REVCOVI®
PRODUCT FACT SHEET

Revcovi®
elapegademase-lvlr
injection 1.6 mg/mL

INDICATION

Revcovi® (elapegademase-lvlr) is indicated for the treatment of adenosine deaminase severe combined immune deficiency (ADA-SCID) in pediatric and adult patients.

IMPORTANT SAFETY INFORMATION

WARNINGS AND PRECAUTIONS

- Injection site bleeding in patients with thrombocytopenia: Increased risk of local bleeding in patients with thrombocytopenia; should not be used if thrombocytopenia is severe.
- Delay in improvement of immune function: Protect immune deficient patients from infections until improvement in immune function.

ADVERSE REACTIONS

The most commonly reported adverse reactions were cough (50%) and vomiting (33%).

In addition, the following post-marketing reports for the same class of enzyme replacement therapy used in the treatment of ADA-SCID may also be seen with Revcovi treatment:

- Hematologic events: hemolytic anemia, autoimmune hemolytic anemia, thrombocythemia, thrombocytopenia and autoimmune thrombocytopenia
- Dermatological events: injection site erythema, urticaria
- Lymphomas

IMPORTANT MONITORING INFORMATION

Treatment with Revcovi should be monitored by measuring trough plasma ADA activity and trough dAXP levels for maintenance of therapeutic targets. If a persistent decline in plasma ADA activity occurs, immune function and clinical status should be monitored closely, and precautions should be taken to minimize the risk of infection.

Please see Important Safety Information throughout and accompanying full Prescribing Information.

SUPPORT for LIFE

ADA-SCID: ULTRA-RARE, GENETIC—AND OFTEN FATAL IF LEFT UNTREATED[1-3]

Adenosine deaminase severe combined immune deficiency (ADA-SCID) is caused by a deficiency in the ADA enzyme.[1] ADA is produced in all cells, and it is most active in lymphocytes, a type of white blood cell that is a key part of the body's immune system.[1] A defective ADA gene reduces or eliminates the amount of enzyme activity, increasing the risk of severe and recurring infection.[1-3]

ADA-SCID primarily affects infants and young children, and it is typically diagnosed within the first few months of life.[1] ADA-SCID is estimated to occur in approximately one in 200,000 to one in 1,000,000 newborns around the world.[3]

> Without early diagnosis and effective treatment, babies with ADA-SCID usually die from infections before they reach 2 years of age.[3]

REVCOVI ADMINISTRATION AND DOSING

Revcovi is for intramuscular injection only.[4] Dose titration along with careful lab monitoring help customize Revcovi treatment to each patient's individual needs.[4]

Recommended starting dose[4]

0.4 mg/kg/wk based on ideal body weight or actual weight, whichever is greater

Divided into 2 weekly doses for a minimum of 12-24 weeks

Until immune reconstitution is achieved

- Once immune reconstitution is achieved, dose may be gradually adjusted down to maintain trough ADA activity >30 mmol/hr/L, trough dAXP levels <0.02 mmol/L, and/or to maintain adequate immune reconstitution based on the clinical assessment of the patient[4]
- Improvement in the general clinical status of the patient may be gradual but should be apparent by the end of the first year of therapy[4]

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THE EFFICACY AND SAFETY OF REVCOVI WAS PROVEN IN 2 CLINICAL STUDIES[4]

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Two prospective, open-label, single-arm, multicenter studies evaluated the efficacy, safety, tolerability, and pharmacokinetics of Revcovi in patients with ADA-SCID.[4]

Study 1: United States study

Study 1 evaluated Revcovi in 6 US patients (aged 8-37 years) with ADA-SCID who were receiving therapy with pegademase bovine, a legacy enzyme replacement therapy (ERT) that is no longer available for use.[4]

Patients had positive results in reduction of dAXP concentrations, stable trough plasma ADA activity, and total lymphocyte count maintenance or improvement.[4]

Study 2: Japanese study

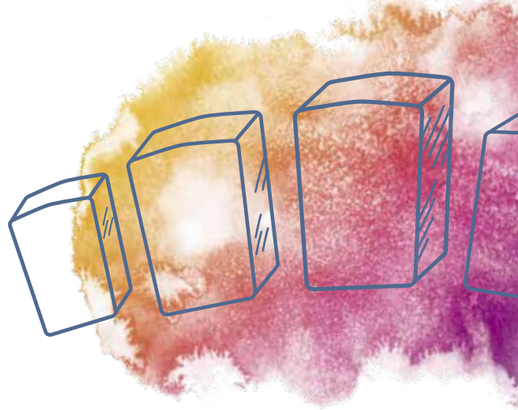
Study 2 evaluated Revcovi in 4 patients (aged 3.4 months to 25 years) with ADA-SCID.[4]

Patients had similar improvements to Study 1 in trough dAXP concentration, trough plasma ADA activity, and total lymphocyte counts.[4]

MECHANISM OF ACTION

Revcovi provides an exogenous source of ADA enzyme, which is deficient in patients with ADA-SCID.[4]

Revcovi treatment is associated with a decrease in toxic adenosine and deoxyadenosine nucleotides levels as well as an increase in lymphocyte number.[4]



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REVCOVI ORDERING & SUPPORT

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Ordering information

Healthcare providers, please visit **ChiesiTotalCare.com** to access the Revcovi Order/Prescription Form and the Chiesi Total Care℠ Enrollment Form.

Please fax completed form to Chiesi Total Care staff at **1-866-272-7079**.

Please call **1-866-272-7078** if you have questions regarding this form or to contact Chiesi Total Care.

One-stop patient support

Chiesi Total Care is a comprehensive support program that provides exceptional service for your patients. A single call to your dedicated Chiesi Total Care Team is all it takes to begin the process of getting a patient started on Revcovi. Chiesi Total Care has the ability to provide assistance to patients with or without commercial insurance, Medicaid, or Medicare. The program also offers injection support for patients who receive Revcovi injections at home.*

Some patients may be eligible for the Revcovi Copay Program, under which they may pay as little as $0 for their prescription.*

Program eligibility

- Patient must be enrolled in Chiesi Total Care. Enrollment form can be mailed to your patient's home.
- Patient has commercial insurance and a valid prescription for a US Food and Drug Administration (FDA)–approved indication for Revcovi.
- Patient must be a resident of the United States or one of its territories.

*Please refer to accompanying full Terms and Conditions for additional eligibility requirements.

Medical information and adverse event reporting

For adverse event reporting, product complaints, or medical information inquiries:

PHONE: **1-888-661-9260**
FAX: **1-866-443-3092**
EMAIL: **us.medical@chiesi.com**

Privacy policy

Where applicable, personal information shared and processed for adverse events reporting, product orders, reimbursement support, and patient assistance is subject to Chiesi's Privacy Policy available at https://www.chiesiusa.com/privacy-policy/.

References

1. Whitmore KV, Gaspar HB. Adenosine deaminase deficiency – more than just an immunodeficiency. *Front Immunol.* 2016;7:314. doi:10.3389/fimmu.2016.00314. **2.** Sauer AV, Brigida I, Carriglio N, Aiuti A. Autoimmune dysregulation and purine metabolism in adenosine deaminase deficiency. *Front Immunol.* 2012;3:265. doi:10.3389/fimmu.2012.00265. **3.** Hershfield M. Adenosine deaminase deficiency. In: Adam MP, Ardinger HH, Pagon RA, et al, eds. *GeneReviews®*. University of Washington, Seattle; 1993-2020. Posted October 3, 2006. Updated March 16, 2017. https://www.ncbi.nlm.nih.gov/books/NBK1483 **4.** Revcovi. Prescribing information. Chiesi USA, Inc.; 2021.

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