CareSource Rxinnovations

PHARMACY POLICY STATEMENT
Indiana Medicaid

DRUG NAME	Alpha$_1$-Proteinase Inhibitor (Aralast NP, Glassia, Prolastin C, Zemaira [human])
BENEFIT TYPE	Medical (Pharmacy allowed for Glassia)
STATUS	Prior Authorization Required

Alpha1-proteinase inhibitor (alpha1 antitrypsin) from pooled human plasma donors acts as augmentation therapy for maintenance treatment in adults with clinical evidence of emphysema due to severe alpha1-antitrypsin deficiency (AATD). The available products are Aralast NP, Glassia, Prolastin C, and Zemaira, with none of them being clinically preferred over the others. Prolastin was the first, approved by the FDA in 1987 (and later replaced by Prolastin C). The goal of this therapy is to restore and maintain alpha1-antitrypsin to protective levels and slow the progression of lung damage and emphysema by inhibiting proteases such as neutrophil elastase.
Alpha-1 antitrypsin deficiency (AATD) is a hereditary disorder caused by pathogenic mutations in the *SERPINA1* gene responsible for producing the protein alpha-1 antitrypsin (AAT) and leads to low levels of AAT. This deficiency results in an imbalance that allows relatively unopposed protease activity to cause destruction in the lungs. The liver, and less likely the skin (panniculitis), can also be affected.

Alpha$_1$-Proteinase Inhibitor (Aralast NP, Glassia, Prolastin C, Zemaira [human]) will be considered for coverage when the following criteria are met:

Alpha$_1$-Antitrypsin Deficiency (AATD)

For **initial** authorization:
1. Member is at least 18 years of age; AND
2. Medication must be prescribed by or in consultation with a pulmonologist; AND
3. Member has a diagnosis of clinically evident emphysema due to severe AATD; AND
4. Member is a never-smoker or has been a non-smoker for at least 3 months; AND
5. Member is in compliance with any prescribed supportive therapy (at least one) (e.g., bronchodilators, pulmonary rehabilitation, oxygen); AND
6. Chart notes must include lab reports showing ALL of the following:
   a) Pre-treatment alpha1-antitrypsin (AAT) serum level less than 11 micromol/L (or equivalent)
   b) High risk genotype (e.g., Pi*ZZ, Pi*ZNull, Pi*NullNull)
   c) Pre-treatment FEV1 is 65% predicted or less; AND
7. Member has NOT had a liver transplant.
   Dosage allowed/Quantity limit: 60 mg/kg once weekly IV infusion.

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:
1. Member continues to abstain from smoking; AND
2. At least ONE of the following:
   a) AAT level at or above protective threshold (11 micromol/L)
   b) Slowed rate of FEV1 decline per spirometry results
   c) CT densitometry demonstrates slowed progression of anatomic lung disease

If all the above requirements are met, the medication will be approved for an additional 12 months.

CareSource considers Alpha$_1$-Proteinase Inhibitor (Aralast NP, Glassia, Prolastin C, Zemaira [human]) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
07/14/2020	Transferred to new template; revised and updated content.
06/29/2023	Transferred to new template. Updated and added references. Removed lower FEV limit and rate of decline. Added liver transplant exclusion.

References:

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4. Sandhaus RA, Turino G, Brantly ML, et al. The Diagnosis and Management of Alpha-1 Antitrypsin Deficiency in the Adult. Chronic Obstructive Pulmonary Diseases: Journal of the COPD Foundation. 2016;3(3):668-682. doi:10.15326/jcopdf.3.3.2015.0182
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6. Gøtzsche PC, Johansen HK. Intravenous alpha-1 antitrypsin augmentation therapy for treating patients with alpha-1 antitrypsin deficiency and lung disease. Cochrane Database of Systematic Reviews 2016, Issue 9. Art. No.: CD007851. DOI: 10.1002/14651858.CD007851.pub3.
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10. Prolastin C [prescribing information]. Grifols Therapeutics LLC; 2020.
11. Zemaira [prescribing information]. CSL Behring LLC; 2022.
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13. Dummer J, Dobler CC, Holmes M, et al. Diagnosis and treatment of lung disease associated with alpha one-antitrypsin deficiency: A position statement from the Thoracic Society of Australia and New Zealand. *Respirology*. 2020;25(3):321-335. doi:10.1111/resp.13774

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