



# CIGNA PHARMACY COVERAGE POLICY

The following Coverage Policy applies to all plans administered by CIGNA Companies including plans administered by Great-West Healthcare, which is now a part of CIGNA.

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Coverage Policy Number ..... 1009

Subject **Idursulfase (Elaprase®)**

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## Hyperlink to Related Coverage Policies

### INSTRUCTIONS FOR USE

Coverage Policies are intended to provide guidance in interpreting certain **standard** CIGNA HealthCare benefit plans as well as benefit plans formerly administered by Great-West Healthcare. Please note, the terms of a participant's particular benefit plan document [Group Service Agreement (GSA), Evidence of Coverage, Certificate of Coverage, Summary Plan Description (SPD) or similar plan document] may differ significantly from the standard benefit plans upon which these Coverage Policies are based. For example, a participant's benefit plan document may contain a specific exclusion related to a topic addressed in a Coverage Policy. In the event of a conflict, a participant's benefit plan document **always supercedes** the information in the Coverage Policies. In the absence of a controlling federal or state coverage mandate, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of 1) the terms of the applicable group benefit plan document in effect on the date of service; 2) any applicable laws/regulations; 3) any relevant collateral source materials including Coverage Policies and; 4) the specific facts of the particular situation. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. Proprietary information of CIGNA. Copyright ©2011 CIGNA

## Coverage Policy

**CIGNA covers idursulfase (Elaprase®) as medically necessary for Hunter syndrome [Mucopolysaccharidosis II (MPS II)].**

**When coverage is available and medically necessary, the dosage, frequency, site of administration, and duration of therapy should be reasonable, clinically appropriate, and supported by evidence-based literature and adjusted based upon severity, alternative available treatments, and previous response to idursulfase (Elaprase®) therapy.**

### FDA Approved Indications

Elaprase is indicated for patients with Hunter syndrome (Mucopolysaccharidosis II (MPS II)). Elaprase has been shown to improve walking capacity in these patients.

### FDA Recommended Dosing

The recommended dosage regimen of Elaprase is 0.5 mg/kg of body weight administered every week as an intravenous infusion.

### Black Box Warning

**Risk of anaphylaxis - Life-threatening anaphylactic reactions have been observed in some patients during Elaprase infusions. Therefore, appropriate medical support should be readily available when Elaprase is administered. Biphasic anaphylactic reactions have also been observed after Elaprase**

administration and patients who have experienced anaphylactic reactions may require prolonged observation. 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.

## Drug Availability

Elaprase is a sterile, aqueous, clear to slightly opalescent colorless solution supplied in a 5 mL Type I glass vial.

## General Background

### Disease Overview

Hunter syndrome is a rare genetic disorder inherited through an X-linked recessive method. Patients with Hunter syndrome have low levels of the enzyme iduronate-2-sulfatase (I2S), and some patients completely lack the enzyme or have inactive I2S. A deficiency of I2S leads to an accumulation of the glycosaminoglycans (GAGs) dermatan sulfate and heparin sulfate in lysosomes in various tissues. The build-up of GAGs, or mucopolysaccharides, ultimately causes damage to major organs in the body.

### Pharmacology

Idursulfase replaces the enzyme iduronate-2-sulphatase (I2S), which catalyses the breakdown of the glycosaminoglycans (GAGs) dermatan sulfate and heparin sulfate in lysosomes. A deficiency of I2S leads to an accumulation of dermatan sulfate and heparin sulfate in lysosomes, and the build-up of these cellular waste products in tissues and organs ultimately results in the inability of tissues and organs to function properly. Idursulfase has a short elimination half-life of 44-48 minutes following intravenous infusion.

### Clinical Efficacy

One published trial of idursulfase is available. In this phase II/III trial, efficacy data for idursulfase were obtained by measuring the change in walking distance during a six minute time period and the percent predicted forced vital capacity (FVC) from baseline to the end of the 53 week study period. The weekly idursulfase group showed a statistically significant 35 meter mean increase in walking distance compared to the placebo group (treatment difference =  $18.96 \pm 6.47$  meters,  $p = 0.0049$ ). The reported changes in percent predicted FVC were not significantly different between the treatment groups, but more patients improved with weekly idursulfase compared to placebo. Spleen volume, liver volume, and urinary GAG levels decreased significantly more in the idursulfase groups compared to placebo ( $p < 0.0001$ ).

### Adverse Reactions

Pyrexia, headache, and arthralgia are the most common adverse effects associated with idursulfase. The most serious adverse reaction to idursulfase is an infusion-related hypersensitivity reaction. The relationship between antibody development to idursulfase and clinical efficacy is unknown. No patients discontinued therapy due to adverse effects during the phase II/III trial.

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## Coding/Billing Information

**Note:** This list of codes may not be all-inclusive.

**Covered when medically necessary:**

| HCPSC Codes | Description                  |
|-------------|------------------------------|
| J1743       | Injection, idursulfase, 1 mg |

| ICD-9-CM Diagnosis Codes | Description           |
|--------------------------|-----------------------|
| 277.5                    | Mucopolysaccharidosis |

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## References

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12. Shire Human Genetic Therapies, Inc. Elaprase® product information. Cambridge, MA: Shire Human Genetic Therapies, Inc. October 2007.

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Connecticut General Life Insurance Company has acquired the business of Great-West Healthcare from Great-West Life & Annuity Insurance Company (GWLA). Certain products continue to be provided by GWLA (Life, Accident and Disability, and Excess Loss). GWLA is not licensed to do business in New York. In New York, these products are sold by GWLA's subsidiary, First Great-West Life & Annuity Insurance Company, White Plains, N.Y.