Kuvan (sapropterin dihydrochloride)

**Override(s)**

<table>
<thead>
<tr>
<th>Override(s)</th>
<th><strong>Approval Duration</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Prior Authorization</td>
<td>Initial requests: 8 weeks</td>
</tr>
<tr>
<td></td>
<td>Continued therapy requests: 1 year</td>
</tr>
</tbody>
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**Medications**

- Kuvan (sapropterin dihydrochloride)

**APPROVAL CRITERIA**

Initial requests for Kuvan (sapropterin dihydrochloride) agents (tablet, oral packet) may be approved if the following criteria are met:

I. Individual has a diagnosis of hyperphenylalaninemia (HPA) due to tetrahydrobiopterin-(BH4-) responsive* phenylketonuria (PKU); **AND**

II. Individual is using in conjunction with a phenylalanine-(PHE-) restricted diet.

*BH4-responsiveness is known or will be determined by a trial of Kuvan

Requests for continued use of Kuvan (sapropterin dihydrochloride) agents (tablet, oral packet) may be approved if the following criteria are met:

I. Individual is using in conjunction with a PHE-restricted diet; **AND**

II. Individual is showing signs of continuing improvement, as evidenced by blood PHE level/dietary PHE allowance. If blood PHE levels do not decrease from baseline at a dose of 10 mg/kg/day administered for up to one month, the dose may be increased to 20 mg/kg/day. Individuals whose blood PHE does not decrease after 1 month of treatment at 20 mg/kg/day are considered non-responders and treatment should be discontinued.

Requests for Kuvan (sapropterin dihydrochloride) agents (tablet, oral packet) may not be approved for the following:

I. Individual is using in combination with Palynziq (pegvaliase-pqpz).
This policy does not apply to health plans or member categories that do not have pharmacy benefits, nor does it apply to Medicare. Note that market specific restrictions or transition-of-care benefit limitations may apply.

### Market Applicability

<table>
<thead>
<tr>
<th>Market</th>
<th>GA</th>
<th>KY</th>
<th>MD</th>
<th>NJ</th>
<th>NY</th>
<th>WA</th>
</tr>
</thead>
<tbody>
<tr>
<td>Applicable</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td>X</td>
</tr>
</tbody>
</table>

**Key References:**


Federal and state laws or requirements, contract language, and Plan utilization management programs or polices may take precedence over the application of this clinical criteria.

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