Louisiana Medicaid
Hereditary Angioedema

The *Louisiana Uniform Prescription Drug Prior Authorization Form* should be utilized to request clinical authorization for medications used to treat or prevent hereditary angioedema (HAE).

Additional Point-of-Sale edits may apply.

*These agents may have **Black Box Warnings** and may be subject to **Risk Evaluation and Mitigation Strategy (REMS)** under FDA safety regulations. Please refer to individual prescribing information for details.*

HAE Medication Use and Minimum Age per Current Drug-Specific Prescribing Information			
Medication	**Brand**	**Use**	**Minimum Age**
Berotralstat	Orladeyo®	Prophylaxis	12
C1 Esterase Inhibitor (Human)	Berinert®	Treatment	5
C1 Esterase Inhibitor (Human)	Cinryze®	Prophylaxis	6
C1 Esterase Inhibitor Subcutaneous (Human)	Haegarda®	Prophylaxis	6
C1 Esterase Inhibitor (Recombinant)	Ruconest®	Treatment	13
Ecallantide	Kalbitor®	Treatment	12
Icatibant	Firazyr®	Treatment	18
Lanadelumab-flyo	Takhzyro™	Prophylaxis	12

Approval Criteria

- The recipient has a diagnosis of HAE; **AND**
- The recipient's age on the date of the request is not less than the minimum age recommended in the prescribing information (see table); **AND**
- The requested medication is used as recommended in the prescribing information for either prevention or treatment (see table); **AND**
- There is no preferred alternative that is the exact same chemical entity, formulation, strength, etc. **AND**
- If the request is for a non-preferred agent - **ONE** of the following is required:
    - The recipient has had a *treatment failure* with at least one preferred drug that is appropriate to use for the condition being treated; **OR**
    - The recipient has had an *intolerable side effect* to at least one preferred drug that is appropriate to use for the condition being treated; **OR**
    - The recipient has *documented contraindication(s)* to the preferred drugs that are appropriate to use for the condition being treated; **OR**
    - There is *no preferred product that is appropriate* to use for the condition being treated; **AND**

- By submitting the authorization request, the prescriber attests to the following:
    - The prescriber is knowledgeable about HAE, is experienced in managing patients with HAE, and is familiar with all HAE treatment options (e.g., allergist, dermatologist, hematologist, immunologist, rheumatologist); **AND**

- The recipient's individual triggers have been identified (where possible) and eliminated (if possible); the recipient has been counseled about HAE triggers and is making appropriate lifestyle changes; and, to the extent possible, lifestyle and activity restrictions are individualized and sensibly applied so that the recipient avoids HAE precipitating factors as primary prevention of HAE attacks; **AND**
- Any C1 esterase inhibitor used for prophylaxis will **NOT** be used in combination with any other C1 esterase inhibitor for prophylaxis (e.g., Cinryze® and Haegarda®); **AND**
- Medications used for treatment of acute attacks will **NOT** be used in combination with any other medication used for treatment of acute attacks (e.g., Berinert®, Firazyr®, Kalbitor® and Ruconest®); **AND**
- Evidence-based recommendations will be used to determine:
  - if long-term prophylaxis, short-term prophylaxis or on-demand treatment will be used for patient-specific triggers and procedures; **AND**
  - if intermittent long-term prophylaxis is appropriate; **AND**
  - the appropriate step-up, stabilize, step-down or intermittent approach to long-term prophylaxis as life events change over time (e.g., changes in stressors or hormonal fluxes); **AND**
- If the request is for Ruconest®, the recipient does not have known or suspected allergy to rabbits and rabbit derived products; **AND**
- If the request is for Ruconest®, the recipient will **NOT** use Ruconest® to treat laryngeal attacks; **AND**
- If the request is for Berinert® or Firazyr®, the recipient has been advised to seek immediate medical attention following treatment of laryngeal attacks; **AND**
- The dose does not exceed the maximum dose recommended in the prescribing information; **AND**
- Kalbitor® will be only be administered by a healthcare professional with appropriate medical support to manage anaphylaxis and HAE; **AND**
- The prescribing information for the requested medication has been thoroughly reviewed, including any Black Box Warning, Risk Evaluation and Mitigation Strategy (REMS), contraindications, warnings and precautions, minimum age requirements, recommended dosing, and prior treatment requirements; **AND**
- All laboratory testing and clinical monitoring recommended in the prescribing information have been completed as of the date of the request and will be repeated as recommended; **AND**
- The recipient has no concomitant drug therapies or disease states that limit the use of the requested medication and will not ~~be~~ receive ~~ing~~ the requested medication in combination with any medication that is contraindicated or not recommended per FDA labeling.

~~**Duration of initial approval: 12 months**~~

Reauthorization Criteria

- The recipient continues to meet initial approval criteria; **AND**
- The prescriber **states on the request** that the recipient is established on the medication with evidence of a positive response to therapy.

Duration of initial and reauthorization approval: 12 months

References

Berinert (C1 Esterase Inhibitor – Human) [package insert]. Kankakee, IL: CSL Behring LLC; April 2019. http://labeling.cslbehring.com/PI/US/Berinert/EN/Berinert-Prescribing-Information.pdf

Cinryze (C1 Esterase Inhibitor – Human) [package insert]. Lexington, MA: Shire ViroPharma Incorporated; January 2021~~une 2018~~. http://pi.shirecontent.com/PI/PDFs/Cinryze_USA_ENG.pdf

Farkas, H. et al. "International Consensus on The Diagnosis and Management of Pediatric Patients with Hereditary Angioedema with C1 Inhibitor Deficiency". Allergy, vol 72, no. 2, 2016, pp. 300-313. Wiley, doi:10.1111/all.13001. Accessed 8 May 2019.

Firazyr (icatibant) [package insert]. Lexington, MA: Shire Orphan Therapies LLC; August 2020. http://pi.shirecontent.com/PI/PDFs/Firazyr_USA_ENG.pdf

Haegarda (C1 Esterase Inhibitor Subcutaneous – Human) [package insert]. Kankakee, IL: CSL Behring LLC; September 2020. https://labeling.cslbehring.com/PI/US/HAEGARDA/EN/HAEGARDA-Prescribing-Information.pdf

Kalbitor (ecallantide) [package insert]. Lexington, MA: Dyax Corp.; December 2020~~March 2015~~. https://www.shirecontent.com/PI/PDFs/Kalbitor_USA_ENG.pdf

Maurer, M. et al. "The International WAO/EAACI Guideline For The Management Of Hereditary Angioedema-The 2017 Revision And Update". Allergy, vol 73, no. 8, 2018, pp. 1575-1596. Wiley, doi:10.1111/all.13384. Accessed 8 May 2019.

Orladeyo (berotralstat) [package insert]. Durham, NC: BioCryst Pharmaceuticals, Inc; December 2020. https://biocryst.com/wp-content/uploads/2020/12/ORLADEYO_PI_V1_2020.pdf

Ruconest (C1 Esterase Inhibitor – Recombinant) [package insert]. Bridgewater, NJ: Pharming Healthcare Inc.; April 2020. https://www.ruconest.com/wp-content/uploads/Ruconest_PI_Apr2020.pdf

Takhzyro (lanadelumab-flyo) [package insert]. Lexington, MA: Dyax Corp.; November 2018. https://www.shirecontent.com/PI/PDFs/TAKHZYRO_USA_ENG.pdf

Revision / Date	**~~Date~~Implementation Date**
Policy created	July 2019
Updated age requirement for Haegarda®, formatting changes, updated references / October 2020	~~October 2020~~April 2021

Added Orladeyo® with reference, added non-preferred wording -/ May 2021	~~January 2021~~ July 2021