HIGHMARK WHOLECARE

Updated: 7/2025
Approved: 8/2025

Prior Authorization Criteria

Alpha-1 Proteinase Inhibitors

All requests for Alpha-1 Proteinase Inhibitors require a prior authorization and will be screened for medical necessity and appropriateness using the criteria listed below.

Alpha-1 Proteinase Inhibitors include: Aralast NP, Glassia, Prolastin-C and Zemaira. New products with this classification will require the same documentation.

Coverage may be provided with a diagnosis of emphysema due to congenital deficiency of alpha-1 proteinase inhibitor (A-1 PI) and the following criteria is met:

- Member has a diagnosis of congenital alpha-1-antitrypsin deficiency (AATD) confirmed by **ONE** of the following:
  - A high risk AATD genetic variant [e.g., Pi*ZZ, Pi*Z(null), Pi*(null)(null), or Pi*SZ protein phenotypes (homozygous)]
  - Other rare AATD disease-causing alleles associated with serum AAT level < 11 µmol/L
- Member has a baseline circulating serum concentration of AATD < 11 µmol/L using rocket immunoelectrophoresis (which corresponds to < 80 mg/dl if measured by radial immunodiffusion or < 57 mg/dl if measured by nephelometry).
- Member has a diagnosis of clinically evident emphysema confirmed by **ONE** of the following:
  - Forced expiratory volume in one second (FEV1) from ≥ 30% to ≤ 65% of predicted, post-bronchodilator
  - FEV1 from > 65% to < 80% of predicted, post-bronchodilator, and a rapid decline in lung function showing a change in FEV1 > 100 mL/year
- Medication is prescribed by or in consultation with a pulmonologist.
- Prescriber attests that member will continue to be on optimal conventional treatment for emphysema (e.g., bronchodilators, supplemental oxygen, etc.)
- Member is currently a nonsmoker or ex-smoker
- Member must not have a contraindication to therapy such as an Immunoglobulin A (IgA) deficiency with antibodies against IgA.
- Must be age-appropriate according to FDA-approved labeling, nationally recognized compendia, or evidence-based practice guidelines
- The requested dose and frequency is in accordance with FDA-approved labeling, nationally recognized compendia, and/or evidence-based practice guidelines
- **Initial Duration of Approval:** 6 months
- **Reauthorization criteria**
  - Documentation of improvement or stabilization of the signs and symptoms of emphysema associated with alpha-1 antitrypsin deficiency including slowed progression of emphysema as evidenced by annual spirometry testing or a decrease in frequency, duration or severity of pulmonary exacerbations
- **Reauthorization Duration of Approval:** 12 months

Coverage may be provided for any non-FDA labeled indication if it is determined that the use is a medically accepted indication supported by nationally recognized pharmacy compendia or peer-reviewed medical literature for treatment of the diagnosis(es) for which it is prescribed. These requests will be reviewed on a case by case basis to determine medical necessity.

When criteria are not met, the request will be forwarded to a Medical Director for review. The physician reviewer must override criteria when, in their professional judgment, the requested medication is medically necessary.

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ALPHA 1 PROTEINASE INHIBITORS PRIOR AUTHORIZATION FORM

Please complete and fax all requested information below including any progress notes, laboratory test results, or chart documentation as applicable to Highmark Wholecare Pharmacy Services. **FAX:** (888) 245-2049

If needed, you may call to speak to a Pharmacy Services Representative. **PHONE**: (800) 392-1147 Mon – Fri 8:30am to 5:00pm

PROVIDER INFORMATION

Requesting Provider:	Provider NPI:
Provider Specialty:	Office Contact:
State license#:	Office NPI:
Office Address:	Office Phone:
	Office Fax:

MEMBER INFORMATION

Member Name:	DOB:	
Member ID:	Member weight:	Height:

REQUESTED DRUG INFORMATION

Medication:	Strength:	
Directions:	Quantity:	Refills:
Is the member currently receiving requested medication? ☐ Yes ☐ No	Date Medication Initiated:	

Billing Information

This medication will be billed: ☐ at a pharmacy **OR**
☐ medically (if medically please provide a JCODE:

Place of Service: ☐ Hospital ☐ Provider's office ☐ Member's home ☐ Other

Place of Service Information

Name:	NPI:
Address:	Phone:

MEDICAL HISTORY (Complete for ALL requests)

Diagnosis: ICD-10:

Does the member have a diagnosis of congenital alpha-1-antitrypsin (AAT) deficiency confirmed by any of the following:

a. Pi*ZZ, Pi*Z(null), Pi*(null)(null), or Pi*SZ protein phenotypes (homozygous)
☐ Yes ☐ No

b. Other rare AAT deficiency disease-causing alleles associated with serum AAT level < 11 μmol/L
☐ Yes ☐ No

Does the member have circulating serum concentration of AAT < 11 μmol/L (which corresponds to < 80 mg/dl if measured by radial immunodiffusion or < 57 mg/dl if measured by nephelometry)? ☐ Yes ☐ No

Does the member have a diagnosis of emphysema confirmed by any of the following:

a. Forced expiratory volume in one second (FEV1) from ≥ 30% to ≤ 65% of predicted, post-bronchodilator ☐ Yes ☐ No

b. FEV1 from > 65% to < 80% of predicted, post-bronchodilator, and a rapid decline in lung function showing a change in FEV1 > 100 mL/year ☐ Yes ☐ No

Does the prescriber attest that the member will continue to be on optimal conventional treatment for emphysema (e.g., bronchodilators, supplemental oxygen, etc.)? ☐ Yes ☐ No

Is the member currently a nonsmoker? ☐ Yes ☐ No

CURRENT or PREVIOUS THERAPY

Medication Name	Strength/ Frequency	Dates of Therapy	Status (Discontinued & Why/Current)

REAUTHORIZATION

Has the member experienced a significant improvement with treatment? ☐ Yes ☐ No

Please describe:

SUPPORTING INFORMATION or CLINICAL RATIONALE

Prescribing Provider Signature	Date