Reference number(s)
1605-A

ENHANCED SPECIALTY GUIDELINE MANAGEMENT

CINRYZE (C1 esterase inhibitor [human])

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications
Routine prophylaxis against angioedema attacks in adults, adolescents and pediatric patients (6 years of age or older) with hereditary angioedema (HAE)

All other indications are considered experimental/investigational and are not a covered benefit.

II. REQUIRED DOCUMENTATION

The following information is necessary to initiate the prior authorization review: C4 levels and C1 inhibitor functional and antigenic protein levels.

III. CRITERIA FOR INITIAL APPROVAL

Authorization of 12 months may be granted for prevention of hereditary angioedema attacks when either of the following criteria is met:
A. Member has C1 inhibitor deficiency or dysfunction as confirmed by laboratory testing.
B. Member has normal C1 inhibitor as confirmed by laboratory testing and meets one of the following criteria:
    1. Member has an F12, angiopoietin-1, or plasminogen gene mutation as confirmed by genetic testing, or
    2. Member has a family history of angioedema and the angioedema was refractory to a trial of high-dose antihistamine (e.g., cetirizine) for at least one month.

IV. CONTINUATION OF THERAPY

Authorization of 12 months may be granted for continuation of therapy when all of the following criteria are met:
A. Member meets the criteria for initial approval.
B. Member has experienced reduction in frequency, severity and duration of attacks since starting treatment.

V. REFERENCES

1. Cinryze [package insert]. Lexington, MA: ViroPharma Biologics; June 2018.
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3. Cicardi M, Bork K, Caballero T, et al. Hereditary Angioedema International Working Group. 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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10. Bernstein J. Update on angioedema: Evaluation, diagnosis, and treatment. *Allergy and Asthma Proceedings.* 2011;32(6):408-412.
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12. Farkas H, Martinez-Saguer I, Bork K, et al. International consensus on the diagnosis and management of pediatric patients with hereditary angioedema with C1 inhibitor deficiency. *Allergy*. 2017;72(2):300-313.