Reference number(s)
6743-A

Standard Medicare Part B 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 will be assessed on an individual basis. Submissions for indications other than those listed in this criteria should be accompanied by supporting evidence from Medicare approved compendia.

Documentation

The following documentation must be available, upon request, for all submissions:

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Adenosine Deaminase Severe Combined Immune Deficiency (ADA-SCID)[1-3]

Initial requests:

Genetic or molecular test results or medical records confirming the diagnosis.

Continuation requests:

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

Coverage Criteria

Adenosine Deaminase Severe Combined Immune Deficiency (ADA-SCID) [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 any of the following:
  - Absent or very low (<1% of normal) adenosine deaminase (ADA) activity in red blood cells.
  - Increased erythrocyte deoxyadenosine triphosphate (dATP) or deoxyadenosine nucleotides (dAXP) concentrations.
  - Genetically confirmed, biallelic variant in the ADA gene.
- 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 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.

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- Member is experiencing benefit from therapy (e.g., maintenance of target trough plasma adenosine deaminase (ADA) activity ≥30 mmol/L, trough erythrocyte (dAXP) levels below 0.02 mmol/L, improved or stabilized total and subset lymphocyte counts and/or immune function).

Summary of Evidence

The contents of this policy were created after examining the following resources:

- The prescribing information for Revcovi
- The available compendium
  - National Comprehensive Cancer Network (NCCN) Drugs and Biologics Compendium
  - Micromedex DrugDex
  - American Hospital Formulary Service- Drug Information (AHFS-DI)
  - Lexi-Drugs
  - Clinical Pharmacology
- Updated Management Guidelines for Adenosine Deaminase Deficiency.
- Consensus Approach for the Management of Severe Combined Immune Deficiency Caused by Adenosine Deaminase Deficiency.

After reviewing the information in the above resources, the FDA-approved indications listed in the prescribing information for Revcovi are covered.

Explanation of Rationale

Support for FDA-approved indications can be found in the manufacturer's prescribing information.

Support for the diagnostic criteria for adenosine deaminase severe combined immune deficiency can be found in consensus guidelines by Kohn, et al. Diagnosis of ADA deficiency is usually established by demonstrating absent or very low (<1% of normal) ADA activity in red blood cells (RBC). An elevated level of dATP (also measured as total dAdo nucleotides, dAXP) in RBC is pathognomonic for ADA deficiency. Demonstrating bi-allelic pathogenic variants in the ADA gene should also be done to further confirm ADA deficiency.

Support for diagnostic criteria can also be found in ADA-SCID Management Guidelines by Grunebaum, et al. Absent or markedly reduced ADA enzyme activity in erythrocytes with increased concentrations in erythrocytes and plasma of Ado, dAdo, dAdo-triphosphate, or dAXP (the sum of dAdo monophosphate, diphosphate, and triphosphate) establish the diagnosis of ADA deficiency.

Support for the use of Revcovi only until definitive therapy with hematopoietic stem cell transplantation (HSCT), when the member is not a suitable candidate for HSCT, or when the member has failed HSCT can be found in consensus guidelines by Kohn, et al. All patients should initially receive enzyme replacement therapy (ERT) followed by definitive treatment with either of two equal first line options (HSCT,

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hematopoietic stem cell gene therapy (HSC-GT)). If HLA matched sibling donor or matched family donor HSCT or HSC-GT are not available or have failed, ERT can be continued or re-instituted.

Support for the use of Revcovi only as bridge therapy in most patients can also be found in ADA-SCID Management Guidelines by Grunebaum, et al. In most ADA-deficient patients, ERT should be used to "bridge" patients until they can receive HCT or gene therapy. ERT should be continued for no more than a few years. This is because decline in T-cell numbers and function is often observed following several years of ERT.

References

1. Revcovi [package insert]. Cary, NC: Chiesi USA, Inc.; August 2022.
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

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