

<b>POLICY NAME</b>	Hereditary Angioedema	<b>POLICY #</b>	2608P
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## Criteria

### Coverage Criteria for Acute Attacks (Berinert, Firazyr, icatibant, Sajazir, Kalbitor, Ruconest)

- ☐ **1.1** Diagnosis of C1INH-HAE (formerly types I/II)
  - Evidence of low C4 level (<14 mg/dL AND evidence of low C1 inhibitor (C1INH <19.9 mg/dL) OR low C1INH functional level (functional C1INH < 72%)
- ☐ **1.2** Prescribed by a(n) allergist (allergy doctor), immunologist (immune system doctor), or rheumatologist (musculoskeletal doctor)
- ☐ **1.3** Appropriate age per approved FDA labeling
- ☐ **1.4** Presence of acute hereditary angioedema (HAE) attack confirmed by one or more symptoms (airway swelling, severe abdominal pain, facial swelling, throat swelling, nausea and vomiting, painful face distortion)
- ☐ **1.5** Medications associated with angioedema (e.g., ACE inhibitors, ARBs, NSAIDs, estrogens) have been evaluated and, if appropriate, discontinued
- ☐ **1.6** Request for coverage is reviewed by both a pharmacist and a medical director
- ☐ **1.7** Initial Approval: 12 months, Reapproval: 12 months with documented clinical benefit

### Coverage Criteria for Prophylaxis (Cinryze, Haegarda, Orladeyo, Takhzyro)

- ☐ **2.1** Diagnosis of C1INH-HAE (formerly types I/II)
- ☐ **2.2** Appropriate age per approved FDA labeling
- ☐ **2.3** History of at least one HAE attack, without the presence of urticaria (raised, itchy rash), per month
- ☐ **2.4** Prescribed by a(n) allergist (allergy doctor), immunologist (immune system doctor), or rheumatologist (musculoskeletal doctor)
- ☐ **2.5** Medications associated with angioedema (e.g., ACE inhibitors, ARBs, NSAIDs, estrogens) have been evaluated and, if appropriate, discontinued
- ☐ **2.6** Trial and failure, intolerance, or contraindication to any one of the following:

**alpha-alkylated androgens (stanozolol, danazol), OR Anti-fibrinolytic agents (aminocaproic acid, tranexamic acid)**

- ☐ **2.7** Request for coverage is reviewed by both a pharmacist and a medical director
- ☐ **2.8** Initial Approval: 12 months, Reapproval: 12 months with documented clinical benefit

**Exclusion Criteria – Any of the following prevents coverage**

- ☐ **3.1** Treatment of any other form of chronic, recurrent angioedema, such as: hereditary angioedema with normal C1 inhibitor (formerly Type III HAE), acquired angioedema, idiopathic angioedema, or recurrent angioedema associated with urticaria and all other indications because its effectiveness has not been established
- ☐ **3.2** Medications for acute attacks will not be approved if being used in combination with another medication used for the treatment of acute attacks because this combination lacks strong evidence for use
- ☐ **3.3** Medications for prophylaxis will not be approved if being used in combination with another medication used for the prevention of attacks because this combination lacks strong evidence for use Drug FDA Approved Dose MDL Berinert (human C1 esterase inhibitor)  $\geq 5$  y/o