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, unless otherwise specified.

- Acute Graft Versus Host Disease (aGVHD): Coverage will be provided for a maximum of 8 doses (4 weeks) and may NOT be renewed.

II. Dosing Limits

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

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

- 2800 billable units every 28 days

aGVHD

- 700 billable units for a total of 8 doses in 28 days (5600 billable units per 28 days)

III. Initial Approval Criteria [1-5]

Site of care specialty infusion program requirements are met (refer to Moda Site of Care Policy).

Coverage is provided in the following conditions:

- Patient is at least 18 years of age; **AND**

Universal Criteria [1-5]

- Patient does not have immunoglobulin-A (IgA) deficiency with antibodies against IgA; **AND**

Emphysema due to alpha-1-antitrypsin (AAT) deficiency † (**Φ –** *orphan designation applies only to Prolastin-C*) **[1-6,8,9,12,16,17]**

- 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 clinical evidence of panacinar/panlobular emphysema or centrilobular emphysema; **AND**

- Patient has low serum concentration of AAT ≤ 57 mg/dL (measured by nephelometry) or ≤ 11 µM/L (measured by ELISA); **AND**
- 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.)

Acute Graft Versus Host Disease (aGVHD) ‡ [13-15]

- Patient has received a hematopoietic stem cell transplant; **AND**
- Used for steroid-refractory acute GVHD; **AND**
- Used in combination with systemic corticosteroids as additional therapy following no response to first-line therapy options

† FDA Approved Indication(s); ‡ Compendia Recommended Indication(s); Φ Orphan Drug

IV. Renewal Criteria [1-5]

Coverage can be renewed based upon the following criteria:

- Patient continues to meet the universal and other indication-specific relevant criteria such as concomitant therapy requirements (not including prerequisite therapy), performance status, etc. identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: severe hypersensitivity reactions, etc.; **AND**

Emphysema due to alpha-1-antitrypsin (AAT) deficiency [1-6,8,9]

- 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

Graft Versus Host Disease (GVHD) [13-15]

- Duration of authorization has not been exceeded (*refer to section I*)

V. Dosage/Administration [1-5,15]

Indication	Dose
Emphysema due to AAT deficiency	Administer 60 mg/kg intravenously once weekly
Acute GVHD	Administer 60 mg/kg intravenously on days 1, 4, 8, 12, 16, 20, 24, and 28 for up to 4 consecutive weeks (maximum of 8 doses)

VI. Billing Code/Availability Information

HCPCS Code(s) & NDC(s):

Drug	Manufacturer	HCPCS code	1 Billable Unit	SDV Size	NDC
Aralast NP (powder)	Takeda Pharmaceuticals USA Inc.	J0256	10 mg	1 g/50 mL	00944-2815-xx
				0.5 g/25 mL	00944-2814-xx
Glassia (solution)	Takeda Pharmaceuticals USA Inc.	J0257	10 mg	1 g/50 mL	00944-2884-xx
Prolastin-C (powder)	Grifols Therapeutics LLC	J0256	10 mg	1 g/20 mL	13533-0700-xx
					13533-0701-xx
					13533-0702-xx
					13533-0703-xx
					13533-0706-xx
Prolastin-C Liquid (solution)	Grifols Therapeutics LLC	J0256	10 mg	500 mg/10 mL	13533-0705-xx
				1 g/20 mL	
				4 g/80 mL	
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

1. Glassia [package insert]. Cambridge, MA; Takeda Pharmaceuticals USA, Inc.; February 2025. Accessed March 2025.
2. Zemaira [package insert]. Kankakee, IL; CSL Behring LLC; January 2024. Accessed March 2025.
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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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14. Referenced with permission from the NCCN Drugs & Biologics Compendium (NCCN Compendium®) Hematopoietic Cell Transplantation (HCT). Version 1.2025. National Comprehensive Cancer Network, 2025. The NCCN Compendium® is a derivative work of the NCCN Guidelines®. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc." To view the most recent and complete version of the Compendium, go online to NCCN.org. Accessed March 2025.
15. Magenau JM, Goldstein SC, Peltier D, et al. α1-Antitrypsin infusion for treatment of steroid-resistant acute graft-versus-host disease. Blood. 2018 Mar 22;131(12):1372-1379. doi: 10.1182/blood-2017-11-815746.
16. 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); 2024 Report.
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Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
D89.810	Acute graft-versus-host disease
D89.812	Acute on chronic graft-versus-host disease
D89.813	Graft-versus-host disease, unspecified

E88.01	Alpha-1-antitrypsin deficiency
T86.09	Other complications of bone marrow transplant

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

The preceding information is intended for non-Medicare coverage determinations. 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 Determinations (NCDs) and/or Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. Local Coverage Articles (LCAs) may also exist for claims payment purposes or to clarify benefit eligibility under Part B for drugs which may be self-administered. The following link may be used to search for NCD, LCD, or LCA documents: https://www.cms.gov/medicare-coverage-database/search.aspx. Additional indications, including any preceding information, may be applied 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
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA
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