



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. If meat is not consumed regularly while on the diet, a supplement prescription carnitine may be helpful. Supplemental carnitine is often advised for patients who are receiving the anti-seizure medication 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 the following two metabolic disorders (epilepsy is common to both); Glucose-1 transporter deficiency syndrome and pyruvate dehydrogenase deficiency. There is 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 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.charlifoundation.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. The Classic KD is gluten-free whereas the MCT oil diet allows more carbohydrate which includes grains. 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.

16. Is it possible to continue the diet during illness such as stomach flu?

- During an illness, we consider the same approach to diet and fluids as we would if the person was on a regular diet. Hydration is the most important goal. Food may be eliminated for 1-2 days without a problem for most people if vomiting or diarrhea is present. A low carbohydrate electrolyte beverage such as PowerAde Zero is a sports drink that has less than 1gm of carbohydrate per 8 ounces. Pedialyte diluted to half strength is an option for young children (who dehydrate

much faster than adults). Dietitians can also create a homemade carb-free electrolyte recipe of Morton's Lite Salt (potassium chloride) and baking soda (bicarbonate). When it is time to re-introduce the diet, it's best to gradually return to solids, starting possibly with sugar-free jell-o or plain gelatin. Re-introducing the ketogenic diet with meals should also be gradual. For example, a meal with ½ the usual fat such as applesauce with chicken sautéed in olive oil.

17. When my son is healthy his is seizure-free. When he gets an illness, he is not. How can I keep him seizure-free when he gets hit with upper respiratory illness?

- Illness can be a huge stressor which can lead to seizures. Parents often suspect that their child is becoming ill with a virus when the child has been seizure-free for a long period then has a sudden increase. The best treatment of viruses is through prevention. Viruses commonly enter the body through the mouth, nose and eyes. This is why hand washing is so important as well as keeping hands out of eyes, mouth and noses. A healthy diet is important for immune function and it is necessary to receive appropriate supplementation of vitamins and minerals with the ketogenic diet. Vitamin D is one micronutrient that is extremely important for immune function. A Vitamin D supplement is advisable for anyone who is receiving anti-seizure medications even if their blood level is normal and especially for someone who does not receive daily sun exposure.

18. Is a lower white blood cell count a side-effect of the diet?

- There is one report that was published in 1989 that described a patient who was on the ketogenic diet and had a severe infection that would not resolve. This patient's white cell count was very low. They discontinued the diet and the infection resolved. They also reported that they checked the white cell count of their other patients on the ketogenic diet and found them to be lower than normal. The issue here is that we cannot assume that the ketogenic diet caused the low white cell count. This is not a problem that has been reported by other centers. It is very possible that these patients were not supplemented adequately and had Vitamin D or other nutrient deficiencies that caused them to be immune compromised. Vitamin D is unique in its role important role in immune function as well as bone health. Most people who take anti-seizure medication do not get enough in from their diet alone and need to be supplemented.

19. How ketotic and how long is diet therapy expected to be needed for other conditions.

- The use of diet therapies for other conditions is in its infancy. One example that I (Beth Zupiec-Kania) have experience with is use of the diet for treatment of brain tumor. The focus for this treatment is on blood glucose rather than degree of

ketosis. The diet would be maintained for as long as the individual was able to follow it and regular MRIs would provide objective feedback as to the effectiveness of this treatment.

20. What are the effects if ketone levels are going up and down.

- I'm assuming that you are referring to urine ketones. They are the waste product of ketones after the kidney cleans the blood. They are diluted by how much the person recently drank. This is why we never make a diet change decision based on one urine ketone reading. It is more helpful to get a series of readings. Typically the urine ketones are weaker in the morning and stronger at the end of the day. When testing urine ketones in a diaper, if the diaper is older than an hour, ketones may have evaporated rendering the reading less reliable.

21. We observe irritability when ketones are very high which also parallels with low CO₂.

We feel our child does better with a ketosis in a lower range.

- First, I would like to explain that CO₂ means carbon dioxide and refers to acid in the blood. Acidosis can cause a couple of problems. It can cause calcium to leach out of bones (weakening bones which can be painful) and it can also impair the ability of hemoglobin to bind to oxygen. It is important to have acidosis evaluated and treated. The most common cause of acidosis with the ketogenic diet is when certain medications are used in combination with the diet. The anti-seizure medications Zonegran (zonisamide), Topamax(topiramate) and Diamox (acetazolamide) can cause acidosis and this effect can be even stronger when used in combination with the ketogenic diet. Another cause for acidosis can be that the person is not receiving sufficient calories.

Please check back for additional questions with answers each week. I have been trying to group questions according to topics. The next set will focus more on the diet itself.

Prepared by Beth Zupec-Kania, RD, CD – Clinical Nutritionist