



Guidelines for Medication Coverage:

C1 Inhibitor (Human) – Cinryze

Background:

Hereditary angioedema (HAE) is a rare genetic disorder resulting from low levels of C1 Inhibitor. C1 inhibitor maintains the natural regulation of the contact, complement, and fibrinolytic systems, that when left unrestricted, can initiate or perpetuate an attack by consuming the already low levels of endogenous C1 inhibitor. The condition is characterized by recurrent, unpredictable and potentially life-threatening episodes of edema that may affect the larynx, abdomen, face, extremities, and urogenital tract.

Cinryze (C1 Inhibitor – Human) is the first product approved for routine prophylaxis against angioedema attacks in adolescent and adult patients with hereditary angioedema.

Administration:

Cinryze is administered as a 1000mg dose (two 500mg vials) administered by intravenous infusion twice a week.

Note: Severe hypersensitivity reactions including anaphylaxis have occurred during and after Cinryze infusions.

Criteria for approval:

1. Candidates for Cinryze should be under the care of an allergist, immunologist or hematologist; **AND**
2. Candidates for Cinryze should be at least 9 years of age and have a confirmed diagnosis of hereditary angioedema; **AND**
3. The dose of Cinryze shall not exceed 1000mg (2 vials) administered every twice a week.
4. The initial authorization period shall be 12 months. Therapy may be re-authorized for 12 month periods for patients who experience a positive response to Cinryze.

Cinryze is administered by intravenous infusion in an ambulatory setting and is covered under the medical benefit for commercial and Medicare enrollees.

Authorization Procedure:

- 1) All courses of Cinryze require prior authorization.
- 2) Authorization Period:
Initial Authorization: Six months
Subsequent Authorizations: 12 months
- 3) Prescribers should contact the Pharmacy Authorization Unit at (330) 996-8805 for authorization or further information. Fax requests may be sent to (330) 996-8580.
- 4) The prescriber and member will be notified of authorization or denial as delineated in the SummaCare Pharmacy Benefit Management policies and procedures. The Pharmacy Management Staff will perform any necessary override/exception entry into the appropriate claims processing system.

Approved: SummaCare Pharmacy & Therapeutics Committee May 7, 2009

Medical Director

Pharmacy Director

References:

Frank J et al. New Therapies for Hereditary Angioedema: Disease Outlook Changes Dramatically. JACI. 2008; 121 (1): 272-280.

Cinryze Prescribing Information. Lev Pharmaceuticals. October 2008