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CARNITINE THERAPY FOR MAPLE SYRUP URINE DISEASE

Details

Written by Susan C. Winter, MD

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Dr. Susan Winter spoke on the topic "Use of Carnitine in MSUD" at the MSUD Symposium 2002. Dr. Winter is the Medical Director for the Department of Medical Genetics/Metabolism at the Valley Children's Hospital in Madera, California. She is also one of the medical advisors for the MSUD Family Support Group. Some families have found a benefit in supplementing the MSUD diet of their child with carnitine. On request, Dr. Winter kindly submitted the following article for this issue of the Newsletter.

Carnitine is a natural substance important to the transport of fat into the mitochondria where it is "burnt" to form chemical energy. Carnitine is also important in removing the chemical "ashes" remaining after fat or protein is burnt in the mitochondria. It does this by binding to the chemical ashes and carrying them out of the mitochondria and then out of the body as carnitine bound "ashes" (acylcarnitine derivatives) dissolved in the urine. Carnitine is obtained from red meats and dairy products in the diet and from breast milk. It is also made in the body by breaking down muscle protein and dietary protein and converting them to carnitine.

As with all natural substances, deficiency can occur. Carnitine deficiency is nearly always secondary to other problems and may often be due to more than one factor. In infants and small children with small muscle masses, carnitine deficiency can develop easily due to a poor supply of protein in the muscles or a decrease in the dietary protein needed for synthesis. These small structured children are very dependent on a supply of carnitine from the diet. A chronic metabolic disorder, such as maple syrup urine disease (MSUD) which requires a special low protein diet, makes these infants and children even more vulnerable.

The majority of body carnitine is found in muscle with only 5 percent in the blood. Carnitine deficiency is best diagnosed with a muscle biopsy to test the levels in muscle tissue, but it is a very invasive test. If the blood level is low, the muscle level is always low. However, if the blood level is normal, it may simply reflect a recent dietary intake of carnitine and be falsely

reassuring. A rapid response with an increase in energy level, muscle strength and stamina with carnitine treatment, even with normal blood levels, suggests that the muscle needs for carnitine were not being met, and the person was probably muscle carnitine deficient.

Sick children with metabolic disorders often rely on total parenteral nutrition (TPN) that is not routinely supplemented with carnitine, and a deficiency can develop within days or weeks. Children and adults with metabolic disorders are very susceptible to infections such as gastroenteritis [an inflamation of the stomach and intestinal tract]. With this condition, malabsorption of carnitine adds to the problems of supply. Increased loss of carnitine from the blood or urine can occur with hemo- or peritoneal dialysis because carnitine is removed by the dialysis fluids. Carnitine deficiency occurs in patients with kidney disorders and affects the reabsorption of needed chemicals from the filtered urine (renal Fanconi syndrome). This syndrome can be a temporary problem resulting in urinary tract infections - frequently a problem in patients with metabolic disorders. In genetic metabolic disorders affecting fat oxidation and organic acid metabolism, carnitine deficiency occurs due to a massive excretion of carnitine in the urine bound to the un-burnt chemical "ashes." High levels of these un-burnt chemicals attached to carnitine can be detected in the blood and urine. This is the basis of the acylcarnitine derivative testing used in the MS/MS method of newborn screening.

In MSUD, the accumulating amino acids, isoleucine, leucine and valine do not form carnitine bound esters, and therefore their removal is not dependent on carnitine nor improved with carnitine supplementation. Even though the child with MSUD cannot use carnitine to detoxify the accumulating metabolites, additional carnitine is needed to generate energy from fat. Individuals with MSUD rely heavily on fat metabolism as the energy source during episodes of decompensation. The presence of ketones in the urine signals this emergency state. The inability to generate energy from fat due to carnitine deficiency can prove life threatening.

Carnitine deficiency is nearly always secondary to another disease process, so the symptoms are often those of the primary disease plus additional problems caused by the deficiency. Carnitine replacement therapy can resolve these problems. A deficiency of carnitine results in decreased energy available to muscle, causing muscle weakness and low muscle tone. Energy is required for the growth of muscle and for weight gain, so the child with carnitine deficiency often fails to thrive. A deficiency can affect cardiac muscle and result in poor cardiac contractions (cardiomyopathy). This leads to heart failure which may respond dramatically to carnitine therapy.

Energy is important for brain function and carnitine deficiency can cause convulsions, lethargy, irritability, and even coma. MSUD patients are very susceptible to infection which requires additional energy to resolve. In a genetic defect, which inhibits the ability to generate energy from protein sources, the problem is made worse by a carnitine deficiency. This results in a decrease of the energy generated from fat.

Carnitine is available as a medication approved by the Food and Drug Administration for treating secondary deficiency due to metabolic diseases. In the U.S.A., only one company, Sigma-tau Pharmaceuticals, Inc., supplies pharmaceutical grade oral L-carnitine (Carnitor). It is available with a prescription as a liquid with 100 milligrams of carnitine in each milliliter

(500 per teaspoon) and as a tablet with 330 milligrams of carnitine per tablet. Intravenous L-carnitine is also available in vials containing 1 gram in 5 milliliters of solution.

The amount of oral carnitine used depends on the reason for treatment and the clinical state of the patient. Higher doses are generally reserved for individuals with serious metabolic disorders during times of metabolic stress and decompensation. Doses range from 25 to 600 milligrams/ kg/day for oral carnitine and 25 to 300 milligrams/kg/day with IV carnitine. Oral carnitine is poorly absorbed with only 25% actually absorbed. The rest is excreted in the stool.

About 10% of patients taking oral carnitine experience side effects of diarrhea and/or stomach upset and in about 7% of individuals, a very fishy body odor develops. This odor is generated in the bowel by normal bowel bacteria converting carnitine to a very smelly, but non-toxic, trimethylamine. This body odor can be treated by taking a low dose of an antibotic, such as metronidazole, to destroy the offending bacteria that makes the trimethylamine. The gastrointestinal upset and diarrhea associated with oral carnitine administration usually improves if the dose is lowered or given with food.

Intravenous carnitine is fully available for body use because it bypasses the bowel absorption problems and for this reason is the preferred route of administration during a life threatening crisis. Intravenous carnitine will burn if infused too quickly and cause pain and irritation if it gets under the skin (interstitial).

In summary, the child with MSUD has decreased intake and synthesis of carnitine and an increased need for carnitine to use in fat oxidation and for the removal of the ketone bodies that are formed. Carnitine is essential for the generation of chemical energy and a deficiency will result in symptoms of decreased energy availability: muscle weakness, poor growth, decreased resistance to infections, cardiac dysfunction and neurologic problems. Treating fatty acid oxidation defects, organic acidurias and amino acid disorders, including MSUD, with L-carnitine has been shown to be safe and also life saving during the times of metabolic stress.

TIPS FOR PREPARING AND TRAVELING

IN A FOREIGN COUNTRY

Details

Written by Joyce Brubacher

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1. Find a hospital treating metabolic disorders close to where you will be staying in case of an emergency. If your child is an infant, look for a pediatrics department.
2. Call the hospital ahead of time to introduce yourselves and the patient (doctors appreciate this gesture). Keep the names and phone numbers of doctors handy during the trip
3. Carry a detailed "emergency letter" from your own Metabolic Clinic and a copy of the letter translated into the language spoken in the country of your destination.
4. Do not send MSUD formula by mail to your destination without first checking on mailing fees, the length of time it takes to get there, and the cost of custom taxes and inspections This can be very expensive.
5. Carry with you enough formula, mixed and ready to use, for the days you will be traveling. The formula and the supplements should be in a cooler with ice packs, and your name and address must be on the cooler. The cooler should be no bigger than the size allowed for a carry-on, otherwise it may be put in a difficult place to reach in the cabin of the plane.
6. Be prepared in case of delays at airports. Carry with you at least a few cans of formula and enough of everything needed to make additional formula. Include some low protein snack foods.
7. Ask your doctor to write a prescription for the MSUD formula and tape it on top of the cans in your carry-on bags.
8. Have your metabolic clinic write a "traveling letter." It should explain your child's MSUD condition and emphasize the importance of the formula for your child's diet. It should include a list of the supplements and other items needed for the care of your child, such as, blender, scales, DNPH, etc. Keep this letter with you during the trip.
9. Take some low protein products with you. This will depend on how much solid food your child eats, the length of your trip, duration of your trip, etc.
10. For a long stay, you might want to do some research on companies from which you can buy low protein products while there. Many of these companies have web sites you can check out before departing. You can keep these addresses, phone numbers or fax numbers with you. Loprofin products can be ordered in numerous European countries.
11. Find a list of leucine values for the foods eaten in the country you are traveling to, or make your own list. Adapt some of the countries recipes for the MSUD diet.

Enjoy your trip!

HAIL TO THE VICTORS

Details

Written by Sarah Foster

Published: 20 July 2009

Sarah Foster was a speaker at Symposium 2002 in Ann Arbor, Michigan. She is employed with Applied Nutrition Corporation, the company which makes the Complex MSUD formula products. She was also a vendor for the company at the Symposium. The following article is condensed from her touching narrative which received a standing ovation. Pictures of teen and adult MSUD "victors" taken at the Symposium are scattered throughout the article.

Friends: Sarah Foster and Shayla Brubacher with Shayla's favorite formula, Complex MSUD.

Although Sarah has PKU, not MSUD, many of the feelings and thoughts she expressed could well be those of a person with MSUD. This candid account of struggling with a metabolic disease, should give us parents some food for reflection. Are we aware of the innermost feelings of our children? May we all learn from Sarah's struggles.

Thank you for allowing me to speak with you today. I am very nervous because the only letters I have behind my name that even remotely qualify me to speak with you are PKU. Why is someone with PKU talking to you? After all, this is an MSUD Symposium, and I had never heard of MSUD until I attended a conference sponsored by the New England Connection for PKU and Allied Disorders in 1996. Even now, there are many things that I don't know about MSUD. I don't have the scientific knowledge of a medical professional, the expertise of a parent of someone with MSUD, or the intimate knowledge of what it is like to live with MSUD. I hope you will not dismiss what I say today just because I don't have MSUD.

One thing that made me more at ease about speaking to you was thinking back to when my parents had the opportunity to meet Erin who had homocystinuria, another metabolic disorder. When Erin met my parents, she said many things that I could relate to and wished I had the ability and courage to say to my parents. It didn't matter then that she didn't have PKU.

I may look familiar to some of you, because I had the honor of supervising your children at the last Symposium in Danvers, Massachusetts. At the end of the symposium, I knew I had to come to the next one here in Michigan. You and your children stole my heart. Luckily, the heart is one of the few things that grows even bigger when given away. When I vowed to come back, however, the last thing I expected was to be standing here talking to you. What I am telling you is based on my own thoughts, feelings, and experiences from growing up and living with PKU. I don't pretend to be unbiased, nor do I pretend to speak for anyone but myself. Everyone, regardless of their metabolic disorder, has different experiences and feelings. For you to understand me, I need to tell you some of my experiences growing up with my metabolic disorder. I have been everything from the star patient to a nasty child. I have gone from loving my formula to dumping it down the drain. I have gone from asking, *why me?* to asking, *why me?*

I was born in Silver Spring, Maryland on August 18, 1970. I was diagnosed with PKU by newborn screening at the age of six days. Thirty-one years ago, my parents, Peter and Judy Foster, were in a state of shock. (I understand many parents are in a state of shock when they first hear their baby has a metabolic disorder.) They had never heard of PKU prior to receiving "the call." After learning that my newborn screening test results were positive, my parents took a picture of me, and my older brother and sister, before they took me to the hospital. They thought I might die.

Shortly after I was diagnosed with PKU, I was enrolled as a participant in the National Collaborative Study of Children Treated for PKU. For those of you who aren't familiar with PKU or the study, it was first thought safe to take children with PKU off the diet at around the age of five or six - when it was thought our brains had stopped developing. The Collaborative Study closely followed 212 people with PKU from birth to age 12. This study ultimately resulted in the current recommendation to continue diet for life. My memories of participating in that study strongly influence my thoughts and feelings today.


Tiffany Gant & Jean Handler

For the most part, my parents describe me as a happy-go-lucky little kid growing up. I don't remember things quite that way. I don't remember the good things, or maybe I should say, I don't recall the good things often enough. There are good things I guess you only truly appreciate when you are 31. I remember having a bottomless supply of jellybeans. I

remember strawberry picking with my Mom every 4th of July. Most of all, despite it all, or perhaps because of it all, I remember the uncon ditional love my parents gave me.

All of this doesn't change the fact that there isn't much positive I remember about having PKU in my youth. I tend to remember the bad things, like Lofenalac [PKU formula], the dietitian coming to take my blood every month, wanting to be like other children, and the children teasing me. I remember my parents being very strict with me - I wanted to eat high protein foods. I remember the EEG's and the endless IQ tests. I remember not wanting to be a guinea pig anymore. Most of all, I remember being scared and lonely.

For a long time I didn't think that I was different from anyone else. Gradually, I figured out that the games the psychologist played with me weren't games. They were IQ tests given so they could assign a number to me that told them how smart I was. I didn't want to be put on display. I was smart enough to know I didn't want to answer question after question from doctors who had never seen someone quite like me. I didn't want to have electrodes stuck to my head for an EEG or for any reason. Why couldn't my doctors look past the numbers and the mountain of medical records to see me? Why weren't they smart enough to see that I was scared and lonely? Why weren't those doctors smart enough to understand and answer my question *why me?*

Around sixth grade, I discovered the library contained books other than "Monster Goes to Park." I started to read as an escape from a world I felt I didn't really fit into or was even sure I wanted to fit into. I began more frequently to eat things I knew I should not eat much and stopped drinking my formula. Cheating on my diet was my way of making my parents hurt as much as I did. It got to the point where I was eating so many things I shouldn't, that it couldn't be called cheating - it was being off-diet.

Cheating and being off-diet for people with PKU is much different than for people with MSUD. When our blood levels rise and get out of control, there is no immediate threat or reaction. There are some consequences for people with PKU, but they pale in comparison to the consequences for people with MSUD. We will not become sick, we will not go into a coma, and we will not die.

I have been asked if I have cheated on my diet so often that it has given me an inherent, internal sense that I am a bad person. Cheating, and ultimately going off the diet, has made me feel like a failure. I can't tell you how many times, when I was a teen, I told myself, "Okay, I am not going to cheat any more. I am going to be good." My secret New Year's resolution every year was always the same. Inevitably, I would fail. You can only fail so many times until you feel that you are a failure. What I didn't understand then is that you never run out of chances to try, and as long as you try, it is impossible to fail. Now each year my only New Year's resolution is to try to make the coming year better than the last.

I remained off-diet all during high school, college, and graduate school. During that time, I didn't allow PKU to be a factor in my life. No one knew I had PKU, and, as far as I was concerned, I didn't have it. Upon completion of my master's degree, my focus turned from my books to myself - the books were easier. Over time, my reasons for being off-diet faded away. I returned to diet because I finally realized that the only person I was hurting was me.

I know returning to diet will never be an issue for people with MSUD as going off-diet is not an option. I think what I have learned may help those with MSUD better adhere to their diets and more easily deal with some of the demands that their metabolic disorder places on them.


Brittany Fuller, Raelynn Safchick, Alana Moceri with Elan Geffan standing in back

While I was off-diet my parents would occasionally send me something about PKU in the mail, such as the National PKU News with "diet for life" highlighted. It only served to make me angry. Oddly enough, the thing that got me thinking about PKU again was mail my parents forwarded to me that they didn't realize was PKU related. It was a letter sent to every patient of Lib Walker, my dietitian when I was a child, letting us know of a retirement party for her. I was unable to attend the party, but I sent a donation towards her retirement gift. The notification made me realize how much I owed her despite the fact that she wasn't exactly my favorite person when I was young.

Even though it was a little more than a year before I made my decision to return to diet, Lib Walker's retirement got me thinking seriously about PKU. There are two things in my life that I regret more than anything. With all my heart I regret having gone off-diet, but even more than that, I regret that I never said thank you to those who helped me in the treatment of my metabolic disorder when I was younger.

For as long as I can remember, I have been angry about having a metabolic disorder. What angers me now is different than when
I was eleven. At that time I felt I was being treated as though I was a lab rat. I remember going to see my doctor at Jackson Labs in Bar Harbor, Maine when he was there for a conference.

I heard that they hoped to soon have a PKU mouse. I was angry because I found it hard to relate to a mouse. I felt as if I was a rare freak of nature. More than anything though, I was angry because I felt no one really understood, and I felt alone.

 Raelynn Safchick &
Sharlene Woorman

Now that I am thirty-one, I get angry at different things. I get angry that there are very few clinics specifically for adults who have metabolic disorders. Even though we have grown into adults, it seems the medical community has not grown with us. I get angry because I am constantly being asked how old my child with PKU is. I get angry at the insurance industry, and the fact that so many adults have problems getting coverage. There isn't a time when I'm not really, really angry that every state isn't screening for MSUD and other treatable metabolic disorders. There are times I get so angry that I want to kick, shout, scream and cry; there are times when I do. My temper tantrums, now that I am grown, are a bit more controlled and refined.

It is difficult to deal with my anger from the past, my frustrations of the present, and my fear of the future. It is hard, but I am dealing with the anger towards myself that I went off-diet, my lack of self-confidence and self-esteem, and my feelings of sadness and loneliness when I was growing up. The most difficult thing of all, however, is dealing with my feeling that I was a burden to my parents. My parents never once said or in any way indicated to me that I was a burden to them, but I still feel this way. I guess now I can appreciate how difficult it must have been for them. How scared and lonely *they* must have felt when they were told I had PKU and there was so little information available. How hard it must have been for *them* when I was fighting them each step of the way.

Now I am dealing with my feelings whereas before I shut them out. When I was eleven, I thought that if I shut out my feelings and didn't talk about them, that I would be safe and protected from them, and I wouldn't be scared and lonely. I only accomplished shutting my feelings in. The only thing I managed to shut out was the outside world and those who cared about me.

 Group of teens and adults with MSUD

It is easy to be angry at a disease, a doctor, a dietitian, a psychologist or anyone who is insensitive. It is much harder to deal with the anger. The major difference between the anger I have now and the anger I had when I was eleven, is that now the anger drives me rather than consumes me.

I'd like to think that I am using my anger constructively. I want to try to make positive changes for those with metabolic disorders. I know I can't change my past, but I can change how my past influences my present and my future. Most people talk about formula and foods being the big issues with respect to metabolic disorders - and they are important issues. However, it seems people ignore the psychological aspects involved. To me, dealing with the food and the formula are a piece of proverbial low protein cake compared to the psychological aspects.

Even though there are many frustrations and some inconveniences associated with having a metabolic disorder, for the most part, managing it doesn't influence my social activities. I think about what I am doing and where I am going and plan accordingly. I have never let my disorder and the diet keep me from going anywhere or doing anything I wanted to do. It does, however, influence my activities and choices in life to some extent. I can't imagine being friends with or caring deeply about someone who isn't flexible and understanding of my disorder and me. I have also chosen to be actively involved in helping others.

Although I have returned to diet very successfully, I am not fully comfortable with my relationship with my parents as it relates to PKU. A lifetime of conflict and old habits aren't erased overnight, but it is getting easier. Gradually I am involving my parents more in things that relate to PKU and my life. Recently, I invited them to come for the day to the Maine Family Metabolic Weekend. It was a strange experience to have my parents there. That weekend made me realize my parents need support too - maybe even more than I do. I have been fortunate. Where would I be without the support and understanding of all the amazing people I have met? Where I would be without all of you?

A few years ago, at the New England Connection for PKU and Allied conference, children eight and older were able to participate in a variety of activities while their parents attended workshops and listened to speakers. At a reception the night before the conference, I overheard a little boy whisper to his mother, after she had pointed out another little boy with PKU, "Mommy, he doesn't look like he has PKU." I think his whisper speaks volumes about how many of us with metabolic disorders feel or have felt in the past. I know I don't have it tattooed on my forehead, but even now I feel I somehow look different because I have PKU. Perhaps it is because I feel different.

My parents wrote many notes and kept records after they were told I have PKU. One question my father asked about six days after I was diagnosed is, "Does she look like a normal PKU kid?" Thirty-one years after the question was asked, the answer is - I am not sure Dad. I am not sure because there isn't one single definition of what a PKU kid looks like. Even we can't tell each other apart from other people. One thing I can tell you, Dad, is that I am beginning to feel more and more like a normal PKU kid. Maybe someday I will even feel like a normal PKU adult. Who knows, some day I might even feel like a normal adult.

The definition of what an adult with a metabolic disorder looks like is changing. Those of us who were diagnosed and treated early for metabolic disorders are now older than the parents giving birth to the next generation of children with these disorders. We are receiving our bachelor's, master's and doctoral degrees. We are getting married and having children of our own. We *are* growing up.

Even now that I am grown up, I still ask, *why me?* The question is the same, but twenty years has changed the inflection and intonation. I used to think that having PKU was the worst thing in the world that could happen to anyone. Now, I only wish that it were true. Shortly after returning to the diet, I met a person who had not been diagnosed until the effects of high levels of phenylalanine had severely taken their toll. It was a strange feeling knowing how easily the tables could have been turned. I was born 10 miles from Washington, D.C., where they didn't do newborn screening at the time. *Why me?* At the first metabolic conference I went to, I learned about diseases that are very similar to PKU, but much more rare and devastating. I met a four year old with one of those diseases who was unable to walk or talk because he had been diagnosed too late. He had suffered a debilitating stroke. *Why me?*

Knowing what I do now, I try to ask the question, *why me?* less often. When you don't know the answer it is sometimes beneficial to stop asking the question. I am beginning to suspect that I can never really understand. In the movie "Patch Adams," the title character, played by Robin Williams, discovers that sometimes when you focus so hard on the question you lose sight of the answer. By focusing a little less on the question, *why me?* I think I have come up with an answer. It is - I don't know. Sometimes I think we fail to see that as a perfectly valid answer.

When I was two, I asked my Dad a billion questions. After a long time of answering fully and completely, he began to answer me by simply saying, "Because." I was just as happy as if he had given me a twenty minute explanation. In my opinion, my Dad's answer of because, and my answer of, I don't know, both truly and completely answer the question, *why me*? This doesn't mean I have stopped asking the question or stopped looking for another answer.


Kylie Williams & friend from Australia

A couple of years ago, I received a call from Dr. Kenneth Wessel, the psychologist who administered the IQ tests to me while I was growing up. He told me about a followup to the Collaborative Study and asked me to participate. After much thought and turmoil, I consented. As part of the study, Dr. Wessel came to Boston and gave me another IQ test. I was also flown to Los Angeles and given an MRI/MRS. During my flight, I sat next to an

elderly couple. I told the lady about PKU and the reason for my trip to LA. Since the weather in both Boston and Los Angeles was expected to reach a high of 60 that day, I mentioned, "Why go to LA?" The lady responded, "To benefit science and humanity." For a long time I thought the lady was correct. However, I decided her answer was incomplete. The complete answer is - to benefit science, humanity, *and me.*

I am slowly realizing the value of research in metabolic disorders. For a long time I hated anyone or anything associated with PKU because of my experiences participating in the National Collaborative Study. I realize that the research benefits me as well as others. I now know it wasn't those doctors who couldn't look past the numbers and the mountains of medical records to see me; it was I who couldn't see myself. It was I who was defining myself by the numbers and the mountain of medical records, not the doctors.

For the last two years I have attended Dr. Rani Singh's Metabolic Camp for girls at Emory University in Atlanta, Georgia. I had the pleasure of spending those weeks with six women who have MSUD, and I have learned a little more about MSUD. Far more importantly, I have come to know a little more about Leslie, Amy Beth, Jessica, Leanna, Alana, and Jean as people. Although MSUD is a very important part of their lives, it is not their total focus in life. If you only see them as persons with MSUD, you do not see them as they really are. You do not hear Leslie's humor, feel Amy Beth's caring touch, hear Jessica's poetry, appreciate Leanna's wit, witness Alana's inquisitiveness, and know that famous people know Jean.

If in the same way, when you look at me, all you see is PKU, then you really haven't seen me at all. I am still in the process of discovering that I define PKU, PKU doesn't define me. It is true that if you look at the DNA in every cell in my body you will see PKU. I haven't reached that point in my life where I am able to see what lies beyond PKU, but I know I will get to that point someday. I am sure that I am much more than a set of numbers and a mountain of medical records.

My cellular structure probably has a college hockey gene right next to the PKU gene and the potato gene. In the past speeches, I have told the story about the 1993 University of Maine hockey team losing to Boston University (a fierce rival) in overtime. And how, after the game, Shawn Walsh, the head coach said to his players, "It's not how hard you fall, it's how fast you get up." The University of Maine hockey team went on to win the national championship that year. They finished the season with a record of 42-1-2. That overtime game was their only loss the entire season.

Now, my story doesn't end there, but ends in April of 2002 with another University of Maine overtime loss and another national championship game. However, this time the overtime loss was to the University of Minnesota. This time Shawn Walsh didn't give his players a motivational speech after the game. At the age of 43, Shawn Walsh died of cancer on September 24, 2001, the first day practice was scheduled to start for the new season. His players dedicated the season to the memory of their much beloved coach. For much of the game it looked as if the players were going to win a national championship for Coach Walsh in a fashion the likes of which only seems to happen in storybooks and movies. But the game took place in real life and the University of Minnesota scored with 52.4 seconds left to send the game into overtime. The University of Minnesota won. And in real life, Coach Walsh died.

Recently I was speaking to an adult with PKU who was very angry that the world had given her PKU. I responded by telling her that the world didn't give PKU to just her, but to me too. The world gave PKU and MSUD and other metabolic disorders to thousands of people like us. I told her that it is much easier to be the victim, but it feels better to be the victor. If any of you are fans of the University of Michigan Wolverines and know their fight song, you know we "Hail the Victors."
The University of Maine lost.

Shawn Walsh died.

You and I and thousand of others like us have metabolic disorders.

But win or lose we are playing the game.

And win or lose, just by staying in the game, we are victors.

Hail to the Victors.

Hail to all of you the conquering heroes.

Hail and farewell.

- Sarah Foster