CareSource

PHARMACY POLICY STATEMENT

Original Effective Date	Next Annual Review		Last Revision
01/18/2013	01/18/2018		11/26/2016
Policy Name			**Policy Number**
Alpha-1 Proteinase Inhibitors			SRx-0056-KY-MCD
Policy Type			
Medical	Administrative	**PHARMACY**	Reimbursement

Pharmacy Policy Statements prepared by CSMG Co. and its affiliates (including CareSource) are derived from literature based on and supported by clinical guidelines, nationally recognized utilization and technology assessment guidelines, other medical management industry standards, and published MCO clinical policy guidelines. Medically necessary services include, but are not limited to, those health care services or supplies that are proper and necessary for the diagnosis or treatment of disease, illness, or injury and without which the patient can be expected to suffer prolonged, increased or new morbidity, impairment of function, dysfunction of a body organ or part, or significant pain and discomfort. These services meet the standards of good medical practice in the local area, are the lowest cost alternative, and are not provided mainly for the convenience of the member or provider. Medically necessary services also include those services defined in any Evidence of Coverage documents, Pharmacy Policy Statements, Provider Manuals, Member Handbooks, and/or other policies and procedures.

Pharmacy Policy Statements prepared by CSMG Co. and its affiliates (including CareSource) do not ensure an authorization or payment of services. Please refer to the plan contract (often referred to as the Evidence of Coverage) for the service(s) referenced in the Pharmacy Policy Statement. If there is a conflict between the Pharmacy Policy Statement and the plan contract (i.e., Evidence of Coverage), then the plan contract (i.e., Evidence of Coverage) will be the controlling document used to make the determination

Contents of Policy

A. INTRODUCTION

Alpha-1 Antitrypsin Deficiency is an inherited condition passed from parents to children that can lead to serious lung disease in adults and/or liver disease at any age. Alpha-1 proteinase inhibitors, also called alpha 1-PI, are used to treat the lung disease (emphysema) caused by the lack of alpha 1-antitrypsin (AAT), which is a protein in the body. Alpha-1 proteinase inhibitors replace the protein when the body does not produce enough.

The intent of CareSource Pharmacy Policy Statements is to encourage appropriate selection of patients for therapy according to product labeling, clinical guidelines, and/or clinical studies as well as to encourage use of preferred agents. The CareSource Pharmacy Policy Statement is a guideline for determining health care coverage for our patients with benefit plans covering prescription drugs. Pharmacy Policy Statements are written on selected prescription drugs requiring prior authorization or step therapy. The Pharmacy Policy Statement is used as a tool to be interpreted in conjunction with the member's specific benefit plan.

NOTE: The Introduction section is for your general knowledge and is not to be construed as policy coverage criteria. The rest of the policy uses specific words and concepts familiar to medical professionals and is intended for providers. A provider can be a person, such as a doctor, nurse, psychologist, or dentist. A provider can also be a place where medical care is given, like a hospital, clinic or lab. This policy informs providers about when a service may be covered.

B. DEFINITIONS

1. None applicable.

C. POLICY COVERAGE CRITERIA

1. Site of Service

Site of Service Administration	Coverage Criteria
Office, Outpatient, Home	**Preferred place of service is in the home.** CareSource supports administering injectable medications in various settings, as long as those services are furnished in the most appropriate and cost effective settings that are supportive of the patient's medical condition(s) and unique needs and condition(s). The decision on the most appropriate setting for administration is based on the member's current medical condition(s) and any required monitoring or additional services that may coincide with the delivery of the specific medication.

2. Coverage Criteria

CareSource will approve the use of Aralast NP, Glassia, Prolastin-C, and Zemaira and consider its use medically necessary when the criteria have been met for each drug/condition listed below. Prior authorization request should be submitted with chart notes and documentation supporting medical necessity.

Condition	Aralast NP, Glassia, Prolastin-C, or Zemaira coverage criteria:
Emphysema due to alpha-1 proteinase inhibitor deficiency	1) Member is age 18 years or older 2) Diagnosis of emphysema due to alpha-1 antitrypsin deficiency. *Include chart notes and imaging studies.* 3) Member has one of the high risk phenotypes (PI*ZZ, PI*Z[null], or PI*[null][null]. 4) Serum plasma levels of alpha-1 antitrypsin (ATT) less than 80 mg/dL (11 micromol/L) measured via radial immunodiffusion 5) Member is currently a non-smoker for 6 or more months 6) One or more of the following: a) Prior to initiation of therapy: Airflow obstruction as evidenced by forced expiratory volume (FEV1) of 30-65% of predicted value b) Rapid decline in lung function as measured by a change in FEV1 greater than 120 mL/year

All other uses of Aralast NP, Glassia, Prolastin-C, and Zemaira are considered experimental/investigational; and therefore, will follow CareSource's off-label policy.

Please note that this policy is reviewed on an annual basis. New drugs and indications receiving FDA approval may not be reflected in this policy immediately.

Notes:

- Documented diagnosis must be confirmed by portions of the individual's medical record which need to be supplied with prior authorization request. These medical records may include, but are not limited to test reports, chart notes from provider's office, or hospital admission notes.
- Member is required to have completed the trial(s) listed in the above criteria unless the member is unable to tolerate or has a contraindication to trial medications. Documentation such as chart notes or pharmacy claims may be requested to verify trial(s), intolerance, or contraindication(s).
- Refer to the product package insert for dosing, administration and safety guidelines.

3. Dosage and Quantity Limits (listed if applicable)

Information for patients with renal or hepatic impairment is not included. See package insert for individual agents.

Condition	Dosage and Quantity Limit of Aralast NP, Glassia, Prolastin-C, or Zemaira
Emphysema due to alpha-1 proteinase inhibitor deficiency	60 mg/kg once weekly

4. Authorization Period

Condition	Approval Period
Emphysema due to alpha-1 proteinase inhibitor deficiency	The initial authorization for Aralast NP, Glassia, Prolastin-C, or Zemaira is valid for 12 months. Continued treatment may be considered when documentation confirms the diagnosis of emphysema due to ATT deficiency, continued non-smoking status, and efficacy of prior treatment (e.g., plasma ATT levels within the normal/protective range. A reauthorization after successful initiation period will be placed for 12 months. **ALL** authorizations are subject to continued eligibility.

5. Coding

HCPCS	
J0256	Injection, alpha 1-proteinase inhibitor (human), not otherwise specified,10 mg
J0257	Injection, alpha 1 - proteinase inhibitor (human), (Glassia), 10 mg

D. RELATED POLICIES

AD-0004: Medical Necessity - Off-Label, Approved Orphan and Compassionate Use Drugs

E. REVIEW/REVISION HISTORY

DATE	ACTION/DESCRIPTION
01/18/2013	Issued, reviewed
02/14/2014	Revised authorization period
01/18/2015	Alpha 1 serum level & FEV value changed
01/13/2016	Revised phenotypes and modified ATT level
11/26/2016	Updated policy format, separated by line of business, updated criteria.

F. REFERENCES

1. Alpha-1 Foundation. Learning About Alpha-1. Available at: https://www.alpha1.org/what-is-alpha1. 2016.
2. Aralast-NP (alpha1-proteinase inhibitor (Human)) [prescribing information]. Westlake Village, CA: Baxter Healthcare Corporation; March 2014.
3. Glassia (alpha1-proteinase inhibitor (Human)) [prescribing information]. Beit Kama, Israel: Kamada; June 2016.
4. Prolastin-C (alpha1-proteinase inhibitor (Human)) [prescribing information]. Research Triangle Park, NC: Talecris Biotherapeutics; October 2009.
5. Zemaira (alpha1-proteinase inhibitor (Human)) [prescribing information]. Kankakee, IL: CSL Behring LLC; September 2015.
6. 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
7. Strange, C., Beiko, T. Treatment of Alpha-1 Antitrypsin Deficiency. Semin Respi Crit Care Med. 2015: 36(4): 470-477.
8. Stockley, RA. The multiple facets of alpha-1 antitrypsin. Ann Transl Med. 2015: 3(10): 130 Stockley, RA. The multiple facets of alpha-1 antitrypsin. Ann Transl Med. 2015: 3(10): 130.
9. Traclet, J., Delval, P., Terrioux, P., et al. Augmentation therapy of alpha-1 antitrypsin deficiency associated emphysema. Rev Mal. Respir. 2015: 32(4): 435-446.
10. Campos, MA., Lascano, J. Alpha-1 antitrypsin deficiency: current best practice in testing and augmentation therapy. Ther Adv Respir Dis. 2014; 8(5): 150-161.
11. MacDonald JL, Johnson CE. Pathophysiology and treatment of alpha 1-antitrypsin deficiency. Am J Health Syst Pharm. 1995;52(5):481-489.
12. Coakley RJ, Taggart C, O'Neill S, et al. Alpha1-antitrypsin deficiency: Biological answers to clinical questions. Am J Med Sci. 2001;321(1):33-41.
13. Parfrey H, Mahadeva R, Lomas DA. Alpha(1)-antitrypsin deficiency, liver disease and emphysema. Int J Biochem Cell Biol. 2003;35(7):1009-1014.
14. Juvelekian GS, Stoller JK. Augmentation therapy for alpha(1)-antitrypsin deficiency. Drugs. 2004;64(16):1743-1756.
15. Abboud RT, Ford GT, Chapman KR. Emphysema in alpha1-antitrypsin deficiency: Does replacement therapy affect outcome? Treat Respir Med. 2005;4(1):1-8.
16. Kerstiens H, Postma D, ten Hacken N. Chronic obstructive pulmonary disease. In: Clinical Evidence. London, UK: BMJ Publishing Group; March 2005.
17. Stocks JM, Brantly M, Pollock D, et al. Multi-center study: The biochemical efficacy, safety and tolerability of a new alpha1-proteinase inhibitor, Zemaira. COPD. 2006;3(1):17-23.
18. Chen S, Farahati F, Marciniuk D, et al. Human a1-proteinase inhibitor for patients with a1-antitrypsin deficiency. Technology Report No. 74. Ottawa, ON: Canadian Agency for Drugs and Technologies in Health (CADTH); 2007.
19. Köhnlein T, Welte T. Alpha-1 antitrypsin deficiency: Pathogenesis, clinical presentation, diagnosis, and treatment. Am J Med. 2008;121(1):3-9.
20. Kalsheker NA. alpha1-Antitrypsin deficiency: Best clinical practice. J Clin Pathol. 2009;62(10):865-869.

21. Chapman KR, Stockley RA, Dawkins C, et al. Augmentation therapy for alpha1 antitrypsin deficiency: A meta-analysis. J Chronic Obstruct Pulm Dis. 2009;6(3):177-184.

The Pharmacy Policy detailed above has received due consideration and is approved.

Independent medical review – 11/15/2012