

Alpha-1 Proteinase Inhibitors (Prolastin-C®, Zemaira®)



Pharmacy Coverage Policy

Effective Date: January 01, 2024
Revision Date: August 28, 2024
Review Date: August 21, 2024
Line of Business: Medicaid - Louisiana
Policy Type: Prior Authorization

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Products Affected

Zemaira intravenous solution
Prolastin-C intravenous solution

Listed Indications

[Congenital Alpha1-antitrypsin Deficiency](#)

Congenital Alpha1-antitrypsin Deficiency

Does the member meet all of the following criteria?

Criteria #1	The member has a diagnosis of congenital alpha1-antitrypsin deficiency with clinically evident emphysema and chronic replacement therapy is needed.
Criteria #2	The member has an alpha1-antitrypsin phenotype of PiZZ, PiZ(null), or Pi (null, null) or phenotypes associated with serum alpha 1-antitrypsin concentrations of less than 57mg/dL if/when measured by laboratories using nephelometry instead of radial immunodiffusion. Otherwise, a deficiency is shown at 80mg/dL. (These products should not be used in individuals with the PiMZ or PiMS phenotypes of alpha1- antitrypsin deficiency because these individuals appear to be at small risk of developing clinically evident emphysema.)
Criteria #3	Member has had previous treatment, contraindication, or intolerance to both of the following: Aralast NP AND Glassia.

Does the member have any of the following exclusions? If yes, approval may not be appropriate.

NOTE: Experimental/Investigational Use – Indications not supported by CMS recognized compendia or acceptable peer reviewed literature.

Exclusion #1	IgA deficient members or presence of antibodies against IgA.
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Approval Duration

Initial	plan year duration
Renewal	plan year duration

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Background

This is a prior authorization policy about Alpha-1 Proteinase Inhibitors (Prolastin-C, Zemaira).

Alpha-1 antitrypsin (a1-PI) deficiency is a chronic and hereditary disorder. It usually manifests in the third or fourth decades of life. The panacinar emphysema that develops is usually worse in the lower areas of the lung. The pathogenesis of emphysema in patients with this deficiency is not well understood. Alpha-1 Proteinase Inhibitors (Prolastin-C, Zemaira) are products that work to replace the anti-elastase activity that is missing in patients with a1-PI deficiency.

Some other key points include: Alpha1-PI is not indicated as therapy for lung disease patient in whom congenital alpha1-PI is not established. In some adults, alpha1-antitrypsin deficiency is complicated by cirrhosis or panniculitis.

Alpha-1 Proteinase inhibitors are contraindicated in IgA deficient patients with antibodies against IgA, since these products may contain trace amounts of IgA and cause an increased risk for severe hypersensitivity.

Alpha-1 Proteinase Inhibitors (Prolastin-C®, Zemaira®)

Effective Date: 1/1/2024

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Line of Business: Medicaid - Louisiana

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The long-term effects of chronic replacement therapy with alpha1-PI in individuals having emphysema due to alpha1-antitrypsin deficiency are not known because of inadequate clinical data (the number of patients is small, and the course of disease is variable and slowly progressive).

	<u>PROLASTIN-C</u>	<u>ARALAST NP</u>	<u>ZEMAIRA</u>	<u>GLASSIA</u>
<u>Entered market</u>	February 1988	May 2003	July 2003	October 2010
<u>Marketed by</u>	Grifols USA	Takeda	CSL Behring	Takeda
<u>Recommended dose</u>	60mg/kg IV weekly	60mg/kg IV weekly	60mg/kg IV weekly	60 mg/kg IV weekly
<u>How Supplied</u>	1G/50ml	1G/50ml and 0.5G/50ml	1G/20ml, 4G/76mL, 5G/95mL	1G/50ml vial
<u>Storage</u>	Refrigerated 2-8° C/36-46° F or at temperatures not to exceed 25° C or 77° F Do not freeze	Refrigerated 2-8° C/35-46° F or at temperatures not to exceed 25° C or 77° F Use product removed from refrigeration within one month Do not freeze	Up to 25°C or 77°F Do not freeze	Refrigerated 2-8° C/36-46° F Do not freeze Brief excursions to room temperature are acceptable
<u>Dilutant (sterile water)</u>	20ml for 1000mg vial	25ml 500mg vial 50ml 1,000mg vial	20ml 1,000mg vial 76 mL 4,000 mg vial 95 mL 5,000 mg vial	None (comes as a liquid ready to use). Volume of product 50ml for 1000mg vial
<u>Infusion rate</u>	0.08 ml/kg per min	0.08 ml/kg per min	0.08 ml/kg per min	0.2 ml/kg per min
<u>Infusion time (approximate)</u>	15 minutes	15 minutes	15 minutes	15 minutes
<u>Contraindications</u>	Individuals with known selective IgA deficiency with antibodies against IgA	Individuals with known selective IgA deficiency (< 15mg/dl) with antibodies against IgA	Individuals with known selective IgA deficiency with antibodies against IgA	Individuals with known selective IgA deficiency with antibodies against IgA
<u>Common side effects</u>	Chills, malaise, headache, rash, hot flush, pruritis	Headache, somnolence, chills, fever, vasodilation, pruritus, (itching) rash, abnormal vision, chest pain, increased cough, dyspnea	Asthenia (weakness) injection site pain, dizziness, headache, paresthesia (abnormal skin sensations) and Pruritis (itching)	Headache, dizziness
<u>Viral inactivation processes</u>	Solvent detergent Nanofiltration	Solvent detergent Nanofiltration	Pasteurization Dual ultra-filtration	Solvent detergent Nanofiltration
<u>Number for reporting adverse events</u>	800-520-2807	888-675-2762	800-504-5434	877-825-3327

Alpha-1 Proteinase Inhibitors (Prolastin-C, Zemaira) are lyophilized preparations of purified human alpha1-proteinase inhibitors (a1-PI), also known as alpha1-antitrypsin.

These products inhibit neutrophil elastase (NE) which degrades protein components of the alveolar walls. Patients with a1-PI deficiency have little protection from the harmful effects of NE. Severe forms of this deficiency lead to panacinar emphysema which significantly shortens life expectancy.

Alpha-1 Proteinase Inhibitors (Prolastin-C, Zemaira) are indicated for chronic augmentation and maintenance therapy of individuals having congenital deficiency of a1-PI with clinically demonstrable panacinar emphysema.

Alpha-1 Proteinase Inhibitors (Prolastin-C, Zemaira) are available in vials containing the labeled amount of functionally active a1-PI, usually 1.0gm in powder form that must be reconstituted before use.

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Provider Claim Codes

For medically billed requests, please visit www.humana.com/PAL. Select applicable Preauthorization and Notification List(s) for medical and procedural coding information.

Medical Terms

Prolastin-C; Zemaira; infusion; Alpha-1 Proteinase inhibitors; antitrypsin; chronic augmentation therapy; chronic replacement therapy; pharmacy

References

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2. [IBM Micromedex® DRUGDEX® \[database online\]. Cambridge, MA: IBM Corporation; URL: http://www.micromedexsolutions.com/. Updated periodically.](#)
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4. [Prolastin-C \[package insert\] Research Triangle Park, NC: Grifols Therapeutics LLC; Jan 2022.](#)
5. [Zemaira \[package insert\] Kankakee, IL: CSL Behring LLC; Jan 2024.](#)

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