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## Eliglustat

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### Related Coverage Resources

#### INSTRUCTIONS FOR USE

The following Coverage Policy applies to health benefit plans administered by Cigna Companies. Certain Cigna Companies and/or lines of business only provide utilization review services to clients and do not make coverage determinations. References to standard benefit plan language and coverage determinations do not apply to those clients. Coverage Policies are intended to provide guidance in interpreting certain standard benefit plans administered by Cigna Companies. Please note, the terms of a customer's particular benefit plan document [Group Service Agreement, 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 customer'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 customer's benefit plan document always supersedes 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 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. In certain markets, delegated vendor guidelines may be used to support medical necessity and other coverage determinations.

### Overview

This policy supports medical necessity review for eliglustat (**Cerdelga**<sup>®</sup>).

Receipt of sample product does not satisfy any criteria requirements for coverage.

### Medical Necessity Criteria

**Eliglustat (Cerdelga<sup>®</sup>) is considered medically necessary when the following are met:**

1. **Gaucher Disease Type 1.** Individual meets **ALL** of the following criteria (A, B, C, and D):
  - A. Individual is age 18 years or older
  - B. There is documentation of **EITHER** of the following (i or ii):
    - i. Deficiency of glucosylceramidase [also known as acid  $\beta$ -glucosidase or glucocerebrosidase] in peripheral blood leukocytes or other nucleated cells
    - ii. Confirmation of biallelic pathogenic variants in the *GBA* gene
  - C. The individual is **ONE** of the following (i, ii, or iii):

- i. CYP2D6 extensive metabolizer (EM)
  - ii. CYP2D6 intermediate metabolizer (IM)
  - iii. CYP2D6 poor metabolizer (PM)
- D. The medication is prescribed by, or in consultation with a geneticist, endocrinologist, metabolic disorder subspecialist, or a physician who specializes in the treatment of Gaucher disease or related disorders.

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

## Reauthorization Criteria

Eliglustat (Cerdelga) is considered medically necessary for continued use when initial criteria are met AND there is documentation of beneficial response.

## Authorization Duration

Initial and reauthorization approval duration: 12 months

## Conditions Not Covered

Any other use is considered experimental, investigational or unproven.

## Background

### OVERVIEW

Cerdelga, a glucosylceramide synthase inhibitor, is indicated for the long-term treatment of adults with Gaucher disease type 1 who are cytochrome P450 2D6 extensive metabolizers, intermediate metabolizers, or poor metabolizers as detected by an FDA-cleared test.<sup>1</sup>

### Disease Overview

Gaucher disease is caused by a deficiency in the lysosomal enzyme  $\beta$ -glucocerebrosidase.<sup>1</sup> This enzyme is responsible for the breakdown of glucosylceramide into glucose and ceramide. In Gaucher disease, deficiency of the enzyme  $\beta$ -glucocerebrosidase results in the accumulation of glucosylceramide substrate in the lysosomal compartment of macrophages, giving rise to foam cells or “Gaucher cells.” Cerdelga is a specific inhibitor of the enzyme glycosylceramide synthase, which is responsible for producing the substrate glucosylceramide; hence Cerdelga functions as a substrate reduction therapy.

## References

1. Cerdelga capsules [prescribing information]. Waterford, Ireland: Genzyme; July 2021.

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