Ashlee Tallis

Hyperinsulinism/Hyperammonemia-HIHA

Triggers for low blood sugar:

- Protein (including any milk and cheese products, meat, eggs). Eating protein alone WILL cause blood sugar to drop. Any protein may be eaten if carbohydrates are given BEFORE (i.e. a juice box or even candy)
- Stress and/or anxiety-good or bad.
- Fasting-going for more than a few hours without eating a carb-loaded snack. Skipping or delaying snacks must not happen.
- Physical Exertion-After extreme exercise or physical activity.

***NOTE-Ashlee does not have any physical restrictions; however, we will be monitoring her after gym class as noted in her Section 504 plan of action.**

Reaction To A Low Blood Sugar

- Headache
- Extremely tired or lethargic
- Crying
- Sudden anger
- Sudden confusion
- Extremely flushed cheeks OR suddenly pale
- Nausea
- Weakness
- Glossy eyes

Please send Ashlee to the office with an adult for a blood sugar check. She must not walk down with a child or by herself, only with an adult. Treat sugar numbers according to provided outline.

Severe Reaction To A Low Blood Sugar

- Nausea with vomiting
- Severe headache
- Severe crying
- Severe lethargy
- Convulsion
- Uncontrollable twitching
- Eyes rolling

Call to office immediately to bring glucometer and glucagon to Ashlee. Please do not move Ashlee if this is happening and make sure an adult is with her at all times. Treat sugar levels according to provided outline. If any doubt about severe symptoms, call 911 immediately.

Hyperinsulinism and Insulin Reactions

We know that Hyperinsulinism/Hyperammonemia (HIHA) is caused by someone's body secreting too much insulin at the wrong time. Children with HIHA need to take medication each day and follow a specific diet. Blood sugar allows the body to feed each cell and gives energy to move and think. Our bodies normally make glucose from the food we eat. In HIHA, the secretion of insulin is not properly regulated causing excess insulin and low blood sugars. When blood sugar is altered, a child's ability to move and think is impaired.

A person with HIHA sometimes has too much insulin. This can happen when they get more exercise or activity than usual; or when they have skipped or delayed a meal or snack, or experience stress (good or bad). This is called an insulin reaction, or hypoglycemia, or LOW BLOOD SUGAR.

Blood sugar is very important to everyone's brain and nervous system. When a person with HIHA has low blood sugar, it is an emergency. Their bodies react to the emergency by releasing the hormone adrenaline or epinephrine into their blood stream.

Scientists say that our bodies react to these chemical danger alarms by FLIGHT OR FIGHT. If you are hiking in the woods and see something scary, or you feel shy before doing a speaking assignment at school, your body prepares you to have the extra energy to run away or to fight.

The body chemical in your blood might give you symptoms like shakiness or butterflies, rapid heart rate, and/or sweating, and you might look pale. The emergency chemicals are trying to raise your blood sugar so the glucose can give you the extra energy you need.

When a person with HIHA has low blood sugar, their body releases the same chemicals. Each person with HIHA may have different signs of the beginning of an insulin reaction, because everyone is different.

Signs of an insulin reaction may be unusual anger, fighting, laughing, or crying; confusion, even when doing everyday things; and also headache, sleepiness, or sudden hunger. The symptoms or signs also depend on how fast the blood sugar is dropping. If it is dropping slowly, signs are more likely to be grouchiness, confusion, sleepiness, or headaches. If the glucose is dropping fast during exercise or activity, then shakiness may be the first sign, perhaps with sweating or paleness.

Sometimes, the person with HIHA is concentrating on something interesting or fun, or they don't notice the signs of low blood sugar quickly enough. The emergency blood chemicals may make the person feel like they should use "flight or fight". Even grown-ups sometimes run away and hide, or fight people who are nearby or who try to help them. No matter how brave or smart a person is, they can be overwhelmed by hypoglycemia. This is very dangerous.

If low blood sugar is not taken care of by taking a quick-acting sugar, followed by a snack, the signs of an insulin reaction get worse. The person will become very drowsy, may fall into a coma, and/or have convulsions (seizures).

It is important that a person having an insulin reaction is not left alone, or allowed to wander off or run away.

If a child is having low blood sugar, grown-ups should be reminded that this person has HIHA and needs a drink of juice, or some candy. Sometimes grown-ups forget a child has HIHA and may react to the behavior instead of the medical emergency. This is dangerous. If they do not take care of low blood sugar, they may need to call for emergency medical help, and/or use the glucagon emergency kit to give an unconscious or unresponsive child a shot to raise their blood sugar.

Hyperinsulinism/Hyperammonemia

The symptoms of hyperinsulinism may vary from episode to episode because low blood sugar can be mild, moderate, or severe. Increasingly severe symptoms appear as the blood sugar level falls.

In healthy people, fasting blood sugar levels are usually between 70 and 99 mg/dL.

Mild hypoglycemia

Symptoms of mild low blood sugar usually occur when blood sugar falls below 70 mg/dL and may include:

- Nausea.
- Extreme hunger.
- Feeling nervous or jittery.
- Cold, clammy, wet skin and/or excessive sweating not caused by exercise.
- A rapid heartbeat (tachycardia).
- Numbness or tingling of the fingertips or lips.
- Trembling.

Moderate hypoglycemia

If blood sugar continues to fall, the nervous system will be affected. Symptoms usually occur when the blood sugar falls below 55 mg/dL and may include:

- Mood changes, such as irritability, anxiety, restlessness, or anger.
- Confusion, difficulty in thinking, or inability to concentrate.
- Blurred vision, dizziness, or headache.
- Weakness, lack of energy.
- Poor coordination.
- Difficulty walking or talking, such as staggering or slurred speech.
- Fatigue, lethargy, or drowsiness.

Severe hypoglycemia

The symptoms of severe low blood sugar develop when blood sugar falls below 35 mg/dL to 40 mg/dL and may include:

- Seizures or convulsions.
- Loss of consciousness, coma.
- Low body temperature (hypothermia).

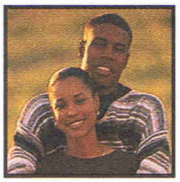
Prolonged severe low blood sugar can cause irreversible brain damage and heart problems, especially in people who already have coronary artery disease. If emergency medical treatment is not provided, severe low blood sugars can be fatal.

How glucagon can save your life.

Glucagon is a medicine that raises blood sugar and is important therapy for low blood sugar (hypoglycemia). It is given as a shot, under the skin. If your blood sugar level gets so low that you pass out or can't swallow, you will need a glucagon shot.

Every person who takes insulin should keep glucagon on hand. If you take insulin, your family, friends, coworkers, and exercise partners should learn how to give you a shot of glucagon. **If you need it, you won't be able to give it to yourself.**

Glucagon is available in a package with the supplies that are needed to take it. This package is called a **Glucagon Emergency Kit.** You need a prescription to buy glucagon or a Glucagon Emergency Kit. Ask your doctor for a prescription. The emergency kit contains a bottle of glucagon (dry powder) and a syringe filled with special liquid.



Why others need to know how to inject you with glucagon.

Show your family, friends, coworkers, and exercise partners how to use the kit. They need to know how to use it **before** you need it. They can practice giving a shot by giving you your normal insulin shots. It's important that they practice. A person who has never given a shot probably wouldn't be able to do it in an emergency.

When others should inject you with glucagon.

Family, friends, coworkers, or exercise partners should give you glucagon if:

1. You are unconscious.
2. You are unable to eat sugar or a sugar-sweetened product.

3. You don't improve after eating a sugar-sweetened product.

Whenever possible, it is important to test blood sugar before giving glucagon.

Glucagon is a safe drug. There is no danger of taking too much. However, it is for emergencies and should be used only under the direction of your doctor. As with hypoglycemia, nausea and vomiting may occur. Generalized allergic reactions have also been reported with glucagon use.

How to use the Glucagon Emergency Kit.

Here are step-by-step instructions for using the Glucagon Emergency Kit. It's important that you, your family, friends, coworkers, and exercise partners read and understand these instructions.

NOTE: Glucagon should **not** be mixed until just before it is to be injected. Important product information for glucagon is available from your healthcare professional.

How to prepare glucagon for injection.

1. • Remove the flip-off seal from the bottle of glucagon. *See Figure 1.*
   • Wipe the rubber stopper on the bottle with an alcohol swab.

1.


2. • Remove the needle protector from the syringe.
   • Inject the entire contents of the syringe into the bottle of glucagon. *See Figure 2.*

   NOTE: Do not remove the plastic clip from the syringe. This clip prevents the plunger from being pulled out of the syringe.

2.


3. • Remove the syringe from the bottle.
   • Shake the bottle gently until the glucagon dissolves and the solution becomes clear. **Glucagon should not be used unless the solution is clear and of waterlike consistency.** *See Figure 3.*
   • Glucagon should be injected immediately after mixing.

3.


How to inject glucagon.

Glucagon is injected the same way as insulin.

4.


1. • Using the same syringe, withdraw all of the solution (1 mg mark on the syringe) from the bottle. For children weighing less than 44 lb (20 kg), withdraw half the solution from the bottle (0.5 mg mark on the syringe). *See Figure 4.*

2. • Cleanse injection site on buttock, arm, or thigh with an alcohol swab.

5.


3. • Insert the needle into the loose tissue at the cleansed injection site.
   • Inject all of the glucagon solution. **There is no danger of overdose.** *See Figure 5.*
   • Apply light pressure at the injection site and withdraw the needle.
   • Press an alcohol swab against the injection site.

4. • Turn the patient on his/her side. When an unconscious person awakens, he/she may vomit. Turning the patient on his/her side will prevent choking.

5. • **Feed the patient as soon as he/she awakens and is able to swallow.** Give the patient a fast-acting source of sugar such as a regular soft drink or orange juice and a longer-acting source of sugar such as crackers and cheese or a meat sandwich.
   • If the patient does not awaken within 15 minutes, give another dose of glucagon and **obtain medical help immediately.**

   WARNING: The patient may be in a coma from severe hyperglycemia (extremely high blood sugar) rather than hypoglycemia. In such a case, the patient will NOT respond to glucagon and will require immediate medical attention.

6. • Even if glucagon revives the patient, his/her doctor should be notified promptly. A doctor should be told whenever severe hypoglycemic reactions occur.

Be prepared! Make sure your family and friends have emergency phone numbers, such as those for your doctor and the nearest emergency room. Also, be sure to take your Glucagon Emergency Kit any time you go away on a business trip or a vacation.

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