



AETNA BETTER HEALTH®  
Coverage Policy/Guideline

Name: Haegarda

Page: 1 of 3

Effective Date: 9/22/2023

Last Review Date: 9/22/2023

Applies to:	<input checked="" type="checkbox"/> Illinois	<input type="checkbox"/> Florida	<input type="checkbox"/> Michigan
	<input type="checkbox"/> New Jersey	<input checked="" type="checkbox"/> Maryland	<input checked="" type="checkbox"/> Florida Kids
	<input checked="" type="checkbox"/> Pennsylvania Kids	<input type="checkbox"/> Virginia	<input checked="" type="checkbox"/> Kentucky PRMD

### Intent:

The intent of this policy/guideline is to provide information to the prescribing practitioner outlining the coverage criteria for Haegarda under the patient's prescription drug benefit.

### Description:

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

#### FDA-Approved Indication

Haegarda is indicated for routine prophylaxis to prevent Hereditary Angioedema (HAE) attacks in patients 6 years of age and older.

All other indications are considered experimental/investigational and not medically necessary.

### Applicable Drug List:

Haegarda

### Policy/Guideline:

#### Documentation:

Submission of the following information is necessary to initiate the prior authorization review:

- A. For initial authorization, the following should be documented:
  1. C1 inhibitor functional and antigenic protein levels
  2. F12, angiopoietin-1, plasminogen, kininogen-1 (KNG1), heparan sulfate-glucosamine 3-O- sulfotransferase 6 (HS3ST6), or myoferlin (MYOF) gene mutation testing, if applicable
  3. Chart notes confirming family history of angioedema and the angioedema was refractory to a trial of high-dose antihistamine therapy, if applicable
- B. For continuation of therapy, chart notes demonstrating a reduction in frequency of attacks

### Prescriber Specialty:

This medication must be prescribed by or in consultation with a prescriber who specializes in the management of HAE.



AETNA BETTER HEALTH®  
Coverage Policy/Guideline

Name: Haegarda

Page: 2 of 3

Effective Date: 9/22/2023

Last Review Date: 9/22/2023

Applies to:	<input checked="" type="checkbox"/> Illinois	<input type="checkbox"/> Florida	<input type="checkbox"/> Michigan
	<input type="checkbox"/> New Jersey	<input checked="" type="checkbox"/> Maryland	<input checked="" type="checkbox"/> Florida Kids
	<input checked="" type="checkbox"/> Pennsylvania Kids	<input type="checkbox"/> Virginia	<input checked="" type="checkbox"/> Kentucky PRMD

### Criteria for Initial Approval:

#### Hereditary angioedema (HAE)

Authorization of 6 months may be granted for prevention of hereditary angioedema attacks when the requested medication will not be used in combination with any other medication used for prophylaxis of HAE attacks and either of the following criteria is met at the time of diagnosis:

- A. Member has C1 inhibitor deficiency or dysfunction as confirmed by laboratory testing and meets one of the following criteria:
  1. C1 inhibitor (C1-INH) antigenic level below the lower limit of normal as defined by the laboratory performing the test, or
  2. Normal C1-INH antigenic level and a low C1-INH functional level (functional C1-INH less than 50% or C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test).
- B. Member has normal C1 inhibitor as confirmed by laboratory testing and meets one of the following criteria:
  1. Member has an F12, angiopoietin-1, plasminogen, kininogen-1 (KNG1), heparan sulfate-glucosamine 3-O-sulfotransferase 6 (HS3ST6), or myoferlin (MYOF) gene mutation as confirmed by genetic testing, or
  2. Member has a documented family history of angioedema and the angioedema was refractory to a trial of high-dose antihistamine therapy (i.e., cetirizine at 40 mg per day or the equivalent) for at least one month.

#### Continuation of Therapy:

Authorization of 6 months may be granted for continuation of therapy when all of the following criteria are met:

- A. Member meets the criteria for initial approval.
- B. Member has experienced a significant reduction in frequency of attacks (e.g.,  $\geq 50\%$ ) since starting treatment.
- C. Member has reduced the use of medications to treat acute attacks since starting treatment.

#### Approval Duration and Quantity Restrictions:

**Approval:** 6 months

**Quantity Level Limit:** 20 vials every 30 days



AETNA BETTER HEALTH®  
Coverage Policy/Guideline

Name: Haegarda

Page:

3 of 3

Effective Date: 9/22/2023

Last Review Date: 9/22/2023

Applies to:	<input checked="" type="checkbox"/> Illinois	<input type="checkbox"/> Florida	<input type="checkbox"/> Michigan
	<input type="checkbox"/> New Jersey	<input checked="" type="checkbox"/> Maryland	<input checked="" type="checkbox"/> Florida Kids
	<input checked="" type="checkbox"/> Pennsylvania Kids	<input type="checkbox"/> Virginia	<input checked="" type="checkbox"/> Kentucky PRMD

## References:

1. Haegarda [package insert]. Kankakee, IL: CSL Behring LLC; January 2022.
2. Maurer M, Magerl M, Ansotegui I, et al. The international WAO/EAACI guideline for the management of hereditary angioedema – the 2021 revision and update. *Allergy*. 2022 Jan 10. doi: 10.1111/all. 15214. Online ahead of print.
3. 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;67:147-157.
4. Bowen T, Cicardi M, Farkas H, et al. 2010 International consensus algorithm for the diagnosis, therapy, and management of hereditary angioedema. *Allergy Asthma Clin Immunol*. 2010;6(1):24.
5. Busse PJ, Christiansen, SC, Riedl MA, et al. US HAEA Medical Advisory Board 2020 Guidelines for the Management of Hereditary Angioedema. *J Allergy Clin Immunol: In Practice*. 2021 Jan;9(1):132-150.e3.
6. Zuraw BL, Bork K, Binkley KE, et al. Hereditary angioedema with normal C1 inhibitor function: consensus of an international expert panel. *Allergy Asthma Proc*. 2012; 33(6):S145-S156.
7. Lang DM, Aberer W, Bernstein JA, et al. International consensus on hereditary and acquired angioedema. *Ann Allergy Asthma Immunol*. 2012; 109:395-402.
8. Cicardi M, Aberer W, Banerji A, et al. Classification, diagnosis, and approach to treatment for angioedema: consensus report from the Hereditary Angioedema International Working Group. *Allergy*. 2014;69: 602-616.
9. Bowen T. Hereditary angioedema: beyond international consensus – circa December 2010 – The Canadian Society of Allergy and Clinical Immunology Dr. David McCourtie Lecture. *Allergy Asthma Clin Immunol*. 2011;7(1):1.
10. Bernstein JA. Update on angioedema: Evaluation, diagnosis, and treatment. *Allergy and Asthma Proceedings*. 2011;32(6):408-412.
11. Longhurst H, Cicardi M. Hereditary angio-edema. *Lancet*. 2012;379:474-481.
12. [Farkas H](#), [Martinez-Saguer I](#), [Bork K](#), et al. International consensus on the diagnosis and management of pediatric patients with hereditary angioedema with C1 inhibitor deficiency. *Allergy*. 2017;72(2):300-313.
13. Henao MP, Kraschnewski J, Kelbel T, Craig T. Diagnosis and screening of patients with hereditary angioedema in primary care. *Therapeutics and Clin Risk Management*. 2016; 12: 701-711.
14. Bernstein, J. Severity of Hereditary Angioedema, Prevalence, and Diagnostic Considerations. *Am J Med*. 2018;24: 292-298.
15. Sharma J, Jindal AK, Banday AZ, et al. Pathophysiology of Hereditary Angioedema (HAE) Beyond the SERPING1 Gene [published online ahead of print, 2021 Jan 14] [published correction appears in *Clin Rev Allergy Immunol*. 2021 Feb 17]. *Clin Rev Allergy Immunol*. 2021;10.1007/s12016-021-08835-8. Doi:10.1007/s12016-021-08835-8.
16. Kanani, A., Schellenberg, R. & Warrington, R. Urticaria and angioedema. *All Asth Clin Immunol* 7, S9 (2011), Table 2.