

The Ketogenic Diet

Introduction:

The Ketogenic Diet is a high fat diet which appears to benefit some people with epilepsy, especially children. It is not a magic cure but one alternative to the various anti-epileptic medications currently available. The ketogenic diet offers the advantage of improved seizure control for some children, and in some cases, improved mental alertness with fewer medications.

The ketogenic diet is often regarded as a difficult regimen to follow, however, with practice, and an understanding what the diet aims to achieve, it can be reduced to a manageable routine. The basic aim is to switch the body's primary fuel source from carbohydrates (like bread and sugar) to fats. This is done by increasing the intake of fats and greatly reducing the intake of carbohydrates. The real difficulty is that the diet is so restrictive, that all foods eaten must be weighed out to a tenth of a gram during meal preparation, and a participant may not eat anything which is not "prescribed" by the dietician. The level of carbohydrates allowed is very low so that even the small amount of sugar in most liquid or chewable medications will prevent the diet from working.

As examples, a typical meal might include some type of meat with green vegetables cooked with a mayonnaise sauce or a lot of butter. Heavy cream may be included on the side for drinking. Another meal might consist of bacon and eggs with a lot of butter or oil added, and heavy cream to drink. A very high ratio of fats to carbohydrates must be maintained with a low total calorie intake for the diet to be successful.

But isn't fat bad for you?

Fats have been the subject of a lot of bad press in recent years. In addition many "healthy" foods are advertised with a low fat content...after all "cholesterol kills" doesn't it? The reality is a lot more complex. It is true that too much fat can lead to arteriosclerosis (blockages of the blood vessels), which can lead to heart attacks or strokes, but fats also have an important role to play in nutritional health. Even cholesterol in controlled amounts is necessary and not as bad as people have been taught. The role of fats and their dietary importance is discussed fully in the section *Understanding the Ketogenic Diet*. For the moment, be reassured, the high fat content of the ketogenic diet has not been shown to be harmful.

Historical Perspective:

The ketogenic diet is not a new treatment. Throughout history it has been recognized that if a person with epilepsy stops eating (fasts) their seizures generally stop. The first scientific study on fasting for the treatment of epilepsy was done in France in 1910. This study reported that seizures stopped during absolute fasting. Later, other investigators also observed cessation of seizures and improvement in mental activity during starvation, however, a person can't fast indefinitely. Therefore, in 1921 Dr. Wilder, at the Mayo Clinic, tried using a ketogenic diet to treat patients with epilepsy. He had been using this same diet to prolong ketosis in diabetic patients. At about the same time Drs Howland and Gamble at the Johns Hopkins Department of Pediatrics observed that "prayer and a water diet which also involved starvation for three to four weeks" reduced seizures in a nephew of a professor of pediatrics. Other investigators like Dr's Lennox and Cobb at Harvard University also began to study the ketogenic diet.

By 1924, Dr. Peterman, at the Mayo Clinic, was using the diet regularly, and the treatment became widely used in the 1930s. After World War II, Dr. Livingston at Johns Hopkins studied almost 1000 patients using the ketogenic diet and reported excellent seizure control. However, as newer and more effective anti-seizure medications were introduced, interest in the ketogenic diet declined.

Toward the end of the 1980s, interest in the diet was revived by Dr. John Freeman at Johns Hopkins, who reported a study in 1992 showing that the diet produced complete seizure control in 30% of children with previously uncontrolled seizures. One of the children treated successfully by the Johns Hopkins team was Charlie Abrahams. In gratitude, his parents have created the Charlie Foundation, which has given widespread publicity to the diet, in part by making available a free video tape.

How does the Ketogenic Diet work?

The food we eat provides the fuel used by our bodies for everyday activities, and the materials needed to help the body grow. Unlike cars which can only run on gasoline, the body is designed to use three main fuels including carbohydrates, fats, and protein. Carbohydrates are the primary component of sugars, starch, and flour which come mostly from plants. Fats come in two broad types: saturated fats, like butter, which mostly come from animals, and unsaturated fats, like corn oil, which mostly come from plants. Finally, protein comes mainly from animals and is represented by meat and fish. The preceding is a broad generalization and there are many variations, such as nuts which contain more than 50% fat.

Carbohydrates, fats, and proteins all undergo the same type of chemical reaction with the oxygen we breathe to produce energy for the body, and waste products including carbon dioxide and water. This is the same reaction seen when a car engine burns gasoline, or when wood or coal are burned for heat, etc. Although all three of the body's fuels are metabolized in the same way, carbohydrates are used preferentially, followed by fats then proteins.

Carbohydrates are used preferentially because they are usually readily available in most people, and the body can metabolize them quickly for energy. Athletes often eat some form of high carbohydrate snack prior to an athletic event to provide extra energy. Typically carbohydrates will be used within a few hours after they are eaten, which is one reason why we eat so frequently. Unused carbohydrates are stored in the liver in the form of glycogen, or converted to fat.

In contrast, the primary role of fats is to store energy. Animals fatten up to prepare for winter. The body normally stores the fats we eat but, if there isn't enough carbohydrate available, the body will break down the fat stores to use as fuel. Fats are metabolized much more slowly and typically it will take a day or so for the fat content of a meal to be used. This is why people feel fuller after a fatty meal as opposed to a low carbohydrate meal.

The third fuel, protein, is primarily used to build and replenish body materials; any excess protein is metabolized as fuel, or excreted. If carbohydrate and fat stores are depleted, the body will begin to breakdown muscle to metabolize the protein for fuel.

In a typical western diet, the proportion (by weight) of the three fuels used will be about 5 – 15% protein, 10 – 20% fat, and 65 – 85% carbohydrate. Any excess “fuel” will be stored as fat by the body, or excreted. By contrast, in the ketogenic diet, the proportion of fats is raised significantly and the proportion of carbohydrate is greatly reduced. It is also necessary to control the total intake of food, since if the body is given excess, it will discard the fats preferentially in an attempt to get back to its preferred balance of fuels. By restricting the total caloric intake, the body is forced to metabolize fat in place of carbohydrate.

The ketogenic diet mimics a starvation or fasting state by denying the body the carbohydrate it requires to function normally, and forcing it to metabolize fat. As the fat is metabolized ketone bodies are produced. It is the production of the ketone bodies which appears to play a central role in the success of the ketogenic diet. When the body begins producing ketone bodies it is referred to as the body being in ketosis. It usually takes 3 – 5 days for the body to go into ketosis after starting the diet. Ketosis is readily recognized, because the ketones can be detected in the urine, and can be recognized by a characteristic smell of the individual’s breath. The prophylactic properties of the ketogenic diet build up with time and it may take several weeks before the full effect of ketogenic diet is achieved.

Who can use the diet?

The diet has mostly been used for children between the ages of one and six years. The diet can be used in adolescents and adults, but is more difficult to maintain in older age groups because of social factors, and the tendency to cheat.

Review of the medical literature (1276 patients totaled from multiple studies) suggests 49.5% of patients will have a greater than 90% reduction in seizure activity. 27% will have a greater than 50% reduction in seizures, and the remainder will have a less than 50% reduction. These results are surprisingly good given two important factors:

1. Compared to taking medications, the ketogenic diet is much more difficult to maintain.
2. The diet is usually used as a last resort in patients who failed essentially all available medications.

Based on the medical literature, individuals with primary generalized seizure types tend to respond better than those with complex partial seizures with or without secondary generalization.

What is a person allowed to eat on the Ketogenic Diet diet?

The ketogenic diet is a precisely prescribed diet. The classical ketogenic diet is calculated at a 4 to 1 ratio, which means it contains four times as much fat by weight as carbohydrate and protein combined. It is important to know that a gram of fat produces more than twice the number of calories produced by a gram of carbohydrate or protein. Therefore the amount of food prepared for each ketogenic diet meal is a much smaller volume than a regular meal. In addition, the total caloric intake is restricted to about three quarters of normal, making the volume appear even less. The amount of calories and the ratio of fats to protein and carbohydrate for each meal are calculated on a case by case basis. Calculations for each day’s meals are based on a participant’s

age, height, weight, and activity level. The calculations and menu plans for the ketogenic diet require the expertise of a registered dietician who is familiar with the diet, and should never be attempted by an unlicensed individual. The dietician makes every attempt to design the menus to resemble the types of foods the participant normally eats, and likes. This is not completely possible in all cases, especially if the participant's favorite foods are almost exclusively made up of foods high in carbohydrates.

The ketogenic diet alone is deficient in vitamins and calcium. Participants are, with few exceptions, prescribed a calcium and vitamin supplement which they will need to take during the time that they remain on the ketogenic diet.

How is a person begun on the Ketogenic Diet?

The diet is normally begun during a 3 to 5 day hospitalization. On the morning of admission we ask the participant to skip breakfast, and begin the "Day 1 Ketogenic Diet Egg Nog or Formula". The day 1 egg nog is at one third of the recommended fat calorie intake targeted. On day 2 the individual will be given egg nog with 2/3 the fat calorie intake targeted. On day 2 the participants parent, guardian, or direct care staff member will be taught how to prepare a regular ketogenic meal, which the participant will eat for lunch if they are handling the diet well to that point. For individuals who are bottle or G-tube fed, the parent, guardian, or direct care staff member will be taught how to mix the ketogenic diet formula. On the morning of day 3 the participant should be to the full ketogenic diet meal or formula which they will remain on for the two year duration. If they are continuing to do well, they are discharged around lunch time.

During the hospitalization, the participant has their blood sugar checked every six hours to watch for hypoglycemia, and treat if indicated. We anticipate that as the body's carbohydrate stores are used up their blood sugar will drop to around 40. At this point, the body's fat stores begin to be metabolized, and the blood sugar levels go back up to the normal range. In addition, whenever the participant urinates we check the urine for the presence of ketones, and check a specific gravity. The specific gravity tells us how concentrated the urine is, and is a good indicator of whether the participant is becoming dehydrated. In some individuals the high fat content of the meals may lead to nausea, and refusal to eat or drink. This can become a bad situation as the individual becomes ketotic as the subsequent ketosis can make the nausea even worse. The nurses will place a heplock (a port for IV access) when the participant is admitted, so that if the individual begins to show evidence of dehydration we can intervene before they are really sick.

Once the Ketogenic Diet is started, how long does it take to work?

The effectiveness of the Ketogenic Diet is variable from person to person. In some the response is immediate once ketosis develops. In others the full effect may not be seen for up to 3 months later. In most individuals, we can predict after one month if the diet is going to be helpful or not.

How long will my child remain on the diet?

The participant is kept on the full ketogenic diet prescription for 2 years. During the third year the dietary ratio is gradually changed back to a normal diet.

When will the medications be stopped?

We don't make any changes in the medications for the first month on the diet. This gives us time to see if the diet is going to be helpful for the participant, and gives the care provider the chance to develop a routine and become comfortable with preparing the meals. Once we can determine as a group (MD, dietician, family, and participant) whether the diet should be continued, we will begin to decrease one of the participant's medications.

Does my child need to return to the clinic during the time I am on the diet?

We ask everyone to return to the clinic on a quarterly basis for evaluation of the status of their seizure control, to perform blood testing, and to re-check their weight. We discuss with the participant, family, or direct care staff if they have any specific questions.

The participant, family, or guardians have the right to request discontinuation of the diet at any time during the course of treatment.

Are there any side effects to the Ketogenic Diet?

Side effects to the ketogenic diet include:

1. Constipation: This is seen in almost everyone who is begun on the diet. We have ways of combating the problem, so that it has never been a reason to discontinue the diet.
2. Hypoglycemia: This can be a problem in children who are independent and active. The family is taught the symptoms of hypoglycemia, and what to do if you suspect the participant is hypoglycemic. If the participant becomes hypoglycemic frequently the dietician should be contacted, to adjust the diet.
3. Hypercholesterolemia: The diet is very high in fat, and in some people, they are unable to metabolize the fat efficiently and their lipid (cholesterol and triglyceride) levels go up. We may recommend a mineral supplement, or attempt to adjust the menus to lower the serum lipid levels. If the lipid levels do not return to an acceptable level, the diet is discontinued.
4. Hyperacidosis: In some participants, if they become ill or overly active resulting in dehydration, their ketones may become too high. In this situation, they may feel nauseated and refuse to eat. They may have some vomiting associated. Treatment would involve re-hydration. If the problem is recurrent, the diet may need to be adjusted or Bicitra given.
5. Osteoporosis: The diet is known to provoke osteoporosis. Everyone who starts on the diet is expected to take a vitamin and calcium supplement to lessen the severity of this problem.
6. Kidney Stones: Many ketogenic diet programs around the country restrict fluid intake for their participants. In New Mexico, we have chosen not to restrict fluids, and have rarely experienced problems with participants developing kidney stones. However, if an individual begins complaining of severe back pain, or difficulty with urination, or blood in their urine, this should be considered. We may recommend Bicitra to try and combat the problem, but if kidney stones recur, the diet may need to be discontinued.

7. **Pancreatitis:** The pancreas is an organ located in the back of the abdomen near the kidneys. The pancreas primary responsibility is to break down fat. In some people, the pancreas can not handle the increased fat of the ketogenic diet. In these instances, the pancreas becomes swollen or inflamed and the entire gastrointestinal (GI) system stops working. The patient experiences severe abdominal discomfort and the abdomen is very tender to touch. Vomiting is often associated. In this situation, the patient needs to be seen by medical personnel immediately for diagnosis and treatment. If a person develops pancreatitis on the ketogenic diet, the diet is stopped immediately, and is never attempted again. Pancreatitis can be fatal.

Summary:

Thank you for your interest in our program. If you choose to pursue the ketogenic diet as an option for treatment, please contact us by calling:

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