Prior Authorization Criteria for Aldurazyme
(laronidase, alpha-L-iduronidase)

Aldurazyme is recombinant human alpha-L-iduronidase, an orphan drug recently approved by the FDA for enzyme replacement therapy in mucopolysaccharidosis (MPS), type I, a lysosomal storage disorder caused by deficiency of the enzyme. Aldurazyme has been shown to improve pulmonary function and walking capacity. Aldurazyme has not been evaluated for effects on the central nervous system manifestations of the disorder.

Aldurazyme is indicated for patients with:
- Hurler form of MPS I
- Hurler-Scheie form of MPS I
- Scheie form of MPS I, only if there are moderate to severe symptoms. The risks and benefits of treating mildly affected patients with the Scheie form have not been established.

Aldurazyme is administered as a once weekly intravenous infusion. It is dosed at 0.58 mg/kg of body weight. The patient should be pretreated with antipyretics and/or antihistamines, 60 minutes prior to the start of the infusion.

Criteria necessary for approval of Aldurazyme:

Documentation of the presence of Hurler or Hurler-Scheie forms of MPS I must be submitted. In patients with the Scheie form of MPS I, documentation of the presence of the disorder, along with the presence of moderate to severe symptoms must be submitted.

When approved, this medication must be obtained through our specialty pharmacy,
**Prior Authorization Criteria for Elaprase**

(idursulfase)

Hunter syndrome is an X-linked, recessive, lysosomal storage disease that is caused by a defect of the iduronate-2-sulfatase gene. Elaprase is indicated for treatment of patients with Hunter syndrome (mucopolysaccaridosis type II [MPS II]), to improve walking capacity in patients 5 years and older. Although it is approved for children down to 5 years of age, children down to 16 months of age have been treated. Safety and efficacy in children younger than 16 months of age has not been established.

Elaprase is dosed at 0.5 mg/kg as an IV infusion once a week. Administer the diluted solution using an infusion set containing a low-protein-binding 0.2 micrometer in-line filter.

In June, 2013, prescribing information was revised to include a boxed warning about life-threatening anaphylactic reactions, presenting as respiratory distress, hypoxia, hypotension, urticaria, and/or angioedema of throat or tongue have occurred in some patients during and up to 24 hours after Elaprase infusions.

**Criteria necessary for approval of Elaprase:**

Documentation of the presence of Hunter syndrome (MPS II) must be submitted. When approved, this medication must be obtained through our specialty pharmacy,