

Pharmacy Medical Necessity Guidelines: Haegarda® (C1 Esterase Inhibitor [Human])

Effective: November 16, 2020

Prior Authorization Required	√	Type of Review – Care Management	
Not Covered		Type of Review – Clinical Review	√
Pharmacy (RX) or Medical (MED) Benefit	RX	Department to Review	RXUM
<p>These pharmacy medical necessity guidelines apply to the following:</p> <p>Commercial Products</p> <ul style="list-style-type: none"> <input checked="" type="checkbox"/> Tufts Health Plan Commercial products – large group plans <input checked="" type="checkbox"/> Tufts Health Plan Commercial products – small group and individual plans <input checked="" type="checkbox"/> Tufts Health Freedom Plan products – large group plans <input checked="" type="checkbox"/> Tufts Health Freedom Plan products – small group plans • CareLinkSM – Refer to CareLink Procedures, Services and Items Requiring Prior Authorization <p>Tufts Health Public Plans Products</p> <ul style="list-style-type: none"> <input checked="" type="checkbox"/> Tufts Health Direct – A Massachusetts Qualified Health Plan (QHP) (a commercial product) <input checked="" type="checkbox"/> Tufts Health Together – MassHealth MCO Plan and Accountable Care Partnership Plans <input checked="" type="checkbox"/> Tufts Health RITogether – A Rhode Island Medicaid Plan 		<p>Fax Numbers:</p> <p>RxUM: 617-673-0988</p>	

Note: This guideline does not apply to Medicare Members (includes dual eligible Members).

OVERVIEW

FOOD AND DRUG ADMINISTRATION-APPROVED INDICATIONS

Haegarda (C1 Esterase Inhibitor [Human]) is a plasma-derived concentrate of C1 Esterase Inhibitor (Human) indicated for routine prophylaxis to prevent hereditary angioedema (HAE) attacks in patients 6 years of age and older.

Hereditary angioedema is a rare, episodic, autosomal dominant, swelling disorder that is characterized by C1 esterase inhibitor (C1-INH) deficiency. C1-INH coordinates the activation of the complement, contact, and fibrinolytic systems. A reduction in the activity of C1-INH may result in an elevated level of bradykinin, which is a key mediator in HAE symptoms. Patients with HAE may experience attacks of swelling and inflammation in the extremities, abdomen, face, urogenital tract, and/or the larynx that are random, recurrent, and potentially life-threatening. The age of onset is variable, ranging from early childhood to adult, with a worsening in frequency occurring around puberty. The age of onset can help to differentiate between HAE and acquired angioedema (AAE), which normally does not present until the fourth decade of life.

Treatment of HAE is divided into acute treatment, short-term/procedural prophylaxis to prevent an attack, and long-term/routine prophylaxis to minimize the frequency and severity of attacks. Long-term prophylaxis is recommended for patients who experience more than one attack per month, or for those who feel the condition is significantly impacting their lives. Short-term prophylaxis is recommended before dental procedures, minor surgery, endoscopy, or any situation where trauma may precipitate an attack; however, there are no FDA-approved agents currently available for procedural prophylaxis. A multifaceted approach that uses both pharmacologic and supportive therapies is required for optimal prevention and treatment of HAE.

COVERAGE GUIDELINES

The plan may authorize coverage of Haegarda (C1 esterase inhibitor) for Members when all of the following criteria are met:

Initial Criteria

1. Documented diagnosis of hereditary angioedema by an immunologist or allergist
- AND**
2. The Member is 6 years of age or older
- AND**
3. Documentation that “on-demand” therapy (e.g., icatibant, Kalbitor, Ruconest, or Berinert) did not provide satisfactory control or access to “on-demand” therapy is limited
- AND**
4. Documentation the Member is NOT concurrently taking an angiotensin converting enzyme (ACE) inhibitor or estrogen replacement therapy

AND

5. Documentation the Member has had an insufficient response or contraindication to both of the following classes of medication:
 - a. 17 α -alkylated androgens (e.g., danazol, stanozolol, oxandrolone, methyltestosterone)
 - b. Antifibrinolytic agents (e.g., aminocaproic acid, tranexamic acid)

Reauthorization Criteria

1. The prescribing physician is an immunologist or allergist

AND

2. The Member is 6 years of age or older

AND

3. Documentation of improvement in severity and duration of attacks has been achieved and sustained

LIMITATIONS

- Haegarda (C1 esterase inhibitor) will be authorized for 12 months.
- Members new to the plan stable on Haegarda (C1 esterase inhibitor) should be reviewed against Initial criteria.

CODES

None

REFERENCES

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2. Cicardi M, Bork K, Caballero T, et al. Evidence-based recommendations for the therapeutic management of angioedema owing to hereditary C1 inhibitor deficiency: consensus report of an International Working Group. *Allergy*. 2012; 67:147-57.
3. Craig T, Riedl M, Dykewicz MS et al. When is prophylaxis for hereditary angioedema necessary? *Ann Allergy Asthma Immunol*. 2009; 102:366-72.
4. Epstein TG, Bernstein JA. Current and emerging management options for hereditary angioedema in the US. *Drugs*. 2008; 68(18):2561-73.
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8. Maurer M, Magerl M, Ansotegui I, et al. The international WAO/EAACI guideline for the management of hereditary angioedema – the 2017 revision and update. *World Allergy Organization Journal*. 2018;11(5):1-20.
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12. Zuraw B, Bernstein J, Lang D et al. A focused parameter update: hereditary angioedema, acquired C1 inhibitor deficiency, and angiotensin-converting enzyme inhibitor-associated angioedema. *J Allergy Clin Immunol*. 2013b; 131(6): 1491-3.

APPROVAL HISTORY

October 17, 2017: Reviewed by Pharmacy & Therapeutics Committee.

Subsequent endorsement date(s) and changes made:

1. January 1, 2018: Administrative update: Added new C code C9015 to the Medical Necessity Guideline.
2. September 8, 2018: No changes. Administrative update to remove the HCPC code C9015 because Haegarda is administered under the pharmacy benefit only.
3. February 12, 2019: No changes.
4. March 12, 2019: Effective July 1, 2019, added an age requirement and allergist provider specialty to the Medical Necessity Guideline.

5. February 11, 2020: Effective April 1, 2020, updated approval duration to 12 months and added Reauthorization criteria. Updated initial therapy criteria to remove documentation of a specific frequency and type of history of attacks. Documentation that “on-demand” therapy (e.g., icatibant, Kalbitor, Ruconest, or Berinert) did not provide satisfactory control or access to “on-demand” therapy is limited is required.
6. November 10, 2020: Updated the age requirements based on expanded indication for use in patients at least 6 years of age.

BACKGROUND, PRODUCT AND DISCLAIMER INFORMATION

Pharmacy Medical Necessity Guidelines have been developed for determining coverage for plan benefits and are published to provide a better understanding of the basis upon which coverage decisions are made. The plan makes coverage decisions on a case-by-case basis considering the individual member's health care needs. Pharmacy Medical Necessity Guidelines are developed for selected therapeutic classes or drugs found to be safe, but proven to be effective in a limited, defined population of patients or clinical circumstances. They include concise clinical coverage criteria based on current literature review, consultation with practicing physicians in the service area who are medical experts in the particular field, FDA and other government agency policies, and standards adopted by national accreditation organizations. The plan revises and updates Pharmacy Medical Necessity Guidelines annually, or more frequently if new evidence becomes available that suggests needed revisions.

For self-insured plans, coverage may vary depending on the terms of the benefit document. If a discrepancy exists between a Pharmacy Medical Necessity Guideline and a self-insured Member's benefit document, the provisions of the benefit document will govern.

Treating providers are solely responsible for the medical advice and treatment of members. The use of this policy is not a guarantee of payment or a final prediction of how specific claim(s) will be adjudicated. Claims payment is subject to member eligibility and benefits on the date of service, coordination of benefits, referral/authorization and utilization management guidelines when applicable, and adherence to plan policies and procedures and claims editing logic.

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