Prescribing Information

Sucraid[®] (sacrosidase) Oral Solution:

DESCRIPTION

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).

Sucraid is a pale yellow to colorless, clear solution with a pleasant sweet taste. Each milliliter (mL) of Sucraid® contains 8,500 International Units (I.U.) 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 (Saccharomyces cerevisiae).

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 g/mole for the glycosylated monomer (range 66,000-116,000 g/mole). Reports also suggest that the protein exists in solution as a monomer, dimer, tetramer, and octomer ranging from 100,000 g/mole to 800,000 g/mole. It has an isoelectric point (pl) 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 duringsub sequent process steps

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 (alvcerin) 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. DO NOT HEAT SOLUTIONS CONTAINING SUCRAID. Do not put Sucraid in warm or hot liquids.

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 distal duodenum and jejunum. Sucrase hydrolyzes the disaccharide sucrose into its component monosaccharides, glucose Skin testing as a rechallenge has been used to verify hypersensitivity in and fructose. Isomaltase breaks down disaccharides from starch into one asthmatic child who displayed wheezing after oral sacrosidase. simple sugars. Sucraid does not contain isomaltase.

from the intestine and their presence in the intestinal lumen may lead to osmotic retention of water. This may result in loose stools.

Unabsorbed sucrose in the colon is fermented by bacterial flora to in each patient. produce increased amounts of hydrogen, methane, and water. As a consequence, excessive gas, bloating, abdominal cramps, nausea, It may sometimes be clinically inappropriate, difficult, or inconvenient and vomiting may occur.

Chronic malabsorption of disaccharides may result in malnutrition Undiagnosed/untreated CSID patients often fail to thrive and fall Sucraid to assess response in a patient suspected of sucrase deficiency. behind in their expected growth and development curves Previously, the treatment of CSID has required the continual use of a strict sucrose-free diet

CSID is often difficult to diagnose. Approximately 4% to 10% of pediatric patients with chronic diarrhea of unknown origin have CSID. See Patient Package Insert. Patients should be instructed to discard 2 mL (17,000 I.U.) (two full measuring scoops or 56 drops) per meal or Measurement of expired breath hydrogen under controlled conditions following a sucrose challenge (a measurement of excess hydrogen excreted in exhalation) in CSID patients has shown levels as great as 6 rinse the measuring scoop with water after each use. times that in normal subjects.

hydrogen of > 10 ppm when challenged with sucrose after fasting and a negative lactose breath test. However, because of the difficulties in diagnosing CSID, it may be warranted to conduct a short USE IN DIABETICS therapeutic trial (e.g., one week) to assess response in patients The use of Sucraid will enable the products of sucrose hydrolysis, suspected of having CSID.

CLINICAL STUDIES

A two-phase (dose response preceded by a breath hydroaen phase) LABORATORY TESTS double-blind, multi-site, crossover trial was conducted in 28 patients (aged 4 months to 11.5 years) with confirmed CSID. During the dose response phase, the patients were challenaed with an ordinary sucrosecontaining diet while receiving each of four doses of sacrosidase: full hydrogen test (high incidence of false negatives) or oral sucrose strength (9000 I,U,/mL) and three dilutions (1:10 (900 I,U,/mL), 1:100 (90 tolerance test (high incidence of false positives). Differential urinary Distributed by: QOL Medical. LLC I.U./mL), and 1:1000 (9 I.U./mL)) in random order for a period of 10 days. disaccharide testing has been reported to show good agreement with Vero Beach, FL 32963 Patients who weighed no more than 15 kg received 1 mL per meal: small intestinal biopsy for diagnosis of CSID. those weighing more than 15 kg received 2 mL per meal. The dose did not vary with age or sucrose intake. A dose-response relationship was DRUG INTERACTIONS shown between the two higher and the two lower doses. The two higher Neither drug-drug nor drug-food interactions are expected or have been

and higher proportions of patients having lower total symptom scores. reconstituted or consumed with fruit juice, since its acidity may reduce the primary efficacy end-points. In addition, higher doses of sacrosidase were associated with a significantly greater number of hard and formed stools as well as with fewer watery and soft stools, the secondary efficacy end-points.

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 Teratogenic effects. Pregnancy Category C. Animal reproduction according to age

A second study of similar design and execution as the first used 4 woman or to affect reproductive capacity. Sucraid should be given to different dilutions of sacrosidase: 1:100 (90 I.U./mL), 1:1000 (9 I.U./mL), 1:10.000 (0.9 I.U./ml.), and 1:100.000 (0.09 I.U./ml.). There were inconsistent results with regards to the primary efficacy parameters.

hydrogen output when they received sacrosidase in comparison to placebo

INDICATIONS AND USAG

ucraid (sacrosidase) Oral Solution is indicated as oral replacement therapy of the genetically determined sucrase deficiency, which is part acquired sucrase deficiency, which is part of CSID. of Congenital Sucrase-Isomaltase Deficiency (CSID).

CONTRAINDICATIONS

Patients known to be hypersensitive to yeast, yeast products, glycerin and were frequently associated with the underlying disease. (glycerol), or papair

necessitated admission into the ICU for a 4-year-old boy. The wheezing abdominal pain (4), vomiting (3), nausea (2), diarrhea (2), constipation was probably caused by sacrosidase. He had asthma and was being (2), insomnia (1), headache (1), nervousness (1), and dehydration (1). treated with steroids. A skin test for sacrosidase was positive.

Other serious events have not been linked to Sucraid.

PRECAUTIONS

few minutes 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 sequelae. symptoms of hypersensitivity appear, discontinue medication and initiate symptomatic and supportive therapy.

not metabolized. Unhydrolyzed sucrose and starch are not absorbed sucrose, it does not provide specific replacement therapy for the diluted with 2 to 4 ounces (60 to 120 mL) of water, milk, or infant 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 heated before or after addition of Sucraid because heating is likely to

> to perform a small bowel biopsy or breath hydrogen test to make a It is recommended that approximately half of the dosage be taken at 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 the meal or snack.

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

INFORMATION FOR PATIENTS

bottles of Sucraid 4 weeks after opening due to the potential for snack for patients over 15 kg in body weight bacterial arowth. For the same reason, patients should be advised to

Sucraid is fully soluble with water, milk, and infant formula, but it is container tip). A generally accepted clinical definition of CSID is a condition important to note that this product is sensitive to heat. Sucraid should characterized by the following: stool pH < 6, an increase in breath not be reconstituted or consumed with fruit juice, since its acidity may HOW SUPPLIED reduce the enzyme activity

glucose and fructose, to be absorbed. This fact must be carefully considered in planning the diet of diabetic CSID patients using Sucraid

The definitive test for diagnosis of CSID is the measurement of intestinal heat and light. haridases following small bowel biopsy

Other tests used alone may be inaccurate: for example, the breath Rx only.

doses of sacrosidase were associated with significantly fewer total stools reported with the use of Sucraid. However, Sucraid should not be NDC# 67871-111-04

the enzyme activity

CARCINOGENESIS MUTAGENESIS IMPAIRMENT OF FERTILITY

ong-term studies in animals with Sucraid have not been performed to evaluate the carcinogenic potential. Studies to evaluate the effect of Analysis of the overall symptomatic response as a function of age Sucraid on fertility or its mutagenic potential have not been performed.

studies have not been conducted with Sucraid. Sucraid is not expected to cause fetal harm when administered to a pregnant a pregnant woman only if clearly needed.

NURSING MOTHERS

The Sucraid enzyme is broken down in the stomach and intestines, and In both trials, however, patients showed a marked decrease in breath 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. vidence in one controlled trial in primarily pediatric patients shows that Sucraid is safe and effective for the treatment of the genetically

ADVERSE REACTIONS

Adverse experiences with Sucraid in clinical trials were generally minor

In clinical studies of up to 54 months duration, physicians treated a total of 52 patients with Sucraid. The adverse experiences and respective Severe wheezing, 90 minutes after a second dose of sacrosidase, number of patients reporting each event (in parenthesis) were as follows:

> Note: Diarrhea and abdominal pain can be a part of the clinical presentation of the genetically determined sucrase deficiency, which is part of Congenital Sucrase-Isomaltase Deficiency (CSID).

Care should be taken to administer initial doses of Sucraid near (within a One asthmatic child experienced a serious hypersensitivity reaction (wheezing) probably related to sacrosidase (see Warnings). The event resulted in withdrawal of the patient from the trial but resolved with no

OVERDOSAG

Overdosage with Sucraid has not been reported.

DOSAGE AND ADMINISTRATION

The recommended dosage is 1 or 2 mL (8,500 to 17,000 I.U.) or 1 or 2 full measuring scoops (each full measuring scoop equals 1 mL; 28 drops from In the absence of endogenous human sucrase, as in CSID, sucrose is Although Sucraid provides replacement therapy for the deficient the Sucraid containertip equals 1 mL) taken orally with each meal or snack formula. The beverage or infant formula should be served cold or at room temperature. The beverage or infant formula should not be warmed or decrease potency. Sucraid should not be reconstituted or consumed with fruit juice since its acidity may reduce the enzyme activity.

the beginning of the meal or snack and the remainder be taken during

The recommended dosage is as follows:

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

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.) sacrosidase. A 1 mL measuring scoop is provided with each bottle. A ful measuring scoop is 1 mL.

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

To order, or for any questions, call 1-866-469-3773 www.sucraid.ne

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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 alleraic 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 sucraseisomaltase 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.

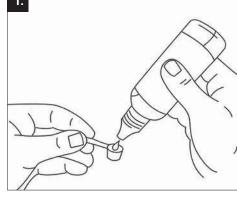


Figure 1. Measure dose with measuring

an accurate dose.

SCOOD.

Side effects to watch for:

your doctor.

the face

Some patients may have worse abdominal pain, dehydration have also occurred. Other side effects at room temperature. may also occur. If you notice these or any other side effects during treatment with Sucraid, check with Figure 2. Mix dose in beverage or infant

Stop taking Sucraid and get emergency help 2. immediately if any of the following side effects occur: difficulty breathing, wheezing, or swelling of

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. 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 scoop with water after each time you finish using it.

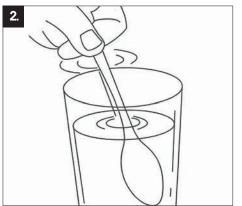
To get the full benefits of this medicine, it is verv important to take Sucraid as your doctor has pre- Storing your medicine: uring scoops (56 drops from the bottle tip).

Measure your dose with the measuring scoop pro- and light. vided (see Figure 1). Do not use a kitchen teaspoon

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 vomiting, nausea, or diarrhea. Constipation, Sucraid causes it to lose its effectiveness. The difficulty sleeping, headache, nervousness, and beverage or infant formula should be taken cold or

formula



the same reason, you should rinse the measuring dosage be taken at the beginning of each meal or snack and the remainder of your dosage be taken during the meal or snack.

scribed. The usual dosage is 1 to 2 milliliters (mL) with Sucraid is available in 4 fluid ounce (118 mL) each meal or snack: 1mL = 1 full measuring scoop see-through plastic bottles, packaged two bottles (28 drops from the bottle tip) and 2 mL = 2 full meas- 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

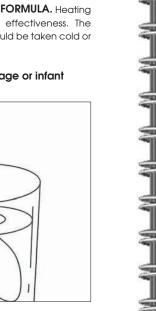
or other measuring device since it will not measure If your bottle of Sucraid has expired (the expiration date is printed on the bottle label), throw it away.

> Keep this medicine in a safe place in your refrigerator where children cannot reach it.

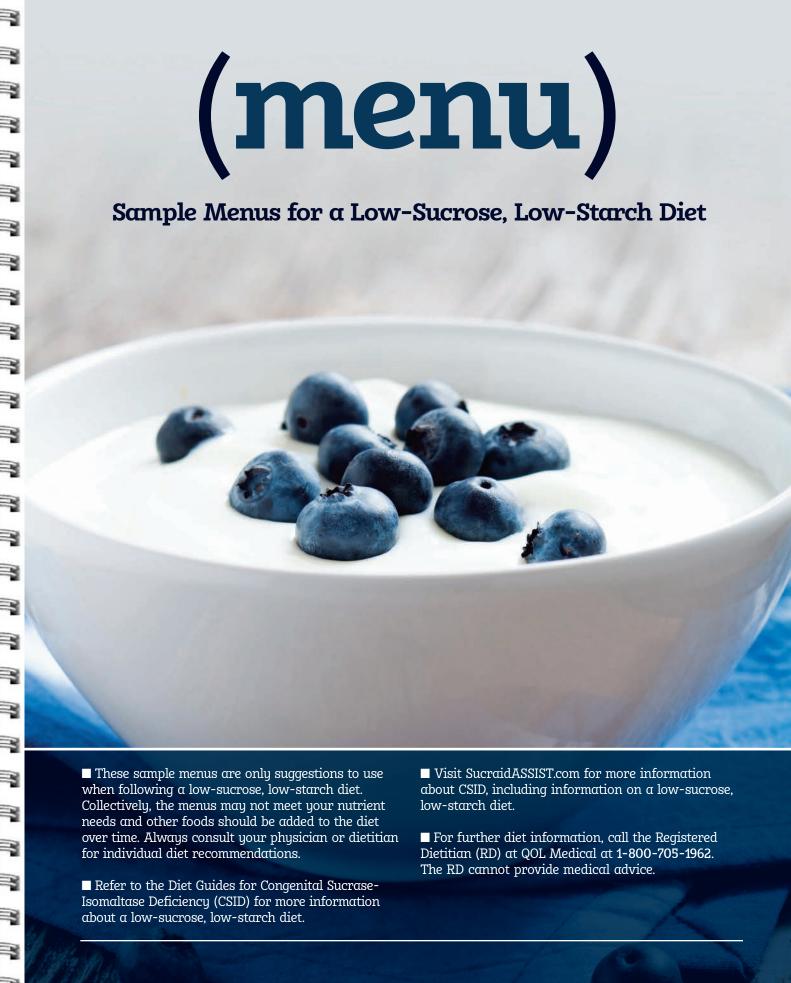
QOL Medical, LLC Vero Beach, FL 32963

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

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it because Sucraid contains no preservatives. For It is recommended that approximately half of your











Breakfast

- Scrambled eggs
- Bacon*
- Blueberries
- Milk
- Egg casserole with sausage*, cheese, green peppers, tomatoes, and mushrooms
- Strawberries
- Milk
- Cottage cheese
- Blueberries
- Milk
- Sliced ham*
- + Cheese cubes
- Grapes
- Milk
- Chicken salad (no sugar)
- + Grapes
- Cheese stick
- Milk
- Plain yogurt (no sugar), sweeten with fructose or dextrose
- Mix in blueberries and finely chopped pecans, almonds. and/or flax seeds
- Milk

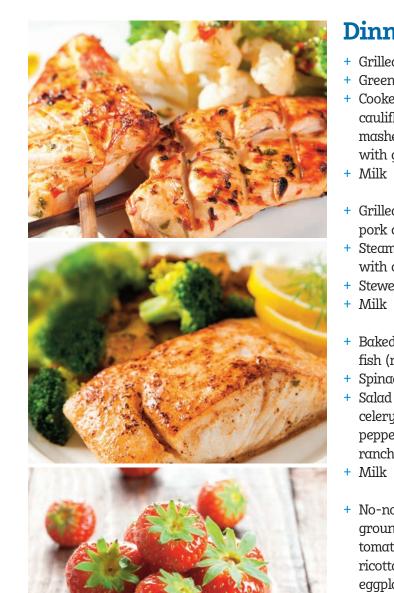
Lunch

- + Plain grilled or baked chicken strips (no breading)
- + Cheese stick or cubes
- + Red, green, or yellow peppers cut into strips + Ranch dressing** for dipping chicken or peppers
- + Milk
- + Deli meat* and sliced cheese
- + Spread mayonnaise*** or cream cheese on the meat/cheese and roll it up
- + Steamed broccoli
- + Mix mayonnaise*** and mustard for a dipping sauce for the broccoli
- + Milk
- + Tuna salad (no sugar) mayonnaise***, eggs, mustard, and dill pickles + Snow peas – steam and
- serve cold + Grapes, cut up
- + Milk
- + Tomato soup made with water, milk, or milk substitute
- + Melt grated cheese into soup
- + Cut up cucumbers, peppers, celery, and broccoli

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- + Italian dressing for dipping (no sugar)
- + Milk





Dinner

- Grilled or baked chicken
- Green beans
- Cooked, mashed cauliflower (looks like mashed potatoes) with grated cheese
- Grilled or baked pork chops
- + Steamed broccoli with cheese
- Stewed tomatoes
- Baked, grilled, or broiled fish (no breading)
- Spinach with butter Salad – cucumbers, celery, broccoli, and peppers with ranch dressing**
- No-noodle lasagna ground beef, canned tomatoes (no sugar), ricotta cheese, zucchini, eggplant, and mushrooms; make layers in a casserole dish, sprinkle with cheese, and bake
- + Tossed salad with dressing** Milk
- Ground turkey or chicken add cumin and chili powder
- Sauté peppers and mushrooms
- Mix the meat and vegetables, top with cheese. sour cream. and avocado
- + Tossed salad with dressing**
- Milk

Snacks

+ Unsweetened yogurt, sweeten with dextrose or fructose + Blueberries or strawberries added

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- + Cottage cheese, sweeten with dextrose or fructose
- + Blueberries or strawberries added
- + Cheese sticks or cubes
- + Meat roll ups (roll up turkey and cheese with cream cheese)
- + Bowl of strawberries, kiwi, grapes, and/or blueberries
- + Celery with natural almond or peanut butter
- + Raw vegetables (cucumbers, celery, peppers, mushrooms, squash, broccoli, cauliflower, snow peas, and/or radishes) and ranch dressing**
- + Mix mashed avocado, salsa (no sugar), and sour cream; use as a dip for raw vegetables

+ Hard-boiled eggs

Notes

- *All meats should be fresh; avoid fillers and sucrose. Meats cured with dextrose would be acceptable.
- **Be sure salad dressings do not contain sucrose or starch.
- ***Choose a mayonnaise that does not contain sucrose or starch.
- Take Sucraid[®] (sacrosidase) Oral Solution as prescribed with all meals and snacks.
- If you do not drink cow's milk, be sure to substitute with Lactaid[™] milk or a sugar-free, plain, or unflavored soy or almond milk.
- Always read food labels. Even if a food is listed here, check the label to make sure it is safe since ingredients are constantly changing.
- Dextrose can be purchased online from NOW[®] Foods (nowfoods.com) or from local breweries. Dextrose can be used in place of sugar.
- Vou can buy fructose at the grocery store on the aisle where other sugars are found. Fructose can also be purchased online from NOW[®] Foods (nowfoods.com). Fructose can be used in place of sugar.
- Talk to your physician or dietitian about the overall completeness of your diet and take vitamin and mineral supplements as recommended.

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

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

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, 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.

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.

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.

Please see full Prescribing Information on back of menu.

