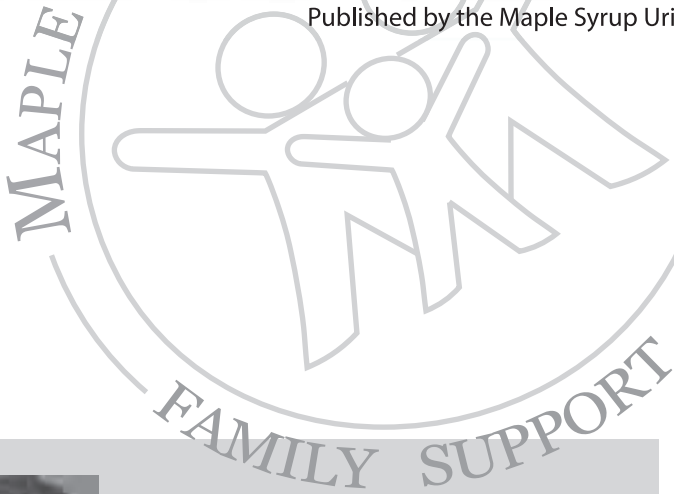


MSUD NEWSLETTER

Published by the Maple Syrup Urine Disease Family Support Group • Volume 26, Number 1 • Spring 2009



MEETING MSUD FAMILIES IN ISRAEL

Karen Dolins, EdD, RD, CDN
Newsletter Editor

As MSUD families from Israel have attended past symposia, I have tried to get an Israeli physician to author an article for our newsletter. Along the way, I made a friend.

My family visited Israel this December. Prior to going, I contacted Dr. Hanna Mandel, at the Rambam Institute in Haifa. Dr. Mandel trained here in the US, and has been a leader in educating professionals and families about MSUD in Israel. I asked her if I would be able to contact her should Hannah become ill during our trip. She kindly gave me all of her contact information, including her cell phone number. Shortly before we left, I received an email from her asking if we would be able to meet. She said she would like to organize a meeting with a group of families. How could we say no? Dr. Mandel and her husband met us at a central location, and she drove us to a Druze village in northern Israel. We were unprepared for the hospitality that greeted us. We were welcomed into the home of a

(Editor's message cont. on page 5)

What is a Support Group? A Perspective from a New Mom with a MSUD Baby By Barbara Mudrick

When our baby was diagnosed with MSUD on the fifth day of his life, our lives were turned upside down. At first we avoided the internet, and were told by close family not to read anything about MSUD. After a few weeks in the hospital, we finally got the courage to go online, and immediately found the MSUD Support Group. Even before our baby was out of the hospital, we mailed our \$10 check to join this support organization. In our hunger for information, we dove head-first and started reading and researching everything we could. We also knew right away that we would be attending the 2008 MSUD symposium to meet other families and hear their stories. I told my husband that it was important for us to have this support group, and become part of the MSUD community. What other choice did we have?

But then at the symposium, I started to question – what is a support group, and what does it really mean for us? I realized that while we are all bound by the same disease, we are all quite different in our approach to this condition, and how we choose to deal with it.

(Support Group cont. on page 2)

Inside This Issue:



<i>Editor's Message</i> cover, 5	<i>Small Town Fundraising</i> 7
<i>What is a Support Group</i> cover, 2	<i>Genetic Disease Foundation</i> 8
<i>Nutrition, Dr. Singh</i> 3	<i>Diet Wise</i> 9,10
<i>Philippine Experience</i> 4,5	<i>Antioxidants & MSUD</i> 11
<i>Valdes Family Story</i> 6	<i>Metabolism Outreach at Tufts</i> 11
	<i>Contacts</i> back page

Page 7 Metabolic Camp at Emory



(Support Group cont. from page 1)

We live all over the country and the world for that matter. We come from different socio- economic and cultural backgrounds. Some of us don't even use the internet. We noticed a few things right away about different ways people manage their kids' MSUD - some eat regular food, and some only eat the medical food. Some of the doctors keep leucine levels higher than others. Some people use filter paper, and some get blood drawn from the vein. We also heard about liver transplants, and noticed right away that some people didn't even want to hear about it, while others couldn't get enough information about it. This was one of the biggest surprises for us. Why aren't people more accepting of the other families? Why weren't there more families who have had a liver transplant at the Symposium?

The more I talked to people, the more varied opinions I heard. But then I realized, that's what a support group is supposed to be - right? Sharing different ideas, different stories, and different suggestions. With such a small number of people with MSUD, we have to remember that we are all part of the MSUD community. It's part of our lives now. Regardless of how you choose to handle the MSUD condition, we should not be judgmental, we should support each other the best we can.

The biggest issue that struck me at the 2008 MSUD Symposium was that nobody talked about fundraising?!?!? We all know that money makes all the difference. Money buys research, money buys treatments, and money buys drugs. Money buys exposure and education. Money is the key to

We met a few families from the Chicago land area that were interested in getting involved in planning a fundraising event. Before we knew it, we had set a date of September 26th. The 4 families involved collected gift certificates and donations to use in our Silent Auction and raffle. A local band donated their time and played for 2 hours, and a local radio DJ was our emcee. We charged \$35 at the door, of which \$15 went to the bar, and \$20 went directly to MSUD. We ended up with over a hundred people and had a great time. The night was a hit, and after all expenses were paid we raised over \$10,000.

How did we do it, you ask? It's really not that hard. You just have to find the right kind of fundraiser for you and your community and dive right in. There is so much information on the internet on how to do fundraising. You don't have to have a huge event at a banquet hall, or bar. Just look at what some of your local schools do or any other organization and you will find something that fits your style.

I realize that we are a very small community, and it is very difficult to raise any significant funds, but if we all got involved we are capable of great things.

The next Symposium will be held July 24-26, 2010, at the Marriott Lincolnshire outside of Chicago. We plan to include a section on "How to put on a fundraiser" and maybe even something on "What it means to be a Support Group." Stay tuned for more information in the next newsletter. If you'd like to help out, please contact me at barbmud@yahoo.com ■

SOCIAL NETWORKING CAN HELP MSUD

Facebook, the social networking website, allows members an opportunity to link their friends, and potentially all of *Facebook*, to a cause or organization they are an activist for. The link occurs by using the application "Causes." Amnesty International, Relay for Life, autism awareness, and more are all linked through *Causes*.

I have started a cause on *Facebook* for MSUD. In doing so I hope to make people more aware of the disorder, and hopefully create fundraising opportunities.

To join the cause, you do need to be a member of *Facebook*, which is free to join. The link is <http://www.facebook.com>.

To join the cause, follow this link

<http://www.facebook.com/p.php?i=509162793&k=5XA4Z3SRT63M5CLGXJ53PQ>

The link to join the cause is on the right. Click on this, and follow the directions.

If you have any questions, please feel free to email Paula Ruter at ruter72@gmail.com.



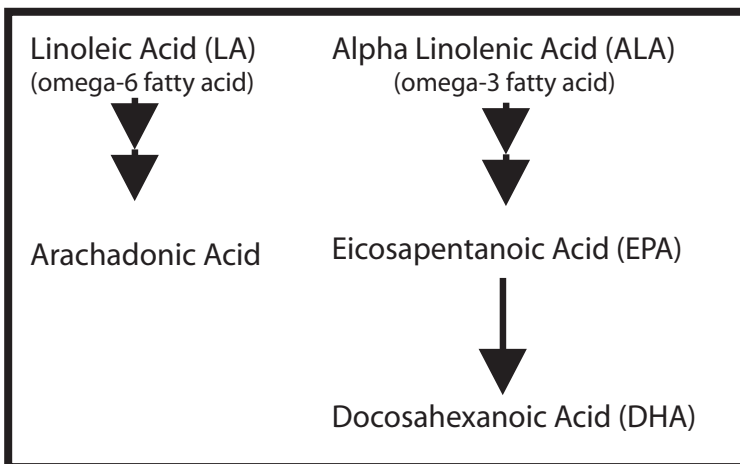
Essential Fatty Acids in MSUD Diets

Rani H. Singh, PhD, RD

Following a protein restricted diet requires patients with MSUD to avoid certain foods, but sometimes avoiding these foods leads to avoiding other important nutrients. An example of this is essential fatty acids.

Essential fatty acids are fats that we must obtain in our diets because our bodies cannot make them. Commonly referred to as omega-3 and omega-6 fats, these nutrients are important for many functions in our bodies, including the formation of healthy skin cells and other cell membranes, brain development, and the achievement of optimum vision. In fact, the brain and retina contain very high amounts of omega-3 and omega-6 fatty acids, which demonstrate their importance in these tissues. Omega-3 fats have also been shown to have help protect against heart disease.

Many types of omega-3 and omega-6 fatty acids exist. Fortunately, the body has the ability to use the essential fatty acids alpha-linolenic acid (in the omega-3 family) and linoleic acid (in the omega-6 family) to make the other omega-3 and omega-6 fatty acids. Alpha-linolenic acid (ALA), which is found in nuts, flaxseed and canola oils, can be converted to other important omega-3 fats like eicosapentaenoic acid (EPA) and docosahexanoic acid (DHA), in small amounts. Linoleic acid (LA), found in corn, sunflower, and safflower oils, can be converted to the omega-6 fatty acid referred to as arachidonic acid (AA) as well as other omega-6 fats.



A recent evaluation of a small group of patients with Maple Syrup Urine Disease showed that the plasma levels of DHA were significantly lower in MSUD patients than in similar individuals without MSUD (submitted for publication). There was no difference in the plasma levels of the essential

omega-3 fatty acid ALA between MSUD patients and those without MSUD. The normal ALA levels and low DHA levels in this small number of MSUD patients raises the question about adequate fat intake. Other studies have shown that plasma DHA levels fluctuate to reflect dietary intake, so the lower levels in MSUD patients reflect a lower intake of DHA. More research is needed with more subjects to evaluate essential fatty acid intake from diet and the choice of medical food.

So in the meantime, what can we eat to make sure we are getting these essential fatty acids in our diets? Omega-6 fats are easily obtained in the diet through corn, safflower, and sunflower oils and should be adequate in MSUD diets where these plant oils are not restricted. The omega-3 fats are more of a challenge to include in the diet. ALA is found in canola and soy oils, which are easily available in marketplaces. Other more expensive sources of ALA include walnut and flaxseed oils; DHA and EPA are found in fatty fish or fish oils. Obviously, fish and nuts are not allowed in MSUD diet, but soy or canola oil can be added to the MSUD diet.

Ongoing clinical monitoring of plasma levels is used to establish adequacy of essential fatty acids in patients with MSUD. Further studies will clarify if supplementation with DHA and EPA ("fish oil" capsules) is needed. Nonetheless, paying attention to total and essential fatty acid intake in the MSUD diet is important for optimum development. Ask your dietitian to calculate total fat and essential fatty acid intake for your diet. Fatty acid data for different foods are available on:

<http://www.nal.usda.gov/fnic/foodcomp/search/> ■

Sources of Essential Fatty Acids¹

Source	Amount (5ml)	Linoleic acid (mg)	Linolenic acid (mg)	Calories
Canola oil	1 tsp	915	420	40
Corn oil	1 tsp	2410	50	40
Safflower oil	1 tsp	3360	0	40
Soy oil	1 tsp	2295	305	40
Walnut oil	1 tsp	2380	470	40
Flax oil	1 tsp	570	2400	40

¹ Data from
<http://www.nal.usda.gov/fnic/foodcomp/search/>

Challenges in a Developing Country: The Philippine Experience

Carmencita Padilla MD, MAHPS

Professor in Pediatrics, UP College of Medicine

Director, Institute of Human Genetics – National Institutes of Health, University of the Philippines Manila

The Philippines is an archipelago of 7,107 islands located in the southeast coast of Asia. It has a total land area of 300,000 square kilometers and separated by three main islands called Luzon, Visayas and Mindanao. It has 17 political regions and Manila is its capital city. It is the twelfth most densely populated country in the world comprising 89 million Filipinos as of August 2007 and 35% of these are children aged 0-14. About 2 million babies are born per year and a family has an average of 3-4 children. The top 10 causes of infant mortality include bacterial sepsis (infections), respiratory distress, pneumonia, prematurity and its complications, congenital malformation of the heart, neonatal aspiration syndrome, other congenital malformations, diarrhea and other gastro-intestinal diseases.

The incidence of MSUD in the Philippines is not known. The first patient with MSUD in the Philippines was diagnosed in 1992. The 7 day old boy presented with the characteristic odor. Of the 76 cases identified since 1992, only 20 are still alive. The majority are diagnosed when signs of metabolic decompensation have set in. One patient was diagnosed as early as the 2nd day of life. Mom had another child with MSUD and she noted the 'typical sweet odor' while breastfeeding this baby.

The first major challenge is the early diagnosis of the babies with MSUD. The early symptoms of poor appetite/feeding, irritability and incessant crying mimic neonatal sepsis, a very common condition among sick neonates in the Philippines. A wrong diagnosis is made unless the physician has a high suspicion of MSUD. Most of the older patients were diagnosed late with onset of complications. The more recently diagnosed patients have better performance.

Most physicians are not aware of MSUD. In the Philippines, more than 60% of newborns are home-delivered and are attended by midwives and traditional birth attendants who also cannot recognize the early signs of MSUD. We suspect that many more babies with MSUD are born but are misdiagnosed as having infection and succumb to death without proper evaluation.

The second major challenge is the cost of the diagnostic examination. MSUD is not yet included in the newborn screening panel. Plasma level determination of leucine, isoleucine and valine is very costly (US\$180). Cheaper alternatives for diagnosis are 1) urine metabolic screening using high voltage electrophoresis which show distinct bands for branched chain amino acids (US\$60); 2) Thin Layer



The Philippine delegation at the MSUD Symposium, 2008

Chromatography (TLC) (US\$6); and 3) Urine screening for 2,4 dinitrophenyl-hydrazine (US\$12). TLC is the most commonly used. All these are generally not affordable for the majority of the families of MSUD patients earning minimum wages of US\$ 8 a day.

Since most families are young, the third major challenge is diagnosis in subsequent pregnancies. MSUD is always considered in succeeding pregnancies of mothers with affected children. Out of the 76 patients identified, 36 families have agreed to undergo mutation analysis. Results showed that 40% of the patients have a novel mutation specific to the Filipino population. Families with known mutations are offered testing for subsequent children at birth. If confirmed positive with mutation analysis, nutritional management is started immediately. If negative for the family mutation, the patient is cleared and started with unrestricted breastfeeding and regular formula. If the family mutation is NOT available, the newborn is subjected to protein challenge guided by leucine level monitoring.

The fourth major challenge is acute and long term management. The majority of the patients undergo peritoneal dialysis as the method for rapid removal of toxic substances. In the Philippines, not all nephrologists are comfortable with dialysis in a neonate and not all tertiary hospitals are capable of handling acute management of MSUD. MSUD milk formula is not readily available in the Philippines. Although Mead Johnson donates several boxes to the group of patients, the supply is not enough and parents have to buy MSUD special formula which costs US\$70 per can, inclusive of taxes and shipping. A major crisis happened last year when Mead Johnson did not provide milk for several months. The donation of the MSUD Support Group USA was very timely.

(Philippines cont. on page 5)

(Philippines cont. from page 4)

A volunteer dietitian assists with the preparation of special low protein menus using local food items. Special food items are likewise not available in the country. And even if made available, the cost of these special food items are not affordable to the families.

The fifth major challenge is follow-up of patients, which is very critical for MSUD management. Not all patients are following up regularly. Not all patients are following the dietary protocol. Not all patients are undergoing therapy. There are only a few clinical geneticists in the country and they are concentrated in Metro Manila. A Metabolic Clinic at the Philippine General Hospital (PGH), the University teaching hospital organizes the follow up of the patients as well as coordinates with the donated milk formula with Mead Johnson. Only 1 other metabolic clinic (Cebu) exists outside of Metro Manila.

The sixth major challenge is securing support from government. The Institute of Human Genetics at the National Institutes of Health- Philippines has been the main institution that has been helping the MSUD families with access to diagnostic and treatment options. In 1993, an informal support group among MSUD family members was established. The families have been supporting each other during crises on MSUD milk shortage. They offer emotional support to new families. Sharing of recipes is an interesting activity of the families. This group was formalized in 2006 as the MSUD Parent Support Group and eventually joined the Philippine Society for Orphan Disorders, an umbrella organization that addresses the needs of patients with rare genetic disorders.

The Future:

The challenges in the Philippines may be difficult to overcome but the professionals and the families have continued to face them with optimism. There are preliminary discussions on the inclusion of MSUD in the newborn screening panel. President and General Manager Paul Richards of Mead Johnson Philippines recently committed to provide a complete supply of MSUD special formula to the 20 living patients. There are efforts to set up an endowment fund to secure the needs of the patients and future babies with MSUD under Philippine Society for Orphan Disorders. ■

(Editor's message cont. from page 1)



Nada, Yuval, Hannah, Shada and Ramzi enjoy a feast of low protein food in Israel

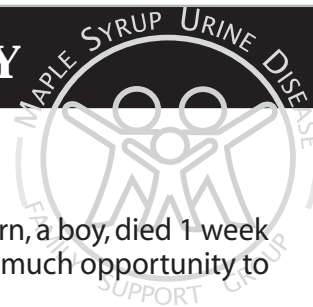
MSUD. One entire table was covered with homemade delicacies, and a second table was covered with low protein goodies. Six families convened from all over northern Israel to meet us. They all wanted to know how we treat MSUD in the US. Most of the children looked healthy and happy. One girl had been diagnosed late and had obvious physical impairments. None spoke more than a few words of English, but the kids all got along fabulously. In fact, 1 girl Hannah's age invited her to sleep over! And she did! The mom told me the next day that it was a wonderful experience for her daughter, and I know it was for mine as well. Dr. Mandel works tirelessly to keep her patients healthy. She tries to get them to visit her monthly for blood tests. Often she travels to them. The dietitian who works with her uses the exchange system for dietary leucine. From what I saw of this group, they're doing a great job.

Israel is more diverse than many think. In our group of 6 families, 3 were Druze, 2 were Arab, 1 was Christian, and 1 was Jewish. As we all know, MSUD unites us all. We are one family in our struggle against this disease. ■

Words to inspire...

"What lies behind us and what lies before us are tiny matters compared to what lies within us."

~ Ralph Waldo Emerson



The Valdes Family

Monterrey, Mexico

We live in Monterrey, Mexico, which is only 250 miles south of San Antonio, Texas. Our first born, a boy, died 1 week after he was born in 1979. He wasn't fed from his mother and he suddenly collapsed without much opportunity to know what had happened. We now imagine that he probably had severe MSUD.

Our daughter Melina was born in 1980. At the age of 6 months she was noted to have developmental delays. Several doctors said the cause was a light cerebral accident which probably occurred at the time of her birth. We went to many places for second opinions, including Houston, Austin, and Mexico City, and Melina underwent many tests. Although she was tested for MSUD, this was done with a urine sample and was negative. Through the years her condition deteriorated badly. When Melina was 9, we knew she had classic MSUD and realized that the improper food she was taking caused her brain damage.

When Daniela was born 9 years later in 1989, she was fed from her mother for the first 6 months. When she started milk from formula and other foods, Daniela began to show the same symptoms as Melina. The doctors asked for complete blood tests on Daniela, and she tested positive for MSUD. We immediately had Melina take the MSUD blood test again, and this time hers was positive too.

After we knew about the MSUD, we got immediate assistance from several doctors at Hospital San Jose (part of the school of medicine from Monterrey Institute of Technology University). Our cases were probably the first in the State, but we had good support from the IUC pediatric director and neurologist director of that Hospital. They researched MSUD extensively, helped our daughters properly, and advised us about MSUD and the strict treatment we must care about.

The girls were medically followed very closely for the first 10 years. Before they were 6 years old, Melina had 2 very serious crises and Daniela only one. As they grew older (10+) they have been fairly well and without a crisis.

In regards to the diet, we attended the Nutritional Clinic of the medical school from the local State University (UANL). They also were willing to help. They researched MSUD and assisted our daughters during the first 10 years.

By now Daniela and our selves have had a lot of practice managing the diet, but we are still very careful about portion sizes and of course the low protein foods. Daniela and Melina have Ketonex 2 complement formula every day, which we get directly from Abbot Laboratories located in Mexico City.

Today we know that the UANL Nutritional Clinic is helping 3 other kids with MSUD (5 to 10 years old). The IMSS (public health system) provided our information to their parents and



Meli & Oscar Valdes, with daughters Melissa, Daniela & Melina

we are sharing with them all we know.

As you can see in the picture, Melina, now 28 years of age, is a very nice but quiet girl. She is always happy and she smiles most of the time to communicate what she needs and wishes, but we must assist her with everything. Sadly, her condition is getting worst every year, and we are now fighting against secondary but terrible problems like epileptic crisis and the most dangerous one, pneumonia due her immobility and spinal scoliosis. We know every year is going to be worse for her and we are doing as much as we can without affecting Melina dramatically.

Daniela, now 20, finished high school last year and is studying to be a kindergarten teaching assistant. Daniela even is assisting my wife's sister kindergarten school and may work there eventually. She loves little kids and she is a very lovely and very happy young girl, caring about her friends, always willing to help and keeping herself healthy dancing and cycling.

Daniela cares a lot for her diet and she is growing almost normally: Daniela surprises us how well she manages her food and diet while at home, school, restaurants, outdoors,... she always carries emergency low protein food bars and candies in her personal bag, just in case. She is not good in mathematics and has difficulty with fine motor skills, but she reads and speaks very well Spanish and understands some English, enough to communicate. Eventually she will be practicing

(Valdes Family cont. on page 8)



15th Annual Metabolic Camp at Emory University, Atlanta, GA

Join us June 22-27, 2009 for the 15th Annual Metabolic Camp at Emory University in Atlanta, GA! This is a model, research-based camp for young women 12 years age and older with PKU and MSUD, which focuses on building social support through a variety of activities including nutrition education, cooking classes, discussion groups, and local field trips. The camp typically accepts 30 attendees on a first-come, first-served basis. Registered dietitians

from across the nation and around the world volunteer their time to serve as camp counselors, and nutrition students provide support as assistant counselors. The cost of the camp is \$325 per person, which includes all sponsored meals, lodging, group activities, field trips, and closing banquet. Partial scholarships are available for certain financial circumstances. Check with your local RD and clinic to see if local sponsorship is available.

For more information about the Metabolic Camp, please contact Rosalynn Borlaza (Camp Coordinator) at (404) 778-8521 or rborlaz@emory.edu .

Small Town Fundraising

My name is Amy Jones. My husband Dean and I have a 6 year-old son, Derek. He was diagnosed with MSUD at 12 days old. We live in Barberton, Ohio. It's just a little town south of Cleveland and Akron. One of those cities where everyone knows everyone and their business. But we don't mind that. We love it here.

In 2005, we decided that we wanted to get a little more involved in searching for a cure for MSUD. We thought that if we held a fundraiser, maybe we would be able to raise some money to help with research projects that we had recently learned about that were going on. For our first annual MSUD Family Support Group Fundraiser, we had a Reverse Raffle at our local Moose Lodge here in town. We advertised in our local newspaper and hung flyers in grocery stores, our library, our relatives' places of employment and more. We had a slow start, but once the word got out about what we were doing and why, we quickly sold all of our tickets and had to turn people away. That year we were able to raise \$4600! Pretty good for our first time!

Our second year, we were approached by a group of classic car collectors here in town, The Magic City Cruisers. They wanted to hold a benefit car show in Derek's name and have all of the proceeds go to the MSUD Family Support Group. We actually did this 2 years in a row. We raised a total of about \$4,000.00 with these 2 events. Just this past year, October 2008, we thought that we would try yet one more type of fundraiser. We opted for a "Night at the Races". Wow, what a great time! Once again, we were sold out and raised around \$4000.

It's amazing what people living in a small town are willing to do when it means helping to improve the quality of life for a child.

Amy Jones



The Genetic Disease Foundation

was formed 11 years ago by several organizations and families affected by genetic diseases sharing a common bond. The diseases involved affected small populations with horrific and life threatening diseases. Some of the family members had succumbed to these rare diseases.

Recognizing the difficulties faced in combating these orphan diseases, the Foundation founders adopted the philosophy to unite resources to raise research funds for genetic diseases in general, while also promoting awareness of these diseases.

The Foundation designated the Department of Genetics and Genomic Science at The Mount Sinai School of Medicine in New York City, one of the largest genetics programs in the US, as its "Center of Excellence" and the recipient of its research grants. In so doing, the GDF has recognized the past accomplishments and dedication of the department to innovative efforts to discover cures and/or treatments for these rare genetic diseases. Since genetic and especially metabolic diseases, share common bases, an advance in one, often leads to advances for the others.

Since our inception, The GDF has raised several million dollars and has used these funds towards the purchase of state-of-the-art research equipment for the Department. Recent purchases have included a mass spectrometer for critical analyses for the diagnosis and monitoring of various metabolic disorders and a NextGen DNA sequencer to identify the genetic mutations causing these diseases. These are the most advanced research instruments for studies of disease pathogenesis and evaluations of new treatment. This equipment will facilitate efforts to prevent and develop new treatments for metabolic diseases including MSUD and PKU. Additionally we have invested in research grants that have been given to scientists involved in genetic research. While raising money for research the GDF has concurrently spearheaded several campaigns reaching tens of thousands of doctors including the "One Test 16 answers" awareness campaign encouraging a simple screening test to identify carriers of gene mutations for 16 genetic diseases.

The continued development and research of genetics has enabled the Foundation to continue its mission through expanded testing and knowledge.

Since the GDF is staffed by volunteers, all funds are directed to our research and awareness efforts. To find out more about our organization, visit our website at <http://www.geneticdiseasefoundation.org>. ■

Submitted by Stan Michaelman

(Valdes Family cont. from page 6)

fluent. So we know she is able to manage 2 languages. Throughout the years she has needed some "growing" assistance but her sister Melissa, 24, helps Daniela a lot. Melissa does not have MSUD. Unfortunately she has optic nerve coloboma in both eyes. But she is so smart. She just graduated from college with the highest honors, she is fully bilingual English-Spanish and now works for me as a SAP certified consultant in my company. Eventually we are planning to start a family business together where Daniela can work productively and independently, and be self motivated.

Meli (my wife) and I do not have cases like these in our respective families; however at the end we are all together a very happy "normal" family, challenging MSUD. We always dream of helping our daughters soon, and were thankful to find the MSUD support group web site 6 years ago. It is our main source regarding this important issue. The impact of this organization, especially when we parents urgently need simple but key information, is extremely valuable. You may not imagine how good it is for many families like ours when it is needed, and how grateful we are. No matter we do not live in the US, we are just one mouse click away. We hope to meet some of you soon and to learn from you. Maybe Chicago is the time. ■

Blessings...

Meli and Oscar Parents to Melina (28) and Daniela (20)
Classic MSUD Monterrey, Mexico

SAVE THE DATE!
MSUD Symposium 2010
July 24 - 26
Marriott Lincolnshire
(outside of Chicago)
More information will follow
in next newsletter...



Great Dishes for SPRING!

Easy Meal Ideas for Busy Families

Here is a collection of ideas for simple meals especially for families on-the-go, from the experts: other families who are members of the PKU Listserv group (to join our large group of about 1400, e-mail Lin at macpku@verizon.net).

Quick meat substitute: Sauté hash brown potatoes or finely chopped fresh mushrooms and add a few tablespoons of Sloppy Joe sauce (Manwich or similar brand). Add a little taco seasoning, and use this as a sandwich filling or as a meat substitute for a taco salad or for tacos.

Onion rings: Use one of the low pro pancake batters for excellent results. Dip the rings in the batter and deep fat fry, then add salt as soon as they're out of the oil.

Mock chicken and rice: Add a little canned mushroom soup and chicken bouillon to low protein rice. Even without the chicken, the rice has great flavor.

"Mock stir-fried chicken": Cut cauliflower into bite-sized pieces and weigh your desired portion for figuring phe (or leucine) content. Stir-fry the cauliflower in hot oil until barely tender. Then sauté a little chopped onion and add a tiny bit of soy sauce for a dish that looks much like stir-fried chicken.

Please send recipes to
Food News Editor

Glenda Groff

515 W. Church Road, Ephrata, PA 17522
Ph: 717-738-4793 • ernieglenda@dejazzd.com

Chex School Fuel- Dawn Hahn

- 3/4 cup brown sugar
- 6 tablespoons butter
- 3 tablespoons corn syrup
- 1/4 teaspoon baking soda
- 8 cups Rice Chex
- 1/4 cup semi-sweet chocolate chips

Melt butter in sauce pan over low heat. Add brown sugar and corn syrup. Stir until well combined. Add baking soda. Mix well. Pour over Rice Chex and stir until coated. Bake at 200 degrees in two 9"-13" baking dishes for 1 1/2 hours. Stir every 15 minutes. Melt chocolate chips; drizzle over Chex. Store in an airtight container.

Makes 8- 1 cup servings

	Protein	Leucine	Calories
per recipe	13.8g	1076mg	2436
per serving	1.7g	135mg	304



Fresh Vegetable Salad

- 2 cups broccoli florets
- 2 cups cauliflower
- 1/2 cup celery, chopped
- 1/2 cup green peppers, chopped
- 1/2 cup onions, chopped
- 1/4 cup grated carrots
- 1 cup Miracle Whip
- 1/4 cup sugar

Toss vegetables together in a large salad bowl. Combine Miracle Whip and sugar together. Pour over vegetables and toss to coat. Cover and chill. Makes 8 2/3 cup servings

	Protein	Leucine	Calories
per recipe	12.2 g	624 mg	1473
per serving	1.5 g	78 mg	184

Note: If desired, remove 2/3 cup salad for low-protein diet, then add 2 bacon strips, fried and crumbled, and 3 tablespoons grated parmesan cheese for the rest of the family. Serve.



Apple Salad

- 5 apples, peeled and diced
- 1- 27 oz can crushed pineapples
- 2 cups miniature marshmallows
- 2 cups Cool Whip

If apples are tart, Sprinkle 1/2 cup sugar over them. Drain pineapples and mix with remaining ingredients. Chill. 10 servings

	Protein	Leucine	Calories
per recipe	9g	570mg	1780
per serving	.9g	57mg	178

Note: after removing portion for low protein diet add 1/4 cup nuts to family portion if desired.



Research Update:**Antioxidants and MSUD****Karen R. Dolins, EdD, RD, CSSD, CDN**

Last winter we told you about the importance of antioxidants, and how they may be particularly helpful for those with MSUD. In the past year, several additional studies have been published on the subject, so we thought it was worth bringing up again.

As a refresher, or for those of you who missed last year's article, antioxidants are naturally occurring substances that battle free radicals. Free radicals are molecules that are very unstable and cause damage to cells in our bodies by reacting with oxygen and other substances. They are formed every time we breathe as a result of normal respiration, and can also be generated by external agents such as smog, tobacco, and stress.

Our bodies have a natural system for battling free radicals, called the antioxidant defense system. Sometimes, though, our ability to neutralize free radicals cannot keep up with the generation of new free radicals. This is called oxidative stress. One study published by Quental and colleagues (Molecular Genetics & Metabolism 2008) reported increased markers of oxidative stress in 7 MSUD patients in Brazil. While all of the children were currently being treated for MSUD, it must be noted that they had been diagnosed at variable ages and that all suffered from neurological complications of the disease. In a personal communication with the study's author, it was learned that compliance with the diet in these patients was poor. Whether the same would be found in children who comply with the diet is unknown.

Clearly more research is needed in this area. Studies of this nature would be strengthened by including a dietary analysis so the antioxidant content of the diets would be known. Meanwhile, we should all be encouraging our children and adults with MSUD to favor fruits and vegetables in their diets. US Dietary Guidelines recommend 2 cups of fruit and 2 cups of vegetables for adults every day.

Interestingly, a paper published in the Journal of the American Dietetic Association (Phillips 2009) reviewed the antioxidant content of various sweeteners. They found that refined sugar, corn sweetener (used in most processed foods) and agave nectar had negligible antioxidant content, while dark and blackstrap molasses had the highest, followed by maple syrup (real maple syrup, not maple flavored syrup such as Aunt Jemima), brown sugar, and honey. As the MSUD diet relies on sugar as a significant source of calories, substituting these less processed sugars for more highly processed forms can improve the antioxidant content of the diet. ■



TEACHING MEDICAL STUDENTS & DOCTORS ABOUT MAPLE SYRUP URINE DISEASE

The Metabolism Service at Boston's Floating Hospital for Children has launched the

METABOLISM OUTREACH SERVICE

This innovative program represents the largest and most comprehensive program to date to educate medical students and physicians about metabolic disease. At this time, it serves 6 academic medical centers in the north-eastern U.S., and provides:

- Seminars & workshops about metabolic disease and abnormal test findings
- Opportunities for patients/families to help teach medical students and doctors
- Consultative assistance to physicians as they assess their patients
- Long distance support for these medical centers during more urgent situations

Participating centers include:

- Boston Medical Center - Boston
- Baystate Medical Center - Springfield, MA
- Vermont Children's Hospital - Burlington, VT
- Dartmouth-Hitchcock Medical Center - Lebanon, NH
- Eastern Maine Medical Center - Bangor, ME
- Maimonides Medical Center - Brooklyn, NY

WE ARE LOOKING FOR PATIENTS AND FAMILIES TO BECOME INVOLVED IN THE TEACHING PROCESS!

If you are interested in speaking to a medical audience as part of the Metabolic Outreach Service, or if you will allow your/your child's record to be reviewed and used as a teaching case (anonymously) during an Outreach workshop, please let us know!

Please call:

Isabel Matos

Tufts Medical Center, Metabolism Service: 617-636-5443

Or visit our website -

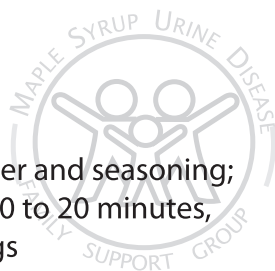
http://www.floatinghospital.org/OurServices/Genetics_Metabolism/theMetabolicOutreachService

Grilled Mushrooms

1/2 pound medium mushrooms
 1/4 cup melted butter
 1/2 teaspoon dill weed
 1/2 teaspoon garlic powder

Thread mushrooms on skewers. Combine butter and seasoning; brush on mushrooms. Grill over hot coals for 10 to 20 minutes, basting and turning every 5 minutes. 4 servings

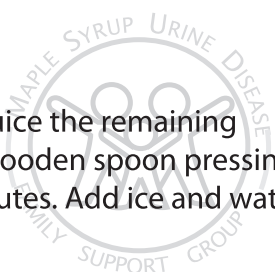
	Protein	Leucine	Calories
per recipe	5.2g	337mg	463
per serving	1.3g	84mg	116

**Old Fashioned Lemonade**

6 lemons
 1 1/2 cups sugar
 water and ice cubes

Wash lemons and thinly slice 2 lemons. Juice the remaining lemons and pour over sugar. Stir with a wooden spoon pressing slices to squeeze out juice. Let set 20 minutes. Add ice and water to make 1 gallon. Stir well. Serves 16

Per serving	Protein 0.09g	Leucine 1.6mg	Calories 77
-------------	---------------	---------------	-------------

**Strawberry Sorbet**

1 1/2 cups water
 1/2 cup powdered sugar
 2 – 10oz packages frozen strawberries
 Blend in blender until smooth. Pour into containers and freeze.
 4 servings

	Protein	Leucine	Calories
Per Recipe	2.9g	166mg	779
Per serving	0.7g	42mg	195

Variation: Substitute 2-10 oz packages peaches for strawberries

Per recipe	3.8g	211mg	775
	1.0g	53mg	194



Tempura veggies: Make a batter using Wel-Plan Baking Mix, cold water, and a little Coffee Rich or reconstituted CoffeeMate non-dairy creamer to a pancake batter consistency. Then dip onion rings, carrot slices, mushrooms, and whatever vegetables you like, and deep-fry them. The batter fries up light, crisp, and just a little bit sweet. Everyone can use chopsticks to dip the veggies into a sauce made from 1/2 teaspoon honey, 1 tablespoon soy sauce, 1 1/2 teaspoons hot pepper vinegar, 1 tablespoon water, and 1 teaspoon sesame oil.

More Easy Meal Ideas

Low protein applesauce or banana muffins are a great addition to many meals. Prebake them and freeze for convenience, then when ready, just thaw and warm in microwave oven. Put on lots of butter and serve warm.

Roasted vegetables: Roast onions, carrots, tomatoes, bell peppers, parsnips, turnips, potatoes, asparagus, or Brussels sprouts: Cut into slices or chunks and coat generously with olive oil. Sprinkle with salt and pepper and add any desired dried herb, then roast in a baking pan or on a baking sheet at 425-450°F. until browned and tender (15 to 30 minutes, depending on the vegetable). The vegetables will be delicious, with a sweet/smoky flavor.

Make a meal in a package: Place a bunch of veggies in aluminum foil (such as 1/4 potato, sliced, a little sliced carrot, some green beans, and sliced onions). Top with butter and seasoned salt, wrap, and bake for about 30 minutes at 350°F.

We Take "Diet For Life"
 to Heart. We Think It's

Essential!

INTRODUCING
COMPLEX
Essential
 MSD
 Vanilla Flavored Drink Mix for
 Maple Syrup Urine Disease



The First and Only
**MSUD Formula with
 Flax & Fiber**



For a **free** sample or more information, contact your dietitian or call

1-800-605-0410 TODAY!

You can also visit us on the web at www.medicalfood.com

Medical Food
 A Division of
 Nestlé Nutrition

© 2008 Nestlé
 All rights reserved.

© 2008 Nestlé
 All rights reserved.

© 2008 Nestlé
 All rights reserved.

More Options for More People™

Organizational Contacts

Contact for general information:
Kay Larsen, Easton, Pennsylvania
 610-252-8572 • kaysfo@aol.com

Contact for organizational information
 & printed material:
Sandy Bulcher, *Director* • Powell, Ohio
 740-548-4475 • dbulcher@aol.com

Contact to be added to the mailing list or
 make a donation:
Dave Bulcher, *Treasurer* • Powell, Ohio
 740-548-4475 • djbulcher@aol.com

Contact for legislative & educational issues
Marcia Hubbard, *Corresponding Sec.*
 Cedar Hill, Missouri
 636-274-2021
 wingman90gl1500@juno.com

Contact for inquiries in Spanish
Monica Falconer, *Spanish Contact*
 Anchorage, Alaska
 907-770-6065 (Alaska time zone)
 monicazf@yahoo.com

Newsletter Editor
Karen R. Dolins, EdD, RD
 33 Valley Road, Scarsdale, New York 10583
 914-723-5458 • krdhed@aol.com

Medical Advisors

Holmes Morton, M.D.
 Clinic for Special Children, Inc.
 535 Bunker Hill Road
 P.O. Box 128, Strasburg, PA 17579
 717-687-9407 / Fax: 717-687-9237
 Clinic Website:
 www.clinicforspecialchildren.org

Vivian E. Shih, MD
 Massachusetts General Hospital
 Amino Acid Disorder Lab
 149 Thirteenth St., Boston, MA 02129
 617-726-3884 / Fax: 617-726-5739

Susan C. Winter, MD
 Dir., Med. Genetics/Metabolism
 Valley Children's Hospital
 9300 Valley Children's Place
 Madera, CA 93638
 559-353-6400 / Fax: 559-353-7213
 winter2571@aol.com

Neil R. M. Buist, MD
 Professor Emeritus
 Pediatrics and Medical Genetics
 Oregon Health & Science University
 buistnrm@aol.com



Nutritional Advisors

Rani Singh, Ph.D., R.D.
 Emory University
 Div. of Medical Genetics
 2040 Ridgewood Dr.
 Atlanta, GA 30322
 404-727-2866 / Fax: 404-727-5783
 rsingh@genetics.emory.edu

Sandy van Calcar, M.S., RD
 University of Wisconsin
 Biochemical Genetics Clinic
 1500 Highland Ave.
 Madison, WI 53705
 608-263-5981
 vancalcar@waisman.wisc.edu

MSUD Family
 Support Group Board
 Wayne Brubacher, *President*
 (Indiana)
 Ivan Martin, *Vice President*
 (Pennsylvania)
 Dave Bulcher, *Treasurer*
 (Ohio)
 Anne Fredericks, *Secretary*
 (Pennsylvania)
 Denise Pinsky
 (Michigan)
 Tony Kohl
 (Arizona)

Nutricia offers a variety of low protein foods and snacks for protein-restricted diets...

...available to purchase at MySpecialDiet.com!

- An interactive, online resource for the metabolic community.
- Offers information, news, recipes and a community forum.
- Features an online metabolic shop with an extensive line of low protein foods, integral to a low protein diet.

US: (800) 365-7354
 Canada: (877) 636-2283
 www.MySpecialDiet.com

This Newsletter does not attempt to provide medical advice for individuals. Consult your specialist before making any changes in treatment.



This publication is supported by a grant from Nutricia North America