



## Pre - PA Allowance

None

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## Prior-Approval Requirements

**Age** 6 years of age and older

### Diagnosis

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

1. Hereditary Angioedema (HAE) with **ONE** of the following:
  - a. Patient has a C1 inhibitor deficiency or dysfunction as confirmed by laboratory testing **AND ALL** of the following:
    - i. C4 level below the lower limit of normal as defined by the laboratory performing the test
    - ii. C1 inhibitor (C1-INH) antigenic level below the lower limit of normal as defined by the laboratory performing the test **OR** 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. Patient has normal C1 inhibitor as confirmed by laboratory testing **AND ONE** of the following:
    - i. F12, angiopoietin-1, plasminogen, or kininogen-1 (KNG1) gene mutation as confirmed by genetic testing
    - ii. Documented family history of angioedema and the angioedema was refractory to a trial of high-dose antihistamine (e.g., cetirizine) for at least one month

**AND ALL** of the following:

- a. Routine prevention of hereditary angioedema attacks
- b. **NO** dual therapy with other agents for the prevention of hereditary angioedema attacks (e.g., Haegarda, Orladeyo, Takhzyro)
- 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



**BlueCross  
BlueShield**

Federal Employee Program.

## **CINRYZE**

**(C1 esterase inhibitor [human])**

- v. Porphyria
- vi. Androgen-dependent tumor
- vii. Active thrombosis or history of thromboembolic disease
- viii. Prepubertal child

## **Prior - Approval Limits**

**Duration** 12 months

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## **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 hereditary angioedema attacks
  - b. Patient has experienced a significant reduction in frequency of hereditary angioedema attacks since starting treatment
  - c. **NO** dual therapy with other agents for the prevention of hereditary angioedema attacks (e.g., Haegarda, Orladeyo, Takhzyro)

## **Prior - Approval *Renewal* Limits**

Same as above