Policy Title:	Alpha-1-Proteinase Inhibitors: Aralast NP, Glassia, Prolastin-C, Zemaira (Intravenous)		
		Department:	PHA
Effective Date:	01/01/2020		
Review Date:	12/13/2019, 1/29/20		
Revision Date:	12/13/2019, 1/29/20		

Purpose: To support safe, effective and appropriate use of Alpha-1-Proteinase Inhibitors.

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

Policy Statement:

Alpha-1-Proteinase Inhibitors are covered under the Medical Benefit when used within the following guidelines. Use outside of these guidelines may result in non-payment unless approved under an exception process.

Procedure:

Coverage of Alpha-1-Proteinase Inhibitors will be reviewed prospectively via the prior authorization process based on criteria below.

Initial Criteria:

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

- Patient is 18 years or older; AND
- 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 ≤ 11 μM/L or ≤ 80 mg/dL (if measured by radial immunodiffusion) or ≤ 50 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; AND
- MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements

Continuation of Therapy Criteria:

- Meets all initial approval criteria AND is tolerating treatment; 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.

Coverage durations:

- Initial coverage: 6 months
- Continuation of therapy coverage: 6 months

*** Requests will also be reviewed to National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) if applicable. ***

Dosage/Administration:

Indication	Dose	Maximum dose (1 billable unit = 10 mg)
All indications	60 mg/kg by intravenous (IV) infusion administered once every 7 days (weekly)	700 billable units every 7 days

Investigational use: All therapies are considered investigational when used at a dose or for a condition other than those that are recognized as medically accepted indications as defined in any one of the following standard reference compendia: American Hospital Formulary Service Drug information (AHFS-DI), Thomson Micromedex DrugDex, Clinical Pharmacology, Wolters Kluwer Lexi-Drugs, or Peer-reviewed published medical literature indicating that sufficient evidence exists to support use. Neighborhood does not provide coverage for drugs when used for investigational purposes.

Applicable Codes:
Below is a list of billing codes applicable for covered treatment options. The below tables are provided for reference purposes and may not be all-inclusive. Requests received with codes from tables below do not guarantee coverage. Requests must meet all criteria provided in the procedure section.

The following HCPCS/CPT codes are:

HCPCS/CPT Code	Description
J0256	Injection, alpha 1-proteinase inhibitor (human), not otherwise specified, 10mg
J0257	Injection, alpha 1 proteinase inhibitor (human), (GLASSIA), 10mg

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.