

## Revcovi® (elapegademase-lvlr) (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, 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]:

- Renvovi 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 <sup>1</sup>

- 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 <sup>1,2,5</sup>

- 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**

- 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); Ⓢ Orphan Drug

#### IV. Renewal Criteria <sup>1,2,5</sup>

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  $\geq 15$  mmol/hr/L)
  - Decrease in red blood cell dATP level (target  $\leq 0.005$  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  $\leq 0.02$  mmol/L)

#### V. Dosage/Administration <sup>1</sup>

Indication	Dose
Adenosine Deaminase (ADA) Deficiency	<p><u>Patients transitioning from Adagen to Revcovi:</u></p> <ul style="list-style-type: none"> <li>• Weekly Adagen dose is unknown or weekly Adagen dose <math>\leq 30</math> U/kg               <ul style="list-style-type: none"> <li>– The recommended minimum starting dose of Revcovi is 0.2 mg/kg, intramuscularly (IM), once a week</li> </ul> </li> <li>• Weekly Adagen dose <math>&gt;30</math> U/kg               <ul style="list-style-type: none"> <li>– An equivalent weekly Revcovi dose (mg/kg) should be calculated using the following conversion formula:                   <math display="block">\text{Revcovi dose in mg/kg} = \text{Adagen dose in U/kg} \div 150</math> </li> </ul> </li> <li>• Subsequent doses may be increased by increments of 0.033 mg/kg weekly if trough ADA activity <math>&lt;30</math> mmol/hr/L, trough deoxyadenosine nucleotides (dAXP) <math>&gt;0.02</math> 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.</li> </ul> <p><u>Adagen-naïve patients:</u></p> <ul style="list-style-type: none"> <li>• The starting weekly dose of Revcovi is 0.4 mg/kg based on ideal body weight<sup>§</sup> or actual weight (whichever is greater), divided into two doses (0.2 mg/kg twice a week),</li> </ul>

	<p>intramuscularly, for a minimum of 12 to 24 weeks until immune reconstitution is achieved.</p> <ul style="list-style-type: none"> <li>The dose may be gradually adjusted down to maintain trough ADA activity &gt;30 mmol/hr/L, trough dAXP level &lt;0.02 mmol/L, and/or to maintain adequate immune reconstitution based on clinical assessment of the patient.</li> </ul>
<p><b>§The Devine formula for ideal body weight:</b></p> <ul style="list-style-type: none"> <li>Ideal body weight (men) = 50 kg + 2.3 kg x ( height, in - 60 )</li> <li>Ideal body weight (women) = 45.5 kg + 2.3 kg x ( height, in - 60 )</li> <li><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></li> </ul>	

## 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

- Revcovi [package insert]. Cary, NC; Chiesi USA, Inc.; December 2020. Accessed January 2023.
- Hershfield, M. Adenosine Deaminase Deficiency. GeneReviews. [www.ncbi.nlm.nih.gov/books/NBK1483/](http://www.ncbi.nlm.nih.gov/books/NBK1483/). Initial Posting: October 3, 2006; Last Update: March 16, 2017. Accessed January 2023.
- 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: <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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PCHP:

Provides free aids and services to people with disabilities to communicate effectively with us, such as:

- Qualified sign language interpreters
- Written information in other formats (large print, audio, accessible electronic formats, other formats)

Provides free language services to people whose primary language is not English, such as:

- Qualified interpreters
- Information written in other languages

If you need these services, contact a Grievance Specialist.

If you believe that PCHP has failed to provide these services or discriminated in another way on the basis of race, color, national origin, age, disability, or sex, you can file a grievance with:

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](mailto:customerservice@preferredone.com)

You can file a grievance in person or by mail, fax, or email. If you need help filing a grievance, a Grievance Specialist is available to help you.

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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ATTENTION: If you do not speak English, language assistance services, free of charge, are available to you. Call 1.800.940.5049 (TTY: 763.847.4013).

ATENCIÓN: si habla español, tiene a su disposición servicios gratuitos de asistencia lingüística. Llame al 1.800.940.5049 (TTY: 763.847.4013).

LUS CEEV: Yog tias koj hais lus Hmoob, cov kev pab txog lus, muaj kev pab dawb rau koj. Hu rau 1.800.940.5049 (TTY: 763.847.4013).

XIYYEEFFANNAA: Afaan dubbattu Oroomiffa, taiaajiila qarqaarsa afaanii, kanfaltiidhaan ala, ni argama. Bilbilaa 1.800.940.5049 (TTY: 763.847.4013).

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ဟ်သ့ဟ်သး- နမၤကတိၤ ကသီၤ ကျိၣ်အယိၤ, နမၤန့ၣ် ကျိၣ်အတၢ်မၤစၢၤလၢ တလၢၣ်ဘျဉ်လၢၣ်စၢၤ နီၣ်တမံၤဘၣ်သ့န့ၣ်လီၤ. ကိး 1.800.940.5049 (TTY: 763.847.4013).

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PreferredOne Insurance Company  
PO Box 59212  
Minneapolis, MN 55459-0212  
Phone: 1.800.940.5049 (TTY: 763.847.4013)  
Fax: 763.847.4010  
[customerservice@preferredone.com](mailto:customerservice@preferredone.com)

You can file a grievance in person or by mail, fax, or email. If you need help filing a grievance, a Grievance Specialist is available to help you.

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  
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