Reference number(s)
6742-A

Specialty Guideline Management
Revcovi

Products Referenced by this Document

Drugs that are listed in the following table include both brand and generic and all dosage forms and strengths unless otherwise stated. Over the counter (OTC) products are not included unless otherwise stated.

Brand Name	Generic Name
Revcovi	elapegademase-lvlr

Indications

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-approved Indications[1]

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

All other indications are considered experimental/investigational and not medically necessary.

Documentation

Submission of the following information is necessary to initiate the prior authorization review:

Adenosine Deaminase Severe Combined Immune Deficiency[1-3]

Initial requests:

- Genetic or molecular test results or medical records confirming the diagnosis.
- Baseline values for plasma adenosine deaminase (ADA) activity, red blood cell deoxyadenosine triphosphate (dATP), trough deoxyadenosine nucleotide (dAXP) levels, and/or total lymphocyte counts.
- Hematologic assessment (e.g., complete blood count) demonstrating absence of severe thrombocytopenia (platelets <50,000/microL).

Continuation requests:

Chart notes, lab values, or medical record documentation supporting positive clinical response.

Prescriber Specialties

This medication must be prescribed by an or in consultation with an immunologist or a physician who specializes in the treatment of metabolic disease and/or lysosomal storage disorders.

Coverage Criteria

Adenosine Deaminase Severe Combined Immune Deficiency [1-3]

Authorization of 12 months may be granted for treatment of ADA-SCID when all of the following criteria are met:

- The diagnosis is confirmed by one of the following:
    - Increased red blood cell deoxyadenosine triphosphate (dATP) or trough deoxyadenosine nucleotide (dAXP) concentrations and ONE of the following:
        - Absent or very low (<1% of normal) adenosine deaminase (ADA) activity in red blood cells
        - Genetically confirmed, biallelic variant in the ADA gene.
- Baseline values for plasma ADA activity, red blood cell dATP, dAXP levels, and/or total lymphocyte counts have been obtained.
- Member meets one of the following:
    - The requested medication will only be used until definitive therapy with hematopoietic stem cell transplantation (HSCT) .
    - Member is not a suitable candidate for HSCT (e.g., matched sibling or family donor not available).
    - Member has failed HSCT.

- Member does not have severe thrombocytopenia (platelets <50,000/microL).
- Member does not have autoimmune disease requiring immunosuppressive therapy.
- Member will be monitored for evidence of treatment efficacy per protocol outlined in the prescribing information during treatment with Revcovi.

Continuation of Therapy

Authorization of 12 months may be granted for continued treatment in members requesting reauthorization for an indication listed in the coverage criteria section when all of the following criteria are met:

- Member meets the criteria for initial approval.
- Member does not have unacceptable toxicity (e.g., severe injection site reactions/bleeding, severe thrombocytopenia).
- Member is experiencing benefit from therapy (e.g., maintenance of target trough plasma ADA activity ≥30 mmol/L, trough erythrocyte dAXP levels below 0.02 mmol/L, improved or stabilized total lymphocyte counts and/or immune function).

References

1. Revcovi [package insert]. Cary, NC: Chiesi USA, Inc.; December 2020.
2. 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
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