Mass General Brigham
Health Plan

Revcovi (elapegademase)
Effective 06/19/2019

Plan	☐ MassHealth UPPL ☒Commercial/Exchange	**Program Type**	☒ Prior Authorization ☐ Quantity Limit ☐ Step Therapy
Benefit	☒ Pharmacy Benefit ☒ Medical Benefit		
Specialty Limitations	N/A		
Contact Information	**Medical and Specialty Medications** All Plans | Phone: 877-519-1908 | Fax: 855-540-3693		
	Non-Specialty Medications All Plans | Phone: 800-711-4555 | Fax: 844-403-1029		
Exceptions	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 my 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 ≥ 15 mmol/hr/L)
4. Red blood cell dATP level decreased (target ≤ 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 ≤ 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 in 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 ÷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/ (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.