

Congenital Sucrase-Isomaltase Deficiency (CSID)

Congenital Sucrase-Isomaltase Deficiency (CSID) is a disorder where individuals affected lack the enzymes to digest sucrose (sugar) and isomaltose (a type of starch). CSID is present at birth, but is often not diagnosed until 4-6 months of age when solid foods containing sugars and starches (cereal, fruits, and vegetables) are first introduced. Diagnosis in adulthood is rare, but does occur.

Individuals with CSID have varying amounts of the enzymes sucrase-isomaltase and maltase-glucoamylase that digest sugar and starch. Because enzyme levels vary, individuals' tolerance to sugar and starch containing foods also varies. Some tolerate more than others, allowing them a little more flexibility with their diet. Lactose is usually digested without any difficulty in all individuals with CSID.

Symptoms

Symptoms of CSID include: abdominal pain, gas, bloating, diarrhea, failure to thrive, weight loss, and malnutrition.

General guidelines

A low-starch, low-sucrose elimination diet is recommended initially to determine tolerance to sugars and starches. After 4-6 weeks, starches can be added back to the diet slowly, with careful attention to symptoms developed after introduction of foods. Start with the foods listed as "tolerated by most," waiting 3 days between food introductions. As tolerance is established, begin to introduce "foods tolerated by some" and "foods tolerated by few" gradually.







Keep a food journal when reintroducing sugars and starches. Record the food item, amount of food consumed, time eaten, and time symptoms occurred (if any) to help determine safe foods and/or safe amounts of certain foods. Portion size is very important to record because sometimes half of a cup of a certain food may be tolerated, but eating more of that same food may cause symptoms.

Dietitian: _____






Phone: _____

Facility: _____

This handout is meant for use during an appointment with a registered dietitian only and is not intended for use by other clinicians.

FOOD GROUP	FOODS RECOMMENDED	FOODS TO AVOID
<p>Milk and Milk Products Limit to 2 cups of milk/day</p> 	<p>Allowed at all phases of the diet, including elimination phase;</p> <ul style="list-style-type: none"> • Cow's milk • Ricotta cheese • Plain cottage cheese • Sour cream • Butter • Cream • Whipped cream • Hard cheeses (cheddar, mozzarella, colby, swiss, parmesan, provolone), • Unsweetened yogurt or yogurt sweetened with dextrose or fructose 	<ul style="list-style-type: none"> • Processed cheese that may contain starch or sucrose (American cheese, Velveeta) <p>Lactose intolerance is rare, but may be present in CSID</p>
<p>Breads</p> 	<ul style="list-style-type: none"> • None 	<ul style="list-style-type: none"> • All grains and starches are avoided during the elimination phase; <p>See 'special considerations' below; add whole grains first</p>
<p>Cereals</p> 	<ul style="list-style-type: none"> • None 	<ul style="list-style-type: none"> • All cereals are avoided during the elimination phase; <p>See 'special considerations' below; add whole grains first</p>
<p>Fruits</p> 	<ul style="list-style-type: none"> • See tables below 	
<p>Vegetables</p> 	<ul style="list-style-type: none"> • See tables below 	
<p>Sweets and Desserts</p> 	<ul style="list-style-type: none"> • See tables below 	

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FOOD GROUP	FOODS RECOMMENDED	FOODS TO AVOID
<p>Potatoes and Substitutes</p> 	<p>May be added back to the diet eventually; see 'special considerations' below</p>	<p>White and sweet potatoes are not well tolerated and are avoided during the elimination phase</p>
<p>Meat and Meat Substitutes</p> 	<p>Allowed at all phases of the diet including the elimination phase;</p> <ul style="list-style-type: none"> • Meats: <ul style="list-style-type: none"> - Beef - Eggs - Fish - Lamb - Plain poultry - Pork - Tofu • Nuts and Seeds: <ul style="list-style-type: none"> - Almonds - Brazil nuts - Flax seeds - Hazelnuts - Macadamia nuts - Peanuts - Pecans - Pumpkin seeds 	<ul style="list-style-type: none"> • Bacon • Breading • Deli meats • Nuts not listed on the 'recommended' list • Processed meats gravies • Sauces • Sausage
<p>Fats</p> 	<p>Allowed at all phases of the diet including the elimination phase;</p> <ul style="list-style-type: none"> • Butter • Olive oil • Vegetable oils • Most margarines (read labels and avoid those with sucrose or starch) 	<ul style="list-style-type: none"> • Margarines with sucrose or starch
<p>Beverages</p> 	<p>Allowed at all phases of the diet including the elimination phase;</p> <ul style="list-style-type: none"> • Milk • Diet soda • Limeade or lemonade made with dextrose or fructose; soda sweetened with high fructose corn syrup may be tolerated. 	<ul style="list-style-type: none"> • All other beverages.
<p>Miscellaneous</p> 	<p>Salt is the only safe seasoning as even pepper contains a small amount of starch and sucrose.</p>	<ul style="list-style-type: none"> • Initially, eliminate all spices and seasonings except salt. • Add seasonings one at a time.

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Foods ‘tolerated by most’ are allowed during the elimination phase, though tolerance may vary among individuals. Use with caution.

Sweeteners

TOLERATED BY MOST	TOLERATED BY SOME	TOLERATED BY FEW
<ul style="list-style-type: none"> • Aspartame (nutrasweet), • Dextrose (glucose), • Fructose • Crystalline fructose • Lactose 	<ul style="list-style-type: none"> • Acesulfame-K (acesulfame-potassiumace-K) • Agave nectar • Corn syrup • High fructose corn syrup • Equal • Honey • Hydrogenated starch, • Invert sugar or syrup • Maltose • Saccharin (sweet n’ low) • Stevia (truvia) • Sucralose (Splenda) • Sugar alcohols (erythritol, lactitol, sorbitol, mannitol, maltitol, xylitol) 	<ul style="list-style-type: none"> • Beet sugar • Brown sugar • Cane juice or sugar • Caramel • Coconut sugar • Confectioner’s sugar • Date sugar • Maple syrup • Molasses • Raw sugar • Sucanat • Sucrose • Sugar • Turbinado sugar

Fruits

TOLERATED BY MOST	TOLERATED BY SOME	TOLERATED BY FEW
<ul style="list-style-type: none"> • Avocado, • Blackberries • Blueberries • Cherries • Cranberries (fresh) • Currants • Figs (raw) • Grapes • Kiwis • Lemons • Limes • Loganberries • Persimmons • Pomegranates • Rhubarb 	<ul style="list-style-type: none"> • Dates • Gooseberries • Olives • Papaya • Plums • Raisins • Raspberries • Strawberries • Watermelon 	<ul style="list-style-type: none"> • Apples • Apricots • Bananas • Boysenberries • Cantaloupes • Grapefruits • Guavas • Honeydew melon • Mangos • Nectarines • Oranges • Passion fruit • Peaches • Pears • Pineapples • Prunes • Tangelos • Tangerines (mandarin, clementine)

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Vegetables

TOLERATED BY MOST		TOLERATED BY SOME	TOLERATED BY FEW	
<ul style="list-style-type: none"> • Alfalfa sprouts • Artichoke • Asparagus • Bamboo shoots • Broccoli • Brussels sprouts • Cabbage • Cauliflower • Celery • Chard • Chicory • Chives • Cress • Cucumber 	<ul style="list-style-type: none"> • Endive • Eggplant • Green beans • Lettuce • Mung bean sprouts • Mushrooms • Okra • Peppers (red, yellow, green) • Radishes • Spinach • Tomatoes • Yellow squash (summer) • Zucchini 	<ul style="list-style-type: none"> • Arugula • Bok choy • Collard greens • Chickpeas (garbanzo beans) • Edamame • Kale • Leek • Rutabaga • Snow peas • Spaghetti squash • Turnips • Yellow wax beans 	<ul style="list-style-type: none"> • Beets • Black beans • Black-eyed peas • Butternut squash • Carrots • Cassava • Corn • Garlic • Green peas • Jicama • Lentils 	<ul style="list-style-type: none"> • Kidney beans • Lima beans • Navy beans • Onions • Parsnips • Pinto beans potato • Pumpkin • Soybeans • Sweet potato • Yams

*Artichokes, asparagus, broccoli, Brussels sprouts, cabbage and cauliflower can cause gas in all individuals, not just patients with CSID, so consumption of these should be monitored closely.

Special Considerations

- Whole grains are often easier to digest than refined carbohydrates because they are digested and move more slowly through the gastrointestinal tract. Oats, barley, bran, quinoa, brown rice, whole wheat pasta, and lentils are good foods to try after the initial 4-6 week elimination diet.
- Read labels carefully to avoid added sucrose (sugar).
- Most foods made with white flour are not well tolerated, but may be added to the diet eventually as starch tolerance improves with age and as the gastrointestinal tract lengthens.
- Chew food well – enzymes in the mouth start the digestion of starch and can help with overall digestion of starches.
- Combine high-starch foods with fatty-foods. This slows the rate of digestion and exposes the starch to pancreatic enzymes that may help with digestion.
- Avoid medications with starch or sucrose.
- When trying new foods, introduce small amounts at a time. Example: try 1 Tablespoon of cereal. If that is tolerated, increase a Tablespoon at a time.
- A Vitamin B Complex and/or Vitamin C may need to be supplemented in the diet – make sure the supplement does not contain starch or sucrose.

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Sample Meal Plan – Elimination Diet

BREAKFAST	LUNCH	DINNER
Smoothie with plain Greek yogurt, blackberries, blueberries, and cherries <p style="text-align: center;">OR</p> Omelet with tomatoes, peppers, spinach, mushrooms, shredded cheddar cheese Milk Coffee	Spinach salad with chicken, tomatoes, cucumber, radishes, avocado, almonds, oil and vinegar dressing <p style="text-align: center;">OR</p> Cabbage/lettuce wraps with chicken, celery, mushroom, peppers, salt for flavoring Limeade	Grilled pork loin or fish fillet, steamed broccoli and cauliflower, fruit salad with grapes and kiwi Milk

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