Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

Haegarda (C1 esterase inhibitor [Human])

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 the following criteria are met:

I. Individual has a diagnosis of hereditary angioedema; **AND**
II. Individual is using for prophylaxis against acute attacks of hereditary angioedema for either of the following:
    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 (such as pregnant, or breastfeeding) to 17 alpha-alkylated androgens (for example, danazol) or antifibrinolytic agents (for example, aminocaproic acid); **AND**
III. Individual is 12 years of age or older; **AND**
IV. Diagnosis is 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. C1 inhibitor (C1-INH) antigenic level below the lower limit of normal as defined by the laboratory performing the test with documentation provided; **OR**
    B. C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test with documentation provided; **OR**
    C. The presence of a known HAE-causing C1-INH mutation; **AND**
V. Individual has a history of moderate or severe attacks such as airway swelling, severe abdominal pain, facial swelling, nausea and vomiting, or painful facial distortion)**.**

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

Requests for Haegarda may not be approved for all other indications not included above.

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

Key References:

1. Clinical Pharmacology [database online]. Tampa, FL: Gold Standard, Inc.: 2019. URL: http://www.clinicalpharmacology.com. Updated periodically.
2. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: July 8, 2019.
3. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
4. Efficacy ans Safety Study of DX-2930 to Prevent Acute Angioedema Attacks in Patients with Type I and Type II HAE. NCT02586805 (HELP Study). Available at https://www.clinicaltrials.gov/ct2/show/study/NCT02586805. Accessed on July 8, 2019
5. Haegarda [Package Insert]. Marburg, Germany. CSL Behring, GmbH.; 2017.
6. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2019; Updated periodically.
7. Riedl MA, Bernstein JA, Craig T, et al. An open-label study to evaluate the long-term safety and efficacy of lanadelumab for prevention of attacks in hereditary angioedema: design of the HELP study extension. *Clin Transl Allergy*. 2017;7:36.
8. Riedl MA. Creating a Comprehensive Treatment Plan for Hereditary Angioedema. *Immunol Allergy Clin N Am*. 2013; 33 (4): 471-485. **doi:10.1016/j.iac.2013.07.003**.
9. Zuraw BL, Banerji A, Bernstein JA, et al. US Hereditary Angioedema Association Medical Advisory Board 2013 Recommendations for the Management of Hereditary Angioedema Due to C1 Inhibitor Deficiency. *J Allergy Clin Immunol: In Practice*. 2013; 1:458-67. **doi:10.1016/j.jaip.2013.07.002.**
10. Zuraw BL, Bernstein JA, Lang DM, et al. A focused parameter update: Hereditary angioedema, acquired C1 inhibitor deficiency, and angiotensin-converting enzyme inhibitor–associated angioedema. *J Allergy Clin Immunol*. 2013; 131(6):1491-1493.e1-e25. Available from: http://www.jacionline.org/article/S0091-6749(13)00523-X/pdf. Accessed on: July 8, 2019.