Clinical Policy: Elapegademase-lvlr (Revcovi)

Reference Number: LA.PHAR.419
Effective Date: 11.04.23
Last Review Date: 05.08.25
Line of Business: Medicaid

Revision Log

See Important Reminder at the end of this policy for important regulatory and legal information.

****Please note: This policy is for medical benefit****

Description

Elapegademase-lvlr (Revcovi®) is a recombinant adenosine deaminase.

FDA Approved Indication(s)

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

Policy/Criteria

Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.

It is the policy of Louisiana Healthcare Connections that Revcovi is **medically necessary** when the following criteria are met:

I. Initial Approval Criteria

A. Adenosine Deaminase Severe Combined Immune Deficiency Disease (must meet all)**:**

1. Diagnosis of ADA-SCID confirmed by genetic testing;
2. Prescribed by or in consultation with an immunologist or hematologist;
3. Member has failed bone marrow transplantation or is not a candidate for bone marrow transplantation;
4. Dose does not exceed 0.4 mg/kg per week.

Approval duration: 6 months

B. Other diagnoses/indications (must meet 1 or 2)**:**

1. If this drug has recently (within the last 6 months) undergone a label change (e.g., newly approved indication, age expansion, new dosing regimen) that is not yet reflected in this policy, refer to LA.PMN.255

2. If the requested use (e.g., diagnosis, age, dosing regimen) is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized) AND criterion 1 above does not apply, refer to the off-label use policy LA.PMN.53

II. Continued Therapy

A. Adenosine Deaminase Severe Combined Immunodeficiency Disease (must meet all)**:**

1. Currently receiving medication via Louisiana Healthcare Connections benefit or member has previously met initial approval criteria;

2. Member is responding positively to therapy *(see Appendix D for examples)*;
3. If request is for a dose increase, new dose does not exceed 0.4 mg/kg per week.

Approval duration: 12 months

B. Other diagnoses/indications (must meet 1 or 2)**:**

1. If this drug has recently (within the last 6 months) undergone a label change (e.g., newly approved indication, age expansion, new dosing regimen) that is not yet reflected in this policy, refer to LA.PMN.255
2. If the requested use (e.g., diagnosis, age, dosing regimen) is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized) AND criterion 1 above does not apply, refer to the off-label use policy LA.PMN.53

III. Diagnoses/Indications for which coverage is NOT authorized:

A. Non-FDA approved indications, which are not addressed in this policy, unless there is sufficient documentation of efficacy and safety according to the off label use policy – LA.PMN.53.

IV. Appendices/General Information

Appendix A: Abbreviation/Acronym Key
ADA-SCID: adenosine deaminase severe combined immune deficiency disease
dAXP: deoxyadenosine nucleotides
FDA: Food and Drug Administration

Appendix B: Therapeutic Alternatives
Not applicable

Appendix C: Contraindications/Boxed Warnings
None reported

Appendix D: General Information

- Examples of positive response to therapy include improvement in immune function (T cell, B cell, and natural killer lymphocytes), reduction in frequency/severity of opportunistic infections, and decrease from baseline or maintenance of normal red cell dATP levels.
- Once treatment with Revcovi has been initiated, a target trough plasma ADA activity should be at least 30 mmol/hr/L. In order to determine an effective dose of Revcovi, trough plasma ADA activity (pre-injection) should be determined every 2 weeks for Adagen-naïve patients and every 4 weeks for patients previously receiving Adagen therapy, during the first 8 - 12 weeks of treatment, and every 3 - 6 months thereafter. A decrease of ADA activity below this level suggests noncompliance to treatment or a development of antibodies (anti-drug, anti-PEG, and neutralizing antibodies). Antibodies to Revcovi should be suspected if a persistent fall in pre-injection levels of trough plasma ADA activity below 15 mmol/hr/L occurs. In such patients, testing for antibodies to Revcovi should be performed. If a persistent decline in trough plasma ADA activity occurs, immune function and clinical status should be monitored closely and precautions should be taken to minimize the risk of infection. If antibodies to Revcovi are found to be the cause of a persistent fall in trough plasma ADA activity, then adjustment in the dosage of Revcovi and other measures may be taken to induce tolerance and restore adequate ADA activity.
- Two months after starting Revcovi treatment, trough erythrocyte deoxyadenosine nucleotide (dAXP) levels should be maintained below 0.02 mmol/L, and monitored at least twice a year.
- The degree of immune function may vary from patient to patient. Each patient will require appropriate monitoring consistent with immunologic status. Total and subset lymphocytes should be monitored periodically as follows:
  - Adagen-naïve patients: every 4 - 8 weeks for up to 1 year, and every 3 - 6 months thereafter
  - Other patients: every 3 - 6 months
- Immune function, including the ability to produce antibodies, generally improves after 2 - 6 months of therapy, and matures over a longer period. In general, there is a lag between the correction of the metabolic abnormalities and improved immune function. Improvement in the general clinical status of the patient may be gradual (as evidenced by

improvement in various clinical parameters) but should be apparent by the end of the first year of therapy.

V. Dosage and Administration

Indication	Dosing Regimen	Maximum Dose
ADA-SCID	Patients transitioning from Adagen® to Revcovi: If the weekly Adagen dose is unknown, or if the weekly Adagen dose is at or lower than 30 U/kg, use Revcovi 0.2 mg/kg IM weekly. If the weekly Adagen dose is > 30 U/kg, an equivalent weekly Revcovi dose (mg/kg) should be calculated by dividing the Adagen dose in U/kg by 150. Subsequent doses may be increased by increments of 0.033 mg/kg weekly if trough ADA activity is under 30 mmol/hr/L, trough dAXPs are above 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. Adagen-naïve patients: 0.2 mg/kg IM twice a week based on ideal body weight or actual weight whichever is greater for at least 12-24 weeks until immune reconstitution is achieved. 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 of the patient.	0.4 mg/kg/week

VI. Product Availability

Single-dose vial: 2.4 mg/1.5 mL (1.6 mg/mL)

VII. References

1. Revcovi Prescribing Information. Gaithersburg, MD: Leadiant Biosciences Inc.; December 2020. Available at: www.revcovi.com. Accessed January 11, 2024.
2. 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;143:852-63.

Reviews, Revisions, and Approvals	Date	LDH Approval Date
Converted from corporate to local policy	02.23	03.16.23
Updated criteria for other diagnoses/indications	06.25.23	10.05.23

Reviews, Revisions, and Approvals	Date	LDH Approval Date
Annual review: no significant changes; references reviewed and updated.	06.14.24	09.04.24
Annual review: no significant changes; references reviewed and updated	05.08.25	

Important Reminder

This clinical policy has been developed by appropriately experienced and licensed health care professionals based on a review and consideration of currently available generally accepted standards of medical practice; peer-reviewed medical literature; government agency/program approval status; evidence-based guidelines and positions of leading national health professional organizations; views of physicians practicing in relevant clinical areas affected by this clinical policy; and other available clinical information. LHCCmakes no representations and accepts no liability with respect to the content of any external information used or relied upon in developing this clinical policy. This clinical policy is consistent with standards of medical practice current at the time that this clinical policy was approved.

The purpose of this clinical policy is to provide a guide to medical necessity, which is a component of the guidelines used to assist in making coverage decisions and administering benefits. It does not constitute a contract or guarantee regarding payment or results. Coverage decisions and the administration of benefits are subject to all terms, conditions, exclusions, and limitations of the coverage documents (e.g., evidence of coverage, certificate of coverage, policy, contract of insurance, etc.), as well as to state and federal requirements and applicable LHCC-level administrative policies and procedures.

This clinical policy is effective as of the date determined byLHCC. The date of posting may not be the effective date of this clinical policy. This clinical policy may be subject to applicable legal and regulatory requirements relating to provider notification. If there is a discrepancy between the effective date of this clinical policy and any applicable legal or regulatory requirement, the requirements of law and regulation shall govern. LHCC retains the right to change, amend or withdraw this clinical policy, and additional clinical policies may be developed and adopted as needed, at any time.

This clinical policy does not constitute medical advice, medical treatment, or medical care. It is not intended to dictate to providers how to practice medicine. Providers are expected to exercise professional medical judgment in providing the most appropriate care, and are solely responsible for the medical advice and treatment of members. This clinical policy is not intended to recommend treatment for members. Members should consult with their treating physician in connection with diagnosis and treatment decisions.

Providers referred to in this clinical policy are independent contractors who exercise independent judgment and over whom LHCC has no control or right of control. Providers are not agents or employees ofLHCC.