Neighborhood Health Plan OF RHODE ISLAND™
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RevCovi™ (elapegademase-lvlr)
(Intramuscular)

Effective Date: 9/01/2019
Review Date: 8/23/2019, 1/29/20, 2/11/2021, 2/10/2022, 1/19/2023, 12/07/2023, 01/10/2024, 01/15/2025

Scope: Medicaid, Commercial, Medicare-Medicaid Plan (MMP)

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. Summary of Evidence

Clinical trials evaluating the efficacy and safety of RevCovi for the treatment of ADA-SCID have demonstrated significant improvements in immune function and overall survival in patients with this life-threatening condition. In a phase 3 clinical trial, RevCovi showed efficacy in restoring immune function, as evidenced by increases in lymphocyte counts and normalization of T-cell receptor excision circle (TREC) levels, a biomarker of thymic output. Patients treated with RevCovi demonstrated a significant reduction in serious infections and hospitalizations compared to historical controls, indicating improved immune function and reduced disease burden. Common adverse events reported being infusion-related reactions, fever, and rash.

IV. Initial Approval Criteria[1,5]

Coverage is provided in the following conditions:

MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements

Universal Criteria

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

Adenosine Deaminase Severe Combined Immunodeficiency (ADA-SCID) † Φ

- Patient 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 mutations in the *ADA* gene by molecular genetic testing; **AND**

- Patient has elevated deoxyadenosine triphosphate (dATP) or total deoxyadenosine nucleotides (dAXP) in red blood cells; **AND**
  - Patient is not a candidate for or has failed definitive therapy with bone marrow transplantation (BMT); **OR**
  - Patient is a candidate for definitive therapy with BMT and Revcovi (elapegademase) will be used as bridge therapy; **AND**
- Patient 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); **Φ** Orphan Drug

V. Renewal Criteria[1,5]

Coverage may 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, delay in improvement of immune function, etc.; **AND**
- Patient 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 levels (target trough level ≤ 0.02 mmol/L)
  - Increase in total lymphocyte counts

VI. Dosage/Administration[1]

Indication	Dose
Adenosine Deaminase Severe Combined Immunodeficiency (ADA-SCID)	Patients transitioning from Adagen to Revcovi: • 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: Revcovi dose in mg/kg = Adagen dose in U/kg ÷ 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. Adagen-naïve patients: • 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 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.
§The Devine formula for ideal body weight: •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) •*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.)*	

VII. 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

VIII. References

1. Revcovi [package insert]. Indianapolis, IN; Leadiant Biosciences; August 2022. Accessed January 2025.
2. 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.
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.

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)

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 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

Policy Rationale:

RevCovi was reviewed by the Neighborhood Health Plan of Rhode Island Pharmacy & Therapeutics (P&T) Committee. Neighborhood adopted the following clinical coverage criteria to ensure that its members use RevCovi according to Food and Drug Administration (FDA) approved labeling and/or relevant clinical literature. Neighborhood worked with network prescribers and pharmacists to draft these criteria. These criteria will help ensure its members are using this drug for a medically accepted indication, while minimizing the risk for adverse effects and ensuring more cost-effective options are used first, if applicable and appropriate. For INTEGRITY (Medicare-

Medicaid Plan) members, these coverage criteria will only apply in the absence of National Coverage Determination (NCD) or Local Coverage Determination (LCD) criteria. Neighborhood will give individual consideration to each request it reviews based on the information submitted by the prescriber and other information available to the plan.