Washington State Health Care Authority

Endocrine and Metabolic Agents – Elapegademase-lvlr (Revcovi)

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

Note: New-to-market drugs in this class are non-preferred and subject to this prior authorization (PA) policy. 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.

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. Client does not have severe thrombocytopenia (<50,000/µL); **AND** 3. Client 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 at least 30 mmol/hr/L; **AND** 3. Trough dAXP levels are below 0.02 mmol/L and monitored at least twice a year; **AND** 4. Prescriber verifies client is still not an eligible candidate for HSCT If **ALL** criteria are met, the request will be **reauthorized for 6 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: 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 and trough dAXP > 0.02mmol/L: Subsequent doses may be increased by increments of 0.033 mg/kg weekly **Clients who are Adagen®-naïve:** • 0.2 mg/kg 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

~~Coding:~~

~~HCPCS Code~~	~~Description~~
~~J3590~~	~~Unclassified biologics~~
~~C9399~~	~~Unclassified drugs or biologicals~~

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.

References

1. ADA deficiency - Genetics Home Reference - NIH. U.S. National Library of Medicine. https://ghr.nlm.nih.gov/condition/adenosine-deaminase-deficiency#diagnosis. Accessed February 5, 2020.
2. Adenosine deaminase deficiency. Genetic and Rare Diseases Information Center. https://rarediseases.info.nih.gov/diseases/5748/adenosine-deaminase-deficiency. Accessed February 5, 2020.
3. Elapegademase-lvlr. Micromedex Solutions. Truven Health Analytics, Inc. Ann Arbor, MI. Available at: http://www.micromedexsolutions.com. Accessed February 5, 2020
4. Grunebaum E, Kohn DB. Adenosine deaminase deficiency: Treatment and prognosis. In: Post T, ed. *UpToDate*. Waltham, MA.: UpToDate; 2019. www.uptodate.com. Accessed February 5, 2020.
5. 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
6. Revcovi Prescribing Information. Gaithersburg, MD: Leadiant Biosciences Inc.; October 2018. Available at: www.revcovi.com. Accessed February 5, 2020.

Washington State Health Care Authority

Endrocrine and Metabolic:
Adenosine Deaminase SCID Treatment Agents

Please provide the information below, please print your answer, attach supporting documentation, sign, date, and return to our office as soon as possible to expedite this request. **Without this information, we may deny the request in seven (7) working days.**

Date of request:	Reference #:	MAS:	
Patient	Date of birth	ProviderOne ID	
Pharmacy name	Pharmacy NPI	Telephone number	Fax number
Prescriber	Prescriber NPI	Telephone number	Fax number

Medication and strength	Directions for use	Qty/Days supply

1. Is this request for a continuation of existing therapy? ☐ Yes ☐ No

2. Is patient's diagnosis adenosine deaminase severe combined immune deficiency (ADA-SCID)? ☐ Yes ☐ No
   If no, specify diagnosis:

3. Does patient have severe thrombocytopenia (platelets <50,000/μL)? ☐ Yes ☐ No

4. Has patient failed, or is not a candidate for, hematopoietic stem cell transplantation (HSCT)? ☐ Yes ☐ No

5. Is patient using elapegademase-lvlr as a bridge to definitive therapy with HSCT? ☐ Yes ☐ No

6. Is this prescribed and will be administered by or in consultation with a physician who specializes in the treatment of ADA-SCID? ☐ Yes ☐ No

7. If approved, does prescriber agree to monitor trough plasma ADA activity, trough deoxyadenosine (dAXP) levels, total lymphocyte counts and neutralizing antibodies? ☐ Yes ☐ No

8. Provide the following for patient:
   Trough plasma ADA activity: _____mmol/hr/L Date taken: _____
   Trough dAXP levels: _____mmol/L Date taken: _____

9. Is patient pegademase bovine (Adagen) naïve? ☐ Yes ☐ No
   If no, what was patient's dose? _____mg/kg

10. What is patient's current weight?
    Actual: _____lb _____kg Date taken: _____
    Ideal: _____lb _____kg

ALL OF THE FOLLOWING ARE REQUIRED WITH THIS REQUEST:

- Genetic testing revealing bi-allelic mutations in the ADA gene or absent or very low (< 1% of normal) ADA catalytic activity at baseline confirming diagnosis
- Most recent CBC and labs
- Documentation of therapy monitoring measuring trough plasma ADA activity, trough dAXP levels, and/or total lymphocyte counts (for reauthorization requests)
- Chart notes

Prescriber signature	Prescriber specialty	Date