Custom Criteria for
BlueCross Blue Shield of Rhode Island Commercial

CINRYZE (C1 inhibitor)

GPI CODING:

85802022002120

DESCRIPTION:

Cinryze is a human plasma-derived C1 esterase inhibitor concentrates. Cinryze is FDA-approved for the treatment of acute abdominal, facial, or laryngeal attacks of hereditary angioedema (HAE) in adult and adolescent patients. HAE is caused by a deficiency in C1 esterase inhibitor. Patients with HAE experience recurrent episodes of angioedema also known as HAE attacks. Management of HAE involves long-term prophylaxis, short-term prophylaxis, and treatment of acute attacks.

APPROVAL DURATION:

Long-term prophylaxis of HAE attacks: 3 months for initial, 1 year for renewals
Quantity Limit: 16 vials (8000 units) per month
Quantity Limit: 20 vials (10000 units) per month only when dosing every 4 days was tried but was inadequate

CRITERIA FOR CINRYZE

I. The following criteria will be applied to all requests for Cinryze:

A. Patient has a diagnosis of HAE that was established by, or in consultation with, an allergist, immunologist or a hematologist **AND**
B. HAE was confirmed by genetic testing for mutations in the C1 esterase inhibitor gene **OR**
C. HAE was confirmed by laboratory testing for C4 and C1 inhibitor levels AND patient had low C4 level and low C1 inhibitor level at time of diagnosis **AND**
D. Patient has been evaluated for potentially treatable triggers of HAE attacks and is maximally managed with respect to avoiding triggers **AND**
E. Patient is not currently taking a medication known to trigger or worsen HAE (eg, estrogen contraceptives, HRT, ACE inhibitors) **AND**
F. Patient has a history of attacks that are considered severe with swelling of the face, throat or gastrointestinal tract (severe is defined as events that significantly interrupt usual daily activity despite short term symptomatic treatment)

II. Additional criteria for use of Cinryze for long-term prophylaxis of frequent and severe HAE attacks:

A. Unless Cinryze is prescribed during pregnancy, patient has tried androgens and/or antifibrinolytics (eg, danazol, tranexamic acid) for long-term prophylaxis **AND**
    a. Androgens and/or antifibrinolytics were ineffective or not tolerated **OR**
    b. Patient has a documented contraindication to androgens or antifibrinolytics **AND**
B. For patients who have previously received Cinryze for long-term prophylaxis (reauthorization), Cinryze was effective as defined by all of the following:
    a. The frequency of HAE attacks decreased by ≥ 50% **AND**
    b. There is significant improvement in severity and duration of HAE attacks **AND**
    c. There is clinical documentation of functional improvement **AND**
C. The requested quantity of Cinryze does not exceed the limit shown in the Quantity Limits above