

Medical Nutrition Therapy Diet

Glycogen Storage Disease

1. Purpose

a. Nutrition Indicators: Glycogen storage diseases are caused by deficiencies of enzymes that regulate the synthesis or degradation of glycogen. (Nelms, Sucher, & Long, 2007, pg 902) There are at least eight different types of glycogen storage disease with respect to clinical and chemical manifestations and according to the enzymatic deficiency. Most of the various types affect the liver and are a consequence of deficient activity of one or the other of the enzymes directly involved in degradation of glycogen to glucose-6-phosphate. (Nelms, Sucher, & Long, 2007, pg 903)

b. Criteria to Assign the Diet: "Diagnosis usually occurs in infancy or childhood, although some milder types of GSD go unnoticed well into adulthood and old age. It is even conceivable that some of the milder GSDs are never diagnosed. The four major symptoms that typically lead a doctor to suspect GSDs are low blood sugar, enlarged liver, retarded growth, and an abnormal blood biochemistry profile. A definitive diagnosis is obtained by biopsy of the affected organ or organs. The biopsy sample is tested for its glycogen content and assayed for enzyme activity. There are DNA-based techniques for diagnosing some GSDs from more easily available samples, such as blood or skin. These DNA techniques can also be used for prenatal testing." ("Glycogen storage diseases,")

c. Rationale for Diet: "The system for glycogen metabolism relies on a complex system of enzymes. These enzymes are responsible for creating glycogen from glucose, transporting the glycogen to and from storage areas within cells, and extracting glucose from the glycogen as needed. Both creating and tearing down the glycogen macromolecule are multistep processes requiring a different enzyme at each step. If one of these enzymes is defective and fails to complete its step, the process halts. Such enzyme defects are the underlying cause of GSDs." ("Glycogen storage diseases,")

2. Population

a. Overview: There is a wide range of incidence in the occurrence of glycogen storage diseases. The more common glycogen storage disease type 1 occurs about one in every 100,000 births, while frequency of less common forms such as type II has not yet been determined. (Nelms, Sucher, & Long, 2007, pg 903)

b. Disease Process: Hepatomegaly is often present at birth and can progress to huge enlargement of the liver if the disease is poorly controlled. The enlargement is due to the accumulation of glycogen as the metabolic block compromises the ability to release free glucose. The chronic lactic acidosis with elevated glucagon and low insulin levels seems to be related to the poor growth seen in children with this condition. Bones may be osteoporotic, and some patients show delayed bone age associated with increased phosphate loss coupled with the acidosis. Longer term complications associated with this condition include liver adenoma, osteoporosis, kidney stones, renal failure, and ovarian cysts. (Nelms, Sucher, & Long, 2007, pg 904)

c. Biochemical and Nutrient Needs: Frequent oral feedings, high in carbohydrate, are recommended to maintain blood glucose levels about 70 mg/dL. Total parental nutrition or continuous nasogastric infusion of glucose correct most of the metabolic abnormalities associated with GSD I. (Nelms, Sucher, & Long, 2007, pg 904)

3. General Guidelines

a. Nutrition Rx: "The nutrition prescription for someone with diet of glycogen-storage disease type I, requires close monitoring and adjustment by a highly specialized nutritionist. The fundamental principle of diet management for these patients is maintenance of a steady-state balance between circulating glucose and existing glycogen stores. Consequently, a chief aim is to avoid excessive carbohydrates and calories while supplying adequate calories and protein for growth. Because of the triglyceridemia characteristic in this disorder, counsel the patient to avoid high lipid intake. Most biochemical parameters can be substantially normalized and liver size can be reduced by approaching glucose homeostasis by means of overnight feeding. Overnight NG feedings should be administered only by a pump equipped with an alarm in case of flow interruption. When pancreatic amylase reaches sufficient activity in children older than 2-3 years, overnight feeding is usually replaced by raw cornstarch at bedtime and early morning hours." (Roth, 2009)

b. Adequacy of Nutrition Rx: Adequate provision of CHO (gCHO/kg/hour from the overnight feed and 2-hourly daytime feeds or UCCS(check dose size and frequency of administration. Nutritional adequacy of vitamin and mineral intake (specifically calcium and vitamin D) (Shaw, & Lawson, 1994)

c. Goals: The goals of nutrition therapy for GSD1 are to prevent hypoglycemia, correct metabolic derangement, and provide optimal nutrition to support growth and development. Carbohydrates should be provided from complex sources and should provide about 60% to 70% of the total caloric intake. Protein should come from lean sources, providing 10%-15% of total kcal. Fat should provide less than 30% of total kcal. (Nelms, Sucher, & Long, 2007, pg 904)

d. Does it Meet DRI: Yes, this goal meets the DRI for a normal individuals.

4. Education Material

a. Nutrition Therapy: "The basis of nutritional therapy is feeding GSD type I patients with foods rich in starches with low concentrations of galactose and fructose during the day and to prevent hypoglycemia during the night." (Heller, 2008)

b. Ideas for Compliance: "Compliance with dietary treatment reduces the risk of developing hypoglycemia. Daytime schedules must be flexible enough to include ingestions of feedings as needed. School-age children need to carry high carbohydrate snacks with them at all times. Infants and toddlers are given formula concentrated in glucose polymers for symptomatic hypoglycemia." (Nelms, Sucher, & Long, 2007, pg 905)

5. Sample Menu

a. Foods Recommended: "The majority of medical centers recommend the use of uncooked cornstarch, mixed in water, soy formula or soy milk (sucrose, fructose and lactose free). Cornstarch should not be mixed in drinks that contain high amounts of ascorbic or citric

acid and the cornstarch drink should not be heated as this may alter its structure rendering it less effective. Cornstarch is digested slowly so it provides a steady release of glucose in between feedings. In some cases, an overnight tube feeding is required to provide a continuous delivery of glucose. Formulas should be sucrose, fructose and lactose free. The rate of the tube feeding is based on the liver's normal glucose delivery rate and the age and weight of the child. Due to the many restrictions for the GSD I diet, it is necessary to supplement the diet with a multivitamin. Additional calcium supplementation may also be required. Because the diet for Type I Glycogen Storage Disease is complex, the ideal team should include a dietitian and a physician familiar with the long-term care and maintenance related to GSD I." ("Type i glycogen," 2006)

b. Foods to Avoid: "People with GSD Type 1a are advised to avoid fruits, fruit juice, high-fructose corn syrup, sorbitol, cane juice, and other foods that cannot be broken down into glucose, and to limit their intake of dairy products." (" glycogen storage,")

c. Example of a meal plan: "Frequent feedings, meals and snacks preferably with precooked cornstarch (PCCS), continuous nocturnal gastric drip feeding (CNGDF) and administration of uncooked cornstarch (UCSS)." (Rake, 2002)

Breakfast :

1 cup oatmeal

1 piece of whole wheat toast with ½ tsp butter

2 scrambled eggs

Water

Lunch:

Baked Potato

Garden Salad, no fruit

1 cupJell-o

Water

Snack:

½ whole wheat bagel with butter

Dinner:

Spaghetti with marinara

1 cup broccoli

1 cup of lentils

Water

Night:

Do an overnight tube feeding

6. Websites

a. Organizations with Websites:

Association for Glycogen Storage Disease- <http://www.agsdus.org/>

b. Government Websites

National Institute of Health-

http://rarediseases.info.nih.gov/GARD/Condition/7864/Glycogen_storage_disease_type_1A.asp

[x](#)

Natinal Institure of Neurological Disorders and Stroke -

http://www.ninds.nih.gov/find_people/voluntary_orgs/volorg137.htm

7. References

a. Journal articles references:

glycogen storage disease type 1a . (n.d.). Retrieved from

<https://www.23andme.com/health/Glycogen-Storage-Disease-Type-1a/howitworks/>

Glycogen storage diseases . (n.d.). Retrieved from <http://medical->

[dictionary.thefreedictionary.com/Glycogen+Storage+Diseases](http://medical-dictionary.thefreedictionary.com/Glycogen+Storage+Diseases)

Heller, S. (2008). Nutritional therapy for glycogen storage diseases. *Journal of Pediatric*

Gastroenterology and Nutrition, 47(p S15-S21), Retrieved from

http://journals.lww.com/jpgn/Fulltext/2008/08001/Nutritional_Therapy_for_Glycogen_Storage_Diseases.6.aspx

Nelms, M., Sucher, K., & Long, S. (2007). *Nutrition Therapy and Pathophysiology*. Belmont, CA: Thomson.

Rake, J. P. (2002). *Guidelines for management of glycogen storage disease type i. european study on glycogen storage disease type i (esgsd i)*.. Retrieved from

<http://dissertations.ub.rug.nl/FILES/faculties/medicine/2003/j.p.rake/c6.pdf>

Roth, Karl. (2009, August 31). *Glycogen-storage disease type i: treatment & medication*.

Retrieved from <http://emedicine.medscape.com/article/949937-treatment>

Shaw, V., & Lawson, M. (1994). *Clinical paediatric dietetics* (Monitoring the diet), Retrieved from

http://books.google.com/books?id=aID9M5A_1-4C&pg=PA397&lpg=PA397&dq=glycogen+storage+disease+adequacy+in+diet&source=bl&ots=U83bjkMJkm&sig=LTrrEI0r8V6dnI5Fo2S1w-pqYE0&hl=en&ei=RWLLS5yOA4Xw9AT7zPjCBA&sa=X&oi=book_result&ct=result&resnum=10&ved=0CDgQ6AEwCQ#v=onepage&q=glycogen%20storage%20disease%20adequacy%20in%20diet&f=false

Type i glycogen storage disease type i gsd. (2006, October). Retrieved from

<http://www.agsdus.org/html/typeivongierke.htm>