

MCAD Communication Network
'All In This Together'

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The *MCAD COMMUNICATION NETWORK Newsletter* was created and is currently edited by Deb and Dan Gould ~ 805 Montrose Drive, Greensboro, NC 27410 (336) 547-8682
Any questions or comments should be directed to them.

From The Editor

Happy Holidays and Happy New Year! It's so hard to believe that 1994 is already here. We hope it has been a safe, healthy, happy, and productive year for all of our MCAD families.

I'm sure all of you have been very busy this past year. The last 6 months for us have been extremely hectic, yet very exciting. So exciting that I wanted to share my news with all of you.

Three years ago I made the decision to return to graduate school to pursue a masters degree in Counseling. My decision was my way of channeling my anger at being a bereaved parent and being away from my home and family in Illinois. The most meaningful reason for me, however, was that it was an active way of keeping my daughter's memory and spirit alive within me.

I took classes for 2.5 years and then this May through December I did a grief counseling internship at Hospice at Greensboro, Inc. It was a very rewarding and fulfilling experience. Then on December 8, 1993, I officially graduated!

Dan and the boys have been very supportive and patient with me and helped me celebrate when my last day arrived. Presently, I am starting up a private practice. In my practice, I will work mostly with individuals and families who have experienced the death of a child.

I was just so excited about finishing one chapter in my life and beginning another that I wanted to share my good news with you. As always, if any of you have your own exciting news, questions, stories, articles, or nutritional ideas, feel free to send them to us. We would love to hear what's going on in other MCAD families across the country. Please don't think your ideas or questions are 'dumb.' Dan and I appreciate whatever you are willing to share. Remember, this is **your** newsletter, too.

In this issue 2 more MCAD families share their stories. Thank you to Randall and Misty McDonald and Simone and Michael Miller for taking the time to write about how their lives have been changed by MCAD. Also in this issue questions are answered, pharmaceutical & medical updates are reported, Love Messages are remembered, and a nutritional guide for healthy low fat eating is shown. Additionally, we have included an article about MCAD for clinicians that may be helpful to your personal physicians, as

well as to emergency room personnel, especially if you are traveling (*see Dr. Roe's article on our website under Medical Information/ MCAD Information for Clinicians), as well as an updated Family List.

Note: MCAD Families cannot be listed until the Family Questionnaire (enclosed in your new Family Packet) is returned to Deb and Dan.

Also, please take a look at the questions at the end of the newsletter (* edited out of this issue). We are trying to determine if there is an interest in having a daylong MCAD Family Network seminar. Families would be able to talk with other MCAD families, as well as gain current information on MCAD research. If you wanted to make a vacation of it, you could take advantage of the beautiful mountains and/or beaches of North Carolina or see other historic sites that are only a few hours from Durham.

Again, we hope that you find this issue informative and helpful in networking with other MCAD families. If you would like to contribute to our June 1994 issue, we would greatly appreciate your suggestions, questions and/or articles. Please include your response to the MCAD seminar.

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Jeremiah Lawton McDonald

My wife and I were very excited about the arrival of our first child. We named him Jeremy. We were both very much in love with our new addition to our family. The physician told us that we had a very healthy 7lb 5oz baby boy with no problems. After spending 2 days in the hospital, we were able to bring our son home.

He was a very healthy and special little boy for eight and a half months. Then on Saturday, December 14, 1991, he started having flu-like symptoms. He ran a fever, had no energy, and didn't want to eat. My wife and I took him to the Dr. and he said it was a virus and gave him some medicine. We thought he would be just fine. Sometime around 3am on Sunday, December 15, he woke up. He would only eat a little. After checking that his temperature was down, I put him back to bed. We didn't know that was going to be the last time we were going to ever see our precious little boy alive.

I woke up the next morning and I could tell something was wrong. My wife called an ambulance, but I knew it was too late. It seemed like we were at the hospital for hours. Yet, it was only a short time. The waiting was more than I could stand.

The Dr. came and said he had done everything that he could do, but that it was too late. They really didn't have an answer. They thought it was crib death. The coroner's autopsy report said it was 'SIDS.' I knew that couldn't be.

At the funeral we realized that our son was really gone. A year later Dr. Pam Loveland contacted us and informed us that from some of our son's blood, his death was determined to be from MCAD. It was such a terrible shock to finally get a cause.

We love Jeremy and, know that he is in heaven, and that we will be there too someday. We now understand that if we have more children, they can be treated. We thank God for this.

Jeremiah Lawton McDonald
Born April 3, 1991
Deceased December 15, 1991

Randall & Misty McDonald
Murphy, NC

Michael Dylan

On August 24, 1991, our son, Michael 'Dylan,' was born at 5:39pm. He got a 9 on the first Apgar test given seconds after he was born, and a perfect 10 on one given a few minutes later. In other words, he was born very strong, healthy, and responsive. I'll never forget the look of pride on my husband's face. And I'll never forget how nothing seemed more important to me than the life and happiness of this child.

We took Dylan home the morning of August 26. He seemed fine! He slept all day in my arms because I just felt like holding him close. I called the nursery at the hospital several times to make sure I was doing everything right. They assured me that we were both doing fine, and that he was too little to be spoiled by holding him all day.

Unknown to us at the time, Dylan had MCAD. The current theory is, due to the long labor and long periods of time (for an MCAD newborn) that Dylan went between feedings, he built up fat toxins in his cells and became weaker and weaker. The symptoms that occurred went undetected because they were similar to a sleepy, worn out newborn.

The details of discovering Dylan was dead by 4:30am, the morning of August 27, 1991, are a permanent part of my memory. For the purposes of this article, however, I prefer to jump ahead several months, to finding out while I was 6 months pregnant with our second child that Dylan had died from MCAD and not Reye's Syndrome, as previously diagnosed. I was relieved to know more about what happened to our son, but petrified with the possibility of our next child having this condition I knew nothing about. The doctors were hesitant to give me much information on MCAD because they felt there was no need to worry me in case our next child was not affected.

Chloe Monet Miller was born at 4:37pm on September 23, 1992, and she too has MCAD. In retrospect, I wish I was more informed about what we were dealing with because now I was terrified!

Luckily for us, Chloe was a very healthy baby with a big appetite. She has the greatest personality a parent can hope for, and she went through her first year without any episodes. Right around when Chloe turned 13-months she started showing signs of getting an ear infection. I immediately took her to the doctors and she was put on antibiotics for her one ear infection. Chloe's caloric intake did decrease slightly and she was not her usual bubbly self. After being on the antibiotic for several days Chloe did not seem to be getting better and she threw up after drinking her morning bottle. We rushed her back to the doctor's office and they changed the antibiotic. This was not enough to reverse the process that occurs when an MCAD child is trying to fight an ear infection.

To make what is a long story for me short for you, Chloe ended up in the hospital on IV glucose. She was stabilized and then sent home the next day. She is still doing great and back to being her adorable 15-month self. However, as a result of this happening I decided it was too scary and foolish for me to wait for information about MCAD to come to us. I called a lady at the lab at Washington University in Missouri where a lot of the research on our family had been done. She was very helpful and sent me the information on Deb Gould and her MCAD support group. I called Deb immediately and we talked for quite awhile. She told me about Carnitor® and also about Dr. Roe and his research efforts at Duke University Medical School (* in 2000, now at Baylor in Dallas). And once again, to make a long story for me short for you, I flew down to Duke, December 10, 1993, and learned as much as a parent can with the knowledge available in one long, wonderful, information-exchanging day. I was amazed at the time and energy these people were willing to devote to me in explaining what they know about MCAD. And I left there feeling optimistic about MCAD children's futures and the ability of Dr. Roe and his staff to continue learning more about MCAD and other rare metabolic disorders so we will understand even better how to help our kids.

Simone and Michael Miller
New Brighton, PA

Son of the Sea

I am a very spiritual person. I have my very own philosophy about God and the workings of the Universe. Much like the American Indians, I believe that animal spirits guide us/me.

A number of months after we lost our first child, Michael Dylan, I had a dream. During my pregnancy with Dylan and up to this dream, I had a series of dreams with water in them. The water took different forms. Sometimes it was wild and scary, sometimes it was calm and clear, or dark and dreary, or cool and soothing, or sometimes it was just there.

In this particular dream, the water was lively and exciting. All kinds of things were happening in the water, and I was watching these events from the shore. All of a sudden, this little brown bear Cub came swimming up to me. I remember thinking to myself in

the dream I should be scared of this wild creature, but something seemed familiar and gentle about him.

The Cub had a leather harness strapped to him with long reins trailing behind. The next thing I knew I was in the water with the Cub, swimming behind him and holding onto the reins. It was an exciting, adventuresome feeling .We swam towards a gate and I got scared and tried to stop the Cub and myself from going through it. The Cub led me back to the shore. When we got there, I was very confused. I wanted to keep the Cub and protect him and play with him and be close to him. But something inside me told me I must take the harness off him and let him go. So I took the harness off and said goodbye to the Cub.

I remember as he swam away feeling a little empty and lonely, but in my heart I knew I had done the right thing. When I woke up the next day, I couldn't stop thinking about the dream. As time passed, I started to understand the symbolism in the dream. In my heart and soul, I know that the little bear Cub was my Dylan coming to me to say Goodbye and to let me know it was time to let go.

Thank you Dylan/God for sending me a symbolic message in such a beautiful way!
Dylan is a Gaelic name which means 'Son of the Sea.'

Written in honor of my son,
Michael Dylan Miller

Always your loving Mother,
Simone Suzanne Miller

Questions and Answers

[Please Note: This question and answer column is designed to answer questions, both medical and practical, on MCAD and its treatment. Answers to questions are solicited from those who have had firsthand experience dealing with MCAD. These include physicians, parents of MCAD children and children themselves. It is our hope to provide general guidelines in responding to questions posed as opposed to specific foolproof solutions. Additionally, it is especially important to note that our Medical Advisor, Dr. Charles Roe, (at printing of this newsletter in 1994, he was at Duke University Medical School and now, in 2000, at Baylor in Dallas) has read and approved responses to all medical questions. However, because of the individual nature of each case, it is always important to discuss these guidelines with your physician before making any changes.]

Question: What is carnitine?

Answer: Carnitine is a naturally occurring substance required for energy metabolism in mammals. It is produced by the body and is also available in the diet (particularly in red meats and dairy products). Carnitine transports fats into the mitochondria, the cellular furnace, where these fats are converted into an energy source. Carnitine also helps to get

rid of the metabolic toxins that that process produces. Our heart and skeletal muscle tissue rely on fat utilization as their major source of energy. Oftentimes, MCAD children/adults also have a secondary carnitine deficiency and supplementation is recommended (*learn more about Carnitor® on our website under Pharmaceuticals).

Question: Why is it important?

Answer: Without carnitine the utilization of fatty acids as an energy source for all tissues, particularly heart and skeletal muscle tissue, is impaired. This impairment in certain individuals can be life threatening.

Question: What causes carnitine deficiency?

Answer: Carnitine deficiency can be due to decreased natural synthesis of carnitine in the body or altered transport of carnitine across the muscle cellular membrane, where carnitine is known to be active. Any situation where the diet lacks carnitine or any state that causes abnormal loss (i.e. taking seizure meds) or over-utilization of carnitine can result in deficiency.

Pharmaceutical News

Some of you have requested information about Sigma-Tau Pharmaceuticals, Inc., the producer of Carnitor® and the financial backer of our newsletter. If you would like to correspond with them, contact them at 1-800-447-0169.

Medical Update

Ross Laboratories has developed a low fat formula (ProViMin) for fat-intolerant infants and children. They are interested in determining if it would be useful for MCAD children in terms of keeping them balanced. The formula has the same calories and sugar content as regular formulas, but with a lot less fat.

Through funding by Ross Laboratories, Dr. Kim Iafolla (formerly of Duke University), is looking for 6 MCAD families with an infant/child between the ages of birth to 3 years (*this study is no longer active in 2000). The child would use the special formula as their primary source of milk for 6 months. A nutrition diary would be kept to document the child's food/drink intake and to check for adequate fat/carbohydrate/protein intake. Periodic measurements (height, weight, blood sample) would be taken at the beginning, middle, and end of the study. The first and last measurements, however, would have to be taken at the Children's Research Unit (CRU) at Duke University Medical Center. The middle measurements could be taken at your local Dr.'s office and then sent to Duke. While at the CRU, the grant money would pay for all costs pertinent to the study for the mother and child. Families would have to provide their own transportation to and from Duke and housing/food for other family members, however.

If you might be interested in participating in this study, please contact Dr. Kim Iafolla at

Duke University Medical Center at 919-684-2036 and mention you are calling about the study.

Carolyn Stamm and her very intelligent and energetic class from Virginia Beach, VA (*see June 1993 issue), were on the road this November. Dr. Roe (*at Duke University Medical Center at the time of this issue), invited the class to participate in a half-day MCAD discussion and tour of the Mass Spectrometry Facility in Durham, NC. The class updated the audience on their accomplishments and what they hope to achieve in the upcoming year. They are continuing their efforts toward educating a variety of groups in VA, as well as getting MCAD as part of the newborn screening tests. They are even planning to write President and Mrs. Clinton!

Dr. Roe then discussed some of the obstacles of getting a newborn test, yet stressed that they are also continuing their efforts of developing an inexpensive (\$15) screening test that could be done nationally. He and his colleagues are determined to succeed. It could take some time, but they are optimistic.

A family panel, consisting of the Ross family from South Carolina and myself (Deb) personalized the session by telling our stories of how MCAD came into our lives. We then answered questions from the audience.

Before going to lunch and for a tour of the Duke campus, the audience toured the Spectrometry Lab. The staff demonstrated how specimens were tested for biochemical and DNA diagnosis and what they looked for on the printouts to determine MCAD, as well as other specific diseases.

It was a very informative and enjoyable day. It was great to finally meet people you've talked with several times over the phone!

Love Messages

(Please see our most current online issue)

Low Fat/No Fat Food Guide

	OKAY ANYTIME (Every day) Low fat or no fat	NOW AND THEN (Once a week) Some fat or cholesterol	ONCE IN A WHILE (Once a month) High fat or cholesterol
Meat, fish poultry eggs No more than 5 to 7 ounces lean meat, fish, or poultry a day	Fish -Tuna (water- packed), flounder, cod, haddock, halibut, perch, mackerel, salmon, sole. Poultry -Chicken or turkey boiled, baked, or	Fish -Tuna (oil- packed), other fried in vegetable oil. Poultry -Chicken fried in vegetable oil. Meat -(Outside fat cut	Fish -Commercially fried. Poultry -Creamed or fried (in butter) chicken, duck, goose. Meat -Bacon, liver,

	roasted (no fat added). Egg whites or substitutes.	off) Lean beef, pork, lamb, veal. Egg -Limit to 2 egg yolks a week.	lunchmeats, hot dogs, sausage, and other high-fat meats.
Milk and dairy products 1 serving = 1 cup or 1 ounce (hard cheese) (At least two servings a day low-fat dairy)	Milk -1% milk fat, non-fat dry milk, skim milk, butter or evaporated milk made from skim milk. Yogurt -Low-fat Cheese -Low-fat cottage cheese, skim milk cheeses.	Milk -2% milk fat, buttermilk, cocoa made with skim milk, ice milk. Yogurt -Frozen Cheese -Regular cottage cheese, mozzarella cheese (part skim type only), Parmesan, grated, low-fat American.	Milk -Whole milk, eggnog, ice cream. Yogurt -Whole milk. Cheese -Hard or processed cheese spreads. Whipped toppings, whipped cream, most artificial creamers- a few brands are okay (check labels).
Bread, grain, cereals, beans 1 serving = 1 slice or .5 cup (At least 3-4 servings a day)	Bread -White and whole grain bread and rolls, matzoh. Cereal -Whole grain hot and cold cereal. Pasta -Whole wheat pasta, brown rice. Beans -Beans of all kinds, peas, and sprouts.	Bread -Waffles, pancakes, pizza with lean meat or vegetable topping. Pasta -Regular pasta, white rice.	Bread -Croissants, doughnuts, stuffing made with butter. Commercial snack foods-Granola bars, corn chips, potato chips (watch for palm and coconut oil).
Fruits and vegetables 1 serving = .5 cup or 1 medium fruit or vegetable (3 servings fruit and 3 servings vegetables a day)	Vegetables -All vegetables juices and vegetables, including white or sweet potatoes. Fruits -All fruits.	Vegetables -Avocado, coleslaw, French fried potatoes, fried eggplant, guacamole, potatoes au gratin.	Vegetables -Fast food French fries and onion rings.
Fat and oil 1 serving = 1 teaspoon No more than 5-8 teaspoons unsaturated vegetable oil a	Vegetable Oils -Safflower, sunflower, corn, soybean, olive, peanut. Salad dressing -Low-fat dressing, mayonnaise.	Salad dressing -Regular dressing. Margarine -Stick margarine.	Gravy, butter, coconut oil, palm oil, sour cream, and dips with sour cream.

day)	Margarine -Soft (tub).		
Sweets, snacks, desserts Ice cream = .5 cup Pudding = .5 cup Cake = 2-inch slice (As treats, used sensibly)	Sherbet, tapioca, sorbet, water ice, gelatin, sugarless gum, hard candy, jelly beans, low-cal soda, banana bread, fig bars, graham crackers, ginger snaps, angel food cake, ladyfingers.	Devil's food cake, fruit muffins, pound cake, pop tarts, fruit pies, peanut brittle, white cake with icing, vanilla wafers, oatmeal cookies.	Chocolate (one ounce), commercially baked cookies, coffee cake, Danish, cheesecake, cake with icing, toffee.

(This chart was on a handout that I received from the 'Citizens for Public Action on Cholesterol' in Washington, D.C. It was produced from a grant supported by Health Information Services, Merck Sharp & Dohme in West Point, PA). * **Please remember these are just guidelines and you should consult your child's/your Dr. or nutritionist about food/fat recommendations in regard to the specific FOD.**

Resources

Low fat Recipes: **Fit or Fat Target Recipe Book** by Covert Bailey
Submitted by: Simone Miller

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[Please Note: Our Group began in 1991 as the MCAD Family Support Group ~ in 1996 we expanded to include all of the Fatty Oxidation Disorders (FODs). Please be sure to read the most current newsletters to get the most updated information on FOD diagnosis, treatment recommendations, research, and names of FOD researchers/Labs.]