Fall 1996

CHERUBS

The
Association
of
Congenital
Diaphragmatic
Hernia
Research,
Advocacy,
and
Support

Dear Members,

I hope this newsletter finds you all doing well. Since our summer newsletter, we have a few new members and have started our Medical Reference Library. I wish we could offer copies of research articles for free, but we have to cover copy and postage costs (we still don't have a copy machine so we have to travel to make copies). I hope in the future donations will cover these costs but I know the price we are charging is much less than the price of driving to your nearest medical libraries, spending hours searching, and paying for copies. Included with this newsletter you will find a current list of the journal articles we have available. As our library grows we will update this list. If any of you have articles out there that we do not have listed and want copies of articles that we do have, we will be happy to make a trade; for each new article you add to our library we will send you a free copy of an article we do have. I hope all of you will take advantage of our library!! Also in this newsletter you will find an advertisement for Exceptional Parent Magazine. As a fund-raiser for CHERUBS, Exceptional Parent has agreed to donate proceeds of subscriptions to their magazine. Many of you have probably never heard of this magazine, as it is not sold on newsstands. It is a great resource for parents of children of all disabilities and lists therapy aids and medical equipment companies. If any of you have questions about this magazine or wish a trial copy please give me a call. We do have to charge $3.00 for a free issue of Exceptional Parent Magazine to cover the cost of postage.

I would like to say thank you to our members who are spreading the word about CHERUBS on the internet. Hopefully someday soon we will have our own page on-line. Also thank you to those members who have sent in their surveys. As I said in an earlier issue of our newsletter, I want to print our survey results in our Spring, 1997 newsletter. I hope we have enough surveys then to report results. I wish we had more pictures and stories of CHERUBS in this newsletter, but we just haven't received that many this quarter. We have a new address!! I love getting CHERUBS' mail at home and it certainly is more convenient but we needed a more professional and permanent address. My family is moving soon and we will also have a new phone number. I will send out our new number soon, if it is not included in our update page. Our ultimate goal is to have a "1-800 number". As promised in our last newsletter, here's the update from our trip to the American Pediatric Surgical Association's 1996 Annual Meeting and the summaries of presentations made at the meeting by pediatric surgeons. Welcome to all our new members and I hope you all have wonderful Thanksgivings!

Sincerely,
Dawn

CHERUBS Would Like To Thank The Following For Their Help and Support:

Association of Birth Defect Children, Inc.
Mrs. Wendy Barkley
Butner-Creedmoor News (Creedmoor, North Carolina)
Exceptional Parent Magazine
Michael Harrison, MD
Mrs. Brenda Slavin
Lesli Ann Taylor, MD
Claudine P. Torfs, PhD
Mr. Jeremy D. Torrence

CHERUBS Would Like To Thank The Following For Their Generous Donations:

Julianne Brian
Exceptional Parent Magazine
Michael R. Harrison, MD
David W. Kays, MD
Kevin and Brenda Lane, in honor of their daughter, Alysha Lane
Measurement, Inc. Printing Co.
Midwest Pediatric Surgical Associates, P.C.
The North Carolina Assistive Technology Program
Robert and Corinne Porter, in honor of their son, Max Robert Porter
Lesli A. Taylor, MD
Mary Von Tungeln
The University of North Carolina Holiday Card Project Fund
The University of North Carolina Health Sciences Library

CHERUBS Wish List

* A laser jet computer printer
* A computer scanner
* A fax/modem system
* A copy machine capable of making 11" x 17" dual-sided copies
* An office phone
* Computer software and clip art
* A telephone answering machine
* Computer paper
* Envelopes, all sizes
* Postage stamps, 19 and 32 cents
* 11" x 17" light colored paper

MEET OUR BOARD MEMBERS

Dawn M. Torrence- President and Founder
Granville Co., N.C. Parent Advisor for Disabled Children

Lesli A. Taylor, MD- Vice-President
Assistant Professor of Surgery and Pediatrics
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Jeremy D. Torrence Board Member
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Assistant Professor of Surgery, Harvard Medical School
Associate in Surgery; Director, ECMO Program
Boston's Children's Hospital, Massachusetts

Beth Zimmerman, RN, MS- Medical Advisor
Clinical Nurse Specialist
Section of Pediatric Surgery
Wyler Children's Hospital, University of Chicago, Illinois

Claudine P. Torfs, PhD- Honorary Medical Advisor
Epidemiologist
California Birth Defects Monitoring Program

Wendy Barkley- Honorary Medical Advisor
Parent and Registered Dietician
Lawton, Oklahoma

CHERUBS is an international organization for families and care-givers of children and adults who are diagnosed with Congenital Diaphragmatic Hernia (CDH). CHERUBS provides information, support, parent-to-parent matches, a congenital diaphragmatic hernia research survey, and medical research. There is no cost to parents for services provided by CHERUBS. We are volunteer organization founded in February of 1995 and an Internal Revenue Service recognized Non-Profit Association. Donations are very welcomed and tax-deductible. The livlihood of CHERUBS depends on donations from our professional members, families, and the public. Checks can be made out to CHERUBS. The opinions shared in this newsletter do not necessarily represent the views and opinions of all members or staff. The information in this newsletter is by no means to be substituted for proper medical advice. Remember, every child is different. You can not compare the progress of another CDH child to the progress of your own child. They are all little angels.....CHERUBS.

Letters to CHERUBS

March 26, 1996

Dear Ms. Torrence:

Thank you so much for your kind letter and copy of your informative newsletter. I am excited about the organization and its role. I feel that the information, services and support you provide to your families and to the community are invaluable.

I am very interested in assisting CHERUBS as a volunteer medical advisor and look forward to obtaining furthur information from you relative to my future involvement in this wonderful organization.

Very truly yours,
Aviva L. Katz, MD
Pediatric Surgeon
Alfred I. du Pont Institute

July, 1996

Dear Dawn,

Thank you for your recent letter. Enclosed is my membership fee and a small donation. Please keep me informed of your activities. I will publishing our data very soon, and I will send you an advanced copy of the manuscript.

Congradulations on the success of your efforts.

Sincerely yours,
David W. Kays, MD
Assistant Professor of Surgery
Director, ECMO Program
University of Florida,
Shands Hospital

June 9, 1996

Dear Dawn,

I am a speech-language pathologist at Children's Hospital & Medical Center in Seattle Washington. I had never heard of CDH until my very good friend gave birth to her firstborn last summer; this sweet little girl was born with CDH. Since then, I have been fortunate to be part of her life and consequent struggles and triumphs. I'll let her mom tell her story, but I am very interested in CHERUBS. I read your last two newsletters at my friend's home, and was very impacted by them.

Enclosed is $25.00 which I hope will cover the cost of mailing your newsletters to me, and perhaps a bit extra. I am a full-time PhD student now, but hope to finish soon, and would love to contribute more to your wonderful and necassary association.

Please let me know if I need to pay more to receive your newsletters. Keep up the great work. My life has changed as a result of my association with little Hayley; I am anxious to stay informed and do what I can to be helpful.

Best of luck and blessings to you.

Very Truly Yours,
Julianne Brian,MA, CCC-SpL
Speech language pathologist
Children's Medical Center

July 22, 1996

Dear Dawn,

I think it is wonderful that you are starting a research library for families who have been touched by a child with a congenital diaphragmatic hernia. Please find enclosed some materials which I hope can help your efforts. If I can be of any assistance in the future please do not hesitate to contact me or my nurse coordinator Jody Farrell.

Sincerely,
Michael R. Harrison, MD
Professor of Surgery and Pediatrics
Director, Fetal Treatment Center
University of California,
San Francisco
June 9, 1996

New Members and Updated Pictures

Kaylee Lyne Bentz
June 20, 1994
Sioux Falls, South Dakota

Naomi Nagurski
July 22, 1995
Alburquerque, New Mexico

Jeremy "Shane" Torrence
January 28, 1993
Franklinton, North Carolina

Letters from Members

July, 1996

Dawn,

Reading the Cherubs newsletter was like traveling back through a time warp, and was a bit disturbing recalling all we had been through and tried to forget. It is great to know that if this awful thing happens now, there is support out there.

Our daughter Angela is now 12 years old, and is a beautiful survivor of CDH. She is exactly where she should be developmentally. She has earned a trophy and 4 blue ribbons for her piano playing, plays sax in the school band, and takes ballet lessons. She does well acedemically, carrying a B average. When she was born she had almost no diaphragm, she was given an artificial on the day she was born. She needed a central line and gastrostomy tube for feedings. She had multiple surgeries, pnuemonia, emphysema (from the ventilator), etc. during her first three month stay at the hospital. For the next two years she ate totally by G-tube, had frequent hospital stays, therapies, surgeries, etc.. Feeding therapy did not work, but at age 2 and a half she decided to eat on her own, and ate Kentucky Fried Chicken!! Several months later she was eating well enough to have her tube removed, and has eaten well since then. She is unable to burp, has many huge scars, and is quite small for her age, but those are the only physical problems she continues to deal with. She handles the burping problem by eating small frequent meals, and avoids carbonated drinks. The scars do not bother her, in fact she sometimes jokes about them (she has a great sense of humor). Her size has worked out great for ballet, and piano does not require a person to be a certain size to play. We were surprised that she took up the sax with her lung problem. She can only play a few songs before getting winded, but continues to practice and perform. The biggest hurdle we now face is the emotional scars that her life has left on her. She has a family that loves her, including her dad (Jim), brother Jimmy (14 years old with Autism), her mom (Roxane), and many loving friends and relatives, who are doing all they can to support her whenever she needs it.

Angela Barricklow
February 27, 1984

God bless you and the work you are doing! It is very important and much needed.

Roxane Barricklow
3011 Snow Hill Road
West Harrison, Indiana 47060

June 3, 1996

Dear Dawn,

Thank you so much for your response and the parent matches. I plan to contact many. I want to thank you also for the time and effort you put into helping others. It is really nice to talk to someone who has "been there". I must admit I am really scared! I am now 29 weeks. My doctor plans to do another amniocentesis test at 36 and a half weeks to test for lung maturity. If lung maturity is thought to be, then they will send me to Gainesville, Florida at Shands Hospital to deliver two weeks early at 38 weeks. I am not sure how I will deliver. One of my doctors says it would be best to deliver by cesarean and the other doctor says an induced vaginal delivery would be fine. I guess they will get together when I travel to them at 38 weeks and we will all decide together. I really don't care, I just want what ever is best for my baby. At this point and time the only thing we know is that my baby has a left-sided CDH, and his heart is displaced to the right. We also know that his stomach is in the chest. My doctor says if the stomach is up there then lots of bowels are up there with it. He also says there is no way to see how much lung has grown on either side because of the compression on both lungs. So far I do not have polyhydramnios!!! I go back to my doctor this week for another ultrasound. Hopefully they will say I still don't have it. I pray to the good lord above that this baby will be okay. I love this child more than I can ever express in words. I know this baby has a hard road ahead of him. I pray I have made the right decision to have him. I feel we made the right decision. I couldn't dismiss this child just because he was not perfect. This is our first child. My husband and I waited nine years to start our family. My doctors say this baby has a chance to live, I feel I have got to give him that chance. I am really scared of ECMO. I pray he will not need it. I have heard and read too many scary things about it. I know ECMO saves lives but it also damages babies sometimes too.

I thank God for people out there such as yourself that reach out to others the way you do. I will keep in touch with you and let you know how things are going. Thank you so much for your kind words and thoughts. God bless you and your husband, and your beautiful little cherub, Shane.

Sincerely,
Suzanne E. Robinson
4334 Wycliff Dr.
Pensacola, Florida 32514

Dear Dawn: July 19, 1996

The only way I can start this letter is to say God Bless you for the work you are doing. You spoke to my husband Bob back in June when we first found out our baby boy has right sided CDH. At that point when he spoke to you, we had only known for about 3 days (if I'm remembering correctly!). As you can imagine, we were terrified, confused, UPSET, and really did not know what we were facing. We had managed to get some information from the University of Washington medical library, and well as from the Children's Hospital in Seattle. Unfortunately, all the articles we first read scared us even more as they were above our understanding, and the first line in virtually all of them was that the mortality rate for these babies was great. When we received the copies of the newsletters you sent us, we were comforted by the stories of all of the cherubs who pulled through. However, to be honest, it was a shock to learn of the additional problems that CDH can cause. We quickly realized that the road ahead for all 3 of us was not going to be easy!

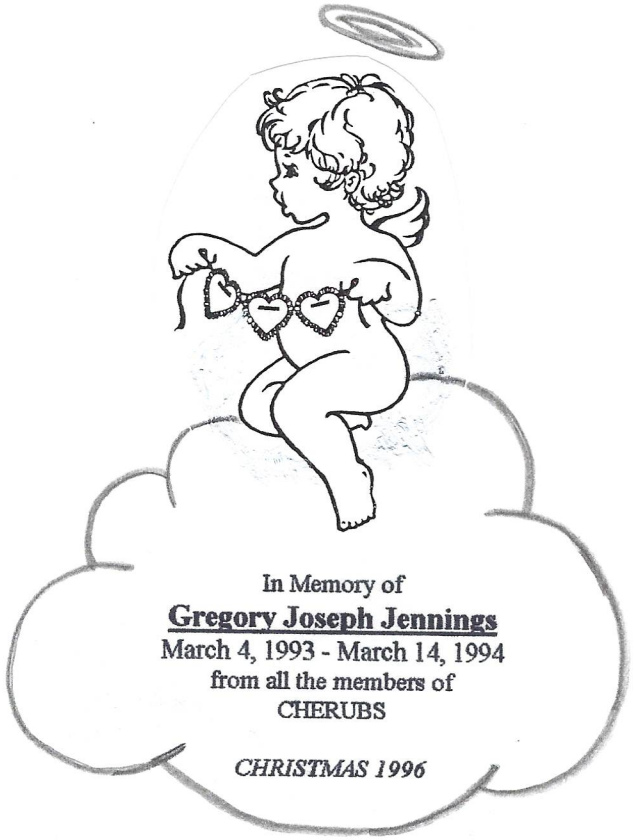
I would like to tell you a little about Bob and I. We live in Kent, Washington, which is a few miles south of Seattle. We are newlyweds, just having been married August 12, 1995. The pregnancy was kind of a surprise, but we were thrilled because we knew even before we were married that we both wanted children. The early pregnancy was uneventful, I was blessed with an easy time. I had only a touch of morning sickness and that did not even last very long. We had our fist ultrasound on May 15th. During the ultrasound, the technician was able to tell us it was a boy and we were so in awe of seeing him move around! However, towards the end, she did call in the doctor on duty to look at our baby. We were scared, but they explained to us that because of the baby's position, there were a few measurements they were unable to get. We made arrangements to come back on Monday the 20th for a quick ultrasound before my regular monthly doctor's appointment. That morning, the news was not so great. They were able to see that the heart was slightly displaced, but they could not tell exactly why. We scheduled a follow up ultrasound for 4 weeks later. We began with an ultrasound at the office we had been going to previously. After scanning for about an hour, they sent us to their main hospital in Seattle. We were scanned for about 2 hours there by 2 doctors. Later that day, we were back at my regular OB's office, where we finally heard the diagnosis . It was as if we ran smack into a cement wall, it took the breath right out of us. As the doctor was describing what she knew about the condition, I felt like she was talking in some foreign language and I just couldn't seem to grasp what she was saying. Her suggestion was that we speak to the team at UCSF. She sent them a video of an ultrasound and they contacted her with the news that we would not be candidates for the actual repair surgery as the liver was herniated into the chest. They explained to her that we would be eligible for the tracheal occlusion. We flew to San Francisco for evaluation and to meet with the team. We left San Francisco with so much information regarding care of the baby when he is born (if no intervention in utero is done) and about the in uterine surgery, that we were simply overwhelmed. After spending all weekend deep in prayer and discussion between the two of us, we decided we will opt for the in uterine surgery. It was such a huge decision, but we feel God is guiding us and we will make it through. The surgery is scheduled for July 30th.

We will keep in touch and let you know how everything goes. I'm praying every second that we will have a miracle to share with you and the CHERUBS family. Thank you so much Dawn, for being an angel yourself and doing so much to reach out to all of us who must deal with this experience. We are enclosing a small donation to hopefully help in postage, paper, etc.. God Bless You and Your Cherub and Your Family!

Sincerely Yours,
Corinne and Bob Porter
26026 184th Ave. SE
Kent, Washington 98042

CHRISTMAS CLOUDS

In memorial of all the CHERUBS we've lost and in honor of all survivors, for the holidays this year we are going to sell "clouds" in our newsletter. This is way for family and friends to remember losses and triumphs. It is a way to include our real cherubs in Christmas and Chunnakah celebrations through remembrance. For parents, grandparents, family, friends, and even doctors (yes, parents, doctors do feel the losses too), this is a way to say to our cherubs, "We miss you, we love you, we remember you. You will always be in our hearts." For family and friends of survivors, this is a way to say; "We remember those children who did not survive and we remember the pain of those first months". For a minimum donation of $10.00, we will print a "cloud", such as the one to the right, suitable for decorating or coloring, cutting out and placing on the Christmas tree. There is no limit to the number of angels you can order. All requests must be submitted by November 7th. Just write down what you would like on a piece of paper and send it, alond with your check or money order, to our new address. You and your loved ones can purchase clouds for your child and for children you met on the way. I hope all of you will join us.



VAGINAL OR CEASERIAN DELIVERY FOR CDH BABIES?

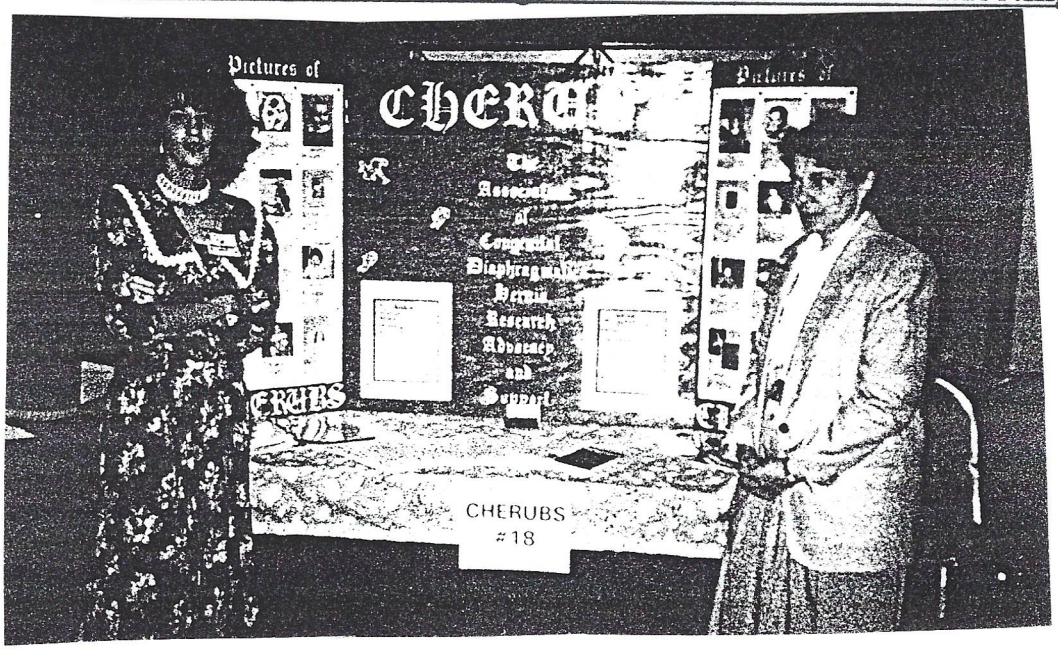
Quite a few of our expectant parents have asked whether it is best for babies with congenital diaphragmatic hernia to be born vaginally or be delivered by C-Section. Well we have yet to find a journal article on this subject so we decided to ask our medical advisors on their thoughts about this. Here is what a few of them said:

Dr. Jacob Langer; "There is no evidence that cesarian delivery is advantageous for babies with diaphragmatic hernias. There may be obstetric indications for caesarian section which are separate from the diaphragmatic hernia issue but in the absence of specific obstetric indications I always advise vaginal delivery. Most prenatally diagnosed babies with diaphragmatic hernias will be immediately intubated at the time of delivery so the use of pain medication for the mom is really irrelevant and that decision should not be made on the basis of whether there is a diaphragmatic hernia or not".

Dr. Vincent Adolph; "I don't think there is any evidence that vaginal or cesarean delivery directly affects the outcome in antenatally diagnosed CDH patients. Delivery in a center with Neonatal and Surgical expertise in CDH should be the absolute minimum and the neonatalogy team should attend the delivery. Many people believe such antenatally-diagnosed children should be delivered in a center where ECMO is available, but I don't know that there is any evidence that the outcome is better".

So all in all, the choice belongs to the patient and her obstetrician. If there are no other complications or reasons why the mother needs a C-section, then there is no reason why a vaginal delivery can not be the best choice if CDH is the only factor. Of course, listen to your own doctor, your own instincts, and the neonatal team.

The 1996 American Pediatric Surgical Association Annual Meeting



Dawn M. Torrence, President (on the left) and Lesli A. Taylor, MD, Vice-President at our booth at this year's A.P.S.A. meeting

Between May 19th and 22nd of this year my husband, Jeremy (one of CHERUBS' board members), and I attended the 27th annual meeting of the American Pediatric Surgical Association as representatives for CHERUBS. Also in attendence were Lesli A. Taylor, MD, our Vice-President and several of our medical advisors; Jacob C. Langer, MD, FRCS(C), Prem Puri, MS, FACS, and Jay Mark Wilson, MD. We met quite a few doctors and nurses (some members) and learned quite a lot about the newest research on congenital diaphragmatic hernia. I would like to say thank you to Dr. Jacob Langer and the executive board of the A.P.S.A. for allowing us to attend this very important and informative meeting and to American Airlines for donating airline tickets. We handed out quite a few Parent Reference Guides in the hopes that many more doctors will refer families to us. Here are the summaries of those reports on CDH and tracheal ligation as written in meetings guidebook:

FAMILIAL CASES

Familial Congenital Diaphragmatic Agenesis: An Autosomal Recessive Syndrome With A Poor Prognosis. David Gibbs, Henry Rice, Jody Farrell, N. Scott Adzick, Micheal Harrison; University of California, San Francisco.

Purpose: Congenital diaphragmatic hernia (CDH) can occur as an isolated defect or as part of a multiple anomaly syndrome. Typically this defect occurs in patients with no prior family history of CDH. Several hereditary syndromes such as Fryn's syndrome have been reported, and some authors have suggested that the agenesis of the hemidiaphragm may be associated with an autosomal recessive inheritance. Prenatally diagnosed CDH has been associated with a poorer prognosis likely related to an increased incidence of unilateral diaphragmatic agenesis. Our large referral base for prenatally diagnosed CDH includes 142 cases in the last three years. During this time we have managed six of these unusual families with a history of CDH and carrying a fetus with agenesis of the diaphragm.

Methods: Patients with prenatally diagnosed CDH and a previous history of CDH pregnancies were evaluated. Previous pregnancies were also evaluated by reviewing available medical records. Characteristics studied included extent of diaphragmatic defect, associated anomalies, fetal sex and chromosomes, and fetal outcome. Analysis by population statistics as described by Li, et al. was used to evaluate a possible autosomal recessive mode of inheritance. Spontaneous and therapeutic abortions for which no pathological information was available were excluded from analysis.

Results: Each of our six families had multiple pregnancies with CDH. Of a total of 28 pregnancies in these families, fouteen fetuses had a diaphragmatic hernia. One of our six CDH fetuses died in utero, one was terminated at the family's request, two died postnatally, one is alive, and one is in utero at the time of this report. Absence of the hemidiaphragm was determined ineach of our six cases by direct operative or post-mortem exam or inferred by the presence of massive liver herniation in the chest on ultrasound examination. Surprisingly, only one fetus had another significant anomaly (holoprosencephaly). Four of our six fetuses were female. Chromosome analysis was normal in each case. We calculated the corrected incidence of all CDH cases in these families. Although the expected incidence of an autosomal recessive trait is 25%, our corrected incidence was 32% (+/-7%).

Conclusion: Prenatally diagnosed familial CDH is a distinct clincal entity associated with isolated diaphragmatic agenesis and poor prognosis. Associated anomalies are uncommon in this group, but should be ruled out. An autosomal recessive mode of inheritance, while not statistically confirmed, was supported by our analysis. This information should be useful in counseling these families.

PRENATAL DIAGNOSIS

Prenatal Ultrasound Frequently Fails To Diagnose Congenital Diaphragmatic Hernia; Dorothy Lewis, Richard Bowerman, Ronald B. Hirschl, Department of Pediatric Surgery and Radiology, University of Michigan School of Medicine, Ann Arbor, Michigan

Purpose: Prenatal ultrasound (USN), which is performed routinely today, has resulted in the frequent in-utero diagnosis of congenital diaphragmatic hernia (CDH). However, in many patients the diagnosis is not made despite performance of a prenatal ultrasound. The purpose of this study was to determine the reason for missed ultrasound diagnosis of CDH.

Methods: The charts of CDH neonates treated at our instituuion between 1985 and 1995 were reviewed. Available prenatal ultrasounds were also reviewed by a radiologist and the following parameters were evaluated: cardiac deviation, absence of stomach in the left upper quadrant, and umbilical vein deviation. Ultrasounds performed prior to 17 weeks gestation were excluded because they were too early for thorough review. Possible reasons for failure to diagnose CDH were divided into 4 categories: technical difficulties; failre to follow established guidelines for prenatal exams; missed abnormalitites; and misdiagnosis of observed abnormalities.

Results: There were 127 neonates with CDH, who were symptomatic within the first 6 hours of life, treated at our institution between 1985 and 1995. Of these, 72 (56.7%) underwent prenatal ultraound, and in 30 (41.7%) the diagnosis of CDH was missed. In 17 of these 30 patients, a total of 31 ultrasounds (1 USN, n =6; 2 USN, n= 9; 3 USN, n = 1; 4 USN, n= 1) were available for review. Of the 31 studies, 6 were performed at a gestational age of less than or equal to 16 weeks. Of the remaing 25 exams, six were associated with technical difficulties (maternal obesity, chest overmagnification, suboptimal fetal position, and/or poor quality study). In 17 of the ultrasounds, the established guidelines for obstetrical ultrasonography were not followed, and this resulted in failure to identify the stomach in 10 and failure to fully visualize the 4 chambers of the heart in 14. Nine ultrasounds had abnormalities consistent with CDH which were not recognized; namely, cardia deviation in 9 and umbilical vein deviation in 1. A single ultrasound clearly demonstated a cardia sihouette in the right chest, but this was interpreted as dextrocardia. In only one study did a thorough examination fail to demonstrate any findings consistent with CDH.

Conclusion: These date demonstrate: 1) the diagnosis of CDH is missed at the time of prenatal ultrasound in more then 40% of patients, and 2) the most important factor which appears to contribute to the missed diagnosis is a failure to follow established guideline for prenatal ultrasound evaluation, primarily localization of the stomach as well as complete visualization of the heart.

Fetal Diaphragmatic Hernia: Echocardiography and Clinical Outcome; Karen VanderWall, Thomas Kohl, N. Scott Adzick, Michael Harrison, Norman H. Silverman; University of California, San Francisco

Purpose: Predicting which fetuses with prenatally diagnosed congenital diaphragmatic hernia (CDH) will survive after birth remains an enigma. Several studies suggest that left heart underdevelopment is associated with poor outcome, but detailed echocardiographic data on fetuses with CDH is lacking.

Methods: We retrospectively reviewed detailed fetal echocardiography studies in 30 patients with CDH who were referred to our center. In twelve fetuses (17-25 week gestational age) the study was adequate for careful measurement and analysis. These patients had follow-up to determine outcome after planned delivery and optimal postnatal care. Patients with other anomalies, and those who chose termination or fetal surgery were excluded. Echocardiographic variables were examined from a four chamber view and corrected for gestational age by comparing the variables to normal controls. The variables examined were: maximum length of the left and right ventricular chamber during diastole (LV Length, RV Length), maximum width of the left and right venticular chamber during diastole (LV Width, RV Width), left ventricular wall, right ventricular wall and septal thicknesses during diastole (LV Wall, RV Wall, VS). The data for survivors (n =6) was compared to non-survivors (n =6) and plotted on regression analysis curves consisting of normal controls at different gestational ages.

Results: There were no differnces between the groups in the 7 parameters analyzed. Although the LV Width and RV Wall were less in non-survivors compared to survivors, the differences were not significant when corrected for gestational age.

Conclusion: Prenatal echocardiographic evaluation of fetuses with CDH is important to exclude associated congenital heart defects. Although left heart underdevelopment may be evident in fetuses with CDH as a group, our findings suggest that there are no specific parameters on echocardiography that can be used to predict outcome after birth.

TRACEAL LIGATION

Tracheal Ligation Increases Cell Proliferation But Decreases Surfactant Protein in Fetal Murine Lungs In Vitro. Kelli Bullard, Johnathan Sonne, Samuel Hawgood, Michael Harrison, N. Scott Adzick, University of California, San Francisco

Purpose: Tracheal occlusion affects both fetal lung growth and maturation. We used a murine in vitro whole organ culture model to investigate these effects. We hypothesized that tracheal ligation would increases cell proliferation and would change surfactant protein expression.

Methods: Lungs were removed from 14 d. murine fetuses (term= 21 d.). Tracheas were ligated with 10-0 dermalon suture and explants cultured in serum free media (BGJb plus vitamin C and antibiotics) at 37 degrees Celcius, 21% oxygen, 5% carbon dioxide for 1, 3, 5, 7, or 14 days. DNA synthesis and cell division were assessed by incubating the explants with 5-Brumo-2'-deoxy-uridine (BrdU) labelling reagent for one hour prior to fixation. Immunohistochemistry was performed to detect BrdU [N = 62; 30 ligated, 32 unligated]; positive cells per 1000X field were counted. To detect surfactant protein, lungs were cultured 3, 4, 5, or 7 days. immunohistochemistry was performed to detect surfactant proteins A (SP-A) and B (SP-B) {N=45; 21 ligated, 24 unligated]. Nonimmune goat serum as a negative control. A Student's t-test was performed.

Results: Ligated lungs showed significantly more BrdU labelled cells/hpf at every time point. Ligated lungs at d.1 showed 27% more cells/hpf than unligated (17.1 vs. 13.5; $p < .05$), d.3 21% more (16.2 vs. 13.4; $p < .05$), d.5 54% more (19.1 vs. 12.4; $p < 00.1$), d.7 60% more (16.2 vs. 10.1; $p < .001$), and d.14 123% more (13.5 vs 6.1; $p < .001$). Unligated lungs consistently showed greater staining for surfactant proteins than did ligated lungs. SP-A was first detectable after three days in culture; at this time point unligated lungs showed 15% of airways positive while ligated lungs showed only 3% ($P = .15$). Differences were statistically significant after four days, with unligated lungs showing 59% of airways positive on d.4 and 71% positive on d.5, versus 33% on d.4 and 59% on d.5 in the ligated group ($p < .05$). The staining pattern for SP-B was similar. This protein was first detectable on d.4, with 15% of airways positive in the unligated group versus 8% in the ligated group ($p = .21$). After 5 days, unligated lungs showed 66% of airways positive versus 40% in the ligated group ($p < .01$). By d.7 all lungs showed 70 to 100% of airways positive for both surfactant proteins.

Conclusion: We conclude that tracheal ligation increases cell proliferation but decreases surfactant protein in murine fetal lungs maintained in serum free whole organ culture. Increased cell proliferation, indicated by increased BrdU uptake, is detectable as early as 24 hours after ligation; tracheal ligation decreases staining for surfactant proteins A and B, suggesting that ligation may delay lung maturation. This model provides a powerful tool for investigating the mechanisms that underlie fetal lung development and tracheal occlusion induced pulmonary hyperplasia.

Fetal Lung Growth After Tracheal Ligation Is Not Solely A Pressure Phenomenon. K. Papadakis, F. I. Luks, M. E. DePaepe, G. Peasecki, Conrad W. Wesselhoeft, Brown University of Medicine, Hasbro Children's and Rhode Island Hospitals, Providence, Rhode Island

Purpose: Fetal tracheal ligation increases lung growth in utero making it potentially applicable for antenatal treatment of diaphragmatic hernia. This phnomenon has been ascribed to increased intratracheal pressure which activates as yet unidentified pulmonary stretch receptors. The purpose of this study was to determine whether the composition of lung fluid has any effect on fetal lung development.
Methods: Five sets of fetal lamb twins underwent tracheal ligaiton with placement of intratracheal catheters at 122 d gestation (term = 145 d). In group I (n =5), tracheal fluid was aspirated daily and replaced with equal volumes of saline. Their respective twins (group 11, n =5) had daily re-infusion of their own tracheal aspirates. Unobstructed fetal lambs (n = 7) were used as negative controls. A Cesarean section was performed at 137 d (post-ligation day 15), and the fetuses were sacrificed. Their lungs were weighed, perfusion-fixed with 10% formaldehyde at 25 cm water and analyzed using standard morphometric methods. Single factor ANOVA and Wilcoxon rank sum test were used for statistical analysis where appropriate.
Results: In all 10 fetuses, tracheal ligation resulted in an almost threefold increase in lung fluid volume by day 1 (from 12.1 +/- 4.1 to 32.3 +\- 7.6 mL/kg birth weight); a slight decrease at a mean of 2.4 days (nadir 28.2 +/- 7.3 mL/kg); and a second surge from days 4 to 14 (range 28.2 +/- 7.3 mL/kg); and a second surge from days 4 to 14 (range 38.4 to 46.1 mL/kg). Mean lung fluid volume was higher in group II than in group I at all measured time-points ($p < 0.05$). Lung weight/body weight (LW/BW) at delivery was 0.046 =/- 0.008 in group I vs. 0.055 +/- 0.010 in group II ($p= 0.06$). In unobstructed lambs, LW/BW was 0.037 +/- 0.007.
Conclusion: Occlusion of the fetal trachea causes an initial decrease in lung fluid volume, followed at 72 hours by lung growth and/or increased fluid production. This lung growth is at least partially dependent on humoral factors present in lung fluid, since replacing this fluid with saline results in smaller lung weight and mean 'alveolar' surface. The immediate, as well as sustained, blunting of the fluid volume curve after saline replacement suggests that the factors present in lung fluid are not merely activated by an increase in intratracheal pressure, but play a primary role in stimulating pulmonary growth after tracheal ligation in the fetus.

Tracheal Ligation Improves Development Of Nitrofen Induced Hypoplastic Fetal Murine Lungs In Organ Culture. Robert E. Cilley, Steven E. Zgleszewski, Mala R. Chinoy, Thomas M. Krummel; Thr Pennsylvania State University, Milton S. Hershey Medical Center, Hershey, Pennsylvania

Purpose: To compare the development of Nitrofen (2,4-dichlorophenyl-p-nitrophenylether) induced hypoplastic lungs to normal fetal murine lungs in vitro.
Methods: Nitrofen (10 to 35 mg in 0.5 ml olive oil, per mouse) was given intragastrically to time-dated pregnant CD-1 mice on gestational days 8 and 9. At day 14 of gestation the mice were sacrificed and fetuses were removed. Lungs were excised and placed in organ culture (BGJb media at 37 degrees Celcius, 95% air, 5% carbon dioxide). Some lungs were cultured after tracheal ligation (10.0 nylon suture), while others were unligated, allowing free egress of fluid from the airway space. After 7 days in culture, lungs were fixed in 10% formalin, paraffin embedded and processed for routine H&E staining. Normal fetal lungs were cultured and processed similarly.
Results: Nitrofen exposed lungs were significantly smaller, in a dose dependent manner, as compared to normal lungs at gestational day 14. The H&E sections revealed that both lungs were in the pseudoglandular stage. After 7 days in culture the normal unligated lungs were underdeveloped, as were the hupoplastic lungs which had undifferentiated parenchyma. Ligated fetal lungs in culture, normal and nitrofen exposed, showed parallel patterns of development with structurally better developed lung architecture when compared to unligated lungs.
Conclusion: 1) Tracheal ligation accelerates lung development in normal and Nitrofen induced hupoplastic lungs in organ culture. (2) Nitrofen induced hypoplastic fetal lungs in organ culture show parallel development to normal whole lungs after tracheal ligation. (3) Our whole lung organ culture model may be useful in designing studies to reverse lung lypoplasia and delineating the mechanisms involved in normal lung development.

FETAL THERAPY

Prenatal Glucocorticoids Improve Pulmonary Morphometrics In Fetal Sheep With CDH: Holly L. Hedrick, Jody M. Kaban, Bella A. Pacheco, Paul D. Losty, Daneil P. Doody, Daniel P. Ryan, Thomas F. Manganaro, Patricia K. Donahoe, Jay J. Schnitzer; Pediatric Surgical Research Laboratories, Pediatric Surgical Services, Massachusetts General Hospital, Harvard Medical School, Boston, MA

Purpose: Prenatal glucocorticoids reverse pulmonary immaturity in a rodent model of congenital diaphragmatic hernia (CDH). We applied quantitative sterologic morphometric techniques to test whether these effects could be reproduced in a large animal model as a prelude to proceeding with human clinical trials.
Methods: CDH was created surgically in fetal lambs at gestational day 80. They were treated with intravenous cortisol (n = 6) or normal saline control (n = 5) from days 133-136. Lungs distended at 15 cm pressure from each group were harvested at day 136, processed histologically, and studied by brightfield microscopy at 400X using a 42-point equidistant counting grid. Ten morphometric parameters (Mean =/- SEM) were measured by point-counting 60 fields/lung, and analysis of variance was performed.
Results: Histologically, the CDH-cortisol treated lungs revealed striking maturational changes compared to CDH-normal saline controls . There were significant improvements in seven morphometric parameters. Also significant, was a reduction in volume fractionof ducts after cortisol treatment ($p = 0.0189$).
Conclusion: 1) Prenatal glucocorticoids accelerate lung maturity in fetal lambs with CDH by seven quantitative morphometric parameters. 2) The observation that prenatal glucocorticoid therapy improves measures of maturity for CDH rodent and sheep models supports proceeding with a Phase I human clinical trial in ultrasound confirmed CDH.

Operating on Placental Support: the Ex Utero Intrapartum Treatment (EXIT) Procedure. George Mychaliska, John Bealer, Joy Graf, Michael Harrison, University of California, San Francisco

Purpose: Airway obstruction at birth is life-threatening and in some casescan be predicted by diagnosis before birth. We have developed and refined a novel technique, Ex Utero Intrapartum Treatment (EXIT), for securing the airway in neonates with threatened airway obstruction at birth while they are maintained on placental support.
Methods: After developing this technique experimentally, we applied this stategy to six cases with predictable complete airway obstruction at birth. We have reviewed these cases noting prenatal diagnosis, fetal surgical procedures, maternal and fetal outcomes, gestational age at delivery, procedures performed and duration of the EXIT procedure.
Results: Five patients underwent in utero tracheal occlusion for congenital diaphragmatic hernia (CDH); 2 with an internal plug and 3 with an external clip. The sixth patient had a huge cystic hygroma of the neck, upper thorax, and floor of the mouth so that securing an airway rapidly at birth could be nearly impossible. The following factors proved crucial for the success of the EXIT procedure: 10 multidisciplinary team approach including pediatric surgeons, anesthesiologists, obstetricians, radiologists, and neonatologists, 2) accuate pre-operative assessment of the airway anatomy, 3) high doses of inhaled halogenated agents to provide uterine relaxation, 4) fetal monitoring with cardiac sonography and pulse oxymetry, and 5) airway evaluation with laryngoscopy and bronchoscopy. EXIT procedures included orotracheal intubation, tracheostomy, trachoplasty, reversal of tracheal occlusion by removal if internal plugs or external clips, and placement of a central venous line. In the CDH patients, lung fluid was aspirated and surfactant given prior to giving the first breath. The mean gestational age at delivery was 32.5 weeks (range 28.1-36.9). In all cases , the airway was secured in a controlled manner with the fetus maintained on placental circulation and there was no significant episodes of fetal bradycardia, hypoxia, or other untoward events. There were no maternal complications.
Conclusion: Our experience with in utero tracheal occlusion prompted us to develop the EXIT procedure to deal safely and effectively with completeor impending airway obstruction at birth. Accurate prenatal diagnosis, a multidisciplinary approach, intensive maternal-fetal monitoring and complete uternine relaxation with the maintenance of feto-placental circulation offers a safe and controlled approach tothe challenging problem of prenatally diagnosed airway obstruction.

Dear members, I came across this article and thought it would be beneficial to all our members, our doctors, nurses, bereived parents, and parents of children who have survived. For our bereived parents, this article can provide a guide for friends and family; for our other parents, this article will help you to better help those bereived families that you will come into contact with. For the parents of survivors, you will unfortunately meet many children who will not survive during your hospital stays. I encourage you to support befriend this parents just as you would if their children survived. Go to the child's funeral if you can, but ask the child's parents first if it would be too hard on them if you attended. Remember, you might represent to them all they have lost, this is a normal emotion that you should be sensitive to. Above all, for all parents, it is important not to abandon these friendships out of feelings of being uncomfortable. I hope we all can learn from this article:

How to Help Grieving People

by Alan D Wolfelt fron News from the Breavement Foundation Vol. 9, No. 2, May 1996
(taken from Pen-Parents of Canada Summer 1996 Newsletter)

Relatives, friends and neighbours are supportive at the time of death, during the wake and funeral. Food, flowers and their presence are among the many thoughtful expressions. After the funeral, many grieving people wonder what happened to their friends. They need their support and caring even more when the reality begins to hit and the long process of grief begins. Their help is essential since immediate family members have their hands full with their own grief and may find it difficult ot give support to one another, or may not live nearby. your help and understanding can make a significant difference in the healing of your friend's grief. A grieving person needs friends who are willing to: LISTEN; cry with them; sit with them; reminisce; care; have creative ideas for coping; be honest; help them feel loved and needed; believe that they will make it through their grief. Ways of helping grieving people are as limitless as your imagination.

How can you help?

A friend had experienced the death of someone loved. You want to help, but you are not sure how to go about it. Hopefully the following will guide you in ways to turn your cares and concerns into positive actions.

Listen With Your Heart

Helping begins with your ability to be an active listener. Your physical presence and desire to listen without judging are critical helping tools. Don't worry so much about what you will say. Just concentrate on listening to the words that are being shared with you. Your friend may relate the same story about the death over and over again. Listen attentively each time. Realize this repetition is part of your friend's healing process. Simply listen and understand.

Be Compassionate

Give your friend permission to express his or her feelings without fear of criticism. Learn from your friend; don't instruct or set expectations about how he or she should respond. Never say, "I know just how you feel." You don't. Think about your helper role as someone who "walks with" not "behind" or "in front of" the one who is bereaved. Allow your friend to experience all the hurt, sorrow, and pain that he or she is feeling at the time. Enter into your friend's feelings, but never try to take them away. And recognize tears are a natural and appropriate expression of the pain associated with the loss.

Avoid Cliches

Words, particularly cliches, can be extremely painful for a grieving friend. Cliches are trite comments often intended to diminish the loss by providing simple solutions to difficult realities. Comments like, "you are holding up so well", "Time will heal all wounds", "Think of all you still have to be thankful for", or "just be happy that he is out of his pain" are not constructive. Instead, they hurt and make a friend's journey through grief difficult.

Understand the Uniqueness of Grief

Keep in mind that your friend's grief is unique. No one will respond to the death of someone loved in exactly the same way. While it may be possible to talk about similar phases shared by grieving people, everyone is different and shaped by experiences in his or her life. Because the grief experience is unique, be patient. The process of grief takes a long time, so allow your friend to proceed at his or her own pace. Don't force your own timetable for healing. Don't criticize what you believe is inappropriate behavior. And, while you should create opportunities for personal interaction, don't force the situation if your grieving friend resists.

Offer Practical Help

Preparing food, washing clothes, cleaning the house, or answering the telephone are just a few of the practical ways of showing you care. And just as with your presence, this support is needed at the time of the death and in the weeks and months ahead.

Make Contact

Your presence at the funeral is important. As a ritual, the funeral provides an opportunity for you to express your love and concern at this time of need. As you pay tribute to a life that has been lived, you have a chance to support grieving friends and family. At the funeral, a touch of your hand, a look in the eye, or even a hug, often communicated more than words could ever say. Don't just attend the funeral, then disappear. Remain available afterwards as well. Remember your grieving friend may need you more in the days and weeks after the funeral than at the time of the death. A brief visit or telephone call in the days that follow are usually appreciated.

Write a Personal Note

Sympathy cards express your concern, but there is no substitute for your personal written works. What do you say? Share a favorite memory of the person who died. Relate the special qualitites that you valued about him or her. These words will often be a loving gift to your grieving friend, words that will be reread and remembered for years. Use the name of the person who has died either in your personal note, or when you talk to your friend. Hearing that name can be comforting, and it confirms that you have not forgotten this important person that was so much a part of your friend's life.

Be Aware of Holidays and Anniversaries

Your friend may have a difficult time during special occasions like holidays and anniversaries. These events exphasize the absence of the person who has died. Respect this pain as a naturalextension of the grief process. Learn from it. And, most importantly, never try to take away the hurt. Your friend and the family of the loved one who has died sometimes create special traditions surrounding these events. Your role? Perhaps you can help organize such a remembrance, or attend one if you are invited.

Understand the Importance of the Loss

Remember that the death of someone loved is a shattering experience. As a result of this death, your friend's life is under reconstruction. Considering the significance of the loss, be gentle and compassionate in all your helping efforts. While the above guidelines will hopefully be helpful, it is important to recognize that helping a grieving friend will not be an easy task. You may have to give more concern, time, and love than you ever knew you had. But this effort will be more than worth it.

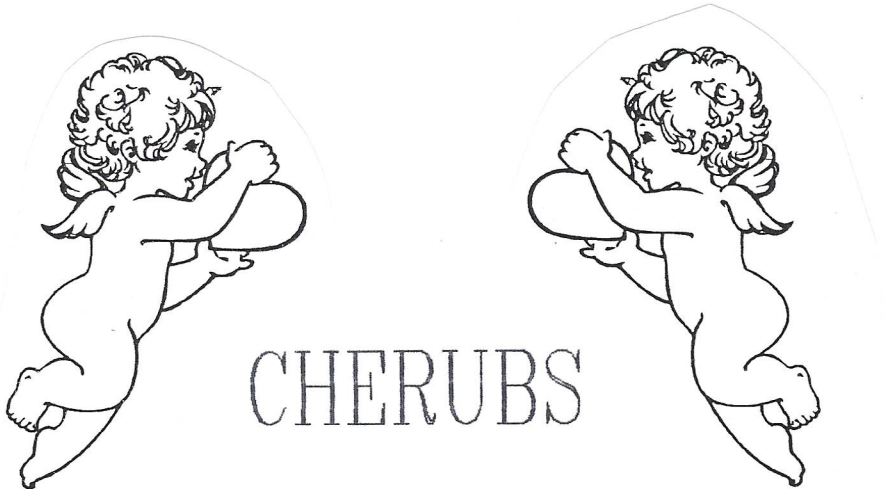
By "walking with" your friend in grief, you are giving one of life's most precious gifts--yourself.

REMINISCING THE PAIN

by Brenda Slavin*

As the summer has passed us all by, some of us reminisced about a previous summer that has resurfaced memories that are very painful. I have been reminiscing about last summer and the summer before. Last summer I was pregnant with our son Nicholas. In June all I could think of was "this is the month we found out about his diaphragmatic hernia". Or the summer before thinking Amanda was just learning how to hold her head up. On the sixth of each month I think of how old they would both be now. Although it has been one and a half years since Amanda died and eight months since Nicholas died, the emptiness remains the same. My grieving didn't stop one month after they died. It didn't stop at one year or almost two years. Maybe some of us don't shed the amount of tears we did in the beginning, but the pain is still very sharp and the emptiness is still there. Sometimes I can talk about the babies and other times I choke just trying to say their names. Grief definitely has no sense of timing. A couple of months ago my parents were in a Christian book store buying my nephew a bible for his confirmation. As they passed through the aisles they noticed several pictures of angels on the cover of several books. They just stopped and began sobbing in the store. They weren't embarrassed that there were several people in the store. They just held onto each other, each helping the other through the moment. My heart broke when my Mom told me about that. She couldn't even tell me without crying. Grandparents should not be forgotten in this web of grief. They not only miss their grandbabies but also hurt seeing the pain their child is going through. My Mom used to think she needed to be "strong" for me and my husband. She would bury her own hurt thinking her pain would make mine worse. Well finally she realized my pain couldn't possibly get any worse and I wanted her to grieve with me not without me. I think the word "strong" is a very funny word. I have been told by many people that I am a very "strong" person. The reason people think that is because they don't always see the real pain that lies in my heart. They are not there when I cry myself to sleep at night. Or they didn't notice that I walked out of the office when someone brought their baby in. I find it very difficult to look at another baby. I absolutely adore and love children. But I can't help but feel jealous of the mother who has her healthy baby and has no idea what losing a child feels like. I cry on every television program that shows a baby born and the baby cries in the delivery room. I never heard that sound from either of my children in the delivery room. The reason I don't grieve in front of certain people is that they make grieving seem like a contagious disease. We "the grievers" are accused of withdrawing from society. When in reality it's mostly society that has withdrawn from us. They don't want to see it, hear it or be around it. these are not bad people. They are just naive to how to handle someone that has been through this kind of tragedy. I have found my lifetime friends and relatives. I had to "discard" those "friends" and family members who chose to distance themselves from me. I try not to be angry with them for hurting me this way. Sometimes I have to remember they do not understand what I have been through. They are embarrassed because they don't know what to say, what to do, or what not to do. But in order for me to move forward and handle this the best way I know how I am choosing to be with the people who have supported me and loved me through this difficult time. Many people have said that I have changed since the babies died. I can't say that I'm not slightly bitter about what has been taken away from me. I had two beautiful children and now I have none. I have become a more serious person. I get angrier when I see unappreciative parents, child abuse and neglect (right down to a parent allowing a child to stand in the seat of a car because the child doesn't feel like being put into a car seat) I want to scream at them "don't you realize how quickly you can lose that child? Don't you know how precious their little lives are?" Overall I think I have grown into a more compassionate person. I find myself to be a much better listener. And I'm now a better friend to those I do have left. So as we reminisce on the sad memories or we are facing the anniversary of our children's journey to heaven, we need to remember to listen and comfort each other. I know a lot of the parents of the Cherubs that are still with us want to be there for us. They may not fully understand our pain but they want to help us and comfort us. I pray that God grants us with the peace and the love that we all need to help us through this roller coaster of emotions. We must remember our children are always with us. We may not see them or hear them but we can always feel them inside our hearts.....loving us forever.

*Brenda and her husband, Bob, are the parents of Amanda (Nov. 6, 1993-Nov. 1, 1994) and Nicholas (Nov. 6, 1995). Brenda is our parent advisor for berieved parents and columnist for berievement. Both of Bob and Brenda's children suffered from Fryn's syndrome and congenital diaphragmatic hernia. If anyone can be labeled as "strong", Brenda certainly qualifies for sharing her feelings and grief in order to help others.



The Association of Congenital Diaphragmatic Hernia, Research, Advocacy, and Support

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