



## **Third International Symposium: dietary therapies for epilepsy & other neurological disorders**

*Family Day, September 22, 2012*

Questions & Answer Session: During the Family Day we held a Q & A session with the Steering Committee; Dr. Elizabeth Thiele, Dr. Eric Kossoff, Dr. Liu Lin Thio and Beth Zupec-Kania. There was not enough time to respond to all of the questions therefore the committee has provided written responses to the remaining questions below.

1. Question: Does it matter what type of seizures you have for the diet to be recommended for an adult?
  - Response: The diet works for all seizures types.
2. Has there been a study to measure efficacy of the diet with medications either taken pre-ketogenic or while on the diet?
  - No, there has not been a study of this type.
3. Do you see instances when the diet spontaneously stops working, even when used properly?
  - This has been reported to occur with the diet. It has also occurred with anti-seizure medications. It is not known why this happens in either case.
4. Could a drug be made that inhibits glycolysis?
  - There is a drug designed to prevent glycolysis; it has been in development for several years and is currently in animal trials. There is hope of using it someday to prevent seizures in people.
5. Is the blood sugar level important if the patient is not symptomatic?
  - The blood glucose (sugar) levels are in a range below the normal range while on ketogenic diet therapies. In this lower range, symptoms of low blood glucose are typically not present.

6. Does the Ketogenic Diet or Modified Atkins Diet affect carnitine in the body.
  - Carnitine is an amino acid that our body can create from other amino acids which we get in our diet (found mainly in meat and dairy). The role of carnitine is to carry the microscopic fat molecules (called fatty acids) into our cells to make energy (called ATP). If there is not enough carnitine, the cells will make less energy. It is theorized that a high-fat diet will use more carnitine than a regular one, therefore, carnitine may become depleted during diet therapy. Supplemental carnitine is often advised particularly for patients who are receiving the anti-seizure medication called Depkote (valproic acid).
  
7. Dr. Thiele, please expand on how breast feeding is used with the ketogenic diet.
  - Breastmilk is the best food for an infant for multiple reasons and it is very high in fat. For this reason, we encourage mothers to continue to breastfeed their infants during diet therapy. We can still make their diets ketogenic by offering the infant ketogenic formula between breastfeedings or if the mother has pumped her milk, it can be mixed together with the ketogenic formula.
  
8. If the ketogenic diet is used for Juvenile Myoclonic Epilepsy, how long does the diet need to be followed?
  - We would approach the diet for this condition very similar to the consideration of anti-seizure medication. What is the best dose and frequency to prescribe to a young adult so that they will be able to be compliant? Any type of diet restriction is difficult during the teenage years. It would be worthwhile to try some of the basic elements of these diet therapies to see if seizure control is better initially. Eliminating simple sugars (sweets, candy, cookies, pastry, soda) is one major step that could be trialed. Eating meals that include a source of fat and protein is another step. Choosing unprocessed or 'whole foods' is another to approach to trial. Processed foods are typically high in carbohydrate, especially the fast absorbing carbohydrate (also called high glycemic index foods).
  
9. What are the absolute contraindications of diet therapies?
  - The following conditions are contraindications to ketogenic diet therapies.
    - Carnitine deficiency (primary)
    - Carnitine palmitoyltransferase (CPT) I or II deficiency
    - Carnitine translocase deficiency
    - beta-oxidation defects
    - Medium-chain acyl dehydrogenase deficiency (MCAD)
    - Long-chain acyl dehydrogenase deficiency (LCAD)

- Short-chain acyl dehydrogenase deficiency (SCAD)
- Long-chain 3-hydroxyacyl-CoA deficiency
- Medium-chain 3-hydroxyacyl-CoA deficiency.
- Pyruvate carboxylase deficiency
- Porphyrin

10. Can the diet be used as a first-line treatment?

- The ketogenic diet is currently first-line treatment for two metabolic disorders; Glucose-1 transporter deficiency syndrome and pyruvate dehydrogenase deficiency because there has been no other therapy that has been effective at treating these conditions. There are a couple of barriers to using the diet first-line for other conditions. One is the above contraindications. The tests for these conditions are expensive and take weeks to obtain final results. Another barrier is that there is limited access to diet therapy programs, largely due to the fact that there is currently little or no insurance reimbursement to the facility for providing diet therapy services. As a healthcare consumer, you have the right to request diet therapy at a facility that provides it.

11. How long do you recommend that a child is seizure-free before weaning drugs?

- Typically we wait 1 month before weaning anti-seizure medication. If there is an adverse effect from the medication during the month such as unresolved sleepiness, then we often reduce the medication.

12. You've talked about the food portion of the diet, what about the soaps, lotions and other things that can soak through the skin that has sugar in it?

- It is not known if sugar (carbohydrate) can soak through skin and effect ketosis or blood sugar in the body. There are reports of children who have had seizures soon after the application of sunscreens that contain carbohydrate. Since there are sunscreens that don't contain carbohydrate, it makes sense to use these products instead. The Charlie Foundation has created a list of these low carbohydrate and carbohydrate-free products which you may download from the Resources link of the web-site: [www.charliefoundation.org](http://www.charliefoundation.org)

13. Is there evidence to suggest going from the Classic Ketogenic diet to the MCT oil Diet would improve seizure control.

- There are no studies to date that compare the crossover in diets. There is one study that showed that the Classic Ketogenic Diet had the same results as the MCT oil Diet (Neal et al. Lancet/Neuro. 2008). Some patients tolerate one better

over the other which may be the reason that seizure control is better. The Classic KD is gluten-free whereas the MCT oil diet allows grains. This may be another reason why one diet would work better than the other. There are several factors that could be the reason why one may be better for a given individual. Many centers use a combination of the MCT oil Diet and Classic Ketogenic. This is a great solution in getting the best from both therapies.

14. Our son is almost 3 years old and is on 1200 calories a day of the Classic Ketogenic Diet. He is begging for food every 1 ½ to 2 hours. What recommendations do you have? He has been on a 4:1 ratio for 15 months.

- Have you increased his calories during the 15 months? Most children his age require 2-3 calorie increases over the course of that time. A growth chart with both weight and height should be plotted to assist with determining when to increase calories. Normal growth is the goal although it is not uncommon to have slower gains in height while on diet therapy. Ketogenic snacks are an easy way to allow flexibility in calories. If a child is very active one day, the next day they are typically hungrier and may need 2 ketogenic snacks instead of one. If a child has seizures on a particular day and is not very active; snacks may be omitted that day.

15. What can be the reason for varied ketone levels when the diet is followed consistently? We have readings of 40 in the morning and 160 in the evening.

- Ketones levels can be measured in the urine and also in the blood. The readings that you have reported are urine. The ketones that are tested in the urine (called acetone) are ketone waste product after the blood has been cleaned by the kidneys. This is an estimate of ketosis and not completely indicative of the degree of ketosis in the blood that is affecting seizures. Ketosis is generally higher after the diet is consumed and lower in the morning after a long period of rest.

Please check back for additional questions with answers each week.

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