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

Effective Date: 01/01/2020
Review Date: 12/13/2019, 1/29/2020, 01/28/2021, 4/22/2021, 4/14/2022, 3/2/2023, 12/07/2023, 01/10/2024, 04/24/2024

Scope: Medicaid, Commercial, Medicare-Medicaid Plan (MMP)

I. Length of Authorization

Coverage will be provided for 6 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. Summary of Evidence

Numerous clinical studies support the efficacy of alpha-1 proteinase inhibitors in reducing the rate of lung function decline, decreasing the frequency and severity of exacerbations, and improving quality of life in patients with AATD. These therapies have been shown to increase serum levels of alpha-1 antitrypsin and restore the protease-antiprotease balance in the lungs.

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

MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements.

- 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**

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
- For commercial patients ONLY when requesting Aralast, Glassia, or Zemaira, they must have a documented failure, intolerance or contraindication to Prolastin
- For MMP patients ONLY when requesting Aralast or Glassia they must have a documented failure, intolerance, or contraindication to Prolastin or Zemaira

† FDA Approved Indication(s); **Φ** Orphan Drug

V. 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.

VI. Dosage/Administration[1-5]

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

VII. 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
Prolastin-C (powder)	Grifols Therapeutics Inc.	J0256	10 mg	1 g/20 mL	13533-0700-xx
					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

VIII. References

1. Glassia [package insert]. Lexington, MA; Baxalta US Inc.; September2023. Accessed November 2023.
2. Zemaira [package insert]. Kankakee, IL; CSL Behring LLC; November 2022. Accessed November 2023.
3. Aralast NP [package insert]. Lexington, MA; Baxalta US Inc.; May 2023. Accessed November 2023.
4. Prolastin-C Liquid [package insert]. Research Triangle Park, NC; Grifols Therapeutics, Inc.; February 2022 . Accessed November 2023.
5. Prolastin-C [package insert]. Research Triangle Park, NC; Grifols Therapeutics, Inc.; February 2022. Accessed November 2023.
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); 2019.
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. 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.

10. Stocks JM, Brantly M, Pollock D, et al. Multi-center study: the biochemical efficacy, safety and tolerability of a new α1-proteinase inhibitor, Zemaira. COPD. 2006;3:17–23.
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.
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

Policy Rationale:

Alpha-1-Proteinase Inhibitors (Aralast NP, Glassia, Prolastin, and Zemaira) were reviewed by the Neighborhood Health Plan of Rhode Island Pharmacy & Therapeutics (P&T) Committee. Neighborhood adopted the following clinical coverage criteria to ensure that its members use Aralast NP, Glassia, Prolastin, and Zemaira according to Food and Drug Administration (FDA) approved labeling and/or relevant clinical literature. Neighborhood worked with network prescribers and pharmacists to draft these criteria. These criteria will help ensure its members are using this drug for a medically accepted indication, while minimizing the risk for adverse effects and ensuring more cost-effective options are used first, if applicable and appropriate. For INTEGRITY (Medicare-Medicaid Plan) members, these coverage criteria will only apply in the absence of National Coverage Determination (NCD) or Local Coverage Determination (LCD) criteria. Neighborhood will give individual consideration to each request it reviews based on the information submitted by the prescriber and other information available to the plan.