

MSUD NEWSLETTER

Published by the Maple Syrup Urine Disease Family Support Group • Volume 26, Number 2 • Fall 2009



MSUD Symposium 2010

June 24 - 26

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A Few Words From the Editor

Karen Dolins, EdD, RD, CDN
Newsletter Editor

This edition of our newsletter has a number of heartening pieces. In our cover piece, Dr. Zinnanti describes his work with Dr. Skvorak and their progress using norleucine to help prevent the brain damage that can occur when leucine levels are high. Inside, Dr. Skvorak further describes this work and also her successes transplanting healthy liver cells into MSUD mice. In years to come this may provide an alternative to whole organ liver transplant. We hope that their work continues to go well and that they are able to conduct human studies soon. In another treatment advance, Coram, Inc. describes their BCAA-free TPN and its uses in critical care. I am hopeful that one day in the not-to-distant future we will have protection against the decompensation that occurs when those with MSUD become ill.

We also hear from several of our older members about their experiences with college life and their advice
(Editor's message cont. on page 3)

Norleucine to prevent brain injury in MSUD

By Dr. William Zinnanti
SUNY Downstate Children's Hospital, Department of Pediatrics



Dietary control of branched-chain amino acid (BCAA) intake is critical for protection of the brain in maple syrup urine disease (MSUD). Usually this is accomplished with life-long adherence to special diets. However, simple colds and flu-like illnesses can stimulate large accumulations of BCAA that can be difficult to manage. Recent work with MSUD mice has provided clues about future alternatives to enhance brain protection during illness and potentially loosen restrictions on dietary adherence.

Classic MSUD mice do not survive for more than three days after being born. We are very fortunate that Drs. Greg Homanics, Kristin Skvorak and Harry Paul, of the University of Pittsburgh, developed an intermediate

(Norleucine cont. on page 2)

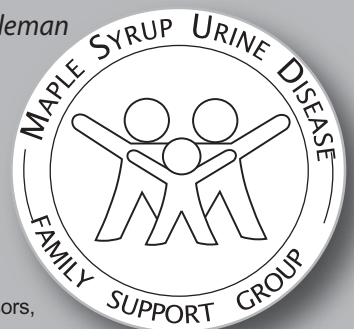
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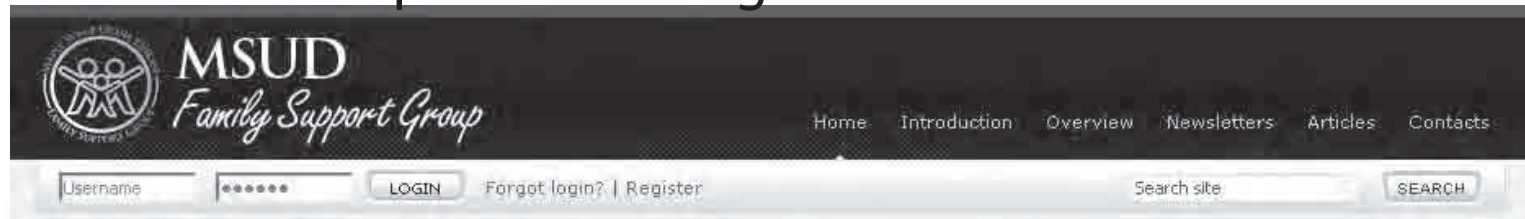
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The information contained herein does not necessarily represent the opinions of the MSUD Board, Medical or Nutritional Advisors, or all of our members. Before applying any of the information contained in this newsletter, you must consult a MSUD specialist.



New Website Up and Running... *check it out!*



The MSUD Family Support's website at www.msud-support.org has recently been re-designed to better serve our visitors. A number of enhancements have been made. The most exciting of all is the ability to register online to get member's only features. These include the ability to post new articles, post your physician (under Physician Network), post new links (under Related Links), post your events (under Events & Resources) and profile updates which include a member gallery. If privacy is of concern, there is also a way to limit what's visible on your profile. Also, global search has been added to scour through our entire archive of articles.

Currently, the contents of the website duplicate the contents previously found on the old website. In the near future, additional sections will be added as needed. You can still visit the old website by going to <http://www.msudsupport.org/index.html> or clicking the link on the new homepage.

Please visit the website regularly to see what has changed.

We hope to see many more parents, kids, professionals and companies register.

Note on registration: The donation is an optional element if you've already made a contribution of \$10 to receive our newsletter.

Eddy Wang

(Norleucine cont. from page 1)

MSUD mouse model as well a classic MSUD mouse and have allowed us to use them in our experiments. The intermediate MSUD mouse was developed with a small amount of enzyme activity allowing some to survive to adulthood. This allowed us to introduce a low BCAA diet for these mice that allows them to live a normal life span. Using both models we have been able to watch events that cause brain injury quickly in classic MSUD mice and more slowly in the intermediate mouse model. Giving intermediate MSUD mice a high protein diet results in the development of brain injury within a few days, similar to the human disease.

We found that brain injury occurs in both models when the amino acid leucine accumulates in the brain rapidly and is converted to an acid, known as alpha-keto-isokaproate (αKIC). This finding gave us an idea about how we might be able to stop the brain injury from occurring. Leucine travels into the brain via a specific transporter known as the large neutral amino acid transporter. An atypical amino acid, norleucine, can also occupy this transporter because it has the same structure as leucine only slightly longer. This amino acid is "atypical" because it is not normally found in our bodies, it does not have a natural degradation pathway, and it is not used to make proteins. Therefore, a low quantity of norleucine may compete with leucine for access to the brain. If the amount of leucine going into the brain can be controlled, then perhaps the effect of too much leucine in the brain can be controlled as well.

In MSUD mice, norleucine works to prevent or delay brain injury. Intermediate MSUD mice placed on a high protein diet became encephalopathic and died within 2-3 days. When 5% norleucine was added to the high protein diet, most of the mice survived 5-8 days with a delay in onset of encephalopa-

thy of at least 2 days. Lower amounts of leucine and αKIC were found in the brains of these mice. Surprisingly, only 1% norleucine given to the mothers of classic MSUD mouse pups provided a substantial increase in survival for these mice. Norleucine limited the amount of leucine found in the mother's breast milk, resulting in a low-BCAA meal for the dependent pups. Additionally, norleucine transferred to the classic MSUD pups reduced brain leucine levels as well.

Although these initial results are exciting and promising, norleucine has not been tested or approved for human use. An immediate goal is to begin clinical trials of norleucine, so it can eventually be provided as an additional tool for brain protection in human MSUD.

How might norleucine be used in human MSUD? The first goal for use of norleucine would be for children in acute encephalopathy or to prevent encephalopathy during flu-like illness when amino acids are known to accumulate. If norleucine works similarly in humans as in mice, we hope that administration of norleucine can delay encephalopathy as efforts are undertaken to control blood levels of BCAA. Additional goals for norleucine include a loosening of dietary restrictions as norleucine may be given as a supplement to prevent brain accumulation of BCAA. Norleucine may also potentially be used by mothers of MSUD infants to allow breastfeeding. Breastfeeding provides infants with their initial immunity from the mother in addition to nutrition. Unfortunately, breastfeeding is currently contraindicated in MSUD because of the inability to control BCAA content of the breast milk.

I am grateful for the collaboration with Dr. Homanics group and the MSUD family support group. Without the awareness and support this work would not be possible. Thank you. ■

The 2010 MSUD Symposium *Lincolnshire, Illinois*

June 24 - 26* (outside Chicago)

We've been planning for a year, and now it's your turn to plan! There will be exciting new features, and we hope to see everyone in the MSUD community attend.

Chicago is a great place for a family vacation, so save the dates and make a whole trip out of the MSUD Symposium. We have an amazing rate of \$104 a night at the beautiful Marriott Lincolnshire Resort.

**Please note: the previous newsletter listed the incorrect dates*

Hotel Information:

Marriott Lincolnshire, 10 Marriott Drive, Lincolnshire, IL 60069

Room rate \$104 (Available 3 days before and 3 days after Symposium)

Reservations can be made on line at:

<http://www.marriott.com/hotels/travel/chiln?groupCode=msumsua&app=resvlink&fromDate=6/24/10&toDate=6/27/10>

The MSUD code is already listed in the group code so all you have to do is book your dates.

Agenda Highlights

**Motivational Speaker • Fundraising • Research Update • MSUD Management • Nutritional Update
Liver Transplant Update • Cooking Demonstration**

Transportation

Chicago O'Hare (ORD)	18 Miles
Chicago/Midway (MDW)	45 miles
Greyhound (Downtown Chicago)	18 Miles

Hope to see you there.

(Editor's message cont. from cover)

to others who are contemplating taking this step. How amazing is it that our children have this opportunity, and are able to manage it safely! This information is particularly reassuring for me, as Hannah is a high school sophomore this year. She is now preparing her own formula and keeping track of her leucine intake. She has had the benefit of a second summer at the Emory Metabolic Camp, which has also helped her achieve greater independence, and is very much looking forward to a college career!

Two heartwarming accounts of living with MSUD under less-than-optimal circumstances are related by the Blaus and Joyce Brubacher. The Blau's daughter Cindy is the oldest person living with MSUD, and her story shows us how far we've come and how we all benefit from the experiences of those who came before us. The Brubacher's trip to Central America also shows us here in the US how others manage under adversity, and remind us of how fortunate we are.

As for our family, we stayed home this summer. After her week at Emory, Hannah has spent the summer in a theater arts camp and loves it. Weather permitting, she rides her bike there and back with her formula and food for the day packed in her backpack. In between those two experiences, we were visited by the Bulcher family. We all went out for a wonderful pasta dinner complete with Italian ices in New York City's Little Italy. A great time was had by all.

As we say goodbye to summer, be sure to make note of the 2010 MSUD Symposium to be held in the Chicago area next June. Also, please be sure to take a look at our new website at **www.msud-support.org** which Eddy Wang has been working so hard on. It has a link to my email address, so please drop me a line and let me know how you like the newsletter and whether you have any ideas for future stories.

Warmly,
Karen Dolins

MSUD NUTRITION WORKSHOP AND FAMILY DINNER



Sponsored by



Sandy Simons, MA, RD, LDN, CHES
Director of Events & Marketing
Applied Nutrition Corp.

On July 29, 2009, MSUD families and patients of the Clinic for Special Children came from all over central Pennsylvania for a day of nutrition education and cooking workshops led by parents and Applied Nutrition staff.

The program began with Brenda Wenger, mom of Karen and Rachel, providing a step-by-step demonstration of low protein bread recipes. Brenda's creativity in the kitchen resulted in a variety of items including loaf bread for sandwiches, low protein breadsticks, cinnamon sticks, donuts and dinner rolls. She explained some tricks of the trade, like adding extra wheat starch on a humid summer day to combat the moisture in the air.

Applied Nutrition's Bridget Wardley, MS, RD, Director of Nutrition and Erica Novack, demonstrated how Maddy's Homestyle low protein baked goods can be both nutritious and delicious. They made Double Banana Muffins, Thumbprint Cookies and Fruit Pizza. Their presentation included many interesting points about health and nutrition. As an example, using canola oil in the recipe for Maddy's low protein muffins can boost intake of omega-3 fatty acids by up to 1/3 of daily needs.



This event would not have been possible without the help of Glenda Groff, mom of Jordan. She taught the group how to make low protein egg rolls and onion rice casserole. Lorraine Martin, mother of Crystal, shared her expertise on how to make chocolate sandwich cookies. All the dishes were served as part of the main dinner and were a huge hit.

While Glenda and Lorraine shared their recipes with the adults, Sandy Simons, MA, RD, CHES, along with Maureen Finkel and Dana Corby from Applied Nutrition led a group of creative young chefs in a culinary challenge to create new low protein recipes using only the food supplied specifically for the event. There were 3 judges, Dr. Rider, Glenda Groff and Bridget Wardley, who tasted and judged each dish based on appearance, taste, originality and of course, leucine content!

The evening portion of the event featured talks by the three doctors from The Clinic for Special Children: Dr. Morton, Dr. Rider, and Dr. Strauss. Dr. Morton spoke first, noting that the MSUD population is getting older and that the clinic now sees more healthy teens and adults with MSUD than ever before. To address this increasing population, Dr. Rider joined the Clinic for Special Children's team a year ago. He will make sure the clinic continues to be the 'medical home' for teens and adults with MSUD.

Dr. Strauss discussed how the need for an improved MSUD infant formula led to the collaboration with Applied Nutrition to create a more nutritious MSUD formula. He took this opportunity to share his research on the new infant formula, still in the developmental stage, and the improved nutrition and health status of the infants in the study. His most dramatic slide showed a significant decrease in hospital admissions and stays for those using the new product. Although the infant product is not yet commercially available, the research used to design the infant formula was used to create the Complex MSD Essential formula that was launched last year.

Over 120 people attended the event and had a fun time sharing ideas, recipes and learning how to be more creative in low protein cooking. Thank you to everyone who attended and contributed to help make this event a success! ■

Try some of the recipes from the workshop. See DIETwise, pages 9 & 10.



By Jordan Bulcher

In June 2009, I completed my first year of college at Ohio State University as a molecular genetics major. I lived on campus in a four-person freshman dorm room, about a half-hour drive from my current residence in Powell, Ohio.

I managed my diet with minimal parental support and monthly contact with my doctor and dietitian. That being said, I did not mix my formula myself, as there was not sufficient space in my small room. My parents came every three days to bring my drink and check my urine DNPH (a compound with a similar purpose as keto sticks).

I used OSU's meal plan and the many restaurants that line nearby High Street to allow me to have plenty of food options making the issue of finding something to eat relatively non-existent. The food issue was also made easier because of my leucine tolerance of 1100 mgs per day. For those days that I ate food higher in protein, I kept a stockpile of candy and Gatorade to curb my appetite in the evening without increasing my protein intake. Through all this, I experienced very few setbacks during autumn and spring quarter (a few minor leucine elevations here and there, but nothing major). However, during the winter quarter, my leucine became elevated numerous times requiring one trip to the ER and multiple trips home to consume more medical formula and low protein food under the watchful eyes of my parents. We didn't determine the cause of the elevations, but my parents believe that stress was to blame. I am not convinced of that, however.

Next year, I will be living off-campus in a larger space and will purchase the meal plan that allows me to eat lunch on campus five times per week. In addition, I will be taking care of myself, including making my own formula, cooking some meals, checking my urine DNPH, and getting monthly blood draws. More than likely, my parents will be contacting me periodically to check up on me.

Overall, I found my first collegiate adventure to be painless in regards to MSUD. The amount of schoolwork I needed to do, however, was a different story. ■

Living at College with MSUD: Advice From One Who's Been There

By Nikolai Rudd

I started looking at colleges at age 9. Two years earlier, my parents made my brother and I sign a contract that stipulated that we would attend four years of college. My parents didn't want me to live in fear or feel restricted by my diet. They wanted me to feel empowered. I was put in charge of my diet at age 7. I was responsible for making my drink and finishing it. I had always been very conscientious about eating the correct foods. I rarely ate foods or amounts of foods that I wasn't allowed. My parents made sure that I was aware of the consequences that could have on my body, my mind, and my life. I had to own up to my actions—and take responsibility for my choices.

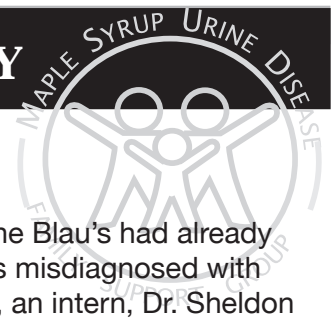
I was used to managing a heavy schedule from high school. Since I did both sports and acting, I was at school from 7:15 am until 6:30 pm. I had a large volume of formula to drink, so I brought my drink with me. I kept it in the nurse's refrigerator and went in there periodically to fill up a bottle that I could carry around with me easily.

When I first started high school I got into trouble because I burned through my calorie intake playing both soccer and tennis. My leucine level rose to 1390. My diet had to be restructured to allow more protein from natural foods. But because of the mess up, not only did I almost die, but I missed my last growth spurt. I was catabolizing (breaking down) protein, which is just as bad, if not worse, than cheating. Your body burns up all the calories, and in order to keep functioning, it gets energy by burning up your stored protein.

My experiences in high school prepared me well for college. College is a lot of work and provides a lot of academic pressure, along with temptations from peers, parties, etc. both in the way of food and in the way of alcohol. Managing the stress of school along with managing your diet will take some getting used to, even if you are extremely responsible about it.

For me, going to a smaller college made a lot of sense since the class sizes are smaller, with more one-on-one attention. This makes it easier to get help as your teachers notice when you are there and not, and when you might be sick. Not only that, but they are more apt to be able to accommodate you if you find you are behind. To avoid getting lost in the crowd—and to develop a personal connection with your teacher, I suggest sitting close to the front of the class. Participate, and often speak with your teacher afterwards.

(College Advice cont. on page 13)



Cindy Blau

Classical MSUD – 50 years old

By Marilyn Blau as told to Karen Dolins

Cindy was born on June 16, 1959 to Marilyn and Seymour Blau of Petaluma, California. The Blau's had already experienced MSUD with their son Gary who was born in 1955. When he became ill he was misdiagnosed with everything from a pituitary disease to childhood retardation. During his last hospitalization, an intern, Dr. Sheldon Miller, noticed a maple syrup odor on him. Remembering a medical school lecture, he contacted New York Hospital (NYU) and it was determined that Gary had MSUD. Sadly, nothing could be done for him at that time and he died at 21 months of age. An autopsy confirmed MSUD as the cause.

When Cindy was born, her doctor sent a blood sample to NYU. It tested positive for MSUD, and Cindy was flown to New York where researchers were making progress in understanding the disease. She lived at Bellevue Hospital, associated with NYU, for 4 1/2 years under a federal grant. Her doctors, Emmet Holt, Selma Snyderman and Joseph Dancis, developed the diet which kept her alive.

At that time, little was known about managing infections and every illness was life-threatening. The Blau's recall being called time after time to be told that they weren't sure Cindy would make it. But she did. The family moved from California to New York to be with her, living first in Brooklyn and then in Long Island. Still, they were told that she wouldn't live past 10 years of age.

Finally they were able to take her home, but the problems of course persisted. The formula did not have flavoring and tasted terrible. Cindy would vomit at will when forced to eat the foul-tasting formula, and become dehydrated. She wouldn't eat regular food, and frequently needed to return to the hospital for further care. Gradually Cindy's condition stabilized. She was able to go to school, and attended special education classes. When she was 13, the family moved to Columbus, Ohio, where she attended a special needs girl's school. She graduated from high school.

Cindy now lives independently, with help from Creative Living, a non-profit organization which provides housing and assistance for physically disabled adults. Nurses come twice a day to monitor her diet and her health, and take her to doctor's appointments. She has a job which she obtained through the Association for Retarded Citizens. She is covered for her medical care and formula through Medicaid and Social Security. She has a long time boyfriend.

"I'm just glad that things turned out so well for Cindy" Marilyn says. She remembers the early years, trying to care for Cindy along with her brother and sister at a time when there was little information about the disease and certainly no support group.

Cindy's diet remains limited. For a long time, she lived on French fries and applesauce. She loves her low protein foods, though, and her weight has significantly outgrown her short stature. Now she struggles with issues familiar to many her age. Fifty years ago the Blau's couldn't imagine that they would see their daughter live to this age and enjoy a life with companionship, independence, and productive work. "I couldn't ask for anything more," says Marilyn.

Dear Sandy,
6-20-09

When I was born, my mother and father were told that I had Maple Syrup Urine Disease and I spent most of my childhood in New York Bellevue Hospital. When I grew up, I was on a special diet. Right now I have a very good job. I work at night Monday through Thursday 5:00PM – 10:45PM. I enjoy my night job. I have Fridays and the weekend off and I also have a very good boyfriend. I am going to close for now.

Your friend,
Cindy Blau



Cindy (front, left) & family

WHERE PERSEVERANCE GOT ME

By: Susan Lynne Needleman

Classic MSUD

After I was born, on July 24, 1990, in Malden, Massachusetts, I had to spend my first week in the hospital for swallowing muconium during birth. After antibiotics I was sent home and everyone thought that there were no other problems. I was a happy baby. Shortly afterwards I stopped drinking my bottle, in fact I would take my hand and literally hit it away. When my mom contacted the hospital in the middle of the night they just said she was feeding me wrong. In the morning she called my pediatrician, who was about to call her to come in. At the time of the visit he informed my parents that one of my newborn screening blood tests had come back positive for a rare metabolic condition, Maple Syrup Urine Disease. My parents then had to take me to Massachusetts General Hospital (MGH) right away where I underwent dialysis. During this time my parents quickly were informed about the dietary restrictions that I would have to live by and my mom had to then find a pharmacy that knew what MSUD was and that would order formula for me. After this surgery I spent the next month in the hospital, before finally going home. Since that time I have been hospitalized three times. The first one was when I was two and a half, which was the first time I was sick. At that time I was in the hospital for approximately a month. I was then hospitalized for three weeks when I was five and a half, for a sore throat that prevented me from drinking. The next time that I would have to face an IV and everything else that comes with being hospitalized was when I was eight and a half, for a week, but it still took my body months to fully recover. I think I have been able to keep my hospitalizations to a minimum by taking a lot of precautions in school. The schools I have been in always sent letters home to the school families about me and how I needed to be careful about not getting sick. The parents would usually let the school know. The school would then tell my mom if my classmates were sick and the kids would usually let me know if they didn't feel good. I would then sit on another side of the room or the school would make other arrangements for me.



(Perseverance cont. on page 8)

Treating MSUD: The Early Years

By Karen Dolins

I was inspired by Cindy Blau's story to look in the scientific literature for publications related to her case. Sure enough, "Maple Syrup Urine Disease with Particular Reference to Dietotherapy" was published in 1964 by Selma Snyderman, MD, Patricia M. Norton, Ellen Roitman, and L. Emmett Holt Jr. in the journal *Pediatrics*. This article details the journey taken by Cindy and her medical team as they tried new therapies to keep her alive.

As Cindy's older brother had died of MSUD, when she became difficult to feed and started to exhibit the classic maple syrup odor at the end of her first week of life, she was transferred to Bellevue Hospital in New York City where her amino acids were analyzed and levels of the branched-chain amino acids (BCAAs)

were found to be elevated. By her 17th day of life, she was started on a special formula which omitted the BCAA along with methionine, which at that point was thought to also be involved in the metabolic abnormality.

At that time, formulas such as we have today did not exist. Cindy was given a synthetic mixture of amino acids along with vitamins, minerals, sugar, and corn oil. Within 3 days of starting this diet, Cindy began to improve.

With time, the team noticed that even slight elevations in temperature would cause Cindy's BCAA levels to rise and symptoms to recur. This was treated by removing the BCAAs along with methionine from the diet and adding fluids.

At about 6 months of age, Cindy stopped growing. The medical

(The Early Years cont. on page 8)

(Perseverance cont. from page 7)

As I grew up, I have learned to accept MSUD as just a way of life, even though it has not always been easy for me. For me, MSUD also led to some neurological impairments, including learning disabilities and a speech disorder. When I was three and a half I was diagnosed with a severe articulation disorder and started Speech Therapy. I remember as a toddler I used to use hand movements like charades in order to even communicate with my parents. Now after fifteen years of speech therapy I am now hopefully understood by all, as long as I slow down and articulate each word. I have even spoken on live TV through my local cable station, where I have volunteered for five years and am now interning.

When I was in third grade I had testing done and it was found that I had learning disabilities and started a phonetic reading program. Since then I have had extensive lessons going back and forth between the Orton-Gillingham and the Wilson Reading System. It was at this point that I actually started to learn to read and spell. Later on it was found that I had a form of dyslexia, an executive functioning disorder, which is when it is hard for someone to organize and keep track of things. In addition, I had fine and gross motor skills problems, which I was able to improve with the help of eight years of occupational therapy.

When I entered high school, I could not read the majority of the materials on my own. For those first few years my mom would read me my homework. In school I was not able to complete much work as teachers would just say, "try" and "try harder". Then I started to read by means of books on CD and a computer software program that reads to listeners whatever they scan into it. I have always been a hard worker and it has paid off. I have been able to earn high grades in school and was an advocate in bringing the National Honor Society to my high school, which I am now a member of.

Throughout high school I was in many clubs: Math Club, Science Club, Moot Court, Movie Production Club, Student Outreach Group, Class Rep. and more. I also created my class's senior DVD yearbook on my own and helped out with the computers in my high school. On June 10, 2009 I graduated high school two days after completing the reading program that I had been learning from for ten years. In the fall I will be a commuter student, at Wentworth Institute of Technology in Boston, Massachusetts, where I will be majoring in Computer Networking and Information Systems (BCNS). ■

(The Early Years cont. from page 7)

team felt that most likely there was an unknown nutritional factor that was missing from her synthetic diet, and decided to try her on a regular milk formula. Within 24 hours she became lethargic and lost muscle control. She did have a spurt of weight gain, but it was clear that she could not be managed on a regular formula, and her artificial formula was resumed.

When Cindy was 23 months of age, her doctors decided to give her a normal dose of the amino acid methionine along with a supplement of Brewer's yeast. This was effective at stimulating growth. Using a new method of amino acid analysis, it was determined that what was previously thought to be an elevation of methionine was actually alloisoleucine, a previously unknown amino acid made from isoleucine.

By manipulating the diet, Cindy's doctors were able to determine that isoleucine is responsible for the maple syrup smell while leucine is primarily responsible for neurological damage.

Cindy remained in Bellevue Hospital for four years, at which point she was finally discharged to her parent's care. The doctors noted that due to the unusual nature of her early years, which were spent in the hospital environment, she was not exposed to the language and experiences of other children. At the time this paper was written, she had recently been returned home and was attending a special needs school. At 5 years of age, Cindy was "sociable, articulate, energetic, and at times mischievous," and they held great hope for her future development.

MSUD treatment has come a long way since the 1960's, and we all owe a great deal to Cindy and her parents for everything they endured as they and their medical team learned together about how to treat this disease. ■



Recipes from the Nutritional Workshop (See page 4)

Chocolate Sandwich Cookies

Ingredients:

Cookie dough:

- 2/3 cup (11Tbs) margarine
- 1 cup sugar
- 1 Tablespoon water
- 1 teaspoon vanilla extract
- 2 Tablespoons unsweetened cocoa powder
- 230 grams (approx. 2 cups) wheat starch
- 1 teaspoon baking powder
- 1/2 teaspoon baking soda
- 1/4 teaspoon salt
- 1 teaspoon xanthan gum
- 1/2 teaspoon methylcellulose

Method of Preparation:

Cookie dough:

1. Preheat oven to 350°F.
2. In large bowl, combine all cookie dough ingredients.
3. Mix well.
4. Roll into 3/4 inch balls.
5. Place on cookie sheet, at least two inches apart.
6. Bake for 8 minutes. Cookies will puff up then flatten.
7. Remove from cookie sheet and allow to cool completely.

To assemble cookies:

- Arrange half the cookies on cookie sheet.
- Place 1 Tablespoon filling on each cookie.
- Top with second cookie. Keep in airtight container and store in refrigerator or freezer.
- Makes 24 sandwich cookies

NUTRITION/Per cookie (1/24th recipe)

Calories 145 Protein, 0.4 g PHE, 11 mg
LEU, 17 mg

Please send recipes to
Food News Editor

Glenda Groff

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Maddy's Fruit Pizza

Ingredients:

- 1 can Maddy's Homestyle Sugar Cookie Mix
- 1 stick (8 Tbs) unsalted margarine
- 1/4 cup water
- 1 pudding cup (3 1/2 oz), Hunt's® Snack Pack® Lemon Pudding
- 1/2 cup Cool Whip® Original
- 1/2 cup fresh blueberries
- 1/2 cup fresh strawberries, sliced thin
- 1/2 cup Mandarin oranges, canned in light syrup, drained
- 1 medium kiwi, peeled, sliced and halved

Method of Preparation:

1. Preheat oven to 375°.
2. Line bottom of 9x2 inch baking dish with parchment paper.
3. Prepare Maddy's Homestyle Sugar Cookie Mix as directed on can. Initially dough will look dry, continue to mix until a smooth ball of dough forms.
4. Press dough into bottom of baking dish. Use spatula or spoon to flatten evenly.
5. Bake 20-25 minutes or until lightly browned on top.
6. Allow cookie crust to cool to room temperature.
7. In large bowl, fold Cool Whip® into Hunt's® Snack Pack® Lemon Pudding.
8. Spread pudding mixture evenly over cooled cookie crust.
9. Arrange fruit on top.
10. To serve, cut into 12 slices.

* Decorating tip: Use thin strawberries slices as first layer, then arrange kiwi halves and mandarin orange halves, and finally disperse blueberries throughout.

* Convenience tip: Cookie crust may be made ahead of time and frozen. Allow crust to come to room temperature before decorating. Pudding topping and sliced fruit can be made the day before and refrigerated.

NUTRITION/Per slice (1/12th pizza)

Calories 225 Protein, 0.9 g PHE, mg 10 LEU, mg 16.5

Maddy's Double Banana Muffins

Ingredients:

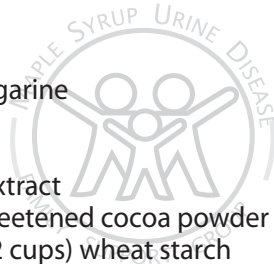
- 1 Full Can Maddy's Homestyle Banana Muffin Mix
- 1/3 cup canola or vegetable oil
- 1 cup water
- 1 medium banana

Method of Preparation:

1. Pre-heat oven to 375°F.
2. Lightly spray 12 muffin cups with non-stick cooking spray.
3. In small bowl, mash 1 medium banana.
4. Stir Maddy's Homestyle Banana Muffin Mix, water, oil and mashed banana in large bowl. Fill each muffin cup 2/3 full.
5. Bake 25-27 minutes or until toothpick inserted in center comes out clean.
6. Cool 10 minutes before removing from muffin pan.
7. Cool completely; store in airtight container.

NUTRITION/Per cookie (1/18th batter)

Calories 209 Protein, 0.4 g PHE, 12.6 mg LEU, 24.8 mg



Onion Rice Casserole

Ingredients:

- 2 Tablespoons canola oil
- 2 Tablespoons canned mushrooms, chopped
- 1/4 cup onions, chopped
- 1 cup low protein rice (dry)
- 6 cups water
- 1 Tablespoon Major Beef Seasoning



Method of Preparation:

1. In large pan, sauté onions and mushrooms in oil.
 2. In separate pot, bring water to a boil.
 3. Add rice to boiling water, lower heat and simmer for 5-8 minutes.
 4. Drain and rinse rice with warm water.
 5. Add rice and seasoning to cooked vegetables.
 6. Add 1-2 tablespoons water to rice if too sticky.
- Makes 2 servings

NUTRITION/Per serving (1/2 recipe)

Calories 449 Protein, 0.7 g PHE, 22 mg LEU, 33 mg

Cambrooke Foods – Summer 2009

Our new **Mini Pockets** are filled calzone-style snacks or meal items that are quick, heat-and-eat items featuring a golden brown seasoned crust filled with savory pizza ingredients or kid favorite peanut butter and jelly. They're great for busy families looking for homemade taste in a hurry. Bake and refrigerate to take along to work or school for a change-of-pace lunch.

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View the new three-minute Camino pro™ product line video on the Cambrooke web site. Learn in three minutes how the modern format of Camino pro™ metabolic formula products can make your MSUD diet more manageable – a tasty way to improve formula compliance. Request your free Camino pro™ sample or place your order today.

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Parenteral Nutrition for Patients with MSUD

Tanya Abel, Pharm.D.
Pharmacy Manager
Mendota Heights, MN Branch
Coram, Specialty Infusion Services



Children and adults with inborn errors of metabolism such as Maple Syrup Urine Disease, Phenylketonuria (PKU), Mehtylmalonic Aciduria (MMA), among others, have special nutritional needs. Diet manipulation is a reality in the lives of these children and adults, requiring special metabolic formulas, low protein recipes, reading product labels and purchasing specialized low protein food products. In writing this article I recognized some similarities with my life.

My son was diagnosed with celiac disease when he was 18 months. Watching him fail to thrive and waste away before the diagnosis was found was heart breaking. I can only imagine what many of you have gone through in the diagnosis of MSUD, as doctors ruled out this and that until they finally found the culprit. Nick started his new diet and the turn around was amazing, as I believe it is for many of you. We read labels, purchase special foods, order special gluten free products that are kid friendly and draw labs annually to follow compliance of his diet. He attends a celiac camp for kids so he can participate in camp without worrying about the food, we go to support groups and have picnics to share recipes. One major difference is that if Nick were to become critically ill and require intravenous nutrition, the products available in hospital pharmacies would give him what he needs.

For patients with inborn errors of metabolism, intravenous nutrition is more complicated. Intravenous nutrition provides nutrients to the body when the gut can't be used (unable to take the nutrition by mouth or tube). This can be life-saving for the MSUD patient. Total parenteral nutrition (TPN – dextrose, amino acids, lipids) is easily provided by hospital pharmacies when a normal formula is required. When patients with metabolic disorders such as Maple Syrup Urine Disease become critically sick and cannot take their nutrition by mouth or by tube, it becomes more of a crisis. There are no commercially prepared formulas that omit the amino acids not tolerated by patients with metabolic disorders. In those situations physicians try their best to manipulate the nutrition solutions, perhaps providing the lipid and dextrose portions of the solution without amino acids to meet some of the nutritional needs of the patient.

Pharmix™ is a special process which creates individual patient-specific amino acid profiles for intravenous use. The process of Pharmix™ compounding uses sterile commercial products and non-sterile powdered amino acids, dextrose and electrolytes. Components are mixed in a tank by trained, skilled technicians. The resulting solution is sterilized through filters to a volume that is required as ordered by the patient's physician. There are a number of quality assurance steps and tests that are in place during compounding to assure a sterile end product. This type of compounding process can provide certain advantages over traditional TPN compounding for specific patient populations. Patients requiring a specific amino acid profile not available commercially, such as the omission of the branched-chain amino acids isoleucine, leucine and valine for MSUD, can be managed using this individualized amino acid mixing procedure. Alternatively, if a patient has an increase need for specific, individual amino acids addition adjustments can also be made.

Compounding from non-sterile powders requires specific quality assurance monitoring, which is regulated by the United States Pharmacopeia document 797. These regulations assure that high risk compounding meets all standards to protect patients receiving products compounded from non-sterile powders.

Most MSUD patients in crisis can be managed short term with commercially available products and may not have any need for this special compounding. In some cases, though, this technology can be life-saving. There are many success stories, such as helping a child through an MSUD crisis or supporting a bone marrow transplant patient's nutrition until they are able to consume enough nutrition by mouth. Just knowing that this specialty amino acid solution is available is reassuring to families of critically ill patients.

This physician directed compounding is available from Coram, Specialty Infusion Services and you can direct questions to our Clinical Operations department, or access on line at www.coramhc.com. Coram staff work directly with the hospital pharmacy to help provide for the critically ill patient and continue to provide services in the home setting. Please feel free to contact me as well for questions at 1-800-542-6726.

I sincerely hope that none of you would ever have to call Coram, Specialty Infusion Services but if you do, know that we are continuing our efforts to maintain this orphan-like compounding in support of metabolically challenged patients. ■



Early Successes in Treating a Mouse Model of Intermediate MSUD - a summary of recent publications.

Kristen J. Skvorak, Ph.D.

University of Pittsburgh, Pediatrics Department
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Editor's Note: Dr. Kristen Skvorak recently received a doctoral degree from the University of Pittsburgh. Her research focused on treatment of MSUD. In this article she describes her work and the possible implications for those living with this disease.

As a University of Pittsburgh graduate student studying under Dr. Gregg Homanics, my main focus was to correct a mouse model of intermediate MSUD (iMSUD). I last reported my progress on achieving that goal in the MSUD Family Support Group newsletter in August of 2007. Now, only two short years later, I have a great deal of exciting results to share!

Hepatocyte (Liver Cell) Transplantation

As you know, transplanting a healthy liver into a patient can essentially cure MSUD. Therefore, it was reasonable to think that transplanting healthy cells into an MSUD liver may also improve disease symptoms. Cell transplant has many advantages over whole organ transplant: it is less invasive with fewer and less serious complications, less expensive, repeated treatments are possible, and cryopreservation of donor cells could allow them to be ready "on demand." For these studies, I collaborated closely with Dr. Stephen Strom of the University of Pittsburgh, a leading expert in the field of hepatocyte (i.e., liver cell) transplant (HTx). Prior to this, Dr. Strom had mediated the clinical application of HTx to patients of liver failure and liver-based metabolic disease, though not MSUD. This new direction proved very fruitful, and in August of 2008 I was awarded my doctorate for my research in the field of MSUD.

Our animal studies were conducted on mice bred to have intermediate MSUD. Since MSUD patients typically begin treatment at birth, we also treated iMSUD mice at this time. Two separate injections of 100,000 healthy liver cells were injected directly into the livers of newborn mice. The mice were then allowed to develop normally and without any other treatments. Although only ~3% of the MSUD liver was repopulated by healthy cells, there were many improvements in the health of these animals when compared to untreated iMSUD mice. Circulating BCAAs were reduced an astounding 75% and body weight was increased 30% at 20 days of age (weaning). At 37 days of age (early adult), liver BCKDH activity, the enzyme which is compromised in MSUD, was more than doubled from 6% (untreated) to 14% (HTx), and improvements were found in some neurotransmitters, neurotransmitter metabolites, and amino acids (most notably dopamine and alloisoleucine) in the brains of HTx-iMSUD

mice. Excitingly, these neurological improvements are the first demonstration that HTx can have therapeutic effects on extrahepatic organs outside of the liver. Finally, the survival of transplanted iMSUD animals was greatly improved; however, long term survival was not determined since the experiment was terminated at 37 days. At this time, experiments are planned to improve liver repopulation and determine long term survival.

Norleucine

One of the major causes of brain injury in MSUD is thought to be related to leucine accumulation. Dr. William Zinnanti, a collaborator at The Penn State University, hypothesized if movement of leucine into the brain was impeded, disease symptoms would improve. Norleucine is an atypical amino acid shown to compete with leucine at the blood-brain barrier for brain access. To accomplish this, norleucine, an atypical amino acid shown to compete with leucine at the blood-brain barrier for brain access, was supplemented in mouse chow. Mice with intermediate MSUD (iMSUD) were fed a high protein diet with or without norleucine. When norleucine was supplemented with a high protein diet, survival was improved and the onset of encephalopathy was delayed in iMSUD mice compared to those only receiving a high protein diet. Norleucine-supplemented feed given to nursing mothers also resulted in improved survival in classic MSUD (cMSUD) mouse pups. Based on these data, norleucine should be further tested as a potential treatment for MSUD children admitted during catabolic stress. Dr. Zinnanti hopes to start a clinical trial sometime next year. (*Editor: see Dr. Zinnanti's article on the cover*)

In addition to norleucine studies, Dr. Zinnanti characterized a great deal of neurological changes in the brains of

(Mouse Model cont. on page 13)

(College Advice cont. from page 5)

That way you won't be just a name to them.

Here are some suggestions for teens planning to live at college and their parents:

1. Make sure you have a cell phone or a pager to use for emergencies.
2. Have your doctor write down a protocol for doctors to follow in case of an emergency AND have a list of IMPORTANT NUMBERS to CALL. If there is an emergency, make sure that any doctor in any hospital knows that they MUST call your MSUD doctor to help treat him.
3. Start getting in the habit of conducting a DNPH test—so that you know how it looks when levels are under control and when they are off. I got to be so good, that I could tell how off my levels were by how cloudy the test was.
4. Implement splitting up and taking formula and any low protein snack whenever hungry, even if it isn't meal time. Make sure that key people in the school (high school and college) know about the illness. In college, the key people are the teachers, counselor, the nurse, roommate and most importantly, the Residential Assistant (R.A.). In high school, it's the teachers, nurse, and principal.
5. Give each of these people a copy of the protocol and make sure that they know what to do in case of an emergency. This is essential if you lose the ability to communicate to EMTs and doctors. In college, this happened to me at least once a year, with the exception of my senior year.
6. Be sure that your teachers understand that there will be days when you might just be fighting something, and might not be totally there. If you get sick, you must be responsible for letting the school or teacher know, and making up the work.

It is important that anyone planning to live independently know as much as he or she can about their health. Ask your doctor questions about what happens when—give examples and hypotheticals. Become as knowledgeable as possible so that you will be able to become your best advocate.

Personally, I love learning—and work my tail off to meet every challenge. I knew that if I could manage my diet at home along with school work and activities, then I would probably be able to do it on my own. And that was something I strived to do. I wanted to be an actor. To live in L.A. And I had a great support system.

I still think that that is the most important thing.

Sincerely,
Nikolai

(Mouse Model cont. from page 12)

both iMSUD and cMSUD mice. These new data have greatly contributed to our general understanding of the pathophysiology of MSUD. Dr. Zinnanti proposes two convergent mechanisms of brain injury resulting from BCKA accumulation: (1) neurotransmitter deficiencies and growth restriction, and (2) Krebs's cycle disruption and disruption of energy metabolism leading to energy deprivation. This new brain injury proposal could illuminate new solutions to prevent such injuries from occurring.

Dr. Zinnanti can be reached by email at wjz105@psu.edu.

Getting the Word Out

Scientific meetings are an excellent venue for a young scientist. It's a place to establish connections and collaborations, share and discuss new data, and discover new technologies from other like-minded researchers from around the world. I have had the incredible opportunity to present my HTx data at two international meetings in the past year. Two talks were given at the Society for the Study of Inborn Error of Metabolism (SSIEM) Symposium in Lisbon, Portugal, and one talk at the Cell Transplantation Society Symposium in Okayama, Japan. All presentations were very well received, and I have been contacted many times by investigators interested in applying cell therapies to their own research. Dr. Zinnanti has presented his norleucine data at the Grand Rounds at Kennedy Krieger Institute (Johns Hopkins University) and will present at the SSIEM Symposium in La Jolla, CA on August 29th, 2009. In addition, an in depth characterization paper on the iMSUD mouse was recently published which describes its remarkable similarity to the human disease. The iMSUD mouse is currently maintained at The Jackson Laboratories, an animal model "zoo" allowing open access to researchers around the world.

We hope the public availability of the iMSUD mouse, as well as the newly published characterization and therapeutic data, will serve to encourage others to forge ahead into the study of MSUD. A greater number of analytical minds studying a problem should yield a more complete understanding of the pathophysiology behind the human disease. In time, this will undoubtedly lead to new therapies to treat or possibly cure the disorder. ■

Publications:

1. KJ Skvorak, et al. Hepatocyte transplantation (HTx) corrects selected neurometabolic abnormalities in murine intermediate maple syrup urine disease (iMSUD). *BBA - Molecular Basis of Disease*. Submitted
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3. Skvorak KJ. Animal Models of Maple Syrup Urine Disease. *Journal of Inherited Metabolic Disease*. Apr 2009; 32(2):229-246
4. Zinnanti WJ, et al. Dual mechanism of brain injury and novel treatment strategy in maple syrup urine disease. *Brain*. Apr 2009; 132(Pt 4):903-18

OUR VISIT WITH MSUD FAMILIES IN CENTRAL AMERICA

By Joyce Brubacher

At the end of January Wayne and I, along with another couple, left for Central America. Our main objective was to visit 2 of my cousins and 3 MSUD families. The one cousin of mine and her husband in Guatemala are the grandparents of a child with MSUD. Their son Frank Wade Martin and his family flew to Pennsylvania as soon as their suspicions were confirmed that their newborn daughter had MSUD. They moved their family from Guatemala to Pennsylvania soon after to be close to Dr. Morton's clinic so they could continue treatment for their daughter. They know of no other families in Guatemala with MSUD. There is no screening or treatment for MSUD in that country.

We stopped in Honduras for a quick visit with the Luis Villafranca family. Luis kindly picked us up at the airport in the evening and spent time with us in spite of an extremely busy work schedule. The second night we met with him, his wife Ivonni, and their son Mario, 5, who has MSUD. Although somewhat disabled, Mario was a very pleasant, happy boy, and it was good to see him looking so well. It was a pleasure to visit with this family and see a little of Honduras.

We then visited the Kropf family in Costa Rica. In this household, there are three older, unmarried girls who care for special needs children given to them by the country's welfare department. Most of the 6 children they are caring for now are severe cases needing total care. One of these caregivers is Rebecca, who received Dwight, an Indian boy with MSUD, when he was 4 months old. He had an older sister Karla with MSUD who was in a foster home at the time. Rebecca did her best to care for Dwight with very little information on MSUD and called me frequently for advice. We advised her to attend a Symposium. The MSUD Family Support Group paid her expenses with the money allotted for that purpose from the United Services Foundation. She learned much at the Symposium in 2006 that helped her improve Dwight's care and also has been instrumental in improving the care of other children in Costa Rica. She plans to write more details for a future Newsletter.



Luis & Ivonni with son, Mario



Rebecca (right) & Karla at Dwight's grave

After Rebecca attended our Symposium, and learned about isoleucine and valine supplementation, she took some valine along to try. Dwight was already getting isoleucine but valine was not available in Costa Rica. Dwight made rapid progress after the supplementation. The doctors were impressed. It is still a problem to get valine, but when they can get some, the children all do so much better. Rebecca made some diet changes on her own, and Dwight developed into a very active, happy boy who was adored by the Kropf family and their friends. A school teacher who worked with him told me he was above average in some areas which was attested to by others. He delighted all those who knew him.

Last year Rebecca was asked to care for Dwight's sister, 7 year old Karla. She now had two children with MSUD and another severely handicapped child in her care. Dwight loved to play outside but was restricted to a certain area of the yard. In December, he was playing in the yard when Rebecca had to take Karla and another of their girls to the bathroom. It only took a few minutes, but long enough for energetic Dwight to get to a pond a little distance away and fall in. It was too late to revive him when they found the missing boy. No child can take his place in the family and especially for Rebecca who deeply mourns for her precious Dwight. He would have been 4 in January.

We are thankful we had the opportunity to visit this family and share in their grief. When Dwight died, Karla lost her appetite and lost weight. Currently she has gained a little weight but may have to have a G-tube inserted. She has had no hospitalizations since she is under Rebecca's care. Karla is a sweet, pleasant child. She is somewhat disabled, hard to understand, and not walking yet. We are sure she will make progress under Rebecca's good care.

Rebecca and her father stayed with us on our second to last night in Costa Rica in the capital, San Jose. We appreciated having

(Families in Central America cont. on page 15)

(Families in Central America cont. from page 14)

them along to translate since we stayed with Emilia Diaz and her daughter, Andrea, who speak Spanish. Emilia was so generous to invite all six of us to her house for meals and overnight. Andrea is 19 years old and has MSUD. She is a beautiful girl with a great smile. She has had extensive leg surgery and can only walk with leg braces which she does not like to wear. She gets around well on her knees and her mother does a good job of carrying her when necessary. She also uses a wheelchair outside the home. Andrea finished high school and lives with her mother and her mother's cousin, Gaby. Gaby is a wonderful caregiver for Andrea, since Emilia works full time to support them. In spite of the language barrier, we enjoyed sharing time with them and eating Emilia's beautifully presented and tasty meals. She is a wonderful cook.



Gaby (left), Emilia (right) and Andrea (middle)

In Costa Rica there are currently 22 children living with MSUD. Emilia scheduled a tour of Tamizaje, the newborn screening center, and a meeting with their metabolic doctor, Dr. Saborio. He is quite knowledgeable about MSUD and acquainted with doctors involved with MSUD in the states. The three hours we visited passed quickly, and we enjoyed every minute of it.

The country's screening program was impressive. Over 20 diseases are screened for throughout the country including MSUD. In remote areas where children are born in poverty,



Dr. Saborio (left), Joyce & Wayne. Notice photo on wall of Dr. Guthrie who developed the newborn screening test and visited this center at one time.

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there are people designated to get blood from all newborns. At this center in San Jose they have the latest technology in processing the blood dots and detailed record keeping. Their screening program could be a model for other national screening programs. However, most of the children have some damage in spite of early detection. Personally I would attribute the damage to a lack of MSUD dietary knowledge and problems getting formula at times. We hope this will change, especially now that they have had their first dietary meeting with all the families. Rebecca will report on this recent event.

Shayla stayed home when we traveled and did well in spite of a urinary infection. Her levels were good the whole time. Our trip was well worth the effort. The countries were beautiful and the weather comfortable. Our hosts all did a great job of sacrificing time and resources to make our stay a memorable one. May God bless them richly. We pray that all those with MSUD will have the opportunity to grow and develop as normally as possible in these countries.

Update: Mead Johnson was not able to send formula to Costa Rica in July and August. They have promised to resume shipments in September. Most of the families' supplies were depleted and the children were in a desperate situation. Rebecca was again instrumental in getting an emergency supply of formula donated by Applied Nutrition. However, it took weeks before this could all take place and many children suffered. ■

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This publication is supported by a grant from Nutricia North America