ea|tef

Child and Family Support Connection, Inc.

Joining families and resources in support of children born with Esophageal Atresia and Tracheoesophageal Fistula

INSIDE

The following article was published in the June 2002 issue of the *Journal of Pediatric Surgery* about a less invasive surgical technique to repair tracheoesophageal fistula in newborns. The author, Dr. Steven S. Rothenberg, is a pediatric surgeon who practices in Denver, Colorado. The article is meant for medical professionals, but we publish it here to help parents learn about surgical advances in the area of EA|TEF. A brief question and answer section with Dr. Rothenberg follows the article on page 13.

—The Editors

Thoracoscopic Repair of Tracheoesophageal Fistula in Newborns

By Steven S. Rothenberg
Denver, Colorado

Background: Advancements in minimally invasive surgery in neonates have allowed even the most complex neonatal procedures to be approached using these techniques.

Methods: During a period of 15 months, 8 patients born with a proximal esophageal atresia and a distal tracheoesophageal fistula underwent repair thoracoscopically. Weights ranged from 2.1 to 3.4kg and operating times ranged from 55 to 120 minutes.

Results: All procedures were completed successfully thoracoscopically, and there were no operative complications. One patient had a small leak on day 4 that resolved spontaneously on day 8. All other patients were shown to have a patent anastomosis with no leak by Barium swallow on day 5.

Conclusion: This initial report shows that esophageal repair in the neonate is technically feasible and may provide advantages in terms of exposure and esophageal length, as well as the recognized advantages of avoiding a thoracotomy.

INDEX WORDS: Tracheoesophageal fistula, esophageal atresia, thoracoscopy.

RECENT ADVANCEMENTS in technique and instrumentation in pediatric endoscopic surgery have allowed significantly more complex and delicate procedures to be performed, even in small neonates. Over the last 5 years the number and breadth of minimally invasive surgical (MIS) procedures performed in infants has increased dramatically.[1-3] However, 1 procedure, successful ligation of a tracheoesophageal fistula with repair of the esophageal atresia, has remained relatively elusive. In 1999, a stepping stone was laid when a successful thoracoscopic repair of a pure esophageal atresia was completed in a 2 month–old boy.[4] One year later we reported on the first successful repair of an esophageal atresia with tracheoesophageal fistula (TEF) in a newborn using a completely thoracoscopic approach.[5] These accomplishments provided the needed experience to allow us to undertake a prospective study to thoracoscopically repair all TEFs in hemodynamically stable patients.

MATERIALS AND METHODS

From March 2000 to July 2001, 8 consecutive patients with esophageal atresia and a distal tracheoesophageal fistula were referred to the author for repair. Four diagnoses had been made prenatally, and the babies were delivered at the high-risk, perinatal/neonatal center. Four others had postnatal diagnosis and were transferred after birth.

Gestational age of the patients ranged from 31 weeks to 40 weeks at the time of delivery. Three other infants with TEF also were referred during this period but were excluded because of size and associated anomalies.These 3 weighed 800g, 1,100g with a tetralogy of Falot, and 1,800g with an omphalocele.The other patients ranged in weight from 2.1 to 3.4kg (mean, 2.6). Preoperative evaluations found small ASD and PDA in 2, a VSD in one, and a tetralogy of Falot with a right–sided aortic arch in 1. There were no other major congenital anomalies. Two patients required intubation before surgery for

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From the Chairman's Desk

Thank You…

I've said that many times over the past year. In May of 2001, Cindy and I welcomed a beautiful, healthy baby girl, Clare Elizabeth, to our family. Forget the 10 fingers and 10 toes, just suction her and let us know she has a normal esophagus.

Thank you, God, for making this baby complete…

A week after bringing our new daughter home, I came down with a case of strep throat I just couldn't shake. The fever wouldn't break. I was dehydrated and couldn't get any rest. One night, I awoke with cramping in my arm and pains in my chest. My doctor said it was most likely indigestion from the massive amounts of ibuprofen prescribed to break my fever, but told me to go to the ER just to make sure. An hour later, I was told I'd suffered a heart attack. I was 41 years old.

Thank you, doctor, for erring on the side of caution.

Lying on the gurney I began to cry. This couldn't be happening to me, I thought. I had a 13–day–old daughter I barely knew. Our son, Jonathan, was only 8 years old. Cindy was still recovering from her C–section. My mother, up from Arizona to help with Clare, extended her stay for an extra month.

Thank you, Mom…

Over a year has now passed. My diagnosis is a rare heart condition called viral myocarditis. The good news is I don't have coronary heart disease and I am close to a full recovery. The bad news is my activities were severely restricted while my heart healed. The ea|tef Child and Family Support Connection suffered from my lack of attention.

Thank you, families, for your patience while things get back to normal…

Finally, a special note to the big guy upstairs: All those months of bed rest offered me the chance to do a lot of reflection. I have a loving wife, two healthy children, wonderful friends and family, and a career I truly enjoy.

No thank you, God, I don't need another wake–up call.

—Bruce Davis
Chairman, Board of Directors

Letter from the Editors

Whew! For a while there, it seemed this issue would never get done. But here it is and we hope that it helps parents new to the EA|TEF world, others whose children have been living with EA|TEF for a few years, and even some adult EA|TEF–ers themselves. As we wrote several months ago, the organization has had a tumultuous time of it, with health problems and staff changes, but we have regained our footing. Thanks, everyone, for your patience.

As some of you may recall, last year former editor Laura Novak (who made it look SO easy!) passed her job over to two other EA|TEF moms, Elizabeth McNamara and Heather Fowlie. Elizabeth lives in East Greenwich, Rhode Island, and is mother to three children: Catherine (11), Aidan (9) and James (5). James was born with Down syndrome and long–gap EA in 1997 (his story is told in the Summer 2001 issue). While James takes all of his nutrition by mouth now, his esophagus is less than perfect (i.e. he chokes a fair bit) and he is prone to respiratory infections. He is on very good terms with his pediatrician.

(Continued on page 6)

In Coming Issues...

In the next few issues, we want to cover topics ranging from reflux and feeding problems to sibling issues and traveling with a medically fragile child. If you have something to say on any of these topics, or just want to share your story, we'd love to hear from you.

Lane's Story

By Amy Dulworth

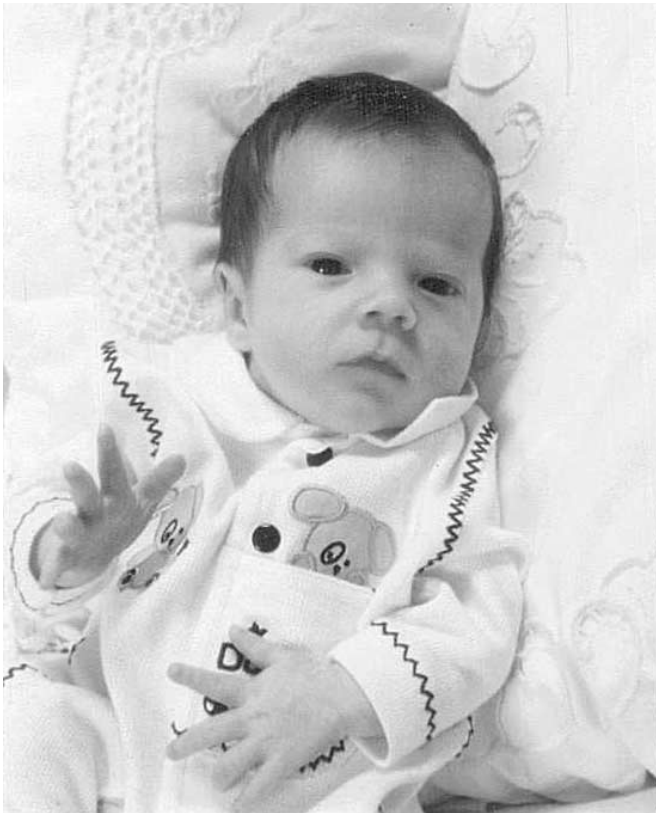
Our precious baby boy was born. Oh, I felt so much relief that he was finally here, September 20, 1998, after a long 36 weeks. As I received a thumbs up from the observing pediatrician, I felt relieved and ready for rest. As I was moved into a private room, my husband went with the baby to the nursery. After 20 minutes went by I was so ready to see my new bundle of joy! No one was around, no nurse, no relatives —it seemed as if I was the only one in the hospital.

A few minutes later my husband, David, came in my room. He had tears in his eyes, a scared look on his face, and he was very silent. I thought he was overwhelmed with all the excitement. A few minutes later my mother–in–law came in the room with the same stale look on her face and still I just thought everyone was exhausted from the long night of laboring. Then the pediatrician who had given me the thumbs up came rushing in. I thought that I was in the middle of a horrible nightmare but I was awake and this unbelievable chain of events was really happening.

Lane was beautiful, with a full head of jet black hair, nicely plump at 6 pounds, 5 ounces, and 19 inches long. A perfect baby, right? Until the frightening moment when they tried to feed him, and he turned blue. Several attempts were tried and Lane turned blue each time. An x–ray machine was brought in and the picture it showed changed our lives. Lane was diagnosed with esophageal atresia and tracheoesophageal fistula (other birth defects were found later).

Lane was born at Mesquite Community Hospital, in Mesquite, Texas. The nursery had no other babies in it, and the labor and delivery floor was empty. When the medical team finally decided to transport Lane to Methodist Hospital in Dallas were he would undergo immediate surgery, David went with him, leaving me behind in the private room with little more than empty arms and tears. Before the transport, they wheeled Lane in so I could say goodbye. He did not look like any baby I'd seen before. There were tubes, wires and machines everywhere. He was discolored and swollen, not the perfect looking baby I had seen only a short time before. They told us we could touch him before he left, and as we laid our hands on him all we could do was cry. Never before had I such despair, having to entrust four strangers to transport our baby to a hospital 30 miles away, without me. All I had was a Polaroid picture of him to cling to.

At Methodist Hospital, Lane was under the care of Dr. Klein, a neonatologist, and Dr. Jeorge Uceda, a pediatric surgeon. On arrival Lane was prepped and taken into surgery.

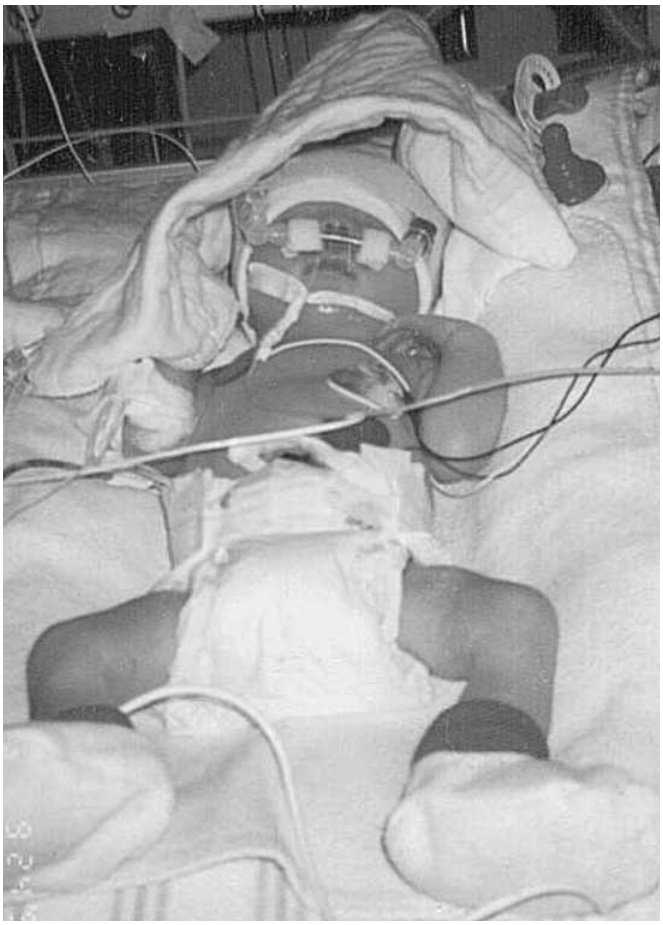


Since we didn't get hospital pictures, this is Lane when we got home, 30 days later.

He was six hours old. The surgery, to close up the fistula between the esophagus and the trachea, was a success and Lane was taken to the critical care nursery where he stayed for 20 days. The white crib we had picked out for him was home, empty. Instead, Lane lay in an incubator, with tubes running everywhere. My husband and mother–in–law stayed with him through that first night. Nurses at Methodist called faithfully through the night to update me on his condition. I could not comprehend everything that was going on. All I knew was that I wanted to be there. The next day, I checked myself out of Mesquite Community Hospital and went straight to Methodist to be with Lane.

As I entered the hospital, I had a huge lump in my throat— Was this really happening? Did I do something wrong during my pregnancy? I did everything by the book. As I entered the hall of the neonatal intensive care unit, my heart was beating so fast that I felt faint. I had to push a button on the wall to even enter the nursery where MY child was. Once inside, I had to scrub my hands and arms for five minutes and put on protective clothing before I could enter the room where my child lay. All I wanted to do was hold Lane, my newborn. In the NICU, my breath was taken from me. None of the babies looked like babies, they looked like play dolls hooked up to machines, noisy machines that kept beeping.

And then there was Lane, the biggest baby there. He didn't look like anything I had imagined. He had so many tubes and machines hooked up to him, there was no way that I could



Lane at 12 hours old, right after his surgery, in the critical nursery.

hold him, The tears just rolled down my face. I just stood there. The nurse introduced herself, but I couldn't hear her name, I couldn't hear anything. I was numb.

How could I touch Lane, how could I hold him, how could I do anything? Slowly I learned. We spent day in and day out there, setting up camp right outside the NICU doors. My husband never left the hospital, but sent me home periodically to get some rest. We spent every minute with Lane that we could.

Finally, the day came and we got to bring him home. As we were leaving, one of the doctors who had cared for Lane told me that even though they had repaired his esophagus, there could be problems in the future. BOY—that was a statement of truth. But after 20 days in the hospital Lane came home! We were so excited.

At home Lane was bottle feeding and doing well. At three months, Lane was hungry all the time, so I decided to add cereal to his feedings. I sat on the couch to give him his first feeding of formula–and–cereal mix, and a minute later I was dialing 911. Lane was blue, not breathing, lifeless. I was in such a panic. You can never prepare yourself for something like that. This episode was the first of several of what we came to call "blue spells."

Over the next six months we spent countless hours at Children's Hospital of Dallas. Lane would have a blue spell and we'd rush him to the hospital, where he'd receive lots of oxygen and undergo test after test. He was not gaining any weight. He was not tolerating his feedings. What could be wrong? Lane underwent dilations, bronchoscopes, and pH probes. He was tested for cystic fibrosis, whooping cough, Down syndrome—all negative. What then, why was he having such a hard time eating and breathing? You can't live without those two things!

By this time Lane had undergone 12 major and minor surgeries. His little body looked like a road map from all of the scarring. Finally, in April 2000, Lane underwent a Nissan fundoplication to see if that would help. The doctors thought that Lane might be aspirating even though the studies didn't indicate it. The surgery went well and soon Lane was able to come home. But two weeks later, we were back at the hospital with another blue spell. The doctors had some more bad news.

They explained that Lane's esophagus was not effectively moving food down to the stomach. As a result, the food was creating a bulge in the esophagus and pressing into the trachea, causing the trachea to collapse. That resulted in the blue spells and pneumonia after pneumonia. The doctors recommended a Mic–Key button. A what? we wondered. It was a feeding tube that would be placed in Lane's stomach, bypassing his esophagus and giving him a chance to grow and mature. My husband and I were not sure we wanted to put Lane through another experiment. The doctors convinced us that this would help Lane gain weight and give his esophagus and trachea a break. So finally we agreed to the surgery and Lane got a Mic–Key button. The post–op period was horrible. Lane was having a hard time breathing on his own. His oxygen saturation rates were in the 80s. Back he went to the ICU.

My husband and I just didn't know what to do any more. We felt lost. This time, when Lane came home, lots of equipment came home too: feeding pump, oxygen, oxygen saturation (pulse ox) machine, portable suction kit, nebulizer, and a vibrating vest. His nursery looked like a hospital room. My husband and I received *Doctor 101* classes very quickly in order to take care of our son. The hardest thing I had to do was to stick an 8 French suction catheter down Lane's nose to help him breath. Lane needed this every 4 hours, along with

The hospital was our second home. We would joke with the staff and tell them that we owned room 456 and when we were not there we would rent it out to others to make some money.

vest and breathing treatments, and feedings through his new Mic–Key button.

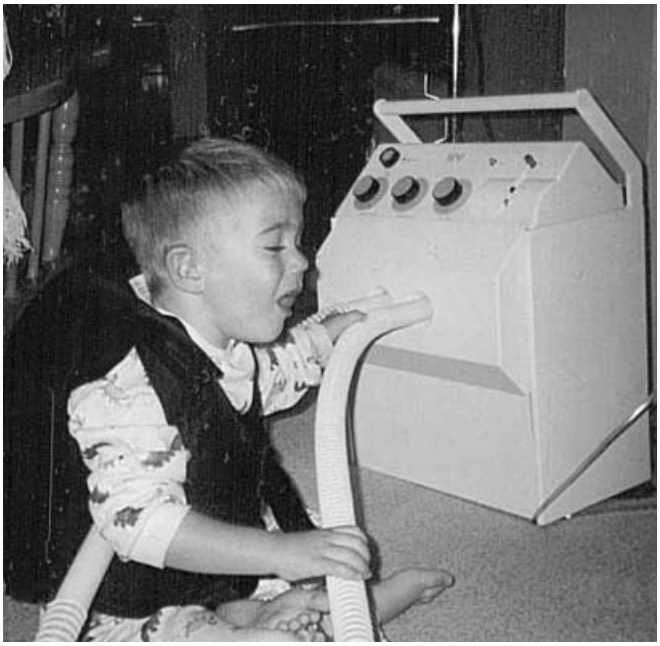
Over the next year Lane remained very ill. He still had blue spells and feeding problems, and he spent months in and out of the hospital. The hospital was our second home. We would joke with the staff and tell them that we owned room 456 and when we were not there we would rent it out to others to make some money. (You have to have a little humor or you will go crazy!)

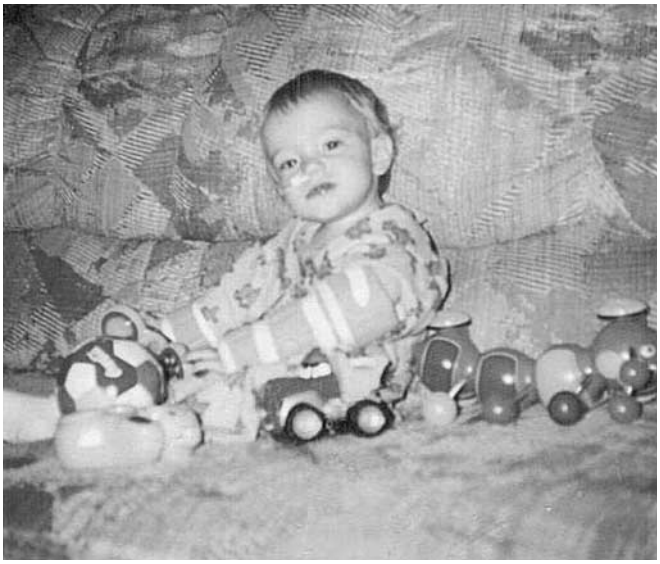
We'd been told before that Lane had tracheomalacia (a weakening of the tracheal wall, particularly the cartilage rings). After reviewing the last two years, Lane's pulmonologist suggested that Lane's tracheomalacia was severe and recommended an aortapexy. *[Editor's note: An aortapexy is a surgical procedure in which stitches (sutures) are placed between the front wall of the aortic arch (the part of the aorta that crosses the midline as it leaves the left heart) and the breastbone (sternum). The trachea lies between the aortic arch and the esophagus and all three are intimately attached to each other. By pulling the aorta forward, it pulls the front wall of the trachea forward as well, preventing collapse.]*

Again, another surgery. But this time the surgery, on Sept. 7, 2001, went great. Lane is post–aortapexy almost a year now and he has had only one hospitalization. We were home for the very first time for Christmas, New Year's, and Easter! We are praying that next winter is as good as this summer has been.

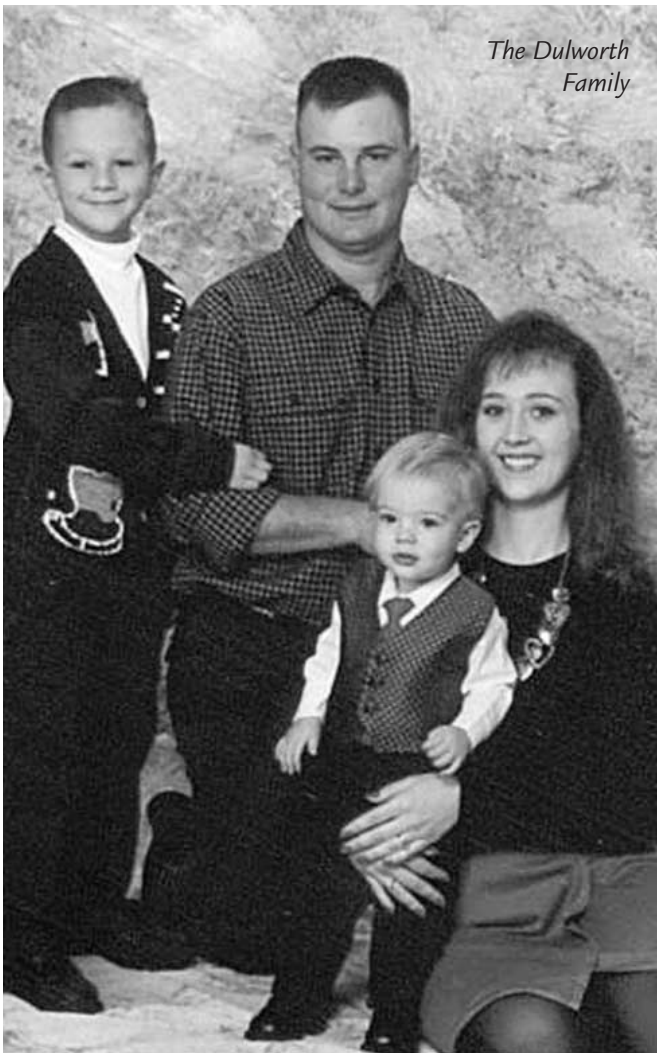
It has been a long three years but Lane is doing great now. He still has his Mic–Key button but that doesn't slow him down and he has begun to eat table foods. He is in a gymnastics class, and when he is not tumbling there, he is running and climbing at full speed at home. In fact, he has mastered climbing our refrigerator to the top to get his own bananas! Despite all the scarring, which will be a bragging tool for him to use when he is older, Lane is going at full speed.

Lane has showed and taught me things I never thought I would be able to do. God has given me a miracle, and has given my husband and me the strength each and every day to take on the challenges. When things were so low and I thought that things could not get worse, God would show me that all things are possible through Him.





Lane getting his pulmonary vest treatment (above) and a pH probe test (below) with no, nos on!!



The Dulworth Family

PS: I did not want to take away from Lane's story, but there is more to this story. Lane lay in a hospital bed fighting for air each and every time we went to the hospital. I could never leave him. However, in December 1999, when Lane was 15 months old, I was diagnosed with colon cancer. I was hospitalized and underwent radiation and chemotherapy for six months. My husband would have both of us in the hospital, 20 miles apart. Thankfully, things are better now, if not completely normal: I am healthier, Lane is healthier.

If you are a mom, dad, brother, sister, grandma, or grandpa of a child like Lane and you are feeling lost, take a minute to stop and savor the moment. That has been a hard lesson for us to learn, but through all the hardships and lessons over the last three years, I have learned to take each day, each minute for what it is worth, to stop and take a breath, to enjoy what I have instead of pining for what I wished I had. I don't know what is in store for us each day, but I do know that God always has our best interest at heart.

If anyone out there is going through a hard time please feel free to contact me at anytime, day or night, email, or even call me collect. I will try my hardest to help in any way that I can.

If you want to contact Amy Dulworth, you can write to her at 213 Larkspur Drive, Forney, Texas 75126; or call her at (972) 564-9068. Her email address is davamkala1@aol.com.

(Editors' Letter, continued from page 2)

Heather Fowlie lives outside of Vancouver, British Columbia, Canada, and is the mother of two gorgeous children. Quinn (5) who was born with EA|TEF and a laryngotracheoesophageal cleft, type III, and his baby sister Laine, who was born April 12, 2002. (Quinn's story was featured in the Summer 1999 issue.) Quinn is about to rock the local school as he enters kindergarten. They have never had a student with a tracheostomy and gastrostomy before, let alone a student who came to school with a full–time registered nurse.

We are both looking forward to sharing stories, information and resources with you. We can't wait to get your letters and emails with your personal stories and suggestions for topics you'd like to see in the newsletter. Do you have questions about EA|TEF and its related challenges that you think other families might be able to answer? Have you found helpful web sites or places on the Internet where EA|TEF families are meeting and exchanging information? We'd love to hear those too. ea|tef Child and Family Support Connection is all about making connections between families and we hope this journal helps that happen.

One last request. Please email us (eatef@msn.com) your current email address. Also, if you know any EA|TEF families who aren't on our mailing list, please have them contact us so that we can send them this journal and upcoming issues.

—Elizabeth McNamara and Heather Fowlie

"Nobody can really guarantee the future. The best we can do is size up the chances, calculate the risks involved, estimate our ability to deal with them and make our plans with confidence."

—Henry Ford II

The Little Bird:
Jenna's Story

By Linda Dolezan

Jenna was born on July 12, 1999, with EA|TEF. There are several types of EA|TEF; Jenna was born with the most common variety, in which the lower part of her esophagus (the distal esophagus) grew into her trachea during the early weeks of development in the womb. This is her story.

Monday, July 12, 1999: Jenna was born at Lakeland Hospital, in St. Joseph, Michigan. Transferred to Bronson Hospital in Kalamazoo.

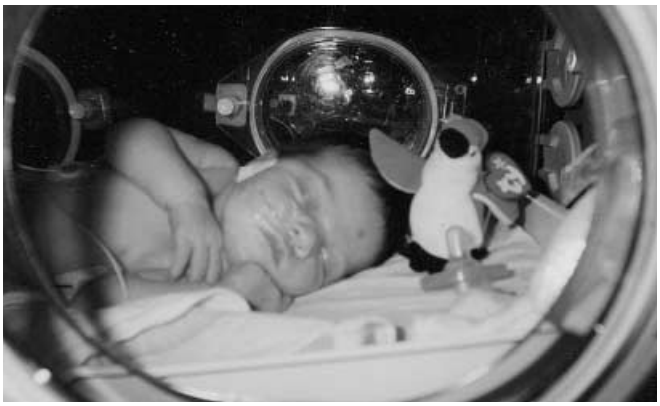
Tuesday, July 13: Tests were run to check for other defects common with TEF babies—heart, kidneys, intestines—aside from the TEF, Jenna was a perfectly normal little girl. At four o'clock, my husband, Leonard, called me at Lakeland Hospital to say that Jenna had gone in for surgery. The prayers were intensified and so were the tears. Her surgery took about five hours. She did pretty well considering she was only one day old. Her surgeon, Dr. Dahman, was concerned with the connection site. He said that he did not have much to work with in connecting the esophagus and that he had to really stretch the tissue.

Dr. Dahman went into Jenna's little body under her right arm near her ribs. After surgery, Jenna was on a neuromuscular paralysis agent to prevent her from moving and endangering the stitches that were holding her esophagus together. She was also put on morphine for pain. Jenna also had to have her chin placed down completely on her chest. She was placed on a ventilator, fed through an I.V. and had tubes going everywhere. We were told that she would remain like this for five days.

Leonard was there for the surgery. I was still in the postpartum unit in St. Joseph, going crazy. Leonard warned me what to expect, but nothing could prepare me for what I would see.

Wednesday, July 14: I was released from the hospital. We went home to pick up a few things and then drove straight to Bronson. I needed a wheelchair because I hadn't completely regained my strength. It was painful for me to take more than a couple of steps at a time.

Leonard rolled me into the NICU. I looked all around the room. There were three other babies in the unit. I looked in all directions. Where is my baby? I was so nervous, I didn't realize that my Jenna was the only full–term baby there. Gretchen, Jenna's nurse, met me about 5 feet from Jenna's



bed, a heating table. Gretchen explained that my first impression of what I saw would be scary, but to realize that all the tubes and wires were helping her. I got up from my chair and walked closer to Jenna. What a sight! There was this little angel with wires connected to her from her head to her ankles. Tubes for everything—breathing, feeding, medicines, drains, tubes, tubes, tubes! Wires were monitoring everything from her heartbeat to her temperature. She was a brownish– yellow color. Her eyes were puffed up with fluid, her whole face was swollen. Because she couldn't move, her body was retaining fluid. A ventilator tube and a suction catheter were taped across her little lips and her mouth was puffy too. I couldn't hold back the tears. There was my little angel, helpless. I felt helpless as well. I couldn't even feed her.

Jenna's name means "Little Bird" in Arabic. She looked like a little bird, a very sick little bird. One thing I could do was to pump my breast milk so that Jenna would at least have that nutritional advantage. It also gave me some feeling of importance—I was doing this for my baby. I needed to give her every fighting chance, and with what I had read, breast milk is the best thing you can do for your baby. If I could do it, I would.

I looked at all of the equipment. Wow, I would need four years of school to fully understand it all! I looked over at the ventilator and noticed that it was marked with the words "VIP BIRD." I guessed that was some kind of identification from the manufacturer, but to me it was a sign.

My "little bird" was a "VIP BIRD." I was still crying as Gretchen put an arm around my shoulder and began to explain all of the equipment. After that, I just stood there and stared at Jenna, tears pouring down my face. I glanced over at Gretchen for a moment and realized that she was also tearing up. I realized that Gretchen was a very special person with a big heart. I believe she was an angel disguise. Leonard was right there with me and we comforted each other and stayed for hours as we continued to do each day she was there.

Initially, we were told that we could expect Jenna to be hospitalized for at least a month. The nurses in the neonatal intensive care unit (NICU) said that most babies stay longer. I considered going back to work so that I wouldn't lose my Family Medical Leave Act time, but it was more important to me to be with Jenna.

One thing I can do is to pump my breast milk so that Jenna will at least have that nutritional advantage.

Thursday, July 15: This day was not good. Jenna developed pneumonia and one of her lungs was filled with fluid. The neonatologist told Leonard and me that Jenna was in very fragile condition. As soon as we saw her, we knew that something was wrong. Her color was grayish. Jenna had to be turned from side to side by the hour so that more fluid did not accumulate in her lung. Although we hadn't stopped praying, at this point we prayed even harder. Jenna stabilized by evening. This was Jenna's worst day.

We visited every day for eight to ten hours. We invited family to come visit Jenna and let her know that we all loved her very much and that she had a reason for being on this earth.

While Jenna was on the paralyzing medication (Pavulon), we watched her intensely. Each day that passed, they had to increase her dose and frequency. Because Jenna's little body was becoming used to the medication, what started as a dose every three or four hours ended up being given every one to two hours. If she were to move during this healing process, she could tear her stitches and have to go back into surgery. We watched for signs that Jenna was coming out of the medication. First we would notice her tongue moving ever so slightly in a sucking motion. Then it would progress to twitching toes and fingers. We would alert the nurses and another dose would be quickly administered.

Friday, July 16: No change today, except for the diapers! The nurses allowed Leonard and me to become more involved by changing Jenna's diapers, taking her temperature and blood pressure and assisting with suctioning out her lungs. Changing Jenna's diapers was a tough task because she was not to change positions. Jaundice levels were up, but still no need for alarm, the doctors said. Today one of the other babies in the unit passed away. I prayed for the family.

Saturday, July 17: Still no change, just watching her. She was so beautiful. Jaundice levels still up. Family members of the other babies in the unit looked over at Jenna and appear puzzled. I imagined they were wondering what could be wrong with such a big, "healthy looking" baby.

Sunday, July 18: Finally, the surgeon decided that Jenna could be taken off the Pavulon. We did not expect the surgeon to be concerned on a Sunday. But he made the phone call, and the Pavulon was stopped. We would be able to see our little girl move. That would make us feel much better. She came off of the medication very slowly, as expected. But with each move and twitch, we were that much closer to holding our little girl. She would still be on morphine however. She needed to be weaned off it because her little body had become dependent.

Monday, July 19: We arrived today and it took us a minute to realize they had taken Jenna off the ventilator. No more breathing tube! She was strong enough. Dr. Dahman came in to look at Jenna. Her chest x–rays looked good.

He took off the bandages to check the incision site. He looked at it for a second, got some suture scissors and removed the stitch that held the drain tube in place, and now it too came out. They also wanted to remove her naval I.V. but they needed to make sure that they could feed a tiny little tube from one of the large arteries to her main artery to get her the medication she still needed. They made us leave the room for the procedure. When we were called back in, they told us that they could not place the tube, Jenna's veins were rolling. Thank goodness. I wasn't comfortable with the procedure anyway. But when I looked at Jenna's little hands and feet, I realize that they had taped her down so that she would not move during the procedure. I sort of understood, but I regreted the layer of skin that came off when they removed the tape. Poor baby, she had been through enough already. It just didn't seem fair that they would even want to put her through that.

Today they also placed a feeding tube in her nose down to her stomach, what they call a nasal–gastric (NG) tube. Starting with 15 ccs (1 tablespoon), Jenna began to take my milk.

And finally I got to hold Jenna for the first time since she was born. Wow, what an incredible feeling! All the monitors and things made it a little difficult and I had to be sure to keep her head in a down position (I was not to stretch her neck), but, finally, my baby was in my arms.

Tuesday, July 20: Everything looked so good! I couldn't believe they thought Jenna would have to be here for another three weeks. That's what we were initially told, that Jenna would be here for a month or more. But today they increased

(Continued on page18)

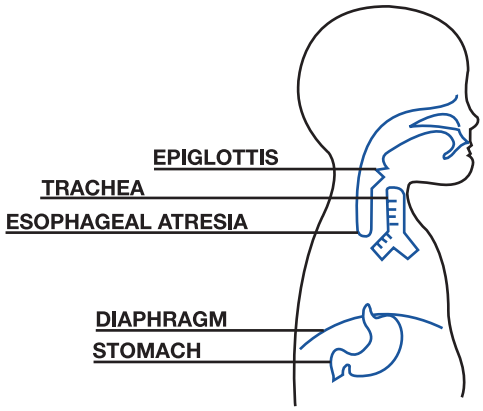
What is EA|TEF ?

First, esophageal atresia (EA): The esophagus (or food-pipe) carries food from the mouth to the stomach. An atresia is an absence or closure of a natural passage of the body. So, in EA, there is a gap in the esophagus so that food and saliva can not pass into the stomach.

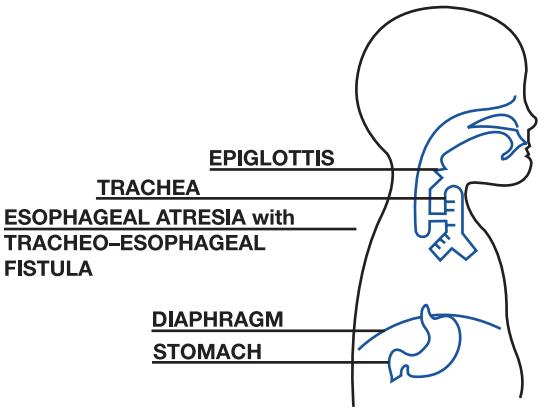
Second, tracheoesophageal fistula (TEF): The trachea (or windpipe) is the passageway through which the air we breath travels to the lungs. In TEF, there is a connection between the trachea and the esophagus, so that potentially either air can pass from the airway into the stomach, or food can pass from the esophagus into the lungs.

Approximately 1 in 3,000 babies is born with one or both of these life–threatening defects. Nobody knows what causes EA|TEF.

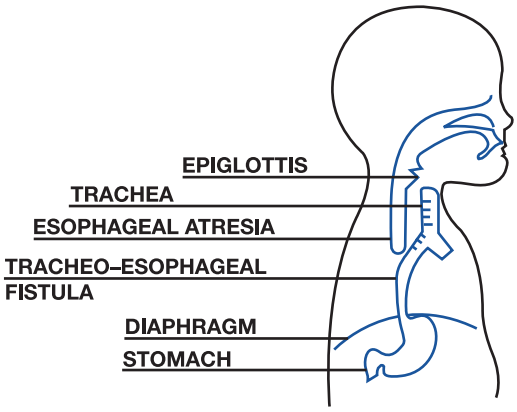
Below are diagrams of the five different types of EA|TEF.



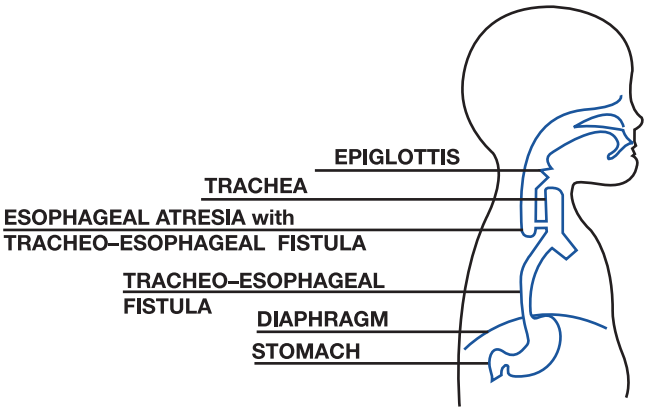
Type A Esophageal Atresia (7.7%) Both segments of the esophagus end in blind pouches. Neither segment of esophagus is attached to the trachea.



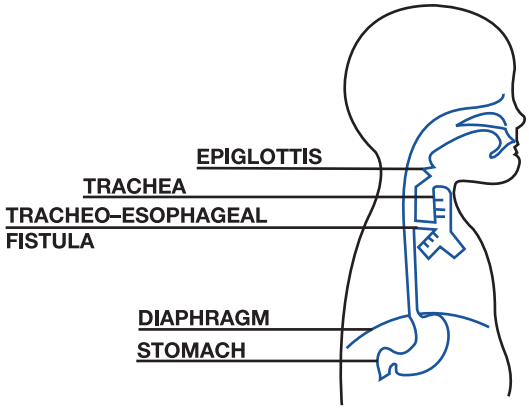
Type B Esophageal Atresia with Tracheoesophageal Fistula (0.8%) The upper segment of the esophagus forms a fistula to the trachea (TEF). The lower segment of the esophagus ends in a blind pouch (EA). This is a very rare form of EA|TEF.



Type C Esophageal Atresia with Tracheoesophageal Fistula (86.5%) The upper segment of the esophagus ends in a blind pouch (EA). The lower segment of the esophagus is attached to the trachea (TEF). This is the most common type of EA|TEF.



Type D Esophageal Atresia with Tracheoesophageal Fistula (0.7%) Both segments of the esophagus are attached to the trachea. This is the rarest form of EA|TEF.



Type H Tracheoesophageal Fistula (4.2%) There is no esophageal atresia as the esophagus is continuous to the stomach. However, fistula is present between the esophagus and the trachea.

(Rothenberg article, continued from page 1)

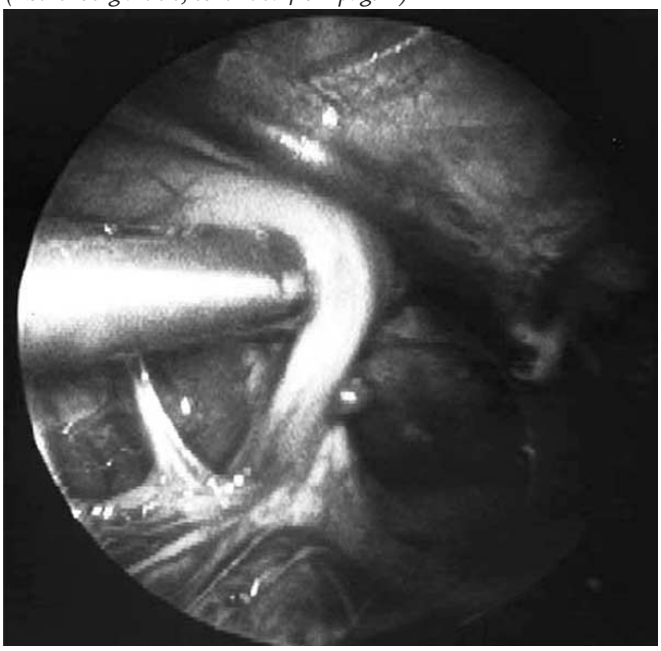


Fig 1. Mobilization of the lower esophageal segment. Fistula is seen going into the membranous trachea.

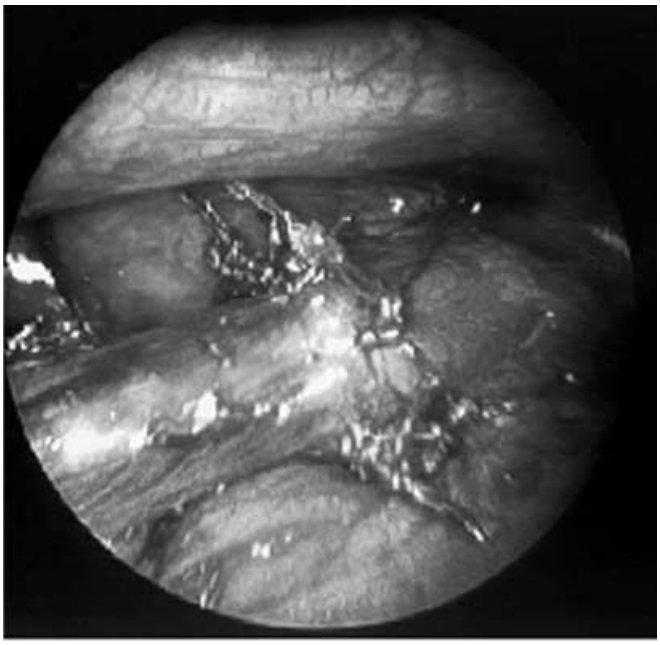


Fig 2. Completed anastomosis.

increasing respiratory distress.

The gap length was estimated preoperatively based on the position of the tip of the nasogastric tube and the apparent bifurcation of the trachea as seen on the chest x–ray. This ranged from 2 to 3½ vertebral bodies. At the time of surgery the longest gap was closer to 4 vertebral bodies because this patient had a trifurcation type fistula.

Six other patients with TEF were referred to our practice during the study period and underwent repair via a standard thoracotomy by surgeons not currently performing the procedure thoracoscopically. All patients underwent standard tracheal intubation with no attempt made to obtain single lung ventilation. Each patient was placed prone with the right side elevated 30°. One patient,with a right–sided aortic arch, was approached through the left chest with the left side elevated. This positioning provided access to the area between the anterior and posterior axillary line for trocar placement while allowing gravity to retract the lung. The chest was entered in the fifth intercostal space between the mid and posterior axillary line with a veres needle. The pleural space was insufflated to a pressure of 4mm Hg at a flow of 1L/min of CO_2. This caused the collapse of the right lung and essentially achieved one–lung ventilation. An initial 3mm port was placed, and a 2.7mm, 30° lens was used to survey the thoracic cavity. The Azygos vein was visualized and used to determine the relative position of the fistula.

A total of 3 ports, 2, 3mm and 1, 5mm were placed to perform the operation. In the initial case a fourth port was necessary to place a lung retractor. The 5mm port was placed superiorly and was used to introduce the Ligasure (Valleylab, Boulder, CO) device, surgical clips, and suture for the anastomosis.

The operation proceeded in a similar fashion to that in the

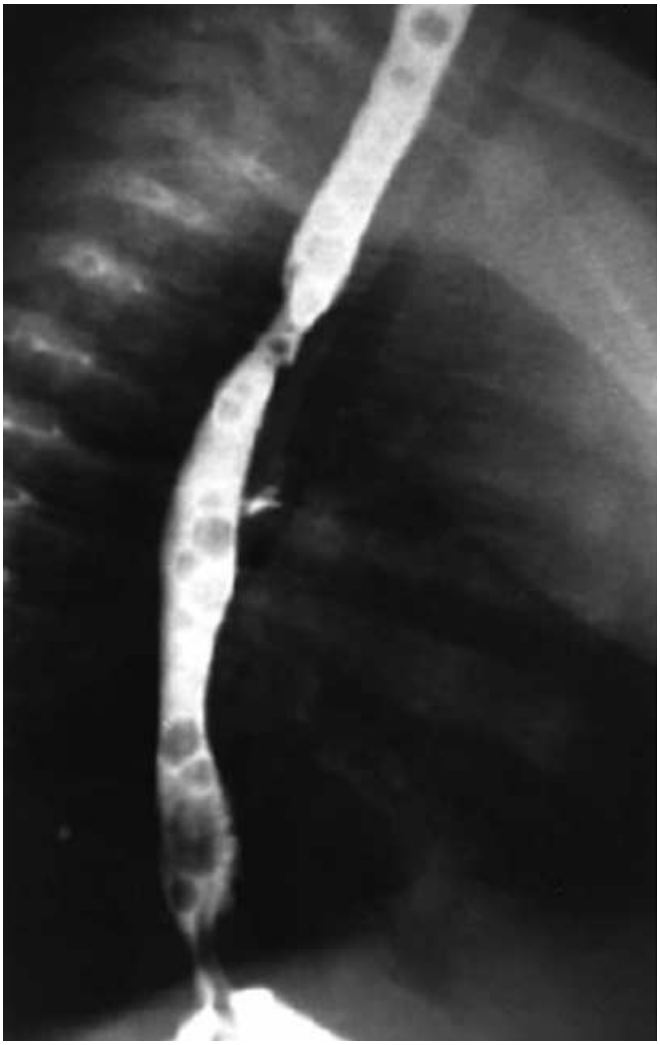


Fig 3. Esophagram 5 days postoperatively.

open technique. The Azygos vein was sealed with the Ligasure and divided. The pleura was incised, and the upper and lower esophageal segments identified. The fistula was mobilized (Fig 1) and closed proximally with two 5mm titanium clips. The fistula was divided distally, and the lower esophageal segment was mobilized for a short distance.

The upper pouch was identified with the help of the anesthesiologist who placed downward pressure on the nasogastric tube. The common wall between the trachea and esophagus was divided sharply taking care not to injure the membranous portion of the trachea. The upper pouch was mobilized up to the thoracic inlet. An initial stay suture of 4-0 Ethibond was placed between the 2 esophageal ends and was used to approximate the upper and lower pouch. An esophagotomy was made in the upper pouch so that the lumen could be visualized in both segments. A posterolateral row of approximately 4 interrupted sutures was placed. With the posterior row intact, the nasogastric tube was passed under direct vision into the lower segment and on into the stomach. By placing traction on the previously applied stitch, the esophagus was rotated right and left to provide exposure of the medial and anterior walls so that the anastomosis could be completed (Fig 2). A single 10F chest tube was placed through the lower trocar site with the tip near the anastomosis. The other trocar sites were closed with absorbable suture.

RESULTS

All procedures were accomplished successfully thoracoscopically. The average operative time was 95 minutes (range, 55 to 120 minutes). Four patients were extubated on postoperative day (pod) 1, two on pod 2, and 1 each on pod 3 and 4. One patient required reintubation 4 hours postextubation for increasing respiratory distress (patient 1). He was reextubated on pod 3. The fifth patient had an extremely long gap (4 vertebral bodies) with significant tension on the anastomosis. He was kept sedated and intubated for 4 days. Esophageal contrast studies were obtained on pod 5 in 7 patients, and the anastomosis was patent with no evidence of a leak in each case (Fig 3). The first patient had clinical evidence of a leak, saliva in the chest tube, on pod 4. He was kept on "nothing by mouth" status, and drainage stopped after 24 hours. He was studied on day 8 with no evidence of a leak.

At initiation of oral feedings 2 patients were found to have poor oral motor coordination resulting in primary aspiration. These 2 underwent a laparoscopic Nissen fundoplication and placement of a gastrostomy tube at 3 weeks of age to protect their airway. Each later required 2 esophageal dilations during the period when their oral intake was limited. Both were on full oral feedings with no evidence of anastomotic stricture at 3 and 6 months of age, respectively. The other 6 patients all tolerated initial oral feedings without problems, but 1 required a single esophageal dilatation and a second, now 4 months out, still is requiring intermittent dilations. Both of these patients have gone on to need a laparoscopic fundoplication because of severe gastroesophageal reflux but did not require gastrostomy tubes. The eighth case, the patient with the right–sided arch and tetralogy of Falot, initially did well but had a cardiac decompensation on the eighth postoperative day requiring emergent shunt surgery. Initially he required nasogastric feedings for approximately 2 weeks postoperatively but now is tolerating full oral feedings.

DISCUSSION

Recent advances in minimally invasive surgery (MIS) in infants and children have allowed for a wide expansion of applications over the last few years. Procedures that were thought impossible previously in children, let alone neonates, have become commonplace at a number of pediatric centers. We first reported our experience in infants less than 5kg 4 years ago. That study found that complex procedures such as Nissen fundoplications, bowel resections, and thoracic procedures, such as lung biopsy and PDA ligation,were not

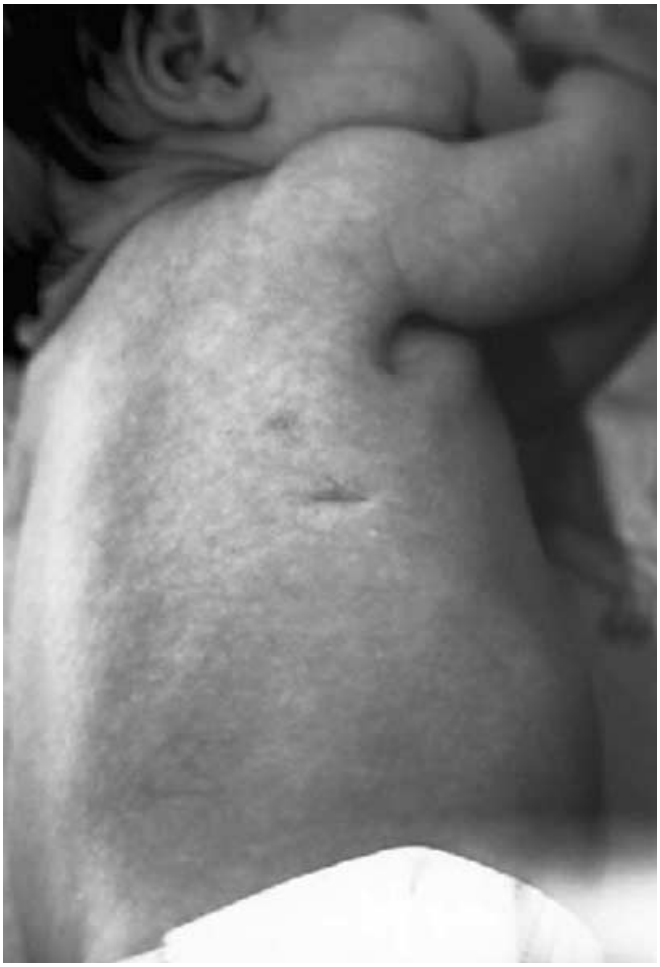


Fig 4. Incisions 10 days postoperatively. The lower trocar site is where the chest tube was placed.

The benefits of performing a TEF using minimally invasive techniques are obvious, but the technical hurdles are many.

only possible but were associated with less morbidity than standard open techniques. Others have reported similar positive findings. Fujimoto et al[2] reported that there is a decreased stress response as measured by interleukin-2 and other stress mediators when MIS techniques are used in infants, and this correlated with improved clinical outcomes. Concerns that neonates could not tolerate, or would be adversely affected by, abdominal insufflation or single–lung ventilation has not been borne out.

With these encouraging results, the impetus to develop the tools and techniques to perform even the most complicated procedures in neonates became a driving force. Much of the needed technological development has been directed at providing the instrumentation to perform minimally invasive cardiac bypass surgery (Mini-CAB). The use of robotics also has been touted as a way to perform these finer and more delicate dissections and anastomoses, but the current models are much too bulky and expensive to warrant their routine use in neonates.

The benefits of performing a TEF using minimally invasive techniques are obvious, but the technical hurdles are many. The greatest advantage is avoiding a thoracotomy in a neonate. This has been shown to be associated with a high degree of scoliosis and shoulder girdle weakness later in development.[6] Another is the improved cosmetic result (Fig 4). An unanticipated benefit was the superior visualization of the anatomy and especially the fistula. Because the fistula was visualized perpendicular to its connection to the membranous trachea, the exact site for ligation could be identified easily, minimizing the residual pouch attached to the trachea. The use of the 5mm titanium clips has proven to be simple and effective with no evidence of tracheal leak or recurrent fistula. We have adopted this method of fistula ligation in the few open procedures we have performed in the last 6 months.

A recognized advantage after the first case was performing the dissection and anastomosis in situ. Because the separation of the fistula and the upper pouch from the trachea was performed under direct magnified vision from a lateral approach there was little manipulation or force applied to the trachea itself. This may help diminish the degree of tracheomalacia that these children have postoperatively. Also, the plane between the upper pouch and trachea was more obvious, making injury to the membranous wall of the trachea less likely.

Another advantage of performing the anastomosis in situ may be less tension on the esophageal ends allowing longer gaps to be brought together without tearing. This appeared to be the case in one patient with a trifurcation fistula and long gap, although an anastomotic stricture that required dilation did develop.

The major technical hurdle in this operation is the suturing of the anastomosis. The placement of the sutures and knot tying are technically demanding and relatively imprecise. Also, as opposed to the open technique in which the entire posterior row of sutures can be placed and then brought together to disperse the tension along multiple points during knot tying, this method places all the tension on one suture at a time. So far,this has not been a significant problem, but it could prove to be. For this procedure to become more widely accepted it maybe necessary to develop a mechanical anastomotic device or self–knotting suture.

The rate of anastomotic narrowing requiring at least 1 dilatation in this series is 50%, a rate somewhat higher than historical controls. This may be secondary to inadequate approximation of the mucosal ends or an insufficient opening being made in the upper pouch. The type of suture being used also may be contributing to the problem. All of these variables are being reviewed.

Clearly, the technical and physiologic hurdles to accomplish this type of repair are many, and it will require continued advances before this surgery becomes commonplace. However,the ability to perform this complex reconstruction without a thoracotomy lays further ground work in minimizing surgical morbidity in even the smallest pediatric patients.

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Q&A with Dr. Rothenberg

Q *Is this surgery for everyone?*

A Probably not but we are doing smaller and sicker babies with good results. A thoracotomy [Ed. Note: the standard repair technique made with an incision under the arm] is still the gold standard for repair of TEF. Early results with thoracoscopy are promising and I think eventually this will be the way we routinely perform the procedure. But for now I would not recommend that the parents go to extremes to get to someone who can do the procedure thoracoscopically. If they live in a community where a surgeon is experienced in the thoracoscopic technique I certainly think it is reasonable for them to seek out or request this person to determine the risks and benefits in their child's case.

If they know of a TEF through prenatal diagnosis then it is up to them how agressive they want to be in pursuing this option. Remember, the relative outcome of the esophagus is probably going to be the same. The major advantage is avoiding the thoracotomy and its associated long–term risks.

Q *Is it feasible to transport a newborn with EA/TEF to a hospital where there's a doctor well–versed in thoracoscopy?*

A A newborn could be transported if the baby were stable. Babies with TEF are transported all the time from hospitals that do not have adequate pediatric surgical and neonatal expertise. They should not, however, transport the baby any significant distance to a different center as the risk–benefit ratio is just not there yet.

Q *How many of these surgeries have you done?*

A The number I've done is now 16. As many as 80 to 100 babies have undergone the surgery worldwide.

Q *Your article suggests that there may be a link between tracheomalacia and a thoracotomy. Isn't tracheomalacia caused in utero?*

A The link isn't between thoracotomy and tracheomalacia, it's between manipulation of the trachea during surgery and tracheomalacia. I think a thoracoscopic approach allows us to manipulate the trachea to a lesser degree.

Q *Are there any indications so far that thoracoscopically repaired EA/TEF babies need fewer esophageal dilitations post op?*

A There is not enough data yet to comment on the need for dilation.

Q *Is it yet known whether a thoracoscopically repaired child will on average have a shorter hospitalization?*

A The advantage of a thoracoscopic approach may not be a decreased hospital stay (you still have to wait for the esophagus to heal before you can start feeds) but the long–term benefits of avoiding a thoracotomy as an infant. Thoracotomy risks include an increased risk of scoliosis, muscle weakness and chest wall asymmetry.

Mail Bag

I am a 42-year-old "more than just a survivor" of EA|TEF. I have had a wonderful life. Normal for me has been pretty close to normal for most others. Yes, I have had a lot of surgeries, but mostly as a baby and so I do not remember the trauma. I have developed some complications in the last few years, I developed diverticulitis in my colon transplant and was aspirating a lot. It caused me to be short of breath, very limited in my activity, and of course, if not treated, it was life threatening. But I have been able to apprehend a very simple and acceptable solution. I had surgery to place a jejunostomy feeding tube *[editor's note: a J-tube is a feeding tube that goes directly into the jejunum, in the small intestine, bypassing the stomach]* in December of 2000.

I have since been able to resume a normal lifestyle and balance tube feeding with normal eating so as to enjoy life and health. I seem to be an oddity to the doctors, but I wonder if there are not others out there who could benefit from my experience. I have never met another person with this condition. It might be interesting, encouraging, and helpful if I did. My main interest is to share what has helped me because I have run into some specialists in this medical field who gave me no hope. It seems very strange to me that the solution is so simple and successful, yet was viewed as not feasible by one renowned gastroenterologist I consulted.

I have had such a normal life for the most part that I want to encourage the parents of these children. I have two grown sons!

Recently I got the chance to visit with the doctor (Dr. David Akers) who did all my surgeries when I was a baby. He is now 94 (or 95?) and of course has been retired for a long time, but we have sporadically kept in touch. I remember as I was growing up that I'd get a birthday and Christmas card from him every year. I hope that it blesses him as much as it does me to be able to see the quality of life that I've had, and to know that God used him in a significant way to give it to me.

I've had 11 surgeries. I know that some of those were to repair things that didn't work out well and caused problems or to try something different. Dr. Akers told me they learned a lot on me. The colon transplant procedure was still being pioneered. I have a lot of questions about the procedures done now and how they have changed. I was told by a gastroenterologist in Denver that the colon transplant is still done quite often.

When I was trying to find information on my condition and the complications I have experienced as an adult, I was unable to find anything. Are there other adult survivors out there? I have already received a welcome packet from your organization and I very much appreciate it. When I read through it, it was the first time in my life that I heard of other people and families who I knew could relate to that part of my life. But I have had such a normal life for the most part that I want to encourage the parents of these children. I have two grown sons! I was always told that I would not be able to carry a child to full term because of the heavy scarring on my abdomen (I tend to scar heavily), but other than the minor problem of having to go on liquids for the last two months of each pregnancy due to the looping of my esophagus, I had no problems, having what my doctor called "textbook" births and healthy babies. I know that I have been very fortunate and I am grateful. I hope that this reaches those who may be helped by my story. Thank you for your help.

—Patty Nickolaus
genesisbotan@juno.com

Due to my recent "retirement," I am now in a position to help out the organization by donating time in some way. I am located in North Carolina. I'm not even sure if we have a state rep anymore or not. I am also a registered pharmacist if that will help out in some way. I've spent the past 18 years working for a pharmaceutical company and would be able to research any inquiries on medications if I didn't know the answers.

I would love to send in my story on my 18-month-old Jack someday when I've got more time. He was a long–gap pure atresia and after some talk of a possible gastric tube or pull–up by our local physician, we did the "long distance dance" to Minnesota, and Dr. John Foker. Thank goodness! The end result was a primary end–to–end anastomosis and Nissen fundoplication when Jack was 10 months old. Jack started eating by mouth and had his g–tube removed two months later. He's now 18 months old and weighs in at 23 pounds, a very long way from his birth weight of 2 pounds!

An interesting note, I ordered the book *The TOFS Child* from the British family support group web page on EA|TEF. The book was fabulous and gave excellent information on sham feeding. Unfortunately I found the book after Jack's spit fistula was already closed. I felt like we were flying by the seat of our pants when we started sham feeding Jack because no one gave us any ideas. This book could really help out a lot of people. (See related item on page 16.)

Let me know what I can do to help.

—Jo Bishop
JoBishop@carolina.rr.com

Several months ago you sent me the EA|TEF packet that had lots of useful info. My son is 20 months old w/VACTERL Association. I am now looking for information on choking. He choked on watermelon recently and had to be Life Flighted to the hospital. It was so terrible. I'm a physical therapist and have way too much medical knowledge.

Thankfully he is fine but I'm still recovering. Anyway, he had a modified Barium swallow and it looked great! Okay, so why did he choke? The gastroenterologist said we could go ahead and feed him crackers, cookies, and solids but I went back to baby foods. What if it happens again? I did get the Baby Safe Feeder at Toys–R–Us. Do you know of any resources or other moms I can connect with that have had more experience with this? I would appreciate any input.

—Fran Grann
egrann@net1plus.com

Hi! My name is Dana Hall. My son, Brandon, was born with long–gap EA in July 1992. He had surgery to construct an esophagus (with the distal portion of his stomach), in Jan. 1994, at 18 months of age. He is now a healthy, happy (and, might I say, handsome and intelligent) skateboarding fifth grader!

It's hard to believe he was a tiny, g–tube–fed baby. I found a support group shortly before his major surgery in 1994, but lost contact with them. Prior to that my husband and I felt so alone and uninformed!

I pray that no parents have to endure that experience—life with a child needing special care, surgeries, etc., is so difficult without feeling like you're all alone and that no one could possibly understand what you're going through!

Recently, when I discovered your web site I cried! I was so thankful to see that information and support exists for new parents! Keep up the great work! I would like to be involved in some way. Please email me if I can be of assistance.

—Dana Hall
ddhall@worldnet.att.net

Well, we don't know where to start… Our grandson Brody will be 5 years old on Nov. 8. Of course he is an EA|TEF child. He has had four or five major operations, is tube fed, has a port for intravenous, etc. Brody also has had at least 70 dilations, most recently today. The surgeon got him to 44 *[Ed. Note: 44 Fr., the width of the dilator]*, but said he cannot understand how Brody is able to eat at all as the stricture is so narrow. He still eats baby food, although he does nibble on other foods a little, cookies, chips, ice cream, etc. He is also fed Pediasure, two cans during the day and four at night. We do not understand why his esophagus is not improving. We have heard of other children whom only need a very few of these dilations. Brody has them every seven weeks. Can you offer any advice at all? We know his surgeon is doing his best. Please help. Thank you.

—R.A. Rozon, Saskatoon, Sk. Canada
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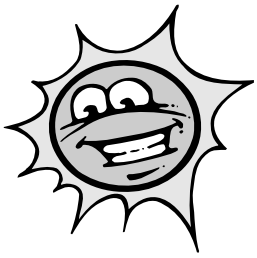
rbrozon@sasktel.net

ea|tef

Wants to Send A Little Sunshine Your Child's Way!



The ea|tef Child & Family Support Connection has a **Sunshine Program** to help bring a little happiness to the families in our group. Part of this program includes sending a special gift to children who are ill or hospitalized. If your child could use a little sunshine, please let us know (by phone, email, even U.S. Post!).

This is a special program for ALL of the familes in our group, from New York to California and beyond! The money we work so hard to raise is best spent when it warms the heart of a child and puts a smile on a parent's face. So, if you have sunshine news about your family or another family, we want to hear from you!

Call us at **(312) 987-9085**, email us at eatef@msn.com, or send us a card at 111 W. Jackson Blvd., Suite 1145, Chicago, Illinois 60604-3502.

We have a new Email address! Do you?

If you've tried to email us and your email keeps being returned to you, don't despair. We have a new address:

eatef@msn.com

Drop us a line and try it out! And while we're on the topic, we may not be the only one whose email has changed. If you too have made a switch please send us your new address. Thanks!

"If you worry about what might be, and wonder what might have been, you will ignore what is."

—Anonymous

Surfing the Web

OUR NEW & IMPROVED WEBSITE!

www.eatef.org

We are happy to announce that our website is revamped and better than ever, thanks to some hard work by EA|TEF parent Joan Crook. The website includes basic information on the different types of EA|TEF, information about the organization's resources (including Family Orientation Packets and booklets on topics like *A Guide to Caregivers* and *Switching from Tube Feedings*), family stories, and web links. Check it out and send us your feedback!

WEB PAGES FOR PATIENTS' FAMILIES

www.caringbridge.org

You and your child are stuck in the hospital, your family and friends are constantly trying to reach you to find out how everything's going and you've explained what a "fistula" is so many times you'll scream if you have to do it once more. There's help: a Minnesota–based nonprofit organization, Caring Bridge, provides technical support for people who want to set up Web pages to keep family and friends updated on a patient's progress.

The free service collects basic information to start a web page, then lets you select page templates for newborn, child or adult, and requires you to log in each time you post an update to the page. Password–protected, the site also allows people to post messages to individual pages, making it easier for those monitoring the Web page to communicate with you and your child.

TOFS: A BRITISH EA|TEF WEBSITE

www.tofs.org.uk

A terrific support group for families and caregivers of children with EA|TEF exists in England, but through its web site, families worldwide can benefit. TOFS stands for Tracheo–Oesophagael Fistula Support—note the difference in the spelling of esophagus.

The TOFS website is packed with information and the organization offers a variety of leaflets, including those with basic information about EA|TEF, ones that discuss more general concerns ("children in hospitals," for instance), leaflets that focus on more complex EA|TEF issues and ones that discuss related syndromes, such as VACTERL.

TOFS also publishes a newsletter cleverly titled "Chew," which you can find on the web site. It is also available in hard copy if you are a member of the organization.

Vicki Martin, the TOFS founder and herself an adult survivor of EA|TEF, has written a great book titled *The TOFS Child*. It covers everything from prenatal diagnosis to eating to long–term prospects. It is available through Amazon.com but you may have to go through the TOFS website to find it on Amazon. A basic Amazon title search came up empty, but clicking to Amazon via the TOFS site yielded results.

ASSOCIATION FRANÇAISE de L'ATRÉSIE de L'OESOPHAFE (AFAO)

www.afao.asso.fr

Hello! We are very pleased to announce the founding of the first French support group for families concerned with esophageal atresia: the AFAO (Association française de l'atrésie de l'oesophage). We have set up a website with the address: www.afao.asso.fr. (Ed. Note: use Internet Explorer.)

At the moment, our main target is to become well known and to get information about EA|TEF. (French doctors and surgeons are not very helpful and cooperative in suppling us with information. They are often very busy.)

Your site is a real gold mine for us! Here is the principal information about our group:

Name: Association française de l'atrésie de l 'oesophage (AFAO)
Languages: French and English
Address: 56 rue Cécile, 94700 Maisons ALfort, France
Telephone: 0143754249
Email: f.armand@afao.asso.fr or armafred@aol.com

Yours sincerely,
Frédéric et Viviane Armand

The inclusion of any website link (or resource accessed through a link) does not imply endorsement by ea|tef Child and Family Support Connection. *Seek the advice of your child's health care provider before you act or rely upon any information from these resources.*

Autumn's Story

Autumn was born on Nov. 15, 1999, with EA|TEF. The film Autumn's Story *was written, directed and produced by Stephanie Scholz Neurohr, Autumn's mother, for Discovery Health Channel and it premiered Feb. 3, 2002. It is available for purchase through www.motherof7.com.*

My name is Stephanie and I am the mother of seven children, a published author and a film producer who began documenting my tenth pregnancy and second set of twins for an educational video about pregnancy, birth and the art of nursing in May of 1999. I wanted to share my wealth of knowledge about motherhood with the world.

During the pregnancy, Twin B died in utero and my high–risk pregnancy began to unravel but I continued on with my documentation.

Minutes after birth, Autumn entered the world naturally and I immediately knew she had a problem after I nursed her. The colostrum backwashed and I yelled for help. My doctor was busy with paper work and answered with haunting words across the room, "You're probably drowning her with your milk."

The obstetrician seemed unimpressed, suctioned Autumn, and left the hospital. The nurse was concerned, called the pediatrician for help and Autumn was rushed to the neonatal intensive care unit where an EA|TEF was found.

Autumn's repair surgery proved to be a success but her left vocal cord was paralyzed when she stopped breathing and they had to perform an emergency intubation. All liquids silently aspirated into her lungs. The neonatologists and other doctors felt it would be unlikely that I would ever be able to nurse Autumn. EA|TEF babies often have discoordinated swallowing patterns and a paralyzed vocal cord compounded the problem. I was brokenhearted. Nursing had always been my natural way of meeting my babies' needs.

Days before Christmas, 1999, my husband and I finally took Autumn home from the NICU. I pumped milk every two hours so my milk would not dry up and I seldom slept; Autumn needed 24 hours around–the–clock care. Postpartum recovery was replaced by postpartum depression, panic and post–traumatic stress syndrome. Even though my life had fallen apart, God gave me the strength to help Autumn survive, hour by hour. I never left my baby for more than a few minutes each day when my husband was able to come home from work.

I would sometimes go out and sit in the car so I could sob or yell out in frustration for help. Six other children also needed us, but we struggled to pull together as a family to make it through.

After four months of nasal–gastic tube feedings with my breastmilk, a modified barium swallow showed remarkable improvement. I convinced the doctors to give us a trial run of breastfeeding. Autumn eagerly latched on and I carefully held her in a position in which the left paralyzed cord was up and away from harm. I pulled her off the breast when the flow became to strong. Autumn gradually learned to protect her airway.

Months after birth, I worked up the courage to watch Autumn's birth tape. I knew then that a documentary about the NICU had emerged and I vowed to finish the film for Autumn and for me. Discovery fell in love with *Autumn's Story* and I worked very hard to meet their high standards for network broadcast. I would drop six children off at school and take Autumn to work. She never left my side and grew up watching her story evolve.

Autumn is now 2½ years old. She is doing better but still chokes several times a day on solid food, has had two dilations of the esophagus, survived the RSV virus and pneumonias and needs to eat small amounts, frequently; her tracheomalacia is quite severe. I still nurse Autumn every two to three hours and that seems to keep both mother and child more relaxed and moving forward in a bonded, positive direction, especially after Autumn chokes and needs comfort and warmth from Mommy. I continue to struggle with post–traumatic stress syndrome and work to reduce my anxiety attacks which doctors tell me naturally follow any chronic medical crisis that is long and on–going.

Autumn's Story is honest, deep and heart rendering. I am just now, for the first time since Autumn's birth, able to reach out and find the strength to talk to other parents of children born with EA|TEF. My website is www.motherof7.com. I am available as a spokesperson and hope that this letter gives strength, compassion and inspiration to someone who is going through a similar situation.

On a happy note, I am beginning my educational film about motherhood and will parallel it with a book publication.

(Jenna's Story, continued from page 8)
Jenna's feedings by 5 ccs a feeding and Jenna was doing well. No vomiting at all.

Wednesday, July 21: Jenna was still doing very well taking my milk. The nurses even let me push her feedings (through the NG tube). Jenna's eyes were still too puffy for her to open them. I wanted so badly to look into her eyes!

Thursday, July 22: Today they let me attempt breastfeeding but it didn't go well. That made me very sad, but I won't give up yet. I'll try again tomorrow. Happily, she finally was able to open her eyes. They were still a beautiful blue. She looked at me as if she knew me.

Friday, July 23: No more naval I.V.! Yea! I breastfed Jenna five times today and she did much better with it. I had to take her off the breast every few seconds though because she would get too much and choke on it. She did not spit up once. In addition to that, the nurses had her all dressed up in one of the outfits I had brought. She looked so adorable! Finally she got to wear something more than a diaper!

Saturday, July 24: Leonard and I were getting a late start today. We needed some rest and since we felt that Jenna was out of the woods, we decided to sleep in. The house was a mess. The last 12 days had been spent getting up early, driving to Bronson, staying there the whole day, returning home exhausted and falling into bed. At 10:30 AM the phone rang. There was a pleasant voice at the other end of the line. It was Jenna's nurse, Gretchen. "Linda?" she said. "Yes," I replied nervously. Gretchen said she wanted to catch us before we left home because we needed to bring the infant car seat.

We could come and get our little Jenna. All I could say was, "Thank you," and I began to cry. Finally we could bring our little bird home! I could not believe it. I was shaking I was so happy. It had only been 12 days, but Jenna had been such a fighter that she was coming home more than two weeks earlier than anyone believed she would. They felt that Leonard and I had proved our dedication and that they were comfortable releasing her early.

We drove straight to Bronson. The one–hour drive felt like three. We were completely elated. We signed some discharge papers, received a few instructions, took some pictures of Jenna's friends the nurses, and brought our little girl home. As we pulled into the driveway, we looked up to see an "It's a Girl!" banner posted across the balcony. This was her welcome home party. Thanks to family our house was spotless, and everything was ready for Jenna.

We've had only one setback since Jenna came home. Three weeks later, when I was breastfeeding her and stopped to burp her, I placed her on my leg and leaned over her has I patted her back. At first it looked as if she was going to burp, but all of a sudden I realized that Jenna was trying to get a breath. I called for Leonard, who was napping. It took him a minute to realize that I really needed him and I didn't want to yell because it could startle Jenna and panic her even more. She kept looking at me with such frightened eyes. I panicked. I started crying hysterically. Her eyes appeared to say, "Help me, Mommy!" But I felt helpless. I didn't know what to do.

By the time Leonard came, I had laid Jenna on her back and was preparing to do CPR. I wondered how I would do it if she really had not stopped breathing but simply could not get her breath. Was there a difference? My mind filled with different thoughts in that short period of time. Then suddenly Jenna turned black around the mouth and eyes and as soon as that happened, she let out a cry. I knew that crying was good, that she could not cry without breathing. Her color returned immediately. Oh, thank God, Jenna was breathing! But what had caused this?

I called her doctor right away and we went for a trip back to Bronson. Jenna was placed under observation for two days. Her doctors said that some of the milk Jenna had swallowed had come up and tickled her vocal cords. That caused her vocal cords to clamp down on her windpipe, cutting off air flow. To try to avoid this from recurring, Jenna's surgeon instructed us to thicken my breast milk with cereal. This meant that I would have to pump for the duration of my breastfeeding, but at that point I would do anything to ensure that Jenna never did that again!

July 12, 2000: Jenna was now one year old. At her one–year visit, she weighed in at 26 lbs., 4 oz., and measured 32 inches. She was off the charts in all of her measurements. Jenna was healthy.

We still had to be careful with what we gave Jenna to eat, but don't all parents? She would have to learn to take small bites and to chew her food very well. She would need to drink a lot of fluids with her meals and eat slowly.

May 12, 2001: At 22 months old, Jenna gave no hint she'd had such a battle early on. The only visible proof was the scar under her arm. For the most part, Jenna ate like any other toddler. We did have to cut up her food into small pieces and she stayed away from "crunchy" vegetables and fruit.

But Jenna taught herself to chew her food well and she insisted on showing us when we asked her if she was chewing (fun, fun!).

May 7, 2002: Jenna is healthy and nearly 3 years old. I need to get her an identification card because she is bigger than most 4 year olds. Buffet restaurants that advertise children under 3 eat free want to charge us for her to eat. She occasionally has trouble with reflux and there are still some foods she struggles with, but all in all we are very fortunate.

I didn't submit this story to brag about Jenna's success with EA|TEF. I simply want to give hope to new parents of children with EA|TEF. There are success stories.

One final note: We would like to thank everyone for all the prayers Jenna received. We believe in the power of prayer.

Recent Donations:

We are grateful to all those who made donations to ea|tef Child & Family Support Connection during the past year. Your dollars make it possible to produce and distribute this journal and to send out New Family Packets. They make it possible to maintain our website and to bring a little "Sunshine" into our children's lives. Many thanks to those listed below:

Adam and Keelin Weiss

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Lynn Leatherman-Zimmershead

James Traina

Richard and Sharon Kenneavy

Ned and Delores Gruca

David and Cherie Leatherman

In Memoriam

We remember two beautiful boys who were born with EA|TEF and who died in the past year. Our condolences to the Testolin and Leatherman families.

Anthony Michael Testolin
Joshua Leatherman

The Cough

We thought it would go away.
Doctors said James would grow out of it.
Five years later, it's as strong as ever.

"Oh, he's a sick one, isn't he?" people will say.
"Shouldn't he be home in bed?" asks the supermarket cashier.
This in July, when James hasn't been sick in two months.

A faraway friend hears the telltale cough on our
answering machine.
"Did you get a dog?" she asks in her message.
It takes a few minutes to grasp her meaning.
No. No dog.
That's just James.

Follow the cough… he's a hard one to hide.
He'll grow out of it, they say.
Maybe.
Someday.

In the meantime,
we celebrate what our priest calls
James's
joyful
noise.

—E. McNamara

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