Custom Criteria for
BlueCross Blue Shield of Rhode Island Commercial

BERINERT (C1 inhibitor)

GPI CODING:

85802022006420

DESCRIPTION:

Berinert is a human plasma-derived C1 esterase inhibitor concentrates. Berinert 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. These attacks usually involve the arms, legs, hands, feet, bowels, genitalia, trunk, face, tongue, or larynx. Management of HAE involves long-term prophylaxis, short-term prophylaxis, and treatment of acute attacks.

APPROVAL DURATION:

Acute treatment of HAE attacks or short-term prophylaxis
One-time approval
Quantity Limit:

Weight	Max. # Vials	Max. # Units
> 50 kg	3 vials	1500 units
> 26 kg to ≤ 50 kg	2 vials	1000 units
≤ 25 kg	1 vial	500 units

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

CRITERIA FOR BERINERT

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

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 Berinert for treatment of acute HAE attacks:

A. Berinert is the requested product OR the prescriber is willing to prescribe Berinert as an alternative to Cinryze **AND**
B. For patients who have previously received Berinert for treating acute HAE attacks (reauthorization):
   i. Documentation of the date(s), number of injections used, and description of attack(s) since the last prescription was filled **AND**
   ii. If the patient has had 2 or more attacks per month, he/she has been evaluated for prophylactic HAE treatment **AND**
C. The requested quantity of Berinert does not exceed the limit shown in the Quantity Limits above

III. Additional criteria for use of Berinert for short-term prophylaxis prior to a medical procedure, dental work, or other triggering situation:

A. Berinert is the requested product OR the prescriber is willing to prescribe Berinert as an alternative to Cinryze **AND**
B. The requested quantity of Berinert does not exceed the limit shown in the Quantity Limits above

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

A. Unless Berinert 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 Berinert for long-term prophylaxis (reauthorization), Berinert 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 Berinert does not exceed the limit shown in the Quantity Limits above