Pediatric Endocrinology Fact Sheet
Ketotic Hypoglycemia: A Guide for Families

What is ketotic hypoglycemia?

Ketotic hypoglycemia (low blood sugar) is the most common type of hypoglycemia in toddlers. Ketotic hypoglycemia is the term used for episodes of low blood sugar with elevated blood or urine ketones occurring in some children if they have not eaten over a long period of time or when ill. It almost always goes away when the children are a little older and almost never causes any permanent harm.

The main fuel for the body is a sugar called glucose. Glucose comes from the breakdown of carbohydrates that we eat, such as sugars, breads, cereals, and pasta. In healthy adults and children, after food is digested, the body stores the extra carbohydrate in the liver, muscle and fat. Some of this stored carbohydrate will be broken down into glucose and released into the blood stream to keep blood glucose at a normal level between meals. During longer periods of fasting when the body starts running out of stored carbohydrate, it will release other stored energy from fat. Before fat is used for energy it is broken down into smaller chemicals. Some of these chemicals are called free fatty acids. Free fatty acids can be broken down even more to form ketones. Ketones can also be used for energy. The ketone level can be measured in blood and urine samples.

The body always tries to keep blood glucose levels in the normal range. If the blood glucose drops below normal levels (less than 70 mg/dL), this is called hypoglycemia. Healthy children and adults usually keep blood sugar above 70 mg/dl while fasting, but children with ketotic hypoglycemia cannot always do so, especially when ill or eating poorly. Effects of hypoglycemia can produce symptoms including sluggishness, tiredness, irritability, shakiness, becoming unconscious, or seizures.

Children with ketotic hypoglycemia develop both low blood glucose and high levels of ketones after 6-12 hours of fasting, and sometimes aren't hungry or start vomiting as a result of the ketones. Most children outgrow this condition by 5-6 years of age. Children who still have hypoglycemia after this age are more likely to have an underlying and more serious problem.

How is ketotic hypoglycemia diagnosed?

Ketotic hypoglycemia often is seen when a toddler has not eaten for many hours due to illness, especially a vomiting illness. Ketotic hypoglycemia is usually suspected after a toddler has had an episode of severe tiredness or unresponsiveness and is taken to an emergency department for testing. Sometimes the parents will smell ketones on their child's breath—they smell like acetone nail polish remover or rotten apples. A blood glucose measurement less than 70 mg/dL at the time of symptoms proves the diagnosis of hypoglycemia. Blood and urine tests will show the presence of ketones and sometimes signs of dehydration during the hypoglycemia. Other blood tests are usually normal. Symptoms will go away if the child is able to eat or drink something containing carbohydrates (sugar) or receives fluids containing glucose given into a vein.

Ketotic hypoglycemia is the most common cause of low blood sugar in an otherwise healthy toddler or young child, however a few children may have a more serious condition. Pediatric

endocrinologists sometimes recommend additional tests to check for this possibility. Many of these tests must be done at the time of a low blood glucose. If these low blood sugar spells keep happening or there are other clues to suggest another problem (slow development and learning, poor growth, an enlarged liver, or a slow recovery from low blood sugar) a pediatric endocrinologist may recommend additional testing.

How is ketotic hypoglycemia treated?

There is no specific treatment for ketotic hypoglycemia except for giving sugar. Luckily, the most severe hypoglycemic spell for most children is usually their first one. Families should learn when ketotic hypoglycemia might develop and how to check blood glucose levels in these situations. It is important to recognize that the strips used to check blood glucose and blood and urine ketones go bad fairly quickly and will give the wrong answers when used, so once a bottle of strips is opened it should be replaced after a month.

If you suspect your child may be having an episode of hypoglycemia, check your child's blood glucose. If it is less than 70 mg/dl, your child should be given juice, candy or other sugar-containing food or drink to raise the blood glucose. The child's blood glucose should increase within 15-20 minutes after eating or drinking something containing sugar. If your child's blood glucose does not improve or your child cannot eat or drink because of vomiting, tiredness, or seizures, then your child should be brought to the closest emergency room for intravenous fluids containing dextrose (sugar). Your doctor can give you a letter to explain to the emergency room what your child needs.

How is ketotic hypoglycemia prevented?

A child who has had one spell of ketotic hypoglycemia may have another one. You can do several things, as suggested by your pediatric endocrinologist, to reduce the risk of another spell:

1. Limit how long your child is allowed to fast. For example, sleeping in on weekends without eating breakfast may need to be limited.
2. If your child is ill, it is important to offer sips of sugar-containing beverages to avoid long periods of time without glucose.
3. Check blood glucose levels during illnesses to be sure it is staying above 70 mg/dl. Cake icing, regular soda, juice, popsicles, are examples of sugars that can help maintain blood glucose levels and decrease the risk for developing high ketones. This helps prevent ketones causing nausea or vomiting.

What causes ketotic hypoglycemia?

The cause of ketotic hypoglycemia in most children is unknown. Children with ketotic hypoglycemia have two problems: (1) they tend to use up energy stored in the liver and switch to making ketones for energy sooner than other children, and (2) they are sometimes unable to use stored fat and muscle energy effectively to keep their blood sugar up. These problems usually improve as children get older and become more grown up in the way the store and use fules like carbohydrate and fat.