Drug Coverage Policy

Effective Date.....08/01/2024
Coverage Policy Number.....IP0447
Policy Title..... Sucraid

Enzyme Replacement Therapy – Sucraid

- Sucraid® (sacrosidase oral solution – QOL Medical)

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. Each coverage request should be reviewed on its own merits. Medical directors are expected to exercise clinical judgment and have discretion in making individual coverage determinations. 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.

Cigna Healthcare Coverage Policy

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.[3-5]

Medical Necessity Criteria

Sucraid is considered medically necessary when the following criteria are met:

FDA-Approved Indication

1. **Congenital Sucrase-Isomaltase Deficiency.** Approve for 1 year if the patient meets ALL of the following (A, B, and C):
   - **A)** The diagnosis is established by ONE of the following (i or ii):
     - **i.** Patient has endoscopic biopsy of the small bowel with disaccharidase levels consistent with congenital sucrose-isomaltase deficiency as evidenced by ALL of the following (a, b, c, and d):
       - **a)** Decreased (usually absent) sucrase level (normal reference: > 25 U/g protein); AND
       - **b)** Decreased or normal isomaltase (palatinase) level [normal reference: > 5 U/g protein]; AND
       - **c)** Decreased maltase level (normal reference: > 100 U/g protein); AND
       - **d)** Decreased or normal lactase level (normal reference: > 15 U/g protein); OR
     - **ii.** Patient has a molecular genetic test demonstrating homozygous or compound heterozygous pathogenic or likely pathogenic sucrase-isomaltase gene variant; AND
   - **B)** Prior to starting therapy with Sucraid, patient had symptomatic congenital sucrose-isomaltase deficiency (e.g., diarrhea, bloating, abdominal cramping); AND
   - **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.

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

Conditions Not Covered

Any other use is considered experimental, investigational, or unproven (criteria will be updated as new published data are available).

References

1. Sucraid® oral solution [prescribing information]. Vero Beach, FL: QOL Medical; May 2023.
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.
5. Chey, W.D. Congenital sucrase-isomaltase deficiency: What, when, and how?, *Gastroenterology Hepatology.* October 2020. Available at: https://www.gastroenterologyandhepatology.net/supplements/congenital-sucrase-isomaltase-deficiency-what-when-and-how/. Accessed: April 15, 2024.

Revision Details

Type of Revision	Summary of Changes	Date
Annual Revision	**Policy Name Change: Updated** Policy Name from "Sacrosidase" to "Enzyme Replacement Therapy – Sucraid." **Congenital Sucrase-Isomaltase Deficiency:** Disaccharidase levels consistent with congenital sucrase-isomaltase deficiency was changed from "decreased to normal" to "decreased or normal" isomaltase and lactase levels**. Updated** duration approval from 6 months to 12 months.	08/01/2024

The policy effective date is in force until updated or retired.