BlueCross BlueShield Arizona

An Independent Licensee of the Blue Cross Blue Shield Association

PHARMACY COVERAGE GUIDELINE

SUCRAID® (sacrosidase) Generic Equivalent (if available)

This Pharmacy Coverage Guideline (PCG):
- Provides information about the reasons, basis, and information sources we use for coverage decisions
- Is not an opinion that a drug (collectively "Service") is clinically appropriate or inappropriate for a patient
- Is not a substitute for a provider's judgment (Provider and patient are responsible for all decisions about appropriateness of care)
- Is subject to all provisions e.g. (benefit coverage, limits, and exclusions) in the member's benefit plan; and
- Is subject to change as new information becomes available.

Scope
- This PCG applies to Commercial and Marketplace plans
- This PCG does not apply to the Federal Employee Program, Medicare Advantage, Medicaid or members of out-of-state Blue Cross and/or Blue Shield Plans

Instructions & Guidance
- To determine whether a member is eligible for the Service, read the entire PCG.
- This PCG is used for FDA approved indications including, but not limited to, a diagnosis and/or treatment with dosing, frequency, and duration.
- Use of a drug outside the FDA approved guidelines, refer to the appropriate Off-Label Use policy.
- The "Criteria" section outlines the factors and information we use to decide if the Service is medically necessary as defined in the Member's benefit plan.
- The "Description" section describes the Service.
- The "Definition" section defines certain words, terms or items within the policy and may include tables and charts.
- The "Resources" section lists the information and materials we considered in developing this PCG
- **We do not accept patient use of samples as evidence of an initial course of treatment, justification for continuation of therapy, or evidence of adequate trial and failure.**
- Information about medications that require prior authorization is available at www.azblue.com/pharmacy. You must fully complete the request form and provide chart notes, lab workup and any other supporting documentation. The prescribing provider must sign the form. Fax the form to BCBSAZ Pharmacy Management at (602) 864-3126 or email it to Pharmacyprecert@azblue.com.

Criteria:

➢ **Criteria for initial therapy:** Sucraid (sacrosidase) and/or generic equivalent (if available) is considered ***medically necessary*** and will be approved when **ALL** of the following criteria are met:

1. Prescriber is a physician specializing in the patient's diagnosis or is in consultation with a Pediatric Gastroenterologist or Specialist in Genetic Disorders

2. Individual is 5 months of age or older

3. Individual has a confirmed diagnosis of genetically determined sucrase deficiency as part of congenital sucrase-isomaltase deficiency (CSID) in an individual having osmotic (watery) fermentative diarrhea, abdominal pain, cramps, bloating, and gas

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4. Congenital sucrase-isomaltase deficiency was confirmed by **ONE** of the following **baseline tests**:
    a. Genetic test that reveals pathogenic or likely pathogenic mutation in the sucrose-isomaltase (SI) gene on chromosome 3 (3q25-q26)
    b. Measurement of intestinal disaccharides after small bowel biopsy consistent for congenital sucrose-isomaltase deficiency (see Definitions section for required results)

5. **If available**: Individual has failure after adequate trial, contraindication per FDA label, intolerance, or is not a candidate for a **generic equivalent** [Note: Failure, contraindication or intolerance to the generic should be reported to the FDA] (see Definitions section)

6. Individual has been adherent with a sucrose free, low starch diet

7. Individual does not have secondary (acquired) disaccharidase deficiencies (see Definitions section)

8. There are **NO** FDA label contraindications such as hypersensitivity to yeast, yeast products, glycerin (glycerol), or papain

Initial approval duration: 1 box (containing 2 bottles) per month x 3 months

- **Criteria for continuation of coverage (renewal request):** Sucraid (sacrosidase) and/or generic equivalent (if available) is considered ***medically necessary*** and will be approved when **ALL** of the following criteria are met **(samples are not considered for continuation of therapy)**:

1. Individual continues to be seen by a physician specializing in the patient's diagnosis or is in consultation with a Pediatric Gastroenterologist or Specialist in Genetic Disorders

2. **Medical record documentation of a routine re-evaluation of continued need for the medication, as affected individuals tend to experience spontaneous improvement of their symptoms with age, as colonic bacteria become able to metabolize non-absorbed carbohydrates into organic acids (lactic acid and short chain fatty acids), most of which are then absorbed** (*Documentation of re-evaluation must be sent with renewal requests*)

3. Individual has documentation of positive clinical response to therapy defined as at least a 50% improvement in **ALL** of the following:
    a. Symptoms of abdominal pain, cramps, bloating, gas
    b. Number of stools per day
    c. Watery, loose stool consistency
    d. Number of symptomatic days

4. Individual has been adherent with the medication

5. **If available**: Individual has failure after adequate trial, contraindication per FDA label, intolerance, or is not a candidate for a **generic equivalent** [Note: Failure, contraindication or intolerance to the generic should be reported to the FDA] (see Definitions section)

6. Individual has not developed any contraindications or other significant adverse drug effects that may exclude continued use such as:

a. Contraindications as listed in the criteria for initial therapy section
b. Severe wheezing

7. Individual continues to be adherent with a sucrose free, low starch diet

8. Individual does not have secondary (acquired) disaccharidase deficiencies (see Definitions section)

Renewal duration: 1 box (containing 2 bottles) per month x 6 months

- Criteria for a request for non-FDA use or indication, treatment with dosing, frequency, or duration outside the FDA-approved dosing, frequency, and duration, refer to one of the following Pharmacy Coverage Guideline:

1. **Off-Label Use of a Non-cancer Medications**

2. **Off-Label Use of a Cancer Medications**

Description:

Sucraid (sacrosidase) chemically is beta D-fructofuranoside fructohydrolase. Sacrosidase, derived from Baker's yeast (*Saccharomyces cerevisiae*), hydrolyzes sucrose. Sucraid (sacrosidase) is indicated as oral replacement therapy of genetically determined sucrase deficiency, which is part of congenital sucrase-isomaltase deficiency (CSID). The effects of sacrosidase have not been evaluated in patients with secondary (acquired) disaccharidase deficiencies. Sucraid (sacrosidase) does not contain isomaltase.

CSID is a carbohydrate intolerance disorder characterized by malabsorption of oligosaccharides and disaccharides. It is also known as disaccharide intolerance I, congenital sucrase-isomaltose malabsorption, congenital sucrose-isomaltose malabsorption, SI deficiency, sucrose-isomaltase deficiency, sucrase-isomaltase deficiency, and congenital sucrose intolerance. It is an autosomal recessive disorder. Onset usually occurs during infancy after weaning from breast milk or lactose-only formula onto foods containing sucrose or starch. Clinical manifestations include osmotic-fermentative diarrhea, abdominal distension and discomfort, flatulence and vomiting. Severe symptoms may lead to failure to thrive, dehydration and malnutrition. The gastrointestinal symptoms associated with CSID are nonspecific; the diagnosis is often delayed, and patients can be misdiagnosed with irritable bowel syndrome, cystic fibrosis, celiac disease or other causes of chronic diarrhea.

With CSID there is complete or almost complete lack of endogenous sucrase activity, marked reduction in isomaltase activity, moderate decrease in maltase activity, and normal lactase levels. Sucrase is naturally produced in the brush border of the small intestine, primarily the distal duodenum and jejunum. Sucrase hydrolyzes sucrose (a disaccharide) into its component monosaccharides, glucose and fructose. Isomaltase breaks down disaccharides from starch into simple sugars.

In the absence of endogenous human sucrase, sucrose is not metabolized. Unhydrolyzed sucrose and starch are not absorbed from the intestine and their presence in the intestinal lumen leads to osmotic retention of water resulting in loose watery stools. Unabsorbed sucrose in the colon is fermented by bacterial flora to produce increased amounts of hydrogen, methane, and water resulting in excessive gas, bloating, abdominal cramps, nausea, and vomiting.

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CSID is inherited as an autosomal recessive genetic trait. The faulty gene has been identified to chromosome 3 (3q25-q26). The *SI* gene provides instructions for producing the enzyme sucrase-isomaltase. Mutations that cause this condition alter the structure, disrupt the production, or impair the function of sucrase-isomaltase. More than 25 mutations within the human sucrase gene are responsible for these CSID phenotypes. Sucrase-isomaltase variants can occur on either sucrase or isomaltase subunits, resulting in varied effects on sucrase-isomaltase enzyme activity.

CSID is difficult to diagnose. Approximately 4-10% of pediatric patients with chronic diarrhea of unknown origin have CSID. Several tests can be used to diagnose CSID tests but when used alone may be inaccurate.

Stool pH < 6 is not always a reliable screening test for the diagnosis of sugar malabsorption as stools with a pH < 6 may not have sugar detected and stools with a pH > 6 may have substantial amount of sugar present.

A hydrogen breath test or a sucrose hydrogen breath test showing an increase in breath hydrogen after a sucrose challenge is not specific for CSID; the test may have a high incidence of false-positive results due to villous injury, dumping syndrome, and the presence of small bowel bacterial overgrowth (SIBO) where unabsorbed sugar is converted to hydrogen gas by colonic bacteria.

Use of a differential urinary disaccharide ratio of sucrose to lactulose relies on obtaining an accurate 10-hour urine collection.

A sucrose breath test for screening and confirmation of CSID using a novel non-invasive ^{13}C-sucrose labeled substrate has been developed and validated and is said to be accurate and specific for CSID, however getting breath samples may be difficult in small children. ^{13}C-sucrose breath testing with infrared spectrophotometry, requires 2 breath tests (one with labeled ^{13}C-sucrose and another with labeled ^{13}C-glucose) $^{13}CO_2$-enriched breath samples are collected for each. The results are expressed as a coefficient of glucose oxidation (CGO) and using a cutoff of < 79% CGO, it yields 100% sensitivity and specificity for CSID. But secondary sucrase deficiency cannot be excluded without clinical evaluations and biopsy.

The definitive test for diagnosis of CSID is the measurement of intestinal disaccharidases following small bowel biopsy. The small bowel biopsy should show normal villous architecture. The biopsy specimens should be assessed for lactase, sucrase, isomaltase (palatinase) & maltase activities. The activity assay should show complete or almost complete lack of endogenous sucrase activity, variable isomaltase activity that ranges from no activity to full activity, decrease in maltase activity, and normal lactase levels (within 1 standard deviation of the mean) or a sucrase:lactase ratio of < 1 in the setting of reduced lactase. In addition, there should be a normal lactose breath hydrogen test to rule out a diagnose lactose intolerance. It is important to rule out secondary (or acquired) forms of disaccharidase deficiencies as the effects of Sucraid (sacrosidase) have not been evaluated in these patients with secondary (acquired) disaccharidase deficiencies.

Treatment of CSID involves following a strict sucrose- and starch-restricted diet, and an oral solution of Baker's yeast-derived enzyme replacement.

Definitions:

U.S. Food and Drug Administration (FDA) MedWatch Forms for FDA Safety Reporting
MedWatch Forms for FDA Safety Reporting | FDA

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Tests that aid in the diagnosis of CSID:

- Small bowel biopsy – considered the gold standard
- Genetic tests – buccal swab, saliva, or blood
- Breath tests – hydrogen-methane and ^{13}C-sucrose
- Sucrose challenge – simple test but not validated

Small bowel biopsy shows ALL of the following:

- Normal villous architecture
- Absent or markedly reduced sucrase activity
- Isomaltase (palatinase) activity ranging from none to full activity
- Normal or reduced maltase activity
- Normal lactase activity or a sucrase:lactase ratio of < 1 if have reduced lactase

Potential causes of secondary or acquired sucrose-isomaltase deficiency or malabsorption:

Villous atrophy or alteration	Celiac disease Non-tropical sprue Chemotherapy and radiation therapy Crohn's disease Allergic enteropathy Immunodeficiency Malnutrition
Infection	Acute gastroenteritis Giardiasis Tropical sprue HIV enteropathy SIBO
Rapid transit	Rapid gastric emptying Chronic non-specific diarrhea Dumping syndrome Ulcerative, microscopic, and lymphocytic colitis

Available treatment options for disaccharidase deficient patients

Name	Component	FDA approval
Baker's Yeast	7,500 IU sucrase	No
Sucrase	8,500 IU sacrosidase	Yes

Resources:

Sucraid (sacrosidase) oral solution product information, revised by QOL Medical, LLC 08-2024. Available at DailyMed http://dailymed.nlm.nih.gov. Accessed June 27, 2025.

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