

Cinryze (C1 esterase inhibitor [human])
Effective 07/01/2023

Plan	<input type="checkbox"/> MassHealth UPPL <input checked="" type="checkbox"/> Commercial/Exchange	Program Type	<input checked="" type="checkbox"/> Prior Authorization <input type="checkbox"/> Quantity Limit <input type="checkbox"/> Step Therapy
Benefit	<input checked="" type="checkbox"/> Pharmacy Benefit <input checked="" type="checkbox"/> Medical Benefit (NLX)		
Specialty Limitations	These medications have been designated specialty and must be filled at a contracted specialty pharmacy.		
Contact Information	Specialty Medications		
	All Plans	Phone: 866-814-5506	Fax: 866-249-6155
	Non-Specialty Medications		
	MassHealth	Phone: 877-433-7643	Fax: 866-255-7569
	Commercial	Phone: 800-294-5979	Fax: 888-836-0730
	Exchange	Phone: 855-582-2022	Fax: 855-245-2134
	Medical Specialty Medications (NLX)		
	All Plans	Phone: 844-345-2803	Fax: 844-851-0882
Exceptions	N/A		

Overview

Cinryze is indicated for routine prophylaxis against angioedema attacks in adults, adolescents and pediatric patients (6 years of age or older) with hereditary angioedema (HAE).

Coverage Guidelines

Authorization may be granted for members new to the plan who are currently receiving treatment with the requested medication, excluding when the product is obtained as samples or via manufacturer's patient assistance programs.

OR

Authorization may be granted for treatment of Hereditary Angioedema (HAE) when all the following criteria are met:

1. The member is using requested medication for the treatment of acute HAE attacks
2. The requested medication will not be used in combination with any other medication used for the prophylaxis of HAE attacks and ONE of the following criteria is met at the time of diagnosis:
 - a. Documentation that the member has C1 inhibitor deficiency or dysfunction as confirmed by laboratory testing and meets ONE of the following criteria:
 - i. C1 inhibitor (C1-INH) antigenic level below the lower limit of normal as defined by the laboratory performing the test, or
 - ii. Normal C1-INH antigenic level and a low C1-INH functional level (functional C1-INH less than 50% or C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test).
 - b. Documentation that the member has normal C1 inhibitor as confirmed by laboratory testing and meets ONE of the following criteria:

- i. Member has an F12, angiopoietin-1, plasminogen, kininogen-1 (KNG1), heparan sulfate-glucosamine 3-O-sulfotransferase 6 (HS3ST6), or myoferlin (MYOF) gene mutation as confirmed by genetic testing, or
 - ii. Member has a documented family history of angioedema and the angioedema was refractory to a trial of high-dose antihistamine therapy (i.e., cetirizine at 40 mg per day or the equivalent) for at least one month.
- 3. Member has had inadequate response, adverse reaction or contraindication to generic Firazyr (icatibant)
- 4. This medication is prescribed by or in consultation with a prescriber who specializes in the management of HAE.

Continuation of Therapy

Reauthorization will be granted for HAE when provider submits the following:

- a. Member meets the criteria for initial approval.
- b. Physician attestation that the member has experienced a reduction in severity and/or duration of acute attacks (e.g., $\geq 50\%$) since starting treatment.
- c. Prophylaxis should be considered based on the attack frequency, attack severity, comorbid conditions, and member's quality of life.

Limitations

- 1. Initial approvals and reauthorizations will be granted for 6 months

References

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3. Cicardi M, Bork K, Caballero T, et al. Evidence-based recommendations for the therapeutic management of angioedema owing to hereditary C1 inhibitor deficiency: consensus report of an International Working Group. *Allergy*. 2012;67:147-157.
4. Bowen T, Cicardi M, Farkas H, et al. 2010 International consensus algorithm for the diagnosis, therapy, and management of hereditary angioedema. *Allergy Asthma Clin Immunol*. 2010;6(1):24.
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14. Bernstein, J. Severity of Hereditary Angioedema, Prevalence, and Diagnostic Considerations. *Am J Med*. 2018;24:292-298.
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Review History

05/10/2023 – Created for May P&T; switched from CVS SGM to custom. Effective 7/1/23

