

Revcovi™ (elapegamase-ivlr) (Intramuscular)

Document Number: IC-0400

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Date of Origin: 10/30/2018

Dates Reviewed: 11/2018, 02/2019, 02/2020, 02/2021

I. Length of Authorization

Coverage will be provided for 12 months and may be renewed.

II. Dosing Limits

A. Quantity Limit (max daily dose) [NDC Unit]:

- Revcovi 2.4 mg/1.5 mL single-dose vial: 20 vials per 7 days

B. Max Units (per dose and over time) [HCPCS Unit]:

- 23 mg twice weekly

III. Initial Approval Criteria^{1,5}

Coverage is provided in the following conditions:

Universal Criteria

- Must not be used in combination with pegademase-bovine; **AND**
- Patient does not have severe thrombocytopenia (<50,000/microL); **AND**

Adenosine Deaminase (ADA) deficiency † Φ

- Patient has severe combined immunodeficiency disease (SCID) with a definitive diagnosis of adenosine deaminase deficiency as determined by one of the following:
 - Deficient ADA catalytic activity (<1% of normal) in hemolysates (in untransfused individuals) or in extracts of other cells (e.g., blood mononuclear cells, fibroblasts); **OR**
 - Detection of biallelic pathogenic mutations in the *ADA* gene by molecular genetic testing; **AND**
- Patient has a marked elevation of the metabolite deoxyadenosine triphosphate (dATP) or total deoxyadenosine nucleotides (dAXP) in erythrocytes; **AND**
- Patient is not a candidate for or has failed bone marrow transplantation (BMT); **AND**

- Baseline values for trough plasma ADA activity, red blood cell deoxyadenosine triphosphate (dATP), trough deoxyadenosine nucleotide (dAXP) and/or total lymphocyte counts have been obtained

† FDA Approved Indication(s); Ⓞ Orphan Drug

IV. Renewal Criteria^{1,5}

Coverage can be renewed based on the following criteria:

- Patient continues to meet universal and other indication-specific relevant criteria such as concomitant therapy requirements (not including prerequisite therapy), performance status, etc. identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: injection site bleeding in patients with thrombocytopenia, severe thrombocytopenia, etc.; **AND**
- Adequate documentation of disease stability and/or improvement as indicated by one or more of the following:
 - Increase in plasma ADA activity (target trough level ≥ 15 mmol/hr/L)
 - Red blood cell dATP level decreased (target ≤ 0.005 to 0.015 mmol/L)
 - Improvement in immune function with diminished frequency/complications of infection as evidenced in improvement in the ability to produce antibodies
 - Improvement in red blood cell dAXP levels (target trough level ≤ 0.02 mmol/L)

V. Dosage/Administration¹

Indication	Dose
Adenosine deaminase deficiency	<p><u>Patients transitioning from Adagen to Revcovi:</u></p> <ul style="list-style-type: none"> • If a patient’s weekly Adagen dose is unknown, or a patient’s weekly Adagen dose is at or lower than 30 U/kg, the recommended minimum starting dose of Revcovi is 0.2 mg/kg, intramuscularly, once a week • If a patient’s weekly Adagen dose is above 30 U/kg, an equivalent weekly Revcovi dose (mg/kg) should be calculated using the following conversion formula: $\text{Revcovi dose in mg/kg} = \text{Adagen dose in U/kg} \div 150$ • Subsequent doses may be increased by increments of 0.033 mg/kg weekly if trough ADA activity is under 30 mmol/hr/L, trough deoxyadenosine nucleotides (dAXP) are above 0.02 mmol/L, and/or the immune reconstitution is inadequate based on the clinical assessment of the patient. The total weekly dose may be divided into multiple intramuscular (IM) administrations during a week. <p><u>Adagen-naïve patients:</u></p> <ul style="list-style-type: none"> • The starting weekly dose of Revcovi is 0.4 mg/kg based on ideal body weight§ or actual weight (whichever is greater), divided into two doses (0.2 mg/kg twice a

	<p>week), intramuscularly, for a minimum of 12 to 24 weeks until immune reconstitution is achieved.</p> <ul style="list-style-type: none"> The dose may be 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 reconstitution based on clinical assessment of the patient.
<p>§The Devine formula for ideal body weight:</p> <ul style="list-style-type: none"> Ideal body weight (men) = 50 kg + 2.3 kg x (height, in - 60) Ideal body weight (women) = 45.5 kg + 2.3 kg x (height, in - 60) <i>Note: this formula is only an approximation, and is generally only applicable for people 60 inches (5 foot) tall or greater. For patients under 5 feet, one commonly-used modification is to subtract 2-5 lbs for each inch below 60 inches (Devine BJ. Gentamicin therapy. Drug Intell Clin Pharm. 1974;8:650-655.)</i> 	

VI. Billing Code/Availability Information

HCPCS Code:

- J3590 – Unclassified biologics
- C9399 – Unclassified drugs or biologicals (Hospital Outpatient Use ONLY)

NDC:

- Revcovi 2.4 mg/1.5 mL single-dose vial: 57665-0002-xx

VII. References

- Revcovi [package insert]. Indianapolis, IN; Lediant Biosciences; December 2020. Accessed January 2021.
- Hershfield, M. Adenosine Deaminase Deficiency. GeneReviews. www.ncbi.nlm.nih.gov/books/NBK1483/. Initial Posting: October 3, 2006; Last Update: March 16, 2017. Accessed January 2020.
- Gaspar HB, Aiuti A, Porta F, et al. How I treat ADA deficiency. Blood. 2009 October 22; 114(17): 3524-3532.
- 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>
- Flinn AM, Gennery AR. Adenosine deaminase deficiency: a review. Orphanet Journal of Rare Diseases 2018. <https://doi.org/10.1186/s13023-018-0807-5>

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
D81.31	Adenosine deaminase (ADA) deficiency with severe combined immunodeficiency

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD), Local Coverage Determinations (LCDs), and Local Coverage Articles (LCAs) may exist and compliance with these policies is required where applicable. They can be found at: <http://www.cms.gov/medicare-coverage-database/search/advanced-search.aspx>. Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD/LCA) – N/A

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	Applicable State/US Territory	Contractor
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto Government Benefit Administrators, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC