Protocol for Revcovi® (elapegademase-lvlr)
Approved April 2022

Background:
Inherited deficiency of adenosine deaminase (ADA; now often referred to as ADA1) causes a subtype of severe combined immunodeficiency (SCID) characterized by unique effects on lymphoid and nonlymphoid cells.

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

Criteria for approval:
1. Diagnosis of ADA-SCID is confirmed by the following:
   a. Absent or very low (<1% of normal) ADA activity in RBCs, which is accompanied by increased levels of adenosine; AND
   b. Elevated deoxyadenosine triphosphate (dATP) levels in erythrocyte lysates compared to
   laboratory standard; OR
   c. Genetic testing confirming biallelic mutations in the ADA gene
2. Medication is prescribed by or in consultation with an immunologist, hematologist/oncologist, or a physician who is an expert in adenosine deaminase severe combined immune deficiency (ADASCID) or related disorders
3. Medication is prescribed in accordance with Food and Drug Administration (FDA) established indication and dosing regimens or in accordance with medically appropriate off-label indication and dosing according to American Hospital Formulary Service, Micromedex, Clinical Pharmacology, Wolters Kluwer Lexi-Drugs (Lexicomp), national guidelines, or other peer-reviewed evidence

Initial Approval: 6 months

Continuation of therapy:
1. Patient has experienced a positive clinical response to elapegademase as demonstrated by the following:
   a. Adequate trough plasma ADA activity levels have been maintained; OR
   b. Adequate deoxyadenosine levels, and/or total lymphocyte counts have been maintained; OR
   c. Decreased frequency of infections
2. Medication is prescribed in accordance with Food and Drug Administration (FDA) established indication and dosing regimens or in accordance with medically appropriate off-label indication and dosing according to American Hospital Formulary Service, Micromedex, Clinical Pharmacology, Wolters Kluwer Lexi-Drugs (Lexicomp), national guidelines, or other peer-reviewed evidence.

Renewal Approval: 6 months

References:

1. Revcovi [prescribing information]. Chiesi USA, Inc. Cary, NC 27518. December 2020
2. Clinical Pharmacology® Gold Standard Series [Internet database]. Tampa FL. Elsevier 2019. Updated periodically
3. Kohn DB, Hershfield MS, et al. Consensus approach for the management of severe combined immune deficiency caused by adenosine deaminase deficiency. J Allergy Clin Immunol 2019;143:852-863