

Cinryze (Human C1 Esterase Inhibitor)

Cinryze will be covered when all of the criteria below are met, confirmed with supporting medical documentation.

I. Criteria for Initial Approval

- Diagnosis of hereditary angioedema (HAE) confirmed by one of the following:
 - Confirmed monoallelic mutation known to cause HAE in either the SERPING1 or F12 gene; OR
 - A C4 level below the lower limit of normal and either C1 inhibitor (C1-INH) antigenic level below the lower limit of normal or C1-INH functional level below the lower limit of normal; AND
- Used for prophylaxis of acute HAE attacks; AND
- Patient is at least 6 years of age; AND
- Patient has experienced the following:
 - History of recurrent laryngeal attacks; OR
 - > 2 severe episodes/month (ex. debilitating GI or cutaneous effects); OR
 - > 5 days/month of debilitating symptoms; AND
- Prescribed by an allergist, immunologist, hematologist or other appropriate specialist; AND
- Medications known to cause angioedema (ex. ACE-Inhibitors, estrogens, angiotensin II receptor blockers) have been evaluated and discontinued when appropriate

II. Criteria for Continuation of Therapy

- All of the criteria for initial therapy in Section I. must be met; AND
- Documented improvement in frequency, severity and/or duration of attacks has been achieved and sustained

III. Dosing/Administration

Cinryze must be administered according to the current FDA labeling guidelines for dosage and timing. The recommended dosing is as follows:

- Ages 6-11 years: 500 units IV every 3-4 days (doses up to 1000 units every 3-4 days may be considered based on individual patient response)
- Ages 12 and older: 1000 units IV every 3-4 days (doses up to 2500 units every 3-4 days may be considered based on individual patient response, not exceeding 100 units/kg)

Maryland Medicaid Medical Drug Policy

IV. Length of Authorization

For initial therapy, Cinryze will be authorized for 6 months when criteria are met. Continuing therapy with Cinryze will be authorized for 12 months.

Maryland Medicaid considers Cinryze investigational and not medically necessary when the criteria above are not met and for all other indications.