Helpful Information for **Patients**



Sucraid®

(sacrosidase) Oral Solution Information about Sucraid® can be found at Sucraid.com

and Sucraid.com/patient-support/ nutritional-support

Specialty Pharmacy

For drug delivery information, contact the specialty pharmacy at

> **Frontier Therapies - Optum** Phone: 1-833-800-0122 Fax: 1-866-850-9155 frontiertherapies.optum.com

CSID

For information on Congenital Sucrase-Isomaltase Deficiency (CSID) and Sucraid® visit Sucraid.com

Main: 1-800-705-1962 • Fax: 1-866-777-7097 sucraid@golmed.com

Sucraid® Digests Sucrose

Sucraid® (sacrosidase) Oral Solution is an FDA-approved enzyme replacement for sucrase to aid in the digestion of sucrose in patients diagnosed with CSID. Sucraid® can help improve the breakdown and absorption of sucrose (table sugar) from the intestine and can help relieve the gastrointestinal (GI) symptoms of CSID. Sucraid® does not break down some sugars resulting from the digestion of starch. Therefore, you may need to reduce the amount of starch in your diet.

Do I Need to Change My Diet?

Diet is specific to each individual and depends on many factors, such as: How much sucrose and starch are currently in your diet; If you have other health issues that require a special diet; How well your digestive enzymes and your GI tract are working. It is important to know which foods are high and low in sucrose and starch.

If you have been on Sucraid® for several weeks and continue to have some lingering GI symptoms, you should talk to your doctor about whether you may need to adjust your starch intake. You should discuss dietary changes with your physician or registered dietitian prior to making any changes to your diet. If you are ever without Sucraid®, you should avoid foods high in sucrose. See Red Flag Foods below.

Foods High in Sucrose - Red Flag Foods!

- Apples
- Apricots
- Bananas
- Cantaloupe Clementine
- Dates
- Grapefruit
- Guava Honevdew melon Mandarin
- oranges Manao
- Nectarine
- Oranges
- Passion fruit Peaches

- Persimmon Pineapple
- Plums Tanaelos
- Tangerines Watermelon

VEGETABLES

- Beets Carrots
- Cassava (vucca)
- Chickpeas (aarbanzo beans)
- Coleslaw
- Corn • Edamame

Green peas

- Jicama
- Kidney beans • Lima beans
- Okra
- Onion
- Parsnips Pumpkin
- Snow peas Split peas
- Sweet pickles Sweet potatoes. vams

DAIRY

 Flavored milks containing

Muffins (chocolate milk)* Pancakes.

- Milk shakes sweetened with condensed milk.
- malted milk* Yoaurt*
- Yogurt containing fruits from the hiah-fructose fruits listed above

BAKED AND PROCESSED FOODS*

 Breakfast cereals Granola bars

Sorbet Brownies Chocolate

pastries, and

desserts: cake.

Sweets and

pie, cookies

waffles

Candy

Ice cream

Popsicles

Pudding

Sherbet

• Pie

SWEETENERS AND INGREDIENTS

- Sucrose (table) sugar)
- · Brown sugar
- Granulated sugar
- · Powdered and raw sugar
- Beet sugar · Cane sugar/syrup
- Cane juice
- Coconut sugar
- Date sugar
- Maple svrup/sugar Molasses
- Syrup
- Jellv. jam

Bold foods are especially high in sucrose. * Sweetened with sucrose

Created with Nutrition Data System for Research® (Regents of the University of Minnesota, 2017). High sucrose defined as ≥1 g sucrose per 100 g food

NOTE: This information is provided for educational purposes only and is not a substitute for talking with your doctor. You should consult with your healthcare provider if you have questions or concerns about your diet and/or the use of Sucraid®

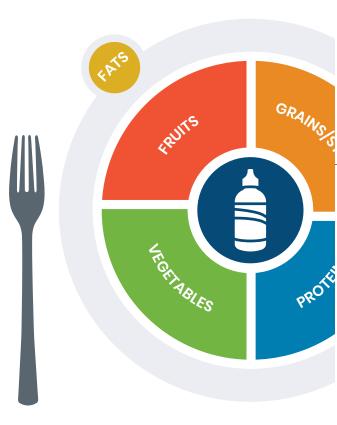
ADDITIONAL IMPORTANT SAFETY INFORMATION

- Some patients treated with Sucraid® may have worse abdominal pain, vomiting, nausea, or diarrhea. Constipation, difficulty sleeping, headache, nervousness, and dehydration have also occurred in patients treated with Sucraid®. Check with your doctor if you notice these or other side effects.
- Sucraid® has not been tested to see if it works in patients with secondary (acquired) sucrase deficiency.
- NEVER HEAT SUCRAID® OR PUT IT IN WARM OR HOT BEVERAGES OR INFANT FORMULA. Do not mix Sucraid® with fruit juice or take it with fruit juice. Take Sucraid® as prescribed by your doctor. Normally, half of the dose of Sucraid® is taken before a meal or snack. and the other half is taken during the meal or snack.
- Sucraid® should be refrigerated at 36°F-46°F (2°C-8°C) and should be protected from heat and light.

Please see additional Important Safety Information on Sucraid® Digests Sucrose 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.

Sucraid® and **Your Diet**

This Guide is Intended for Patients on Sucraid®



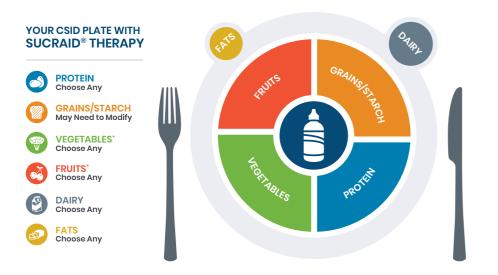


Sucraid® and Diet

The goal of Sucraid® therapy is to help you eat as near normal and as healthy a diet as possible without the return of aastrointestinal (GI) symptoms.

With Sucraid® therapy, you can include foods that are higher in sucrose, because Sucraid® helps you diaest these foods. Because **Sucraid**® therapy does not break down some sugars that come from the digestion of starch, you may need to restrict or experiment with the amount of starch in your diet to prevent any lingering GI symptoms.

Many of you with Congenital Sucrase-Isomaltase Deficiency (CSID) who are on Sucraid® therapy can choose freely from all food groups, but some of you may need to be cautious about foods that are high in starch. Go to choosemyplate.gov to help plan your meals.





Foods High in Starch

STARCHY VEGETABLES

- Beans (black, kidney, lima)
- Corn

INDICATION

- Peas (black-eved, areen)
- · Potatoes (white, red, golden)
- Sweet potatoes Yams

WHOLE GRAINS

- Brown rice

your diet. Your doctor will tell you if you should restrict starch in your diet.

REFINED STARCH

 Cakes Bran cereal, oats

IMPORTANT SAFETY INFORMATION FOR SUCRAID® (SACROSIDASE) ORAL SOLUTION

- Popcorn

which is part of congenital sucrase-isomaltase deficiency (CSID).

Quinoa

Cookies

 Whole grain bread, cereal, crackers, pasta

Created with Nutrition Data System for Research® (Regents of the University of Minnesota, 2017), High starch defined as > 2.5 a starch per 100 a food or > 2.5 a starch

Sucraid® (sacrosidase) Oral Solution is an enzyme replacement therapy for the treatment of genetically determined sucrase deficiency,

Sucraid® may cause a serious allergic reaction. If you notice any swelling or have difficulty breathing, get emergency help right away.

■ Sucraid® does not break down some sugars that come from the digestion of starch. You may need to restrict the amount of starch in

■ 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

- - Pasta
- Cereal, granola bars Chips (corn, potato,
- tortilla)
 - Muffins, pastries
 - Pancakes, waffles
 - Refined cereal
- Saltine crackers
- White bread · White rice
 - closes. In a world give closed is actually were associated with given total stoke and higher propriations of patients having lower total symptom scores, the primary efficacy end-points. In addition, higher doses of sociadase were associated with a significantly general runder of hard and formed stoke as well as with fewer watery and soft stock, the secondary efficacy end-points.

Analysis of the overall symptomatic response as a function of age indicated that in CSID patients up to 3 years of age, 86% became asymptomatic. In patients over 3 years of age, 77% became asymptomatic. Thus, the therapeutic response did not differ significantly according to age.

A second study of similar design and execution as the first used 4 different dilutions of socrosidase: 1:100~(90~LU/mL), 1:1000~(9~LU/mL), 1:10,000~(0.9~LU/mL), and 1:100,000 (0.09 LU./mL). There were inconsistent results with regards to the primary

Prescribing Information

Sucraid (socrosidase) Oral Solution is an enzyme replacement therapy for the treatment of genetically determined sucrase deficiency, which is part of congenital sucrase-isomathase deficiency (CSD).

Sucraid is a pale yellow to colorless, clear solution with a pleasant sweet taste. Each millifer (mL) of Sucraid contains 8,500 International Units (LU.) of the enzyme

sacrosidase, the active ingredient. The chemical name of this enzyme is B.D.-fructofuranoside fructohydrolase. The enzyme is derived from baker's yeast (Sac

It has been reported that the primary amino acid structure of this protein consists of

513 amino acids with an apparent molecular weight of 100,000 a/mole for the

glycosylated monomer (range 66,000-116,000 g/male). Reports also suggest that the protein exists in solution as a monomer, dimer, fetramer, and octomer ranging from 100,000 g/male to 800,000 g/male. It has an isoelectric point (pl) of 4.5.

Sucraid may contain small amounts of papain. Papain is known to cause allerain 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

Sucraid contains sacrosidase in a vehicle comprised of glycerol (50% wt/wt), water, and citric acid to maintain the pH at 4.0 to 4.7. Glycerol (glycerin) in the amount

This enzyme preparation is fully soluble with water, milk, and infant formula. DO NOT

Congenital sucrase-isomatiase 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 endoaenous

sucrase activity, a very marked reduction in isomaltase activity, a moderate de-

Sucrase is naturally produced in the brush border of the small intestine

primarily the distal duodenum and leiunum. Sucrase hydrolyzes the disaccharide

sucrose into its component monosaccharides, glucose and fructose. Isomalitase breaks down disaccharides from starch into simple sugars. Sucraid does not contain

In the absence of endogenous human sucrose as in CSD sucrose is not

and their presence in the intestinal lumen may lead to associate in the intestinal lumen may lumen may lead to associate in the intestinal lumen may lumen may l

Unabsorbed sucrose in the colon is fermented by bacterial flora to produce increased amounts of hydrogen, methone, and water. As a consequence, excessive gas, bloating, abdominal cramps, nausea, and vamiling may occur.

Chronic malabsoration of disaccharides may result in mainutrition. Undiagnosed

untreated CSD patients often fail to thrive and fall behind in their expected growth and development curves. Previously, the treatment of CSD has required the

CSD is often difficult to diagnose. Approximately 4% to 10% of pediatric patients with chronic diarrhea of unknown origin have CSD. Measurement of expired breath hydrogen under controlled conditions following a sucrose challenge (a

measurement of excess hydrogen excreted in exhalation) in CSID patients has show

A generally accepted clinical definition of CSID is a condition characterized by the

following: stool pH < 6, an increase in breath hydrogen of > 10 ppm when challenged

with sucrose offer fasting and a negative lactose breath test. However, because of the difficulties in diagnosing CSID, it may be warranted to conduct a short therapeutic trial (e.g., one week) to assess response in patients suspected of having CSID.

A two-phase (dose response preceded by a breath hydrogen phase) double-blind, multi-site, crossover trial was conducted in 28 patients (aged 4 months to 11.5 years) with confirmed CSD. During the dose response phase, the patients were challenged

with an ordinary sucrose-containing diet while receiving each of four doses of

sorrosidose: full strenath (9000 LLL /ml.) and three dilutions (1-10 (900 LLL /ml.) 1-100

sociosase: tul strength (VUU LU/ML), and three dullards (LIU VUU LU/ML), LIUU (90 LU/ML), and 1:1000 (9 LU/ML) in random order for a period of 10 days, Patients who weighed no more than 15 kg received 1 mL per med; those weighing more than 15 kgreceived 2 mL per med. The dose did not vary with age or sucrose intake.

A dose-response relationship was shown between the two higher and the two lower

doses. The two higher doses of sacrosidase were associated with significantly fewer

consumed in the recommended doses of Sucraid has no expected toxicity.

Sucraid® (sacrosidase) Oral Solution:

completely removed during subsequent process steps.

crease in maltase activity, and normal lactase levels.

continual use of a strict sucrose-free diet.

CLINICAL STUDIES

levels as areat as 6 times that in normal subjects.

CHEMISTRY

haromyoes cerevisiae).

CUNICAL PHARMACOLOGY

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 sucrase deficiency, which is part of congenital sucraseisomottose deficiency (CSD)

CONTRAINDICATIONS

Patients known to be hypersensitive to yeast, yeast products, alycerin (alycerol), or nanain

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

Other serious events have not been linked to Sucraid

of travel) a facility where acute hypersensitivity reactions can be adequately treated. Alternatively, the patient may be tested for hypersensitivity to Sucraid through skin abrasion testing. Should symptoms of hypersensitivity appear, 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

Although Sucraid provides replacement therapy for the deficient sucrase, it does not provide specific replacement therapy for the deficient isomatose. Therefore restricting starch in the diet may still be necessary to reduce symptoms as much a

possible. The need for dietary starch restriction for patients using Sucraid should be

small bowel biopsy or breath 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 sucrase

The effects of Sucraid have not been evaluated in patients with secondary

See Patient Package Insert. Patients should be instructed to discard bottles of Sucroid diversity offer angoing due to the protection for bootstand and the Sucretical Company.

reason, patients should be advised to rinse the measuring scoop with water after

Superaid is fully soluble with water milk, and infant formula, but it is important to note

ie use of Sucraid will enable the products of sucrose hydrolysis, glucose and

The definitive test for diagnosis of CSID is the measurement of intestinal

Other tests used alone may be inaccurate: for example, the breath hydrogen test

(high incidence of false negatives) or oral sucrose tolerance test (high incidence of

false positives). Differential urinary disaccharide testing has been reported to show good gareement with small intestinal biopsy for diagnosis of CSID.

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

rcinogenic potential. Studies to evaluate the effect of Sucraid on fertility or its mi

reconsult. I Tearloganic effects. Pregnancy Cafegory C. Animal reproduction studies have not been conducted with Sucrial. Sucrial is not expected to cause fetal harm when administered to a pregnant woman or to affect reproductive capacity. Sucrial should be given to a pregnant woman only if clearly needed.

The Sucraid enzyme is broken down in the stomach and intestines, and the

Sucraid has been used in patients as young as 5 months of age. Evidence in one

for the treatment of the genetically acquired sucrase deficiency, which is part of

Adverse experiences with Sucraid in clinical trials were generally minor and were

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 parenthesis) were as follows: abdominal pain (4), vomitina (3), nause (2), diarrhea (2), constipation (2), insomnia (1), headache (1), nervousness (1), and dehydration (1).

Note: Diarrhea and abdominal pain can be a part of the clinical presentation of the

genetically determined sucrase deficiency, which is part of congenital sucrasi isomaltase deficiency (CSD).

One asthmatic child experienced a serious hypersensitivity reaction. (wheezing

robably related to socrosidase (see Warnings). The event resulted in withdrawal of the patient from the trial but resolved with no sequelae.

The recommended dosage is 1 or 2 mL (8,500 to 17,000 LU.) or 1 or 2 full measuring

scops (each full measuring scopp equals 1 m1; 28 drops from the Sucraid contain tip equals 1 m1;) taken orally with each meal or snack diluted with 2 to 4 ounces (é

to 120 mL) of water, milk, or infant formula. The beverage or infant formula should be

served cold or at room temperature. The beverage or infant formula should not be

warmed or heated before or after addition of Sucraid because heating is likely to

decrease potency. Sucraid should not be reconstituted or consumed with fruit juice

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

1 mL (8,500 I.U.) (one full measuring scoop or 28 drops) per meal or snack for patient

2 mL (17,000 I.U.) (two full measuring scoops or 56 drops) per meal or snack for

Dosage may be measured with the 1 mL measuring scoop (provided) or by drop

Sucraid (sacrosidase) Oral Solution is available in 118 ml (4 fluid ounces) translucen

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

Store in a refrigerator at 2° - 8° C (36° - 46° F). Discard four weeks after first opening due to the potential for bacterial growth. Protect from heat and light.

t method (1 mL equals 28 drops from the Sucraid container tip)

component amino acids and pentides are then absorbed as nutrients

with fruit juice, since its acidity may reduce the enzyme activity.

CARCINOGENESIS, MUTAGENESIS, IMPAIRMENT OF FERTILITY

ana-term studies in animals with Sucraid have

that this product is sensitive to heat. Sucraid should not be reconstitutions with fruit juice, since its acidity may reduce the enzyme activity.

INFORMATION FOR PATIENTS

of diabetic CSID patients using Sucraid.

disaccharidases following small bowel biopsy

LABORATORY TESTS

NURSING MOTHERS

ADVERSE REACTIONS

frequently associated with the underlying disease

Overdosage with Sucraid has not been reported.

since its acidity may reduce the enzyme activity.

The recommended dosage is as follows:

up to 15 kg in body weight.

patients over 15 kg in body weight.

bottle. A full measuring scoop is 1 mL.

To order, or for any questions, call 1-866-469-3773

Rx only.

QOI Medical III

Vero Beach, FL 32963

NDC# 67871-111-04

DOSAGE AND ADMINISTRATION

PEDIATRIC USE

INFORMATION FOR PATIENTS It may sometimes be clinically inappropriate, difficult, or inconvenient to perform a

Patient Package Insert Sucraid® (sacrosidase) Oral Solution

BEFORE TAKING SUCRAID

Please read this leaflet carefully before you take

information or advice, ask your doctor or pharmacist

of travel) just in case there is an allergic reaction.

Sucraid is an enzyme replacement therapy for the

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

The symptoms of CSID often include frequent watery

diarrhea, abdominal pain, bloating, and ags. In many

cases, the symptoms of CSID are similar to other

Sucraid can help improve the breakdown and absorption

of sucrose (table sugar) from the intestine and can help

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

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,

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

Some patients may have worse abdominal pain,

vomiting, nausea, or diarrhea. Constipation, difficulty

sleeping, headache, nervousness, and dehvdration

have also occurred. Other side effects may also occur.

If you notice these or any other side effects during

if any of the following side effects occur: difficulty

cap which covers a dropper dispensing tip. Remove

treatment with Sucraid, check with your doctor.

breathing, wheezing, or swelling of the face

yeast products, papain, or glycerin (glycerol).

relieve the gastrointestinal symptoms of CSID.

medical problems. Only your doctor can make a

reatment of the genetically determined sucrase

INFORMATION AROUT VOLID MEDICINE

prescription from your doctor.

The purpose of your medicine:

definite diagnosis of CSID.

need to be changed.

Side effects to watch for:

How to take your medicine:



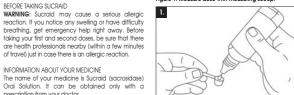
Sucraid (sacrosidase) Oral Solution or give Sucraid to a Measure your dose with the measuring scoop provided child. Please do not throw away this leaflet. You may (see Figure 1). Do not use a kitchen teaspoon or other need to read it again at a later date. This leaflet does measuring device since it will not measure an accurate not contain all the information on Sucraid. For further

each meal or snack: 1ml = 1 full measuring scoon (28

drops from the bottle tip) and 2 mL = 2 full measuring

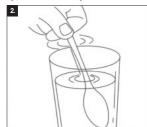
Figure 1 Measure dose with measuring scoop

scoops (56 drops from the bottle tip).



Mix your dose in 2 to 4 ounces of water, milk, or infant

NEVER HEAT SUCRAID OR PUT IT IN WARM OR HOT



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

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

If your bottle of Sucraid has expired (the expiration date is printed on the bottle label), throw it away.

Keen this medicine in a safe place in your refrigerator where children cannot reach it. Stop taking Sucraid and get emergency help immediately

www.sucraid.com Each bottle of Sucraid is supplied with a plastic screw

the outer cap and measure out the required dose. Reseal 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 i 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

formula (see Figure 2). Sucraid should not be dissolved in or taken with fruit juice.

BEVERAGES OR INFANT FORMULA. Heating Sucraid causes it to lose its effectiveness. The beverage or infant formula should be taken cold or at room temperature

Figure 2 Mix dose in beverage or infant formula

during the meal or snack.

Storing your medicine:

QOL Medical, LLC Vero Beach, FL 32963

For auestions call 1-866-469-3773

Rev 06/20

■ Tell your doctor if you have diabetes, as your blood glucose levels may change if you begin taking Sucraid®. Your doctor will tell you if your diet or diabetes medicines need to be changed.

Please see additional Important Safety Information on Sucraid® and Diet page and in enclosed full Prescribina Information.