Revcovi (elapecagademase)
Effective June 19, 2019

<table>
<thead>
<tr>
<th>Plan</th>
<th>Program Type</th>
</tr>
</thead>
<tbody>
<tr>
<td>☒ MassHealth</td>
<td>☒ Prior Authorization</td>
</tr>
<tr>
<td>☒ Commercial/Exchange</td>
<td>☐ Quantity Limit</td>
</tr>
<tr>
<td>☒ Pharmacy Benefit</td>
<td>☐ Step Therapy</td>
</tr>
<tr>
<td>☒ Medical Benefit (NLX)</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Specialty Limitations</th>
<th>N/A</th>
</tr>
</thead>
<tbody>
<tr>
<td>Specialty Medications</td>
<td></td>
</tr>
<tr>
<td>All Plans</td>
<td>Phone: 866-814-5506</td>
</tr>
<tr>
<td>Non-Specialty Medications</td>
<td></td>
</tr>
<tr>
<td>MassHealth</td>
<td>Phone: 877-433-7643</td>
</tr>
<tr>
<td>Commercial</td>
<td>Phone: 800-294-5979</td>
</tr>
<tr>
<td>Exchange</td>
<td>Phone: 855-582-2022</td>
</tr>
<tr>
<td>Medical Specialty Medications (NLX)</td>
<td></td>
</tr>
<tr>
<td>All Plans</td>
<td>Phone: 844-345-2803</td>
</tr>
</tbody>
</table>

| Exceptions | N/A |

Overview
Elapegademase is an exogenous source of adenosine deaminase enzyme that reduces levels of toxic adenosine and deoxyadenosine and increases lymphocytes. Adenosine deaminase is an enzyme that catalyzes the deamination of both adenosine and deoxyadenosine. Hereditary lack of adenosine deaminase activity results in severe immunodeficiency disease. Elapegademase is FDA indicated for treatment of adenosine deaminase severe combined immune deficiency (ADA-SCID) in pediatric and adult patients.

Coverage Guidelines
Authorization may be granted for members who are currently receiving treatment with Revcovi, excluding when the product is obtained as samples or via manufacturer’s patient assistance programs OR
Authorization may be granted for treatment of ADA-SCID when the following criteria are met, and documentation has been provided:
1. Member has confirmed severe combined immunodeficiency disease (SCID) with a definitive diagnosis of adenosine deaminase deficiency as determined by one of the following:
   a. Deficient ADA catalytic activity (<1% of normal) in hemolysates (in untransfused patients) or in extracts of other cells (e.g., blood mononuclear cells, fibroblasts) OR
   b. Detection of mutations in the ADA gene by molecular genetic testing AND
2. Member has a marked elevations of the metabolite dATP or total dAdo nucleotides in erythrocytes AND
3. Member is not a candidate for or has failed a bone marrow transplant
4. Baseline values for plasma ADA activity red blood cell deoxyadenosine triphosphate (dATP), trough deoxyadenosine nucleotide (dAXP) and/or lymphocyte counts have been obtained.
Continuation of Therapy
Reauthorization may be granted when the following criteria/conditions have been met:
1. Member continues to meet initial criteria
2. Documentation of disease stability and/or improvement as evidenced by one or more of the following:
   - Increase in plasma ADA activity (target trough level $\geq 15$ mmol/hr/L)
   - Red blood cell dATP level decreased (target $\leq 0.005$ to $0.015$ mmol/L)
   - Improvement in immune function with decrease in frequency of infections
   - Improvement in red blood cell dAXP levels (target trough level $\leq 0.02$ mmol/L)

Limitations
Approvals will be granted for 12-month intervals

Dosing

| For Adagen-naïve patients | • Starting dose of Revcovi is $0.2$mg/kg twice a week IM for minimum of 12 to 24 weeks  
|                           | • Dose maybe gradually adjusted down to maintain trough ADA activity over $30$ mmol/hr/L  
|                           | • trough dAXP level under $0.02$ mmol/L and/or to maintain adequate immune function  
| For patients transitioning from Adagen to Revcovi | • If a patient’s weekly Adagen dose in unknown or is at or lower than $30$U/Kg, recommended starting of Revcovi is $0.2$mg/kg, IM once a week  
|                           | • If a patient’s weekly Adagen dose is above $30$ U/kg, the equivalent Revcovi dose should be calculated as follows:  
|                           | Revcovi dose in mg/kg =Adagen dose in U/kg ÷150  

References
4. Adenosine Deaminase Deficiency-genetic and Rare Diseases Information Center. US Department of health and human services-NIH. Available at: https://rarediseases.info.nih.gov/diseases/5748/adenosine-deaminase-deficiency

Review History
06/19/2019 – Reviewed
05/20/2020 – reviewed May P&T Mtg; no clinical updates

Disclaimer
AllWays Health Partners complies with applicable federal civil rights laws and does not discriminate or exclude people on the basis of race, color, national origin, age, disability, or sex.