SUMMER 1995

CHERUBS

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

CHERUBS
c/o Dawn M. Torrence
3671 Bruce Garner Rd.
Franklinton, NC 27525

Thank you all for making our first newsletter such a success! All of the letters and stories sent were so touching, I know they will each be an inspiration to all other parents. CHERUBS has just filed for non-profit status so soon all donations will be tax-deductible! Please continue to share your newsletters with other parents and doctors. I need to ask all of you to please fill out the new Parent Worksheet. Thanks again! Dawn

CHERUBS Would Like To Thank The Following People For Their Help:

Lesli Taylor, MD - Asst. Professor of Pediatric Surgery, University of North Carolina
Iley Baker Browning, MD - Pediatric Pulmonary, Duke University
Prem Puri, MBBS, MS - Director of Research, Children's Research Centre, Dublin, Ireland
John A. Harris, MD, MPH - Chief, California Birth Defects Monitoring Program
Jacob C. Langer, MD - Pediatric Surgery, Washington University of St. Louis
Betty Mekdeci - Executive Director, Association of Birth Defect Children, Inc.
Liz Stierman, MS - Editor, Perspectives in Genetic Counseling
Tim Safley, CRT - Ambulatory Services of America
Rachel L. King, RN - Duke University P.I.C.U.
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Ann Worthington - In Touch Trust, Great Britain
Mr. Jeremy D. Torrence
Julie Gordan - MUMS
Mrs. Michelle Y. Riley
Mrs. Elena Powell
Ms. Susan Pate

CHERUBS Would Like To Thank The Following People For Their Generous Contributions:

Measurement Inc Printing
B. Mayes Marks, Jr., Esq.
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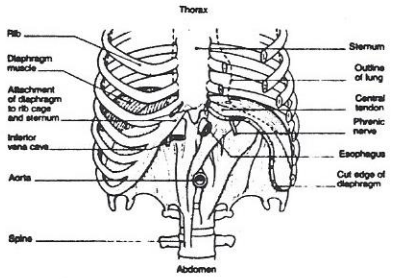
Drew & Jennifer Torrence
Todd & Melissa LePage
Prem Puri, MBBS, MS
Lesli Taylor, MD
Mrs. Mita Patel
Mr. Matthew Torrence
Mr. Michael Torrence
CCB Bank of Oxford, NC

ANATOMY OF THE DIAPHRAGM



* *From the American Medical Association Home Medical Encyclopedia; 1989; medical editor - Charles B. Clayman, M.D.; Random House; NY, NY*

CHERUBS is an international organization for families and caregivers of children who are diagnosed with Congenital Diaphragmatic Hernias (CDH). CHERUBS provides information, support, and parent-to-parent match-ups. There is no cost to parents for services provided by CHERUBS. We are a volunteer organization. CHERUBS was founded in February 1995. Donations and very welcomed and checks can be made out to CHERUBS. The opinions shared in this newsletter do not necessarily represent the opinions of all members or staff. The information in this newsletter is by no means to be substituted in place of proper medical advice. Remember, every child is different. You can't compare the progress of another CDH child to the progress of your child. They are all little angels . . . . . .CHERUBS.

GOALS FOR THE FUTURE OF CHERUBS!!!!!

* Providing information to parents immediately following their child's diagnosis.
* To advocate for prenatal diagnosis and research on the causes of CDH
* To recruit more members

This newsletter is dedicated to the memories of:

Harold Jennings Mitchell, III
April 5, 1995 - April 17, 1995

Amber Joyce Moore
August 14, 1994 - September 8, 1994

Gregory Joseph Jennings
March 4, 1993 - March 14, 1994

THE ASSOCIATION OF BIRTH DEFECT CHILDREN, INC.

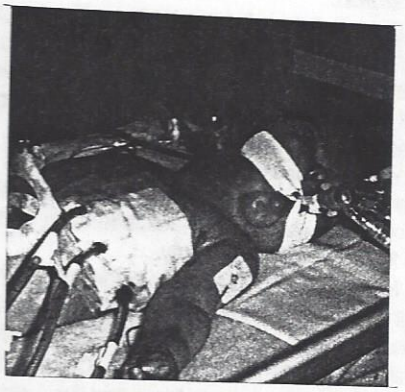
The Association of Birth Defect Children, Inc. is a national organization that conducts extensive research on birth defects, including Congenital Diaphragmatic Hernias. I urge all members of CHERUBS to register with the Association of Birth Defect Children, Inc. You will be asked to fill out forms regarding the family, prenatal and postnatal history of your child. Isn't the amount of time it will take to fill out these forms worth finding a cause and prevention of CDH? For more information write to:

The Association of Birth Defect Children, Inc.
827 Irma Street * Orlando, FL 32803
or call: (407) 248-7035

FOR PARENTS WHO ARE EXPECTING A CHILD WITH CDH

The University of California in San Francisco conducts in-utero CDH repairs. The doctors there are working miracles! For information call: 1-800-RX-FETUS

YOU'VE COME A LONG WAY, BABY!



Anna Tina
April 6, 1992
(left-sided CDH, Congestive Heart Failure, Hydrocephalus, 2 Shunts, G-tube, 2 Nissens, Pulmonary Hypertension, 13 chest tubes, and 6 surgeries)
ANNA IS DEFINITELY A CHERUB!
Joan Tijan
Rt. 2, Box 986 * Crandon, WI 54520

FOR DOCTORS AND PARENTS OF CHILDREN RECENTLY DIAGNOSED WITH CDH

CHERUBS will soon publish a pamphlet especially for parents of children who are recently diagnosed with CDH; at birth or in-utero. It will be valuable to medical professionals to help them explain to the parents their child's diagnosis. It will include medical terms, information on dealing with emotions, breastfeeding, financial help, grief, and words of wisdom from our veteran parents. For information call: (919) 528-1544.

Letters To CHERUBS

April 18, 1995

Dear Ms. Torrence:

Thank you for the CHERUBS newsletter. I was greatly touched by your commitment, energy, and courage.

I direct the California Birth Defects Monitoring Program (CBDMP). Our program does research to try to find causes of birth defects. To do this research we collect and analyze data in California about children with birth defects, including CDH. No one unfortunately knows the causes of CDH so there is not much that I can tell you at this time. As a pediatrician scientist I wish there was more known to help people like you.

I'm really glad that there are people like you advocating to help children with birth defects. If you would like to talk, please do not hesitate to call me.

Sincerely,

John A. Harris

John A. Harris, MD, MPH, Chief
California Birth Defects
Monitoring Branch

April 5, 1995

Dear Ms. Torrence:

Thank you for your letter. I really like the name of your organization. I would be happy to print an item about CHERUBS on the Parents' Page in our next newsletter so families of children with CDH will be able to contact you.

Our mailing list exceeds 12,000 so you may hear from quite a few families.

We would appreciate it if you would let your group know about our National Birth Defect Registry. We would like to have as many cases as possible of different kinds of birth defects in our database.

Sincerely,

Betty Mekdeci

Betty Mekdeci
Executive Director
Association
of Birth Defect Children, Inc.

May 30, 1995

Dear Ms. Torrence:

I recently received information about your new organization through my job at the California Birth Defects Monitoring Program. I thought your newsletter was quite impressive - very well done.

I am also editor of the quarterly newsletter for the National Society of Genetic Counselors. I'd like to include information about your organization in the research section of our next issue-genetic counselors often meet families who have just learned their child has CDH, either prenatally or in the newborn period, and I think they would be prolific source of referrals to CHERUBS.

I look forward to hearing from you soon.

Sincerely,

Liz Stierman

Liz Stierman, MS
Editor, Perspectives in
Genetic Counseling

April 12, 1995

Dear Ms. Torrence:

I received your letter of February 27 concerning the activities of your organization, CHERUBS, and your request that we refer patients with CDH to your organization.

The Division of Birth Defects & Developmental Disabilities is a part of the Centers for Disease Control & Prevention. Our mission is to prevent birth defects & developmental disabilities. Our activities involve monitoring the rates of about 150 birth defects, including CDH, and conducting applied research to find the causes of birth defects. We conduct similar programs for developmental disabilities such as mental retardation, cerebral palsy, hearing and vision deficits. Most of this research is conducted by search of medical records, rarely involving contact with the patient or their family.

I congratulate you for founding CHERUBS to provide support to the families of children with CDH. I know how valuable this type of support is to families as they struggle to understand the nature of this birth defect and how to help their child learn to live with it. Organizations such as yours can be a tremendous help to these families. I wish you success as your organization and your children grow.

Sincerely yours

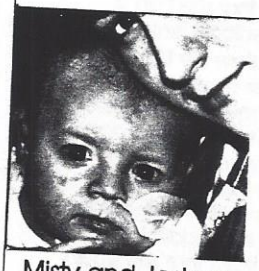
Godfrey P. Oakley, Jr., MD
Director
Division of Birth Defects &
Developmental Disabilities,
National Center for
Environmental Health

Letters From Members

Dear Dawn,

It was so nice of you to write and introduce yourself. I always enjoy hearing from moms who have been lucky enough to share in one of God's miracles. We have been very blessed with our daughter, Hannah, and her surgical scars remind me every day of God's promise and faithfulness. I. enjoyed your newsletter. It was very informative and contained so much. I must agree with the need for support groups. I went into the hospital to have a baby and came home without one. I didn't feel very well informed of Hannah's condition and most of my education came from other moms. Every parent of a critically ill newborn needs to know where they can turn to talk to others who have been there . . someone who is not part of the medical team but just another parent.

Sincerely,
Jackie Horn
4212 Green Park Rd. * St. Louis, MO 63125


Misty and Joshua

Dear Dawn,

My nightmare began five months ago. I got pregnant at 18 and gave birth at 19. All of my doctor visits were normal, even my ultrasound. I had one 2 days before I delivered. On November 13, 1994, seconds after the umbilical cord was cut, my son was laid on my chest for a split second until he turned blue. The nurse took him from me to the corner of the room where everyone crowded around. Scared and crying, I repeatedly asked what was wrong. No one would tell me. My son coded and was brought back. When I was allowed to see my son, he looked scared and we both felt helpless. He was full of tubes, wires, and everything else. He was taken away by helicopter to Johns Hopkins Hospital in Baltimore. My family and I chose to go ahead with surgery instead of taking him off the vent. He was ECMO for 14 days until his brain began to bleed. On December 21, 1994, they made him a diaphragm out of Gortex. They gave him a 5% chance. I was so proud of my baby, I traveled 2 hrs. every day to see him. My parents were always there. Joshua has had many ups and downs. I knew he was meant to be here for a reason. I thank God every day that he is alive. I was allowed to hold my son when he was 3 1/2 months old. At 3 months he had a tracheostomy. He is still on the vent and soon he will get a G-tube. I have been told that he will be vent dependent for several years - I have had training and am ready and willing to take care of him. My son, Joshua, is now 5 months old and is the best thing that ever happened to me.

Sincerely,
Misty D. Weller
15207 National Pike, Apt * Hagerstown, MD 21740

Dear Dawn,

Thank you very much for contacting me and my husband, Randy, to join CHERUBS. I am very grateful to you for establishing this support organization. There is nothing out there to specifically support CDH parents - so thanks! My son, Dallas, was diagnosed in-utero with his hernia because I was suffering with polyhydramnios. It turned out to be such a blessing because we had a "heads up" and the doctors were prepared also. Dallas had an ECMO of 10 days and then had his surgery. He had to be on a ventilator for about 5-6 weeks. The doctors tried several times to take him off but he couldn't maintain his oxygenation well enough. When he finally came off, he had nasal cannulas for about 7-8 more months. On his first birthday, the oxygen was finally taken out of our home. What a present! He is presently doing really well. He is walking, climbing, running, and behaving like a 22 month old. He has a few developmental delays but more in his fine motor skills. He's beginning to talk also but not as soon as my other 3 kids. His first year was pretty rough because every little cold landed him in the hospital. With 3 other siblings it was a sure bet we were going to get sick! We praise God for all the miracles we have seen in Dallas' life and in the surrounding circumstances. Dawn, thank you once again, for starting this organization. We would love to be able to help other parents who are in the same boat. I hope you don't mind but I copied your first newsletter and sent it to a friend of mine who also has a CDH child. She is such a source of encouragement to me. I also took the newsletter to Madigan's NICU so they can use it for other parents. Take care and may God bless you and keep you!

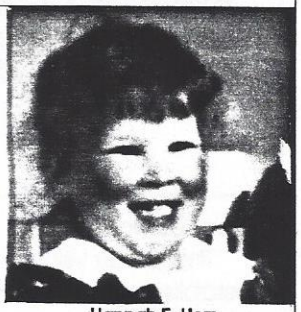
Sincerely,
Diane Cox
4032 Hemlock St. SW * Tacoma, WA 98439

Stories of CHERUBS

Sarah Harrison
August, 13, 1990

I had an "ideal" pregnancy, complete with routine ultrasounds, all assuring me I had a healthy child on the way. How they could fail to see that Sarah's intestines were up in her left lung cavity where her lung should have been is beyond me. The world came crashing down when I went into premature labor 3 weeks early and the fetal heart monitor recorded that each time I had a contraction, her heart rate would cut in half. The doctors immediately surmised that she wasn't getting enough oxygen and began prepping me for a C-section. I was so scared. I told them they couldn't do one because I hadn't read that chapter in the book yet! Ridiculous, huh? Of course we proceeded with the C-section. When Sarah was born I remember waiting to hear her cry that would let me know she had arrived. That sound never came. Within seconds she was surround by specialists and whisked away. I kept asking, "Is she dead?". The doctor had managed to say, "It's a girl". As I was being stitched up, I was told that "you have a very sick baby". The recovery room can be a place of despair when left alone with no information and only you own imagination running wild. Two hours later, barely hanging on to my sanity, my husband came in, weeping uncontrollably, and told me that she had a Diaphragmatic Hernia and they were going to fly her to the University of Tennessee Medical Center. I had never even seen her! About then, the doctor came and explained what the condition was and that I could see her briefly before the Life Star team took her. I will never, ever forget his next words to me. He said, "Karen, take a good look, you will probably never see her alive again". Could it truly be that the child we had prayed for would come into our lives only to be taken so quickly? She was so tiny and I know all the tubes and equipment she was hooked to weighed more that her, but to me she looked like an angel. I stroked her cheek and a moment later they took her. My arms ached to hold her and I felt so empty. Just then, a member of the Life Star team came back and handed me a Polaroid picture of her. He said, "I know you can't hold her, but you can hold onto this". That is still my favorite picture. So much happened in the following days that there is no way I can recount it all unless I write a book. But quickly let me say that although it was uncertain whether Sarah would live at first she it made through surgery and was weaned off the ventilator in 9 days, which is miraculous. During those days I wanted to help her all I could. There wasn't much I could do, but giving her my breast milk was something I was committed to. I knew that she would need the nutrients from it. So I pumped and pumped. Was it easy? Never. I would hold her picture (tubes and all), and cry and pray, and pump. The hospital froze it until she was able to have it. It made me feel like I was "making up" to her for "allowing" her life to begin this way. Although the doctors assured me that there was nothing I did to cause this, I felt an overwhelming sense of guilt. For new parents out there: please realize that those feelings are normal and unfounded. The good news is that they passed, and were replaced by many other feelings that come and go at random, even now, after 5 years. As I write this, my daughter Sarah, is doing her best to clean out my cupboards, rearrange the booksheives, and drag out every toy she owns. And what do I say to that? "Good girl, Sarah". It tickles me pink to see her able to do things that we never dreamed she would live to do. (Sarah was born with left-sided CDH, Trisomy 14-Partial, Agenesis of Corpus Collusum, and enlarged kidneys on 8/13/90).

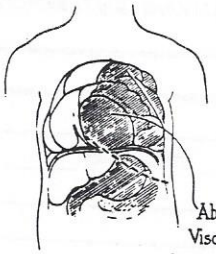
Karen Harrison * 86 Gefellers Dr. * Greeneville, TN 37743



Hannah E. Horn
December 12, 1990

It was 10:02 pm on December 12, 1990 when Hannah made her grand entrance into this world, just seconds after the doctor arrived. After 2 sons, I was blessed with a daughter. My sister, Lu, was on a conference call to our mom and older sister, bragging about her new niece and how she practically single-handedly delivered the baby. Lu stood in as proxy for my husband who had recently suffered a serious knee injury that left him incapacitated. I couldn't wait until things settled down so I could introduce Hannah to her father via the telephone. But some things weren't meant to be. Hannah was in respiratory distress and she was whisked away for evaluation. Lu and I waited in silence. A short distance down the hall, in another birthing room, we heard an excited cry from a father who had just witnessed the birth of his son. His jubilant cry pierced my heart as I waited on word about my own daughter. The wait seemed forever but it was actually only 30 minutes before the pediatrician entered my room. I held my breath expecting the worst when the doctor took my hand in hers and spoke. "You have a very,, very sick little girl. The x-rays show that Hannah has a diaphragmatic hernia. This means she has a hole in her diaphragm which allowed the intestines to move up into the chest cavity, hindering the growth of her left lung and caused displacement of her heart. We placed her on a ventilator and a helicopter was called to transport her to St. Louis Children's Hospital for immediate surgery". In less than an hour the helicopter was gone, carrying my little girle to the hospital. In the quiet of my room, I called my husband and told him about his beautiful but very critical daughter as I gazed at the Polaroid given to me by the helicopter team. The dawn greeted me with a phone call from Hannah's surgeon. He said, "She came through like a champ. Her condition is stable but very critical. We'll just have to wait ands see what happens in the next 48 hours". I thanked him and he told me that Hannah is a beautiful baby. That morning, I made my first of many trips to the Children's Hospital. Hannah was an extension of wires and tubes. Her heart monitor begged for attention while her vent forced air into her lungs. Just seeing her like this left me emotionally exhausted. It was here that I introduced my husband to his daughter. It was very difficult to go home that night and leave my baby but I had to extablish some degree of normalcy before my husbands's scheduled knee surgery the next morning. At 1:00 am, less than 24 hours after Hannah's surgery, her doctor called to tell me that Hannah needed another surgery in order to sustain her life. ECMO would replace the work of her heart and lungs, allowing her own organs to rest. The side effects could include deafness, blindess, brain hemorrhage, even death. But this was our only hope. Family and friends babysat my sons, chauffeured me to the hospitals, brought food, cleaned my house, and stayed with me through the endless nights. The new week brought good news. Hannah was successfully weaned off ECMO and my husband was released from the hospital. Hannah depended less and less on the vent and on December 23rd, she was completely weaned off and breathing on her own. After a month in the hospital, Hannah came home. That was over four years ago and Hannah is doing great. She walked before her first birthday and at 18 months, passed her neurological exam with flyng colors. By three she was rollerblading and at four she rides her two-wheeler without training wheels. She has a passion for books and loves to read. She has a thirst for learning and enjoys doing her letters and simple math with our computer. This January she started ballet and tap lessons. Some people think Hannah is quite amazing, I myself think she is quite normal. I attribute her quick learning to the fact that she tries to keep up with her older brothers. I thank God for sharing the miracle of Hannah with us. I'll never forget the support, the prayers of Job (Job 42:10 The Lord restored his . . . happiness. In fact, the Lord gave him twice as much as before).

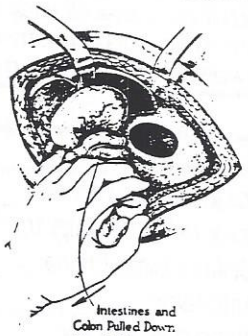
Jackie Horn * 4212 Green Park Road * St. Louis, MO 63125



Abdominal Viscera Herniated Into Left Pleural Cavity

The Surgical Evaluation and Management of Diaphragmatic Hernia

by Lesli A. Taylor, M.D.



Intestines and Colon Pulled Down Out of Thorax

The diaphragm is a broad muscle that creates respiration of the lungs by contracting and relaxing. The diaphragm forms at 8 weeks of gestation. This occurs by merging of two membranes to close off the open area between the chest and the abdomen. If it does not close completely, a defect, called a diaphragmagic hernia, is created. The term "hernia" refers both to the hole in the diaphragm muscle and the protrusion of the abdominal organs into the chest.

The most common defect occurs on the left toward the back of the body. This is called a Bochdalek hernia. The hole may be nearly as large as the diaphragm itself. When the diaphragm is completely absent, this is call agenesis of the diaphragm. A less common hernia, called a Morgagni hernia, occurs near the front of the body, near the breast bone.

The stomach, small and large bowel and other organs, such as kidney and liver can herniate up into the chest. Hernia on the left tend to allow many of the abdominal organs into the chest. On the right, the liver covers the hole and there is less chance of bowel or other organs entering the chest.

The biggest problem caused by this herniation of the bowel is that the lung on the affected side and even the lung on the opposite side cannot grow to its fullest capacity. The extent of the lung deficiency becomes apparent at birth when the umbilical cord is clamped and the infant must oxygenate its blood by breathing.

Diaphragmatic hernia can be diagnosed during pregnancy with ultrasound. This allows the pregnancy to be closely monitored. The mother can be delivered at a specialized center for high risk pregnancies where the pediatric surgeon and ECMO (extra corporeal membrane oxygenation) are available.

Diaphragmatic hernia can be repaired in the womb by fetal surgery, but this is still considered high risk and experimental. It is avaiable in San Francisco for selected patients.

The severity of problems the child will have depends on how big the hole in the diaphragm is, how much of abdominal contents have entered the chest, and at what point in time in gestation the herniation occured. These factors determine the lung capacity.

Shortly after birth, the baby swallows air while crying and the bowel in the chest inflates with air and further compromises the insufficient lung. Children with severe lung defiency require intubation and ventilation in the delivery room to survive. Children with better lung capacity may not be detected for sevral weeks, months or even years, perhaps by a chest x-ray taken for other reasons.

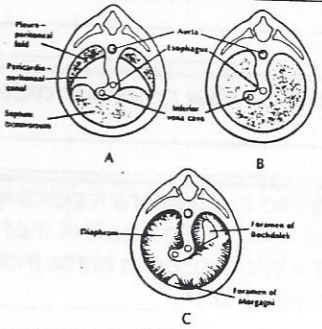
Surgery is required to return the abdominal organs to the abdomen and to repair the diaphragm. There is no surgery at present to repair the lung insufficiency. Lung transplant has been used to replace deficient lungs in diaphragmatic hernia, but this is experimental and obtaining donor lungs for newborns is extremely difficult.

An abdominal chest incision can be used to repair the diaphragm. Many pediatric surgeons prefer to use an abdominal approach as a plastic tent (Silastic silo) can be sewn to the abdominal will to hold the bowl temporarily in the rare event that the abdominal organs will not fit back in the abdomen. Timing of the repair depends on the patient's stability and the patient's need for ECMO. Bowel injury due to the herniation is rare and is not a reason to operate immediately.

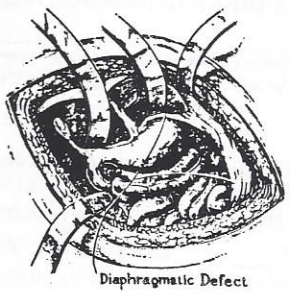
Sometimes the defect is large enough to require synthetic material to close the diaphram. This will keep the bowel out of the chest, but will not grow with the child. This may lead to detachment of the graft in children with a very large defect. Graft material is a foreign body to the patient and has a greater risk of infection than natural tissues. Muscle flaps have been devised to repair the defect. while there is less risk of infection, the flank will be weak due to loss of two layers of muscle.

Children with severe lung insufficiency can be placed on ECMO if they qualify. This allows circulation of the blood through an external oxygenator pump. Timing of surgery to close the defect in a baby on ECMO is at the discretion of the surgeon.

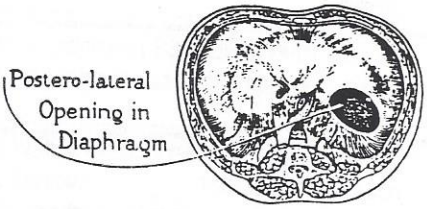
Recurrence of diaphragmatic hernia is rare. The most important problem the child has after surgery is attaining adequate lung capacity by gradual, though minimal, expansion of the deficient lung, and lung growth with time.



A B C



Diaphragmatic Defect



Postero-lateral Opening in Diaphragm

Bear ye one another's burdens. Galatians 6:2 KJV

Even if you have already filled out the last form, please fill out this new form also. New members must complete and return this form to be put on our mailing list. Mail to: CHERUBS, 3671 Bruce Garner Road, Franklinton, NC 27525. If you have any questions or want to talk, call me at (919) 528-1544.

Parent Worksheet

Your name: _____ Title: _____
Your child's name: _____ Male _____ Female _____
Your relationship to this child: _____
Child's Birth Date: _____ Date of Death (if appl.) _____
Address: _____
Phone Number: _____ Number of Siblings: _____
Is there any family history of CDH? _____ If yes, who? _____
How did you learn about CHERUBS? _____

Pregnancy History

AFP Test _____
Ultrasound _____
Amniocentesis _____
Age of mother at delivery _____

(Did you, the mother, have any of the following:)
Prenatal diagnosis of CDH _____
Polyhydramnios _____
Any Abnormal Findings _____
Age of father _____

(You are not obligated to answer any questions that you are not comfortable with.)

Birth History

Birth Weight _____
Side of Hernia _____
Was your child early? _____
Type of Hernia _____
Age child was diagnosed _____
How do you feel about the birth experience? _____
Was your child delivered vaginally or by C-section? _____

MEDICAL HISTORY

At what hospital(s) was your child treated? _____
How much lung capacity does your child have? _____
What was the length of your child's hospital stay? _____
Does your child have any other birth defects? _____
Was your child ever on ECMO? _____ How many diaphragm repairs has your child had? _____
Did your child have any complications? _____
For how long did your child receive ventilator assistance? _____
For how long did your child need oxygen? _____
Has your child had difficulty eating by mouth? _____
At what stage is your child, developmentally? _____

Do you give CHERUBS permission to publish your name, your child's name, your address, phone number, your child's case history, and/or picture? _____
Signed _____ Date _____

Have you registered your child with the Association of Birth Defect Children, Inc.? _____ Do you give CHERUBS permission to release information and/or your name to research organizations? _____
Signed _____

Do you give CHERUBS permission to give out your name, address, and/or phone number to other parents in order to make a parent-to-parent match-up? _____
Signed _____ Date _____

Do you feel that the hospital staff that cared for your child kept you informed and involved in decisions regarding your child's care? _____ Would you recommend your child's doctor to other parents? _____ Do you feel that you were given enough information about CDH? _____ Did your child's doctor explain this information in terms that you could understand? _____ Do you have any advice for new parents of children born with CDH? _____

_____