Prior Authorization Group Description	**Treatment of Hereditary Angioedema (HAE)**
Drugs	**Preferred:** Berinert (C1 esterase inhibitor, human) Ruconest (C1 esterase inhibitor, recombinant) **Non-preferred:** Cinryze (C1 esterase inhibitor, human) Kalbitor (ecallantide)
Covered Uses	Medically accepted indications are defined using the following sources: the Food and Drug Administration (FDA), Micromedex, American Hospital Formulary Service (AHFS), United States Pharmacopeia Drug Information for the Healthcare Professional (USP DI), the Drug Package Insert (PPI), or disease state specific standard of care guidelines.
Exclusion Criteria	N/A
Required Medical Information	See "Other Criteria"
Age Restrictions	According to package insert
Prescriber Restrictions	Prescriber must be an immunologist, allergist, rheumatologist, or hematologist
Coverage Duration	If criteria are met, the request will be approved as follows: • Acute treatment: 1 + 5 refills • Pre procedural prophylaxis: 1 treatment • Long-term prophylaxis: o Initial: 6 months o Reauthorization: 12 months
Other Criteria	**Initial Requests:** • Documentation submitted indicates the medication is being prescribed at FDA approved dose. • The patient is not taking ACE inhibitors or estrogen containing oral contraceptives/hormone replacement therapy • Diagnosis of one of the following**:** o HAE with deficient or dysfunctional C1INH (e.g. type I, type II, or acquired C1NH deficiency o HAE with normal C1INH: ▪ If known origin, documentation of results of confirmatory genetic test (e.g. mutations in gene for factor XII, angiopoietin-1, plasminogen, kininogen-1) ▪ If unknown origin (U-HAE), documentation of a prolonged trial of high-dose non-sedating antihistamines **For acute treatment:** o The patient is receiving only one agent for the treatment of acute attacks o If the request is for a non-preferred agent, the member has documented trial and failure of, or intolerance to a preferred

Revision/Review Date: 4/2023	agent or medical reason why the member cannot use a preferred agent **For prophylaxis:** • Pre-procedural o Documentation that patient will be undergoing a medical, surgical, or dental procedure associated with mechanical impact to the upper aerodigestive tract • Long-term o The patient has a history of at least two severe attacks/month (e.g. with swelling of the face, throat, or GI tract) or at least one laryngeal attack, and chart notes have been submitted indicating the date and severity of attack. o The patient is only receiving one medication for long-term prophylaxis • If the request is for a non-preferred agent o And the patient has a C1INH deficiency or dysfunction, documented trial and failure of or medical reason why patient cannot use a preferred agent o And the patient has HAE with normal C1INH, documented trial and failure of, or documented medical reason why patient cannot use danazol **<u>Renewal Criteria:</u>** **For acute treatment:** • Documentation was submitted that the patient has clinically benefited from medication • The patient is receiving no other medications for acute treatment • The medication is being prescribed at FDA approved dose. **For prophylaxis:** • Documentation was submitted that the patient has clinically benefited from prophylactic therapy as demonstrated by a reduced number of attacks • The medication is being prescribed at an FDA approved dose • The patient is receiving no other medications for prophylaxis . **NOTE: Medical Director/Clinical Reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary**