Texas Prior Authorization Program
Clinical Criteria

Drug/Drug Class

Hereditary Angioedema (HAE)

Clinical Criteria Information Included in this Document

Agents for the treatment of HAE

- **Drugs requiring clinical prior authorization**: the list of drugs requiring prior authorization for this clinical criteria
- **Clinical prior authorization criteria logic**: a description of how the prior authorization request will be evaluated against the clinical criteria rules
- **Logic diagram**: a visual depiction of the clinical criteria logic
- **Supporting tables**: a collection of information associated with the steps within the criteria (diagnosis codes, procedure codes, and therapy codes)
- **References**: clinical publications and sources relevant to this clinical criteria

**Note**: Click the hyperlink to navigate directly to that section.

Revision Notes

- Annual review by staff
- Updated references, page 6
Hereditary Angioedema (HAE) Agents

Drugs Requiring Clinical Prior Authorization

The listed GCNs may not be an indication of TX Medicaid Formulary coverage. To learn the current formulary coverage, visit TxVendorDrug.com/formulary/formulary-search.

<table>
<thead>
<tr>
<th>Label Name</th>
<th>GCN</th>
</tr>
</thead>
<tbody>
<tr>
<td>BERINERT 500 UNIT KIT</td>
<td>31159</td>
</tr>
<tr>
<td>CINRYZE 500 UNIT VIAL</td>
<td>10495</td>
</tr>
<tr>
<td>FIRAZYR 30 MG/3 ML SYRINGE</td>
<td>14778</td>
</tr>
<tr>
<td>HAEGARDA 2,000 UNIT VIAL</td>
<td>37478</td>
</tr>
<tr>
<td>HAEGARDA 3,000 UNIT VIAL</td>
<td>43356</td>
</tr>
<tr>
<td>KALBITOR 10 MG/ML VIAL</td>
<td>28088</td>
</tr>
<tr>
<td>RUCONEST 2100 UNIT VIAL</td>
<td>30182</td>
</tr>
<tr>
<td>TAKHZYRO 300 MG/2 ML VIAL</td>
<td>45184</td>
</tr>
</tbody>
</table>
1. Is the client ≥ 5 years of age?
   - [ ] Yes – And request is for Berinert, go to #6
   - [ ] Yes – And request is for another agent, go to #2
   - [ ] No – Deny

2. Is the client ≥ 6 years of age?
   - [ ] Yes – And request is for Cinryze, go to #6
   - [ ] Yes – And request is for another agent, go to #3
   - [ ] No – Deny

3. Is the client ≥ 12 years of age?
   - [ ] Yes – And request is for Haegarda, Kalbitor or Takhzyro, go to #6
   - [ ] Yes – And request is for another agent, go to #4
   - [ ] No – Deny

4. Is the client ≥ 13 years of age?
   - [ ] Yes – And request is for Ruconest, go to #6
   - [ ] Yes – And request is for another agent, go to #5
   - [ ] No – Deny

5. Is the client ≥ 18 years of age?
   - [ ] Yes – And request is for Firazyr, go to #6
   - [ ] No – Deny

6. Does the client have 2 claims for the requested agent in the last 180 days?
   - [ ] Yes – Approve (365 days)
   - [ ] No – Go to #7

7. Does the client have a diagnosis of hereditary angioedema in the last 730 days?
   - [ ] Yes – Approve (365 days)
   - [ ] No – Deny
Hereditary Angioedema (HAE) Agents

Clinical Criteria Logic Diagram

- **Step 1** Is the client ≥ 5 years of age? [Yes → Step 2, No → Deny Request]
- **Step 2** Is the client ≥ 6 years of age? [Yes → Deny Request, No → Go to Step 3]
- **Step 3** Is the client ≥ 12 years of age? [Yes → Deny Request, No → Go to Step 4]
- **Step 4** Is the client ≥ 13 years of age? [Yes → Deny Request, No → Go to Step 5]
- **Step 5** Is the client ≥ 18 years of age? [Yes → Deny Request, No → Go to Step 6]
- **Step 6** Does the client have 2 claims for the requested agent in the last 180 days? [Yes → Go to Step 7, No → Deny Request]
- **Step 7** Does the client have a diagnosis of hereditary angioedema in the last 730 days? [Yes → Approve Request (365 days), No → Deny Request]

- Approve Request (365 days) [Yes → Approve Request, No → Deny Request]
<table>
<thead>
<tr>
<th>ICD-10 Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>D841</td>
<td>DEFECTS IN THE COMPLEMENT SYSTEM</td>
</tr>
</tbody>
</table>
Hereditary Angioedema (HAE) Agents

Clinical Criteria References


Publication History

The Publication History records the publication iterations and revisions to this document. Notes for the most current revision are also provided in the Revision Notes on the first page of this document.

<table>
<thead>
<tr>
<th>Publication Date</th>
<th>Notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>04/10/2014</td>
<td>Presented to the DUR Board</td>
</tr>
<tr>
<td>04/25/2014</td>
<td>Initial publication and posting to website</td>
</tr>
<tr>
<td>07/31/2015</td>
<td>Review of ICD-9 and ICD-10 codes</td>
</tr>
<tr>
<td>06/25/2018</td>
<td>Removed ICD-9 codes</td>
</tr>
<tr>
<td></td>
<td>GCNs for Haegarda added to ‘Drugs Requiring PA’, page 2</td>
</tr>
<tr>
<td></td>
<td>GCN for Firazyr removed from ‘Drugs Requiring PA’, page 2</td>
</tr>
<tr>
<td></td>
<td>Logic and logic diagram updated, pages 3-4</td>
</tr>
<tr>
<td></td>
<td>Updated references, page 7</td>
</tr>
<tr>
<td>03/29/2019</td>
<td>Updated to include formulary statement (The listed GCNS may not be an indication of TX Medicaid Formulary coverage. To learn the current formulary coverage, visit TxVendorDrug.com/formulary/formulary-search.) on each ‘Drug Requiring PA’ table</td>
</tr>
<tr>
<td>05/01/2019</td>
<td>Addition of Tahkzyro and agents for treatment of acute HAE attacks (Berinert, Firazyr, Kalbitor and Ruconest). Criteria approved by the DUR Board on April 26, 2019</td>
</tr>
<tr>
<td>07/15/2019</td>
<td>Annual review by staff</td>
</tr>
<tr>
<td></td>
<td>Updated references, page 6</td>
</tr>
</tbody>
</table>