MEDICAL MUTUAL®

Drug Policy

Policy:	Sucraid (sacrosidase)	Annual Review Date: 08/22/2024 Last Revised Date: 08/22/2024

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).

POLICY STATEMENT

This policy involves the use of Sucraid. Prior authorization is recommended for pharmacy benefit coverage of Sucraid. Approval is recommended for those who meet the conditions of coverage in the **Criteria and Initial/Extended Approval** for the diagnosis provided. **Conditions Not Recommended for Approval** are listed following the recommended authorization criteria. Requests for uses not listed in this policy will be reviewed for evidence of efficacy and for medical necessity on a case-by-case basis.

Because of the specialized skills required for evaluation and diagnosis of patients treated with Sucraid as well as the monitoring required for adverse events and long-term efficacy, initial approval requires Sucraid be prescribed by or in consultation with a physician who specializes in the condition being treated. All approvals for initial therapy are provided for the initial approval duration noted below; if reauthorization is allowed, a response to therapy is required for continuation of therapy unless otherwise noted below.

RECOMMENDED AUTHORIZATION CRITERIA

Coverage of Sucraid is recommended in those who meet the following criteria:

1. Congenital Sucrase-Isomaltase Deficiency

Criteria. Approve for 1 year if the following criteria are met (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 (normal reference: > 25 U/g protein); AND
  - **b)** Decreased or normal isomaltase (palatinase) [normal reference: > 5 U/g protein]; AND
  - **c)** Decreased maltase (normal reference: > 100 U/g protein); AND
  - **d)** Decreased or normal lactase (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) Sucraid 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.

CONDITIONS NOT RECOMMENDED FOR APPROVAL

Sucraid has not been shown to be effective, or there are limited or preliminary data or potential safety concerns that are not supportive of general approval for the following conditions. (Note: This is not an exhaustive list of Conditions Not Recommended for Approval).

1. Coverage is not recommended for circumstances not listed in the Recommended Authorization Criteria. Criteria will be updated as new published data are available.

Documentation Requirements:

The Company reserves the right to request additional documentation as part of its coverage determination process. The Company may deny reimbursement when it has determined that the drug provided or services performed were not medically necessary, investigational or experimental, not within the scope of benefits afforded to the member and/or a pattern of billing or other practice has been found to be either inappropriate or excessive. Additional documentation supporting medical necessity for the services provided must be made available upon request to the Company. Documentation requested may include patient records, test results and/or credentials of the provider ordering or performing a service. The Company also reserves the right to modify, revise, change, apply and interpret this policy at its sole discretion, and the exercise of this discretion shall be final and binding.

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.