Hereditary Angioedema Acute Treatment: Berinert (C1 esterase inhibitor [human]), Ruconest (C1 esterase inhibitor [recombinant]) Firazyr (Icatibant, Sajazir) and Kalbitor (ecallantide)

Products for treatment of acute attacks for the treatment of HAE:

- **Berinert** (C1 esterase inhibitor [human])
- **Firazyr** (Icatibant, Sajazir)
- **Ruconest** (C1 esterase inhibitor [recombinant]) **Ruconest will not be covered for prophylaxis of HAE.**
- **Kalbitor** (ecallantide)

Coverage Criteria:

Berinert (C1 esterase inhibitor [human]), **Firazyr** (icatibant, Sajazir), **Ruconest** (C1 esterase inhibitor [recombinant]) or **Kalbitor** (ecallantide) are considered medically necessary for the treatment of acute attacks of hereditary angioedema when the following criteria are met:

1. Prescribed by a specialist such as a hematologist, allergist or immunologist; **and**,
2. Appropriate age of member (≥ 6 for Berinert, ≥12 for Kalbitor, ≥ 13 for Ruconest, ≥ 18 for Firazyr (icatibant, Sajazir); **and**,
3. Diagnosis of hereditary angioedema (HAE) with laboratory confirmation including one of the following:
    A. Type I defined as serum C4 < 14 mg/dL and C1 inhibitor (C1INH) < 19.9 mg/dL; **or**
    B. Type II defined as functional C1INH < 72%; **or**,
    C. A known HAE-causing C1INH mutation; **and**,
4. Patient must be experiencing or have a history of attacks with at least one symptom of a moderate or severe attack including one of the following:
    A. Airway swelling; **or**,
    B. Nausea and vomiting or severe abdominal pain; **or**,
    C. Facial swelling or painful distortion of the face; **and**,
5. Patients prescribed Ruconest are prescribed a regimen for treatment of acute attacks, not prophylaxis; and,
6. A hereditary angioedema management plan must be in place, provided to HealthPartners and use of this therapy is in accordance with that plan; **and**,
7. The drug is prescribed according to the FDA approved regimen.

Required Medical Information:

1. Lab documentation confirming HAE diagnosis (see criteria 3).
2. Weight documented in medical record (within past 30 days)

P&T Date: 5/6/19
Effective Date: 7/1/19

Coverage Duration:

Initial authorization and reauthorizations will be provided for 12 months

Renewal Criteria:

1. Patient has been seen by provider in the past 12 months and medical chart documentation that markers of disease are improved by therapy. These include but may not be limited to a reduction in the number and/or severity of attacks.