**Cinryze**

**Description**

Cinryze (C1 esterase inhibitor [human])

**Background**

Cinryze is a C1-esterase inhibitor used for the treatment routine prophylaxis against angioedema attacks with hereditary angioedema (HAE). Hereditary angioedema is caused by having insufficient amounts of a plasma protein called C1-esterase inhibitor. People with HAE can develop rapid swelling of the hands, feet, limbs, face, intestinal tract, or airway. These acute attacks of swelling can occur spontaneously, or can be triggered by stress, surgery or infection. Swelling of the airway is potentially fatal without immediate treatment. Cinryze is intended to restore the level of functional C1-esterase inhibitor in a patient’s plasma, thereby preventing the acute attack of swelling (1-4).

**Regulatory Status**

FDA-approved indication: Cinryze is a C1 esterase inhibitor indicated for routine prophylaxis against angioedema attacks in adults, adolescents and pediatric patients (6 years of age and older) with Hereditary Angioedema (HAE) (2).

Hypersensitivity reactions may occur. Epinephrine should be immediately available to treat any acute severe hypersensitivity reactions following discontinuation of administration (2).

Thrombotic events have been reported at the recommended dose of C1 Esterase Inhibitor (human) products, including Cinryze, following treatment of HAE. Monitor closely patients with known risk factors for thrombotic events (2).
Cinryze is made from human plasma and may contain infectious agents, e.g., viruses and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent (2).

**Related policies**
Berinert, Firazyr, Haegarda, Kalbitor, Ruconest, Takhzyro

**Policy**
*This policy statement applies to clinical review performed for pre-service (Prior Approval, Precertification, Advanced Benefit Determination, etc.) and/or post-service claims.*

Cinryze may be considered **medically necessary** in patients 6 years of age or older for the routine prevention of hereditary angioedema (HAE) attacks and if the conditions indicated below are met.

Cinryze may be considered **investigational** in patients less than 6 years of age and for all other indications.

**Prior-Approval Requirements**

**Age**
6 years of age and older

**Diagnosis**

Patient must have **ALL** of the following:

1. Hereditary Angioedema (HAE)
   a. Routine prevention of angioedema attacks
   b. **NO** dual therapy with other agents for the prevention of hereditary angioedema attacks
   c. Inadequate treatment response or intolerance to a short-term course (5-days or less) of an androgen such as danazol, or a contraindication to one such as:
      i. Undiagnosed abnormal genital bleeding
      ii. Markedly impaired hepatic, renal, or cardiac function
      iii. Pregnancy (member is currently pregnant or may become pregnant)
   iv. Breast feeding
   v. Porphyria
   vi. Androgen-dependent tumor
vii. Active thrombosis or history of thromboembolic disease  
viii. Prepubertal child

Prior – Approval Renewal Requirements

Age  6 years of age and older

Diagnosis

Patient must have ALL of the following:

1. Hereditary Angioedema (HAE)
   a. Routine prevention of angioedema attacks
   b. NO dual therapy with other agents for the prevention of hereditary angioedema attacks

Policy Guidelines

Pre - PA Allowance

None

Prior - Approval Limits

Duration  12 months

Prior – Approval Renewal Limits

Duration  12 months

Rationale

Summary
Cinryze is a C1 esterase inhibitor indicated for routine prophylaxis against angioedema attacks in adolescent and adult patients with Hereditary Angioedema (HAE). HAE symptoms include episodes of edema (swelling) in various body parts including the hands, feet, face, and airway. HAE is caused by mutations to C1-esterase-inhibitor (C1-INH). Serious arterial and venous thromboembolic (VTE) events have been reported at the recommended dose of plasma derived...
C1 esterase inhibitor products in patients with risk factors. The safety and efficacy of Cinryze in children less than 6 years of age has not been established. Persons who experience frequent and/or severe episodes may be candidates for prophylactic treatment (2).

Prior authorization is required to ensure the safe, clinically appropriate and cost effective use of Cinryze while maintaining optimal therapeutic outcomes.

References

Policy History

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<td>Addition of no dual therapy with other C1-esterase inhibitors for the</td>
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**Keywords**

This policy was approved by the FEP® Pharmacy and Medical Policy Committee on September 13, 2019 and is effective on October 1, 2019.