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From the
Editor

2014 FOD/OAA Conference in July!

The FOD/OAA National Metabolic Conference in Arlington, VA is fast approaching on July 25-26, 2014! Online and phone HOTEL reservations at the Crystal Gateway Marriott [which is where the Conference meetings will be] are being accepted now. A dedicated booking website has been created for our FOD/OAA event so you will be able to make, modify and cancel your hotel reservations online, as well as take advantage of any room upgrades, amenities or other services offered by the hotel.

For ONLINE hotel reservations for our Conference Hotel, please click the following link:  
[https://resweb.passkey.com/Resweb.do?mode=welcome_ei_new&eventID=10749619]  

If you choose to make a PHONE hotel reservation, please use the following dedicated Group Reservations phone numbers in order to access special block rates and ensure you book within the block.

Reservations Toll Free: 877-212-5752  
Reservations Local Phone: 1-506-474-2009

If you call by PHONE be sure to mention this is for the Fatty Oxidation Disorders and Organic Acidemia Association Conference so you get the DISCOUNTED GROUP RATE of $119 [plus tax] per night - and be sure to ask about a REFRIJ [or other amenities like a crib etc] if you need one.

We will have the Speaker Agenda up on our site soon as well as our ONLINE Eventbrite Conference REGISTRATION FORM [every person age 5 and older will NEED to register]—we are hoping to offer free registration for Families Only [Professionals pay $50 per person], but it depends on how much funding we can raise. If we need to have a Registration fee it will be $50 per registrant to cover food costs for the 2 days—Breakfast and Lunch on both days are provided for all REGISTRANTS, and that includes registered sitters and anyone age 5 and older]. We will also post a printed copy of the Registration Form in case you feel more comfortable mailing it to me. FRIDAY the 25th is dedicated to FOD specific speakers/topics [and OAA has their own speakers] and SATURDAY will be our JOINT day with topics similar to both FODs and OAs. Keep in mind we can’t speak on EVERY SINGLE FOD on Friday . However, most of our speakers will mention some specific ones throughout their talk, but because we want to get as much info out as possible and reach as many as Families as possible we try to broaden the main talks. BUT we ALWAYS have breakout sessions for specific FODs and ALL of the SPEAKERS are available on Friday and Saturday for individual questions etc at the FRIDAY NIGHT RECEPTION for Families, Professionals and Families and Professionals and e breakfast and lunch on both days. We may also have a child activity room [activities will include videos, coloring, games etc], but that too depends on our funding and availability of local volunteers. Each Family would still be responsible for having a designated REGISTERED person [ie., grandparent, sitter] for their child/ren in the room, however. The talks and open times with our FOD experts are INVALUABLE to our Families ~ along with the fantastic opportunity to network and get to really know and talk with IN PERSON other FOD [and OAA] FAMILIES from all over the world!

START SAVING and we hope to see some NEW FACES as well as ‘old’ ones in July! DC would be a wonderful Family vacation too—the discounted Hotel rate is also good 2 days before the Conference begins! We are also hoping to offer some Family Scholarships to help with travel/hotel costs—again that depends on our funding. Last Conference we were able to assist at least 6-7 Families with some of their costs. As for DONATIONS that can help with Conference costs ~ We truly appreciate every penny that our members and their Family members donate via cash, buying FOD Awareness items, and doing your own fundraisers. We also have some NEW Awareness Items for sale ~ the FOD Ribbon Car Magnet and the FOD Car Sticker! So please keep us in mind if you are able to donate anytime throughout 2014!

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...

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

‘We Are All in This Together!’
Dear Everyone:

Our 2014 FOD/OAA National Metabolic Conference is for parents and family members, affected individuals, professionals and exhibitors that have an interest in Fatty Oxidation Disorders and Organic Acidemias.

It is only through generous donations and sponsorships that the FOD Group and OAA are able to offer such an important opportunity every two years to Families and Professionals eager to learn more about our disorders. If you are interested in helping our efforts as a donor or a Conference Sponsor please refer to each Group's website linked above or contact Kathy Stagni [OAA Director] directly at mkstagni@gmail.com!

Hotel & Conference:
Crystal Gateway Marriott 1700 Jefferson Davis Highway · Arlington, Virginia 22202 USA
For ONLINE Hotel reservations for our Conference Hotel, please click the following link: https://resweb.passkey.com/Resweb.do?mode=welcome_ei_new&eventID=10749619
PHONE Hotel reservation, please use the following dedicated Group Reservations phone numbers in order to access special block rates and ensure you book within the block.

Conference REGISTRATION FORM: All teen/adult registrants get a FREE FOD or OAA tshirt—be sure to list your size! Other Awareness items and extra adult tshirts and a limited amount of child tshirts will also be for sale AT the Conference.

ANYONE over age 5 MUST register. ONLINE Registration will open soon—the link will be posted on www.fodsupport.org There is NO COST for registration for Families [at this time] but you MUST still Register every person over age 5 to help provide accurate meal counts. Professionals will pay $50 per registrant.

Printed REGISTRATION FORMS [posted on fodsupport.org and oaanews.org] can be mailed to: Deb Lee Gould/FOD Group PO Box 54 Okemos, MI 48805 Professionals using the printed Form please make your checks out to ‘OAA’ and mail everything to me/Deb [Deb handles the Registration but Kathy handles the financial end! Online Registration accepts your credit card.]

Hotel Online Reservations: Attendees are responsible for making their own TRAVEL and HOTEL reservations. Hotel prices will be in effect 2 days before and 1 day after the conference dates of July 25 and 26, 2014. If you have special requirements [e.g., refrigerator, microwave, crib, etc.] please call to check availability and/or extra cost.

Airport: Crystal Gateway Marriott is @2 miles from Reagan National Airport [DCA] Airport shuttle service, scheduled, complimentary Shuttle Phone: 1 703 417 8000 Courtesy phone available Subway service, fee: 1.5 USD [one way] Estimated taxi fare: 10 USD [one way] Please check here for other airport and transportation information

Meals: The hotel chef will be coordinating with us to provide a wide variety of food options for conference meals [Friday and Saturday Breakfast and Lunch and the Friday night Reception are for all Registered attendees ONLY] to include low-fat and low protein foods to accommodate our families’ needs. All other meals are your responsibility. The online and printed Registration Form will allow you to make a list of special dietary requirements needed. The hotel chef will make accommodations whenever possible.

Child Activity Room: We will have an activity room for children - however, each family will be responsible for providing someone to supervise their child(ren) [e.g., grandparent/sitters] and those persons must also be registered. We are also looking for local volunteers to help in the room by providing drawing activities, games, videos, etc. Children are allowed in the conference sessions only IF they can do so quietly. Disruptive, noisy children must be removed from the session as a courtesy to all other participants. Older children and teens are encouraged to attend sessions - every child intending to dine at the conference meals [Friday and Saturday breakfast and lunch and Friday night reception] MUST be registered. ANYONE over age 5 MUST register. All sitters MUST be registered. This is to provide advance accurate numbers for meal preparations.

Kathy and I are hoping to see MANY of you in July ~ you will be empowered to spread even MORE Awareness of FODs and OAs!!!

~ DLG
Nine years ago, I was diagnosed with a disease that has forever shaped my life. Carnitine Palmitoyl Transferase Deficiency Type 2, also known as CPT2, is a disorder that changed every part of who I was, every dream, every goal, and every vision I had ever created. I was a young twelve-year-old girl who was focused on track, the sport that was my everything, fitting in middle school, and having as many friends as possible.

I would have never imagined that my whole world could change in just one doctor appointment, which to this day I remember every detail about. My diet was quickly changed to only consuming nutrients that were low in fat and high in carbohydrates. Gatorade became my new best friend, which I carried with me everywhere, and hospitals and IVs became second nature whenever I got sick. The careless, spontaneous, and adventurous young girl I was rebelled against every aspect of this new life. It has been nine years since I sat in that doctor’s office at OHSU and was given this new future. At that point in time, I cried for nights thinking my whole life was over.

Now, I look back and think I was given this huge opportunity to change my life as well as impact others. I am a twenty-year-old college student that has learned you will never really understand this disease, but you do learn how to cope with it and find different ways to accommodate. Quickly, I realized that my siblings and parents were my main support system and they were being affected by CPT2 in similar ways to how I was. I had to deal with friends who didn’t believe my pain, and coaches who told me to stretch it out and go back in the game. My normal adolescence was not taken, but snatched away from me.

Yes, I will never be a normal young woman, but I have adapted to this new life by keeping a consistent diet, staying away from the germs, participating in as many studies as possible, and surrounding myself with people who are there to lift me up, not bring me down. By having an FOD, you will face obstacles you didn’t even know existed, and find strength within yourself that you never knew you had. I have accepted that I have CPT2, which is something I never thought I would say and because of this disease I am the strong, independent, fighter that I will forever be.

Carmen Shahtout
carmen4life@comcast.net

Our son, Nathan, was born on December 6, 2007. I had an awful pregnancy and was in the hospital a lot hooked up to IVs because of morning sickness. On the day I had him, I weighed the same as when I got pregnant. Nathan was born at 38 weeks weighing 6 pounds, 10 ounces and was 21 inches long. Nathan was an awful eater; he would only nurse for a little bit and when we gave him a bottle, he would only drink a couple of ounces. It did not get better when he started solid foods; he would only take a couple bites. Nathan did not sleep through the night until after he was one.

When he was 18 months old, I was getting Nathan and his older sister ready for daycare and I noticed something was wrong with Nathan. He was so weak. He was not able to hold his head up and his eyes were rolling back into his head. I rushed him to the ER and was told it was inner ear infection and to go home. I was also sick at this time so I thought maybe we had the same thing. He was better within a couple of days. A month later, the same thing happened again. I took him into the clinic and was told to go home and come back when he is better. He started having these “spells” on and off for the next couple of weeks. Another month later, he had a huge “spell” and we took him in again. This time, the doctor finally believed us that something was wrong with Nathan and sent us to the Mayo Clinic. Over the next 3 ½ years, Nathan had tons of ultrasounds, a scope of his stomach, and blood work, but all they came up with was they thought he had migraines, so he got medication to help him with that.
Nathan...cont’d

They decided that he needed a MRI of his brain and spine. On December 26, 2012, we were driving the 40 minute trip from our home to the Mayo Clinic. During the drive, I was noticing that Nathan was not feeling the best, but I thought it was just because he was tired from celebrating Christmas the day before. Our MRI appointment was at 9:30 a.m., and when I got there, they told me that they were running 2 hours behind. I was not happy to hear this because Nathan has not ate anything since supper the day before, but he said that he was okay. While we were waiting, Nathan asked if he could take a nap. I said okay, thinking it would help time pass more quickly for him. After two hours of waiting, I heard a knocking noise. I looked over at Nathan and he was having a seizure. They called a Code and within couple of minutes, there were about 30 medical personal in the room working on him. I was in the corner not able to see him or do anything. He was transferred to the ER across the hall; within 15 minutes, I got to see him and I heard him crying. That was the best noise I ever heard. I was told that Nathan did stop breathing for a little bit and his blood sugar was 25. Nathan was in the PICU for the next 4 days. They did a lot of tests while he was there, but they still did not know what was wrong with him.

During the next month we had a lot of appointments. It is sad to see Nathan not cry anymore when he is getting poked with needles. One of his blood results came back that he might have a FOD. At that time, they did a skin biopsy and we started him on cornstarch. In March 2013, we were told he has SCAD. Since his seizure, Nathan has been a really sick boy. We have had 4 ER visits, a lot of muscle pains, crying and screaming, and a lot more appointments. We have been told by a couple of doctors that Nathan should not be getting that sick from SCAD, so now we are waiting for more results from his skin biopsy. They are thinking that he might possibly have a Mitochondrial Disease.

Nathan has started kindergarten. His teacher says he is doing good at school, but when he gets home he is so tired that he has bags under his eyes and he is limping because has muscle pains that make him whine, scream, or cry. My heart breaks to watch him be so sick, but we take one at a day and I am blessed to have him as my child, along with his brother and sister. Thank you to all of the parents out there on this support group who have helped me. THANKS!

Beth beth_ball@hotmail.com

Emma’s Story ~ TFP

I gave birth to Emma the usual way. Sunday September 2, 2012 at 4:44 am. She is our third child and second daughter. All looked perfect. We were blessed with three beautiful healthy children so we thought. By 6 pm the next night she was lethargic and unresponsive. I told my husband to get the nurse and her BS was 31. They took her to Nicu where she ended up staying for 4 days. On my arrival to get her on that 4th day there were 4 nurses working with her. She had an IV back in, feeding tube oxygen, etc. I had been pumping to give her breast milk and they supplemented with formula and her body couldn’t handle it and she crashed.

Within an hour they redid her NBS and she was put on a respirator. I thought I would die. Her test came back for a fatty oxidation disorder and she was transferred to Mount Sinai NY where she got stronger everyday and came home September 19. The best day ever!!!

We still hadn’t had a diagnosis except that she had a long chain fatty oxidation disorder. I went online and found this support group. All was well and manageable. We thought ok we can handle this. January to February 14, 2013 Emma had slowly stopped eating and got very sick. My husband and I put in an NG feeding tube but her CK levels were very high. Going to hospital. She ended up in the hospital in March and April for bronchitis and never lost the NG. She wouldn’t eat orally at all. The end of April she ended up getting a GTube - best decision we made. It has actually kept her out of the hospital because we can put it on a drip. By then we had a diagnoses.

Our LCHAD came back negative so we knew she had the more severe rare TFP. My husband and I got tested and we both have a mutated gene. We just started researching and trying to find out all about her condition. Here comes the month of May and it’s her first echo since the NICU. I’m at the hospital by myself and her Dr tells me her heart function doesn’t look good. She has cardiomyopathy. My 8-month-old has cardiomyopathy. I don’t know how I made it home through my tears!

We put her on analapril and things seemed to stabilize. Our awesome metabolist found this Dr In Pittsburgh who has had patients with this for the past 25 years. The beginning of July Emma started the new c7 oil. Her next echo was awesome. Her function was normal. We couldn’t believe it. August we headed to Pittsburgh and Emma was officially enrolled in this experimental trial. The experimental oil is called trihepinion. As of now she is still only tube-fed and behind in motor skills, but we are in therapy. And she is making progress. It is going to be a long emotional journey but Emma is a happy joy. The best baby ever. She chose us to be her parents and we are so lucky and blessed to have three beautiful happy children.

Noreen and Robert Weidener Wyckoff, NJ snooks74@hotmail.com Marissa 6 and Andrew 3 unaffected
[Mom, Noreen, will be participating in a triathlon in June and raising money for the FOD Group through crowdrise!]
Medical ‘Bits of Info’

Sudden Cardiac Arrest may be a complication of LCHAD deficiency in young adults [late teens and early 20’s]

We have recently become aware of several cases of sudden cardiac arrest in young adults with LCHAD deficiency. We wanted to make the FOD Family Support Group aware of this abstract and provide some important additional information. The cardiac arrests occurred without obvious signs of rhabdomyolysis or other muscle symptoms. After reviewing the medical records, the patients were not taking significant amounts carnitine or MCT at the time of the cardiac arrest. One had recently seen a geneticist but other patients were not closely followed by their metabolic health care team. This is certainly a potentially concerning late complication of LCHAD deficiency, especially for those not following a prescribed treatment protocol. The best course of action for young adults with LCHAD deficiency is to remain under the care of their metabolic specialist, continue on their prescribed diet and supplements, and follow up regularly with the genetics team. We are continuing to investigate these events in hopes of gaining more insight and potential prevention/treatment options.

Melanie Gillingham, PhD, RD
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Abstract from the ICIEM meeting:

SUDDEN CARDIAC DEATH AMONG YOUNG ADULTS WITH LONG-CHAIN 3-HYDROXY ACYL-COA DEHYDROGENASE (LCHAD) DEFICIENCY
Gillingham MB1, Strauss A2, Arch E3, Arnold GL4, Aleck, K5, Harding CO1

1Oregon Health & Science University, Portland, USA; 2University of Cincinnati, Cincinnati, USA; 3AI duPont Hospital for Children, St. George, USA, 4University of Pittsburgh, Pittsburgh, USA, 5Phoenix Children’s Hospital, Phoenix, Arizona, USA

Three LCHAD deficient young adults (19-21 years) presented with sudden cardiac arrest; one female died and two males were resuscitated. Subjects were homozygous or heterozygous for 1528G>C with a history of classical LCHAD deficiency presenting in infancy. Per family report all subjects had been relatively healthy with no hospitalizations for several years prior to the event; subjects were not taking oral carnitine and were consuming minimal or no medium chain triglyceride (MCT) at that time. Autopsy records of one subject indicate clear vessels and normal cardiac size, suggesting an arrhythmic event as the cause of death. She had a low free carnitine (7 μM) with elevated straight and hydroxyl acylcarnitines measured 2 months prior to her death. Emergency room records indicate two subjects were transported to the hospital in ventricular tachycardia, were cardioverted, and had episodes of ventricular fibrillation before a normal sinus rhythm was established. One subject had a low free carnitine (9 μM) with elevated straight and hydroxy acylcarnitines. No carnitine profile was measured in the other. Sudden cardiac death among young adults with LCHAD deficiency may be an unrecognized late complication of the disease. It is unknown if treatments such as carnitine and MCT supplements could prevent these events.

Deb, here is an example of a letter we wrote for a 4-year-old who was having a colonoscopy. We allowed him to stay home for the three days of Miralax as he could continue to eat his regular diet but admitted him before he started the “day before procedure prep” when he could only have clear liquids. It is similar to our perioperative recommendations for surgery, but for planned surgeries we just admit late in the day the day before your procedure. Let me know if you have any questions.

Judy Miller ARNP
Children’s Hospital of Iowa
Division of Medical Genetics
Iowa City, IA 52242
319-356-2674
Judith-miller@uiowa.edu
~ Colonoscopy Protocol Example ~

*** is scheduled for a colonoscopy on 6/27/11. His diagnosis of medium chain acyl-CoA dehydrogenase (MCAD) deficiency puts him at risk for hypoglycemia and metabolic decompensation with any illnesses, fasting or significant stress which includes surgery and in this case bowel prep. The disease has significant perioperative implications, and awareness about the condition and simple measures can significantly affect outcome.

We have assembled the following recommendations to assist with the pre and post colonoscopy management for ***’ upcoming procedure.

Colonoscopy prep (days 1 - 3) - Specific directions for the prep will be provided by the Pediatric GI service. The first three days of the prep will take place at home and will include TID Miralax. During the three days of home prep, *** will be able to continue on his regular diet and regular dose of Carnitor. Mrs. *** knows to contact the Peds GI service for possible admission with any concerns about his food and fluid intake during the prep.

Colonoscopy prep (day 4)
- Take regular Carnitor dose in the morning
- *** will be admitted by the Peds GI service on the morning.
- **Start IV D10 1/2 NS with appropriate electrolytes at 100% maintenance.**
- Monitor glucose levels using Accucheck.

Colonoscopy (day 5)
- Baseline glucose check before the procedure
- For the procedure switch IV fluids to D10 1/2 NS at 100% maintenance (we suggest to remove potassium from the IV fluids in case the IV infusion rate needs to be increased)

Post-op
- Continue IV D10 1/2 NS at 100% maintenance, but start decreasing the rate after he wakes up and his oral intake progresses.
- Start with calorie containing fluids such as Sprite, juices or sports drinks. Advance diet as tolerated
- Resume regular dose of Carnitor
- Continue to monitor glucose levels

Discharge criteria
- IV discontinued and *** is tolerating his regular feedings (no nausea or vomiting) and maintaining normal blood glucose levels.
- If discharge is delayed due to poor intake, consider adding electrolytes back to IV.

Please do not hesitate to call if you have any questions. You can reach us at [319] 356-2674, Division of Medical Genetics at the University of Iowa Hospital. After hours and on weekends, call The University of Iowa Hospital operator at [319] 356-1616 and ask to speak to the Geneticist on call.

Medical Tidbits....cont’d
Reach for the Stars!

Out of 100 kindergarten kids, Nathan is 1 out of 8 kids who is now in the Gifted Program at his school and he has one of the highest test scores in his grade!!!
Mom Beth beth_ball@hotmail.com

♥ ♥ ♥

This is Tara, mom to Karina, 6-years-old with MCADD! Just wanted to share with you that Karina gave a speech at the Children's Hospital of Michigan specialty center to a group of about 20 doctors, nurses etc., on bringing awareness to MCADD. She also created a book for kids. It's her story and she gave it to the hospital to share with other kids and families with MCADD ~ I am so proud! Her title of the book is 'I have MCADD and it's ok!' She is also bringing awareness to newborn screening in the book by thanking them :)
Mom loves her quote, "the best part about having MCADD is that you can Just be You!"
Tara Vicencio tkgj4@yahoo.com

♥ ♥ ♥

Tanna has been hospital free for 1 year! She was so excited to meet that mark. Now she is going for year 2. Tanna is also a Jr High cheerleader and on October 18th, 2013, she and some friends cut their hair for Pantene Shampoo. They take the hair and make wigs free of charge for other children that have cancer or something else that may cause them to lose their hair. I'm a proud mom for her doing this at such a young age!

Tammy Wheless twheless697@yahoo.com

♥ ♥ ♥

Tommy Waananen, brother to Nicklas (LCHAD) is a level 9 gymnast in northern California. He took first place on the pommel horse and 3rd All Around in season opener meet in Petaluma, California.
Sirpa [mom] jswaananen@comcast.net

♥ ♥ ♥
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.

‘The Hospital Bag’

Having a bag packed for the hospital at all times helps relieve the stress if it is necessary to suddenly go. My hope is that this will be a guide, and that each family can personalize for their specific needs. Having a medic alert tag or other type of similar service in case your loved one is transported to the hospital without you is an important consideration. Medic alert (other) should have the latest protocol letter, a list of allergies (including no propofol use if surgery is necessary), current medications, contacts for doctors and families. Be sure to check that this information is current periodically.

**Paperwork**
- 5-10 copies of the emergency protocol letter (leave original at home)
- Identification and Insurance card
- 3-5 copies of current list of medications, medical equipment (Continuous Glucose Monitor, Cane and CPAP in our case) and allergies (needed for both the ER and the floor)
- List of menus from previous hospitalizations (if applicable—this will help the nutritionists)

**Food (please pack to your loved one’s needs—this is only one example)**
- Argo cornstarch, tablespoons, cinnamon
- Extend shakes w/manual blenders
- Beef jerky
- Licorice
- Gatorade

**Medications/medical supplies (to be packed immediately before leaving)**
- One week supply of carnitine (do not keep this in your car long term as it is temperature sensitive). We have had issues with our hospital having this available.
- Glucometer
- Other (check your medication list above before leaving to make sure nothing is forgotten—it can get complicated for an adult or a child with a g-tube, talk to your doctor if you have questions about what you should bring and what the hospital will provide)

**Clothes and Other Necessities**

△ Clothes for loved one
- If this is a child, set a reminder in your phone/calendar to check sizes every 3-6 months to be sure they still fit.
- For a child, 7 sets of clothing that may or may not come home.
- For an FOD adult, extra socks and underwear and a set of clothes to wear home

△ Clothes for you
- Lots of underwear and socks
- Mix and match yoga pants/sweats
- Mix and match Shirts
- Zipper sweatshirt (to layer for warmth)
- Toiletries for loved one
- Toiletries for you (toothbrush, deodorant, hair ties and feminine hygiene)
- Phone & phone charger
- I keep separate lists for the kids and our pet that also need care during a hospital stay

If you have a baby
- Diaper bag, with enough supplies to get through the travel time and ER wait.
- Breast Pump (if used)

Other suggestions from FOD mom Barb Richmire:
- Pillow and pillow case of my own, 2 fleece blankets or flat sheets
- Easy slip on shoes like crocs, nonslip and washable
- Activity Bag: coloring books, crayons, markers, stickers, other washable toys
- EVERYTHING is either washable or tossable, suitcase has 4 wheels and rolls/spins 360 (Canadian tire $60 for set) and hose it down in shower when home
- A binder with hospital info such as parking rules, cafeteria hours and menu, local restaurants that will deliver, Tablet or computer with books and Netflix, cash/credit cards/or pay as you go refillable MC or Visa.

**Restock the bag when you get home, so it is ready for next time.**
‘Surviving the ER’

The bag has been packed by the door since getting the diagnosis, and it was finally necessary to do the most feared and dreaded thing: Head to the hospital. I’d like to share some tips with you from my experiences as a trained hospital chaplain and someone who loves a person with FOD.

Sometimes contacting your physician and asking them to call ahead will smooth the way. In reality, this may or may not actually help. In my experience hoping for it, but not expecting it has been a solid way to proceed.

Have a copy of the protocol letter in your hand when you reach the check-in desk. As you sign in, be sure to mention that you have the protocol letter. If your physician was supposed to call ahead, mention that also. This person will likely tell you to just hold on to it until someone with medical expertise can help you. The reason to mention it now is that often there is a nurse within hearing distance of the front desk. If so, they will take note. Ask what the current wait time is. Choose a seat near the reception desk, but away from anyone very obviously sick if at all possible. This body language communicates urgency. Children’s ERs tend to be much better than adult ERs, but one day these kids will be adults. Of course, some hospitals are small enough to only have one.

Ideally, in 5-15 minutes, you will be called to a triage nurse who will ask the reason for the visit. Hand this person a copy of the protocol letter, while stating that the patient has a rare condition where the body does not process fat for energy and this condition becomes potentially life threatening when there are other complications in the body. Next, give as many details as you can about what your specific concerns are about the current illness/injury. Be as calm and clear as possible. Explain what steps have been taken at home prior to ER presentation and why they weren’t enough. The triage nurse will take vitals (temp, blood pressure, oxygen saturation levels). At our hospital, the nurse usually asks if he/she can make a copy of the protocol letter. To speed things up, I tell the nurse to keep it, I have extra copies.

Often times the next step is again more agonizing waiting. If there is no room in the ER because it is full, they have no choice but to keep you waiting until one is free. For the staff in the ER, this requires that A) a patient is being moved to the floor which requires a ton of paper work and phone calls as the ER staff must communicate with the floor nurse about the patient they are about to get. B) a patient is being discharged from the ER which also takes paper work. C) If there has been a death, the process is even more complicated and time consuming. Regardless, the room must be thoroughly cleaned and prepped for a new patient.

This does not mean you are helpless. Continue to treat with Gatorade or other supplies you have brought with you. If it is clear that waiting is no longer an option and your loved one requires immediate attention, calmly walk back to the triage nurse and explain that the patient’s condition is deteriorating, and immediate assistance is needed. Be sure to use I statements throughout this process (Learn more about I language here: http://www.dealingwithdifficultpeople.org/i-language-2). For example, “I am concerned that...or...in my experience....” It is normal to feel anxious, and that will likely come out in your voice. This is fine. Getting angry or hostile with the hospital staff will likely not accomplish the goals you seek, however. I can’t emphasize enough how important it is to stay clear and focused. This can be a monumental task after possible hours or days of no sleep and trying to manage at home, on top of the very real fear for the loved one. Breathe. Stay in the line of sight of a nurse until they call you back. It may be reassuring to know that there is a crash cart nearby, but the entire reason you are in the ER is to avoid this.

Once someone finally comes to take the patient to an ER space, we start over from the beginning, explaining that the patient has a genetic condition where the body does not process fat for energy, and the specific details of the illness or injury that brought the patient to the ER. I hand another copy of the protocol letter to the nurse. Most of the time, this nurse will say, “I’ve worked in the hospital X number of years, and have never heard of this.” Affirm that yes, this disease is rare, and it can be deadly. This is why it is important to carefully follow the protocol letter getting labs and D10, ASAP. Try to answer the nurse’s questions as quickly and carefully as possible. If she asks for current medications, I pull out the list I carry in my purse at all times—anything to speed the process.

It is important to remember throughout the time in the hospital that most doctors and nurses have not heard of fatty oxidation disorders—let alone treated a patient with one. During the hospital stay it will be necessary to educate absolutely everyone that interacts with your loved one exactly what VLCHAD, LCHAD, MCAD, SCAD, GAZ, etc. is and how it affects their body. Do not expect the hospital staff to know. They will not. This can create tension as doctors do not like not knowing. They invested years of their lives in school so that they would know. Consequently, careful communication is key.

Another wait, and finally, the doctor will come. If he or she does not walk in carrying a copy of the protocol letter, I hand over yet another copy. It can feel maddening to explain yet AGAIN why you are in the ER. At this step, though, it is imperative that you are concise, and articulate. I usually have to explain to the doctor that while his glucose number would be considered “safe” in a normal patient, it does not reflect the whole picture. It is simply the value that can be measured. What is actually happening is that the patient’s body is being poisoned due to the fat that is not being processed properly in the cell. This is usually enough explanation to have the doctor begin carefully reading the protocol letter, if it has not yet happened.

Then the doctor will need to put in the order for the D10 and other medication as needed, as well as order the labs. D10 will take time as it has to come up from the pharmacy—we have had ER nurses that have gone down to get it to speed the process. The nurse will come to put in the IV, if you have a child they may offer EMLA cream which numbs the area so the insertion isn’t as painful. For those with more hospital experience, where blown veins have caused long term issues in being able to get an IV started, ask if they would consider using ultrasound to get the IV started. Usually at this point, it is more critical to get the IV started. ALWAYS check what is hung on the IV pole, to confirm that is exactly what the doctor ordered. Learn what the flow rate should be for your loved one, and check that it is correct.

Finally, when the doctor returns to see how things are progressing and to discuss whether admittance or discharge is in order don’t be afraid to express concerns. Continue to speak using “I” language. Don’t agree to discharge until patient maintenance without the IV has been established. In some cases, treatment of the symptoms and a bag of D10 may restore your loved one to a more normal equilibrium. In others, admittance may be required. I’ll cover admittance in a future article.

Deb Porter porter.deb@sbcglobal.net
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.

We NEED your help NOW and in the FUTURE so our children will thrive and grow into adulthood with the best of ongoing care!

FOD Awareness project

My 4 year old cousin Maggie has VLCAD, which is part of the FOD metabolic disorders. We are blessed to say that while she has had battles with this disorder, she hasn’t had too many. My aunt and uncle do a great job of making sure she eats properly and doesn’t get sick. This year I joined a group called the National Honor’s Society through my high school which is a group that helps our communities. I saw this as a great opportunity to make a donation to FOD research and awareness. This is why I am making the blue and yellow colored bracelets - to sell so that the NHS can gain money to donate to the FOD Family Support Group. [The donations will be donated through the National Honor’s Society, this is not a fundraiser through the FOD Family Support Group organization].

Brooke Cantrell
bcantrell24@yahoo.com

Ways to Raise FOD Awareness and Funds for the FOD Group!

Over the past several years, we had some Families plan their own FOD Awareness Projects and not only raised awareness but funds for our Group! We even had Families have friends send donations into the Group in honor of their child’s birthday or baptism. Another Family, in honor of their granddaughter, hosted several Silpada jewelry parties and raised over $1000!

If you don’t feel comfortable doing your own fundraiser/project, you can help raise FOD funds by using Goodsearch as your browser or shop online using the iGive site – a portion of your purchases benefit our Group.

As for raising awareness – sharing your story at a local hospital or teaching hospital during grand rounds would be terrific ~ we NEED more clinical professionals in the field of metabolism and this would be a way of exposing them to the challenges of this exciting field!
Q: I am mom to a 4-year-old who has SCAD, and to be honest I don’t completely understand FODs and SCAD. He starts preschool soon and I am concerned he is going to end up in the hospital all the time due to parents sending their kids to school sick. When he gets sick, he doesn’t eat. He has been in and out of the hospital a lot in the past 4 years. I do everything I know to do as far as keeping him away from sick people and watching what he eats, but he goes between my house and his dad’s and his father acts like nothing is wrong. Am I overreacting? I am a young mom and I don’t know everything and I worry all the time about him. Please any advice will help!

♥

A from Rosie ~
First, congratulations on being a great mom. You are aware of your son’s needs, doing your best to meet them and you are trying to get more information. That is all any parent can do. It sounds like what happens when you are caring for your son as good as it can be. For the preschool, your son’s dad and other relatives/caregivers, I suggest:

1. Get simple and easy to understand information about SCAD printed and hand it out. You might be able to find this on the FOD website or from a doctor. The United Mitochondrial Diseases Foundation (www.umdf.org) also has information about mitochondrial diseases, including the fatty acid disorders.
2. Make sure the preschool workers and directors understand your son’s condition and the impact colds and flu will have on him. They can remind the other parents not to bring sick kids to class. Make sure written information about how to care for your son is in his file at preschool and the workers understand the information. Ask the school to have and use hand sanitizer on the kids when they arrive and during the day.
3. Is there any type of food that he will eat when he is sick or not feeling well? I know he is 4, but would he drink formula (maybe from a bottle even)? If you can keep him eating during sickness, you might be able to cut down on hospitalizations which would be good all around. We use chocolate milk, popsicles or sprite when one of our kids is not feeling well (they have to eat frequently). Sometimes that works, but sometimes they still end up in the hospital. Is feeling like throwing up a frequent problem? If so, there is medication that will stop that feeling so he will still eat. That really made our lives easier, since it was a frequent problem here.
4. If you find your son is frequently going into crisis and going to the hospital when he is being taken care of by his dad, perhaps custody arrangements need to be changed.

I was frequently very sick as a child [now I know I may have a mitochondrial disorder] and was raised by a single mother. It’s hard, but definitely doable. Let me know if you need any more help or support.

Rosie
near St. Louis MO
mom to Sarah, 13, carnitine def + mito disorder?
and four other similar kids
jimrosiesarah@gmail.com

Q: Why is using the anesthetic propofol a problem with FODers?
A1 from Jeannine ~
Propofol is as a very short acting drug but it’s ultimate excretion is metabolism dependent and thus dependent on cellular energy production [which is where we all have trouble]. Observations are that propofol is well known to inhibit mitochondrial function at the level of complex I function as well as uncoupling oxidative phosphorylation. In addition, propofol infusion syndrome is thought to result from mitochondrial dysfunction by inhibition of the transport of long-chain acylcarnitine esters with an indirect effect on complex II. Thus, at present, propofol has been shown to affect mitochondrial function by at least four different mechanisms. It is likely that patients with mitochondrial defects may be at increased risk from this drug.

Several reports document that patients with mitochondrial defects may face an increased risk in the operating room with such perioperative complications as respiratory depression, cardiac depression and arrhythmias, metabolic disturbances and occasionally severe neurological deficits. It’s recommended to be used less than 60 minutes on mito patients and only if necessary. There are other choices that can be used. This is type of drug that is given in OR and ICU units only. What you are describing with your daughter sounds like they did conscious sedation [at dentist] and for that they would have used Versed. Versed is quite normal for this type of thing. The time needed to make suture repairs would require a propofol drip because it is so short lasting. They could not have put her on a propofol drip and done such repairs without putting her on a ventilator.
Jeannine, CPT II
jeanninemesler@gmail.com
Michigan

A2 from a medical professional via phone to DLG ~
Some of the other anesthetics that MAY be used for FODers [*of course you want to double-check with YOUR Drs and also depends on the individual child/adult’s specific FOD and other factors]: versed and fentanyl—anesthetic gases might be Sevoflurane and isoflurane - the best anesthetic is one that isn’t metabolized by the liver.

Presidex is a newer anesthetic and can take the place of propofol. [Note from Deb—I HOPE i got this info correct since I was taking notes from a phone conversation. I didn’t get ALL the names but this at least can start a conversation with the Drs. But again, please TALK with your Drs and make sure the anesthesiologist UNDERSTANDS that the fat binding and fat producing anesthetics NEED to be avoided, especially for longer surgeries!}
It is with great sadness that we learned of a death within our ‘FOD Family’ in the last several months—please send your prayers and thoughts to Caroline Dugdell’s [adult GA2] Family in the UK

“It is with immeasurable sadness I must report to all the passing of one of my family’s dear friends, Caroline Dugdell. I’ve just spoken with Carl Dugdell, who has asked me to report this to our support groups. Caroline was an adult onset GA2 patient—who was horribly ill the entirety of her remaining years, after onset. Her case was one of the greatest arguments for NEVER referring to a case as “mild” as many medical professionals wish to claim. Certainly those who like to assert the adult onset patients have much milder cases should take a moment to revisit their positions.

Caroline was always available to me, Mallory Faulkner, and the rest of our family, providing great information on the disease, her experiences—both successes and failures—her time, and all the energy she could possibly muster, to help others with this disease. She was selfless in her sharing and gave incomparable support.

She was one of the sweetest people I have ever had the pleasure of calling friend.

Please keep her family in your thoughts & prayers. Carl & James will need our prayers to face the future without their beloved wife & mother.”

[taken from Terrie Breedlove facebook post]

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

~ Please also send your prayers & thoughts for Dr Stephen Kahler [our very special FOD expert] and his Family ~


Pem was born Julia Pemberton Kelly in Berkeley, CA, to Graham MacDonald Kelly and Julia Ferguson Kelly. She was the oldest of eight. She grew up in San Diego where she met her future husband Stephen Kahler. After graduation from Pt. Loma High School she attended Bryn Mawr College, Yale University, and the University of North Carolina at Chapel Hill (M.D.). She commenced further training in psychiatry, but illness curtailed this in 1985.

She and Steve have one daughter, Jean Stirling Kahler. Jean and her wife Jessica Rowe are the mothers of Graham Curtis Kahler-Rowe. Pem and Steve were married in 1970 in San Diego. They lived in Durham, NC, San Diego, Chapel Hill, NC, Melbourne, Australia, and Baltimore, before moving to Little Rock in 2005, when Steve took a position as Professor of Pediatrics at University of Arkansas for Medical Sciences and Arkansas Children’s Hospital.

She was passionate about all of nature, with a deep love and knowledge of birds and flowers, especially roses; she loved music and was a member of the St. Mark’s choir and the University of Arkansas at Little Rock Community Chorus. She kept up an extensive correspondence (and phone conversations) with dozens of relatives and friends. She read extensively (especially poetry and literature). Curiosity about color vision and perception led to a deep interest in textiles, especially Japanese silk. She was passionate about civil rights and marriage equality. And she maintained an active medical license via continuing education.

She will be greatly missed and lovingly remembered by her family, her siblings Graham Kelly, Jr., Susan Kelly, Molly Kelly, Nancy Kelly Johnson, John Kelly, Sarah Kelly and Anne Kelly, and numerous nieces, nephews, cousins and friends in Little Rock and around the world.

She died suddenly and unexpectedly. Memorial gifts can be made to the Nature Conservancy, the Lupus Foundation of America, or St. Mark’s Episcopal Church. Ruelbel Funeral Home

~ Please also remember Dr Proud’s Family ~ she treated many of our Families ~

Virginia Beach - Virginia K. Proud “Ginny”, 68, passed away on Monday July 15, 2013 in the company of her family after a brave fight with multiple myeloma. Ginny was born in Flourtown, Pennsylvania, and moved to Virginia Beach in 1996 with her husband Ken to be near the ocean. She had an extremely busy and prolific career as a physician, and worked as a Clinical Geneticist at The Children’s Hospital of the King’s Daughters. She showed amazing determination, drive, intellect, and compassion in the care of all of her patients. She could examine a baby without making them cry, showed unerring scientific curiosity and love of learning, was a skilled and gifted teacher, and has been a mentor to many women in medicine. At home she found great joy in roller blading along the Virginia Beach boardwalk, which she did even up until two months ago. She also spent many hours outdoors with her husband Ken riding bikes, hiking along the beach with her adopted geriatric Irish setters, and had just acquired a sea kayak which she did not have a chance to test drive before her passing. She was a beacon of energy and strength for her family, and is survived by her husband Ken, her daughter Laura and her husband Philip, her son Mark and his wife Crystal, and her grandson Huntley. She will be dearly missed. In lieu of flowers, please send donations to the Dana-Farber Cancer Institute for research into multiple myeloma. An informal memorial gathering will be held at the amphitheater on the bay side at First Landing State Park on Shore Drive in Virginia Beach at 7 p.m. on Friday July 19.

Published in The Virginian Pilot on July 18, 2013
EMMA ROSE HERR  
Born: AUGUST 20, 2013 at 9:00 AM.  
Weight: 8 lbs. 15 oz

Emma was a scheduled repeat cesarean delivery. She had low blood sugar in the hospital while mom was attempting to breastfeed. One nurse suggested supplementing her with formula to give her a boost. After a 4 day stay because of the cesarean, we were informed that something turned up on her newborn screening. Instead of being discharged as planned, our baby was whisked away to the NICU. It was a very emotional time for our family. We feel very blessed to have found out about her MCADD when we did. We are following up with Dr Strauss at the Clinic for Special Children in Strasburg, PA. Emma is now 4 months old. We have not had any other hospital stays yet, and are hoping to get through her first year without any problems.

Benjamin and Christine Herr (parents)  
Joshua Herr, 10 years old - waiting on test results  
Samantha Herr, 7 years old - waiting on test results  
benandchris9@frontiernet.net

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.bgoodmanjuiceplus.com  
doublebn@aol.com

Mom to  
Kayla, 11y, SCADD, Unidentified Mito, Pulmonary Valve Stenosis (repaired), Epilepsy, SLD, PDD-NOS, SID...who knows what else!!!  
Naomi, 15y, unaffected, untested, GIFTED-HIGH HONORS STUDENT!  
Pepper Pike, OH, USA
Ashton 5, Hayden, 5 [MCAD], and Kellan, 7-mos-old
Soon to be moving to Germany!

Hayden
5 yrs old
MCAD

Jackson
2 months old
LCHAD
Rhode Island

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

~ NEEDED FOR THE JULY 2014 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?’
DONATIONS  
[since our July 2013 Newsletter]

Family Donations:

T-shirts, Bracelets, Ribbons, CafePress, GoodSearch browsing, or iGive shopping:  

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: Erin Conway via KPMG Community Giving Campaign/Bergen County United Way. Microsoft Matching Gifts Program/Virginia Luchau. Anonymous United Way/Truist donations. Anonymous donation [The FOD Group has chosen to use this donation for 2014 Conference Scholarships - travel and hotel costs for several FOD Families]. 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

We also have a Secure PayPal link on our Donate page
ALL US donations are tax-deductible.
Our 501c3 Tax ID # is 83-0471342.

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.

Families - Please send TYPED (preferably in word document) stories etc, by July 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 July 15, 2014 to Deb.

‘Challenges are what make life interesting and overcoming them is what makes life meaningful’

~ Joshua J. Marine

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