UnitedHealthcare Pharmacy
Clinical Pharmacy Programs

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| Program Number | 2024 P 2210-5 |
| Program | Prior Authorization/Medical Necessity |
| Medication | Ruconest® (C1 esterase inhibitor [recombinant]) |
| P&T Approval Date | 6/2020, 4/2021, 4/2022, 4/2023, 4/2024 |
| Effective Date | 7/1/2024 |

**1. Background:**
Ruconest (C1 esterase inhibitor [recombinant]) is indicated for the treatment of acute attacks in adult and adolescent patients with hereditary angioedema (HAE).

Effectiveness was not established in HAE patients with laryngeal attacks.[1]

**2. Coverage Criteria [a]:**

**A. Initial Authorization**

1. **Ruconest** will be approved based on **all** of the following criteria:

   a. Diagnosis of hereditary angioedema (HAE) as confirmed by **one** of the following:

      (1) C1 inhibitor (C1-INH) deficiency or dysfunction (Type I or II HAE) as documented by **one** of the following (per laboratory standard):

         (a) C1-INH antigenic level below the lower limit of normal
         (b) C1-INH functional level below the lower limit of normal

**-OR-**

      (2) HAE with normal C1 inhibitor levels and **one** of the following:

         (a) Confirmed presence of variant(s) in the gene(s) for factor XII, angiopoietin-1, plasminogen-1, kininogen-1, myoferlin, or heparan sulfate-glucosamine 3-O-sulfotransferase 6
         (b) Recurring angioedema attacks that are refractory to high-dose antihistamines with confirmed family history of angioedema
         (c) Recurring angioedema attacks that are refractory to high-dose antihistamines with unknown background de-novo mutation(s) (i.e., no family history) (HAE-unknown)

**-AND-**

   b. **Both** of the following:

      (1) Prescribed for the acute treatment of HAE attacks

**-AND-**

      (2) Not used in combination with other products indicated for the acute treatment of

HAE attacks (e.g., Berinert, Firazyr)

**-AND-**

c. Prescribed by **one** of the following:

(1) Immunologist
(2) Allergist

**Authorization of therapy will be issued for 12 months.**

**B. Reauthorization**

1. **Ruconest** will be approved based on **all** of the following criteria:

a. Documentation of positive clinical response to Ruconest therapy

**-AND-**

b. **Both** of the following:

(1) Prescribed for the acute treatment of HAE attacks

**-AND-**

(2) Not used in combination with other products indicated for the acute treatment of HAE attacks (e.g., Berinert, Firazyr)

**-AND-**

c. Prescribed by **one** of the following:

(1) Immunologist
(2) Allergist

**Authorization of therapy will be issued for 12 months.**

[a] State mandates may apply. Any federal regulatory requirements and the member specific benefit plan coverage may also impact coverage criteria. Other policies and utilization management programs may apply.

**3. Additional Clinical Programs:**

- Notwithstanding Coverage Criteria, UnitedHealthcare may approve initial and re-authorization based solely on previous claim/medication history, diagnosis codes (ICD-10) and/or claim logic. Use of automated approval and re-approval processes varies by program and/or therapeutic class.
- Supply limits may be in place.

**4. References:**

1. Ruconest [package insert]. Warren, NJ: Pharming Healthcare, Inc.; April 2020.

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 Jan 10.
3. Wu, E. Hereditary angioedema with normal C1 inhibitor. In: UpToDate, Saini, S (Ed), UpToDate, Waltham, MA, 2024.
4. Busse, P., Christiansen, S., Riedl, M., et. al. "US HAEA Medical Advisory Board 2020 Guidelines for the Management of Hereditary Angioedema." *The Journal of Allergy and Clinical Immunology.* 2020 September 05.
5. Maurer M, Magerl M, Betschel S, et al. The international WAO/EAACI guideline for the management of hereditary angioedema-The 2021 revision and update. Allergy. 2022;77(7):1961-1990. doi:10.1111/all.15214

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| **Change Control** | |
| 6/2020 | New program. |
| 4/2021 | Added diagnosis criteria. Updated references. |
| 4/2022 | Updated references. |
| 4/2023 | Annual review. Updated references. |
| 4/2024 | Annual review with update to examples of genetic variant(s) and diagnostic criteria with normal C1 inhibitor levels. Updated language for reauthorization criteria. |