I. Length of Authorization

Coverage will be provided for 12 months and may be renewed.

II. Dosing Limits

A. Quantity Limit (max daily dose) [Pharmacy Benefit]:
   - Haegarda 2000 IU SDV kit: 16 kits per 28 days
   - Haegarda 3000 IU SDV kit: 8 kits per 28 days

B. Max Units (per dose and over time) [Medical Benefit]:
   - 5,600 billable units per 28 days

III. Initial Approval Criteria

Coverage is provided in the following conditions:

**Prophylaxis to prevent Hereditary Angioedema (HAE) attacks †**

- Must be prescribed by, or in consultation with, a specialist in: allergy, immunology, hematology, pulmonology, or medical genetics; AND
- Patient must be at least 12 years of age; AND
- Patient has a history of one of the following criteria for long-term HAE prophylaxis:
  - History of two (2) or more severe HAE attacks per month (i.e., airway swelling, debilitating cutaneous or gastrointestinal episodes); OR
  - Patient is disabled more than 5 days per month by HAE; OR
  - History of at least one laryngeal attacks caused by HAE: AND
- Treatment of patient with “on-demand” therapy (i.e., Kalbitor, Firazyr, Ruconest, or Berinert) did not provide satisfactory control or access to “on-demand therapy” is limited: AND
- Not used in combination with C1 inhibitor prophylaxis (e.g., Cinryze or Takhzyro): AND
- Confirmation the patient is avoiding the following possible triggers for HAE attacks:
  - Estrogen-containing oral contraceptive agents AND hormone replacement therapy; AND
  - Antihypertensive agents containing ACE inhibitors: AND
- Patient has one of the following clinical presentations consistent with HAE subtype, which must be confirmed by repeat blood testing:

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**HAE I (C1-Inhibitor deficiency)**
This HNE clinical criteria is only a screening tool. It is not for final clinical or payment decisions. All care decisions are solely the responsibility of your healthcare provider. This HNE clinical criteria is confidential and proprietary. It applies only to this review.

- Low C1 inhibitor (C1-INH) antigenic level (C1-INH antigenic level below the lower limit of normal as defined by the laboratory performing the test): **AND**
- Low C4 level (C4 below the lower limit of normal as defined by the laboratory performing the test): **AND**
- Low C1-INH functional level (C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test): **AND**
  - Patient has a family history of HAE: **OR**
  - Onset of HAE symptoms occurred before age 30: **OR**
  - Normal C1q level

**HAE II (C1-Inhibitor dysfunction)**
- Normal to elevated C1-INH antigenic level: **AND**
- Low C4 level (C4 below the lower limit of normal as defined by the laboratory performing the test): **AND**
- Low C1-INH functional level (C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test)

**HAE with normal C1INH (formerly known as HAE III)**
- Prophylaxis for HAE with normal C1-INH is not routinely recommended and will be evaluated on a case by case basis

† FDA Approved Indication(s)

**IV. Renewal Criteria**

Coverage can be renewed based upon the following criteria:

- Patient continues to meet the criteria in section III: **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include the following: severe hypersensitivity reactions, thromboembolic events, etc.: **AND**
  - Significant improvement in severity and duration of attacks have been achieved and sustained: **OR**
  - Patient requires dose titration due to an inadequate response to therapy (> 1.0 HAE attack/month, regardless of severity/duration)

**V. Dosage/Administration**

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
</tr>
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<tbody>
<tr>
<td>Prophylaxis of Hereditary Angioedema (HAE) attacks</td>
<td>60 IU/kg body weight injected subcutaneously twice weekly (every 3 or 4 days)</td>
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</tbody>
</table>

**VI. Billing Code/Availability Information**

**Jcode:**
- J0599 – Injection, c-1 esterase inhibitor (human), (haegarda), 10 units: 1 billable unit = 10 IU (effective 1/1/19)
- J3590 – Unclassified Biologics
• C9015 – Injection, c-1 esterase inhibitor (human), Haegarda, 10 units: 1 billable unit = 10 IU. (inactive 1/1/2019)

NDC:
• Haegarda 2000 IU single-use vial kit: 63833-0828-xx
• Haegarda 3000 IU single-use vial kit: 63833-0829-xx

VII. References


Appendix 1 – Covered Diagnosis Codes

<table>
<thead>
<tr>
<th>ICD-10</th>
<th>ICD-10 Description</th>
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<td>C9015</td>
<td>Injection, c-1 esterase inhibitor (human), Haegarda, 10 units: 1 billable unit = 10 IU.</td>
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Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. They can be found at: http://www.cms.gov/medicare-coverage-database/search/advanced-search.aspx. Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD):

N/A

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<td>Noridian Healthcare Solutions, LLC</td>
</tr>
<tr>
<td>F (2 &amp; 3)</td>
<td>AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ</td>
<td>Noridian Healthcare Solutions, LLC</td>
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<tr>
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<td>KS, NE, IA, MO</td>
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</tr>
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<tr>
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<td>First Coast Service Options, Inc.</td>
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<tr>
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<td>Novitas Solutions, Inc.</td>
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