VOLUME 10-1

Spring/Summer, May 1992

FAMILY HISTORY - CHAD & HIS LAST HOSPITALIZATION

Details

Written by Lynn & Skip Adams

Published: 14 July 2009

None of us like to hear sorrowful news. It was great not to have to report any deaths of children with MSUD in our last several newsletters. We always hope medical care has reached the point where these children and adults can live normal lives. However, other children die from various causes, so death is a reality for all of us. Our third history is a mother's touching account of the last illness of her son. The Skip Adams family is from Florida. We extend our sincere sympathy to this bereaved family.

Chad was diagnosed with MSUD approximately two weeks after his birth. At that time, he was having seizures and was in a coma-like state. After two months in the hospital, we brought him home.

Chad had not been sick to the extent that he had to be hospitalized since July of 1988. We felt very fortunate. We were always able to adjust his formula and control his diet. The doctor was just the best we could have asked for. He was a metabolic specialist at Tampa General Hospital. He was always there for us. We took blood from Chad once a month and had his amino acids checked. We had been doing this since he first came home from the hospital in November of 1987.

Chad turned four on Sept. 3, 1991. Everything was going fine. He was enrolled in the local preschool program for disabled children. Although Chad was four, he was unable to walk without a posture walker. He had received physical therapy twice a week since he was about a month old. We felt Chad was fine mentally. Actually we felt he was very intelligent. His delay was mainly in his physical development. He was such an outgoing child.

Earlier this past year we were notified by the local Tampa Shriner's Hospital that he was accepted for surgery on his legs, but the waiting list was about four months. On September 23, 1991, we admitted him to the Shriners Hospital for surgery. The purpose of the surgery

was to lengthen the tendons, heel cords and abductors in his legs. The surgery went great. He was put on an IV the night before, and was off his formula less than 24 hours.

He was to be in the hospital a total of eight weeks for therapy. I remained at the hospital with him the first week, and then returned to work. We spent the weekends at the hospital with him.

After four weeks, they took the cast off his legs to below his knees. During this time, they worked with him standing and walking. Then his amino acid levels got out of control. His doctor worked with the Shriner's Hospital adjusting his formula as we always did when his levels changed. Slowly Chad got so that he didn't want to eat his food or drink his formula.

On the fourth weekend, we brought him home from the hospital and set up his therapy with his private therapist, because the hospital felt that he would do better at home. I think they were also a little nervous about his condition.

By Sunday night, Chad was throwing up and wanted to sleep most of the time. He had lost a considerable amount of weight. We took him to the emergency room at Tampa General that evening, and his doctor, Dr. Terry DeClue, admitted him and immediately started to work on getting the levels down.

After the second day, the doctor had the levels down enough to start adding the branched chain amino acids back into his formula. Chad vomited most of that up, so they had to put in a feeding tube. Later on that third day, Chad had a seizure. A short time after that, on Oct. 23, he died in his sleep.

FAMILY HISTORY - OMAR FROM SAUDI ARABIA

Details

Written by Amal Al-Sibai

Published: 14 July 2009

Our family histories reveal some interesting variations. The first is a history from the Sibai family who now live in Saudi Arabia. While living in Kansas, where Omar was born, they attended our second symposium held in Kentucky in 1984. Omar's father, Atef, is a doctor. Omar's sister writes this history of her brother.

Omar was born in 1983. We all rejoiced when we heard that we had a baby brother. My mother came home two days after his birth. Of course we had no idea that he had MSUD, so my mother

was breast-feeding him.

One night, not knowing what was in store for us, my father checked on Omar. My sister and I were also awake at that time. My father found Omar's face blue and he wasn't breathing. My father breathed for him as he called an ambulance. A few minutes later the sound of the deafening ambulance siren awakened my mother. My father went with Omar to the hospital, because my mother was confused and couldn't recover from the shock. A friend of my mother came to our house. My mother spent the whole night praying and crying.

From Hutchinson, Omar was transferred to Wesley Medical Center in Witchita. My father lost hope for Omar's survival, but my mother kept striving strongly until finally Omar recovered. Doctor Cho discovered that Omar had been a victim of MSUD.

We would take Omar frequently to Doctor Cho. We would watch his diet at home, and we put him through physical therapy.

In 1985 we moved to Riyadh, Saudi Arabia. Omar was two years old at that time. We registered him in the King Faisal hospital. There are a lot of children with MSUD here. The problem is that they are diagnosed late, so their cases are more severe.

For the past five years we have avoided taking any blood tests, and he's been doing fine. We take a urine test by the strip so we'll know if he has been taking too much protein.

For a year now there was only one episode in which Omar became very ill. We do not take him to the hospital; my father puts an IV in for him. My mother devotes all her attention to him if he happens to get sick. His main problem is that he gags, usually in the morning. We sometimes give him medicine that will help him cope with this feeling.

Omar has miraculously improved. He is in first grade and goes to a normal school. He is a very bright boy and does not hesitate to study. He also has many friends. He drinks his Milupa formula, but we encourage him very much.

Just recently my mother gave birth to a baby girl. She was born on October 31st, 1990. We were extremely worried about the fact that she could also have MSUD. Praise be to God, after some lab tests, we discovered she did not have MSUD. Although we have a new baby, I think Omar will remain greatly special to all of us.

FAMILY HISTORY - AMOS AND THE BRAIN TUMOR

Details

Published: 08 July 2009

Our second history is actually a report of a very successful major surgery on Amos Fox who has MSUD. This family has experienced much sorrow. Their first children with MSUD were born before the disease was recognized in the Lancaster County area of PA. Their first diagnosed daughter succumbed to the brain edema that has taken the lives of so many of these children.

Amos had the advantage of the new TPN (a special intravenous nutritional therapy), and the constant day and night care of a dedicated doctor, Dr. Morton from the Clinic For Special Children.

Dr. Morton planned to prepare an account of the operation for our newsletter. He was unable to complete it for this issue. He learned some pertinent information through this experience which may be valuable in helping other children with MSUD during hospitalizations. We are eager for him to share with us and hope to have it for the next newsletter.

Amos is our 13 yr. old child with MSUD. He has five brothers and two sisters living. Three sisters have gone to their reward. His two oldest sisters died in infancy, undiagnosed. The first, born in 1963, died at the age of 5 weeks and another sister died at 13 days of age in 1967. In 1971 Susan was diagnosed at 9 days of age. She died from a viral infection when she was almost old 4 yrs old. Since then much has been learned about MSUD.

Amos was doing good this past summer and early fall. Dr. Morton thought maybe we could cut down on his formula intake since it is so expensive and increasing in price all the time. So we cut down on formula and substituted coffee creamer and cornstarch for calories. I guess we didn't add enough calories since he didn't gain weight all summer. In November he started vomiting in the mornings, sometimes just gagging. He would be all right the rest of the day and kept going to school.

We took him to see Dr. Morton the last week in November. His levels were all right that day, but he did vomit a little in the doctor's office. Our first thought was that we shouldn't have changed his formula. The vomiting continued and Dr. Morton did some tests.

The last week, before entering the hospital, we had trouble keeping his levels down. If I put him back on protein, his 2-4 DNPH would turn cloudy. On his bad days his vomiting would last about two hours. We always got him settled in time for school. Finally the last day he was home, he was bringing up his formula most of the day.

On Dec. 14, a Saturday evening, Amos was admitted to the Lancaster General Hospital. We thought he probably had the flu that was going around. Dr. Morton thoroughly examined him. Amos didn't want to lay his head on the examination table. Dr. Morton was already suspicious of a brain tumor. Amos' refusal to lay his head down plus his vomiting in the mornings seemed to be clues to Dr. Morton. All other tests were clear. Dr. Morton kept saying, "I don't think it is his MSUD, do you?" As far as we know he had no double vision or headaches. His level was still only 7 mg./dl.

After my husband left, Dr. Morton said he still thinks there is something in the back of Amos' head. Oh, what a blow.

The next day, Sunday, they did a brain scan. Dr. Morton was with us most of the afternoon. After checking the reports of the scan, he returned saying there is trouble. I asked him if it is a tumor and he answered, "Yes".

A surgeon came to talk to us. We had a decision to make. We asked him a lot of questions, and he told us it was a very serious operation. I asked if it could paralyze him. He said it could, but the chances weren't too great. I asked what his chances were if we didn't do anything. He said they would give him three weeks to live. We knew that was in the hands of the Lord.

The tumor was close to the brain stem. We chose surgery. Then we had to decide in which hospital to have the surgery because of his MSUD condition. I asked Dr. Morton if he would consider doing it in our local hospital. He was willing, but said it would make him very busy. (He is a very busy man to start with.) Dr. Morton would take care of the MSUD and the surgeon would operate on the tumor.

On Dec. 18 Amos had surgery. It took seven hours. It was a long day for us, but he came through surgery with a leucine level of 6 mg/dl. He was in the trauma unit for 48 hrs. The day before Christmas Amos was taken off his IV, and the next day he was supposed to drink 60 oz. of formula, if I remember correctly. That evening his leucine had gone up to 31 mg/dl. He was in real trouble. I would like Dr. Morton to write about how he quickly got it corrected. He worked hard, spending many hours in the lab running tests and traveling to the hospital.

The day after New Year, Amos came home from his 19-day stay in the hospital. They had removed only 80% of his brain tumor. It was a low grade cancer tumor. He needed radiation 5 days a week for 6 weeks. Twelve days after he was home, he began treatments. We wondered if Amos could tolerate radiation. Some of the doctors were concerned, but I am happy to write that he had very little side effects. At times his appetite was poor, and he lost some hair behind his ears. Other than that, he did real well.

After four weeks of treatments, an MRI showed the tumor was gone. What a relief! We remember being told it could grow back. His future is in the hands of the Lord.

The last two weeks of his treatments he went full days to school, getting his radiation in the morning before school. I would say Amos is as well as he was before he was sick.

I want to thank all of you for your prayers sent heavenward during Amos' illness. And a special thanks to Dr. Morton for all his time and efforts. We were richly blessed again.

Weaver & Alma Fox
NOTE: In the year 2000 Amos is a strong, healthy, active young man. See Vol. 10 No.2, Dec. 1992 for Dr. Morton's account of the surgery.