Choose Foods That Are Low in Sucrose and Starch

**Dairy**
Cow’s milk
Cream cheese
Half and half
Hard cheeses (cheddar, colby, mozzarella, swiss, parmesan, provolone)
Plain cottage cheese
Plain yogurt sweetened with fructose or dextrose
Ricotta cheese
Sour cream
Whipping cream

**Vegetables**
Alfalfa sprouts
Artichoke
Asparagus
Bamboo shoots
Bok choy
Broccoli
Brussels sprouts
Cabbage
Cauliflower
Celeriac
Cucumber
Eggplant
Green beans
Greens (collards, kale, mustard, turnip, and chard)
Lettuce (arugula, endive, iceberg, romaine)
Mung bean sprouts
Mushrooms
Peppers (red, green, and yellow)
Radishes
Rutabaga
Spaghetti squash
Spinach
Tomatoes
Turnips
Yellow squash
Zucchini

**Protein**
Beef
Chicken
Eggs
Fish
Lamb
Pork
Tofu
Turkey

**Fats**
Any vegetable oils
Butter

**Sweeteners**
Granulated fructose
Granulated dextrose

**AVOID Foods High in Starch**
Baked goods
Beans, peas, & lentils
Breads
Cereals
Corn
Crackers
Gluten-free starches & grains
Grains (wheat, oats, rice)
Pasta
Potatoes

**Note:** These diet recommendations are for general guidance only. Every case of Genetic Sucrase-Isomaltase Deficiency is unique. Food consistency should be age appropriate. All diet plans should be developed in conjunction with a physician and Registered Dietitian (RD).

Always read nutrition labels, as ingredients may change.

**Fruits**
Avocado
Blackberries
Blueberries
Cherries
Coconut (fresh or dried, unsweetened)
Cranberries
Currants
Figs
Grapes
Kiwi
Lemons
Limes
Loganberries
Olives
Papaya
Pears
Pomegranate
Prunes
Raspberries
Rhubarb
Strawberries

**Formula**
Seek advice from a physician or dietitian.
Breast milk is the best choice for infants.

**ADDITIONAL IMPORTANT SAFETY INFORMATION**
- Sucraid® may cause a serious allergic reaction. Patients should stop taking Sucraid® and get emergency help immediately if any of the following side effects occur: difficulty breathing, wheezing, or swelling of the face. Care should be taken when administering initial doses of Sucraid® to observe any signs of acute hypersensitivity reaction.
- Adverse reactions as a result of taking Sucraid® may include worse abdominal pain, vomiting, nausea, diarrhea, constipation, difficulty sleeping, headache, nervousness, and dehydration.
- Before prescribing Sucraid® to diabetic patients the physician should consider that Sucraid® will enable sucrose hydrolysis and the absorption of those hydrolysis products, glucose and fructose.
- The effects of Sucraid® have not been evaluated in patients with secondary (acquired) disaccharidase deficiency.

Please see additional Important Safety Information on back page and in enclosed Full Prescribing Information. You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.FDA.gov/medwatch or call 1-800-FDA-1088.
(Child Diet Guide)

Guidelines for Children with Genetic Sucrase-Isomaltase Deficiency (GSID)
See Sucraid Important Safety Information below and on the back page

Starting Sucraid® (sacrosidase) Oral Solution Therapy with a Low-Sucrose, Low-Starch Diet

After a diagnosis of Genetic Sucrase-Isomaltase Deficiency has been made:

**Weeks 1 and 2:**
Avoid sucrose and starch for two weeks only
- Start Sucraid® therapy as prescribed.
- It is recommended that both sucrose and starch be restricted initially. If disaccharidase levels are available, further guidance for sucrose and starch restrictions may be provided.
- The reverse side of this card lists foods that are low in sucrose and starch. Choose a variety of foods from each of these food groups every day.
- Foods high in starch should be avoided. Examples of high-starch foods are listed on the reverse side of this card.

**Week 3:**
Begin reintroducing sucrose and then starch back into the diet
- Step one: Start by gradually adding more sucrose back into the diet. This step may take many weeks. Once the sucrose tolerance level is determined, move to step two.
- Step two: Begin adding foods higher in starch back into the diet.
- Follow these tips when reintroducing foods higher in sucrose and starch:
  - Continue taking Sucraid® as prescribed with meals and snacks.
  - Add only one new food every three to five days, gradually increasing the amount of the new food each day before adding the next new food.
  - Refer to the CSIDcares.org website for additional help.
  - Keep detailed records of food intake, noting the new foods added and any symptoms that may result.
  - If symptoms are noted, remove the last new food added and wait three days before trying another new food.
  - Read food labels carefully and note portion sizes: note the portion size, the amount of carbohydrates, fiber, and sugars, and the ingredient list.
- Failure to add foods back to the diet could lead to nutrient deficiencies. A Registered Dietitian can ensure the diet meets overall nutrient needs and can recommend appropriate vitamin and mineral supplements, if needed.

For further diet information, call the Registered Dietitian at QOL Medical at 1-800-705-1962. The RD cannot provide medical advice but can answer questions about diet and the safe and effective use of Sucraid®.

**Key Points:**
- Weeks 1 and 2: Follow a low-sucrose, low-starch diet.
- Week 3: Reintroduce sucrose-containing foods and then foods higher in starch.
- Consult with a Registered Dietitian (RD) for diet assessment, education, and follow up.

**Note:** These diet recommendations are for general guidance only. Every case of Genetic Sucrase-Isomaltase Deficiency is unique. Food consistency should be age appropriate. All diet plans should be developed in conjunction with a physician and Registered Dietitian (RD).

**INDICATION**
Sucraid® (sacrosidase) Oral Solution is an enzyme replacement therapy for the treatment of genetically determined sucrase deficiency, which is part of congenital sucrase-isomaltase deficiency (CSID).

**IMPORTANT SAFETY INFORMATION FOR SUCRAID® (SACROSIDASE) ORAL SOLUTION**
- Although Sucraid® provides replacement therapy for the deficient sucrase, it does not provide specific replacement therapy for the deficient isomaltase.
- Do not use Sucraid® (sacrosidase) Oral Solution with patients known to be hypersensitive to yeast, yeast products, papain, or glycerin (glycerol).

**WWW.SUCRAID.NET**

Please see additional Important Safety Information on last page and in Full Prescribing Information on adjacent pages.

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**Prescribing Information**

**Sucraid** (sacrosidase) Oral Solution:

**DESCRIPTION**

Sucraid (sacrosidase) Oral Solution is an enzyme replacement therapy for the treatment of genetically determined sucrose deficiency, which is part of congenital sucrase-isomaltase deficiency (CSID).

**CHEMISTRY**

Sucraid is a pale yellow to colorless solution of a pleasant sweet taste. Each milliliter (mL) of Sucraid contains 8,500 International Units (I.U.) of the primary enzyme, the active ingredient. The chemical name of this enzyme is β-D-fructofuranoside fructohydrolase. The enzyme is derived from baker's yeast (Saccharomyces cerevisiae).

It has been reported that the primary amino acid structure of the protein consists of 513 amino acids with an apparent molecular weight of 100,000 g/mole for the glycylated monomer (range 65,000-116,000 g/mole). Reports also suggest that the protein exists in solution as a monomer, dimer, tetramer, and octamer ranging from 100,000 g/mole to 800,000 g/mole. It has an isoelectric point (pI) of 4.5.

Sucraid may contain small amounts of papain. Papain is known to cause allergic reactions in some people. Papain is a protein-cleaving enzyme that is introduced in the manufacturing process to digest the cell wall of the yeast and may not be completely removed during subsequent process steps.

Sucraid contains sacroside in a vehicle comprised of glycerol (50% w/v), water, and citric acid to maintain the pH at 4.0 to 4.7. (Glycerol (glycerin) in the amount consumed in the recommended doses of Sucraid has no expected toxicity.

This enzyme preparation is fully soluble with water, milk, and infant formula.

**CLINICAL PHARMACOLOGY**

Congenital sucrase-isomaltase deficiency (CSID) is a chronic, autosomal recessive, inherited, phenotypically heterogeneous disease with very variable enzyme activity. CSID is usually characterized by a complete or almost complete lack of endogenous sucrase activity. A very marked reduction in isomaltase activity, a moderate decrease in maltase activity, and normal lactase levels.

Sucrase is naturally produced in the brush border of the small intestine, primarily the duodenum and jejunum. Sucrose hydrolyzes the disaccharide to its component monosaccharides, glucose, and fructose. Isomaltase breaks down disaccharides from starch into simple sugars. Sucraid does not contain isomaltase.

In the absence of endogenous human sucrase, as in CSID, sucrose is not metabolized. Unhydrolyzed sucrose and starch are not absorbed by the small intestine. Malabsorption of disaccharides may result in malnutrition. Undiagnosed/untreated CSID patients often fail to thrive and fall behind in their expected growth and development curves. Previously, difficulties in diagnosing CSID, it may be warranted to conduct a short therapeutic trial (e.g., one week) to determine if the patient has sucrase-isomaltase deficiency. In both trials, however, patients showed a marked decrease in breath hydrogen output when they received sacrosidase in comparison to placebo.

**INDICATIONS AND USAGE**

Sucraid (sacrosidase) Oral Solution is indicated as oral replacement therapy of the genetically determined sucrose deficiency, which is part of congenital sucrase-isomaltase deficiency (CSID).

**CONTRAINdications**

Patients known to be hypersensitive to yeast, yeast products, glycerol (glycerin), or papain.

**WARNINGS**

Severe wheezing, 90 minutes after a second dose of sacrosidase, necessitated admission into the ICU for a 4-day-old baby. The wheezing was probably caused by sacrosidase. He had asthma and was being treated with steroids. A skin test for sacrosidase was positive.

Other serious events have not been linked to Sucraid.

**PRECAUTIONS**

Care should be taken to administer initial doses of Sacraad near (within a few minutes of travel) a facility where acute hypersensitivity reactions may be treated. Allergic reactions in patients with severe sacrosidase reactions may be seen in patients with Sacraad, but the reactions are usually milder.

Alternatively, the patient may be tested for hypersensitivity to Sacraad through skin abrasion testing. Should symptoms of hypersensitivity disappear, discontinue medication and initiate symptomatic and supportive therapy.

Skin testing as a rechallenge has been used to verify hypersensitivity in one asthmatic child who displayed wheezing after oral sacrosidase.

**GENERAL**

Although Sucraid provides replacement therapy for the deficient sucrase, it does not provide specific replacement therapy for the deficient isomaltase. Therefore, restricting starch in the diet may still be necessary to reduce symptoms as much as possible. The need for dietary starch restriction for patients using Sucraid should be evaluated in each patient.

It may sometimes be clinically inappropriate, difficult, or inconvenient to perform a small bowel biopsy or biopsy the hydrogen test to make a definitive diagnosis of CSID. If the diagnosis is in doubt, it may be warranted to conduct a short therapeutic trial (e.g., one week) with Sucraid to assess response in a patient suspected of sucrose deficiency.

The effects of Sucraid have not been evaluated in patients with secondary acquired disaccharidase deficiencies.

**INFORMATION FOR PATIENTS**

See Patient Package Insert. Patients should be instructed to discard bottles of Sucraid 4 weeks after opening due to the potential for bacterial growth. For the same reason, patients should be advised to rinse the measuring scoop with water after each use. Sucraid is fully soluble with water, milk, and infant formula, but it is important to note that this product is sensitive to heat. Sucraid should not be reconstituted or consumed with fruit juice, since its acidity may reduce the enzyme activity.

**USE IN DIABETICS**

Sucraad will enable the products of sucrose hydrolysis, glucose and fructose, to be absorbed. This fact must be carefully considered in planning the diet of diabetic CSID patients using Sucraid.

**LABORATORY TESTS**

The definitive test for diagnosis of CSID is the measurement of intestinal disaccharidases following small bowel biopsy.

**DRUG INTERACTIONS**

Neither drug-drug nor drug-food interactions are expected or have been reported with the use of Sucraid. However, Sucraid should not be reconstituted or consumed with fruit juice, since its acidity may reduce the enzyme activity.

**CARCINOGENESIS, MUTAGENESIS, IMPAIRMENT OF FERTILITY**

Long- term studies in animals with Sucraid have not been performed to evaluate the carcinogenic potential. Studies to evaluate the effect of Sucraid on fertility or its mutagenic potential have not been performed.

**PREGNANCY**

Teratogenic effects. Pregnancy Category C. Animal reproduction studies have not been conducted with Sucraid. It is not known whether Sucraid can cause fetal harm when administered to a pregnant woman or to affect reproductive capacity. Sucraid should be given to a pregnant woman only if clearly needed.

**NURSING MOTHERS**

The Sucraid enzyme is broken down in the stomach and intestines, and the component amino acids and peptides are then absorbed as nutrients.

**PEDIATRIC USE**

Sucraid has been used in patients as young as 5 months of age. Evidence in one controlled trial in primarily pediactric patients shows that Sucraid is safe and effective for the treatment of the genetically acquired sucrose deficiency, which is part of CSID.

**ADVERSE REACTIONS**

Adverse experiences with Sucraid in clinical trials were generally mild and were frequently associated with the underlying disease.

In clinical studies of up to 54 months duration, physicians treated a total of 52 patients with Sucraid. The adverse experiences and respective number of patients reporting each event (in parentheses) were as follows: abdominal pain (4), vomiting (3), nausea (2), diarrhea (2), constipation (2), insomnia (1), headache (1), rashes (1), and dermatitis (1).

Note: Diarrhea and abdominal pain can be a part of the clinical presentation of the genetically determined sucrose deficiency, which is part of congenital sucrase-isomaltase deficiency (CSID).

One asthmatic child experienced a serious hypersensitivity reaction during a challenge probably related to Sucraid (see Warnings). The event resulted in withdrawal of the patient from the trial but resolved with no sequelae.

**OVERDOSAGE**

Overdosage with Sucraid has not been reported.

**DOSAGE AND ADMINISTRATION**

The recommended dosage is 1 mL or 2 mL (9,500 to 17,000 I.U.) or 1 or 2 full measuring scoops (each full measuring scoop equals 1 mL). 28 drops from the Sucraid container to equal 1 mL taken every meal or snack with Sucraid. Sucraid should not be reconstituted or consumed with fruit juice since its acidity may reduce the enzyme activity.

It is recommended that approximately half of the dosage be taken at the beginning of the meal or snack and the remainder be taken during the meal or snack.

The recommended dosage is as follows:

- 1 mL (9,500 I.U.) (one full measuring scoop or 28 drops) per meal or snack for patients up to 15 kg in body weight.
- 2 mL (17,000 I.U.) (two full measuring scoops or 56 drops) per meal or snack for patients over 15 kg in body weight.

Dosage may be measured with the 1 mL measuring scoop (provided) or by drop count method (1 mL equals 28 drops from the Sucraid container tip).

**HOW SUPPLIED**

Sucraid (sacrosidase) Oral Solution is available in 118 mL (4 fluid ounces) translucent plastic bottles, packaged two bottles per box. Each mL of solution contains 8,500 International Units (I.U.) of sacrosidase. A 1 mL measuring scoop is provided with each bottle.

A full measuring scoop equals 28 drops from the Sucraid container tip.

Store in a refrigerator at 2°-8° C (36°-46°F). Protect from light and freeze.

To order, or for any questions, call 1-866-409-3773.

www.sucraid.net

NDC #7871-111-04

1114-VR8021
Patient Package Insert

INFORMATION FOR PATIENTS

Sucraid® (sacrosidase) Oral Solution

Please read this leaflet carefully before you take Sucraid® (sacrosidase) Oral Solution or give Sucraid to a child. Please do not throw away this leaflet. You may need to read it again at a later date. This leaflet does not contain all the information on Sucraid. For further information or advice, ask your doctor or pharmacist.

BEFORE TAKING SUCRAID

WARNING: Sucraid may cause a serious allergic reaction. If you notice any swelling or have difficulty breathing, get emergency help right away. Before taking your first and second doses, be sure that there are health professionals nearby (within a few minutes of travel) just in case there is an allergic reaction.

INFORMATION ABOUT YOUR MEDICINE

The name of your medicine is Sucraid® (sacrosidase) Oral Solution. It can be obtained only with a prescription from your doctor.

The purpose of your medicine:

Sucraid is an enzyme replacement therapy for the treatment of the genetically determined sucrase deficiency, which is part of congenital sucrase-isomaltase deficiency (CSID). CSID is a condition where your body lacks the enzymes needed to break down and absorb sucrose (table sugar) and other sugars from starch.

The symptoms of CSID often include frequent watery diarrhea, abdominal pain, bloating, and gas. In many cases, the symptoms of CSID are similar to other medical problems. Only your doctor can make a definite diagnosis of CSID.

Sucraid can help improve the breakdown and absorption of sucrose (table sugar) from the intestine and can help relieve the gastrointestinal symptoms of CSID.

Sucraid does not break down some sugars resulting from the digestion of starch. Therefore, you may need to restrict the amount of starch in your diet. Your doctor will tell you if you should restrict the amount of starch in your diet.

Discuss the following important information with your doctor before you begin to take Sucraid:

Tell your doctor if you are allergic to, have ever had a reaction to, or have ever had difficulty taking yeast, yeast products, papain, or glycerin (glycerol).

Tell your doctor if you have diabetes. With Sucraid, sucrose (table sugar) can be absorbed from your diet and your blood glucose levels may change. Your doctor will tell you if your diet or diabetes medicines need to be changed.

Side effects to watch for:

Some patients may have worse abdominal pain, vomiting, nausea, or diarrhea. Constipation, difficulty sleeping, headache, nervousness, and dehydration have also occurred. Other side effects may also occur. If you notice these or any other side effects during treatment with Sucraid, check with your doctor.

Stop taking Sucraid and get emergency help immediately if any of the following side effects occur: difficulty breathing, wheezing, or swelling of the face.

How to take your medicine:

Each bottle of Sucraid is supplied with a plastic screw cap which covers a dropper dispensing tip. Remove the outer cap and measure out the required dose. Rinse the bottle after each use by replacing and twisting the cap until tight.

Write down the date the sealed bottle is first opened in the space provided on the bottle label. Always throw away the bottle four weeks after first opening it because Sucraid contains no preservatives. For the same reason, you should rinse the measuring scoop with water after each time you finish using it.

To get the full benefits of this medicine, it is very important to take Sucraid as your doctor has prescribed. The usual dosage is 1 to 2 milliliters (mL) with each meal or snack: 1 mL = 1 full measuring scoop (28 drops from the bottle tip) and 2 mL = 2 full measuring scoops (56 drops from the bottle tip).

Measure your dose with the measuring scoop provided (see Figure 1). Do not use a kitchen teaspoon or other measuring device since it will not measure an accurate dose.

Figure 1. Measure dose with measuring scoop.

1. Measure dose with measuring scoop.

Mix your dose in 2 to 4 ounces of water, milk, or infant formula (see Figure 2). Sucraid should not be dissolved in or taken with fruit juice.

NEVER HEAT SUCRAID OR PUT IT IN WARM OR HOT BEVERAGES OR INFANT FORMULA. Heating Sucraid causes it to lose its effectiveness. The beverage or

Figure 2. Mix dose in beverage or infant formula.

It is recommended that approximately half of your dosage be taken at the beginning of each meal or snack and the remainder of your dosage be taken during the meal or snack.

Storing your medicine:

Sucraid is available in 4 fluid ounce (118 mL) see-through plastic bottles, packaged two bottles per box. A 1 mL measuring scoop is provided with each bottle. Always store Sucraid in a refrigerator at 36°F - 46°F (2°C - 8°C). Protect Sucraid from heat and light.

If your bottle of Sucraid has expired (the expiration date is printed on the bottle label), throw it away. Keep this medicine in a safe place in your refrigerator where children cannot reach it.

QOL Medical, LLC
Vero Beach, FL 32963

www.sucraid.net
For questions call 1-866-469-3773

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