HARVARD PILGRIM HEALTH CARE
RECOMMENDED MEDICATION REQUEST GUIDELINES

ALPHA-1 PROTEINASE INHIBITORS

Generic	Brand	HICL	GCN	Exception/Other
ALPHA-1-PROTEINASE INHIBITOR	ARALAST NP, PROLASTIN C, ZEMAIRA		47570, 47571, 44288	

GUIDELINES FOR USE

1. Is the request for a patient diagnosed with emphysema due to alpha-1 antitrypsin (AAT) deficiency?

   If yes, continue to #2.
   If no, do not approve. Please use status code #238 and the denial text provided.
   DENIAL TEXT: Per your health plan's Alpha-1 Proteinase Inhibitors guideline, this medication is only covered for emphysema due to alpha-1 antitrypsin (AAT) deficiency. Your provider did not indicate that you have this condition and therefore your request was not approved.

2. Does the patient meet **ALL** of the following criteria?
   - Patient displays clinically evident emphysema
   - Pretreatment serum alpha-1 antitrypsin (ATT) level is less than 80 mg/dL (11 µMl/L)
   - CT scan confirmation of panacinar emphysema
   - Patient has one of the high-risk phenotypes (PI*ZZ, PI*Z[null], or PI*[null][null])
   - Patient is currently a non-smoker
   - Airflow obstruction evidenced by **ONE** of the following:
     - Forced expiratory volume (FEV1) of 30-65% of predicted value; **OR**
     - Rapid decline in lung function as measured by a change in FEV1 greater than 120 ml/year

   If yes, **approve open ended by HICL.** Please use status code #057 and the following approval language:
   APPROVAL TEXT: Your request for _____ has been approved.

   If no, do not approve. Please use status code #238 and the denial text provided.
   DENIAL TEXT: Per your health plan's Alpha-1 Proteinase Inhibitors guideline, this medication is only covered for emphysema due to alpha-1 antitrypsin (AAT) deficiency if you met all of the following criteria:
   - You have clinically evident emphysema
   - Pretreatment serum alpha-1 antitrypsin (ATT) level is less than 80 mg/dL (11 µMl/L)
   - CT scan confirmation of panacinar emphysema
   - You have one of the high-risk phenotypes (PI*ZZ, PI*Z[null], or PI*[null][null])
   - You are currently a non-smoker
   - Airflow obstruction evidenced by **ONE** of the following:
     - Forced expiratory volume (FEV1) of 30-65% of predicted value; **OR**
     - Rapid decline in lung function as measured by a change in FEV1 greater than 120 ml/year

   Your provider did not indicate that **[criteria not met leading to denial]** and therefore your request was not approved.

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RATIONALE

Ensure appropriate utilization of Aralast NP, Prolastin-C, or Zemaira for the treatment of panacinar emphysema due to alpha-1 antitrypsin deficiency when such use (including dosage, frequency, site of administration, and duration of therapy) is reasonable, medically necessary, clinically appropriate, and supported by evidence-based literature.

FDA APPROVED INDICATIONS

- Aralast NP [alpha$_1$-porteinase inhibitor, human (alpha$_1$-antitrypsin)]:
  - **Alpha-proteinase inhibitor deficiency:** Long-term augmentation and maintenance therapy in adults with congenital deficiency of alpha-proteinase inhibitor with clinically evident emphysema.
  - **Limitations of use:** Clinical data demonstrating the long-term effects of chronic augmentation or maintenance therapy of individuals with alpha$_1$-proteinase inhibitor are not available. The effect of augmentation therapy with alpha$_1$-proteinase inhibitor on pulmonary exacerbations and on the progression of emphysema in alpha$_1$-proteinase inhibitor deficiency has not been demonstrated in randomized, controlled clinical trials. Alpha$_1$-proteinase inhibitor is not indicated as therapy for patients with lung disease in whom congenital alpha$_1$-proteinase inhibitor deficiency has not been established.

- Prolastin-C [alpha$_1$-porteinase inhibitor, human (alpha$_1$-antitrypsin)]:
  - **Alpha-proteinase inhibitor deficiency:** Long-term augmentation and maintenance therapy in adults with congenital deficiency of alpha-proteinase inhibitor with clinically evident emphysema.
  - **Limitations of use:** Clinical data demonstrating the long-term effects of chronic augmentation or maintenance therapy of individuals with alpha$_1$-proteinase inhibitor are not available. The effect of augmentation therapy with alpha$_1$-proteinase inhibitor on pulmonary exacerbations and on the progression of emphysema in alpha$_1$-proteinase inhibitor deficiency has not been demonstrated in randomized, controlled clinical trials. Alpha$_1$-proteinase inhibitor is not indicated as therapy for patients with lung disease in whom congenital alpha$_1$-proteinase inhibitor deficiency has not been established.

- Zemaira [alpha$_1$-porteinase inhibitor, human (alpha$_1$-antitrypsin)]:
  - **Alpha-proteinase inhibitor deficiency:** Long-term augmentation and maintenance therapy in adults with congenital deficiency of alpha-proteinase inhibitor with clinically evident emphysema.
  - **Limitations of use:** Clinical data demonstrating the long-term effects of chronic augmentation or maintenance therapy of individuals with alpha$_1$-proteinase inhibitor are not available. The effect of augmentation therapy with alpha$_1$-proteinase inhibitor on pulmonary exacerbations and on the progression of emphysema in alpha$_1$-proteinase inhibitor deficiency has not been demonstrated in randomized, controlled clinical trials. Alpha$_1$-proteinase inhibitor is not indicated as therapy for patients with lung disease in whom congenital alpha$_1$-proteinase inhibitor deficiency has not been established.

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REFERENCES

- Aralast NP [package insert]. Westlake Village, CA: Baxter Healthcare Corporation; March 2014.
- Glassia [package insert]. Westlake Village, CA: Baxter Healthcare Corporation; March 2014.
- Prolastin-C [package insert]. Research Triangle Park, NC: Grifols Therapeutics, Inc.; November 2013.
- Zemaira [package insert]. Kankakee, IL: CSL Behring LLC; September 2015.
- 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.
- Marciniuk D, Hernandez P, Balter M, et al; Canadian Thoracic Society COPD Clinical Assembly Alpha-1 Antitrypsin Deficiency Expert Working Group. Alpha-1 antitrypsin deficiency targeted testing and augmentation therapy: a Canadian Thoracic Society clinical practice guideline. *Can Respir J.* 2012;19(2):109-116.

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