

## PHARMACY POLICY STATEMENT

### Georgia Medicaid

<b>DRUG NAME</b>	<b>Revcovi (elapegademase-ivlr)</b>
<b>BENEFIT TYPE</b>	Pharmacy
<b>STATUS</b>	Prior Authorization Required

Revcovi, approved by the FDA in 2018, is a recombinant adenosine deaminase (rADA) indicated for the treatment of adenosine deaminase severe combined immune deficiency (ADA-SCID) in pediatric and adult patients. It acts as an enzyme replacement therapy (ERT).

SCID associated with a deficiency of ADA enzyme is a very rare, inherited, and often fatal purine salvage pathway defect. ADA deficiency is a primary immunodeficiency caused by a genetic mutation that affects white blood cell (WBC) production. ADA-SCID is the most complete form of ADA deficiency and typically leads to a severe combined immunodeficiency with dysfunction of T, B, and natural killer cells (T-B-NK-SCID), presenting in the first few months of life. Non-immune symptoms also occur.

Revcovi provides an exogenous source of ADA enzyme that is associated with a decrease in toxic adenosine and deoxyadenosine nucleotide levels as well as an increase in lymphocyte number. It is a second-generation ADA replacement therapy, taking the place of the discontinued first-generation ADA replacement product, Adagen. Most newly diagnosed patients should initially receive ERT. Ideally, it is used as a bridge prior to allogeneic hematopoietic stem cell transplantation (HSCT) when that is an option.

Treatment with Revcov provides extensive monitoring. Immune function, including the ability to produce antibodies, generally improves after 2 - 6 months of therapy, and matures over a longer period. Improvement in the general clinical status of the patient may be gradual but should be apparent by the end of the first year.

Revcovi (elapegademase-ivlr) will be considered for coverage when the following criteria are met:

#### **Adenosine Deaminase Severe Combined Immune Deficiency (ADA-SCID)**

For initial authorization:

1. Medication must be prescribed by or in consultation with an immunologist, geneticist, or hematologist/oncologist; AND
2. Member has a diagnosis of ADA-SCID confirmed by at least one of the following:
  - a) Absent or very low ADA activity (<1%) in red blood cells or in extracts of dried blood spots
  - b) Molecular genetic testing showing bi-allelic pathogenic variants in the ADA gene; AND
3. Baseline lab values for monitoring of all the following parameters must be provided:
  - a) ADA activity level
  - b) Erythrocyte dAXP levels
  - c) Total lymphocyte counts; AND
4. Member meets one of the following:
  - a) Revcov is being used as a bridge prior to undergoing HSCT
  - b) Member is not a candidate for HSCT or does not have a matched donor available
  - c) Member has failed HSCT; AND
5. Member does not have severe thrombocytopenia (platelet count <50 x 10<sup>9</sup>/L).
6. **Dosage allowed/Quantity limit:** The starting weekly dose is 0.4 mg/kg intramuscularly, divided into two doses (0.2 mg/kg twice a week), for a minimum of 12 to 24 weeks until immune reconstitution is achieved.

Then, 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.

Note: It is generally not advised to continue ERT for more than a few years.

***If all the above requirements are met, the medication will be approved for 6 months.***

For reauthorization:

1. Chart notes must show at least one of the following:
  - a) Trough plasma ADA activity level of at least 30 mmol/hr/L
  - b) Trough erythrocyte dAXP levels below 0.02 mmol/L
  - c) Improved lymphocyte counts
  - d) Improved clinical immune status (i.e., fewer/less severe infections compared to baseline); AND
2. Member meets one of the following:
  - a) Revcovi is being used as a bridge prior to undergoing HSCT
  - b) Member is not a candidate for HSCT or does not have a suitable donor
  - c) Member has failed HSCT.

***If all the above requirements are met, the medication will be approved for an additional 12 months.***

**CareSource considers Revcovi (elapegademase-lvlr) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.**

DATE	ACTION/DESCRIPTION
11/23/2022	New policy for Revcovi created.
01/08/2025	Updated references. Removed note about Adagen. Added note about duration of use. Minor wording change in diagnostic requirement (Hershfield 2024).

#### References:

1. Revcovi [prescribing information]. Chiesi USA, Inc.; 2020.
2. Dorsey MJ, Rubinstein A, Lehman H, Fausnight T, Wiley JM, Haddad E. PEGylated Recombinant Adenosine Deaminase Maintains Detoxification and Lymphocyte Counts in Patients with ADA-SCID [published correction appears in *J Clin Immunol*. 2023 Oct;43(7):1671-1672. doi: 10.1007/s10875-023-01531-6]. *J Clin Immunol*. 2023;43(5):951-964. doi:10.1007/s10875-022-01426-y
3. 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):852-863. doi:10.1016/j.jaci.2018.08.024
4. Grunebaum E, Booth C, Cuvelier GDE, Loves R, Aiuti A, Kohn DB. Updated Management Guidelines for Adenosine Deaminase Deficiency. *J Allergy Clin Immunol Pract*. 2023;11(6):1665-1675. doi:10.1016/j.jaip.2023.01.032
5. Hershfield M, Tarrant T. Adenosine Deaminase Deficiency. 2006 Oct 3 [Updated 2024 Mar 7]. In: Adam MP, Feldman J, Mirzaa GM, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2024. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK1483/>

Effective date: 02/01/2026

Revised date: 01/08/2025