



FOD

Support

**fatty oxidation disorder
communication network**

FOD™
All in This Together
Fatty Oxidation Disorders
Family Support Group

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From the Editor



2014 FOD/OAA Conference SUCCESS!



The FOD/OAA National Metabolic Conference was July 25-26 in Arlington, VA and we had our largest participation ever!



Out of our 145 FOD Attendees (and 90 OAA), we saw some NEW FACES as well as 'old' ones at the [Crystal Gateway Marriott](#). Our 2-day Conference was packed with wonderful Speaker Presentations, Networking time, tasty meals, a Pasta Bar and Cheese Reception so Families could get to know our Speakers and other attendees on a one-on-one basis, specific FOD Breakout sessions, a SibShop seminar offered for 8-12 yr old siblings, Vendor booths from some of our Sponsors, and a touching ending ceremony video. Many made their trip to Arlington and DC a Family trip ~ lots of great sightseeing and food!

As fantastic as everything was, the cost of having the Conference on the east coast was quite high. So if you are able to DONATE to the FOD Group at any time throughout the year, it would be GREATLY appreciated. We truly appreciate every penny that our members and their Family members donate via cash, buying FOD Awareness items, and doing your own fundraisers. Thank you to all the members that purchased Awareness items at the Conference as well. We also have a NEW Awareness Item for sale ~ an FOD baseball cap ~ every FOD attendee received one! I will post it on the Awareness Item page soon.



So please keep us in mind if you are able to donate anytime throughout 2014 and beyond!

Please also continue to create awareness of FODs with your family, friends, and medical professionals, as well as create your own ways to raise funds, via 'Family Fundraisers,' so we can continue to spread the word about FODs via our website, Conferences, speaking at hospitals, and other various ways that allow us to offer all of our services free of charge. Also, when buying online please remember when you use the [iGive link](#) on our site, the FOD Group gets a percentage of your sale. We also earn funds by using [GoodSearch](#) as a search engine, or using the [Donate button](#) on our site or on our [facebook Cause page](#).

Families ~ We welcome ALL new or updated Family Stories and pictures and we encourage Families dealing with the less common FODs [i.e. HMG, GA2, Carnitine Uptake Defect, TFP, CPT 1&2 etc.] to share their experiences. We're also always looking for more low fat recipes, poems, 'Silver Linings,' pictures, and 'Reach for the Stars' accomplishments of our kids/adults/families.

Professionals ~ we need to hear from you too! New Medical, Research, Nutritional, Counseling/Coping, etc articles are always appreciated.

Whether you're a Family or a Professional, we are all striving to create awareness, education, screening and diagnosis, long-term clinical treatment, and research ~ by sharing your story or your expertise...

'We Are All in This Together!'



Take care...
 Deb Lee Gould, MEd, Director



Editorial

Connecting with others and creating FOD Awareness

Dear Everyone ~

I had the wonderful opportunity to create FOD Awareness by connecting with a 4th grade classroom in Georgia that has an FOD child—what a wonderful opportunity to help celebrate Rare Disease Day 2014! The students were able to ask me any question they would like about FODs and living with a chronic disorder. It was wonderful to actually see everyone via skype. The teacher initiated contact with me and I am so grateful that she did !



Another way to CONNECT with other FOD Families and some Professionals is to join our [facebook Group](#) and/or our [google Email List](#). We have over 1300 members on facebook and 1200 members on the google List.

♥ Please be sure you have completed the '[JOIN OUR GROUP](#)' form BEFORE you request to join either group. ♥

To help EDUCATE and CREATE AWARENESS please also share our [website](#) and [brochure](#) with ALL in your Family and your Professional contacts! I often mail extra brochures when I mail out FOD Awareness items [bracelets, magnets, tshirts etc] that members have purchased that can be shared with their medical professionals or friends.

Professionals ~ I can mail a larger # of brochures if you [contact me](#) and send your address with the # of brochures you'd like for your office or clinic.

And for those that would like to create FOD Awareness in your own town by having [your own fundraiser](#), PLEASE DO — [donations](#) to the FOD Group are tax-deductible! Please be sure to complete the Family Fundraiser form so you are aware that it is your own fundraiser and not one endorsed or solicited by the FOD Group. [Contact me](#) if you have any questions!

Make a CHOICE to SHARE your experiences with others ~ it MAY SAVE A LIFE!

~ Deb Lee Gould, MEd FOD Director



JULY is FOD Awareness Month and we once again submitted our Banner to USA Today's Charity Spotlight. It was printed in black and white on July 22nd just in time for our Conference! This year we honored Austen (GA2/MADD) and Kayla (SCAD/mito) on the Banner. Thank you Keith Widmann (MCAD dad) for updating our Banner.

JULY is Fatty Oxidation Disorders Awareness Month

Expanded Newborn Screening Saves Lives!

FOD
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For more info or to donate:
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www.bereavedparent.com
www.fodsupport.org/donate.htm
'All in This Together'

28 MONEY
Queasy over meat scare
McDonald's, Yum can't ignore overkill in U.S.

CHIPOTLE INVESTORS FEAST ON RESULTS

Apple's quarterly results out today

How to Tell Time Like a Man

CHARITY SPOTLIGHT
JULY is Fatty Oxidation Disorders Awareness Month

Since 1993, the jewelry industry has donated nearly \$4 million to help support children in need.

What all children do to help families you did together?
No answer to know. Visit www.starsforchildren.org

The weekends of war are not always easy to see.

Give \$4 an hour

For advertising details call 703-854-5325

2014 FOD/OAA Conference Pictures

[More on <http://photos.fodsupport.org/GalleryThumbnails.aspx?gallery=4517120>]



SibShop Seminar



Kathy Stagni (OAA) and Deb



Deb and Mary (FOD Webmaster)



Goodtime Downtime after full day session



Paladiy Family



Brenda and Christine



Grodskis and full FOD room



Harrys and Helds



FOD Banner and Brenda's JuicePlus display



Friday FOD session Professional Panel



Family Stories

Adam Update ~ Adult LCHAD

I can't believe it's been so long since I've updated Adam's Story! The last story was when he was 19 years old (Jan 2010 Newsletter). Now he's 23. Since then he finished his senior year at high school, graduated, and attended West Valley community College for 3 years before transferring to the University of California at Santa Cruz, where he is now a junior majoring in Bioengineering. It is a very difficult major and **we are so proud of what he has achieved so far**. Because of his health issues he will probably take 5 years to graduate but that is fine with us. He worries that he won't graduate with his friends but **his health is more important than graduating on time**.

Last year, his first at UCSC, he lived in the dorm. We thought this would be a big challenge for him to stay healthy and eat properly along with the demands of his major and the 'partyiness' of dorm life. He did well having a great supportive roommate. I just don't get it that now in the dorms the men and women live on the same floor and share the same bathroom but I guess I'm just too old to understand.!

Unfortunately, Adam has had 3 hospitalizations since starting college. They were mostly due to letting himself get into a fasting state, not getting enough sleep, and drinking alcohol. Of course he knows that alcohol inhibits fat metabolism, therefore depleting his energy sources more quickly but at his age, reason isn't always in the forefront of his thinking. He has though learned from the severity of the last two hospitalizations, in September and October of this year, that he really can't drink anymore. Usually one beer or a glass of wine will be ok but occasionally it will put him over the edge and he must be hospitalized. Fortunately UCSC is only about a 45-minute drive from our home (over the Santa Cruz Mountains). We have been able to go pick him up there and drive him back to San Jose and Kaiser Hospital, as there are no Kaisers in Santa Cruz.

Hospitalizations at his age are pretty scary. And as of January 2, he's in the hospital again! He just doesn't bounce back as quickly as he did when he was younger. During the first of the 3 recent hospitalizations it took days before his CK levels started to go down and the usual hospitalization time seems to be 5 or more days. Hospitalization #2 he was released too soon and had to return late evening of his release date. Besides his slow recovery time, we must still stay with him as much of the time as possible. We don't sleep overnight in the room as we did when he was younger but need to be in his room to **advocate for him and make sure he is receiving the right treatment**. His metabolic doctor is WONDERFUL still. We call him on his cell when Adam is going to the hospital and he calls the ER and gets things rolling, cluing in the ER doctors and alerting them about his symptoms and treatment. However, even with this any time of day or night direct communication with his doctor (usually via text) on site, still mistakes are made. Since the hospital staff hasn't ever treated an LCHADer, they really have no clue as to his treatment. Usually D-10 is administered too slowly or at too slow a drip rate. Or in September by mistake they took him off his IV drip for 6 hours. My husband is usually the wonderful parent who stays with Adam and follows his treatment.

When Adam is in a severe LCHAD Episode now, he for the most part loses his ability to talk clearly. He thinks it might be weak larynx muscles due to his continuous vomiting. He can't enunciate consonants and he becomes very difficult to understand. That's why a **parent advocate must be present as much time as possible**. And in the hospital it seems like the nurses make less frequent visits and when we push a call light, sometimes it can be almost an hour before someone responds. Usually the doctor on call only visits once a day. Adam's metabolic doctor is in another city 2+ hours away so he can't visit and just keeps up with test results on line and talks to the on call doctor. I'm terrified that once Dr Lipson retires and Obama Care goes into effect, that there will be even less doctors and nurses available to care for Adam. Don is now 69 and I turned 67 this week. I don't know how many more years we can stay with Adam in the hospital.

Adam's eyes are worse. Every LCHAD episode affects them more. He still drives a car well, even at night. And he's still in the monthly autocrosses (individual timed driving between cones in a parking lot) and occasional Track Days (driving around a professional race track with a group of similar cars at the fastest speed that is safe for each individual), but when he studies, he must have all the lights on at their brightest. A sad note, he can't see stars in the night sky. It just looks black. This is especially sad because his older brother is an Astronomer and now in the doctorate program at the University of Hawaii. One astronomer in the family, and one who can't even see the stars!

We hope Adam will continue for the most part to do well. Now that his age is closer to 25 years when hopefully the adult brain is fully developed, **he will continue to make more good decisions**. He does take his 1T of MCT Oil and 1t Carnitine in a 6 oz bottle of fat free milk at least 3 times a day. He also drinks Gatorade when he knows he will be stressed mentally or physically. He also eats an occasional NUGO protein bar (nugonutrition.com), which has 11 grams of protein. One choice he made after he was 21 was to get a 'Medical Marijuana' card. He smokes pot, he says, to relieve his anxiety. He has discussed this with his doctor who didn't ban its use. I don't know if it is helpful or harmful. We old-timer parents HATE this and HATE the smell in our house but can do little about it. We need some up and coming nutrition/metabolic graduate student to do research on Marijuana intake and Effects on the LCHAD patient. But I guess that most likely won't happen with such a rare disease.

Adam's present hospitalization is the same ole, same ole. He had severe stomach pains that led to vomiting continuously, dehydration, and an inability to eat anything by mouth. He's put on several pain meds once in the hospital for the pain. He had a CT Scan that didn't show any abnormalities in his stomach. We're beginning to wonder if he's developed a food allergy. He will probably need further testing to see if this is the case. He will probably go home tomorrow. And life moves forward...





Medical Update

Professionals: Please contact [Deb](#) if you'd like to write an article/summary for our Jan 2015 issue. Our Families are really interested in learning what research or clinical issues you are working on!



Medical 'Bits of Info'

Fatty Acid Oxidation and Retina

Webpage with info:

http://www.ohsu.edu/xd/education/schools/school-of-medicine/departments/basic-science-departments/molecular-and-medical-genetics/index.cfm?WT_rank=2

One of the complications of LCHAD deficiency is vision loss because of a degeneration of the retina, a part of the eye that is essential for us to see. This degeneration of the retina is called retinopathy and the cause of retinopathy in children with LCHAD is not known.

Our previous study found a correlation between blood levels of an LCHAD metabolite, long chain 3-hydroxyacylcarnitines and progression of retinopathy. We followed 14 children with LCHAD deficiency over 5 years. Subjects who had high long chain 3-hydroxyacylcarnitines in their blood had a greater progression of retinopathy associated with loss of night and color vision. Long-chain 3-hydroxyacylcarnitines are observed only in patients with LCHAD deficiency. Our hypothesis is that metabolites are toxic to the retina.

We have not been able to test this hypothesis because an LCHADD animal or LCHADD retinal cell culture model do not exist. Recent advances in science have made it possible to directly reprogram cultured skin cells or fibroblasts into stem cells; cells that have the potential to become any type of cell in the body. Fibroblasts are frozen from skin biopsies of patients obtained to diagnosis LCHADD. If patient fibroblasts were reprogrammed, the stem cells would have LCHAD deficiency like the original fibroblast from which they were derived. The LCHAD deficient stem cells can then be programmed to become retina cells.

In this project we propose to generate stem cells from the cultured skin cells of patients with LCHADD. We will then program the LCHAD deficient stem cells to become retinal cells to create an LCHAD deficient retinal cell. We hypothesize that the LCHADD retinal cells will accumulate long-chain 3-hydroxyacylcarnitines. The accumulation of these metabolites will result in cell death. If our hypothesis is true, we will have the first direct evidence for the cause of retinopathy of LCHAD deficiency and we can develop potential treatments. **We believe this project will further our understanding of retinal cell energy metabolism and provide insights to develop new treatment options for this LCHAD retinopathy.**

Our progress to date is summarized below:

1. We have obtained Institutional Review Board (IRB) approval for this project
2. We have obtained skin cells to use in our experiments from patients with LCHAD deficiency that had skin biopsies at the time of diagnosis.
3. We are testing these skin cells to see how much fat they can burn for energy and how much sugar they burn compared to other skin cells.
4. We have finalized our protocol to transform the skin cells into stem cells and then retinal cells, and we have received Institutional Biosafety Committee (IBC) approval to conduct the experiments.
5. We recently hired a laboratory technician to help conduct these experiments.

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Medical Tidbits....cont'd

Note from Deb: Additional information can be found on the National Institutes of Health's "Clinical Trials" site

Contact Info:

Elizabeth McCracken, MS, CGC at 412.692.5662 or Elizabeth.McCracken@chp.edu

Long Chain Fatty acid Oxidation Disorder (LC-FAOD) Study

A clinical trial in long-chain fatty acid oxidation disorders (LC-FAOD) is looking for participants:

The goal of this 18-month trial will be to study the safety and efficacy of the investigational product Triheptanoin or C7 on people who experience persistent symptoms of rhabdomyolysis, hypoglycemia, or cardiomyopathy related to LC-FAOD.

To be eligible for this trial, participants must:

- Be 6 months to 35 years old
- Have a confirmed diagnosis of VLCAD, LCHAD, CPT2, or TFP
- Experience persistent symptoms of rhabdomyolysis, hypoglycemia, or cardiomyopathy related to LC-FAOD
- Be willing to travel to a clinical site approximately every 4-6 weeks for the first 6 months of the study, and every 12-18 weeks for remaining 12 months

Participants will be REIMBURSED for all travel and study related expenses.

To find out more about the study and to see whether you or your family member might be eligible to participate, see above.



Research Opportunity: Adults with MCAD Deficiency

Dr. Jerry Vockley and his colleagues at the Children's Hospital of Pittsburgh of UPMC are conducting a 7-week evaluation of safety and biochemical changes of the drug Ravicti™ in MCAD patients. Ravicti™ is currently approved for treatment of urea cycle disorders, but laboratory studies in cells have suggested that Ravicti™ may also increase the amount of MCAD enzyme activity.

To be eligible for this study, you **must be 18 years or older and have MCAD deficiency caused by two copies of the 985A>G mutation.** Patients who have kidney or liver failure, or are pregnant or breastfeeding are not eligible. You must also be able to travel to Pittsburgh on four occasions and will be required to stay overnight in the Clinical Research Center for your first visit.

For more information, contact the research coordinator, Elizabeth McCracken, MS, CGC at (412)692-5662 or Elizabeth.McCracken@chp.edu.

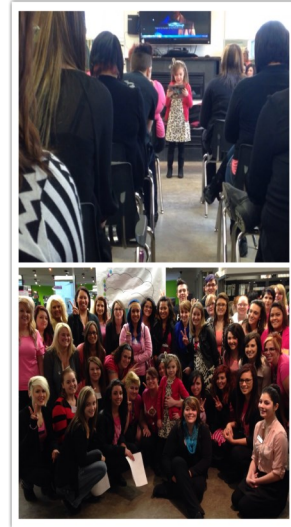




Reach for the Stars!

6 year Karina was a guest speaker for the Paul Mitchell School! She spoke about having MCADD and also her experience with children's hospital ☺she was amazing! So proud of her!!!

Tara Vicencio tkgi4@yahoo.com



Dear Friends,

For all those who have read my book and are on Facebook, kindly "Like" the link <https://www.facebook.com/pages/The-Survivor-The-Hero-The-Angel/52561967419> and "Share" on your own page. My website <http://maryannraccosta.com/> will promote my book, myself as a speaker - to parents, churches, teachers, and healthcare workers, and bring awareness to other "matters of the heart" - all positive things that will inspire and encourage us to love one another.

Thank you for those who have shared the journey . Keep the prayers coming. And, have hope and peace of mind as you find balance in each day!

I am sincerely grateful.

All the Best, MaryAnn jraccosta@hotmail.com

Also, for those that came to the Conference and may have met Maryann ~ she did a webinar on July 23 with National Spinal Cord. At 37 minutes or so she talks about FODs. It's a wonderful and inspiring talk!

<http://www.spinalcord.org/video-raising-a-child-with-special-needs-a-mothers-story/>



Hi everyone,

I thought I would share a blog post that I wrote about my daughter Jordan (21, VLCAD). We just found out that she was accepted to the Disney college program, and this post is about why I am so happy and proud, I hope it resonates with you in some way...

<http://sisterofminehomeagain.wordpress.com/2014/03/08/my-girls-goin-to-disney-world/>

Best,

Dawn, mom to the amazing Jordan (VLCAD) dawnd39@comcast.net

The 'Silver Linings' of FODs ~ What is *your* 'Silver Lining?'

I think most of us have a few of those "moments" when it all kind of hits you and makes you tear up. Usually it's things like the first step, first day at school, graduation. The more time, emotion, and energy invested, the greater the moment. Today wasn't one of the big

milestones, but it was still a moment for me. Stephen is 12 now. He is 5' 9", 160 lbs. and growing, growing, growing. He wears size 13 shoes. He has dark peach fuzz on his upper lip and has fun playing with his changing voice. He used to seem so fragile, so anxious, so fearful. Now he smiles and laughs, makes corny puny jokes, and seems pretty resilient, as long as there is not too much metabolic stress at once. I feel wonder at the tall handsome, and NICE kid he has become. We are one month away from the end of 6th grade, and the end of elementary school. Transitioning to middle school next year. (Not looking forward to that)



As I was dropping Stephen off for school this morning, watching him cross the road and bounce down the sidewalk, I teared up with joy of feeling our successes so far, and felt so happy about this moment in time, when he was so excited for his day. We have a lot of things to deal with next year. But for now, I am so grateful Stephen has come to this point, and that I have made it to this point with him. So much work, so much worry, so much paying attention, so much figuring out. It has all been worth it. Today. Hoping for many more moments like today.

Have a nice day, Diane, Mom to Stephen, age 12, TFP/LCHAD Kent (Seattle), WA dnielsen6@gmail.com

Special Family Article

[Deb Porter](#) has written some wonderful articles based on her husband's [Rodney, adult MCAD, diagnosed at age 36] experience and their experience as a Family, as well as her experience as a former hospital chaplain. She will be contributing several articles over time and they will also be posted on the FOD website in the future on the [Education](#) page. Hopefully, Families can learn from what the Porters have experienced and make some of their own journeys less stressful.

On The Floor Part 1: Nurses

So the ER doctor admitted your loved one with an FOD. What next? This article will be broken down in three parts: Nurses, Doctors, and Food & Nutrition/Nutritionists. Hopefully these will help you through the next part of the process of being in the hospital. The most important thing to remember is that you and the hospital staff are all working for the same thing: to get your loved one well and home again.

Once the doctor has decided on admittance, I explain to both the Dr. and ER nurse that I will not leave my loved one's side unless I am absolutely sure he will be safe. Parents may be expected to stay with their children, but once you have an adult FOD patient, especially one being cared for by a spouse, it raises issues for the hospital. The hospital is technically not supposed to allow a person of opposite sex to be in the room all night. This must be handled with delicate care in order to avoid being bounced by security. I start with, "I understand the hospital rule, however, I have a loved one with a unique set of circumstances. I know exactly what to check for to make certain he is safe. Most hospital staff has never heard of his disorder, let alone what to look for." I explain that I will not leave my loved one's side unless I am absolutely sure he will be safe, because as it says on the protocol letter if we're in the hospital it's because things are bad. While I understand that they also consider their job to ensure my loved one's safety, in this particular case they could miss important cues and by that time it may be too late. This conversation starts in the ER, as the ER nurse talks with the floor your loved one is being moved to next. More than once this has meant that I spend the night in an upright chair, and the next day my loved one gets moved to a private room. Staying calm and being clear while communicating this is essential. I explain the other option to me not staying is a higher level of care (ICU—or other floor where glucose is checked more than once every 4 hours). This typically gets communicated to the floor. Eventually once home and dealing with the financial side of this, it is possible to have insurance pay for the private room, pointing out the other option was the ICU, and next time that will become the choice.

Once on the floor, the RN (registered nurse) assigned will immediately begin asking all the same questions just dealt with down in the ER at least 3 times. Why does this happen? It is to ensure that nothing is missed in the care of the patient. It is to triple check that nothing was wrongly recorded (typos happen, and this can be dangerous in the medical business). So while highly frustrating to go through it all *again* (especially at 4am), know that the reason is to give your loved one the best care and ensure no mistakes are made.

During this time of questions with the first assigned RN, one of the concerns we raise is an immediate request for a nutritionist consult, understanding it won't happen until likely late the next day. We also explain the difficulty of proper food & nutrition and the timing of food. This way, the nurse knows from the start we will likely need RN assistance with food, and that it's a critical part of care. (More about food and nutrition and nutritionists in part 3.)

RN nurses on a regular floor (meaning non-critical care patients, not ICU or post surgical) usually carry a load of between 6-8 patients for a shift. This number continues to rise as hospitals try to operate on trimmed budgets. Nurses start their day with report. This means the nurse that is going off duty explains the needs—medications, upcoming tests, general information, possible dismissal, concerns, etc—to the person taking over. Hopefully, they cover a basic explanation of the fatty oxidation disorder your loved one has. This may or may not happen. This is why it is important to ask the first time the next nurse comes into the room, "Did you get an explanation of (specific FOD) during report?" It is difficult to get a nurse to come to the room during report. Basically an RN won't, unless it is a real emergency. Therefore, be sure needs are addressed with the outgoing nurse by around 5:30am and 5:30pm. Be sure to stockpile any juices or foods your loved one may require while they wait for breakfast or dinner, and make sure that meds including the next bag of D10 are on the floor. The oncoming RN may not appear until 8 or 9am or in the case of evening shift 8 or 9pm—depending on what order patients are seen and their needs. If an IV blows during the report time window, it will be necessary to walk down to the nurses' station, and explain that immediate attention is required, and cannot wait. It depends on the hospital, but report typically happens between 6 and 7am, and again between 6 & 7pm. The RN now begins her initial rounds with patients.

Typical times of stress for an RN are: morning/evening rounds, being assigned a new patient, dismissing a patient, and sending a patient down for tests. With 6-8 patients to care for, a nurse has to continually prioritize. Most are committed to doing a good job, and don't like to disappoint. The RN may need to run interference with food and nutrition. Especially on the first day it can be very difficult to get the proper nutrition for your loved one. Be sure to let both the RN and the CNA (certified nursing assistant) know how much you appreciate their efforts. Some compassion for the difficulty of their job can get you a long way in making sure your loved one gets solid care. It is NOT an RN's job to be counselor or therapist. They may willingly offer to listen and if they do it's fine to accept that. Just try to remain aware that the time they spend in one room means they are not in another.

Both a CNA and an RN are assigned during both shifts. At our local hospital they have gone to a system of cell phones to communicate with nurses. Both the numbers for the CNA and the RN are posted on a white board. The CNA is assigned tasks such as keeping fresh water available, changing bed linens & gowns, emptying and/or measuring bodily waste, assistance with hygiene and showers, getting to the bathroom, and getting other juice or snacks as needed as long as the RN has already communicated approval for it. The CNA can be a good resource to communicate with the RN if she is busy and the CNA is available. The CNA is NOT allowed to touch the IV or dispense any medications. The RN must do these tasks. Additionally, there are some things an RN can't do without Dr. permission. See Part 2 for that discussion. Using this system of calling ahead for the appropriate nursing staff for the need, or using the intercom call button system will make sure they arrive in the room ready to respond, and saves them running down the hall an extra time.

The Charge Nurse is the RN that oversees the entire floor. The charge nurse assigns patients to rooms, nurses to patients, as well as handling a huge other number of tasks (If you don't believe their job is ridiculously hard, google their job description.) Be sure to tell the Charge nurse when people do things right on the floor. Be clear and concise. This builds credibility if something should go wrong. Some charge nurses will keep you with the same nurses, others want to teach their staff about FODs and thus will continually assign a new nurse each shift.

Special Article...cont'd

I let the RN know if I am leaving the floor for any reason, and make sure they have my cell number. I also make sure they see me when I return. This way, they can trust that they have a solid back up with a complicated patient. Finding ways to be a helpful part of the team, yet not intrusive is very important.

There are usually several glucometers used on the floor. Use your own glucometer when they take the first blood stick to compare readings. They can differ by as many as 30 points, even if they have been properly calibrated (though that is an appropriate question to ask). We ask that the same glucometer be used for each glucose reading to ensure as much accuracy as possible.

Finally, if your loved one needs to be transported for any type of medical test that requires they leave the floor, go along if they will allow it. This tends to be less of an issue for children, but can be a huge issue for an adult. If they will NOT allow it, explain to the person doing the transport that they MUST be certain that they hand your loved one off to a trained medical professional who will be doing the testing. Emphasize that your loved one is extremely vulnerable without either a call button or you to alert if immediate help is needed. Before they leave, be certain that the D10 is flowing properly and there is enough in the bag to get them through the time of the testing plus extra. Ask how long they will be gone, and if your loved one is more than 15 min. past that time, explain your concern to the nurse.

Written in support of the FOD community

Deb Porter

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Fatty acid oxidation disorders and athletes

By Chrystine Bliton acbliton@gmail.com

[Note: I'm a bioengineer, not a doctor or nurse, so this info is what I have gathered through reading articles, personal experience, and listening to other peoples' experiences, not medical experience. Still, I think it is sound.]

Fatty acid oxidation disorders (FODs) can lie hidden in an athlete until a combination of metabolic stressors cause a life-threatening situation. This family of genetic disorders can cause sudden death in a healthy athlete. However, there are warning signs of a problem with an FOD, and once the defect is known, there are precautions an athlete can take to stay healthy. These disorders are also one cause of Sudden Infant Death Syndrome (SIDS). The FOD's include beta-oxidation defects in proteins of the mitochondria and peroxisomes, and a few others.

An athlete with an FOD could fall victim to:

- Hypoglycemia resulting in coma and death
- Heart attack from dyslipidemia and enlarged heart
- Dehydration or heat stroke
- Kidney failure from rhabdomyolysis
- Liver failure from fatty liver

Beta oxidation is the primary way that the body turns fats into energy. Slow twitch fibers of the spine, neck, shoulder, and stomach muscles use fats as their primary energy source. The heart is also very dependent on beta oxidation for energy. Consequently, these muscles are most negatively affected by FOD's. FOD patients may also be prone to hypoglycemia under fasting conditions, because the body can't make enough ketones from fats to supply the brain's energy needs once glycogen stores run out. Any metabolic stressor such as extended intense exercise, excessive heat, high altitudes, etc. can speed up the depletion of glycogen stores.

There are a number of clues that an athlete may have an underlying FOD:

- Hypoglycemic episodes
- Difficulty with strengthening back, neck, stomach muscles.
- Neck, back, or stomach muscles get unusually "floppy" after extended exercise
- Neck, back, stomach muscles recovery time is unusually long
- Delayed Onset Muscle Pain (DOMS) is more severe or takes longer to resolve than usual
- Unusual sensitivity to dehydration
- Unusual sensitivity to exercising in hot temperatures
- Difficulty exercising at high altitude
- High triglycerides
- Enlarged heart
- Transient heart failure symptoms
- Fatty liver
- Kidney disease
- Lipomas
- Myalgias from medications that have myopathy as a possible side effects

Special Article...cont'd

Muscle weakness from NSAIDS (Advil)

Increased muscle pain after high-fat meals

Increased muscle pain or hypoglycemic episodes before and during menstruation and ovulation.

(The body favors using fats for energy at these times.)

Muscle pain at night that is severe enough to prevent sleep

The different FOD's may have different combinations of these symptoms. MCAD and LCHAD are two of the most common. Hypoglycemia is often a problem in MCAD, and heart disease in LCHAD. A list of FOD's can be found at this website - <http://fodsupport.org/list.htm> . There are many other resources where FOD information can be found, e.g. NIH, the Muscular Dystrophy Association, umdf.org and elsewhere. The FOD's fall under the general category of "metabolic myopathy". These diseases have been recently added to newborn screening programs, however teenagers and adults will not have had these tests, and so cases that may be 'mild' or usually asymptomatic have been missed. However the diseases can still be deadly or permanently disabling if the person is exposed to intense metabolic stress. One reason why the FOD's have been added to newborn screening programs is because disease symptoms can often be prevented with specific dietary and exercise interventions.

Examples of interventions include:

Low-fat diet

Higher consumption of complex carbohydrates

Avoidance of fasting

Frequent ingestion of carbohydrates especially when exercising

Frequent hydration

Frequent meals

Eating before bed

B-vitamin supplementation, especially riboflavin

Prescription levocarnitine and acetyl-L-carnitine supplementation for all muscles

Creatine supplementation for core and heart muscles

Focused training of fast-twitch muscle to compensate for core muscle weakness.

Monitoring with CK-kinase blood tests if rhabdomyolysis is suspected

Avoidance of alcohol

Suppression of menstruation with hormones

The interventions vary according to the specific FOD, and so it is important to identify which one it is. This is commonly done with an acyl-carnitine blood test followed up by either a genetic test or skin biopsy or both. More information can be found here- <http://fodsupport.org/clinicians.htm>



URGENT NEED for Medical Professionals

With more Families being identified with an inborn error of metabolism (through expanded newborn screening), **our Families will need ongoing Clinical Care from knowledgeable and caring professionals.** In addition to our Newborn Screening Advocacy by many of our Families, our Group is hoping to also bring awareness to medical schools and other medical organizations and facilities the **need for educating and training new Professionals (physicians, metabolic nutritionists etc) in the field of Medical Genetics and Metabolism to treat our children, as well as our FOD adults.** We are also raising funds for Clinical Training.

[see our website for the [donation box](#)]

Once we raise enough Funds we will be able to offer grants to
US Clinical Training institutions.



Parent to Parent Q & A and Suggestions

[Always check with your specialists before making any changes to your/your child's diet or supplements]



Q: How do you explain to a new doctor that we know what needs to be done and the procedures of handling an FOD child without coming across as a 'know it all' because I know I'm not ?

A: from Stephanie ~

I feel like I have navigated similar waters over the years. Not with our metabolic team because that has always stayed the same, but every time Christopher has entered into the hospital I get nervous...new doc, new team...and many of them have not heard of LCHADD before...so how do I not piss them off because I want us to work together as a team.

We have been really lucky because his metabolic dietician has really treated us as a "team member" in Christopher's care, and so she has been much more approachable and I think this has enabled me to get more confident and at times be a little more aggressive when I need to.

All this to say, you definitely want to do what feels most comfortable and in character for yourself. When the clinician comes in I wouldn't hesitate to put yourself out there a bit and while you are introducing yourself take a minute to share some of your background. "I am so excited to meet you, and I am excited to get to know your staff and clinic and how you do things here! We have been immersed in the FOD world for quite a while now, this is what we have been doing _____." It puts you in the lead. It forces them to hear your voice first without having to cut them off because you have already heard that! (We have all done our fair share of nodding I suppose;) Perhaps creating a brief document of previous protocols or what you found helpful at your last clinic and if you can think of any question to engage them (perhaps about new research within the FOD community or concerns)...this will help them to get the feel that you are a proactive parent. Hope that helps a little. Let us know how it goes! And trust your gut!

Stephanie

Christopher, LCHADD 5.5-baseball lover!

srharry374@hotmail.com

Q: What can we do about our adult kids away from home—as far as in an emergency?

A: from Jeannine ~

I am a CPT II patient. Dehydration is a killer for us. Chances are he needed more complex carbs. ALOT more! I don't think the oil would have been tolerated at all given the nausea. Second an "episode" is when the muscles lack so much energy that they can't function, it shouldn't be surprising that his speech muscles can be affected as well. A rhabo flare fills the body with toxins. Anything can happen.

I work in a hospital as well. I am in the same situation of questioning my safety since my husband passed. I worry about what will happen to me in an emergency. I know first hand how quickly things can go bad. Here is some advice:

A: cont'd from Jeannine ~

1. I drafted a protocol letter with instructions about my disorder as well as explicit instructions. I listed all my doctors with their specialties and info on it as well. Attached to it, is my medication list. I then gave a copy of this to all my doctors to have in their file. They all understand, including my family physician, that they could be called upon in an emergency and they will need this. They were all pleased to have it since most of them admitted they don't know anything about this.

2. I wear a medical bracelet that says: CPT II Myopathy, Keep sugar above 90, No propofol/Lact Ringers, See med list in purse. My family also knows these papers can be found in my purse and on my fridge.

3. As many of you are aware there are certain drugs we need to avoid. Propofol is one of them. This can be a problem. Many people think, okay, I will let the doc know I shouldn't have this before surgery. HOWEVER, if there is any kind of emergency and you end up on a ventilator, the first thing they are going to do is put you on a continuous propofol drip! You could be intubated in ER and on propofol before any family members ever arrived at the hospital. They don't bother telling you or your family you are on this drug because it's standard use. I am a respiratory therapist and I know this for a fact. Every one of my vent patients is on a propofol drip.

Take nothing for granted. Preparation is key. He should wear a medical alert bracelet and keep an informational paper in his wallet.

Good luck!

Jeannine, CPT II jeanninemesler@gmail.com Michigan



Recipe Suggestions from Kristy



I am an adult, partially diagnosed FOD. As all of us in this boat, I have to keep careful track of the food. I started using a FREE service that is excellent. I recommend it for everyone, even if you are not FOD.

I used several sites, and settled on this one:

<http://www.fatsecret.com/Default.aspx?pa=m>

Kristy mrsratfire@gmail.com

I also have a blog – check it out! <http://mrsratfirelosesweight.com/>

I like recipes that are simple, quick and can use multiple ingredients items



Love Messages



Please remember our Families in your thoughts and prayers throughout the year
[For entire memory list please refer to our Jan 2010 issue]



Condolences...

'The best and most beautiful things in the world cannot be seen or even touched ...they must be felt with the heart'
~ Helen Keller

It is with great sadness that we learned of two deaths within our 'FOD Family' in the last several months...please send your prayers and thoughts to the Boyer and Cianchette Families

Travis and Krystal Boyer have suffered an unimaginable loss on June 13, 2014 ~ Maisyn (CACT) was 22 mos old



NEWPORT - Emily Hope Cianchette 20, (adult VLCAD) died unexpectedly May 23, 2014 at Portland hospital following a brief illness with her family at her bedside. Emily was born Oct. 18, 1993 in Waterville, the daughter of Christopher A. and Valerie L. (Vicnaire) Cianchette. She was a graduate of Maine Central Institute, Pittsfield in the Class of 2012.

Emily is survived by her parents, Christopher and Valarie Cianchette of Newport, her brother, Matthew P. Cianchette of Newport, her sister, Audra L. Cianchette and her fiancé, Andrew deBethune of St. Louis, MO, her grandparents, Joseph and Greta Cianchette of Detroit, her special aunts and uncles; uncle Tony Vicnaire and family, auntie Laura Cianchette and family, uncle Jim Cianchette and family, uncle Mike Cianchette and family, aunt Tricia Skirnik and family, auntie Amy Donaghy and family, three special friends, Pam Pomerleau, Sandra Neville, Becky Fitts. She will be missed by her special furry friends, Tucker, Flynn and Cooper. She was predeceased by a brother, Benjamin, grandparents, Paul and Florence Vicnaire and uncle Joe.

Private graveside services will be conducted at the Maloon Cemetery in St. Albans. A celebration of Emily's life, with refreshments and fellowship, will be conducted from 1-3 p.m. Saturday, May 31 2014 at Emily's home at 27 Maplewood Road, Newport. In lieu of flowers, friends and family are encouraged to make memorial donations to the Make-A-Wish Maine, 477 Congress St., Suite M1, Portland, ME 04101. Arrangements are entrusted to the care of the Brown Funeral Home and Cremation Service Center, 34 High St., Newport. To sign and online guest book and leave written condolences please visit: [Philip Brown Funeral Home](#)

~ All of our FOD children/adults will ALWAYS be with us in our hearts ~



~ NEEDED FOR THE JAN 2015 ISSUE ~



Medical Update ~ Please Submit to Deb

PROFESSIONAL ABSTRACTS/ARTICLES OF ALL KINDS (Drs, Nutritionists, Genetic Counselors, Social Workers, etc.)

FAMILY STORIES & Pictures for KidsKorner



The 'Silver Linings' of FODs ~ What is your 'Silver Lining?'



Family Fundraisers to Raise FOD Awareness



Hello family and friends ~

This June I am participating in the Franklin Lakes Triathlon, in hopes to raise money for the FOD(Fatty Oxidation Disorder) Family Support Group. Due to a loss of a child this special group was formed. They reach out to families, provide information and spread awareness of rare and dwarf metabolic conditions. This organization is very dear to me because my third child Emma has a very rare metabolic condition called TFP.

Emma's story:

Our 15-month-old daughter, Emma, has a very rare metabolic disorder called Trifunctional Protein Deficiency. TFP it is part of a group of disorders called FODs– Fatty Oxidation Disorders. She cannot have long chain fat. This is the fat that your body needs to feed your organs and keep you stabilized, so you can fast and have reserve energy when you are ill etc. It's found in all oils, egg yolks and all dairy with fat. She cannot have this fat or use fat stored in her body, so her body is in constant overdrive. Her only source of energy is food. A common cold can knock her down and her body will start using muscle for energy, which is dangerous and can be life threatening. Persons with these rare metabolic disorders have to be fed often and/or around the clock (for some), tend to tire quickly, and can have major health issues. Emma is currently tube fed and is suffering from Cardiomyopathy, which is a thickening of the heart muscle. I am hoping to raise money for this amazing Group. If you could help out that would be AWESOME. Thank you from the bottom of our hearts.

Noreen and Rob Weidener XXOO [Emma's Energy](#)

[Note from Deb ~ THANK YOU so much for your generous donations!!!]

ATTENTION FOD FAMILIES ~ FUNDRAISING EFFORT AT ITS BEST!

“The Next Best Thing to Fruits and Vegetables”



Any orders from our FOD members and their families
will benefit [the FOD Group](#)

Whole Food Nutrition is extremely beneficial to those affected with FODs
and those that are not!

Please take a look at my website and click on “watch the video”
beneath the Juice Plus bottles.

Then give me a call or an email to place your order.

Please be sure to tell me you are an FOD family!

CALL OR EMAIL ME WITH QUESTIONS!

Have a healthy and blessed day!

Brenda Goodman

"Sharing Health Wealth!"

(866)280-5726

www.bgoodman.juiceplus.com

doublebn@aol.com



Mom to

Kayla, 13 y, SCADD, Unidentified Mito, Pulmonary Valve Stenosis (repaired), Epilepsy, SLD, PDD-NOS, SID...who knows what else!!!

Naomi, 17y, unaffected, untested, GIFTED-HIGH HONORS STUDENT!

Pepper Pike, OH, USA

Kids Korner



Ben
15 yrs old
MCAD
Michigan



Kayla
13 yrs old
SCAD/mito
Ohio

Briar
7 mos old
MCAD
Ontario Canada



Please note that we also have an FOD KidsKorner/Adults Gallery and other Pictures on our homepage. To submit a pic please email Deb.



- I thought I should share the new **Long & Very Long Chain FOD Food Group**: <https://m.facebook.com/groups/689967947735974/>. This group was made specifically for those that are living with (or know someone who is) a long or very long chain fatty acid oxidation disorder. We discuss tips, tricks and brands that pertain to the often severe fat restricted diets of long and very long chain disorders. Whereas some need to avoid medium chain fats, we are actually looking for medium and small chain fats.

It is a closed group, but I'll get you approved quickly so that our discussions will remain private in case personal information is shared. We can all still meet here for all non food related issues. **Jill O'Malley**

- Ours Family Blog is:** www.harryfamilyblog.blogspot.com

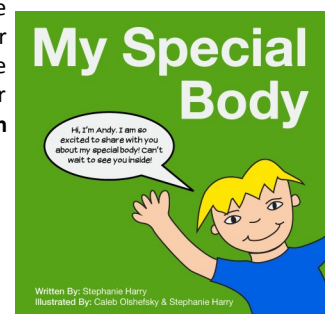
Our blog was originally created when we ran a sprint triathlon to help raise awareness and money for Christopher's clinic. My purpose was to educate as many people as I could about LCHADD and Fatty Oxidation Disorders. Since our race, I have continued our blog with the hopes of bringing hope to other LCHADD/FOD families by sharing our life and experiences. Recently, I have also begun adding recipes and hope to share helpful activities in educating one's child about the body and their condition. **Steph R. Harry** srharry374@hotmail.com

- Rachel Ragosa Quapp – GA 2/MADD Blog** <http://themaddlife.blogspot.com/>

- Emergency Management of Metabolic Crisis slideshows** http://www.slideshare.net/atit_ghoda/emergency-management-of-metabolic-crisis

"My Special Body" is a children's book that was **written for children with LCHAD, TFP and VLCAD deficiency** and was published over a year ago [Written by LCHAD mom, Stephanie Harry]. If your child has been diagnosed with one of these conditions and you have not yet received a copy please visit <http://www.fodsupport.org/book.htm> on our FOD website to fill out an order form. Currently, Stephanie [the author] is trying to reach out to the metabolic clinics and make them aware that the book exists! Her desire is that all families with these metabolic conditions can utilize the book and she can sure use your help! **If your child visits a metabolic clinic throughout the year, if you would consider sharing with them information about the book or Stephanie's contact information this would be very helpful!**

You and/or your clinicians are welcome to contact Stephanie at srharry374@hotmail.com
Stephanie Harry blog www.harryfamilyblog.blogspot.com





DONATIONS

[since our Jan 2014 Newsletter]

Family Donations: Rob and Noreen Weidener (Emma, TFP). Arnie M. Anonymous in honor of McKenna Widmann. Angela Ragosa. Greg S. Andy Driska. Dr Jerry Vockley in memory of Emily Cianchette. Stephanie Harry. Liying Chen & Yong Xie in honor of son, Jianhua D. Xie. Missy Homer, Christine Cox. Chad and LeAnne Pence. William Halvorsen.

Tshirts, Bracelets, Ribbons, CafePress, GoodSearch browsing, MissionFish/eBay selling, or iGive shopping. Brianna Robinson. Jessica Buckley. Jessica Pallett. Joanie Segovia. Kristina Hexamer. Kati Lavi-Waltari. Carol Linthicum. Dustin Harper. Christopher Abrams. Kerry Abrams. Many Conference Attendees.

Thank you to all that have bought products from companies on the Internet that support the iGive and CafePress.com program of donating a certain percentage to Groups like ours. All of those links are on www.fodsupport.org/Donate.htm

Professional Donations: Chuck Hehmeyer. BP America, Inc. Microsoft Matching Gifts Program/Virginia Luchau. Anonymous United Way/Truist donations. Erin Conway via KPMG Community Giving Campaign/Bergen County United Way. Microsoft Matching Gifts Program/Virginia Luchau. Anonymous United Way/Truist donations. Angela Beasley thru United Way of KY. Family Fundraiser thru Everyday Hero program. Network for Good donations as part of Emma's Energy Campaign. Anonymous donations thru AmazonSmile.org. eBay iGiving donor. PayPal Giving Fund. Dr Jerry Vockley in memory of Emily Cianchetti.

We greatly appreciate donations to help with daily costs, website fees, supplies, Conference costs, phone calls around the world, rent for the Grief Consult office, and raising funds for FOD Clinical Training and FOD Research and long-term investments. ALL donations go toward FOD efforts and programs.

US Checks can be made payable to 'FOD GROUP' and mailed to:

FOD Group PO Box 54 Okemos, MI 48805



Reminders

Families - Please send **TYPED (preferably in word document)** stories etc, by **Dec 15, 2014 to Deb.** Continue to spread the word about FODs and the need for screening ~ it will SAVE LIVES!

Professionals - Please let us know about your research and/or clinical work with FOD Families. Send articles, summaries, etc by **Dec 15, 2014 to Deb.**

'We must be willing to relinquish the life we've planned, so as to have the life that is waiting for us'

~ Joseph Campbell

The 2013 FOD Group 990 tax return is on our [Financial page](#)

The bulk of Expenses are for monthly phone, website fees, supplies, Conferences, and for our Grief Consultation office (rent, advertising, etc) to offer pro bono grief support to local Bereaved Parents & Families (and also via Skype to others in the US). We also donate FOD funds from undesignated donations to various FOD related entities (ie., for NBS issues, outreach) to support their efforts.

All Undesignated and Grief Consult donations are deposited into the General Fund or Gen Trust Fund, as are Bracelet and Ribbon Sales, CafePress.com, iGive, GoodSearch, and any donation that isn't specifically designated for the other Funds. Once the Research and Clinical Funds reach a substantial amount (@\$50,000) we will be able to offer grants to clinicians and researchers in the US.

Additionally, we have 1yr & 3yr certificates and long-term stocks/bonds earning interest and dividends for future FOD endeavors and programs.



Thank you to Erika Wallace - erikawallacepa@yahoo.com (Mailing Lists), Mary Lingle - Mcartwrite@aol.com (Website Designer) and Brian Gould - (newsletter consulting) for all your hard work, and to Mark Heinz mark@markheintz.com & Matt Pfeiffer mpfeiffer@danlhos.com for their pro bono consulting expertise on email/website information. Keith Widmann 4wdesign@gmail.com for our website slideshow pictures, the FOD Banner for booth displays, and the USA TODAY Charity Spotlight banner ad. Eileen Shank eshank@helmsbriscoe.com for helping us event plan FOD/OAA Conferences. And all of our Conference volunteers.

The views expressed in the FOD Communication Network Newsletter do not necessarily represent the views of our Advisors or all of our members. Before trying anything new with your child or yourself in regard to treatment, please discuss matters with your doctor or specialist. Please read our Disclaimer on our website ~ it also applies for all communications.



Communicate With Us

Please **ADD** me to your mailing list [Conference years]
 Family Professional (**please circle one**)
 Name/Address or Address Correction (**circle one**)

Please **REMOVE** me from your mailing list:
 Name/Address:

Please include ideas for future issues or your questions
