

Dietary advice for Congenital Sucrose- Isomaltase deficiency

Information for patients, parents,
and carers



leeds children's
hospital

caring about children

Congenital Sucrose-Isomaltase Deficiency

Congenital Sucrose-Isomaltase Deficiency (CSID) is a rare genetic disorder that impairs the ability to digest certain disaccharides, specifically sucrose and starch. In individuals with this condition, the body is unable to properly break down these carbohydrates.

Sucrose, commonly known as table sugar, is found naturally in fruits and vegetables, while starches are carbohydrates which are present in grains and tuberous foods like potatoes.

Children with CSID often present with symptoms that can mimic irritable bowel syndrome (IBS), including abdominal pain, bloating, diarrhoea, and occasionally nausea and vomiting. These symptoms result from the body's inability to digest and absorb the affected sugars properly.

Diagnosing CSID

CSID often becomes evident during infancy, particularly after the introduction of fruits and grains into the diet. Ingesting sucrose and maltose can trigger rapid gastrointestinal reactions, including cramps, bloating and diarrhoea.

In some cases, these symptoms may contribute to poor weight gain or failure to thrive. However, many children with CSID tend to develop better tolerance of sucrose and maltose as they grow older.

The diagnosis of CSID is confirmed through a small intestinal biopsy, where a test called a disaccharidase assay is performed. This assay measures the activity levels of key enzymes, including sucrase, maltase, and lactase, to identify any deficiencies that would indicate CSID.

CSID treatment and management

CSID cannot be cured, and nutritional management remains the primary treatment. The severity of enzymatic deficiency varies from person to person, leading to different tolerance levels for certain carbohydrates. Whilst it is nearly impossible to completely eliminate sucrose, maltose, and isomaltose from the diet due to their presence in a wide range of foods, limiting their intake can effectively control symptoms.

Sucrose is commonly found as table sugar, which is added to processed foods such as biscuits, yoghurts, and soft drinks. Additionally, it occurs naturally in fruits and vegetables. Foods containing starch, such as bread, pasta, cereals, and potatoes, may also need to be restricted or avoided, depending on individual tolerance. However, many children experience improved tolerance of these carbohydrates by the age of three to four years.

For some individuals, enzyme replacement therapy with Invertase or sacrosidase can be beneficial. This enzyme specifically breaks down sucrose into simpler sugars that can be absorbed, though it does not affect starch digestion. This treatment can help manage dietary intake of sucrose more effectively, reducing symptoms associated with CSID.

How to reduce sucrose in the diet?

- Reduce the number of processed foods containing sugar that your child eats
- Read food labels to avoid large sources of sucrose (check labels for sugars and syrup)
- Limit the amount of high sucrose fruit and vegetables eaten

How to reduce starch in your food / meal preparations?

By incorporating culinary modifications, you can effectively reduce starch in your diet while still enjoying flavourful and satisfying meals.

1. Use vegetable substitutes

- Replace potatoes, rice, and pasta with lower carbohydrate alternatives like cauliflower rice, courgette noodles or spaghetti squash
- Try using mashed cauliflower instead of mashed potatoes

2. Incorporate more leafy greens

- Use lettuce, cabbage, or spring greens as wraps for sandwiches and tacos instead of bread or tortillas
- Add more leafy greens like spinach, kale, and Swiss chard to your meals to bulk them up without the added starch

3. Modify baking recipes

- Substitute all-purpose flour with almond flour, coconut flour, or other low carbohydrate flour alternatives in baking recipes
- Use psyllium husk or flaxseed meal to add bulk and texture to baked goods without adding starch

4. Increase protein and fat content

- Add more protein sources like chicken, fish, tofu, or legumes to your meals, which can help you feel full and reduce the need for starchy sides
- Incorporate healthy fats like avocado, nuts, seeds, and olive oil to enhance flavour and satiety

5. Use non-starchy thickeners

- Instead of using flour or corn-starch to thicken soups and sauces, use alternatives like pureed vegetables (e.g., cauliflower or courgette), Greek yoghurt, or double cream

6. Experiment with cooking methods

- Try grilling, roasting, steaming, or stir-frying vegetables instead of boiling them. These methods can help retain their nutrients and provide better texture and flavour.
- Use a spiralizer to create vegetable noodles from courgette, carrots, or cucumbers

7. Create low-starch sauces and dressings

- Make your own sauces and dressings using low carbohydrate ingredients like Greek yogurt, mustard, herbs, and spices instead of store-bought options that may contain hidden starches
- Use avocado or nut-based sauces as creamy alternatives

8. Explore new recipes

- Look for recipes specifically designed to be low-starch or low carbohydrate. There are many creative and delicious recipes available that focus on reducing starch

9. Utilize beans and legumes wisely

- While beans and legumes contain some starch, they are also high in fibre and protein. Use them in moderation and combine them with vegetables and lean proteins for a balanced meal

How long do I have to follow a low sucrose diet?

CSID symptoms can vary greatly between individuals. While some may experience relief soon after starting a restricted diet, others may require more time to see improvement. Once symptoms begin to subside, foods can gradually be reintroduced into your child's diet.

It is important to reintroduce one new food at a time, with a waiting period of two to three days between each new addition. If your child remains symptom-free, you may continue introducing more foods.

However, if symptoms recur, revert to the last food that was well-tolerated without symptoms. Maintain this restricted diet until your child is symptom-free again, and then cautiously resume the process of reintroducing new foods. This stepwise approach can help ensure that the diet remains manageable while avoiding symptom flare-ups.

The tables below show which foods to choose to help you follow a low sucrose diet.

Dairy and dairy alternatives

Foods allowed	Foods to avoid
<ul style="list-style-type: none"> • Plain, unflavoured milks • Unsweetened milk alternatives • Plain, natural or Greek yoghurt • Cheese or cream cheese • Sugar free ice cream 	<ul style="list-style-type: none"> • Flavoured or sweetened milks • Sweetened milk alternatives • Sweetened yoghurts • Ice cream, sorbet • Frozen yoghurt

Fats

Foods allowed	Foods to avoid
<ul style="list-style-type: none"> • Margarine • Butter • Lard • Vegetable oils 	<ul style="list-style-type: none"> • None

Breakfast cereals

Foods allowed	Foods to avoid
<ul style="list-style-type: none"> • Cereals with no added sugar, e.g., Weetabix, Shreddies, Shredded Wheat • Rolled oats 	<ul style="list-style-type: none"> • Cereals with added sugar • Porridge sachets or porridge pots with added sugar

Biscuits / cakes / snack bars / puddings

Foods allowed	Foods to avoid
<ul style="list-style-type: none">• Homemade low sugar biscuits, cakes, or muffins• Savoury biscuits• Sugar free jelly	<ul style="list-style-type: none">• Biscuits, cakes, muffins, pastries, snack bars with added sugar• Jelly with added sugar

Spreads

Foods allowed	Foods to avoid
<ul style="list-style-type: none">• Marmite• Honey (not suitable for children under one)• Peanut butter (not suitable for children with nut allergies)	<ul style="list-style-type: none">• Jam• Marmalade• Chocolate spread• Peanut butter (not suitable for children with nut allergies)

Drinks

Foods allowed	Foods to avoid
<ul style="list-style-type: none">• Diet, zero sugar cordial and soft drinks• Tea (with unsweetened milk)• Coffee (with unsweetened milk)	<ul style="list-style-type: none">• Full sugar cordial and soft drinks• Flavoured water• Sweetened juices• Energy drinks and isotonic drinks, e.g., Lucozade

Vegetables

Foods allowed	Foods to avoid	
<ul style="list-style-type: none"> • Most vegetables (apart from those listed in the foods to avoid section) 	<ul style="list-style-type: none"> • Parsnip • Carrot • Peas • Onion • Sweet potato 	<ul style="list-style-type: none"> • Sweetcorn • Beetroot • Okra • Legumes and beans • Lentils

Fruits – initially use fruits less than 1g sucrose per 100g

If tolerated, you may be able to include fruits higher in sucrose - discuss with your dietitian.

Less than 1g sucrose		
<ul style="list-style-type: none"> • Avocado • Bilberries • Blackberries • Blackcurrants • Cherries • Cooking apples • Damsons 	<ul style="list-style-type: none"> • Dates • Gooseberries • Grapes • Lemons • Loganberries • Lychees • Melon (cantaloupe, honeydew) 	<ul style="list-style-type: none"> • Pears • Raisins • Raspberries • Redcurrants • Rhubarb • Strawberries • Sultanas

Less than 3g sucrose		
<ul style="list-style-type: none"> • Galia melon • Grapefruit 	<ul style="list-style-type: none"> • Kiwi fruit • Passion fruit 	<ul style="list-style-type: none"> • Plums • Watermelon

Less than 5g sucrose

- Apples
- Apricots
- Clementines
- Oranges
- Satsumas

Alternative sweeteners that can be added to foods

It is advisable to prioritise natural sources of sweeteners whenever possible

- **Alternative sweeteners:** crystalline glucose, dextrose, corn syrup and crystalline fructose, honey, glucose, fructose, maltose, agave syrup.
- **Artificial sweeteners:** e.g., aspartame, sucralose
- **Natural sweeteners** e.g., Stevia
- **Sugar alcohols:** e.g., sorbitol, xylitol, mannitol (these can have a laxative effect if eaten in large quantities)

Please note: when checking labels, if the word 'sugar' is higher up the list of ingredients, the more added sugar there is in the product.

Example meal plan

Breakfast

- Porridge made with milk

Or

- Toast with margarine and marmite

Mid-morning

- One portion of fruit (less than 100g portion) from the less than 1g sucrose list: e.g., Blackberries, pear, cantaloupe or honeydew melon

Lunch

- Sandwich with cheese, meat, or marmite
- Crisps
- Natural yoghurt

Mid-afternoon

- One portion of fruit (less than 100g) from the less than 1g sucrose list: e.g., grapes, strawberries, raisins

Evening

- Chicken, beef, pork, lamb, sausages or fish
- Potatoes, rice or pasta
- Vegetables e.g., broccoli, cauliflower, green beans
- Sugar free jelly with cream



What did you think of your care?
Scan the QR code or visit bit.ly/nhsleedsfft
Your views matter



© The Leeds Teaching Hospitals NHS Trust • 1st edition Ver 1.0
Developed by: The Paediatric Dietitians and Paediatric Gastroenterology
team at The Leeds Teaching Hospitals NHS Trust
Produced by: Medical Illustration Services • MID code: M20241106_019/DG

LN005940
Publication date
03 / 2025
Review date
03 / 2028