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) [NDC Unit]:

- 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: 3 vials per week
- Zemaira 4 g/76 mL: 1 vial per week
- Zemaira 5 g/95 mL: 1 vial per week

B. Max Units (per dose and over time) [HCPCS Unit]:

- 700 billable units every 7 days

III. Initial Approval Criteria[1-6,8,9,12]

- Patient is at least 18 years of age; AND

Universal Criteria

- Patient is not a tobacco smoker; AND
- Patient is receiving optimal medical therapy (e.g., comprehensive case management, pulmonary rehabilitation, vaccinations, smoking cessation, self-management skills, etc.); AND
- Patient does not have immunoglobulin-A (IgA) deficiency with antibodies against IgA; AND

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Emphysema due to alpha-1-antitrypsin (AAT) deficiency †, Φ (applies only to Prolastin-C)

- Patient has an FEV_1 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 ≤ 57 mg/dL or ≤ 11 µM/L as measured by nephelometry

† FDA Approved Indication(s); Φ Orphan Drug

IV. Renewal Criteria[1-6,8,9]

Authorizations can be renewed based on the following criteria:

- Patient continues to meet universal and other indication-specific relevant criteria such as concomitant therapy requirements (not including prerequisite therapy), performance status, etc. 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_1, 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[1-5]

Indication	Dose
Emphysema due to AAT deficiency	60 mg/kg by intravenous (IV) infusion administered once every 7 days (weekly)

VI. Billing Code/Availability Information

HCPCS Code & NDC:

Drug	Manufacturer	HCPCS code	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
	Grifols Therapeutics Inc.	J0256	10 mg	1 g/20 mL	13533-0700-xx

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Prolastin-C (powder)					13533-0701-xx
					13533-0702-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
				4 g/76 mL	00053-7202-xx
				5 g/95 mL	00053-7203-xx

VII. References

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2. Zemaira [package insert]. Kankakee, IL; CSL Behring LLC; April 2019. Accessed March 2021.
3. Aralast NP [package insert]. Lexington, MA; Baxalta US Inc.; December 2018. Accessed March 2021.
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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.
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9. 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):109-16.
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11. 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); 2020.

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12. Miravitlles M, Dirksen A, Ferrarotti I, et al. European Respiratory Society statement: diagnosis and treatment of pulmonary disease in α1-antitrypsin deficiency. Eur Respir J 2017; 50.

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 Biologicals. In addition, National Coverage Determination (NCD), Local Coverage Determinations (LCDs), and Local Coverage Articles (LCAs) 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/LCA): N/A

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
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

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