PROCEDURE

ORIGINAL DATE: 12/94
Revised Date: 09/02

SUBJECT: CEREDASE ALGLUCERASE INFUSION

PURPOSE: To safely administer Ceredase intravenously to patients with Type I Gaucher's disease.

BACKGROUND

Ceredase is a modified form of the enzyme glucocerebrosidase and is purified from a large pool of human placental tissue collected from selected donors. Ceredase catalyzes the hydrolysis of the glycolipid, glucocerebroside, to glucose and ceramide as part of the normal degradation pathway for membrane lipids. Glucocerebroside is primarily derived from hematologic cell turnover. Gaucher Disease is characterized by a functional deficiency in B-glucocerebrosidase enzymatic activity and resultant accumulation of lipid glucocerebroside in tissue macrophages which become engorged and are termed "Gaucher cell". Gaucher cells are typically found in the liver, spleen and bone marrow and occasionally, as well, in the lung, kidney and intestine. Secondary hematologic sequelae includes severe anemia and thrombocytopenia in addition to the characteristic progressive hepatosplenomegaly. Skeletal complications including osteonecrosis and osteopenia with secondary pathological fracture are a common feature of Gaucher Disease.

INDICATIONS

Ceredase is indicated for use as long term enzyme replacement for patients with a confirmed diagnosis of Type 1 Gaucher Disease who develop one or more of the following conditions:

- Moderate to severe anemia
- Thrombocytopenia with bleeding tendency
- Bone disease
- Significant hepatomegaly or splenomegaly

CONSIDERATIONS

1. The patient must meet requirements to be admitted to Home Health VNA's IV Therapy Team.
2. micron filters shall be used during administration.
3. Patients who meet criteria for self-administration of Ceredase may do so after instruction by a VNA nurse.
4. LPN's may not administer Ceredase

PROCEDURE

1. Prepare work area aseptically with soap and water.
2. Assemble equipment:
   - Ceredase vials, sterile water for injection, 100-200cc bag NS, syringes, tubing.
   - Filter
   - Venous access device materials
3. Reconstitute Ceredase vials per manufacturer's instructions, once reconstituted, add the Ceredase to the bag of normal saline.
4. Avoid shaking the medication bag since shaking may de-nature the glycoprotein rendering it biologically inactive.
5. Inspect visually for discoloration or particular matter. If noted, do not use; inform Pharmacist.
6. Position patient comfortably.
7. Perform venipuncture/venous access.
8. Begin infusion at initial dose of up to 60 units/kg (dosage will be individualized for each patient) in no more than 100cc of normal saline. This infusion will last from one (1) to four (4) hours depending on patient tolerance. Frequency is determined by disease severity and physician order. After patient response is well established, dosage may be adjusted downward for maintenance therapy at 3-6 month intervals. Evidence suggests that glucocerebroside lipid storage may respond to doses as low as 1 unit/kg.
9. Although adverse reactions are not readily reported or in need of medical interventions, instruct patient to report any of the following symptoms.
   - Fever
   - Chills
   - Abdominal discomfort
   - Nausea and vomiting
10. When infusion is complete, discontinue and dispose needles in sharps container and call IV vendor for pick up.
11. Document in patient record:
    - Dosage
    - Length of infusion
    - Response to therapy i.e.; decrease of bone pain, bleeding episodes, hepatomegaly
    - Frequency of infusion
    - Any noted adverse reactions
    - Weight
    - Absence or presence of bone pain