



CSID

Congenital Sucrase-Isomaltase Deficiency

A Patient and Caregiver's Guide



ALASKA NATIVE
TRIBAL HEALTH
CONSORTIUM



Program supported by a grant
from QOL Medical, LLC



CSID

Congenital Sucrase-Isomaltase Deficiency *A Patient and Caregiver's Guide*

Contents

What is Congenital Sucrase-Isomaltase Deficiency?	2
What does CSID mean for my body?	
How does CSID develop?	3
How is CSID diagnosed?	
How common is CSID?	4
Symptoms of CSID	
Managing symptoms of CSID	5
Managing CSID with medication	6
What if my infant has CSID?	7
How to determine food tolerances	8
Reading food labels	9
Medications and dietary supplements	
Traditional Foods Guide	10
Food Guide	12
Food log	15
References and resources	17



What is Congenital Sucrase-Isomaltase Deficiency?

Congenital Sucrase-Isomaltase Deficiency (CSID) is a genetic change that makes it hard for some people to digest sucrose (table sugar) and starch. “Congenital” means that a person is born with this condition.

You may also see CSID called any of the following: Genetic Sucrase-Isomaltase Deficiency (GSID), Sucrose Intolerance, Disaccharide Intolerance I, Congenital Sucrose Intolerance, or SI Deficiency. All of these names refer to the same condition.

What does CSID mean for my body?

Our bodies use enzymes to help digest, or break down, the food we eat.

Sucrase is an enzyme needed to break down sucrose.

Isomaltase is one of many enzymes needed to break down starch.

These two enzymes work together in a complex known as the **sucrase-isomaltase** enzyme complex. In a person with CSID this enzyme complex is not present in the amount the body needs to be able to completely digest sucrose and starch.

How does CSID develop?

CSID is a result of a genetic change in your body. Everyone has genes that are unique, and contain growth and development instructions. Sometimes genes develop differently and change the way the body works. The genetic change in CSID makes it hard for the body to digest sucrose and starch.

Genes are genetic data passed from parents to their children. For most Alaska Native people with CSID, the gene is passed down by both parents. Sometimes, symptoms of CSID may still occur if the gene is passed down by just one parent. If you would like more information about how CSID develops, talk to your health care provider, or you can find more information at CSIDcares.org.

How is CSID diagnosed?

Your health care provider may ask you to give a blood sample to determine if you have CSID. The blood sample will then be sent to an out-of-state lab to be analyzed. It may take 4 weeks until your provider gets the results. Children and adults can be tested this way.

There is also a breath test available that can be used to test people 18 years of age or older. The test requires drinking a sucrose solution and breathing into several tubes over two hours. These tubes will be sent to an out of state lab to be analyzed. It may take a week or longer for your health care provider to receive the results.

While you wait for your results you can learn about managing CSID with diet.





How common is CSID?

The number of Alaska Native people who have CSID is not fully known; however, it is estimated that 3-10 percent may have it. Health care providers have not been able to identify CSID in Alaska Native people for very long.

Symptoms of CSID

Symptoms occur when foods that contain sucrose and/or starch are eaten. Every person may have different symptoms, ranging from mild to severe, including:

- Watery diarrhea
- Increased gas
- Stomach pain
- Malnutrition
- Bloating
- Weight loss

In infants, symptoms often begin when table foods are introduced around 4-6 months of age. See page 7 for more information on CSID and infants.

Managing symptoms of CSID

CSID can be managed through diet by avoiding or limiting sucrose and starch intake. Every case of CSID is unique. One person with CSID may be able to tolerate different foods than another person with CSID.

A traditional Alaska Native diet is excellent for managing CSID symptoms because it is naturally low in sucrose and starch. See the Traditional Foods List on page 10.

Foods common in the Western diet can be very high in sucrose and starch.

Common sources of sucrose include:

- Baked goods
- Candy
- Jams/jellies
- Processed foods
- Some fruits and vegetables
- Sweetened beverages

Common sources of starch include:

- Bread
- Cereals
- Corn
- Crackers
- Flour
- Oats
- Pasta
- Potatoes
- Rice

For a person with CSID, eating these foods may cause diarrhea, stomach pain and/or bloating. You can work with your health care team to help determine which foods are better tolerated than others.

TIP: Chew food as much as possible. Enzymes in the mouth begin the process of breaking down starch.





Managing CSID with medication

CSID symptoms can also be partially managed with a medication called Sucraid®.

Sucraid® is an enzyme replacement that helps break down sucrose; however, it does not help break down starch. Sucraid®, in combination with diet modifications can help control symptoms of CSID.

Talk to your health care provider about a prescription for Sucraid®.

What if my infant has CSID?

Infants with CSID usually tolerate lactose. Lactose is a type of sugar found in milk and milk products. Breast milk contains lactose and does not contain sucrose or starch, making it a great option for your infant. If formula is needed, standard lactose-based infant formulas such as Enfamil® Infant, Similac® Advance®, or other similar brands, should be tolerated. Work with your health care providers to find the best formula for your infant.

Introducing table foods:

- When your infant is ready to start eating table food, begin with low sucrose and low starch foods.
- Use the food lists in this book to help guide you. Do not start with baby cereal because it is high in starch.

NOTE: Since CSID is a genetic condition it cannot be “outgrown.” Symptoms and food tolerances may change as children grow and their digestive system develops. Children with CSID may be able to eat different foods as they age.



How to determine food tolerances

CSID can affect people in different ways. Some people will be able to tolerate foods that others cannot. To determine what foods are tolerated or are not tolerated, follow these steps:

1. Start by completely eliminating sucrose and starch from the diet until symptoms have improved.

2. Determine sucrose tolerance.


- Introduce sucrose-containing foods to the diet starting with foods that are usually tolerated by people with CSID. Use the provided Food Guides to help find these foods.
- Try one new food at a time and wait three days before trying another food.
- If symptoms, such as stomach pain, bloating, or diarrhea occur, stop eating that food.

3. Determine starch tolerance.

- Most grains contain starch. Grain based starches include foods such as rice, wheat, corn, pasta, oats, and crackers.
- Some foods are high in starch, but are not a grain. These include beans, potatoes, green peas, bananas, and yams.
- Try one starch at a time starting with starches that are not a grain. Wait three days before trying another food.
- Stop eating any food that causes symptoms.

Tips:

- Keep a food journal. Write down foods that are tolerated and any foods that cause symptoms. Use the food log provided in this booklet on page 15.
- Include details such as amount of food eaten, symptoms, and other relevant information.



REMEMBER: When determining food tolerances, limit spices and seasonings as they may contain sucrose and/or starch.

Reading food labels

Reading the food label and the ingredients list is helpful in determining if a food contains sucrose and/or starch.

There are a lot of different names for sucrose and starch. It is important to become familiar with these ingredients, so CSID symptoms can be minimized.

Here are some names for **sucrose** to look out for:

- Beet sugar
- Brown sugar
- Cane juice
- Cane sugar
- Caramel
- Coconut sugar
- Date sugar
- Evaporated cane juice
- Maple syrup
- Molasses
- Powdered sugar
- Raw sugar
- Sucanat
- Sugar
- Table sugar
- Turbinado sugar
- White sugar

Here are some names for **starch** to look out for:

- Arrowroot
- Corn starch
- Dextrin
- Flour
- Food starch
- Malt
- Maltodextrin
- Modified food starch
- Potato starch
- Starch
- Tapioca starch
- Wheat starch



Medications and dietary supplements

Sucrose is an ingredient in some prescription and over the counter medications and dietary supplements. To minimize symptoms of CSID, review all medications and dietary supplements you are taking with your health care provider and pharmacist.

Traditional Foods Guide

A traditional Alaska Native diet is usually well tolerated. It is low in sucrose and starch, and high in healthy fats and proteins. Most wild game, seafood, fish, birds, and marine mammals are tolerated. Most Alaskan plants, seaweed, and berries are tolerated. Some common examples are found below.

Meat & Protein		
USUALLY TOLERATED		LEAST TOLERATED
Black bear	Octopus	<div>Abalone</div> <div>Clams</div> <div>Some considerations:</div> <div>Traditional meats, fish, and seafood that are breaded or prepared with seasonings, sauces, or any ingredients that contain sucrose and/or starch, may cause symptoms of CSID.</div> <div>Organ meats, herring eggs, and salmon eggs contain some starch and may not be tolerated, especially in large amounts.</div>
Blackfish	Ptarmigan*	
Caribou	Salmon	
Chiton/	Sea cucumber	
Gumboots	Sea lion	
Cod	Seal	
Crab	Sheep	
Duck	Smelt	
Eggs (bird)	Walrus	
Goose	Whale	
Moose	Whitefish	
*indicates sucrose and starch content has not been analyzed, but is likely tolerated based on sucrose and starch content of similar foods		

Vegetables		
USUALLY TOLERATED		LEAST TOLERATED
Beach greens*	Roseroot greens	Masru/Eskimo potato Willow leaves
Coltsfoot*	Sea lovine*	
Dandelion greens	Seaweed*	
Fireweed (leaves)	Sourdock	
Oysterleaf*		
*indicates sucrose and starch content has not been analyzed, monitor closely for tolerance		



Fruits and Fruit Dishes

USUALLY TOLERATED

Blueberry
Cloudberry
Cranberry
Crowberry (blackberry)
Rhubarb
Salmonberry

LEAST TOLERATED

Akutuq (Eskimo ice cream) if made with sugar



IMPORTANT SAFETY REMINDER:

Many plants have lookalikes that can be toxic and even poisonous. Know your traditional plants well. If unfamiliar, harvest and prepare traditional foods with an Elder or plant expert.

Other

USUALLY TOLERATED

Muktuk (whale blubber with skin)
Seal oil
Tundra tea

LEAST TOLERATED

Fry bread
Sailor Boy Pilot Bread
White rice

Food Guide

Use this guide to help figure out what foods have sucrose and/or starch in them and how they may be tolerated.

Note: In general, foods low in sucrose and/or starch are better tolerated by people with CSID. Foods that have higher amounts of sucrose and/or starch are less tolerated by people with CSID.

Fruits				
USUALLY TOLERATED		SOMETIMES TOLERATED	LEAST TOLERATED	
Avocado	Lemon	Persimmon	Apple	Mandarin orange
Blackberry	Lime	Plum	Apricot	Mango
Blueberry	Loganberry	Raisins	Banana	Nectarine
Boysenberry	Olives	Watermelon	Cantaloupe	Orange
Cherry	Papaya		Clementine	Passion fruit
Cranberry, fresh	Pear		Dates	Peach
Currants	Pomegranate		Grapefruit	Pineapple
Fig, raw	Prunes		Guava	Tangelo
Gooseberry	Raspberry		Honeydew melon	Tangerine
Grapes	Rhubarb			
Kiwi	Strawberry			

Meat & Protein		
USUALLY TOLERATED*		LEAST TOLERATED
Beef	Lamb	Breaded meat
Chicken	Pork	Meat and protein cooked with sauces and seasonings
Eggs	Turkey	Processed meat (e.g bacon, sausage, deli meat)
Fish		

*Most plain and unseasoned meats are tolerated.

NOTE: Nuts contain various amounts of sucrose and starch. Use caution when determining tolerance.

Dairy		
USUALLY TOLERATED*		LEAST TOLERATED
Butter	Milk	Flavored milk
Cottage cheese	Plain yogurt	Flavored yogurt
Cream	Sour cream	Processed cheeses (e.g. American cheese, Velveeta cheese)
Hard cheeses		

*Most plain/unsweetened dairy products are tolerated

Vegetables & Legumes

USUALLY TOLERATED		SOMETIMES TOLERATED	LEAST TOLERATED	
Alfalfa sprouts	Endive	Edamame (soybeans)	Beets	Kidney beans
Artichoke*	Green beans	Jicama	Black beans	Lentils
Arugula	Kale	Leek	Black-eyed peas	Lima beans
Asparagus*	Lettuce	Okra	Butternut squash	Navy beans
Bamboo shoots	Mung bean sprouts	Pumpkin	Carrots	Onions
Bok choy	Mushrooms	Snow peas	Cassava (yuca)	Parsnips
Broccoli*	Mustard greens	Tempeh	Chickpeas	Pinto beans
Brussels sprouts*	Peppers	Tofu	Corn	Potatoes
Cabbage*	Radishes	Yellow wax beans	Garlic	Split peas
Cauliflower*	Rutabaga		Green peas	Sweet potatoes
Celery	Spaghetti squash			Yams
Chard	Spinach			
Chicory	Tomatoes			
Chives	Turnips			
Collard greens	Yellow squash			
Cress	Zucchini			
Cucumber				
Eggplant				

*These foods can cause gas in all individuals, not just those with CSID, so consumption should be monitored closely.

TIP: If buying canned, frozen, or dried food, check the ingredients label to make sure no sucrose and/or starch has been added.

Starches

TOLERANCE VARIES		
Barley	Crackers	Pasta
Bran	Lentils	Quinoa
Bread	Oats	Rice

REMEMBER: Starch tolerance varies greatly. Caution should be used when eating these foods. Use the steps on page 8 to determine starch tolerance.



Continued on the next page

Beverages

USUALLY TOLERATED	SOMETIMES TOLERATED	LEAST TOLERATED
100% Grape juice Coffee Lemonade* Limeade* Milk Tea Unsweetened flavored water Water *Homemade with fructose or dextrose	Diet beverages Beverages with high fructose corn syrup	Apple juice Orange juice Beverages sweetened with sucrose, such as: Energy drinks Flavored milk Juice cocktails (e.g. cranberry, fruit punch) Powdered drink mixes Regular soda/pop Specialty coffees Sports drinks Sweet iced tea

Sweeteners

USUALLY TOLERATED	SOMETIMES TOLERATED	LEAST TOLERATED
Dextrose Fructose Glucose Lactose	Agave Corn syrup High fructose corn syrup Honey Invert sugar Maltose Acesulfame-K (Sweet One) Equal Saccharin (Sweet'N Low) Stevia Sucralose (Splenda) Sugar alcohols	Beet sugar Brown sugar Cane sugar/juice Caramel Coconut sugar Date sugar Maple syrup Molasses Powdered sugar Raw sugar Sucanat Sugar Turbinado sugar White sugar

TIP: Dextrose, fructose, and glucose can be purchased from online suppliers and large grocery stores. These sweeteners can be used in place of sucrose.



Food Log

[illegible]

Notes



ALASKA RESOURCE:

CSID Outreach and
Education Coordinator
csid@anthc.org
(907) 729-3628
anthc.org/csid

NATIONAL RESOURCES:

CSID Cares: csidcares.org
Sucraid Information: sucraid.net

References:

- About CSID and GSID. Retrieved January 2018 from: <https://www.sucraid.net/about-gsid>
- About CSID Archives. Retrieved December 2017 from: <https://www.csidcares.org/about/csid>
- Alaska Native Tribal Health Consortium. (2008). Traditional Food Guide for Alaska Native Cancer Survivors. Anchorage, Alaska: Office of Alaska Native Health Research, Cancer Program.
- Congenital Sucrase-Isomaltase Deficiency (CSID), (2016, December 27). Retrieved January 29, 2018, from <https://www.iffgd.org/other-disorders/congenital-sucrase-isomaltase-deficiency-csid.html>
- Fact Sheet 7 Autosomal Recessive Inheritance [PDF]. (2018 January). NSW Government.
- Food Composition Database. Retrieved May 2020 from <https://www.csidcares.org/treatment/food-composition-database>
- Jones A. (2010). Plants That We Eat: Nauriat Niġiñaqtuat. Fairbanks: University of Alaska Press.
- The Journey to a CSID Diagnosis. Retrieved May 2020 from <https://www.sucraid.com/about-csid/diagnosis>
- Marcadier, J.L., Boland, M., Scott, C., Issa, K., Wu, Z., McIntyre, A.D., ... Lines, M. A. (2015). Congenital sucrase-isomaltase deficiency: identification of a common Inuit founder mutation. Canadian Medical Association Journal, 187(2), 102-107.
- Schofield J. (1993). Alaska's Wild Plants. Portland: Alaska Northwest Books.
- Treatment Options to be Considered for CSID. Retrieved December 2017 from <https://www.csidcares.org/treatment>



Second edition 2020



ALASKA NATIVE
TRIBAL HEALTH
CONSORTIUM

3900 AMBASSADOR DR, ANCHORAGE, AK 99508

ANTHC.ORG



Program supported by a grant
from QOL Medical, LLC