

Haegarda (C1 Esterase Inhibitor Subcutaneous [Human])
Effective 01/01/2023

Plan	<input type="checkbox"/> MassHealth UPPL <input checked="" type="checkbox"/> Commercial/Exchange	Program Type	<input checked="" type="checkbox"/> Prior Authorization <input checked="" type="checkbox"/> Quantity Limit <input type="checkbox"/> Step Therapy
Benefit	<input checked="" type="checkbox"/> Pharmacy Benefit <input checked="" type="checkbox"/> Medical Benefit (NLX)		
Specialty Limitations	This medication has been designated specialty and must be filled at a contracted specialty pharmacy.		
Contact Information	Specialty Medications		
	All Plans	Phone: 866-814-5506	Fax: 866-249-6155
	Non-Specialty Medications		
	MassHealth	Phone: 877-433-7643	Fax: 866-255-7569
	Commercial	Phone: 800-294-5979	Fax: 888-836-0730
	Exchange	Phone: 855-582-2022	Fax: 855-245-2134
	Medical Specialty Medications (NLX)		
	All Plans	Phone: 844-345-2803	Fax: 844-851-0882
Exceptions	N/A		

Overview

Haegarda (C1 Esterase Inhibitor Subcutaneous [Human]) is to prevent attacks of hereditary angioedema (HAE) in adults and pediatric patients 6 years and older.

Coverage Guidelines

Authorization may be granted for members new to the plan who are currently receiving treatment with Haegarda, excluding when the product is obtained as samples or via manufacturer's patient assistance programs.

OR

Authorization may be granted for members when ALL the following criteria are met, and documentation is provided:

1. The member will be using Haegarda for the prevention of hereditary angioedema attacks
2. Haegarda will not be used in combination with any medication used for the prophylaxis of HAE attacks.
3. Member meets ONE of the following:
 - a. Member has C1 inhibitor deficiency or dysfunction as confirmed by laboratory testing and meets BOTH of the following criteria:
 - i. Member has a C4 level below the lower limit of normal as defined by the laboratory performing the test
 - ii. Member meets ONE of the following criteria:
 - (a) C1 inhibitor (C1-INH) antigenic level below the lower limit of normal as defined by the laboratory performing the test
 - (b) 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:
 - i. Member has an F12, angiopoietin-1, plasminogen, or kininogen-1 (KNG1) gene mutation as confirmed by genetic testing
 - ii. Member has a documented family history of angioedema and the angioedema was refractory to a trial of high-dose antihistamine (e.g., cetirizine) for at least one month.
4. Member has adverse effect or contraindication to icatibant (generic for Firazyr)

Continuation of Therapy

Reauthorization may be granted for members when ALL of the following are met, and documentation is provided:

1. Member meets all initial approval criteria.
2. Member has experienced a significant reduction in frequency of attacks (e.g. $\geq 50\%$) since starting treatment.
3. Member has reduced the use of medications to treat acute attacks.

Limitations

1. Initial approvals and reauthorizations will be for 6 months.

References

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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.
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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. Henaó 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*.



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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.
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Review History

05/19/2021 – Created and Reviewed May P&T. Effective 07/01/2021.

9/21/2022 – Reviewed and Updated for Sept P&T; added requirement of adverse effect or contraindication to icatibant (generic for Firazyr). Separated out MH vs. Comm/Exch. Effective 1/1/2023.

