

**Revcovi (elapegamase)  
Effective June 19, 2019**

<b>Plan</b>	<input checked="" type="checkbox"/> MassHealth <input type="checkbox"/> Commercial/Exchange	<b>Program Type</b>	<input checked="" type="checkbox"/> Prior Authorization
<b>Benefit</b>	<input checked="" type="checkbox"/> Pharmacy Benefit <input checked="" type="checkbox"/> Medical Benefit (NLX)		<input type="checkbox"/> Quantity Limit <input type="checkbox"/> Step Therapy
<b>Specialty Limitations</b>	N/A		
<b>Contact Information</b>	<b>Specialty Medications</b>		
	All Plans	Phone: 866-814-5506	Fax: 866-249-6155
	<b>Non-Specialty Medications</b>		
	MassHealth	Phone: 877-433-7643	Fax: 866-255-7569
	Commercial	Phone: 800-294-5979	Fax: 888-836-0730
	Exchange	Phone: 855-582-2022	Fax: 855-245-2134
	<b>Medical Specialty Medications (NLX)</b>		
	All Plans	Phone: 844-345-2803	Fax: 844-851-0882
<b>Exceptions</b>	N/A		

### Overview

#### FDA-Approved Indication

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 mono nuclear 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 then 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:
3. Increase in plasma ADA activity (target trough level  $\geq 15$  mmol/hr/L)
4. Red blood cell dATP level decreased (target  $\leq 0.005$  to  $0.015$  mmol/L)
5. Improvement in immune function with decrease in frequency of infections
6. Improvement in red blood cell dAXP levels (target trough level  $\leq 0.02$  mmol/L)

## Limitations

Approvals are granted for 12 months.

## Appendix

### Recommended Dosing

- For Adagen-naïve patients;
- Starting dose of Revcovi is 0.2mg/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 is unknown or is at or lower than 30U/Kg, recommended starting of Revcovi is 0.2mg/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  $\div$  150

## References

1. Revcovi [package insert]. Indianapolis, IN; Leadiant Biosciences; October 2018. Accessed January 2019
2. Hershfield, M. Adenosine Deaminase Deficiency. GeneReviews. [www.ncbi.nlm.nih.gov/books/NBK1483/](http://www.ncbi.nlm.nih.gov/books/NBK1483/) (Accessed on September 1, 2017).
3. Gaspar HB, Aiuti A, Porta F, et al. How I treat ADA deficiency. Blood. 2009 October 22; 114(17): 3524–3532.
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>
5. Flinn AM, Gennery AR. Adenosine deaminase deficiency: a review. Orphanet Journal of Rare Diseases 2018. <https://doi.org/10.1186/s13023-018-0807-5>

## Review History

06/19/19 – Reviewed

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