VENTURA COUNTY
HEALTH CARE PLAN

Prior Authorization DRUG Guidelines

ELAPRASE (Idursulfase)

Effective Date: 1/28/14

Date Developed: 1/28/14 by Catherine Sanders, MD

Last Approval Date: 1/26/16, 1/24/17, 1/23/18

Elaprase is a recombinant form of iduronate-2-sulfatase, an enzyme needed to hydrolyze the mucopolysaccharides dermatan sulfate and heparan sulfate in various cells. Accumulation of these polysaccharides can lead to various manifestations of disease, including physical changes, CNS involvement, cardiac, respiratory, and mobility dysfunction. Replacement of this enzyme has been shown to improve walking capacity in patients with a deficiency.

Pre-Authorization Criteria:
Elaprase is used for replacement therapy in Hunter syndrome (mucopolysaccharidosis II) for improvement of walking capacity.

VCHCP requires that Elaprase be prescribed by a physician experienced in the treatment of Hunter syndrome.

Registry: Patients and healthcare providers are encouraged to participate in the Hunter Outcome Survey, intended to monitor disease progression, patient outcomes, and long-term effects of therapy. For more information, refer to www.elaprase.com or call OnePath[sm] at 1-866-888-0660.

Dosing: Adult:
Hunter syndrome (mucopolysaccharidosis II): I.V.: 0.5 mg/kg once weekly

Dosing: Pediatric:
Hunter syndrome (mucopolysaccharidosis II): Children ≥5 years and Adolescents: Refer to adult dosing.

Dosing: Renal Impairment:
No dosage adjustment provided in manufacturer's labeling.

Dosing: Hepatic Impairment:
No dosage adjustment provided in manufacturer's labeling.

Dosage Forms: U.S.:
Excipient information presented when available (limited, particularly for generics); consult specific product labeling.
Solution, Intravenous [preservative free]:
Elaprase: 6 mg/3 mL (3 mL)

Generic Equivalent Available: U.S.-No

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Administration:
Administer using an infusion set containing a 0.2 micron low protein-binding inline filter. Infuse at an initial rate of 8 mL/hour for the first 15 minutes. If tolerated, may increase rate by 8 mL/hour increments every 15 minutes; maximum infusion rate of up to 100 mL/hour. Rate may be decreased, temporarily stopped, or discontinued based on tolerance. Initial infusion should be over 3 hours; if tolerated, subsequent infusions may be gradually reduced to a 1-hour infusion. Total infusion time should not exceed 8 hours.

Compatibility: Stable in NS.

Adverse Reactions:
>10%: hypertension, atrial abnormality, pyrexia, headache, malaise, anxiety, irritability, pruritus, urticaria, pruritic rash, skin disorder, dyspepsia, infusion site edema, arthralgia, limb pain, chest wall musculoskeletal pain, musculoskeletal dysfunction, visual disturbance, wheezing, antibody development, infusion reactions.
Other Serious Less Common Reactions: respiratory failure, cyanosis, pneumonia, pulmonary embolism, cardiac failure.

U.S. BOXED WARNING:
Life-threatening anaphylactic reactions have occurred during and up to 24 hours after infusion regardless of treatment duration; signs and symptoms include respiratory distress, hypoxia, hypotension, urticaria, and/or tongue/throat angioedema; observe patients during and after administration; inform patients of signs and symptoms and to seek immediate medical care if they occur; additional monitoring required for patients with compromised respiratory function or acute respiratory disease due to risk of serious acute respiratory exacerbation from hypersensitivity reactions.

References:

1. Muenzer J, Beck M, Eng CM, et al, "Long-Term, Open-Labeled Extension Study of Idursulfase in the Treatment of Hunter Syndrome," *Genet Med*, 2011, 13(2):95-101. [PubMed 21150784]
2. Muenzer J, Wraith JE, Beck M, wet al, "A Phase II/III Clinical Study of Enzyme Replacement Therapy With Idursulfase in Mucopolysaccharidosis II (Hunter Syndrome)," *Genet Med*, 2006, 8(8):465-73. [PubMed 16912578]
3. www.uptodate.com: Idursulfase: Drug Information
4. www.epocrates.com: Elaprase Drug information

REVISION HISTORY:

Date Reviewed/No Updates: 1/13/15 by C. Sanders, MD
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Revision Date	Content Revised (Yes/No)	Contributors	Review/Revision Notes
1/24/17	No	Catherine Sanders, MD; Robert Sterling, MD	Annual review
1/23/18	No	Catherine Sanders, MD; Robert Sterling, MD	Annual review