Mass General Brigham
Health Plan

Cinryze (C1 esterase inhibitor [human])
Effective 01/01/2024

Plan	☐ MassHealth UPPL ☒Commercial/Exchange	**Program Type**	☒ Prior Authorization ☐ Quantity Limit ☐ Step Therapy
Benefit	☒ Pharmacy Benefit ☒ Medical Benefit		
Specialty Limitations	This medication has been designated specialty and must be filled through a contracted specialty pharmacy.		
Contact Information	**Medical and Specialty Medications**		
	All Plans	Phone: 877-519-1908	Fax: 855-540-3693
	Non-Specialty Medications		
	All Plans	Phone: 800-711-4555	Fax: 844-403-1029
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 prophylaxis 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. 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., ≥ 50%) since starting treatment.

Limitations

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

References

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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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Review History

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

11/15/2023 – Reviewed and Updated for Nov P&T; diagnosis changed from treatment to prophylaxis. Removed requirement of prior use of generic Firazyr as that is only used for treatment. Effective 1/1/2024