

## Alpha-1-Proteinase Inhibitors: Aralast NP®, Glassia®, Prolastin®-C, Zemaira® (Intravenous)

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### I. Length of Authorization

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

### II. Dosing Limits

#### A. Quantity Limit (max daily dose) [Pharmacy Benefit]:

- Aralast NP 1 g/50 mL: 7 vials per week
- Aralast NP 0.5 g/25 mL: 1 vial per week
- Glassia 1 g/50 mL: 7 vials per week
- Prolastin-C 1 g/20 mL: 7 vials per week
- Prolastin-C Liquid 1g/20 mL: 7 vials per week
- Zemaira 1 g/20 mL: 7 vials per week

#### B. Max Units (per dose and over time) [Medical Benefit]:

- 700 billable units every 7 days

### III. Initial Approval Criteria

#### Emphysema due to alpha-1-antitrypsin (AAT) deficiency †

- Patient is 18 years or older; **AND**
- Patient has an FEV<sub>1</sub> in the range of 30-65% of predicted; **AND**
- Patient has alpha-1-antitrypsin (AAT) deficiency with PiZZ, PiZ (null), or Pi (null, null) phenotypes; **AND**
- Patient has AAT deficiency and clinical evidence of panacinar/panlobular emphysema; **AND**
- Patient has low serum concentration of AAT  $\leq 11 \mu\text{M/L}$  or  $\leq 80 \text{ mg/dL}$  (if measured by radial immunodiffusion) or  $\leq 50 \text{ mg/dL}$  (if measured by nephelometry); **AND**

- Patient is receiving optimal medical therapy (e.g., comprehensive case management, pulmonary rehabilitation, vaccinations, smoking cessation, self-management skills, etc.); **AND**
- Patient is not a tobacco smoker

† FDA Approved Indication(s)

#### IV. Renewal Criteria

Authorizations can be renewed based on the following criteria:

- Patient continues to meet the criteria identified in section III; **AND**
- Disease response with treatment as defined by elevation of AAT levels above baseline, substantial reduction in rate of deterioration of lung function as measured by percent predicted FEV<sub>1</sub>, or improvement in CT scan lung density; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include severe hypersensitivity reactions, etc.

#### V. Dosage/Administration

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

#### VI. Billing Code/Availability Information

Jcode & NDC:

Drug	Manufacturer	Jcode	1 Billable Unit	SDV Size	NDC
Aralast NP (powder)	Baxalta US Inc.	J0256	10 mg	1 g/50 mL	00944-2815-xx
				0.5 g/25 mL	00944-2814-xx
Glassia (solution)	Baxalta US Inc.	J0257	10 mg	1 g/50 mL	00944-2884-xx
Prolastin-C (powder)	Grifols Therapeutics Inc.	J0256	10 mg	1 g/20 mL	13533-0700-xx
					13533-0703-xx
Prolastin-C Liquid (solution)	Grifols Therapeutics Inc.	J0256	10 mg	1 g/20 mL	13533-0705-xx
Zemaira (powder)	CSL Behring LLC	J0256	10 mg	1 g/20 mL	00053-7201-xx

## VII. References

1. Glassia [package insert]. Westlake Village, CA; Baxalta US Inc.; June 2017. Accessed March 2019.
2. Zemaira [package insert]. Kankakee, IL; CSL Behring LLC; September 2015. Accessed March 2019.
3. Aralast NP [package insert]. Westlake Village, CA; Baxalta US Inc.; December 2018. Accessed March 2019.
4. Prolastin-C Liquid [package insert]. Research Triangle Park, NC; Grifols Therapeutics, Inc.; August 2018. Accessed March 2019.
5. Prolastin-C [package insert]. Research Triangle Park, NC; Grifols Therapeutics, Inc.; June 2018. Accessed March 2019.
6. American Thoracic Society/European Respiratory Society Statement: Standards for the Diagnosis and Management of Individuals with Alpha-1 Antitrypsin Deficiency. American Thoracic Society; European Respiratory Society. Am J Respir Crit Care Med. 2003 Oct 1;168(7):818-900.
7. Global Initiative for Chronic Obstructive Lung Disease (GOLD). Global strategy for the diagnosis, management, and prevention of chronic obstructive pulmonary disease. Global Initiative for Chronic Obstructive Lung Disease (GOLD); 2018.
8. Sandhaus RA, Turino G, Brantly ML, et al. The diagnosis and management of alpha-1 antitrypsin deficiency in the adult. Chronic Obstr Pulm Dis (Miami). 2016; 3(3):668-682.
9. 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):109-16.
10. Wisconsin Physicians Service Insurance Corporation. Local Coverage Determination (LCD): Drugs and Biologics (Non-chemotherapy) (L34741). Centers for Medicare & Medicaid Services, Inc. Updated on 5/24/2018 with effective date 6/1/2018. Accessed March 2019.

### Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
E88.01	Alpha-1-antitrypsin deficiency

### Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologics. In addition, National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. They can be found at: <http://www.cms.gov/medicare-coverage-database/search/advanced-search.aspx>. Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD):

<b>Jurisdiction(s):</b> 5, 8	<b>NCD/LCD Document (s):</b> L34741
<a href="https://www.cms.gov/medicare-coverage-database/search/lcd-date-search.aspx?DocID=L34741&amp;bc=gAAAAAAAAAAAA">https://www.cms.gov/medicare-coverage-database/search/lcd-date-search.aspx?DocID=L34741&amp;bc=gAAAAAAAAAAAA</a>	

<b>Medicare Part B Administrative Contractor (MAC) Jurisdictions</b>		
<b>Jurisdiction</b>	<b>Applicable State/US Territory</b>	<b>Contractor</b>
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC