

General Nutrition Guidelines for Glycogen Storage Disease Type I

Glycogen Storage Disease Type I (GSDI) is a genetic metabolic disorder of the liver. GSD I causes the inability of the liver to breakdown glycogen to glucose which the body uses as its main source of fuel. Glycogen is a stored form of sugar in the body. As a result of the inability to breakdown glycogen, patients with GSD are at risk for low blood sugars (hypoglycemia) during periods of fasting (i.e. between meals).

The following is a recommended general nutrition guideline for those with GSDI to help maximize blood sugar and lactic acid control, nutrition, and energy.

Carbohydrates

All carbohydrates are classified as complex carbohydrates or simple sugars. Complex carbohydrates take longer to digest than simple sugars and include foods such as breads, cereals, vegetables, grains, rice, pasta, crackers, beans (garbanzo, pinto, kidney for example). The combination of taking cornstarch and eating complex carbohydrates with each meal is important to maintain appropriate blood sugar levels. However, complex carbohydrates should be avoided if they contain added sugar, dried fruits and honey. Read the food label on each package to find brands with the lowest sugar content.

Reminder: It is recommended that **each meal needs to contain less than 5 grams of simple sugar.**

Reminder: It is recommended that each meal is **between 10- 15 grams of carbohydrates.**

Reminder: If blood sugars are between **85-100 mg/dL (4.7-5.6 mmol/L)** a snack is not needed.

<u>Cereals low in sugar</u>	<u>Sugar (grams per serving)</u>
Puffed Rice Cereal	0
Cheerios	1
Corn Chex	2
Kix, Rice Krispies	3

Simple sugars include:

Glucose, Galactose (dairy sugar), Lactose (galactose + glucose), Fructose (fruit sugar) and Sucrose (fructose + glucose).

When a person has GSDI, the enzyme that converts galactose and fructose to glucose is defective. This means that glucose can immediately be used by the body, but any sugar containing fructose or galactose cannot be used by the body. Instead, galactose and fructose are converted to unwanted glycogen stores, lactic acid, fatty acids, and uric acid which can be harmful to the body in large quantities.

Therefore, patients with GSD Type Ia or Ib should not consume foods that contain fructose and galactose as the goal is to try to limit the amount of non-usable sugars at all times.

Our team's recommended diet allows small amounts of fructose and galactose to be consumed in order to diversify the diet and improve nutrition, but it is important to keep the amount of fructose + galactose to **less than 2.5 grams per meal.**

Table 1. List of sugars that are and are not allowed for the GSDI diet.

Sugars allowed	Sugars Not Allowed
Glucose	Fructose (natural sugar found in fruit), also a component of sucrose
Dextrose	
Dextrin	
Maltose	Galactose (a component of lactose), Lactose (sugar found in milk)
Maltodextrin	Honey
Corn Syrup	High Fructose Corn Syrup
Aspartame (NutraSweet, Equal)	Brown Sugar
Acesulfame K (Sunett, Sweet One)	Invert Sugar
Saccharin (Sweet'N low, Sugar Twin)	Powdered Sugar, Confectioners' Sugar
Stevia	Maple Syrup
Sucralose (Splenda)	Molasses
Truvia	Sorghum
Barley Syrup	Sorbitol (Sugar Alcohol)
Brown Rice Syrup	Sucrose
Tapioca Syrup	
Mannitol (Sugar Alcohol)	
Xylitol (Sugar Alcohol)	

Foods and the Type of Sugar Each Contain

Fruits

Fruits contain **fructose**. Many fruits contain significant amounts of fructose and should be avoided. If fruit is consumed, it should be consumed in very small amounts and should not be consumed in a meal that includes foods that contain galactose. **Add lemon to the water you drink to prevent kidney stones.**

Vegetables

Most vegetables are low in sugar and low in calories. However, it is necessary to avoid using more than 2 packets of ketchup at a time and limiting tomato/marinara sauces on foods to no more than ¼ cup per meal because of the fructose in the tomato (remember tomatoes are actually classified as a fruit). Vegetables provide many nutrients and vitamins that we all need and should be included in the diet of every GSD Type I patient.

Milk/Dairy

With GSD I we recommend to avoid dairy products as much as possible. If milk must be consumed, it is suggested to limit the intake to 2 oz or less per day. Cow's milk may be appropriately substituted with enriched soy milk (Silk Soy, Eden soy or West Soy plus) or rice milk; however, it is important to be aware of how much sugar is in soy milk. Even some "Plain/Original" soymilks contain added sugars, making them inappropriate for a person with GSD I to consume. Therefore, the best type of soy milk to buy is the "plain, unsweetened" or soy milk that is sweetened with an artificial sweetener.

As a result of the recommended limited intake of milk and dairy, calcium and vitamin D deficiency is likely to occur without adequate substitutions. Please refer to the section titled "Multivitamins and Calcium Supplementation" for suggestions on calcium supplements.

Below are some suggested brands of appropriate dairy and dairy alternatives to consume.

- Two Good Yogurt (4oz: 1.5g CHO, 1g sugar)



- Fairlife Fat free milk (4oz: 3g CHO, 3g sugar)



- Ripple unsweetened milk (4oz: 0g CHO, 0g sugar)



- Hood Calorie Countdown (4oz: CHO, g sugar)



Meats

Most plain meat dishes are low in sugar. Avoid meats cooked in sweet sauces. It is best to cook meats using low-fat methods such as broiling, baking, grilling, steaming, and stir frying (without oil).

Fat

Dietary fat should be limited to 20-30% of total daily calories with equal distribution between monounsaturated, polyunsaturated, and saturated fats. Cholesterol should be limited to <300 mg/day. It is also encouraged to consume fish, especially oily fish, at least twice a week. Check with your dietitian to determine if your child is meeting these recommendations.

Beverages

Those that contain fructose, lactose, or sucrose should be avoided. Choose unsweetened beverages such as unsweetened tea or sugar-free beverages such as crystal light, sugar-free soft drinks, or any type of artificially sweetened lemonade, Kool-aid, or fruit punch.

Snack Ideas

Plain popcorn, Pretzels (hard and soft), Baked Corn Chips or Potato Chips, Bagels, Rice Cakes, Breadsticks, Crackers (rice, oyster, soda, and saltines, low fat Triscuits or Ritz), Melba Toast, Sugar-free Jell-O or Popsicles, Peanuts or other types of nuts (that are not honey roasted or coated with any type of sugar). It is important to read all food labels as many snack items may contain added sugar. **If you have snack ideas that you would like to share with other GSD Patients and families, please send them to the GSD Program Dietitian (contact information can be found at the end of this document).**



The amount of each sugar in common foods can be found on the following website:

<http://www.nal.usda.gov/fnic/foodcomp/search/>

http://www.waldenfarms.com/nutrition_facts.html (site for sucralose products)

Cornstarch Guidelines



Uncooked Cornstarch

Raw, uncooked cornstarch is a complex starch which provides a steady amount of glucose because it is more slowly digested than other complex carbohydrates. Given throughout the day and night, it can help keep blood sugar levels within a normal range. Cornstarch usually provides 30-45% of an individual's daily calories. A schedule will be developed just for you or your child.

Directions for mixing the cornstarch:

- 1.) Place a clean dry container on your gram scale. Press the Zero/Tare button to zero the scale with the container on it.
- 2.) Measure out the prescribed dose of cornstarch into the container. **Cornstarch doses should not be estimated using household measurements such as the Tablespoon as the measurement is not precise enough.**
- 3.) Add fluid (water or a sugar-free beverage) to the container
- 4.) Cover the container and shake well until cornstarch is dissolved into the solution

Important Notes about cornstarch:

- Ⓢ The cornstarch should be mixed with the fluid **just prior to taking it.**
- Ⓢ Heating/cooking will ruin the cornstarch by breaking it down.
- Ⓢ Mixing the cornstarch and beverage with a spoon is not recommended as cornstarch will stick to the spoon and you/your child will not receive the correct dose.

- Ⓢ Not all brands of cornstarch are created equally. In the US, the ARGO or Kingsford brand has been known to last the longest and taste the best.
- Ⓢ The cornstarch cannot be administered through continuous feeds as clogging of the feeding tube will occur.
- Ⓢ Your meal should be consumed prior to taking cornstarch to avoid feeling too full to consume your meal once the cornstarch has been provided. Food and cornstarch combined work to sustain adequate blood sugars and thus the two must be used together for optimal control.
- Ⓢ Cornstarch will go bad/stale after a period of time (approx 1 month). We do not recommend buying cornstarch in large bulk amounts that will last longer than approximately 1 month. Also once a box of cornstarch has been opened, the starch should be kept in a sealed, airtight container.

Multivitamins and Calcium Supplementation

Due to the restrictions on fruit and dairy products and because a large percentage of calories consumed come from cornstarch (which lacks vitamins and minerals and other important nutrients), patients often have very low intakes of vitamins and essential minerals. Calcium supplementation is extremely important in combination with daily multivitamin supplementation. The table below provides some calcium supplement and multivitamin suggestions. If the multivitamin you/your child takes is not listed in the below table, be sure to check with your doctor and/or dietitian if it is appropriate for GSDI.

Calcium Supplement and Multivitamin Suggestions in Type I GSD

Calcium supplement Suggestions	<ul style="list-style-type: none"> ❖ UpCal D (powder) – Global Health Products ❖ Cal-Quick (liquid) – Twin Lab ❖ Calci-Mix (powdered capsule) - Watson Pharmaceuticals ❖ CitraCal (tablet) or any type of calcium citrate ❖ Tums (tablet) Ⓢpareve – available in sugar-free ❖ Solgar Ⓢpareve (adult, in health food stores, online) ❖ Calcium Citrate is better absorbed than Calcium Carbonate
Multivitamin Suggestions	<ul style="list-style-type: none"> ❖ One-A-Day (sugar-free, children's and adult varieties available) or generic equivalent ❖ Nature's Plus Animal Parade(sugar free in health food stores, chewables only) ❖ Flintstone's Complete (sugar-free in CVS, Walgreens) ❖ Natrol for Children (chews, non-sucrose in CVS & Walgreens) ❖ Poly-vi-solⓈ (liquid for infants & children in CVS, Walgreens) ❖ Centrum or generic equivalent (chewable form in Walmart) ❖ Reviva (sucralose, maltodextrin in Sam's Club) ❖ SolgarⓈ Formula VM (tablet in health food stores) ❖ Wellesse multi(adult, sugar free liquid, online) ❖ Dr. Mercola Children's (chewable in Vitamin Shoppe) ❖ FruitVits by VitaFlo ❖ NutriStart Multivitamin Powder

Important Notes about taking Multivitamins and Calcium:

- Ⓢ Better absorption of the multivitamin occurs when it is taken with food.
- Ⓢ Do not take more than 500 mg calcium at one time. If more than 500 mg calcium supplementation needed daily, separate each dose, and take with a meal.
- Ⓢ It is not recommended to take the multivitamin at the same time as the calcium supplement.
- Ⓢ It is also not recommended to take the calcium supplement at the same time as an iron supplement, as iron inhibits the absorption of calcium, causing less to be absorbed.

Sugar Alcohols

Those with GSD types Ia and Ib have dietary restrictions on sugar consumption, particularly the consumption of fructose, galactose, and sucrose. Because sucrose is restricted, it is encouraged for our GSD Type Ia and Ib patients to consume sugar-free foods and beverages. Many of these sugar-free foods and beverages have artificial sweeteners added, mostly in the form of sugar alcohols (or polyols).

What are sugar alcohols?

Even though their chemical make-up partially resembles a sugar and an alcohol, the name is a misnomer, and they are neither a sugar nor an alcohol. They are actually considered a carbohydrate, but they aren't digested completely in the body.

Some very common types of sugar alcohols are sorbitol, xylitol, mannitol, maltitol, lactitol, erythritol, isomalt (note that most sugar alcohol names end with **-ol**). You may also see the term "Hydrogenated Starch Hydrolysates" (HSH), which is a type of sugar alcohol.

What do they do?

They are mostly used to add a sweet flavor to foods, in place of sugar. They also add bulk or texture to foods and help retain moisture. In addition, sugar alcohols do not turn brown when they are heated.

Should they be consumed in GSD Type Ia and Ib?

Due to their incomplete absorption in the body, they do not cause a rise in the blood glucose concentrations as most carbohydrates do. Since they have limited absorption, the sugar alcohol does not enter the metabolic pathways that are affected by GSD Ia and Ib.

Sorbitol is an exception, however. Our body changes sorbitol to FRUCTOSE during digestion. Therefore, if you have type Ia and Ib, please limit and/or avoid foods that have sorbitol listed in the ingredients.

It should also be noted that very high intakes of sugar alcohols may have a **laxative** effect (this warning may actually be listed on the food or beverage package).

If you have any questions, please contact the UCONN GSD Program Dietitian:

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