



Eating Well for Epilepsy

The benefits of the ketogenic diet for children with intractable seizures.

BY ANDREW WILNER, M.D.

From early childhood, John's* life has been full of challenges. When he was just six hours old, his blood sugar dropped and he was rushed to the intensive care unit. He was later diagnosed with cerebral palsy (CP) with underlying epilepsy. Doctors aren't sure what caused John's CP, but they do know that this type of CP is associated with a greater than 50 percent chance of developing epilepsy as well.

John's arms and legs are weak and spastic. He has trouble eating, can't lift his head, and doesn't speak yet. But he also experiences the broad spectrum of emotions and thoughts that all kids experience.

"John is what they call a 'trapped kid,' because he doesn't have the usual tools to express himself," says John's mother, Deborah. "But he has good receptive language, a full range of emotions, likes, dislikes, and episodes of jealousy with his younger brother. He communicates through a computer by hitting a large switch [similar to a mouse] with his hand. He goes to therapy and school every day—a special school for children with cerebral palsy."

When he was 13 months old, John had his first seizure.

"It was Christmas Day 2004," recalls John's father, Paul. "Just after putting him to bed for a nap, my wife and I heard grunting sounds. His left side was twitching, his eyes were blinking, his head moved from left to right, and he was foaming at the mouth. We didn't know what was happening. We called 911 and they came and took him to the ER. We only found out at the hospital from the ER doctor that this was a seizure."

Since then, John has had only 13 seizures, but most of them were episodes of status epilepticus, a life-threatening condition that requires emergency treatment. While most seizures stop after a minute or two, status epilepticus lasts more than 30 minutes: One seizure follows another without recovery in between.

John's first seizure lasted an hour and a half, and the second lasted two hours. Both required hospitalization and use of a respirator. Since then, his parents have been able to stop several of the seizures at home with rectal diazepam (Diastat), but others have required admission to intensive care. After an episode of

status epilepticus, John is lethargic, and it can take as long as two weeks for him to get back to his normal function.

EPILEPSY IN CHILDREN

Epilepsy is a condition of recurrent seizures, which are abnormal electrical discharges in the brain. Seizures may appear as staring spells, episodes of confusion, convulsions, or a number of other symptoms. Epilepsy is the most common major childhood neurologic disorder in the United States, occurring in approximately 1 out of 100 children. Approximately 325,000 American children between the ages of five and 14 have active epilepsy. Doctors diagnose about 45,000 children in the U.S. each year with the condition.

Epilepsy in children has many causes, including birth defects, brain infections, head injuries, heredity—but often the cause is unknown. Some young children only have seizures with fever, known as “febrile seizures.” Epilepsy is more common in children with cerebral palsy, developmental delay, and mental retardation. Conditions such as attention deficit hyperactivity disorder (ADHD), autism, and developmental disabilities commonly occur in children with epilepsy.

For most children who have seizures, the path to successful treatment is the selection of the appropriate antiepileptic drug for the child’s seizure type. Antiepileptic drugs used in children include carbamazepine, levetiracetam, lamotrigine, valproate, and others.

Common seizure types include absence seizures (“petit mal,” or staring spells), partial complex seizures (episodes of confusion), and convulsions (“grand mal,” or generalized tonic clonic seizures). Depending upon the seizure type, different drugs are likely to be more effective. For example, valproate is often effective in absence seizures, while carbamazepine is preferred for partial seizures.

Most children with epilepsy respond to antiepileptic medication treatment or eventually grow out of their seizures. But some children—often those with associated brain

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injury and other neurologic problems like cerebral palsy or mental retardation—have seizures that are difficult to control. This is called “refractory” or “intractable” epilepsy.

BREAKFAST

Scrambled egg and apple sauce

Whole egg	1.6 oz.
Butter	0.5 oz.
Nuts	0.1 oz.
Granny Smith apple	1.06 oz.
Fresh olive oil	0.46 oz.
Regular butter	0.35 oz.
Cream	0.18 oz.

LUNCH

Creamy bacon meal

Green pepper	0.7 oz.
Celery	0.7 oz.
Brown onion	0.7 oz.
Cream	2.26 oz.
Lean middle bacon	0.95 oz.
Margarine	0.77 oz.

DINNER

Chicken and vegetable stir fry

Peeled carrot	1.06 oz.
Green pepper	1.06 oz.
Broccoli	1.06 oz.
Brown onion	0.5 oz.
Lean chicken breast	0.81 oz.
Olive oil	1.5 oz.
Soy sauce	0.18 oz.

SEIZURE TRIGGERS

John’s parents were told to keep track of his seizures by observing when and under what circumstances they occur. They noted that many of his seizures were triggered by a fever. As a result, when John develops an ear infection or a cold, they treat the fever round the clock with acetaminophen and ibuprofen to prevent a seizure. They also realized that the seizures tended to occur when he was waking up or falling asleep, so they keep a close eye on him at those times.

“You’re always on guard,” Paul says.

“You keep living, but you’re always ready.

We don’t go anywhere without Diastat, and we watch him carefully if he’s beginning to develop a cold or fever. We stay at home more. My parents live four hours away, but we learned that long trips can tire him out, and fatigue lowers his seizure threshold, so we wouldn’t risk that. His grandparents come here to visit.”

DRUG TRIALS

John was treated with trials of oxcarbazepine, levetiracetam, and lamotrigine, but he continued to have seizures and suffered medication side effects.

“The medications were too much for him,” Deborah says. “I even wondered whether we should send him to school because he didn’t appear to be able to learn. It was a very frustrating experience watching John become more impaired on the medications. They affected his alertness and his ability to pay attention, caused his muscle tone to decline, and increased his drooling.”

In addition to medication therapy, John’s parents explored the possibility of epilepsy surgery. However, because John’s brain wave studies (electroencephalogram) suggested multiple abnormal areas, he was not considered a good surgical candidate.



Epilepsy is the **most common** major childhood neurologic disorder.

THE KETOGENIC DIET

When antiepileptic drugs failed to control John's seizures, his parents turned to one of the world's oldest epilepsy treatments: the ketogenic diet.

"I read about it in some of the books and pamphlets at the doctor's office," Deborah remembers. "My expectation and hope was that we would become a zero-seizure family, but I know that's hard. We eventually switched to a doctor who specializes in intractable seizures and the ketogenic diet."

The ketogenic diet is a strict, medically supervised nutritional regimen that contains a large amount of fat, adequate protein, and very small amounts of carbohydrates. Typically, the diet requires eating four times as much fat as protein and carbohydrates (for a ratio of 4 to 1).

The high fat content of the diet causes a condition called "ketosis" that appears related to seizure control. Usually the body uses carbohydrates for energy. When fats are used instead, the liver produces substances called "ketones." Higher ketone levels often lead to improved seizure control, though scientists are not sure why.

The amount of ketosis can be gauged in the blood or urine by using test strips similar to those used by people with diabetes. The diet has been popularized in a movie, *First Do No Harm* (1997), starring Meryl Streep, which was nominated for a Golden Globe award.

Successful use of the diet requires motivated patients and caregivers, a knowledgeable dietician, and medical supervision. A lot of work goes into preparing menus in order to make the food taste good.

Shlomo Shinnar, M.D., Ph.D., estimates that less than five percent of children with epilepsy use the ketogenic diet. "It's an effective treatment that is probably underutilized," says Dr. Shinnar, who is a professor of neurology, pediatrics, and epidemiology and director of the Comprehensive Epilepsy Management Center at Montefiore Medical Center and the Albert Einstein College of Medicine in Bronx, NY.

However, Dr. Shinnar cautions that the ketogenic diet, while effective, is not "benign," because it causes metabolic changes that may adversely affect cholesterol, bone development, growth, and weight gain. "Some parents want it as a 'holistic' alternative to drugs. Personally, I reserve its use for

children with refractory epilepsy. In those children, approximately 10 percent will become seizure-free and another 30 percent almost seizure-free—which is an excellent result."

THE LATEST RESEARCH

Adam Hartman, M.D., assistant professor of neurology and pediatrics at Johns Hopkins Medical School in Baltimore, MD, focuses his research on the ketogenic diet.

"No one knows for certain how the diet works," Dr. Hartman says. "It probably has multiple mechanisms of action—those that affect neurotransmission, or communication between brain cells. In addition, it may have various effects on the mitochondria, the energy powerhouses of the cell, and alter the metabolic function of the neurons and their supporting cells."

Dr. Hartman adds: "There is a growing body of literature looking at the use of dietary therapy for other disorders affecting the brain, such as Alzheimer's disease, Parkinson's disease, cancer (brain, gastric, prostate, colon adenocarcinoma), and many others, which is very exciting."

Home Equipment

- Calculator
- Electronic weighing scale
- Measuring cup
- Vitamin supplements
- Urine or blood testing strips
- Computer access with Excel program for meal planning

MAKING THE DIET WORK

Paul and Deborah attended a one-day orientation about the diet at the Children's Hospital of Philadelphia. They learned that a minority of patients become seizure-free on the diet, but that nearly all children will have some benefit. For Deborah, the decision to try the diet was easy.

"I was really prepared to do anything to get John's seizures under control," Deborah says. "The nurse, dietician, and doctor at the Children's Hospital of Philadelphia were amazing. Everyone stuck with us to make it work. We had an incredible amount of support, and you need that."

The diet requires carefully weighing each food portion, and all of the food has to be consumed at each meal. John had a slight advantage in this respect: Because he had trouble eating, he had a feeding tube placed. This has made it easier to administer the diet, because whatever he doesn't finish by mouth can be put in through the tube. In the beginning, Deborah spent two to three hours each day preparing the unusual, high-fat meals. Now that she's familiar with the diet, she spends an extra half hour a day in meal preparation. Some special equipment, including an electronic scale, is necessary in the kitchen. (See box, "Home Equipment.")

Parent to Parent Advice

- Find an experienced epilepsy doctor who is also experienced with, or is comfortable referring patients for, the ketogenic diet.
- Learn about available treatment options.
- Work closely with the dietician, nurse, and doctor.
- Budget extra time to learn how to use the diet.
- Keep Diastat on hand.
- Treat the fever.
- Keep a seizure calendar with notes regarding possible triggers (like fever).
- Watch for side effects from medication and the diet.
- Recruit family support.
- Don't give up.

More than 100 studies document the diet's effectiveness, says Eric Kossoff, M.D., assistant professor of pediatrics and neurology and medical director of the Ketogenic Diet Center at Johns Hopkins Medical Center in Baltimore, MD. "Children with infantile spasms, tuberous sclerosis, and Doose syndrome do best on the ketogenic diet," Dr. Kossoff observes. (Doose syndrome is a rare childhood epilepsy characterized by frequent episodes of atonic and absence seizures. Tuberous sclerosis is a genetic disorder characterized by "tubers" that may grow in different organs, such as the heart, kidneys, eyes, and brain. More than half the people with tuberous sclerosis have seizures.) The ketogenic diet can also be used in adults, he adds, but they may have more trouble sticking to it.

Dr. Kossoff has also pioneered use of the Modified Atkins diet for seizure control, which according to many patients is easier to follow and offers slightly more variety and amounts of foods than the ketogenic diet. In a 2008 paper in the medical journal *Epilepsia*, Dr. Kossoff explained that while the ketogenic diet typically has a fat to protein and carbohydrate ratio of 4 to 1, the modified Atkins diet has a ratio of 0.9 to 1, or about 65 percent of calories from fat. This is still more fat than a typical diet, which has about 35 percent fat and a ratio of 0.3 to 1. Results at this time appear similar to the ketogenic diet, but are still preliminary in comparison. Clinical trials of the ketogenic diet and modified Atkins diet are ongoing and recruiting participants. (See Resource Central, page 36.)

SIDE EFFECTS

Not unlike antiepileptic drugs, the ketogenic diet has side effects. A person may feel sluggish for a few days after starting the diet. Other side effects include kidney stones, high cholesterol levels in the blood, dehydration, constipation, slowed growth or weight gain, and bone fractures. Because it doesn't provide all the vitamins and minerals found in a balanced diet, a dietician will probably recommend vitamin and mineral supplements.

“This diet has unlocked John's potential.”

“There are a lot of things that you have to look for and manage,” Deborah reports. “You have to make sure your child is not dehydrated or too acidotic [has too much acid in the blood], that the blood sugar is not too low and the calories are adequate, and that there are enough vitamins and minerals. John has had problems with vomiting from the diet's high fat content, and we have been struggling with that.”

John gets complete blood work every three months, including triglycerides and cholesterol, which are followed closely because of the diet's high fat content. His parents also check his blood to make sure he stays in “ketosis” at least once week.

Deborah cautions: “If you are wavering about it, I think the likelihood of success is low. You really have to be determined to make it work and stick with it when it gets hard.” She offers a few tips for parents of children with intractable epilepsy. (See box, “Parent to Parent Advice.”)

JOHN'S SUCCESS

John started the ketogenic diet in January 2008. Since then, he has only had one brief seizure and has experienced improvements in alertness and development. He has been weaned off levetiracetam and is due to stop his lamotrigine in a month, if all goes well. John's father, Paul, says, “Since John has been on the ketogenic diet we've been watching him blossom emotionally and improve his concentration and ability to focus. He's happier now. We feel he's able to make better strides in his physical and occupational therapy. He's not in a fog.”

Previously, John had had a problem with excessive drooling and had to wear a special scarf to protect his clothes. Within a month after starting the ketogenic diet, the drooling vastly improved.

“The day I realized we didn't have to use those little scarves any more, it was a wonderful thing. It's pretty remarkable. This diet has unlocked John's potential,” Deborah beams. “He's a beautiful child and I'm so proud of him.” NN

Andrew N. Wilner, M.D., is the author of Epilepsy: 199 Answers. A Doctor Responds to His Patients' Questions, 3rd edition (Demos Health, 2008).



For more information on the ketogenic diet, see RESOURCE CENTRAL on page 36.

**The names have been changed to protect the privacy of the patient and his family. However, Paul and Deborah have offered to talk to any parents who have questions about using the ketogenic diet, and can be contacted by writing to the editor at Neurology Now. (See “@ Your Service,” page 8, for e-mail and postal address.) Please note that all information, including menus, is for educational purposes only and should not be used unless authorized by your doctor.*