Enzyme Replacement Therapy for Gaucher's Disease

**Description:**

Gaucher's disease is a rare inherited enzyme deficiency that manifests itself by a variety of clinical signs and symptoms. It can range from severely affected infants to asymptomatic adults. Many patients suffer from anemia, bone damage, enlarged livers and spleens; a few develop severe central nervous system damage. There are three types of Gaucher's disease.

1. Type I: Chronic non-neuronopathic is the most common and affects both children and adults.
2. Type II: Acute neuronopathic involves the central nervous system and survival is rarely beyond age 2.
3. Type III: Subacute neuronopathic involves the central nervous system, but its course is variable with slower progression.

Imiglucerase (Cerezyme®) and alglucerase (Ceredase®) are modified forms of the enzyme B-glucocerebrosidase which are used for the treatment of Type I Gaucher's disease. The enzyme replacement therapy has resulted in a reduction in liver and spleen volume and consequently improved anemia and thrombocytopenia in most patients. These drugs are administered through an intravenous infusion, usually in a doctor's office, outpatient hospital, or occasionally in the home. The therapy is administered as often as weekly or as infrequently as monthly, dependent on the individual's response and medical indications. Dosing can range from as low a 30U/kg every two weeks to 60U/kg every two weeks initially, depending on the severity of the disease.

**Medical Criteria:**

Not applicable as this is a reimbursement policy.

**Policy:**

The use of imiglucerase (Cerezyme) and alglucerase (Ceredase) in the treatment of Type I Gaucher's is covered for all product lines.

**Coverage:**

Benefits may vary between groups/contracts. Please refer to the appropriate benefit booklet, subscriber agreement, or Rite Care contract for the applicable infusion therapy benefits/coverage.

**Coding:**

Reimbursement for Ceredase and Cerezyme is covered when billed with the appropriate NDC# or J codes listed below:

J0205 Injection, alglucerase, per 10 units
J1785 Injection, Imiglucerase, per unit

**Also Known As:**

Lysosomal storage disease
Ceredase
Cerezyme

**Related topics:**
This medical policy is made available to you for informational purposes only. It is not a guarantee of payment or a substitute for your medical judgement in the treatment of your patients. Benefits and eligibility are determined by the member's subscriber agreement or member certificate and/or the employer agreement, and those documents will supersede the provisions of this medical policy. For information on member-specific benefits, call the provider call center. If you provide services to a member which are determined to not be medically necessary (or in some cases medically necessary services which are non-covered benefits), you may not charge the member for the services unless you have informed the member and they have agreed in writing in advance to continue with the treatment at their own expense. Please refer to your participation agreement(s) for the applicable provisions.

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