

ACDA NOTES

From The Alveolar Capillary Dysplasia Association

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Dear Friends and Family,

On November 15, we had the opportunity to meet with the team of ACD researchers and Craig Snyder. Craig recently founded the 3 Angels Memorial Fund for ACD Research and was instrumental in getting Congressional approval this week for \$250,000 to fund ACD research in fiscal 2008. Craig requested the meeting at Baylor College of Medicine in Houston, Texas for the purpose of discussing the best way to move forward with ACD research. The details of our visit are explained in an article on page 2, but the recurring theme that we heard from the ACD research team was the need for additional research samples from affected children. The main criticism of a prior Baylor proposal to the National Institutes of Health was that the limited number of samples available limited the chance of success. The critical contribution that ACDA members can make to help with this is a donation of tissue samples, paraffin blocks, or blood samples that were collected from your baby. Baylor has less than a dozen complete sets of samples and needs at least a threefold increase to improve the chances for a successful genetics study. Consent forms for this contribution have to be obtained from Baylor. Prior to contacting your hospital, please contact Dr. Partha Sen at psen@bcm.tmc.edu or (832) 824 4764. While samples from affected children are of utmost importance, Baylor reminded us that blood samples from parents and healthy siblings are also needed.

If you haven't donated samples to Baylor, we urge you to do so now. This is an opportunity for something positive to come of your loss and to know that your one contribution can make a difference. In addition to honoring your child lost to ACD, you need to do this on behalf of the healthy children that you have. Only through this research can the cause of ACD be determined which will eventually reveal if your healthy children's offspring may be affected.

Finally, may you create new family traditions this holiday season that include the babies that you have lost to this terrible disease. All our best for a healthy and prosperous 2008.

Happy Holidays,
Steve and Donna Hanson
Executive Directors, ACDA

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ACDA Attends Research Meeting

As we informed you previously, Craig Snyder and his wife, NiCole, established the 3 Angels Memorial Fund for ACD Research in memory of their son, Lincoln, and Craig's two other children, Rebecca and Jonathan who all died from ACD. One of the goals of the 3 Angels Fund is to raise money for ACD research. Craig has spent many hours talking to medical professionals around the United States to help jump start research. On November 15, 2007, he convened a meeting at Baylor College of Medicine, to discuss the on-going research by Partha Sen, Ph.D. and Dr. Claire Langston, a pediatric pathologist. In addition, Drs. Galambos and Barmada attended as they have the skills needed to complete the team and are interested in ACD research. We had the opportunity to attend this meeting.

In addition to us, the following people were in attendance:

- Craig Snyder, founder, 3 Angels Memorial Fund for ACD Research
- Dr. Naomi Bishop, Assistant Professor of Pediatrics at Weill Medical College, Cornell University (Naomi cared for Craig's child Lincoln in the hospital)
- Dr. Claire Langston, Professor of Pathology and Pediatrics, Baylor College of Medicine
- Partha Sen, PhD, Assistant Professor, Dept. of Pediatrics – Nutrition, Baylor College of Medicine
- Dr. Csaba Galambos, Pediatric Pathologist, Children's Hospital of Pittsburgh
- Dr. Michael Barmada, Department of Human Genetics, University of Pittsburgh

Dr. Langston opened the meeting with an overview of her involvement in ACD cases since 1989. She gave an update of the immunohistological study that was funded by a NORD grant. As you may remember, the immunohistological study is trying to identify differences in the expression of proteins in vascular development in the lung in unaffected vs. affected babies. This is proving to be difficult research and they are continuing to evaluate the results.

She gave an overview of work that she and other researchers from the Children's Interstitial Lung Disease Clinical (ChILD) and Research Cooperative involved in to classify lung disorders, including ACD. ACD is categorized as one of the three Diffuse Development Disorders which also includes Acinar Dysplasia and Congenital Alveolar Dysplasia. The lung developmental stages are summarized in the table below.

| Lung Development Stage | Growth that Occurs | In utero period for humans |
|-------------------------------|---------------------------------------|-----------------------------------|
| Embryonic | Larynx, trachea, primordial lung buds | 3-6 weeks |
| Pseudoglandular | Airway development | 6-16 weeks |
| Canalicular | Capillary ingrowth period | 16-24 weeks |
| Terminal sac phase | Gas-exchanging surface area | 24-36 weeks |
| Alveolar | Alveolar development | 36 weeks through adulthood |

It is not yet understood when lung developmental problems begin. It could be the result of arrested development at a particular stage (ie normal development until then and then no further development) or possibly development is abnormal from the beginning and proceeds far more slowly.

Dr. Sen then presented a numerical accounting of the affected babies, parents and healthy siblings that are participating in the ACD studies that have been on-going at Baylor. Doctors and families from around the world have contacted Baylor via the ACDA, Baylor's posting on Gene-Test, and from doctor referrals. There are currently 47 families in the study. As was mentioned in the opening letter, additional family participation is needed to increase the sample size to improve the odds of finding the cause of ACD. The lack of enough subjects was a major reason that the National Institutes of Health denied funding for a recent Baylor proposal. Dr. Sen outlined the areas of ACD research that, if given the resources, they are prepared to support, including:

- Establish a worldwide repository for biomaterials and genetic material
- Genotype all ACD samples
- Collect all ACD samples
- Test more proteins for the Immunohistological Study
- Conduct fine mapping of potential candidates
- Conduct a microarray study of gene expression
- Study knock out animal models
- Perform a descriptive ultrastructure study

Craig Snyder was then given the floor and he stated that the goal of the 3 Angels Memorial Fund was twofold: (1) to raise money for ACD research and (2) to raise awareness in the medical community. Craig shared the following information:

- At least \$200,000 should be available from private donations to support ACD research in 2008.
- The Congress of the United States has just appropriated \$250,000 for fiscal year 2008 for ACD related activities, to be managed by the Centers for Disease Control (CDC). This is an enormous step for ACD as it is the first recognition by the federal government as a matter of public health and a research area worthy of being seeded with taxpayer money.
- The foundation is working with a publicist to raise awareness of ACD.
- Craig requested that Baylor submit a proposal in early 2008 outlining the resources they need to conduct the above mentioned research.
- The Foundation anticipates a grants for ACD research after review of the proposal.
- The 3 Angels website has been launched. Please visit <http://3angelsfund.org/> for more information.

The ACDA is extremely grateful to Craig and all of the other attendees for initiating and supporting this gathering and working together to find the cause and cure for ACD.



My Little Peanut ■'s Story

I had been in labor for probably a week before I went to the hospital. It wasn't pain, just pressure...until that last day. By 9:30, Friday night, December 3, 2004, I couldn't take it. At 11:30, the nurse came in and looked at the fetal monitor. She left the room pretty quick and came back, put me on oxygen and informed me that they were going to have to do an emergency C-Section. I was terrified. ■'s heart rate had dropped to 60 during the last two contractions and I was only dilated to 2. They didn't want to take any chances, since I still had a long way to go. I had ■ at 2:00 a.m. on Saturday morning.

The nurses brought her to my room around 11 that day. I was so drugged up, but still held her and tried to feed her. She wouldn't eat. I wasn't sure if that was because I wasn't doing it right, or what. I have 2 nieces, one 5 and one 2, and helped with both of them as babies. So, it wasn't like I'd never fed a newborn before. Then I figured that maybe I was just too drugged up. So, they took her back to the nursery and the nurse there tried to feed her and had the same problem. She gagged and just wouldn't eat. She switched her formula and that helped. I guess they thought that was the problem.

The next day, Sunday, I called the nursery around 10:30am to have them bring ■ to me. It rolled around noon and they still hadn't. I assumed that they were just really busy. It was time for pain killers anyway, so I called down and told them to not bring her and I would call again when I woke up. The nurse who answered told me that another nurse was on her way to talk to me. It wasn't a good conversation. She came in and said that ■ had turned dusky(bluish) in the nursery and they wanted to do a spinal tap on her. I started bawling. Here, my brand new baby, and a "procedure" already. Of course, I allowed it. They admitted her to the NICU and did the spinal. The results came back clear. I was so happy.

The doctors ended up diagnosing her with pneumonia. They said this was probably a result of my c-section. They needed to keep her in the NICU to continue her antibiotics but as soon as she was finished with those, she would be able to come home. The first time I walked in the NICU and saw my precious baby in the incubator with an oxygen hood over her whole face, was one of the worst sights of my life. It took a few times before I could handle it. They had her on oxygen for a few days and that was it. She seemed to be fine.

I went home on December 7th and they kept ■ until December 15th, which was when she finished her antibiotics and she came home. I was so excited to finally have her home. I found out after she came home that they had done an echocardiogram on her in the NICU. This was an ultrasound of her heart and lungs. They found the pressures in her lungs to be in the 50's. The normal pressure should have been from 15-20, but because of her pneumonia, they weren't too worried.

I had to take her to the cardiologist for a follow-up about a week and a half after she came home. I took her on January 4th and her pressures were still high. Her cardiologist called

me and said that he wanted me to get her to Riley Hospital in Indianapolis, IN as soon as possible. That in itself scared me to death. On January 7th, I took her to Riley Hospital for the first time. The cardiologist, Dr. Ebenroth, decided to do another Echo on her to check her lungs for himself. He backed up exactly what her cardiologist said. Then came the bad news. He then told me that she might have PPH (Primary Pulmonary Hypertension). It is a lung disorder, which causes high pressures, but is also not curable. So, I said, "Basically you're telling me that if this is what she has, that I have to just sit back and wait for my baby to die?" He answered with the most horrifying answer possible. "YES". I could have dropped right there, but I wasn't giving up without a fight.

They admitted her to the heart unit that day and put her on oxygen to see if that might help. The next day, they did another Echo on her and no change in her pressures, but I do think that the oxygen was helping her breathe.

I hadn't actually realized how much of a problem she had breathing until then. I started thinking about how she was when she was home. She would gag during almost every feeding. At times, she would vomit up her whole bottle. I would feed her an ounce and while burping her, she would fall asleep. I would have to wake her up, after almost every ounce. That stopped in the beginning of her being on the oxygen. She actually started eating and sleeping better.

I took her home on the 11th of January and had her home for another week and a half. They had me schedule her for a cardiac catheter on January 20th. This was a test where they would knock her out, intubate her and then put a catheter straight into her lungs. They would test her lungs without anything special. Then they would test her on 100% oxygen, and then add Nitric Oxide to the oxygen and see if her lungs react to that. If they would open up at all. If they do react then they know she can be helped with a medication. My fingers were crossed during the whole procedure.

Well, her lungs did react, but not as much as they would have liked. A little was better than nothing. They had decided to keep her for 24 hours for observation. That night, Thursday, she was having problems. I fed her and within 20 minutes, her oxygen saturation, which should be above 90, was dropping into the 70's. About an hour after she ate, it came back up. This continued to happen all that night and the next day. The doctors just decided to stop feeding her. They put in a feeding tube and wanted to see if that would stop the de-sating. They also decided that in a week they were going to try and do a lung biopsy to see if they could figure out what was wrong with her. They wanted to wait a week just to give her time to rest after her cardiac catheter.

On Saturday morning, January 22nd, around 7:00AM, I got a, not so nice, wake up call. They were rushing ■■■ to the ICU. They couldn't get her oxygen to stay up. I was so terrified. They got her there and told me that they were going to have to intubate her to give her higher amounts of oxygen then she could get through the nasal cannula. They also told me that they were going to try and put in a central line. ■■■ was so sick. Was my baby going to make it through this?

While Michele (my friend who went with me to Riley) and I were in the waiting room, a doctor came in to tell me that ■■■ was doing worse than when I left her. I asked him if she was going to die and he said that it was possible. I lost it. I just couldn't understand what was going on.

They had no idea what was wrong with her and she might die. I didn't know how to deal with that. So, I cried for the next half hour, until the doc came back in and said she was stable. I gave him the biggest hug...probably scared the poor guy. He was an intern. I got to go see her shortly after that. The saddest sight in the entire world. Blood everywhere and my poor baby laying there with tubes coming out all over.

Then, I found out that they were going to have to put a chest tube in because when they were losing her, they had to "bag" her. They had to bag her so hard that they blew a hole in her right lung. I was starting to get numb. I was so scared again. They got the chest tube in with few complications. ■ just didn't want to be messed with. That actually became somewhat of a joke between me and the staff. ■ was always, for the most part, fine until someone "messed with her". I used to say, just like her mother.

The rest of the day Saturday and then on Sunday, were hard. She was up and down all night and the next day. By Sunday night, they decided that there was nothing else they could do for her. They had given her every medication possible and nothing would keep her stable for long. What was I supposed to do? Her wonderful doctor, Dr. Nitu, then decided that she wanted to try her on steroids. She said that if they would have had the chance to do the biopsy, this was one thing they would have probably put her on anyway, so why not try it now. She said it couldn't hurt. And, she was right. Within an hour after her first dose, she was stable.

Then, they came to me with the idea that they need to do the lung biopsy now. They didn't want to wait. They didn't think she could make it too much longer and they knew that only a biopsy was going to give them any information into what was wrong with her. It was so hard to make the decision to do that because they warned me that she could die during the operation. As scared as I was, I had to let them. I then decided to put ■'s life, 100%, in God's hands. It was the weirdest thing, but as soon as I gave God the control, I was fine. I knew in my heart that she was going to come out of the surgery fine. I didn't even cry when I left her room. I was fine the whole two hours she was gone. The doc came to the waiting room to tell me she came through it. I hugged him and told him thank you, but I knew she was going to be fine. He just smiled.

Well, as soon as she got back to her room, the problems started again. They couldn't keep her oxygen level up. About three hours after she came out of surgery, another doctor came to me to tell me that he wanted to put her on the ECMO machine. Another, oh so scary, decision to make, but an easier one than the biopsy. The doctor told me that if I didn't agree to put her on ECMO, she probably wouldn't live another 2 hours. That was all I needed to hear. I couldn't just sit there and watch her die. Her life was still in God's hands and if it was her time to go, then at least I knew I did everything I could for her. When I left her room, right before they put her on the ECMO, her oxygen sats were in the 50's...she was purple. I wasn't as calm this time, but still not as bad as I would think. Again, she came through. What a fighter ■ was.

■'s biopsy results back the next day and her Doctor came in the room, so excited. She informed me that ■ was diagnosed with PPHN (Persistent Pulmonary Hypertension of the Newborn), which was curable. I just couldn't believe it. I have never wanted to scream for joy, so loud, in my life. I almost knocked Dr. Nitu over, jumping up to hug her. She stayed on the ECMO machine for 6 days. That whole week was wonderful. All I could do was think

about all the things that I was going to get to do with my baby, that for a short time, I didn't think possible. I started keeping a journal. I wanted to have everything written down, so when [REDACTED] got older, I could show her how strong she was and all the nasty things she fought through. She did so well. The day they took her off the ECMO machine, her lung pressures had gone down into the 40's...they had actually gone over 130, which is very bad. She had to have felt like she was suffocating. Poor baby.

Then, the day after she came off of the ECMO machine, Dr. Nitu came in with this horrid look on her face. She proceeded to tell me that [REDACTED]'s initial biopsy results were wrong. She doesn't have PPHN, she has ACD(Alveolar Capillary Dysplasia). But, this one isn't curable. I stared at her with the most intense feeling of numbness that I've ever experienced. How could that be? How could they have been wrong about something like this?

I decided to do some reading on ACD and found out that it is a so very rare lung disease. As of now, there are only 116 diagnosed cases in the ENTIRE WORLD. How is this possible? Why my baby? What are the chances? All the questions going through my head. They aren't positive, but they believe that her disease is genetic. She had a 25% chance of receiving both abnormal genes from both her father and I, and she did. I felt so guilty.

Here, I went a week believing that she was going to be fine, and then get smacked in the face with this news. I wanted to hit something. I just kept telling myself and the doctors that God was going to work a miracle. [REDACTED] was going to be fine. God brought her this far. He's not going to take her away from me now. I felt that way until the day she passed away.

Almost 2 weeks after she had come off of the ECMO machine, she started having problems. Two days before she passed away, her lung pressures were over 100 and the day she passed, her pressures were over 125. I knew that there was nothing else I could do for her...except to stop her suffering. It was another hard decision, but one I knew I had to make, if I love my baby as much as I knew I did. It was again in God's hands and knew that is she was meant to live, he would bring her through this too. At 9:30pm on Monday February 14th, 2005, they took [REDACTED] off of her ventilator. 20 minutes later, she passed away in my arms. I was lucky enough to have close friends and family there with me, but nothing could ease the pain I was feeling. I will always love my precious [REDACTED] and know that someday I will see her in Heaven. Without tubes and the happy baby she should have been here on earth.

I wrote this story for [REDACTED]'s website about a month after she passed away. Every time I read it, it feels like someone else's story. Her 3rd Birthday is in two days and right around the corner, it will be three years since she's been gone. There are days where I still sit back and ask, why me? Other days my faith gets me through helping me to see that there is a reason for everything. The pain is still there but it's just not as raw as it was three years ago. [REDACTED] will forever be Mommy's Little Peanut and never forgotten but time does heal. Even if it's just a little at a time.





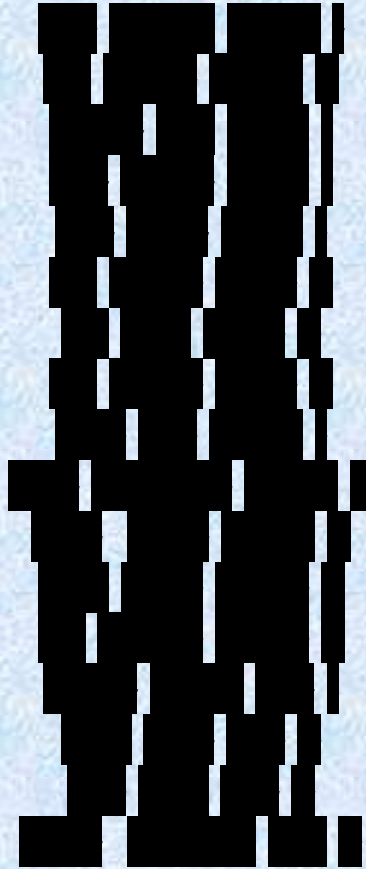
Acknowledgements

The ACDA would like to acknowledge some people for their recent efforts to support the ACDA. As you will read, there are many ways to support the ACDA and its effort to find the cause and cure for ACD.

- The ACDA recently received a very generous donation from an anonymous donor to support expenses of the organization. Some of the money was spent for travel expenses for the 3 Angels Memorial Fund meeting at Baylor Houston in November.
- Brian Robinson, nephew of Steve and Donna Hanson, updated the ACD website to include a new navigation bar, an updated Guestbook, electronic registration and updated newsletters and journal articles. Many thanks to Brian for all the hours he spent developing the new look and updated content!
- [REDACTED] and her family have printed a new ACDA brochure and letterhead. Thanks to [REDACTED], her brother [REDACTED] and the employees of [REDACTED] for their support.
- [REDACTED] and [REDACTED] of New York held their second annual golf outing in memory of their son, Ryan and raised over \$3,500 for ACD research – more than twice what they raised in their inaugural event in 2006.
- If you live in the state of New York (or you know someone who does) and need a mortgage, then contact Albany Funding. Albany Funding is owned by ACDA member Lisa Durand. Every quarter, they donate a portion of their profits to charity. For the last quarter, the Down's Association received over \$2,000. Albany Funding has selected the ACDA as the recipient for the quarter ending December 31, 2007. As you recall, Lisa and her family have been on-going contributors to the ACDA through her various businesses. If you have mortgage needs, please contact Albany Funding at <http://www.albanyfunding.com/>. The ACDA is grateful for the Durand's entrepreneurial endeavors on behalf of ACD research.

Memorial Garden

We are dedicated to remembering the birth dates of our families' babies who are not here to share our lives. Please pause to remember them.



Please let us know if we have inadvertently omitted your baby's name or if you do not wish to have your baby's name included in this section. You can email us at sdesj@verizon.net.

****Announcements****

- Lisa and Harry Durand have cookbooks available for purchase. Many of you remember that they developed the cookbook, *Recipes from the Heart*, in memory of their daughter, Olivia, and to raise money for ACD research. We can attest that these make special gifts at the holidays for friends, family, teachers and other special people in your life. The cookbooks are \$8.00 and you can purchase them by contacting Lisa at casper119@aol.com.
- Please remember to send a change of address (snail mail or e-mail) to us. We have recently lost touch with six families and want to be sure that we keep up with everyone, particularly as ACD research continues. You may be able to help and you would certainly want to hear about any new breakthroughs that could determine if your healthy children might be affected.

Surviving the Holidays

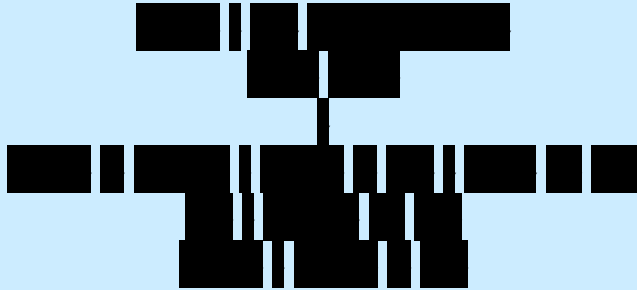
The holidays are a difficult time for all of us as we grieve for our baby who is not there to share in the holiday season. You may wrestle with mixed emotions but set aside some time that accommodate your feelings - even if it means some time in private. And accept that this may be a bittersweet time. Here are a few suggestions on surviving this bittersweet time:

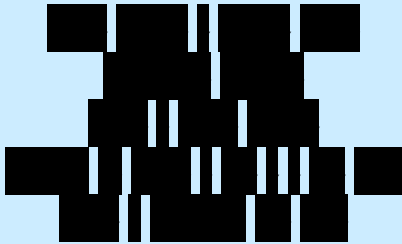
- Buy a beautiful Christmas candle and light it each day through December. It can be a daily, silent reminder of your baby and, maybe, make the whole house seem just a little bit warmer.
- Plan your shopping trips ahead of time or try creative alternatives. Try shopping at a different mall and go early in the day, before bumping into all those merry shoppers.
- Enlist the help of others in wrapping your gifts. Many friends would love to help but need to be asked.
- Consider changing your traditional family Christmas letter. Feel free to mention your loss. You may feel good sharing your feelings and others may benefit from hearing it as a reminder of the "true meaning of Christmas."
- Give yourself permission to change family traditions. Doing things differently this year may help to ward off some bad memories or expectations.
- Try a new idea when you are getting ready to hang up Christmas stockings. Go ahead and hang one up for your baby. Encourage each family member to write a note to the baby that can be read on Christmas Eve. A flower placed in the stocking for Christmas morning is a sweet sight.
- Determine to do one special thing for someone else in December. Maybe you know someone who is hurting like you or a lonely neighbor who could use a short visit. Perhaps you could make cookies to give away, pick out a special card to send or take a plant to someone.
- Contribute to your favorite charity in your baby's name.
- Pamper yourself this month. Be determined to buy yourself a present.
- Plan on taking time to cry. It is normal to have to cry during the holidays. Holidays intensify all emotions-why should your loss be any different?

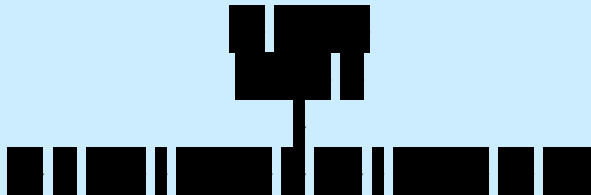
*From "Getting Past Christmas"
by Carlene Vester Eneroth*

WELCOME TO OUR
NEW FAMILIES

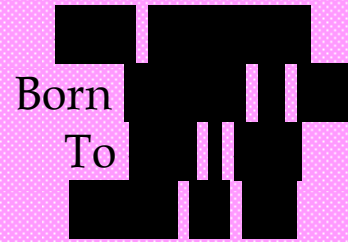
Please reach out to our families that have recently
contacted the ACDA:







Safe Arrival



Congratulations!



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