

(Adult Diet Guide)

Guidelines for Adults 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



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. Diet consultation with a Registered Dietitian (RD) is recommended.

WWW.SUCRAID.NET

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).

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

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®.

(Adult Diet Guide)

Guidelines for Adults with Genetic Sucrase-Isomaltase Deficiency (GSID)



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. Diet consultation with a Registered Dietitian (RD) is recommended.

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.

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

NOTE:

- Full-fat dairy products may be used if more calories are indicated.
- Avoid processed cheeses or cheese products that contain sucrose or starch fillers.
- If lactose intolerant, avoid dairy foods. Substitute lactose-free milk, unsweetened almond milk, or soy milk for cow's milk. A dietitian can provide other dairy alternatives.

Protein

Beef
Chicken
Eggs
Fish
Lamb
Pork
Tofu
Turkey

NOTE:

- All meats above should be plain.
- Avoid breaded meats.
- Avoid processed meats such as bacon, sausage, luncheon meat, pâté, and liverwurst that are cured with sucrose or have starch fillers.

Vegetables

Alfalfa sprouts
Artichoke*
Asparagus*
Bamboo shoots
Bok choy

Broccoli*
Brussels sprouts*
Cabbage*
Cauliflower*
Celery
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

NOTE:

- * These vegetables may cause gas in all individuals, not just patients with Genetic Sucrase-Isomaltase Deficiency, and should be monitored closely.

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

NOTE:

- All fruits should be fresh; not cooked, canned, or dried.
- Some fresh, frozen fruits may be included.
- All fruits should be "unsweetened".

Sweeteners

Granulated fructose
Granulated dextrose

Fats

Any vegetable oils
Butter

Nuts & Seeds

Almonds
Almond butter
Brazil nuts
Flax seeds
Hazelnuts
Macadamia nuts
Peanuts
Peanut butter
Pecans
Pumpkin seeds
Sesame butter (tahini)
Walnuts

NOTE:

- Nuts and seeds can be difficult to digest in general.
- Most nuts and seeds contain varying amounts of sucrose and starch.
- When starting the diet, it is best to avoid nuts and seeds the first two weeks.
- It is important to keep the portion size small (in general a serving is less than 1 ounce for nuts).



Breakfast

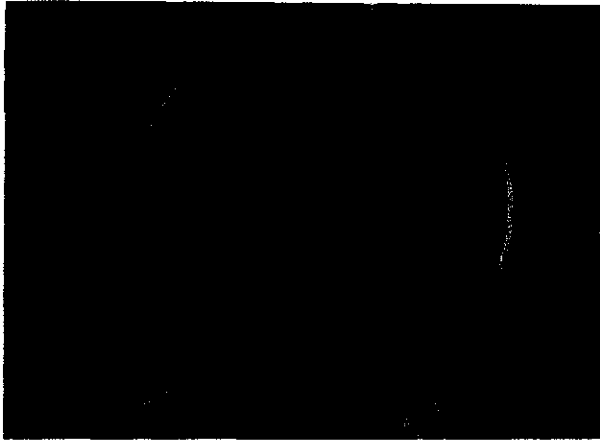
- + Scrambled eggs
- + Bacon*
- + Blueberries
- + Milk
- + Egg casserole with sausage*, cheese, green peppers, tomatoes, and mushrooms
- + Strawberries
- + Milk



- + 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



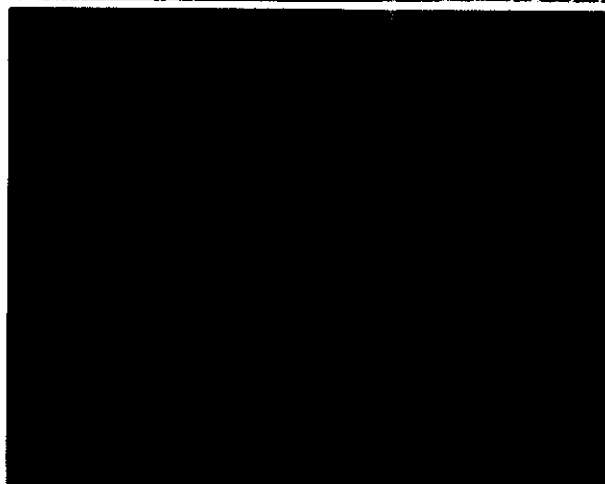
Lunch

- + 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, pepper, celery, and broccoli
- + Italian dressing for dipping (no sugar)
- + Milk



Dinner

- + Grilled or baked chicken
- + Green beans
- + Cooked, mashed cauliflower (looks like mashed potatoes) with grated cheese
- + Milk

- + Grilled or baked pork chops
- + Steamed broccoli with cheese
- + Stewed tomatoes
- + Milk

- + Baked, grilled, or broiled fish (no breading)
- + Spinach with butter
- + Salad - cucumbers, celery, broccoli, and peppers with ranch dressing**
- + Milk

- + 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

- + 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 as 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.
- You can buy fructose at the grocery store on the aisle where other sugars are found. Fructose can also be ordered online from NOW® Foods. 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 even 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 may have worse abdominal pain, vomiting, nausea or diarrhea. Constipation, difficulty sleeping, headache, nervousness and dehydration have also occurred. Check with your doctor if you notice these or other side effects.
- **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.

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.

Sucraid®
(sacrosidase) Oral Solution
WWW.SUCRAID.NET