



*Easy Choice Health Plan*

*Missouri Care*

*'Ohana Health Plan, a plan offered by WellCare Health Insurance of Arizona*

*OneCare (Care1st Health Plan Arizona, Inc.)*

*Staywell of Florida*

*WellCare (Arizona, Arkansas, Connecticut, Florida, Georgia, Illinois, Kentucky, Louisiana, Mississippi, Nebraska, New Jersey, New York, South Carolina, Tennessee, Texas)*

*WellCare Prescription Insurance*

*WellCare Texan Plus (Medicare – Dallas & Houston markets)*

**Prolastin-C**

**Policy Number: HS-333**

**Original Effective Date: 5/3/2018**

**Revised Date(s): N/A**

## APPLICATION STATEMENT

The application of the Clinical Coverage Guideline is subject to the benefit determinations set forth by the Centers for Medicare and Medicaid Services (CMS) National and Local Coverage Determinations and state-specific Medicaid mandates, if any.

## DISCLAIMER

The Clinical Coverage Guideline (CCG) is intended to supplement certain standard WellCare benefit plans and aid in administering benefits. Federal and state law, contract language, etc. take precedence over the CCG (e.g., Centers for Medicare and Medicaid Services [CMS] National Coverage Determinations [NCDs], Local Coverage Determinations [LCDs] or other published documents). The terms of a member's particular Benefit Plan, Evidence of Coverage, Certificate of Coverage, etc., may differ significantly from this Coverage Position. For example, a member's benefit plan may contain specific exclusions related to the topic addressed in this CCG. Additionally, CCGs relate exclusively to the administration of health benefit plans and are NOT recommendations for treatment, nor should they be used as treatment guidelines. Providers are responsible for the treatment and recommendations provided to the member. The application of the CCG is subject to the benefit determinations set forth by the Centers for Medicare and Medicaid Services (CMS) National and Local Coverage Determinations, and any state-specific Medicaid mandates. Links are current at time of approval by the Medical Policy Committee (MPC) and are subject to change. Lines of business are also subject to change without notice and are noted on [www.wellcare.com](http://www.wellcare.com). Guidelines are also available on the site by selecting the Provider tab, then "Tools" and "Clinical Guidelines".

## BACKGROUND

Deficiency of alpha-1 antitrypsin (AAT) is a genetic condition that can cause early onset pulmonary emphysema as well as several forms of liver disease, including cirrhosis, neonatal hepatitis, and hepatocellular carcinoma. Severe deficiency of AAT is defined as a low serum level of AAT, less than 11 micromol/L or <57 mg/dL by immunodiffusion.

Treatment is similar to the treatment of emphysema related to COPD and includes pulmonary rehabilitation, nutritional support, and supplemental oxygen, preventive vaccination such as the flu and pneumococcal vaccines, and prompt treatment of lower respiratory tract infections.

AAT deficiency can be treated with AAT augmentation therapy. The goal of this treatment is to raise the serum AAT level to a level above the protective threshold. For patients with end stage lung or liver disease, transplant can be necessary.<sup>1</sup>

Prolastin-C is an Alpha1-Proteinase Inhibitor that is indicated for chronic augmentation and maintenance therapy in adults with clinical evidence of emphysema due to severe hereditary alpha1-antitrypsin deficiency. The drug works by increasing antigenic and functional serum levels and antigenic lung epithelial lining fluid levels of Alpha1-PI.

Prolastin-C is administered by injection and dosage is calculated based on patient weight. It is contraindicated in patients with immunoglobulin A (IgA) deficiency with antibodies against IgA as well as patients with a history of anaphylaxis or other severe systemic reaction to Alpha1-PI. The most common adverse reaction during clinical trials was upper respiratory tract infection.<sup>2</sup>

## POSITION STATEMENT

### Applicable To:

- Medicaid – All Markets
- Medicare – All Markets

### Exclusions

PROLASTIN-C is **not considered medically necessary** and not a covered benefit when any of the following apply:

1. Members who are IgA deficient with antibodies against IgA
2. Members who are current smokers
3. Members 17 years old or younger

### Coverage

PROLASTIN-C is **considered medically necessary** for members with congenital deficiency of alpha-1 antitrypsin (alpha-1 proteinase inhibitor) and clinically evident emphysema, and a covered benefit when all of the following criteria apply:

#### Initial Authorization

1. Patient is 18 years or older; **AND**,
2. Patient has diagnosis of congenital deficiency of alpha-1 antitrypsin (AAT) as demonstrated by:
  - a. Genetic test indicating Pi\*ZZ, Pi\*Z(null) or Pi\*(null, null) phenotype (homozygous); **AND**,
  - b. Serum AAT level less than 11 micromol/L (equivalent to <80 mg/dl measured by radial immunodiffusion or <50 mg/dl measured by nephelometry); **AND**,
3. Patient has documented clinical evidence of progressive emphysema, as demonstrated by:
  - a. Pulmonary function tests demonstrating airflow obstruction– reduced forced expiratory volume in 1 second (FEV1) from percent of predicted; **AND**,
  - b. A documented rate of decline in FEV1; **AND**,
4. Patient is currently on maintenance therapy of bronchodilators and other supportive therapy as recommended for treatment of Chronic Obstructive Pulmonary Disease (COPD); **AND**,
5. Medication was prescribed by a pulmonologist or in consultation with a pulmonologist who specializes in the treatment of alpha-1 antitrypsin deficiency.

#### Continuation of Care

1. Member meets initial authorization criteria; **AND**,
2. Demonstrates a clinical response to Prolastin-C, defined as:
  - a. Increase in serum AAT levels above protective threshold (>11 micromol/L, equivalent to >80 mg/dl measured by radial immunodiffusion or >50 mg/dl measured by nephelometry); **AND/OR**,
  - b. Improvement in FEV1 rate of decline.

**Initial Authorization:** 6 months

**Reauthorization:** 12 months

## CODING

### Covered CPT/HCPCS Codes

- J0256** Injection, alpha 1-proteinase inhibitor (human), not otherwise specified, 10 mg
- J0257** Injection, alpha 1 proteinase inhibitor (human), (GLASSIA), 10 mg
- S9346** Home infusion therapy, alpha-1-proteinase inhibitor (e.g., Prolastin); administrative services, professional pharmacy services, care coordination, and all necessary supplies and equipment (drugs and nursing visits coded separately), per diem
- 82103** Alpha-hyphen1-hyphenantitrypsin; total
- 82104** Phenotype

### Covered ICD-10 Codes

- E88.01** Alpha-1-antitrypsin deficiency
- J43.0** Unilateral pulmonary emphysema [MacLeod' syndrome]
- J43.1** Panlobular emphysema
- J43.2** Centrilobular emphysema
- J43.8** Other emphysema
- J43.9** Emphysema, unspecified

Coding information is provided for informational purposes only. The inclusion or omission of a CPT, HCPCS, or ICD-10 code does not imply member coverage or provider reimbursement. Consult the member's benefits that are in place at time of service to determine coverage (or non-coverage) as well as applicable federal / state laws.

## REFERENCES

1. Insert Stoller, J.K. Treatment of alpha-1 antitrypsin deficiency. UpToDate Web site [https://www.uptodate.com/contents/treatment-of-alpha-1-antitrypsin-deficiency?source=history\\_widget#H1859655](https://www.uptodate.com/contents/treatment-of-alpha-1-antitrypsin-deficiency?source=history_widget#H1859655) . Published Apr 20, 2018. Accessed May 9, 2018.
2. Highlights of prescribing information: Prolastin®-C (Alpha1-Proteinase Inhibitor [Human]) Initial U.S. Approval: 1987 <https://www.prolastin.com/documents/31477834/0/prolastinpi/b436e646-0787-4aad-a3d5-b5e004cc92c6>.

## MEDICAL POLICY COMMITTEE HISTORY AND REVISIONS

Date	Action
5/3/2018	<ul style="list-style-type: none"> <li>• Approved by MPC. New.</li> </ul>