Clinical Policy: Alpha-1 Proteinase Inhibitors (Aralast NP, Glassia, Prolastin-C, Zemaira)

Reference Number: PA.CP.PHAR.94
Effective Date: 01/18
Last Review Date: 07/18

Coding Implications
Revision Log

Description
The intent of the criteria is to ensure that patients follow selection elements established by of Pennsylvania Health and Wellness ® clinical policy for alpha-1 proteinase inhibitor (human) (Aralast NP™, Glassia®, Prolastin®-C, Zemaira®).

Policy/Criteria
It is the policy of of Pennsylvania Health and Wellness that Aralast NP, Glassia, Prolastin-C, and Zemaira are **medically necessary** when the following criteria are met:

I. Initial Approval Criteria

A. Alpha-1 Antitrypsin Deficiency (must meet all):

1. Diagnosis of severe congenital AAT deficiency;
2. Age ≥ 18 years;
3. Member meets one of the following (a or b):
    1. Documentation of plasma AAT level < 11 micromol/L (approximately 50 mg/dL using nephelometry or 80 mg/dL by radial immunodiffusion);
    2. If member has an AAT level >11 umol/L, then the member must have one of the high-risk phenotypes (i.e. PiZZ, PiZnull, Pi(null, null), or one of a few rare phenotypes [e.g. Pi(Malton, Malton)].
4. Prescribed by or in consultation with a pulmonologist;
5. Clinical evidence of emphysema (a or b):
    1. Forced expiratory volume in one second (FEV_1) from ≥ 30% to < 65% of predicted, post-bronchodilator;
    2. FEV_1 from ≥ 65% to < 80% of predicted, post-bronchodilator, and a rapid decline in lung function showing a change in FEV_1 > 100 mL/year;
6. Dose does not exceed 60 mg/kg/week.

Approval Duration: 6 months

B. Other diagnoses/indications: Refer to PA.CP.PMN.53

II. Continued Approval

A. Alpha-1 Antitrypsin Deficiency (must meet all)**:**

1. Currently receiving medication via of Pennsylvania Health and Wellness benefit or member has previously met all initial approval criteria or Continuity of Care policy applies;
2. Documentation supports positive response to therapy;
3. Prescribed dose does not exceed 60 mg/kg once weekly.

Approval Duration: 12 months

B. Other diagnoses/indications (must meet 1 or 2)**:**
1. Currently receiving medication via of Pennsylvania Health and Wellness benefit and documentation supports positive response to therapy; or
2. Refer to PA.CP.PMN.53

Background
Description/Mechanism of Action:
Aralast NP, Glassia, Prolastin-C, and Zemaira are purified human alpha-1 proteinase inhibitors. Alpha-1 antitrypsin (AAT) is the principle protease inhibitor in serum. Its major physiologic role is to render proteolytic enzymes (secreted during inflammation) inactive. A decrease in AAT, as seen in congenital AAT deficiency, leads to increased elastic damage in the lung, causing emphysema.

Formulations:
Intravenous solution:
Glassia: 1000 mg/50 mL (1 ea)
Reconstituted intravenous solution:
Zemaira: 1000 mg (1 ea)
Aralast NP: 500 mg (1 ea); 1000 mg (1 ea)
Prolastin-C: 1000 mg (1 ea)

Appendices
Appendix A: Abbreviation Key
AAT: alpha-1 antitrypsin
Alpha-1 PI: alpha-1 proteinase inhibitor
FEV_1: forced expiratory volume in one second
IgA: immunoglobulin A
LRTI: lower respiratory tract infection

Coding Implications
Codes referenced in this clinical policy are for informational purposes only. Inclusion or exclusion of any codes does not guarantee coverage. Providers should reference the most up-to-date sources of professional coding guidance prior to the submission of claims for reimbursement of covered services.

HCPCS Codes	Description
J0256	Injection, alpha 1-proteinase inhibitor (human), not otherwise specified, 10 mg Aralast NP; Prolastin-C; Zemaira
J0257	Injection, alpha 1 proteinase inhibitor (human), (Glassia), 10 mg

Reviews, Revisions, and Approvals	Date	Approval Date
Removed requirement for supportive measures (avoidance of cigarette smoking and vaccinations) due to lack of actionability and objectivity. Protective threshold value per nephelometry changed from 57 mg/dL to 50 mg/dL per American Thoracic Society 2003 guidelines. Added "If the member has an AAT level >11 umol/L, then the member must have one of the high-risk phenotypes (i.e. PiZZ, PiZnull, Pi(null, null), or one of a few rare phenotypes [e.g. Pi(Malton, Malton)]" to allow treatment before clinical deterioration due to definite diagnosis. Added prescriber requirement due to the complexity of disease diagnosis and management; Changed minimally significant change in FEV from 120 mL to 100 mL per ATC guidelines and specialist feedback. References reviewed and updated.		

References

1. Aralast NP Prescribing Information. Westlake Village, CA: Baxter Healthcare Corporation; September 2015. Available at http://www.shirecontent.com/PI/PDFs/ARALASTNP_USA_ENG.pdf. Accessed November 20, 2017.
2. Glassia Prescribing Information. Negev, Israel: Kamada, Ltd.; June 2016. Available at http://www.liquidglassia.com. Accessed November 20, 2017.
3. Prolastin-C Prescribing Information. Research Triangle Park, NC: Grifols Therapeutics, Inc.; August 2016. Available at http://www.prolastin.com. Accessed November 20, 2017.
4. Zemaira Prescribing Information. Kankakee, IL: CSL Behring LLC; September 2015. Available at http://www.zemaira.com. Accessed November 20, 2017.
5. American Thoracic Society/European Respiratory Society statement: standards for the diagnosis and management of individuals with alpha-1 antitrypsin deficiency. Am J Respir Crit Care Med. 2003; 168(7): 818-900.
6. Sandhaus RA, Turino G, and Brantly ML, et al. The diagnosis and management of alpha-1 antitrypsin deficiency in the adult. Journal of COPD Foundation. 2016;3(3):668-682.
7. Cazzola M, MacNee W, Martinez FJ, et al.; American Thoracic Society; European Respiratory Society Task Force on outcomes of COPD. Outcomes for COPD pharmacological trials: from lung function to biomarkers. Eur Respir J 2008;31:416–469.