

Revcovi® (elapegademase-lvlr) (Intramuscular)

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I. Length of Authorization

- Initial: Prior authorization validity will be provided initially for 12 months (365 days).
- Renewal: Prior authorization validity may be renewed every 12 months (365 days) thereafter.

II. Dosing Limits

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

- 23 mg twice weekly

III. Initial Approval Criteria

Prior authorization validity is provided in the following conditions:

Universal Criteria ¹

- Member does not have severe thrombocytopenia (i.e., platelet count <50,000/microL); **AND**

Adenosine Deaminase Severe Combined Immunodeficiency (ADA-SCID) † Φ ^{1,2,5}

- Member has adenosine deaminase severe combined immunodeficiency (ADA-SCID) disease 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 (or likely pathogenic) variants in the *ADA* gene by molecular genetic testing; **AND**
- Member has elevated deoxyadenosine triphosphate (dATP) or total deoxyadenosine nucleotides (dAXP) in red blood cells; **AND**
 - Member is not a candidate for or has failed definitive therapy with bone marrow transplantation (BMT); **OR**

- Member is a candidate for definitive therapy with BMT and elapegeademase will be used as bridge therapy; **AND**
 - Member has baseline values for trough plasma ADA activity, red blood cell dATP, trough red blood cell dAXP, and/or total lymphocyte counts
- † FDA Approved Indication(s); ‡ Compendia Recommended Indication(s); Ⓢ Orphan Drug

IV. Renewal Criteria ^{1,2,5}

Prior authorization validity may be renewed based on the following criteria:

- Member continues to meet the 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 members with thrombocytopenia, severe thrombocytopenia, delay in improvement of immune function, etc.; **AND**
- Member has demonstrated a beneficial response to therapy compared to pretreatment baseline in one or more of the following:
 - Increase in plasma ADA activity (target trough level ≥ 15 mmol/hr/L)
 - Decrease in red blood cell dATP level (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
 - Decrease in red blood cell dAXP level (target trough level ≤ 0.02 mmol/L)
 - Increase in total lymphocyte counts

V. Dosage/Administration ¹

Indication	Dose
Adenosine Deaminase Severe Combined Immunodeficiency (ADA-SCID)	<u>Members transitioning from Adagen to Revcovi:</u> • Weekly Adagen dose is unknown or weekly Adagen dose ≤ 30 U/kg — The recommended minimum starting dose of Revcovi is 0.2 mg/kg, intramuscularly (IM), once a week • Weekly Adagen dose > 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 < 30 mmol/hr/L, trough deoxyadenosine nucleotides (dAXP) > 0.02 mmol/L, and/or

	the immune reconstitution is inadequate based on the clinical assessment of the member. The total weekly dose may be divided into multiple IM administrations during a week. <u>Adagen-naïve members:</u> • 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 week), intramuscularly, for a minimum of 12 to 24 weeks until immune reconstitution is achieved. • The dose may be gradually adjusted down to maintain trough ADA activity >30 mmol/hr/L, trough dAXP level <0.02 mmol/L, and/or to maintain adequate immune reconstitution based on clinical assessment of the member.
<u>§The Devine formula for ideal body weight:</u> • 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 members 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(s):

- J3590 – Unclassified biologics
- C9399 – Unclassified drugs or biologicals

NDC:

- Revcovi 2.4 mg/1.5 mL single-dose vial: 10122-0502-xx

VII. References

1. Revcovi [package insert]. Cary, NC; Chiesi USA, Inc.; August 2022. Accessed February 2026.
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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>

6. Dorsey MJ, Rubinstein A, Lehman H, et al. PEGylated Recombinant Adenosine Deaminase Maintains Detoxification and Lymphocyte Counts in Patients with ADA-SCID. *J Clin Immunol* 43, 951–964 (2023).
7. Kohn DB, Hershfield MS, Puck JM, et al. Consensus approach for the management of severe combined immune deficiency caused by adenosine deaminase deficiency. *J Allergy Clin Immunol*. 2019;143(3):852–63.
8. Grunebaum E, Booth C, Cuvelier GDE, et al. Updated Management Guidelines for Adenosine Deaminase Deficiency. *J Allergy Clin Immunol Pract*. 2023 Jun;11(6):1665-1675. doi: 10.1016/j.jaip.2023.01.032. Epub 2023 Feb 1. PMID: 36736952.

Appendix A – Non-Quantitative Treatment Limitations (NQTL) Factor Checklist

Non-quantitative treatment limitations (NQTLs) refer to the methods, guidelines, standards of evidence, or other conditions that can restrict how long or to what extent benefits are provided under a health plan. These may include things like utilization review or prior authorization. The utilization management NQTL applies comparably, and not more stringently, to mental health/substance use disorder (MH/SUD) Medical Benefit Prescription Drugs and medical/surgical (M/S) Medical Benefit Prescription Drugs. The table below lists the factors that were considered in designing and applying prior authorization to this drug/drug group, and a summary of the conclusions that Prime’s assessment led to for each.

Factor	Conclusion
Indication	Yes: Consider for PA
Safety and efficacy	No: PA not a priority
Potential for misuse/abuse	No: PA not a priority
Cost of drug	Yes: Consider for PA

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
D81.31	Severe combined immunodeficiency due to adenosine deaminase deficiency

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

The preceding information is intended for non-Medicare coverage determinations. 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 Determinations (NCDs) and/or Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. Local Coverage Articles (LCAs) may also exist for claims payment purposes or to clarify benefit eligibility under Part B for drugs which may be self-administered. The following link may be used to search for NCD, LCD,

or LCA documents: <https://www.cms.gov/medicare-coverage-database/search.aspx>. Additional indications, including any preceding information, may be applied 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 GBA
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA
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