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5.30.008

Section:	Prescription Drugs	**Effective Date:**	April 1, 2023
Subsection:	Endocrine and Metabolic Drugs	**Original Policy Date:**	September 9, 2008
Subject:	Elaprase	**Page:**	1 of 4

Last Review Date: March 10, 2023

Elaprase

Description

Elaprase (idursulfase)

Background
Hunter Syndrome (mucopolysaccharidosis type II or MPS II) is an X-linked recessive disease caused by the body's inability to break down certain elements in the body called mucopolysaccharides, also known as glycosaminoglycans (GAG) due to insufficient levels of the lysosomal enzyme iduronate-2-sulfatase (I2S). The missing or defective enzyme causes mucopolysaccharides to accumulate in a variety of cells, leading to cellular growth, organ enlargement, tissue destruction, and organ system dysfunction. Elaprase (idursulfase) replaces the deficient or absent enzyme to breakdown the excess buildup within the cells, returning the cells to normal size (1).

Regulatory Status
FDA-approved indication: Elaprase is a hydrolytic lysosomal glycosaminoglycan (GAG)-specific enzyme indicated for patients with Hunter syndrome (Mucopolysaccharidosis II, MPS II) is indicated for patients with Hunter syndrome (Mucopolysaccharidosis II, MPS II). Elaprase has been shown to improve walking capacity in patients 5 years or older. In patients 16 months to 5 years of age, no data are available to demonstrate improvement in disease-related symptoms or long term clinical outcome; however, treatment with Elaprase has reduced spleen volume similarly to that of adults and children 5 years of age and older (1).

Elaprase carries a boxed warning regarding the risk of life-threatening anaphylaxis reactions during infusions. Patients with compromised respiratory function or acute respiratory disease may be at risk of serious acute exacerbation of their respiratory compromise due to infusion reactions, and require additional monitoring. Appropriate medical support should be readily available when Elaprase is administered (1).

Safety and efficacy have not been established in pediatric patients less than 16 months of age of age (1).

Related policies
Aldurazyme, Mepsevii, Naglazyme, Vimizim

Policy

This policy statement applies to clinical review performed for pre-service (Prior Approval, Precertification, Advanced Benefit Determination, etc.) and/or post-service claims.

Elaprase may be considered **medically necessary** for the treatment of Hunter syndrome in patients 5 years of age or older.

Elaprase may be considered **investigational** in patients younger than 5 years of age and for all other indications.

Prior-Approval Requirements

Age 5 years of age or older

Diagnosis

Patient must have the following:

Hunter Syndrome (mucopolysaccharidosis type II)

Prior – Approval *Renewal* Requirements

Same as above

Policy Guidelines

Pre - PA Allowance

None

Prior - Approval Limits

Duration 2 years

Prior – Approval *Renewal* Limits

Same as above

Rationale

Summary

Elaprase (idursulfase) is indicated for patients with Hunter syndrome (Mucopolysaccharidosis II, MPS II). Elaprase carries a boxed warning regarding the risk of life-threatening anaphylaxis reactions during infusions. Patients with compromised respiratory function or acute respiratory disease may be at risk of serious acute exacerbation of their respiratory compromise due to infusion reactions, and require additional monitoring. Appropriate medical support should be readily available when Elaprase is administered. Safety and efficacy have not been established in pediatric patients less than 16 months of age (1).

Prior approval is required to ensure the safe, clinically appropriate, and cost-effective use of Elaprase while maintaining optimal therapeutic outcomes.

References

1. Elaprase [package insert]. Lexington, MA.: Shire Human Genetic Therapies, Inc.; September 2021.

Policy History

Date	Action
September 2011	New Policy aligned with MPRM.
September 2012	Annual editorial review and reference update.
June 2013	Annual editorial review and reference update.
September 2014	Annual editorial review and reference update.
September 2015	Annual review.
September 2016	Annual editorial review and reference update Policy number change from 5.30.08
December 2017	Annual review and reference update
June 2018	Annual editorial review
December 2019	Annual editorial review and reference update. Changed approval duration from lifetime to 2 years

December 2020	Annual review
March 2021	Annual editorial review
March 2022	Annual review
March 2023	Annual review and reference update. Changed policy number to 5.30.008

Keywords

This policy was approved by the FEP® Pharmacy and Medical Policy Committee on March 10, 2023 and is effective on April 1, 2023.