ACDA NOTES

From The Alveolar Capillary Dysplasia Association

Volume 9, Issue 1
http://www.acd-association.com
Spring 2007

Dear Friends and Family,

In this edition of *ACDA Notes*, we continue our memorials to our babies with the story of Olivia Durand. Thank you to the Durand family of New York who has graciously offered to share the story of their daughter who died of ACD in 2002. In the years since Olivia's death, the Durands have engaged in various fundraisers in memory of their daughter and to raise money for the ACD research account at the National Organization for Rare Disorders. Their creative and entrepreneurial spirit has been instrumental in raising over $13,000 in Olivia's memory. A special thanks to the Durands for sharing Olivia with us and for their support of ACD research.

If you would like to share the story of your baby, please email it to us at sdesj@verizon.net.

Fondly,

Steve and Donna Hanson
Executive Directors, ACDA

INSIDE

Olivia's Story

By Lisa Durand

Olivia Leona Durand was born on November 2, 2001 (All Saints Day). She was the third child of my husband Harry and I and finally the little girl we had hoped for. I had Gestational Diabetes with her and my previous pregnancy, but my other two boys were healthy and I had assumed the same would be for her. She was born a few weeks early and still weighed over 8 lbs. I had her cesarean section and was told that she ingested some amniotic fluid during delivery so she would have to go to the NICU unit. I never really worried because they said that was very common and I never expected to have a sick child.

This birth was different then the other two. I did not feel good at all after delivery and was very tired. I could hardly wake up and did not hold her until the next day. When I did see her she had tubes in her head and the oxygen in her nose. She was still so beautiful. The NICU nurses assured me that in a few days she would be fine and I could take her home. Still no worries. As promised I took her home from the hospital and life was good. She was very different then my other two babies. She was a little

Continued on Page 2

smaller then my first son Joseph 10 lbs 7oz and Alexander 10 lbs 6 oz, but that was to be expected because she was a girl.

Olivia was never was a big eater, but she would nurse as expected. When she was around 4 months we were told to introduce her to formula and cereal. She did not do well with either. She could not really keep her food down and would projectile vomit most of the time. I was worried because she was not gaining any weight and she would sometimes turn blue after her nightly bath or when vomiting. The doctor told me not to worry that not all babies are the same and it is ok that she only gained a few pounds since birth. She then had strep throat. My two boys had it and she was acting different so I took her to the doctors. They told me that babies can not get strep and especially when they are nursing. Well, they were wrong. She had it and was treated.

There was something inside me that I did not realize until after she passed, but I knew she was not going to be here long. I bought enough clothes to outfit a dozen babies. I would also buy sizes bigger all the way to size 4 thinking that if I bought the clothes she would wear them. I even told her Godmother that I do not worry about the money I am spending because I feel like I will not have her forever. Stunned and angry at me she questioned why I would say such a thing and I did not know where that came from. I guess a mother always knows.

It was April 7, 2001 and I was at a baby shower for a friend. I had Olivia dressed up like a princess and everyone was looking at her and commenting on her beauty. Someone had asked me if I thought her lips looked blue and I explained that I had taken her to the Doctors and she is little and everything is OK. About 10 minutes later I looked at her again and I realized it was NOT OK to be blue. I abruptly left the shower and told my Mother I was taking her to the hospital. Of course my Mother thought I was over reacting, but was later glad I did.

When we got to the hospital her oxygen level was 47. They thought their equipment was not fitting correctly so they took it again and after 5 tries they realized something was wrong. She was not crying or in pain, just sleepy and a tinge blue. They told me she had broncholitis and she needed to stay. We were in this hospital for a week and she was in an Oxygen tent. When she was in the tent she was pink and happy and beautiful. When we would take her from the tent to feed her or hold her, her lips would turn blue. I knew something was not right. The Doctor insisted that these things can take a few weeks but I had a sick feeling he was wrong. We insisted that we be transferred to a better hospital that had a PICU unit. After resistance from the doctor we transferred to the hospital and were told they thought she had a hole in her heart that never closed up during birth. As bad as that sounded it was fairly common in newborns and a simple operation would help her. We were glad that she was diagnosed and would be fixed.

The next day the heart doctor came in to do an Echocardiogram to assess the situation. When he was doing the procedure we could tell he was surprised and concerned. He told us he would be back and we waited. When they returned they told us that they believed she had Pulmonary

Hypertension. A rare disease in infants that usually is characterized as high blood pressure in the lungs. They said it is a hard thing to treat especially in infants, but they were doing their best. They gave her Viagra to help dilate her veins and then for the first time in that hospital with a baby they tried Nitric Oxide to bring the pressures down. From here she went downhill fast. She went into cardiac arrest five times in one day. I remember watching them bring her back each time thinking it was over. They finally stabilized her and told us that there is nothing more they could do. One of the Doctors had called New York Presbyterian Hospital in New York City and hoped that they may have a solution because they had a group there studying Pulmonary Hypertension. Olivia was very sensitive and when she got mad or was not happy her blood pressure would drop and her heart would race. They were not sure if she could make the trip to New York City, but we felt that we had to at least try.

We made it to New York City and that is where she spent her last 32 days. When we first got there they put her on an ECMO machine, which actually performs the functions of the heart and lungs. They warned us that most babies get large heads full of blood and bleed from their nose and mouth and wanted us to be prepared. When we got there they hooked her up and she responded well, opened her eyes, smiled at us and gave us hope. This machine was very scary and it was manned 24 hours a day. We could not hold her because there were canulas going into her heart and she had to be still. They told us that this would give her time to heal without causing brain damage or hurting her heart or lungs. The Pulmonary Hypertension specialists visited her often and were developing their plan. As the days went on they thought they could remover her from the machine. They did not want to leave her for more then one week because it could cause internal bleeding. When they took her off they had her covered in the blue scrub towels they use in the hospital and had one on her chest and body and one on her head. She reminded me of the Virgin Mary and I still had that feeling like the end was near.

Each day she got worse, not better. They could not keep her blood pressures up and the pressures in her lungs down. The Doctors tried every combination they could think of with medicine and ventilators and they would keep her somewhat stable, but not really get any further ahead with a solution. After 32 days of 24 hours 7 days a week attention Olivia could not fight anymore. The Doctors and Nurses knew that there was nothing they could do and she would be fading away. My husband had left that morning and by 4 they had told me there was nothing left to do. My husband was taking my two sons to their ball game and then was going to come back to the hospital. When I called him with the news he realized he could not leave and make it there in time and wanted me to call him when she passed (we were 3 hours from home). My Mother was on her way up to stay with me until my husband arrived. It was 6:30 or so and I was holding Olivia on her bed. It was the first and last time that I was able to hold her in 38 days. Her heart rate slowly started to drop. I prayed that she would not leave until my Mother arrived. My Mother walked in the room and gasped when she saw me on the bed. She asked what I was doing and if she was doing better. I told her that this was the end of the road for Olivia and my Mother leaned over and kissed her. Within minutes Olivia passed away.

During her stay at the hospital in New York I decided to start emailing my family and friends so they would not bother my husband or my parents while they were trying to get through this. I did not want them having to go over every detail each and everyday. The amazing thing was that I started with a handful of friends and family and each day the group would grow. I would give detailed explanations of what the doctors were thinking and doing and what our hope was for the outcome. People would email me back with their prayers and wishes and ask to be added to the email list. There were people I had never met that were in one way or another connected to someone I knew. Churches were having masses in Olivia's honor, families were having prayer groups and everyone was having faith. By the end of Olivia's life there were over 150 email addresses that I personally sent to and then those recipients would pass it along to their friends. The email list was actually a worldwide chain of love reaching Europe and several other countries.

Several weeks after Olivia died the doctor in New York called me and told me that the autopsy discovered she had Pulmonary Hypertension and that was the cause of death. She told me that she was not really convinced of their findings and wanted permission to send Olivia's slides to some other doctors to get their opinion. Dr. Claire Langston was the doctor who confirmed the New York doctor's suspicion and told her that Olivia had died of ACD. When I heard this, I tried to get as much information as possible on this disease and tried to make senses of it. This is when I found ACDA. I was able to talk to other parents, share my story and make it real. I have met wonderful people all over the world that have lost a child or children and I have a connection with everyone of them.

Olivia was only here 5 ½ months, but impacted many people forever. From the day she was born, she was adored by everyone that met her. I dressed her everyday like she was going to a party. She was with me every minute of everyday from the day she was born until the day she died. She went to work with me every day and everyone loved her. When she got sick, she touched people literally all over the world and all different ways. At her funeral services, people came to say goodbye to her that I did not know. They thanked us for the opportunity to be part of her illness and claimed that she had helped them. My husband and I found ourselves consoling people that came instead of us being consoled. I guess we will never know why this had to happen to Olivia or why it continues to attack other children. I do believe that everything in life happens for one reason or another and that we do not need to define ourselves or our lives by these tragedies. We talk of Olivia often and so do my children. We do not try to hide the fact that we lost her and we try to educate everyone we meet. I volunteer at the Ronald McDonald House at the hospital that helped us so I can let parents know they will get through their hardship, I did. Olivia is gone for now, but will always live in our hearts.

Update on ACD Research

New Research from Germany

Recently, a research team in Germany, headed by Dr. Anita Rauch, claimed that they have found mutations in a gene called STRA6 in a variety of malformations, including ACD. These mutations reported by Dr. Rauch are currently being investigated in DNA samples of ACD families in the Baylor repository.

Mutations in STRA6 cause a broad spectrum of malformations including anophthalmia, congenital heart defects, diaphragmatic hernia, alveolar capillary dysplasia, lung hypoplasia, and mental retardation.
Am J Hum Genet. 2007 Mar;80(3):550-60.
Epub 2007 Jan 29.
PMID: 17273977 [PubMed - indexed for MEDLINE]

To view the abstract of the article published by Dr. Rauch and her team, go to:

http://www.ncbi.nlm.nih.gov/sites/entrez and do a search for 17273977.

Baylor ACD Study Update

In addition to this work, Dr. Clare Langston at Baylor is preparing the manuscript of the immunohistochemcial study that was funded by the $35,000 grant from the National Organization of Rare Disorders.

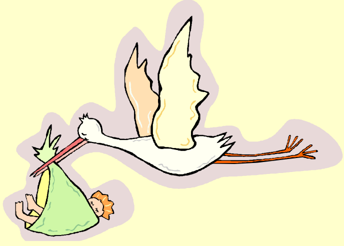
SAFE ARRIVALS

[redacted] [redacted] [redacted]
Born: [redacted]
To [redacted] [redacted] [redacted],
Brothers [redacted] & the late, and sisters [redacted]
and [redacted]

****

[redacted] [redacted] [redacted]
Born: [redacted]
To [redacted] [redacted] [redacted]
and
Sisters [redacted] & the late [redacted]

Congratulations to both families!



Announcements

- Lisa and Harry Durand of New York recently lost a loved one around the same time of year that their daughter, Olivia, passed away from ACD five years ago. In lieu of flowers from friends and family members, the Durands asked for donations to NORD. A total of $1,000.00 was donated to NORD in memory of both Harry's father and Olivia.

- The [redacted] family of [redacted] recently made a $1,000.00 donation to NORD in memory of their daughter, [redacted], who died of ACD in January 2007.

Thanks to both of these ACDA families for their generous donations!

ACDA CONTACT INFORMATION

Website – www.acd-association.com
Steve and Donna Hanson
5902 Marcie Court
Garland, TX 75044-4958 USA
(972) 414-7722
Email – sdesj@verizon.net