group health Cooperative of eau claire	DEPARTMENT:	Utilization Management
	SUBJECT:	C1 Esterase Inhibitors
	PRODUCT LINE:	All
	POLICY NUMBER:	078
	ORIGINAL POLICY EFFECTIVE DATE:	01/01/2016
KMTSJ, Inc.	LAST REVISED DATE:	05/06/2019
	LAST REVIEWED DATE:	02/12/2024

SCOPE: To ensure Group Health Cooperative of Eau Claire (the Cooperative) consistently and correctly administers C1 Esterase Inhibitor benefits to all members according to their policy specifics.

POLICY: It is the policy of the Cooperative to review requests for human C1 esterase inhibitors (e.g., Berinert, Cinryze, Ruconest) for prior authorization according to evidence based medical criteria.

PROCEDURE: Prior authorization required: Yes

Definition:

Hereditary angioedema (HAE) is a rare disease that causes swelling of the face, hands, feet, throat, stomach, bowels, or sexual organs. People who have HAE have low levels of C1 esterase inhibitor in their bodies.

Commercial Members: Covered benefit when medical criteria below are met, and it is delivered in the least restrictive environment.

Medicaid Members: Covered benefit when medical criteria below are met, and it is delivered in the least restrictive environment.

Criteria to determine medical necessity: In order to meet medical necessity guidelines for long term, routine prophylaxis against angioedema attacks in members with hereditary angioedema (HAE), all the following criteria must be met:

1. Diagnosis of hereditary angioedema (HAE) with laboratory confirmation including one of the following:
   A. Type I defined as serum C4 < 14 mg/dL and C1 inhibitor (C1 INH) < 19.9 mg/dL; **or**
   B. Type II defined as functional C1 INH < 72%; **or**
   C. A known HAE-causing C1 INH mutation; **and**
2. Appropriate age of member (≥ 6 for Berinert, ≥12 for Kalbitor, ≥ 13 for Ruconest, ≥ 18 for Firazyr); **and**
3. Use is to prevent future attacks when there is a history of at least two attacks a month with at least one symptom of a moderate or severe attack including one of the following:
   A. airway swelling; **or**
   B. nausea and vomiting or severe abdominal pain; **or**
   C. facial swelling or painful distortion of the face; **and**,

4. Member has tried and failed, has a contraindication to, or intolerance of 17 alpha-alkylated androgens (e.g., danazol and stanozolol) or anti-fibrinolytic agents (e.g., aminocaproic acid [Amikar], tranexamic acid [Cyklokapron]) for HAE prophylaxis

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APPROVED:  DATE: 02/12/2024

REVISION HISTORY:

Rev. Date	Revised By/Title	Summary of Revision
04/20/2018	Michele Bauer, MD, CMO	Reviewed with no changes.
05/06/2019	Michele Bauer, MD, CMO	Updated criteria
04/01/2020	Michele Bauer, MD	Reviewed with no updates
03/10/2021	Michele Bauer, MD, CMO	Reviewed. No updates
02/10/2022	Michele Bauer, MD, CMO	Reviewed. No changes.
02/15/2023	Michele Bauer, MD, CMO	Reviewed. No changes.
02/12/2024	Dakota Rau, PharmD	Reviewed. No changes.