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**

- 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),

	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

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

Nondiscrimination & Language Access Policy

Aspirus Health Plan, Inc. complies with applicable Federal civil rights laws and does not discriminate on the basis of race, color, national origin, age, disability, sex, sexual orientation, or gender identity. *We* do not exclude people or treat them differently because of race, color, national origin, age, disability, sex, sexual orientation, or gender identity.

We will:
Provide 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)

Provide 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 *us* at the phone number shown on the inside cover of this *contract*, *your* id card, or aspirushealthplan.com.

If *you* believe that *we* have failed to provide these services or discriminated in another way on the basis of race, color, national origin, age, disability, sex, sexual orientation, or gender identity, *you* can file a grievance with:

Nondiscrimination Grievance Coordinator
Aspirus Health Plan, Inc.
PO Box 1062
Minneapolis, MN 55440
Phone: 1.866.631.5404 (TTY: 711)
Fax: 763.847.4010
Email: customerservice@aspirushealthplan.com

You can file a grievance in person or by mail, fax, or email. If *you* need help filing a grievance, the Nondiscrimination Grievance Coordinator 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.

Language Assistance Services

Albanian: KUJDES: Nëse flitni shqip, për ju ka në dispozicion shërbime të asistencës gjuhësore, pa pagesë.Telefononi në 1.866.631.5404 (TTY: 711).

Arabic تنبيه: إذا كنت تتحدث اللغة العربية، فإن خدمات المساعدة اللغوية متاحة لك مجاناً . اتصل بن اعلى رقم الهاتف 1.866.631.5404(رقم هاتف الصم والبك : 711)

French: ATTENTION : Si vous parlez français, des services d'aide linguistique vous sont proposés gratuitement. Appelezle 1.866.631.5404 (ATS : 711).

German: ACHTUNG: Wenn Sie Deutsch sprechen, stehen Ihnen kostenlos sprachliche Hilfsdienstleistungen zurVerfügung. Rufnummer: 1.866.631.5404 (TTY: 711).

Hindi: _यान द_: य_द आप ि◌हंदी बोलते ह_ तो आपके ि◌लए मु_त म_ भाषा सहायता सेवाएं उपल_ध ह_। 1.866.631.5404 (TTY: 711) पर कॉल कर_।

Hmong: LUS CEEV: Yog tias koj hais lus Hmoob, cov kev pab txog lus, muaj kev pab dawb rau koj. Hu rau 1.866.631.5404 (TTY: 711).

Korean: 주의: 한국어를 사용하시는 경우, 언어 지원 서비스를 무료로 이용하실 수 있습니다. 1.866.631.5404 (TTY: 711)번으로 전화해 주십시오.

Polish: UWAGA: Jeżeli mówisz po polsku, możesz skorzystać z bezpłatnej pomocy językowej. Zadzwoń pod numer 1.866.631.5404 (TTY: 711).

Russian: ВНИМАНИЕ: Если вы говорите на русском языке, то вам доступны бесплатные услуги перевода.Звоните 1.866.631.5404 (телетайп: 711).

Spanish: ATENCIÓN: si habla español, tiene a su disposición servicios gratuitos de asistencia lingüística. Llame al 1.866.631.5404 (TTY: 711).

Tagalog: PAUNAWA: Kung nagsasalita ka ng Tagalog, maaari kang gumamit ng mga serbisyo ng tulong sa wika nangwalang bayad. Tumawag sa 1.866.631.5404 (TTY: 711)

Traditional Chinese: 注意：如果您使用繁體中文，您可以免費獲得語言援助服務。請 致電 1.866.631.5404 (TTY:711).

Vietnamese: CHÚ Ý: Nếu bạn nói Tiếng Việt, có các dịch vụ hỗ trợ ngôn ngữ miễn phí dành cho bạn. Gọi số 1.866.631.5404 (TTY: 711).

Pennsylvania Dutch: Wann du Deitsch (Pennsylvania German / Dutch) schwetzscht, kannscht du mitaus Koschte ebbergricke, ass dihr helft mit die englisch Schprooch. Ruf selli Nummer uff: Call 1.866.631.5404 (TTY: 711).

Lao: ໂປດຊາບ: ຖ້າວ່າ ທ່ານເວົ້າພາສາ ລາວ, ການບໍລິການຊ່ວຍເຫຼືອດ້ານພາສາ,ໂດຍບໍ່ເສັ້ງຄ່າ, ແມ່ນມີພ້ອມໃຫ້ທ່ານ. ໂທຣ 1.866.631.5404 (TTY:711).