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

Reference Number: CP.PHAR.94
Effective Date: 03/12
Last Review Date: 03/17

Coding Implications
Revision Log

See Important Reminder at the end of this policy for important regulatory and legal information.

Description

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

Policy/Criteria

It is the policy of health plans affiliated with Centene Corporation® 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 alpha-1 antitrypsin (AAT) deficiency;
   a. Plasma AAT level is < 11 micromol/L (approximately 57 mg/dL using nephelometry or 80 mg/dL by radial immunodiffusion);
2. Clinical evidence of emphysema (a or b):
   a. Forced expiratory volume in one second ($FEV_{1)}$ from ≥ 30% to < 65% of predicted, post-bronchodilator;
   b. FEV_1 from ≥ 65% to < 80% of predicted, post-bronchodilator, and a rapid decline in lung function showing a change in FEV_1 > 120 mL/year;
3. Member is being managed with the following supportive measures per chronic obstructive pulmonary disease (COPD) guidelines (a and b):
   a. Avoidance of cigarette smoking;
   b. Supportive care measures that may include use of bronchodilators, inhaled or oral glucocorticoids, oxygen, pulmonary rehabilitation; nutritional support, lower respiratory tract infection (LRTI) management and preventive vaccinations;
4. Prescribed dose does not exceed 60 mg/kg once weekly.

Approval Duration: 6 months

B. Other diagnoses/indications: Refer to CP.PHAR.57 - Global Biopharm Policy.

II. Continued Approval

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

1. Currently receiving medication via Centene benefit or member has previously met all initial approval criteria;
2. Documentation supports positive response to therapy;

3. Member is being managed with the following supportive measures per COPD guidelines (a and b):
   a. Avoidance of cigarette smoking;
   b. Supportive care measures that may include use of bronchodilators, inhaled or oral glucocorticoids, oxygen, pulmonary rehabilitation; nutritional support, LRTI management and preventive vaccinations;
4. 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 Centene benefit and documentation supports positive response to therapy; or
2. Refer to CP.PHAR.57 - Global Biopharm Policy**.**

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)

FDA Approved Indications:

Aralast NP, Glassia, Prolastin-C, and Zemaira are alpha-1 proteinase inhibitors*/injectable products indicated for:

- Chronic augmentation and maintenance therapy in individuals with clinically evident emphysema due to severe congenital AAT deficiency.

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**Also known as alpha-1 PI, alpha-1 antitrypsin, AAT*

Limitations of use:

- The effect of augmentation therapy with alpha-1 proteinase inhibitors on pulmonary exacerbations and on the progression of emphysema in alpha-1 proteinase inhibitor deficiency has not been demonstrated in randomized, controlled clinical trials.
- Clinical data demonstrating the long-term effects of chronic augmentation or maintenance therapy of individuals with alpha-1 proteinase inhibitors are not available.

- Alpha-1 proteinase inhibitors are not indicated as therapy for patients with lung disease in whom congenital alpha-1 proteinase inhibitor deficiency has not been established.

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
Duration of approval has been changed to 12 months	03/13	
Description updated based on Caremark guideline document	04/13	04/13
Converted to Centene policy template	06/13	
atReviewed with only minor language changes	03/14	03/14
References reviewed and updated	02/15	03/15
Corrected FEV1 range from 35 to 65% to 30 to 65% based on Table 9 in the 2003 ATS/ERS AAT guidelines	08/15	
Policy converted to new template. Criteria: added max dose and attestation that member is receiving additional supportive measures per COPD guidelines.	02/16	03/16
Initial criteria: Age removed; conditions representing potential contraindications to therapy are removed.	02/17	03/17

References

1. Aralast NP prescribing information. Westlake Village, CA: Baxter Healthcare Corporation; March 2014. Available at http://www.shirecontent.com/PI/PDFs/ARALASTNP_USA_ENG.pdf. Accessed February 16, 2017.

2. Glassia prescribing information. Westlake Village, CA: Baxalta US, Inc.; June 2016. Available at http://www.shirecontent.com/PI/PDFs/GLASSIA_USA_ENG.pdf. Accessed February 15, 2017.
3. Prolastin-C prescribing information. Research Triangle Park, NC: Grifols Therapeutics, Inc.; August 2016. Available at http://www.prolastin.com/documents/5103363/0/prolastinPI/b436e646-0787-4aad-a3d5-b5e004cc92c6. Accessed February 15, 2017.
4. Zemaira prescribing information. Kankakee, IL: CSL Behring, LLC; September 2015. Available at http://labeling.cslbehring.com/PI/US/Zemaira/EN/Zemaira-Prescribing-Information.pdf. Accessed February 15, 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. Stoller JK. Clinical manifestations, diagnosis, and natural history of alpha-1 antitrypsin deficiency. In: UpToDate, Waltham, MA: Walters Kluwer Health; 2017. Available at UpToDate.com. Accessed February 16, 2017.
7. Stoller JK. Treatment of alpha-1 antitrypsin deficiency. In: UpToDate, Waltham, MA: Walters Kluwer Health; 2016. Available at UpToDate.com. Accessed February 16, 2017.
8. Alpha-1 proteinase inhibitor: Drug information. In: Lexicomp through UpToDate, Waltham, MA: Walters Kluwer Health; 2017. Available at UpToDate.com. Accessed February 16, 2017.

Important Reminder

This clinical policy has been developed by appropriately experienced and licensed health care professionals based on a review and consideration of currently available generally accepted standards of medical practice; peer-reviewed medical literature; government agency/program approval status; evidence-based guidelines and positions of leading national health professional organizations; views of physicians practicing in relevant clinical areas affected by this clinical policy; and other available clinical information. The Health Plan makes no representations and accepts no liability with respect to the content of any external information used or relied upon in developing this clinical policy. This clinical policy is consistent with standards of medical practice current at the time that this clinical policy was approved. "Health Plan" means a health plan that has adopted this clinical policy and that is operated or administered, in whole or in part, by Centene Management Company, LLC, or any of such health plan's affiliates, as applicable.

The purpose of this clinical policy is to provide a guide to medical necessity, which is a component of the guidelines used to assist in making coverage decisions and administering benefits. It does not constitute a contract or guarantee regarding payment or results. Coverage decisions and the administration of benefits are subject to all terms, conditions, exclusions and limitations of the coverage documents (e.g., evidence of coverage, certificate of coverage, policy, contract of insurance, etc.), as well as to state and federal requirements and applicable Health Plan-level administrative policies and procedures.

This clinical policy is effective as of the date determined by the Health Plan. The date of posting may not be the effective date of this clinical policy. This clinical policy may be subject to applicable legal and regulatory requirements relating to provider notification. If there is a discrepancy between the effective date of this clinical policy and any applicable legal or regulatory requirement, the requirements of law and regulation shall govern. The Health Plan retains the right to change, amend or withdraw this clinical policy, and additional clinical policies may be developed and adopted as needed, at any time.

This clinical policy does not constitute medical advice, medical treatment or medical care. It is not intended to dictate to providers how to practice medicine. Providers are expected to exercise professional medical judgment in providing the most appropriate care, and are solely responsible for the medical advice and treatment of members. This clinical policy is not intended to recommend treatment for members. Members should consult with their treating physician in connection with diagnosis and treatment decisions.

Providers referred to in this clinical policy are independent contractors who exercise independent judgment and over whom the Health Plan has no control or right of control. Providers are not agents or employees of the Health Plan.

This clinical policy is the property of the Health Plan. Unauthorized copying, use, and distribution of this clinical policy or any information contained herein are strictly prohibited. Providers, members and their representatives are bound to the terms and conditions expressed herein through the terms of their contracts. Where no such contract exists, providers, members and their representatives agree to be bound by such terms and conditions by providing services to members and/or submitting claims for payment for such services.

Note: For Medicaid members, when state Medicaid coverage provisions conflict with the coverage provisions in this clinical policy, state Medicaid coverage provisions take precedence. Please refer to the state Medicaid manual for any coverage provisions pertaining to this clinical policy.

Note: For Medicare members, to ensure consistency with the Medicare National Coverage Determinations (NCD) and Local Coverage Determinations (LCD), all applicable NCDs, LCDs, and Medicare Coverage Articles should be reviewed prior to applying the criteria set forth in this clinical policy. Refer to the CMS website at http://www.cms.gov for additional information.