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REVCovi (elapegademase-lvlr)

RATIONALE FOR INCLUSION IN PA PROGRAM

Background

Severe combined immune deficiency (SCID) associated with a deficiency of ADA (adenosine deaminase) enzyme is a rare, inherited, and often fatal disease. ADA enzyme is involved in purine metabolism, catalyzing the irreversible hydrolytic deamination of adenosine or deoxyadenosine to inosine or deoxyinosine, respectively, as well as several naturally occurring methylated adenosine compounds. Maintaining a low level of 2'-deoxyadenosine and adenosine is crucial for proper number and function of immune cells as well as decreasing the frequency of opportunistic infections. Elevated adenosine levels, as occurring in ADA deficiency, contribute to apoptosis and a block in the differentiation of thymocytes, causing severe T-lymphopenia (1).

Revcovi (elapegademase-lvlr) provides an exogenous source of ADA enzyme that is associated with a decrease in toxic adenosine and deoxyadenosine nucleotides levels as well as an increase in lymphocyte number (1).

Regulatory Status

FDA-approved indication: Revcovi is a recombinant adenosine deaminase, indicated for the treatment of adenosine deaminase severe combined immune deficiency (ADA-SCID) in pediatric and adult patients (1).

Revcovi is administered by IM injection and thus it should be used with caution in patients with thrombocytopenia due to risk of injection site bleeding, and should not be used if the thrombocytopenia is severe (1).

There may be a delay in the improvement of immune function following administration of Revcovi. Precautions should be maintained to protect immune deficient patients from infections until improvement in immune function has been achieved. The timing and degree of improvement in immune function may vary from patient to patient (1).

Once treatment with Revcovi has been initiated, a target trough plasma ADA activity should be at least 30 mmol/hr/L. A decrease of ADA activity below this level suggests noncompliance to treatment or a development of antibodies. Antibodies to Revcovi should be suspected if a



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persistent fall in pre-injection levels of trough plasma ADA activity below 15 mmol/hr/L occurs. 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. In these situations, testing for Revcovi antibodies should be undertaken (1).

Two months after starting Revcovi treatment, trough erythrocyte dAXP levels should be maintained below 0.02 mmol/L, and monitored at least twice a year. Total and subset lymphocytes should also be monitored periodically (1).

The safety and effectiveness of Revcovi in pediatric patients have been established (1).

Summary

Revcovi (elapegademase-lvlr) provides an exogenous source of ADA enzyme that is associated with a decrease in toxic adenosine and deoxyadenosine nucleotides levels as well as an increase in lymphocyte number. The safety and effectiveness of Revcovi in pediatric patients have been established (1).

Prior authorization is required to ensure the safe, clinically appropriate and cost-effective use of Revcovi while maintaining optimal therapeutic outcomes.

References

1. Revcovi [package insert]. Indianapolis, IN: Chiesi USA, Inc.; December 2020.