



NORWICH FREE ACADEMY

Medical Center

Glycogen Storage Disease Action Plan

Name of student: _____ D.O.B. _____ Class Year: _____

Parent/Guardian name: _____ Phone number: _____

The above mentioned student has been diagnosed with Glycogen Storage Disease (GSD). GSD is an inherited disorder in which an abnormal amount or type of glycogen is stored in the liver. This abnormal storage results from the liver's inability to adequately regulate the metabolism of glycogen and glucose. "Glycogen Storage Disease occurs when an enzyme (proteins produced by the body) that regulates conversion of sugar (glucose) into its storage form (glycogen) or release of glucose from glycogen is missing" (Cincinnati Children's Hospital Medical Center [CCHMC], 2012). GSD occurs when there is an absence or deficiency of one of the enzymes responsible for making or breaking down glycogen in the body.

_____ has the following type of GSD:

___ GSD I. This condition causes the accumulation of glycogen in certain organs and tissues, especially the liver, kidneys, and small intestines, which impairs their ability to function normally.

___ GSD II. This type of GSD presents itself as a deficiency of a lysosomal enzyme. Involvement of skeletal muscles dominates the clinical presentation.

___ GSD III. This occurs due to an enzyme called the debrancher is deficient, causing the body to form glycogen molecules that have an abnormal structure. This abnormal structure also prevents the glycogen form being broken down into glucose. Liver and skeletal muscles are involved in GSD type III.

___ GSD IV. In GSD IV (amylopectinosis) glycogen that accumulates in the tissues has very long outer branches. This is due to a genetic deficiency of the branching enzyme. The main clinical features are liver insufficiency and abnormalities of the heart and nervous system.

Treatment of Glycogen Storage Disease

Treatment of Glycogen Storage Disease depends on the type of GSD. Some GSD types cannot be treated; others can be treated by controlling the presenting symptoms. For the types of GSD that can be treated, occasionally medication is ordered due to an accumulation of uric acid (a waste product) in the body. This can cause gout (painful inflammation of the joints) and kidney stones. In some types of this disease, children must limit their amount of exercise to reduce muscle cramps.

PROVIDING OPPORTUNITIES... PREPARING LIVES

It is vital that the child follow a special diet. Frequent high carbohydrate meals are recommended throughout the day. For some children, eating several small meals rich in sugars and starches every day helps prevent blood sugar levels from dropping (hypoglycemia).

For some children, giving uncooked cornstarch every four to six hours can also relieve the problem, or In order to maintain appropriate blood glucose levels, gastrointestinal tube feedings with solutions containing high concentration of glucose may need to be administered.

The student is encouraged to carry snacks to eat as needed throughout the day, and parents are advised to keep a supply of snacks in the Medical Center as well. Snacks may be eaten while in the classroom to avoid hypoglycemia. At times the child may also need to test his/her blood sugar during class since hypoglycemia can happen very suddenly. Hypoglycemia usually can be treated easily and effectively. If it is not treated promptly, however, hypoglycemia can lead to unconsciousness and convulsions and can be life-threatening. Early recognition of its symptoms and prompt treatment are necessary for preventing severe symptoms that may place the student in danger. Hypoglycemia can impair thinking abilities and sometimes can be mistaken for misbehavior. If there is sudden change in behavior, lethargy, combativeness, or unconsciousness, or if a seizure or convulsion occurs, presume that the student has hypoglycemia. Treat the situation as a hypoglycemic emergency.

Symptoms of hypoglycemia, which are different for each student and may vary from episode to episode, can include:

Mild/Moderate Symptoms

shakiness	sleepiness	changed personality
sweatiness	dizziness	inability to concentrate
hunger	confusion	weakness
paleness	disorientation	lethargy
headache	incoordination	changed behavior
blurred vision	irritability or nervousness	

Severe Symptoms

inability to swallow	having a seizure or convulsions	unconsciousness
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The student should never be left alone or sent anywhere alone when experiencing hypoglycemia.

As soon as symptoms of hypoglycemia are observed, advise the student to check his/her blood sugar level. If blood sugar results are less than 70 or he/she does not have a meter at the location to test then advise the student to take a quick-acting sugar product equivalent to 15 grams of carbohydrate as per licensed care provider's orders. (The best rule is "When in doubt TREAT.") This may include: 3 or 4 glucose tablets, 3 teaspoons (or three-fourths of a tube) of glucose gel, 4 ounces of juice, or 6 ounces (half a can) of non-diet soda. The student must recheck his/her blood glucose level 10 to 15 minutes after treatment and repeat treatment if the

blood glucose level still falls below his/her target range. Contact Campus Safety for escort to the Medical Center for further assessment and treatment.

If written authorization is obtained from the licensed care provider and parent/guardian, Glucagon may be administered when the student's blood glucose level gets so low that the student passes out, experiences seizures, or cannot swallow. Although it may cause nausea and vomiting when the student regains consciousness, glucagon can be a life-saving treatment that cannot harm a student. The student's parent/guardian should supply the school with a glucagon emergency kit. This kit usually contains a bottle (vial) of glucagon in powder form and a pre-filled syringe with special liquid; the two are mixed just before a glucagon injection is given by the nurse or trained individual.

If Glucagon is administered, 911 must be called for ambulance transport to the hospital. The parent /guardian is also contacted to notify of the student's condition and transportation to the hospital.

When documentation is provided from the student's health care provider concerning special meal requirements, this information will be shared with the school food service director. The cafeteria staff will offer food choices that are available as per the needed diet accommodations.