Agalsidase beta: Drug information

Brand Names: US

Fabrazyme

Brand Names: Canada

Fabrazyme

Pharmacologic Category: Enzyme

Mechanism of Action

Agalsidase beta is a recombinant form of the enzyme alpha-galactosidase-A, which is required for the hydrolysis of GL-3 and other glycosphingolipids. The compounds may accumulate (over many years) within the tissues of patients with Fabry disease, leading to renal and cardiovascular complications.

Use: **Fabry disease:** For use in patients with Fabry disease.

Pricing: US

Solution (reconstituted) (Fabrazyme Intravenous)

5 mg (per each): $1,010.40

35 mg (per each): $7,072.80

Pricing: Brasil

Solution (reconstituted) (Fabrazyme Intravenous)

35 mg (per each): R$ 14.047,67

Dosage Forms

Excipient information presented when available (limited, particularly for generics); consult specific product labeling.

Solution Reconstituted, Intravenous [preservative free]:

Fabrazyme: 5 mg (1 ea); 35 mg (1 ea) [contains mouse (murine) and/or hamster protein]

Generic Equivalent Available (US): No

Dosing: Adult

Fabry disease: IV: 1 mg/kg every 2 weeks

Children ≥8 years and Adolescents: IV: Refer to adult dosing

Dosing: Renal Impairment: Adult

No dosage adjustment required.

Dosing: Hepatic Impairment: Adult

There are no dosage adjustments provided in the manufacturer's labeling.

Administration

IV: Antipyretics should be administered prior to infusion. Infuse through a low protein binding 0.2 micron in-line filter. Initial infusion rate should not exceed 0.25 mg/minute (15 mg/hour). Interrupt or decrease rate in the event of an infusion reaction; may be restarted after resolution of symptoms and/or after administration of antipyretics, antihistamines, and/or steroids. After patient tolerance to the infusion is established, rate may be increased in increments of 0.05-0.08 mg/minute (3-5 mg/hour) with each subsequent infusion. Maximum infusion rate: Patients <30 kg: 0.25 mg/minute; patients ≥30 kg: Infuse over at least 1.5 hours. An initial maximum infusion rate of 0.01 mg/minute should be used for rechallenge in patients with IgE antibodies or who have had a positive skin test to agalsidase beta; may increase infusion rate (doubling the infusion rate every 30 minutes) to a maximum rate of 0.25 mg/minute as tolerated.

Adverse Reactions

>10%:

Cardiovascular: Peripheral edema (21%), hypertension (14%)

Central nervous system: Chills (43%), headache (39%), paresthesia (31%), procedural pain (25%), fatigue (24%), dizziness (21%), pain (16%), sensation of cold (11%)

Dermatologic: Skin rash (20%)

Immunologic: Development of IgG Antibodies (69% to 79%)

Local: Infusion site reaction (50% to 55%, severe ≥5%)

Neuromuscular & skeletal: Limb pain (19%), back pain (16%), myalgia (14%)

Respiratory: Upper respiratory tract infection (44%), cough (33%), nasal congestion (19%), lower respiratory tract infection (18%)

Miscellaneous: Fever (39%)

1% to 10%:

Cardiovascular: Tachycardia (9%), bradycardia (≥5%), chest discomfort (≥5%), chest pain (≥5%), facial edema (≥5%), flushing (≥5%), hypotension (≥5%), ventricular hypertrophy (5%)

Central nervous system: Hypoesthesia (9%), anxiety (6%), burning sensation (6%), depression (6%), falling (6%)

Dermatologic: Pruritus (10%), excoriation (9%), pallor (≥5%), urticaria (≥5%), thermal injury (4%)

Gastrointestinal: Toothache (6%), abdominal pain (≥5%), diarrhea (≥5%), nausea (≥5%), vomiting (≥5%), xerostomia (4%)

Hematologic & oncologic: Bruise (4%)

Hypersensitivity: Anaphylaxis, hypersensitivity reaction

Infection: Fungal infection (5%), viral infection (5%), localized infection (4%)

Neuromuscular & skeletal: Muscle spasm (5%)

Otic: Tinnitus (8%), hypoacusis (5%)

Renal: Increased serum creatinine (9%)

Respiratory: Sinusitis (9%), dyspnea (8%), respiratory congestion (8%), pharyngitis (6%), wheezing (6%), pharyngeal edema (≥5%)

Miscellaneous: Procedural complications (postprocedure, 10%)