

Market Applicability														
Market	DC	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	X

\*FHK- Florida Healthy Kids

## Haegarda (C1 esterase inhibitor [Human])

DRUG.00058

Override(s)	Approval Duration
Prior Authorization Quantity Limit	1 year

Medications	Quantity Limit
Haegarda (C1 esterase inhibitor [Human]) subcutaneous injection	May be subject to quantity limit

### APPROVAL CRITERIA

Requests for Haegarda (C1 esterase inhibitor [Human]) may be approved for *prophylaxis* against acute attacks of hereditary angioedema in individuals who meet the following criteria:

- I. Individual is 12 years of age or older; **AND**
- II. Individual has a diagnosis of Hereditary Angioedema (HAE) confirmed by a C4 level below the lower limit of normal as defined by the laboratory performing the test **AND** ANY of the following (A, B, or C);
  - A. C1 inhibitor (C1-INH) antigenic level below the lower limit of normal as defined by the laboratory performing the test; **OR**
  - B. C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test; **OR**
  - C. The presence of a known HAE-causing C1-INH mutation; **AND**
- III. There is a history of moderate or severe attacks (for example, airway swelling, severe abdominal pain, facial swelling, nausea and vomiting, painful facial distortion); **AND**
- IV. Haegarda is being used for prophylaxis for EITHER of the following (a or b) indications:
  - A. Short term prophylaxis prior to surgery, dental procedures or intubation; **OR**
  - B. Long-term prophylaxis and the individual has failed, is intolerant to, or has a contraindication (for example, under the age of 12, or pregnant, or breastfeeding) to 17 alpha-alkylated androgens (for example, danazol) or antifibrinolytic agents (for example, aminocaproic acid).

This policy does not apply to health plans or member categories that do not have pharmacy benefits, nor does it apply to Medicare. Note that market specific restrictions or transition-of-care benefit limitations may apply.

Market Applicability														
Market	DC	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	X

\*FHK- Florida Healthy Kids

State Specific Mandates		
State name	Date effective	Mandate details (including specific bill if applicable)
N/A	N/A	N/A

#### **Key References:**

1. Berinert [Prescribing Information], Kankakee, IL. CSL Behring LLC. September, 2016. Available at: <http://labeling.cslbehring.com/PI/US/Berinert/EN/Berinert-Prescribing-Information.pdf>. Accessed on July 12, 2017.
2. Caballero T, Farkas H, Bouillet L, et al.; C-1-INH Deficiency Working Group. International consensus and practical guidelines on the gynecologic and obstetric management of female patients with hereditary angioedema caused by C1 inhibitor deficiency. *J Allergy Clin Immunol.* 2012; 129(2):308-320.
3. Cicardi M, Aberer W, Banerji A, et al.; HAWK under the patronage of EAACI (European Academy of Allergy and Clinical Immunology). Classification, diagnosis, and approach to treatment for angioedema: consensus report from the Hereditary Angioedema International Working Group. *Allergy.* 2014; 69(5):602-616.
4. Cicardi M, Bork K, Caballero T, et al.; HAWK (Hereditary Angioedema International Working Group). 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(2):147-157.
5. Cinryze [Prescribing Information], Exton, PA. ViroPharma Biologics, Inc. February 11, 2014. Available at: <http://www.fda.gov/downloads/BiologicsBloodVaccines/BloodBloodProducts/ApprovedProducts/LicensedProductsBLAs/FractionatedPlasmaProducts/UCM129918.pdf>. Accessed on July 12, 2017.
6. Craig T, Aygören-Pürsün EA, Bork K, et al. World Allergy Organization (WAO) guideline for the management of hereditary angioedema. *World Allergy Organ J.* 2012; 5(12):182-199.
7. Craig TJ, Bernstein JA, Farkas H, et al. Diagnosis and treatment of bradykinin-mediated angioedema: outcomes from an angioedema expert consensus meeting. *Int Arch Allergy Immunol.* 2014; 165(2):119-127.
8. CSL Behring LLC. A study to evaluate the clinical efficacy and safety of subcutaneously administered C1-esterase inhibitor in the prevention of hereditary angioedema. NLM Identifier: NCT01912456. Last updated June 5, 2017. Available at: <https://www.clinicaltrials.gov/ct2/show/NCT01912456?term=csl+830&rank=3>. Accessed on July 12, 2017.
9. CSL Behring LLC. A study to evaluate the long-term clinical safety and efficacy of subcutaneously administered C1-esterase inhibitor in the prevention of hereditary angioedema. NLM Identifier: NCT02316353. Last updated July 6, 2017. Available at: <https://www.clinicaltrials.gov/ct2/show/NCT02316353?term=csl+830&rank=2>. Accessed on July 12, 2017.
10. Farkas H, Martinez-Saguer I, Bork K, et al. HAWK International consensus on the diagnosis and management of pediatric patients with hereditary angioedema with C1 inhibitor deficiency. *Allergy.* 2017; 72(2):300-313.

This policy does not apply to health plans or member categories that do not have pharmacy benefits, nor does it apply to Medicare. Note that market specific restrictions or transition-of-care benefit limitations may apply.

Market Applicability														
Market	DC	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	X

\*FHK- Florida Healthy Kids

11. Firazyr [Prescribing Information], Lexington, MA. Shire Orphan Therapies, Inc. August 30, 2013. Available at: [http://www.accessdata.fda.gov/drugsatfda\\_docs/label/2013/022150s004lbl.pdf](http://www.accessdata.fda.gov/drugsatfda_docs/label/2013/022150s004lbl.pdf). Accessed on July 12, 2017.
12. Gropmels MM, Lock RJ, Abinun M, et al. C1 inhibitor deficiency: consensus document. Clin Exp Immunol. 2005; 139(3):379-394.
13. Haegarda C1 Esterase Inhibitor Subcutaneous (Human) [Prescribing Information], Kankakee, IL. CSL Behring LLC. June 22, 2017. Available at: <https://www.fda.gov/downloads/BiologicsBloodVaccines/BloodBloodProducts/ApprovedProducts/LicensedProductsBLAs/FractionatedPlasmaProducts/UCM564335.pdf>. Accessed on July 13, 2017.
14. Kalbitor [Prescribing Information], Burlington, MA. Dyax Corporation. March 2014. Available at: [http://www.accessdata.fda.gov/drugsatfda\\_docs/label/2014/125277s070lbl.pdf](http://www.accessdata.fda.gov/drugsatfda_docs/label/2014/125277s070lbl.pdf). Accessed on July 12, 2017.
15. Lang DM, Aberer W, Bernstein JA, et al. International consensus on hereditary and acquired angioedema. Ann Allergy Asthma Immunol. 2012; 109(6):395-402.
16. Mansi M, Zanichelli A, Coerezza A, et al. Presentation, diagnosis and treatment of angioedema without wheals: a retrospective analysis of a cohort of 1058 patients. J Intern Med. 2015; 277(5):585-593.
17. Ruconest [Prescribing Information], Raleigh, NC. Santarus Inc. July 2014. New revision date not provided. Available at: <http://www.fda.gov/downloads/biologicsbloodvaccines/bloodbloodproducts/approvedproducts/licensedproductsblas/fractionatedplasmaproducts/ucm405634.pdf>. Accessed on July 12, 2017.
18. U.S. Hereditary Angioedema Association (HAEA) Advisory Board. HAEA Consensus Document: An approach to diagnosis and treatment of HAE. 2012. Available at: <http://www.angioedemacenter.com/wp-content/uploads/2012/03/HAEA-MAB-Treatment-guidelines-v2.pdf>. Accessed on July 12, 2017.
19. Zuraw BL, Banerji A, Bernstein J, et al. U.S. Hereditary Angioedema Association (US HAE) Medical Advisory Board 2013 recommendations for the management of hereditary angioedema due to C1 inhibitor deficiency. J Allergy Clin Immunol. 2013a; 1(5):458-467. Available at: <http://haei.org/wp-content/uploads/2015/04/Zuraw-B-L-US-HAEA-MAB-2013-Recommendations.pdf>. Accessed on July 12, 2017.
20. Zuraw BL, Bernstein JA, Lang DM, et al. American Academy of Allergy, Asthma & Immunology (AAAAI); American College of Allergy, Asthma & Immunology (ACAAI); and the Joint Council of Allergy, Asthma and Immunology. A focused parameter update: hereditary angioedema, acquired C1 inhibitor deficiency, and angiotensin-converting enzyme inhibitor-associated angioedema. J Allerg Clin Immunol. 2013b; 131(6):1491-1493.

This policy does not apply to health plans or member categories that do not have pharmacy benefits, nor does it apply to Medicare. Note that market specific restrictions or transition-of-care benefit limitations may apply.