Revcovi® (elapegademase-lvlr)
(Intramuscular)

Document Number: IC-0400

Last Review Date: 02/02/2023
Date of Origin: 10/30/2018
Dates Reviewed: 11/2018, 02/2019, 02/2020, 02/2021, 02/2022, 02/2023

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

Coverage is provided in the following conditions:

Universal Criteria [1]

- Will not be used in combination with pegademase-bovine; **AND**
- Patient does not have severe thrombocytopenia (i.e., platelet count <50,000/microL); **AND**

Adenosine Deaminase (ADA) Deficiency † Φ [1,2,5]

- 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 erythrocytes; **AND**
- Patient is not a candidate for or has failed bone marrow transplantation (BMT); **AND**

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- Patient has baseline values for trough plasma ADA activity, red blood cell dATP, trough dAXP and/or total lymphocyte counts

† FDA approved indication(s); ‡ Compendia recommended indication(s); Φ Orphan Drug

IV. Renewal Criteria [1,2,5]

Coverage may be renewed based on the following criteria:

- Patient 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 patients with thrombocytopenia, severe thrombocytopenia, delay in improvement of immune function, etc.; **AND**
- 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)
  - 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)

V. Dosage/Administration [1]

Indication	Dose
Adenosine Deaminase (ADA) Deficiency	<u>Patients 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: *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 <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 patient. The total weekly dose may be divided into multiple IM administrations during a week. <u>Adagen-naïve patients</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),

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	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 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.)*

VI. Billing Code/Availability Information

HCPCS Code(s):

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

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.; December 2020. Accessed January 2023.
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 2023.
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

Appendix 1 – Covered Diagnosis Codes

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

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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: https://www.cms.gov/medicare-coverage-database/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 GBA, 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

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- Qualified sign language interpreters
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Grievance Specialist
PreferredOne Community Health Plan
PO Box 59052
Minneapolis, MN 55459-0052
Phone: 1.800.940.5049 (TTY: 763.847.4013)
Fax: 763.847.4010
customerservice@preferredone.com

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You can also file a civil rights complaint with the U.S. Department of Health and Human Services, Office for Civil Rights, electronically through the Office for Civil Rights Complaint Portal, available at https://ocrportal.hhs.gov/ocr/portal/lobby.jsf, or by mail or phone at:

U.S. Department of Health and Human Services
200 Independence Avenue, SW
Room 509F, HHH Building
Washington, D.C. 20201
1-800-368-1019, 800-537-7697 (TDD)

Complaint forms are available at http://www.hhs.gov/ocr/office/file/index.html.

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PreferredOne Insurance Company
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Minneapolis, MN 55459-0212
Phone: 1.800.940.5049 (TTY: 763.847.4013)
Fax: 763.847.4010
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U.S. Department of Health and Human Services
200 Independence Avenue, SW
Room 509F, HHH Building
Washington, D.C. 20201
1-800-368-1019, 800-537-7697 (TDD)

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