Policy Title:	Haegarda (C1 Esterase Inhibitor Subcutaneous [Human]) (subcutaneous)		
		Department:	PHA
Effective Date:	01/01/2020		
Review Date:	09/25/2019, 12/18/19, 1/22/20		
Revision Date:	09/25/2019, 1/22/20		

Purpose: To support safe, effective and appropriate use of Haegarda (C1 Esterase Inhibitor Subcutaneous [Human]).

Scope: Medicaid, Exchange, Medicare-Medicaid Plan (MMP)

Policy Statement:

Haegarda (C1 Esterase Inhibitor Subcutaneous [Human]) is covered under the Medical Benefit when used within the following guidelines. Use outside of these guidelines may result in non-payment unless approved under an exception process.

Procedure:

Coverage of Haegarda (C1 Esterase Inhibitor Subcutaneous [Human]) will be reviewed prospectively via the prior authorization process based on criteria below.

Initial Criteria

- MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements; OR
- Authorization may be granted for prevention of hereditary angioedema attacks when either of the following criteria is met:
    - Member has C1 inhibitor deficiency or dysfunction as confirmed by laboratory testing; OR
    - Member has normal C1 inhibitor as confirmed by laboratory testing and meets one of the following criteria:
        - Member has an F12, angiopoietin-1, or plasminogen gene mutation as confirmed by genetic testing; OR
        - Member has a family history of angioedema and the angioedema was refractory to a trial of high-dose antihistamine (e.g., cetirizine) for at least one month.

Continuation of Therapy Criteria:

- All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

Coverage durations:

- Initial coverage: 6 months
- Continuation of therapy coverage: 6 months

*** Requests will also be reviewed to National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) if applicable. ***

Dosage/Administration:

Indication	Dose	Maximum dose (1 billable unit = 10 IU)
Prophylaxis of Hereditary Angioedema (HAE) attacks	60 IU/kg body weight injected subcutaneously twice weekly (every 3 or 4 days)	5,600 billable units per 28 days

Investigational use: All therapies are considered investigational when used at a dose or for a condition other than those that are recognized as medically accepted indications as defined in any one of the following standard reference compendia: American Hospital Formulary Service Drug information (AHFS-DI), Thomson Micromedex DrugDex, Clinical Pharmacology, Wolters Kluwer Lexi-Drugs, or Peer-reviewed published medical literature indicating that sufficient evidence exists to support use. Neighborhood does not provide coverage for drugs when used for investigational purposes.

Applicable Codes:
Below is a list of billing codes applicable for covered treatment options. The below tables are provided for reference purposes and may not be all-inclusive. Requests received with codes from tables below do not guarantee coverage. Requests must meet all criteria provided in the procedure section.

The following HCPCS/CPT code is:

HCPCS/CPT Code	Description
J0599	Injection, c-1 esterase

References:

1. Haegarda [package insert]. Kankakee, IL: CSL Behring LLC; October 2017.
2. Maurer M, Magerl M, Ansotegui I, et al. The international WAO/EAACI guideline for the management of hereditary angioedema – the 2017 revision and update. *Allergy*. 2018;00:1-22.
3. Cicardi M, Bork K, Caballero T, et al. 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;67:147-157.
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. 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(5): 458-467.
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; 109:395-202.
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.