Friday, February 02, 2007

KETOGENIC DIET

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The ketogenic diet is a special high-fat, low-carbohydrate diet that helps to control seizures in some people with epilepsy. It is prescribed by a physician and carefully monitored by a dietitian. It is more strict, with calorie, fluid, and protein measurement and occasional restriction than the modified Atkins diet, which is also used today.

The name ketogenic means that it produces ketones in the body (keto = ketone, genic = producing). Ketones are formed when the body uses fat for its source of energy. Usually the body usually uses carbohydrates (such as sugar, bread, pasta) for its fuel, but because the ketogenic diet is very low in carbohydrates, fats become the primary fuel instead. Ketones are not dangerous. They can be detected in the urine, blood, and breath. Ketones are one of the more likely mechanisms of action of the diet; with higher ketone levels often leading to improved seizure control. However, there are many other theories for why the diet will work.

Who will it help?

Doctors usually recommend the ketogenic diet for children whose seizures have not responded to several different seizure medicines. It is particularly recommended for children with the Lennox-Gastaut syndrome.

Doctors seldom recommend the ketogenic diet for adults. However, in the limited studies that have been done, the diet seems to work just as well, although it is very restrictive for most adults. Studies are underway to evaluate the modified Atkins diet in this population.

The ketogenic diet has been shown in case reports and case series to be particularly effective for some epilepsy conditions. These include infantile spasms, Rett syndrome, tuberous sclerosis complex, Dravet syndrome, Doose syndrome, and GLUT-1 deficiency. Using a formula-only ketogenic diet for infants and gastrostomy-tube fed children may lead to better compliance and possibly even improved efficacy. The diet works well for children with focal seizures, but may be less likely to lead to an immediate seizure-free result. In general, the diet can always be considered as long as there are no clear metabolic or mitochondrial reasons not to use it.

What is it like?

The typical ketogenic diet, called the "long-chain triglyceride diet," provides 3 to 4 grams of fat for every 1 gram of carbohydrate and protein. The dietician recommends a daily diet that contains 75 to 100 calories for every kilogram (2.2 pounds) of body weight and 1-2 grams of protein for every kilogram of body weight. If this sounds complicated, it is! That's why parents need a dietician's help.

A ketogenic diet “ratio” is the ratio of fat to carbohydrate and protein grams combined. A 4:1 ratio is more strict than a 3:1 ratio, and is typically used for most children. A 3:1 ratio is typically used for infants, adolescents, and children who require higher amounts of protein or carbohydrate for some other reason.

The kinds of foods that provide fat for the ketogenic diet are butter, heavy whipping cream, mayonnaise, and oils (e.g. canola or olive). Because the amount of carbohydrate and protein in the diet have to be restricted, it is very important that the meals be prepared carefully. No other sources of carbohydrates can be eaten. (Even toothpaste might have some sugar in it!). For this reason, the ketogenic diet is supervised by a dietician. The parents and the child become very familiar with what can and cannot be eaten.

What happens first?

Typically the diet is started in the hospital. The child usually begins by fasting (except for water) under close medical supervision for 24 hours. For instance, the child might go into the hospital on Monday, start fasting at 6 p.m. and continue to have only water until 6 a.m. on Tuesday. The diet is started at that point, either by slowly increasing the calories or the ratio. This is the typical Hopkins protocol. There is growing evidence that fasting is probably not necessary for long-term efficacy, although does lead to quicker onset of ketosis. The primary reason for admission in most centers is to monitor for any increase in seizures on the diet, ensure all medications are carbohydrate-free, and educate the families.

Does it work?

Several studies have shown that the ketogenic diet does reduce or prevent seizures in many children whose seizures could not be controlled by medications. Over half of children who go on the diet have at least a 50% reduction in the number of their seizures. Some children, usually 10-15%, even become seizure-free.

Children who are on the ketogenic diet continue to take seizure medicines. Some are able to take smaller doses or fewer medicines than before they started the diet, however. The time when medications can be lowered depends on the child and the comfort level of the neurologist. Evidence suggests it can be done as early as the diet initiation period safely in many circumstances.

If the person goes off the diet for even one meal, it may lose its good effect. So it is very important to stick with the diet as prescribed. It can be especially hard to follow the diet 100% if there are other children at home who are on a normal diet. Small children who have free access to the refrigerator are tempted by "forbidden" foods. Parents need to work as closely as possible with a dietician.

Are there any side effects?

A person starting the ketogenic diet may feel sluggish for a few days after the diet is started. This can worsen if a child is sick at the same time as the diet is started. Make sure to encourage carbohydrate-free fluids during illnesses.

Other side effects that might occur if the person stays on the diet for a long time are:

- kidney stones
- high cholesterol levels in the blood
- dehydration
- constipation
- slowed growth or weight gain
- bone fractures

Because the diet does not provide all the vitamins and minerals found in a balanced diet, the dietician will recommend vitamin and mineral supplements. The most important of these are calcium and vitamin D (to prevent thinning of the bones), iron, and folic acid.

There are no anticonvulsants that should be stopped while on the diet. Topamax (topiramate) and Zonegran (zonisamide) do not have a higher risk of acidosis or kidney stones while on the diet. Depakote (valproic acid) does not lead to carnitine deficiency or other difficulties while on the diet either. Medication levels do not change while on the diet according to recent studies.

How is the patient monitored over time?

Early on, the doctor will usually see the child every 1-3 months. Blood and urine tests are performed to make sure there are no medical problems. The height and weight are measured to see if growth has slowed down. As the child gains weight, the diet may need to be adjusted by the dietician.

Can the diet ever be stopped?

If seizures have been well controlled for some time, usually 2 years, the doctor might suggest going off the diet. Usually, the patient is gradually taken off the diet over several months or even longer. Just as happens if seizure medicines are stopped suddenly, seizures may become much worse if the ketogenic diet is stopped all at once. Children usually continue to take seizure medicines after they go off the diet. In many situations, the diet has led to significant, but not total, seizure control. Families may choose to remain on the ketogenic diet for many years in these situations.

Where can I find out more information about the diet?

Other than the internet, there are several books about the ketogenic diet available. One is The Ketogenic Diet: A Treatment for Children and Others with Epilepsy, by Drs. Freeman and Kossoff, which discusses the Johns Hopkins approach and experience. The Charlie Foundation at www.charliefoundation.org and Matthew's Friends at www.matthewsfriends.org are parent-run organizations for support.

This site was updated by Dr. Eric Kossoff on July 19, 2006

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