Effective Date **9/15/2023**
Next Review Date **9/15/2024**
Coverage Policy Number **IP0447**

Sacrosidase

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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 sacrosidase oral solution (**Sucraid**®).

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

Medical Necessity Criteria

Sacrosidase (Sucraid) is considered medically necessary when the following are met:

1. **Congenital Sucrase-Isomaltase Deficiency.** Individual meets **ALL** of the following criteria (A, B, and C):
    A. There is documentation of symptomatic congenital sucrose-isomaltase deficiency (CSID) (for example, diarrhea, bloating, abdominal cramping)
    B. The diagnosis is established by **ONE** of the following (i or ii):

i. Endoscopic biopsy of the small bowel with disaccharidase levels consistent with CSID as evidenced by **ALL** of the following (a, b, c, and d):
    a. Decreased (usually absent) sucrase (normal reference: greater than 25 U/g protein)
    b. Decreased to normal isomaltase (palatinase) (normal reference: greater than 5 U/g protein)
    c. Decreased maltase (normal reference: greater than 100 U/g protein)
    d. Decreased to normal lactase (normal reference: greater than 15 U/g protein)
ii. Individual has a molecular genetic test demonstrating homozygous or compound heterozygous pathogenic or likely pathogenic sucrose-isomaltase (*SI*) gene variants

C. The medication is prescribed by, or in consultation with, a geneticist, gastroenterologist, a metabolic disorder sub-specialist, or a physician who specializes in the treatment of congenital diarrheal 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

Sacrosidase (Sucraid) 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: 6 months

Conditions Not Covered

Any other use is considered experimental, investigational or unproven.

Background

OVERVIEW
Sucraid, an enzyme replacement therapy, is indicated for the treatment of genetically determined sucrase deficiency, which is part of **congenital sucrase-isomaltase deficiency** (CSID).[1]

Disease Overview
CSID is an autosomal recessive intestinal disorder characterized by reduced or absent activity of the sucrase-isomaltase complex.[2,3] These enzymes are responsible for the hydrolysis of complex sugars and starches into simple sugars which are absorbed from the gastrointestinal tract. With absent or diminished enzyme activity, complex sugars and starches accumulate in the small intestine and lead to disease manifestations.[2] Symptoms include osmotic diarrhea, vomiting, bloating, abdominal pain, and steatorrhea.[2,3] Patients can occasionally experience dehydration, failure to thrive, developmental delay, and muscular hypotonia.[2] The diagnosis of CSID can be established by testing small intestine biopsy specimens for reduced or absent enzyme activity or by genetic testing to identify a mutation in the sucrase-isomaltase gene.[3,4]

Additional Clinical Information
The gold standard for diagnosis of CSID is the small bowel dissacharidase assay.[5] The sucrose hydrogen breath test is prone to error and a non-specific method to detect CSID.[5]

References

1. Sucraid® oral solution [prescribing information]. Vero Beach, FL: QOL Medical; August 2021.

2. Naim HY, Heine M, Zimmer KP. Congenital sucrose-isomaltase deficiency: Heterogeneity of inheritance, trafficking, and function of an intestinal enzyme complex. *J Pediatr Gastroenterol Nutr*. 2012;55:S13-S20.
3. Cohen SA. The clinical consequences of sucrose-isomaltase deficiency. *Mol Cell Pediatr*. 2016;3:5.
4. Gericke B, Amiri M, Scott CR, Naim HY. Molecular pathogenicity of novel sucrose-isomaltase mutations found in congenital sucrose-isomaltase deficiency patients. *Biochim Biophys Acta Mol Basis Dis*. 2017;1863:817-826.

Supplemental References

5. Treem, W. R. (1995). Congenital Sucrase-Isomaltase Deficiency. Journal of Pediatric Gastroenterology and Nutrition, 21(1), 1–14. https://doi.org/10.1097/00005176-199507000-00001