

## PRIOR AUTHORIZATION POLICY

**POLICY:** Enzyme Replacement Therapy – Sucraid Prior Authorization Policy

- Sucraid® (sacrosidase oral solution – QOL Medical)

**REVIEW DATE:** 04/06/2022; selected revision 08/31/2022

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### 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)**.<sup>1</sup>

### Disease Overview

CSID is an autosomal recessive intestinal disorder characterized by reduced or absent activity of the sucrase-isomaltase complex.<sup>2,3</sup> 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.<sup>2</sup> Symptoms include osmotic diarrhea, vomiting, bloating, abdominal pain, and steatorrhea.<sup>2,3</sup> Patients can occasionally experience dehydration, failure to thrive, developmental delay, and muscular hypotonia.<sup>2</sup> 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.<sup>3,4</sup>

### POLICY STATEMENT

Prior Authorization is recommended for prescription benefit coverage of Sucraid. All approvals are provided for the duration noted below. 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, approval requires Sucraid to be prescribed by or in consultation with a physician who specializes in the condition being treated.

**Automation:** None.

### RECOMMENDED AUTHORIZATION CRITERIA

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

#### FDA-Approved Indication

**1. Congenital Sucrase-Isomaltase Deficiency.** Approve for 1 year if the patient meets the following criteria (A and B):

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 sucrase-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)
  - b) Decreased to normal isomaltase (palatinase) [normal reference: > 5 U/g protein]
  - c) Decreased maltase (normal reference: > 100 U/g protein)
  - d) Decreased to normal lactase (normal reference: > 15 U/g protein); OR

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- ii. Patient has a molecular genetic test demonstrating homozygous or compound heterozygous pathogenic or likely pathogenic sucrose-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**

Coverage of Sucraid is not recommended in the following situations:

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

#### **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.