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

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I. Length of Authorization [13]

- Initial: Prior authorization validity will be provided initially for 12 months (365 days), unless otherwise specified.
  - Acute Graft Versus Host Disease (aGVHD): Prior authorization validity will be provided for a maximum of 8 doses (4 weeks).
- Renewal: Prior authorization validity may be renewed every 12 months (365 days) thereafter, unless otherwise specified.
  - Acute Graft Versus Host Disease (aGVHD): Prior authorization validity 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]

For groups in scope for the Site of Care (SOC) specialty infusion program, all program requirements are met.

Prior authorization validity is provided in the following conditions:

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

Universal Criteria [1-5]

- Member does NOT have any FDA labeled contraindications to the requested agent; **AND**

Emphysema due to alpha-1-antitrypsin (AAT) deficiency † (Φ – *orphan designation applies only to Prolastin-C*) [1-6,7,8,10,14-17]

- Member has an FEV_1 in the range of 30-65% of predicted; **AND**
- Member has alpha-1-antitrypsin (AAT) deficiency with PiZZ, PiZ (null), or Pi (null, null) phenotypes; **AND**
- Member has clinical evidence of panacinar/panlobular emphysema or centrilobular emphysema; **AND**
- Member has low serum concentration of AAT ≤ 57 mg/dL (measured by nephelometry) or ≤ 11 µM/L (measured by Enzyme-Linked Immunosorbent Assay [ELISA]); **AND**
- Member is not a tobacco smoker; **AND**
- Member is receiving optimal medical therapy (e.g., pharmacologic therapy [e.g., bronchodilators, inhaled corticosteroids, etc.], comprehensive case management, pulmonary rehabilitation, vaccinations, smoking cessation, self-management skills, supplemental oxygen, etc.)

Acute Graft Versus Host Disease (aGVHD) ‡ [11-13]

- Member 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]

Prior authorization validity may be renewed based upon the following criteria:

- Member 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**
- Duration of authorization has not been exceeded (*refer to section I*); **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 [1-7,17]

- 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

V. Dosage/Administration [1-5,13]

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 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

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Appendix A – Non-Quantitative Treatment Limitations (NQTL) Factor Checklist

Non-quantitative treatment limitations (NQTLs) refer to the methods, guidelines, standards of evidence, or other conditions that can restrict how long or to what extent benefits are provided under a health plan. These may include things like utilization review or prior authorization. The utilization management NQTL applies comparably, and not more stringently, to mental health/substance use disorder (MH/SUD) Medical Benefit Prescription Drugs and medical/surgical (M/S) Medical Benefit Prescription Drugs. The table below lists the factors that were considered in designing and applying prior authorization to this drug/drug group, and a summary of the conclusions that Prime's assessment led to for each.

Factor	Conclusion
Indication	Yes: Consider for PA
Safety and efficacy	No: PA not a priority
Potential for misuse/abuse	No: PA not a priority
Cost of drug	Yes: Consider for PA

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