

Pharmacy Policy Bulletin: J-0219 Sucraid (sacrosidase) – Commercial and Healthcare Reform	
Number: J-0219	Category: Prior Authorization
Line(s) of Business: ☒ Commercial ☒ Healthcare Reform ☐ Medicare	Benefit(s): Commercial: Prior Authorization (1.) 1. Miscellaneous Specialty Drugs Oral = Yes w/ Prior Authorization Healthcare Reform: Not Applicable
Region(s): ☒ All ☐ Delaware ☐ New York ☐ Pennsylvania ☐ West Virginia	Additional Restriction(s): None
Version: J-0219-007	Original Date: 05/01/2019
Effective Date: 07/18/2025	Review Date: 06/25/2025

Drugs Product(s):	Sucraid (sacrosidase)
FDA-Approved Indication(s):	Treatment of sucrase deficiency, which is part of congenital sucrase-isomaltase deficiency (CSID), in adult and pediatric patients 5 months of age and older.

Background:	- Sucraid replaces deficient sucrase, which is responsible for the metabolism of sucrose. Sucrase hydrolyzes sucrose into glucose and fructose. Unhydrolyzed sucrose is not absorbed, causing water retention which may cause loose stools. The unabsorbed sucrose can be fermented by intestinal bacteria resulting in excessive gas, bloating, cramps, nausea, and vomiting. The only previous treatment was diet restriction of sucrose. - CSID is a genetic condition where the body lacks the enzymes needed to break down and absorb sucrose (table sugar) and other sugars from starch. It is caused by variants in the sucrase-isomaltase (SI) gene. - A small bowel biopsy provides a definitive diagnosis of CSID. The sucrose breath hydrogen test offers a noninvasive alternative; however, it is an indirect test and not specific for CSID and may produce false-positive or false-negative results. Additionally, it can provoke gastrointestinal symptoms in patients due to the amount of sucrose patients must ingest. Sucrase deficiency is evidenced by an increase in breath hydrogen of > 10 ppm when challenged with sucrose after fasting. The ¹³C-sucrose breath test offers more specificity than the sucrose breath hydrogen test; however, it may also produce false-positive or false-negative results and requires further validation. A normal level of sucrose digestion is indicated by a 90-minute breath sample reading of exhaled ¹³CO₂ ≥ 5.10% for women and ≥ 3.91% for men. Other generally accepted tests that support diagnosis of CSID include an acidic stool pH (caused by fermentation of carbohydrates due to malabsorption) and a negative lactose breath test. - Genetic testing of the SI gene may also be utilized to identify homozygous and compound heterozygous mutations responsible for CSID.
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	• Due to the difficulties of diagnosing CSID, it may be appropriate to provide a short therapeutic trial (for example, one week) to assess response in patients suspected of having CSID. • Prescribing Considerations: ○ Sucraid is contraindicated in patients with hypersensitivity to yeast, yeast products, glycerin (glycerol), or papain. ○ Sucraid is not for use in isomaltase deficiency because it does not provide specific replacement therapy for deficient isomaltase. ○ Restricting starch in the diet may still be necessary to reduce symptoms. ○ The effects of Sucraid have not been evaluated in patients with secondary (acquired) sucrase deficiency.
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Approval Criteria

I. Initial Authorization

When a benefit, coverage of Sucraid may be approved when all of the following criteria are met **(A., B., and C.)**:

- A. The member is 5 months of age or older.
- B. The member has a diagnosis of congenital sucrase-isomaltase deficiency (CSID) (ICD-10: E74.31).
- C. There is clinical documentation supporting the CSID diagnosis with one (1) of the following **(1., 2., or 3.)**:
 1. Small bowel biopsy with a disaccharidase assay demonstrating all of the following **(a. through d.)**:
 - a. Absent or reduced sucrase activity
 - b. Reduced or normal isomaltase activity
 - c. Reduced maltase activity
 - d. Reduced or normal lactase activity
 2. Genetic testing indicates a mutation in the sucrase-isomaltase (SI) gene.
 3. The member meets all of the following criteria **(a., b., and c.)**:
 - a. Stool pH < 6
 - b. Negative lactose breath test
 - c. Sucrase deficiency evidenced by one (1) of the following **(i. or ii.)**:
 - i. Sucrose breath hydrogen test (an increase in breath hydrogen of > 10 ppm when challenged with sucrose after fasting)
 - ii. ¹³Carbon-sucrose breath test (CO₂ exhalation over 90 minutes is below the 10th percentile, which is < 3.91% for men and < 5.10% for women)

II. Reauthorization

When a benefit, reauthorization of Sucraid may be approved when the following criterion is met **(A.)**:

- A. The prescriber attests that the member has experienced positive clinical response to therapy.

- III. An exception to some or all of the criteria above may be granted for select members and/or circumstances based on state and/or federal regulations.

Limitations of Coverage

- I. Coverage of drug(s) addressed in this policy for disease states outside of the FDA-approved indications should be denied based on the lack of clinical data to support effectiveness and safety in other conditions unless otherwise noted in the approval criteria.
- II. For Commercial or HCR members with a closed formulary, a non-formulary product will only be approved if the member meets the criteria for a formulary exception in addition to the criteria outlined within this policy.

Authorization Duration

- Commercial and HCR Plans: If approved, up to a 12 month authorization may be granted.

Automatic Approval Criteria

None

Refer to [J-0664](#) for previous Healthcare Reform versions

References:

1. Sucraid [package insert]. Vero Beach, FL: QOL Medical, LLC; August 2024.
2. International Foundation for Functional Gastrointestinal Disorders. Congenital sucrose-isomaltase deficiency (CSID). Available at: <https://iffgd.org/other-disorders/congenital-sucrase-isomaltase-deficiency-csid.html?showall=&start=1>. Accessed April 11, 2025.
3. Puntis JWL, Zamvar V. Congenital sucrose-isomaltase deficiency: diagnostic challenges and response to enzyme replacement therapy. *Arch Dis Child*. 2015;100:869-871.
4. Treem WR. Clinical aspects and treatment of congenital sucrose-isomaltase deficiency. *J Pediatr Gastroenterol Nutr*. 2012 Nov;55 Suppl 2:S7-13.
5. CSIDCares. How Is CSID Diagnosed? Available at: <https://www.csidcares.org/about/how-is-csid-diagnosed/>. Accessed May 4, 2022.
6. CSIDCares. How Do I Interpret Disaccharidase Biopsy Results? Available at: <https://www.csidcares.org/about/biopsy/>. Accessed May 4, 2022.
7. CSID Disease Information. Diagnosing CSID. Available at: <https://www.csiddiseaseinfo.com/diagnosing-congenital-sucrase-isomaltase-deficiency/>. Accessed April 8, 2024.
8. Gastroenterology & Hepatology. Congenital Sucrase-Isomaltase Deficiency: What, When, and How? Available at: <https://www.gastroenterologyandhepatology.net/supplements/congenital-sucrase-isomaltase-deficiency-what-when-and-how/>. Accessed April 11, 2025.
9. Frissora CL, Rao SS. Sucrose intolerance in adults with common functional gastrointestinal symptoms. *Proc (Bayl Univ Med Cent)*. 2022;35(6):790-793.
10. Treem WR. Clinical aspects and treatment of congenital sucrase-isomaltase deficiency. *J Ped Gastro Nutr*. 2012;55 Suppl 2:S7-13.

Pharmacy policies do not constitute medical advice, nor are they intended to govern physicians' prescribing or the practice of medicine. They are intended to reflect the plan's coverage and reimbursement guidelines. Coverage may vary for individual members, based on the terms of the benefit contract.

The plan retains the right to review and update its pharmacy policy at its sole discretion. These guidelines are the proprietary information of the plan. Any sale, copying or dissemination of the pharmacy policies is prohibited; however, limited copying of pharmacy policies is permitted for individual use.