

C1 Inhibitor (CINRYZE, HAEGARDA) for Hereditary Angioedema Prophylaxis Criteria for Use August 2026

VA Pharmacy Services and National Formulary Committee

The following recommendations are based on medical evidence, clinician input, and expert opinion. The content of the document is dynamic and will be revised as new information becomes available. The purpose of this document is to assist practitioners in clinical decision-making, to standardize and improve the quality of patient care, and to promote cost-effective drug prescribing. THE CLINICIAN SHOULD USE THIS GUIDANCE AND INTERPRET IT IN THE CLINICAL CONTEXT OF THE INDIVIDUAL PATIENT. INDIVIDUAL CASES THAT ARE EXCEPTIONS TO THE EXCLUSION AND INCLUSION CRITERIA SHOULD BE ADJUDICATED AT THE LOCAL FACILITY ACCORDING TO THE POLICY AND PROCEDURES OF ITS P&T COMMITTEE AND PHARMACY SERVICES.

The Product Information should be consulted for detailed prescribing information.

Exclusion Criteria

If ANY of the following are selected, the patient will NOT meet criteria for C1 Inhibitor.

- ☐ Angioedema or abdominal pain not associated with C1 inhibitor deficiency, including hereditary angioedema with normal C1 inhibitor (HAE nC1-INH)¹
- ☐ Concomitant use with another prophylaxis agent for HAE (e.g., berotralstat, donidalorsen, or lanadelumab)

Inclusion Criteria

All the following must be selected to meet criteria.

- ☐ Restricted to VA / VA Community Care allergy/immunology provider, dermatology provider, or locally designated expert
- ☐ Diagnosis of Hereditary Angioedema Type 1 or 2 as established by laboratory testing²
- ☐ Long-term prophylaxis for HAE attacks or short-term prophylaxis prior to procedures or intubation

Other Justification

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Footnotes

1. The most common subtype of HAE with C1 inhibitor deficiency (also identified as HAE types 1 and 2) is a result of SERPING1 variants. HAE with normal C1 inhibitor (HAE-nC1-INH) is caused by variants in other genes like FXII, PLG, KNG1, ANGPT1, MYOF, HS3ST6, CPN1, or DAB21P, and unidentified genetic causes.
2. Diagnostic laboratory results include a C4 level below the limit of normal and one of the following: C1 inhibitor (C1-INH) antigenic level below the limit of normal or C1-INH functional level below the limit of normal.

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