

Aunt Cathy's Guide to Nutrition:

Why Supplementation of Carnitine is Important when on the Ketogenic Diet for Seizure Control



Cathy Breedon PhD, RD, CSP, FADA
Clinical and Metabolic Nutrition Specialist
Perinatal/Pediatric Nutrition Specialist
Sanford Medical Center, and UND School
of Medicine, Dept. of Pediatrics, Fargo, ND

Introduction:

It is not uncommon to have to convince insurance companies that they should cover the cost of supplemental carnitine for patients following a ketogenic diet for seizure control. I have written explanations many times for patients and their physicians, so I decided to finally get smart and write it up in this form to make it easily available to other folks working on this issue. (It also helps me a bit because I type with one finger! 😊)

I recommend that people give the information I have written below to the patient's health care provider and have that person submit the request. This is because the insurance company will often only look at things coming from an MD and not from a PhD. But do include the credential part shown above by the picture so the doctor might more comfortably believe me that this is really a big deal, along with giving him/her some ammunition to include in the letter requesting coverage.

As always, nothing I say here takes the place of the advice of your physician or other health care provider. Also, please note that: I have no relationship with manufacturers or sellers of carnitine. All my observations are based on the legitimate scientific literature and my own 30 years of experience as a pediatric nutrition specialist. In other words, the following is just a well-intentioned collection of thoughts on the issue. Good luck! Cathy B.

Carnitine supplementation (a carbohydrate-free form) is a key part of managing the ketogenic diet because:

1. One normally makes carnitine in the liver, kidney and brain, which allows fat to be used for fuel to do things like have a heartbeat, diaphragm function and all movement. It has other important uses as well, but these functions are clearly critical for health.
2. The distorted fuel mix that controls her seizures on the ketogenic diet (WAY more fat and WAY less CHO than the usual dietary mix) means that the amount of carnitine a

person might ordinarily make will simply be no longer adequate to meet her needs because much more is needed to process the high fat content.

The situation is somewhat analogous to the fact that everyone needs to produce insulin to utilize CHO fuels. Children who develop Type I diabetes cannot produce enough of the insulin they need, with potentially injurious and even lethal consequences. Providing insulin to these children is always covered by insurance. It would never be denied.

3. Failure to provide carnitine to children on the ketogenic diet and/or certain seizure medications that impair carnitine production is analogous to telling children with Type-I diabetes to just get busy and make their own insulin, since other people can easily do it. They simply can't, and it is clear that the situation is not at all benign.
4. Failure to provide adequate carnitine to efficiently use fat for fuel will result in seriously dangerous low blood sugar. It can cause brain damage and/or death from hypoglycemia. This is because the body's first need is for energy and it will need to burn glucose it cannot afford to burn if it can't burn fat because of inadequate carnitine. This decreases the blood glucose that is available to fuel the brain.
5. Some seizure medications (valproic acid / Depekene, and likely some others that have not been as closely studied) will actually cause a person to stop making their own carnitine. Because the medication itself REQUIRES carnitine to work, the loss of carnitine results in break-through seizures and the need to continually increase the amount and types of seizure meds required to control seizures. This causes problems like excessive sleepiness and lethargy, etc., and it also results in risk of very dangerous liver toxicity.
6. Assuring adequacy of carnitine is known to prevent the very serious complication of liver toxicity. The emergency treatment for children hospitalized with this kind of liver toxicity includes intravenous carnitine.
7. Failure to provide it and the resultant toxic liver situation has been the cause of admissions to our pediatric intensive care unit which have been very lengthy (several months) and very expensive, and from which the children experienced further (preventable) neurologic injury. It also resulted in their requiring more therapies post discharge, and they often experienced the loss of important skills.

For example, one very memorable child lost her ability to smile, which was especially devastating to her family and to all of us who cared for her. These events were tragedies that were totally preventable by simply assuring adequate carnitine supplementation.

8. When the ketogenic diet is shown to be helpful to a particular patient in terms of seizure control, its use results in significantly reduced overall costs. These include lower cost due to less total medication needed, the commonly found ability to discontinue the use of certain medications altogether, reducing side effects, and improving quality of life.
9. Please my short overview paper on carnitine issues that provides more detail;