Washington State Health Care Authority

Endocrine and Metabolic Agents – Elapegademase-lvlr (Revcovi)

Medical policy no. 30.90.20.30-1 Effective Date: December 1, 2020

Note: *New-to-market drugs included in this class based on the Apple Health Preferred Drug List are non-preferred and subject to this prior authorization (PA) criteria. Non-preferred agents in this class require an inadequate response or documented intolerance due to severe adverse reaction or contraindication to at least TWO preferred agents. If there is only one preferred agent in the class documentation of inadequate response to ONE preferred agent is needed. If a drug within this policy receives a new indication approved by the Food and Drug Administration (FDA), medical necessity for the new indication will be determined on a case-by-case basis following FDA labeling.*

Background:

Adenosine deaminase (ADA) deficiency is an autosomal recessive genetic disorder caused by mutations in the ADA gene and a common cause of severe combined immune deficiency (SCID). Diagnosis of ADA-SCID may be suspected by newborn screening or confirmed by blood and genetic testing. Most patients with ADA-SCID experience complications such as pneumonia, chronic diarrhea, widespread skin rashes, slowed growth and developmental delay before 6 months of age. Patients with ADA-SCID are unable to fight off most types of bacterial, viral, and fungal infections. If undiagnosed, most patients do not survive past two years of age. The annual incidence of ADA-SCID is 1 in 200,000 livebirths affecting both males and females. Enzyme replacement therapy (ERT) is recommended for all patients newly diagnosed with ADA-SCID as an immediate stabilizing measure and as a bridge to curative therapy with hematopoietic stem cell transplant (HSCT). Elapegademase-lvlr (Revcovi) is a recombinant adenosine deaminase indicated for the treatment of ADA-SCID in pediatric and adult patients. Elapegademase-lvlr is an exogenous source of ADA enzyme that reduces levels of toxic adenosine and deoxyadenosine and increases lymphocytes.

Medical necessity

Drug	*Medical Necessity*
Elapegademase-lvlr (Revcovi)	Elapegademase-lvlr may be considered medically necessary when used for the treatment of: • Adenosine deaminase severe combined immune deficiency (ADA-SCID) in pediatric and adult clients

Clinical policy:

Drug	Clinical Criteria (Initial Approval)
Elapegademase-lvlr (Revcovi)	Elapegademase-lvlr (Revcovi) may be considered medically necessary when **ALL** of the following criteria are met: 1. Diagnosis of ADA-SCID confirmed by any **ONE** of the following: a. Genetic testing revealing bi-allelic mutations in the ADA gene; **OR** b. Absent or very low (< 1% of normal) ADA catalytic activity at baseline; **AND** 2. Patient does not have severe thrombocytopenia (<50,000/µL); **AND**

	3. Patient is not a candidate for HSCT, has failed HSCT, or is using elapegademase-lvlr as a bridge to definitive therapy with HSCT; **AND** 4. Prescribed and administered by or in consultation with a physician who specializes in the treatment of ADA-SCID; **AND** 5. Prescriber agrees to monitor **ALL** the following to ensure effectiveness and compliance: **a.** Trough plasma ADA activity (every 2 weeks for 8-12 weeks then every 3-6 months); **AND** **b.** Trough deoxyadenosine (dAXP) levels; **AND** **c.** Total lymphocyte counts; **AND** **d.** Neutralizing antibodies in client with trough ADA activity persistently less than 15 mmol/hr/L If **ALL** criteria are met, the request will be **approved for 12 months.**
	Criteria (Reauthorization) Elapegademase-lvlr (Revcovi) may be reauthorized when **ALL** of the following criteria are met: 1. Documentation of trough plasma ADA activity, trough dAXP levels, and total lymphocyte counts; **AND** 2. Trough plasma ADA activity is greater than or equal to 15 mmol/hr/L; **AND** 3. Trough erythrocyte dAXP levels are below 0.02 mmol/L and monitored at least twice a year; **AND** 4. Client is not a candidate for HSCT, has failed HSCT, or is using elapegademase-lvlr as a bridge to definitive therapy with HSCT If **ALL** criteria are met, the request will be **reauthorized for 12 months.**

Dosage and quantity limits

Drug Name	Dose and Quantity Limits
Elapegademase-lvlr (Revcovi™)	**Clients switching from pegademase bovine (Adagen®) to elapegademase-lvlr:** • If Adagen® dose is unknown or < 30U/kg: 0.2 mg/kg intramuscularly once weekly • If client's weekly Adagen® dose is above 30U/kg: $Elapegademase-lvlr\ dose\ \left(\frac{mg}{kg}\right) = \frac{Adagen\ dose\left(\frac{U}{kg}\right)}{150}$ • If client's trough ADA activity is < 30 mmol/hr/L, trough dAXP > 0.02mmol/L, or immune reconstituation is inadequate based on clinical assessment: Subsequent doses may be increased by increments of 0.033 mg/kg weekly

	Clients who are Adagen®-naïve: • 0.2 mg/kg based on ideal body weight twice a week intramuscularly for a minimum of 12 to 24 weeks until immune reconstitution is achieved • Maximum weekly dose: 0.4 mg/kg intramuscularly *Doses may be divided into multiple injections as long as weekly cumulative dose does not exceed 0.4 mg/kg

History:

Date	Action and summary of changes
02/05/2020	New policy
02/17/2020	Added 0.2 mg/kg dosing for Adagen naïve
02/26/2020	Formatted Adagen dose equation and changed context of the term covered
03/09/2020	Changed approval date from 6 months to 12 months. Changed re-approval date to 6 months.
03/19/2020	Added note to the top of the policy
06/17/2020	Approved by DUR Board

References

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3. Center for Drug Evaluation and Research. Accessdata.fda.gov. https://www.accessdata.fda.gov/drugsatfda_docs/nda/2018/761092Orig1s000MedR.pdf. Published 2020. Accessed June 22, 2020.
4. Elapegademase-lvlr. Micromedex Solutions. Truven Health Analytics, Inc. Ann Arbor, MI. Available at: http://www.micromedexsolutions.com. Accessed February 5, 2020
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6. Kohn DB, Hershfield MS, Puck JM, Aiuti A, Blincoe A, Gaspar HB, etal. Consensus approach for the management of severe combined immune deficiency caused by adenosine deaminase deficiency. J Allergy Clin Immunol. 2018;S0091- 6749(18):31268-5
7. Revcovi Prescribing Information. Gaithersburg, MD: Leadiant Biosciences Inc.; October 2018. Available at: www.revcovi.com. Accessed February 5, 2020.