- IV. Graft versus host disease (GVHD);
- V. Post-lung transplantation for acute rejection or infection episodes.

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. 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. Aralast NP (Baxter Healthcare Corp., Westlake Village, CA) Updated September 2017. Available at: <https://dailymed.nlm.nih.gov/dailymed/drugInfo.cfm?setid=a9a5b46e-04da-41bd-bb5f-c4936b664fef>. Accessed on February 28, 2018.
3. Chen S, Farahati F, Marciniuk D, et al. Human  $\alpha$ 1-proteinase inhibitor for patients with  $\alpha$ 1-antitrypsin deficiency [Technology report no 74]. Ottawa: Canadian Agency for Drugs and Technologies in Health; 2007.
4. Glassia (Kamada Ltd., Israel; Baxter Healthcare Corp., Westlake Village, CA). Updated June 2017. Available at: <https://dailymed.nlm.nih.gov/dailymed/drugInfo.cfm?setid=83473cbb-48e4-42a2-81b6-4c851423da7b>. Accessed on February 28, 2018.
5. Gøtzsche PC, Johansen HK. Intravenous alpha-1 antitrypsin augmentation therapy for treating patients with alpha-1 antitrypsin deficiency and lung disease. Cochrane Database Syst Rev. 2010;(7):CD007851.
6. Marciniuk DD, Hernandez P, Balter M, et al.; Canadian Thoracic Society COPD Clinical Assembly Alpha-1 Antitrypsin Deficiency Expert Working Group. 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. Prolastin C [Product Information]. (Talecris Biotherapeutics, Inc., Research Triangle Park, NC) August 2017. Available at <https://dailymed.nlm.nih.gov/dailymed/drugInfo.cfm?setid=91edab72-c889-470e-8315-1798b5548dca>. Accessed on February 28, 2018.
8. Russi EW, Karrer W, Brutsche M, et al.; Swiss Respiratory Society. Diagnosis and management of chronic obstructive pulmonary disease: the Swiss guidelines. Official guidelines of the Swiss Respiratory Society. Respiration. 2013; 85(2):160-174.

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9. Stoller JK, Lachawan FL, Aboussouan. 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 on February 28, 2018.
10. Zemaira [Product Information]. (CSL Behring LLC, Kankakee, IL). Updated Nov 2017. Available at: <https://dailymed.nlm.nih.gov/dailymed/drugInfo.cfm?setid=0c3354b5-a1d8-4f98-ad55-2eafe4265c4e>. Accessed on February 28, 2018.

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