



## General Nutrition Guidelines For Glycogen Storage Disease Type III

Glycogen Storage Disease Type III (GSDIII) is a genetic metabolic disorder which causes the inability to break down glycogen to glucose. Glycogen is a stored form of sugar in the body. Glucose (sugar) is the main source of fuel for the body and brain.

**GSD IIIa** causes the inability of the liver and muscles to breakdown glycogen to glucose.

**GSD IIIb** causes the inability of the liver to breakdown glycogen to glucose.

As a result of the inability to breakdown glycogen, patients with GSDIII are at risk for low blood sugars (hypoglycemia) during periods of fasting. Patients with GSDIIIa also do not have the ability to access glycogen in their muscles as well. The lack of glycogen access in the muscles causes muscle damage as the muscle do not have a fuel to aid them in working.

The following is a recommended general nutrition guideline for those with GSDIII to help maximize blood sugar control, nutrition, energy, and hopefully minimize muscle damage for those with GSDIIIa.

### Protein

#### What is protein?

Protein is a macronutrient (like carbohydrate and fat) and is required for proper growth and development. In GSDIII, the most important role that protein serves is a way for the body to make glucose since those with GSDIII do not have access to stored sugars (glycogen). Protein also serves as the building blocks for our cells, is also necessary to make antibodies which help our bodies fight off illnesses, make up hormones, enzymes, and even our DNA. With GSD type III, it is very important to make sure you are getting all the protein that your body needs.

#### How much protein do I need?

You will need to follow a “high protein diet” which is defined as **3-4 grams of protein** per kilogram body weight per day. This amount of protein should be spread out throughout the entire day. For example protein should be consumed at the following times: breakfast, mid-morning snack, lunch, mid-afternoon snack, dinner, and snack before bed.

As stated above GSDIII prevents the body from having access to stored sugars and the consumption of protein will aid your body in gaining a source of sugar/fuel. Taking the recommended amount of protein throughout the day will increase your energy, improve your blood lab work, and potentially will help decrease the amount of muscle damage in type IIIa.

**What are sources of protein?**

The best sources of protein come from animals, especially meats, dairy and eggs. Plants do contain some protein; however, plants do not contain as much protein as in animal sources. It is also important to know that our body uses the protein we eat from meat, eggs, and dairy better than protein from plant sources. Other sources of protein include all dairy products, all meats, soy milk, tofu, cooked or baked beans (garbanzo, pinto, kidney, white, black eyed peas, split peas), peanut butter, and the different types of nuts. Most protein sources, with the exception of dairy products and cooked dried beans contain little amounts of carbohydrates.

**How do I know how much protein I am getting?**

**Table 1. List of foods and the amount of protein each contains**

<b>Food</b>	<b>Amount of Protein (grams)</b>
1 ounce meat	7
2 ounces meat (1 small chicken leg or thigh)	14
3 ounces meat (1 medium pork chop, 1 small hamburger, ½ of a whole chicken breast)	21
Cooked dried beans, peas, lentils (1/2 cup)	7
1 large egg	6
1 hot dog	6
1 Tablespoon peanut butter	4
2 ounces fish (tuna)	14
3 ounces fish (unbreaded fish fillet)	21
½ cup cottage cheese	14
1 slice bread	3
1/3 cup pasta	3
½ cup oatmeal	3
1 container yogurt	~7
¼ cup peanuts	9

**What are some tips on how to add extra protein to foods I like?**

- Ⓢ Add extra cheese to casseroles
- Ⓢ Use cream sauces or cream soups
- Ⓢ Incorporate meats into meatloaf, stews, casseroles, burritos, pizzas
- Ⓢ Use nuts and peanut butter in cookies
- Ⓢ Add powdered milk to baked goods
- Ⓢ Use milk to cook hot cereals
- Ⓢ Offer milk-based puddings and custards
- Ⓢ Dry roasted Edamame has 14g of protein in 1/4cup



- Ⓢ Consider adding a protein supplement
  - Carnation Instant Breakfast mixed with 8 ounces milk has ~13 grams protein
  - Beneprotein (protein powder) contains 6 gr protein in 1 scoop or 1 packet
  - Ensure, Boost, and other ready-made nutritional supplements usually contain about 7 grams protein per 8 oz
    - Many also come in the “High Protein” or “Plus” variety which offers more protein
    - Protein bars with low sugars
  - Unjury (protein powder) contains **20 grams** protein in 1 scoop or 1 packet
  - EAS Protein drink low Carbs purchased in Wal-Mart, CVS or Walgreens
  - Rice Protein found in Whole food stores
  - Egg-Pro purchase by NutraBalance
  - Quinoa and Hemp Seed (add to salads and foods)
- Ⓢ Other suggestions: taking Cornstarch (CS) doses with milk also helps meet the elevated protein needs in type III GSD.

## 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, 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. Your diet should include more complex carbohydrates and limited simple sugars, which are defined below.

### Simple sugars include:

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

Although milk and fruits contain simple sugars, they are not discouraged as they contain many important vitamins and minerals that are essential. For example, one should not omit milk or milk products they are excellent sources of calcium and vitamin D. In addition, fruits contain many vitamins and antioxidants which are an important part of any person’s diet.

Foods high in sucrose and/or high fructose corn syrup should be limited with Type III GSD. Some examples are foods such as cookies, cakes, pies, candies, doughnuts, ice cream and other desserts. It is also advised to limit the amount of high-sugar beverages such as soda, Kool-Aid, fruit juices, etc. **Reminder:** It is recommended that **each meal** needs to contain less than **5 grams of simple sugar**.



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

**Fat**

Dietary fat should be limited to 20% 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.

**Calcium Supplement and Multivitamin Suggestions**

<b>Calcium supplement Suggestions</b>	<ul style="list-style-type: none"> <li>❖ UpCal D (powder) – Global Health Products</li> <li>❖ Cal-Quick (liquid) – Twin Lab</li> <li>❖ Calci-Mix (powdered capsule) - Watson Pharmaceuticals</li> <li>❖ CitraCal (tablet) or any type of calcium citrate</li> <li>❖ Tums (tablet) – available in sugar-free</li> <li>❖ Calcium Carbonate</li> </ul>
<b>Multivitamin Suggestions</b>	<ul style="list-style-type: none"> <li>❖ One-A-Day (sugar-free, children’s and adult varieties available) or generic equivalent</li> <li>❖ Flintstone’s Complete (sugar-free available)</li> <li>❖ Natrol for Children (liquid, non-sucrose in CVS &amp; Walgreens)</li> <li>❖ Poly-vi-sol (liquid for infants &amp; Children)</li> <li>❖ Centrum or generic equivalent (liquid form in Walmart)</li> <li>❖ Reviva (sucralose, maltodextrin in Sam’s Club)</li> <li>❖ Bariatric vitamins are in liquid form</li> </ul>
<b>Omega-3 Fish Oil</b>	<ul style="list-style-type: none"> <li>❖ Kids Smart by Bioglan grape flavored (liquid, Xylitol, Sucralose in Walgreens)</li> <li>❖ Omega Smart, Kids DHA, fruit punch flavored softgel chew or swallow (Stevia leaf in Walgreens)</li> </ul>

**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.
- Ⓢ For low iron, pull out the cast iron skillets and cook in them, iron transfers into the food. Better iron absorption occurs with vitamin C foods.



## 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/or night, it can help keep blood sugar levels within a normal range. 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 or milk are suggested) 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.
- ⓐ Mixing the cornstarch and beverage with a spoon is not recommended as cornstarch will stick to the spoon.
- ⓐ 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 that period of time. Also once you open a box of cornstarch keep the starch in a sealed, airtight container.

*If you have any questions please contact the University of Florida GSD Program Dietitian:*

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