ELELYSO™ is an enzyme replacement therapy (ERT) for the long term treatment of adults with type 1 Gaucher disease. ELELYSO is a form of the human lysosomal enzyme, glucocerebrosidase, which is genetically engineered in carrot cells. Taliglucerase alfa is an analogue of glucocerebrosidase; it is produced by recombinant DNA technology using plant (carrot) cell culture. Glucocerebrosidase is an enzyme deficient in Gaucher's disease. It is needed to catalyze the hydrolysis of glucocerebroside to glucose and ceramide, thereby reducing liver and spleen size and improving anemia and thrombocytopenia.

**Pre-Authorization Criteria:**

treatment of long-term enzyme replacement therapy for patients with type 1 Gaucher's disease

Note: VCHCP requires that ELELYSO be prescribed by a Geneticists or Hematologists.

**Dosing: Adult**
The recommended dose is 60 Units/kg administered every other week as a 60-120 minute intravenous infusion.

Dosing is individualized based on disease severity. Dosing range: 11-73 units/kg every 2 weeks.
Note: Pretreatment with antihistamines and/or corticosteroids can be considered for prevention of subsequent infusion reactions in patients with an infusion reaction requiring symptomatic treatment; during clinical studies, patients were not routinely premedicated prior to infusion. Round up to the next whole vial when determining the number of vials needed.

Warnings/Precautions

Concerns related to adverse effects:

• Anaphylactic reactions: Use with caution in patients who have exhibited hypersensitivity reactions to taliglucerase alfa or other enzyme replacement therapies. Anaphylaxis has occurred; appropriate medical support should be readily available in the event of a serious reaction.

• Antibody formation: The development of IgG anti-drug antibodies (ADA) has been reported; the clinical significance is unknown. Patients who develop immune or infusion reactions to taliglucerase alfa or who have had an immune response to other enzyme replacement therapies and who are switching to taliglucerase alfa should be monitored for antibody development; it is unknown if presence of antibodies is related to a higher risk of infusion reactions.

• Infusion-related reaction: Infusion-related reactions (eg, headache, chest pain, fatigue, urticaria, arthralgia), including allergic reactions, occurring within 24 hours were the most commonly reported adverse drug reactions in clinical trials. Management strategies for reactions include symptomatic treatment, pretreatment with antihistamines, antipyretics, and/or corticosteroids, and slowing of the infusion rate.
REFERENCES


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