ea|tef

Child and Family Support Connection, Inc.

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

INSIDE

Madison's Story

by Melissa Orrick

My husband, Donald, and I have two beautiful daughters: Callie and Madison. Callie is 11 and Madi, as we call her, is 7. Callie was born in 1991—she arrived a month early—a little small but otherwise was perfect. And for the first seven months of my pregnancy with Madi, five years later, everything was very normal.

Then, on the night of April 19, 1995, our world started to change in ways we could never imagine. I'd come home early from work because I was not feeling well. I was hurting in my chest and as evening began to fall, I was feeling a lot worse. By 7 p.m. I felt so bad, I was barely aware of anything. It felt as if my mind was coming and going.

My husband and mother decided to take me on to the emergency room to see what was going on. Once there my regular doctor was called but he was out of town, so the doctor on call examined me and decided that I needed to be sent to Vanderbilt Children's Hospital as soon as possible. My blood pressure, platelet count and liver enzymes were all very high. I was taken by ambulance to Nashville.

I was getting worse by the minute. Once at Vanderbilt, things steadily went downhill. They tried to stop my labor but Madi went into distress and so did I. The nurses and doctors had already prepared my family for the worst: we both could die. That's when things went really crazy. I was rushed into emergency surgery and had a C-section. Donald said I wasn't gone for more than 10 minutes before they came back out and said the baby was a girl. She was even bigger than they first had thought, weighing a whopping 3 pounds, 8 ounces, and she was 15 inches long. I was already recovering. Everything seemed wonderful.

Madison arrived at 7:29 a.m. on a very stormy Saturday. Tornados had been sweeping through the area most of the night—we sometimes laugh now that Madi came in a whirlwind and that she's never stopped.

By the next morning, a doctor came by to tell us there was some type of a problem with Madi's feeding. They were doing some test to see what was wrong. I never gave it any serious thought. A few hours later another doctor came by, Dr. George Holcomb, a pediatric surgeon. I knew when he introduced himself that something was wrong. I still had not gotten to see my baby, so I was having a hard time dealing with the fact that my baby girl was sick.

Dr. Holcomb proceeded to tell us that Madison was born with esophageal atresia, without fistula. We had no idea what he was talking about. Just like so many of the parents who's stories we've since read, we had never heard of anything like this. And we too thought we were the only parents in the world having to deal with this. Not knowing what to ask or who to turn too, I just closed my eyes and told God that it was in his hands. Then I looked at the doctor and told him to take care of Madi.

From the very beginning Dr. Holcomb told us Madi was going to be just fine, not to worry. The next morning, Madi went in to have her g-tube placed, and we talked of the plan for the next few weeks. She was going to stay at Vanderbilt to grow—the doctor wanted her to reach a certain weight before he would do the surgery to connect the

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Editor's Letter

When you are a parent of a child with ea|tef, you learn all too quickly that life is very precious. Still, over time, the fear of those first few weeks wears off somewhat. After a surgery or two, you begin to realize that surgery isn't the end of the world. After a year or two, you find yourself secretly scoffing (perhaps) when a friend talks of how scared she is about her otherwise healthy 2-year-old's upcoming ear tube surgery. Or when a colleague complains that his 8-year-old has a stomach bug for the second time in three months.

Occasionally, I've found myself fighting the urge to shout, "Come on! Don't you realize that there are much worse things out there? Don't you know how lucky you are?" But, really, how different am I from every other parent in the world? Do any of us really, truly know how lucky we are? We are reminded of that in a beautifully written story on page 3 by Cherie Leatherman.

Cherie writes the story of her son, Joshua, who was born in 1996 with ea|tef. On March 27, 2002, Josh died in a tragic accident unconnected to his ea|tef. Josh's story reminded me yet again to treasure every moment, even the bad ones.

Shortly after receiving Josh's story, my son James, also an ea|tef child, took ill with what turned out to be a lethal strep infection. What began on a Tuesday with vomiting and diarrhea, turned by Wednesday into double pneumonia and acute respiratory distress. James went into septic shock and lay closer to death than he'd ever been before. All this after a reasonably healthy winter and a regular day at school the Monday before. My husband, Neal, and I were shocked out of our complacency and I felt complete desperation for the first time.

We were fortunate: James pulled through. Like Joshua Leatherman's accident, however, James's illness had nothing to do with his ea|tef. It could have happened to one of our other two children. But maybe we went to the hospital faster because it was James and we've learned the hard way to be more vigilant with him.

In true child form, James has bounced back into his life with vitality. I am left more fearful, perhaps, but with an even greater awareness of the daily miracles of life that surround me. When you read Cherie Leatherman's story, I think you will be too.

You'll learn about a new addition to the ea|tef web site in the "Surfing the Web" column (page 16). New web site guru (and ea|tef dad) Matt Parker has added a glossary of terms—always helpful in trying to put into everyday language the medicalese you learn in the hospital. More significantly, he has established web site discussion forums.

I visited the site recently and was amazed at the wealth of information and solicitude offered by other visitors. I was so embolded, in fact, that I posted my very first web site notice. You can sign in and participate, or you can just read other people's entries. It's a terrific advance, especially since we have been so slow in publishing these journals! We're hoping to improve on that, but by all means, check out the website. And, welcome aboard, Matt!

—Elizabeth McNamara

In Coming Issues...

We are looking for stories about traveling with your medically fragile child. Also, we will be covering reflux in the next issue. If you have any stories that address reflux (and who doesn't!) and want to share them, please send them along. As always, we look forward to reading your stories.

Joshie's Story

By Cherie Leatherman

Who would have ever thought we would endure the loss of a child? This story of Joshua Leatherman is not written in mourning, but in celebration of the wonderful, loving life that Joshua lived. Memories of Josh, which are still painful, will provide comfort for Dave and me in the years to come. Joshua lives forever in heaven and I am certain that one day we will be reunited.

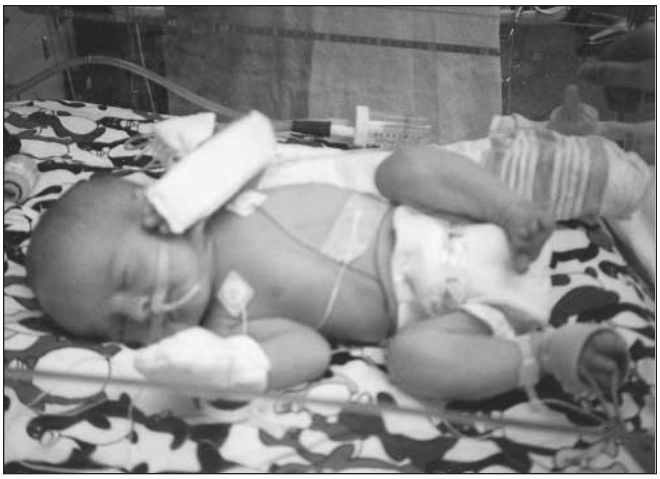
Joshua, our third child, was born on a warm day, June 17, 1996, by Caesarean section. I did not have any idea that he would be born with problems. What parent thinks that their child will be born with a birth defect? The happy birth that we expected turned into an occasion of tears, sadness, disbelief, worry, and loss. Joshua was born with esophageal atresia and tracheoesophageal fistula. I was in shock. I could not even process the information. I broke down crying, but the full reality had not hit.

Joshua was transferred to Lutheran General Hospital in Park Ridge, Illinois, and had surgery to repair his esophagus when he was two days old by Dr. Neuman. The surgery was a success. Dr. Gunasekaran became his gastrointestinal doctor. He was eating and came home after two weeks.

While in the hospital, we were given the phone number for ea|tef Child & Family Support Connection. I thought that our main problems were over. I look back and know how wrong I was. When Joshua was about three and a half weeks old, he started to projectile vomit. He was not gaining weight. I took him in to the doctor and he ended up having pyloric stenosis, which prevented the movement of food from his stomach into his intestines. He had his second surgery to repair the stenosis.

About one and a half weeks later, I was feeding Joshua and he stopped breathing. I became frantic. I tried rescue breathing, but was unable to get any breaths in him. I thought he was choking, so I attempted to perform the infant choking maneuver on him. It did not work. Meanwhile, Dave was on the phone with 911. Joshua lost consciousness, then suddenly the color came back into his face and he started breathing again. He was taken to our local hospital and subsequently transferred to Lutheran General for evaluation. This happened a few more times at home and he always ended up at Lutheran.

We attended an ea|tef meeting in the cafeteria while Joshua was in the hospital, having left one of the nursing assistants to finish feeding Joshua. Apparently, while we were gone, Joshua stopped breathing on her. They called a code and one of the nurses ran upstairs to get us. He was transferred to the Pediatric Intensive Care Unit (PICU).



Joshua was in the PICU for almost the whole month of August. He was on a ventilator for most of that time. He did code when they tried to extubate him. There was so much laryngeal swelling that his airway was cut off and they had a hard time reintubating him. His heart rate dropped to the 20s. They were able to save him but he was a hair away from dying.

He finally came home at the end of August. He had numerous dilations to his esophagus because it kept closing due to scar growth. Still, I again thought that the worst was behind us. Again, I was so, so wrong.

We were at home and I was feeding Joshua one day in the middle of September. He was three months old. He stopped breathing again. Dave called 911 and I attempted rescue breathing, but of course I was unable to get any breaths in. It seemed like forever before the first person arrived, a police officer, Troy Peacock. I was panicking at this time. Joshua was not coming out of this. Troy took over and eventually got Joshua breathing again. Josh was rushed to our local hospital, then to Lutheran General.

Joshua was taken off of oral feedings and a nasogastric tube was placed. Because he was underweight, Joshua was put on a high calorie formula. We were sent home. Joshua stopped going into respiratory arrest. This was the answer. I had been so afraid that he would end up with a tracheostomy. He never needed one.

Josh started to grow and flourish. He was put on continuous tube feedings and he caught up with his growth and development. He had an apnea monitor on him whenever he slept.

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He became a very fat happy baby. He always had a smile on his face. It was really interesting when he learned to scoot and crawl. I felt like I spent my days following Josh with the IV pole to which the feeding pump was attached to so he could move without feeling restraint. I gave him tiny "tastes" of different foods like applesauce and did oral care on him at least two to three times a day. I was so afraid that he would become orally sensitive like so many other children in his situation.

Joshua ate enterally until March 1998, when he was almost two years old. He never had problems eating—he loved it. He continued growing and getting stronger. He did get sick more often than most children. He had the cough that so many ea|tef children have and it seemed that he always had a cold or pneumonia.

His visits to his doctors became fewer and fewer. I was so happy. We were getting through this. It seemed as if the major struggle was over. One would never know Joshua had ever had medical problems by just looking at him. Yes, I knew that he would be on Prevacid and Reglan for years to come, but he did not mind taking the medicines and I knew it was minor compared to what we had gone through before.

In September 2001, Joshua started kindergarten. He was so happy to be in school like his brothers. I was one of the room moms and helped out every week in his class. He loved school and his teacher, Mrs. Heuser. He was so serious about school and so ready to learn. He was so kind to all of his classmates and he made a lot of friends. Josh was a very smart little boy and a very fast learner. He was always the last one done with his work at school because he spent half the time thinking about what he was going to do. Halloween and Christmas (of course!) were his favorite holidays.

Joshua loved the warm weather and the change of seasons. On March 1, Joshua began counting off the days until spring and he was so excited when spring finally arrived.

The last week of March was the kids' spring break. The days were mostly cloudy and cool except for one, Wednesday, March 27. Joshua spent the day playing outside with friends and riding his bike. I hardly saw him that day except when he came in for lunch and when he would check in about every half hour to 45 minutes. He came in at 2:30 in the afternoon for water and quickly went back out to play. Around 3:15 pm, I was startled by kids screaming and crying, "Joshua is dead!" I thought, "No, he must have fallen, but he will be okay." When I got to him I saw his lifeless body on the ground in front of a neighbor's town home. Apparently, Dustin, my son who was 11 years old at the time, found Joshua hanging between two branches of a small crabapple tree. Dustin pulled him out of the tree and tried to carry him home. Joshua did not have a pulse and was not breathing so I started CPR on him. I did rescue breathing and a neighbor did compressions. I remember being fixated on his pupils and screaming, "His pupils are fixed and dilated!" over and over as another neighbor was on the phone with 911. When the paramedics arrived they took over. I remember one of the paramedics shaking his head. He knew as well as I did that Josh was gone. He was taken to Delnor Hospital and they continued the code in the emergency department. They called

As I look back over Joshua's life I see a strong little boy who loved to be loved and to love back.

us back about five minutes after we arrived at the hospital. They were still doing compressions, pacing his heart, and giving him breaths through an endotrachial tube with an ambu bag. At 4:40 p.m. the ER doctor pronounced Joshua dead.

We stayed in the hospital until we were kicked out. Leaving my little boy there was one of the hardest things I have done. I felt as if someone had kicked me in the stomach. I did not sleep at all that night and planned his funeral the next day on about one hour of sleep. The wake and the funeral were beautiful and we received immense support from the community and at our jobs. Meals were sent to us every day for about a month. That certainly helped, but we were still in a state of shock.

We attended our first Compassionate Friends meeting, a support group for bereaved parents, two weeks after Joshie's death, and still attend once a month. Dave and I still feel that there is a big hole in our heart that will never heal. When someone loses a child, it is like a part of you has died also, along with the future hopes and dreams. I have caught myself picking out a toy to buy for Joshua only to remember that he is not here anymore. Some days I feel fine and other days the grief and the sadness is so intense, I feel as if I cannot function. We are both here for Joshua's brothers, Dustin and Nathan. If we did not have them, we would have a very hard time staying alive.

I can ask, "Why us?" Why did he go through so much in his short life and then just die? I will probably never know the reasons until it is my time to go.

As I look back over Joshua's life I see a strong little boy who loved to be loved and to love back. He was a teacher. Everyone was enamored by his bright smile and cute giggle. He had been through more trials in his short five years here than most people ever face. Throughout it all, he never complained. I would have to say that he spent his days here on earth fully, with joy. The anniversary of Joshua's death is two days away as I finish writing this. It is a day of sorrow, loss and remembrance. Joshua was such a happy child that if Joshua were to come back and talk to everyone who sheds tears over his death, this is what he would say:

> I'd like the memory of me
> to be a happy one,
> I'd like to leave an afterglow
> of smiles when life is done.
> I'd like to leave an echo
> whispering softly down the ways,
> Of happy times and laughing
> times and bright and sunny days.
> I'd like the tears of those who
> grieve, to dry before the sun
> Of happy memories that I leave
> When life is done.
>
> *—Author Unknown*

Written in loving memory of Joshua Zachary Leatherman June 17, 1996 - March 27, 2002. We love you Joshie, our sweetie boy.

(Madison's Story, continued from page 1)

two ends of her esophagus. He prepared us for a long hospital stay as well.

As Madi was growing in the neonatal unit, we were making our new home at the Ronald McDonald house. Madison had a long gap atresia, though I can't remember now how long. We developed a routine right away. My husband went back to work at his job an hour and a half away but he drove back and forth almost daily, while I stayed at the Ronald McDonald House. Callie stayed with my parents and came to visit often, spending every weekend with us. I sometimes thought that it was harder on Callie than the rest of us. She was 4 1/2 years old, and her world was turned upside down.

On July 20, Dr. Holcomb came by as he did almost every day, and told us he had scheduled her surgery for July 22. I can't explain all of the feelings that came over me. Terrified, elated, confused… those are some of the emotions we all felt. We had already been told what to except, the long surgery and her transfer to the pediatric intensive care unit, where she would be kept paralyzed until the connection had time to heal.

On the fourth day after the surgery, we noticed that Madi wasn't looking very good. She was very pale and was having trouble breathing. The doctors decided she needed a little blood, but even after a blood transfusion, Madi wasn't breathing normally. The next morning, one of her lungs collapsed. After placing another chest tube, we finally were on the road to recovery again.

Madi was transferred back to her room on the 6th floor after three weeks in the PICU. The nurses even had prepared Madi's room back up for her—after four months, we were like family. On Aug. 15, Dr. Holcomb came by and told us to get ready to go home. The first thing I thought was no—I wasn't ready to go home. I was too afraid to take Madison so far from the hospital. I didn't think there was any way we could take care of her at home.

Dr. Holcomb, however, thought differently. I could, we could, he said, and we had been at the hospital long enough. We needed to get on with our life. I wasn't convinced, and still wanted to argue, but on the morning of Aug. 17, we came home to a very happy Callie. Her family was finally home.

Once home, we had to learn how to use the Kangaroo feeding pump for continuous feeding for Madi and try to teach her how to nurse, keep her g-tube clean and use nitrate sticks to cauterize around her g-tube. We got so good at it that in time we were going to soccer games and church. Three weeks into home life, tests revealed that Madi needed a dilation and that she had severe reflux. A surgery to dilate Madi's esophagus and give her a Nissan fundoplication went great. A dilation tube was left in place so they would not have to put one in every time she needed a dilation. It ran down her nose, through her esophagus and out her g-tube. Each end was taped to her back. We used this process for about eight months. Madi needed dilations every four to six weeks. I remember thinking, it will be so great to actually see her face one day with no tape on it. One thing I have to say, Madi stayed happy and content throughout, always taking in stride everything that was asked of her.

When Madison was 13 months, Dr. Holcomb decided to remove the dilation tube and her g-tube. By now Madi was eating fairly well on her own, and he thought her esophagus would not benefit from any more dilations. This was the day I had been waiting for, to see Madi's face with no tape, her body with no tubes. It was finally here. Afterwards, I couldn't stop looking at her and crying, she was beautiful. She looked free.

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Ethan's Story

by Laura Green

Before I can even begin Ethan's story, I have to tell you about our first son, Jacob. Jacob was born at 27 weeks weighing 2 pounds 4 ounces. Most babies lose weight when they're born, and so did he. Jacob dropped to 1 pound 15 ounces. This was my introduction to motherhood. He was rushed to the NICU, where he spent the next three months of his life. Every day at the beginning was a battle. Each morning began with my daily phone call to the NICU. How was his night? How many apneas, how many bradycardias did he have? How low were his oxygen sats? I vividly remember sitting in the NICU every day and wondering why there were some big babies here with my little preemie. Surely they couldn't be as sick or need the nurses' attention as much as Jacob did.

Flash forward eight years. Jacob is perfect. You would never know that he had been a preemie. He's smart and cute, and a real gift. By then, we had another son, Zachary. When Zach is with his brothers and asks where his scars are, I tell him that Mommy has enough scars for everyone. When he asks if he is a miracle baby too, we tell him that he is the real miracle: It was a miracle he never spent a day in the NICU. My husband, Eric, and I had always wanted three children, so despite the specter of months of bed rest again, as I'd had with the earlier pregnancies, we went for it. My obstetrician was singing the theme song to "My Three Sons" as Ethan was born.

I had an easy labor and a good delivery. Ethan looked beautiful. Then he started grunting. We thought he was making really cute noises. The pediatricians knew better. Once again, my newborn was rushed to the NICU. Honestly, I really thought it was nothing serious. I never imagined that we would be on our way back to the NICU.

I have a very strong attachment to the doctors and nurses at NYU Medical Center. They are the reason that Jacob is the way he is. I had been back to visit many times. Jacob even cut the ribbon on the new NICU and Special Care Nursery when they were dedicated a couple of years ago. I remember saying to Dr. Karen Hendricks how wonderful the place looked, and how pleasant it was to be drinking wine and having hors d'oevres instead of watching the monitors above a sick baby. I never thought I'd be back. But here I was. Some of my favorite nurses were still there—Judy, Joan and Sonia. The neonatologists who I had grown so close to were also there, Dr. Hendricks and Dr. Wasserman. It was one big pity party.

When Eric was told that Ethan had esophageal atresia/trachealesophageal fistula, he was numb. How could he tell me once again that I wouldn't be taking my baby home with me?

Ethan had the most common form of ea|tef. In a way, the doctors said, he was lucky. He showed no signs of cardiac problems, kidney problems, or any of the other things that accompany VATER syndrome, which is what Ethan had been diagnosed with. He did however have a vertebral anomaly, which would need to be watched. Ethan needed surgery to connect his esophagus and repair the fistula. We pushed for it to be done as soon as possible. Since we were a return family to the NICU, Ethan was watched over by everyone. We could not have asked for better care of both child and mother. He was scheduled for surgery the day after his birth.

That first night was probably the worst night of my life. I felt sorry for my little baby, now on a ventilator, with leads all over his little body to monitor every function. It was all too familiar, the beeping, the bells going off all the time. Not even two dozen Krispy Kremes sent by my best friends could help. I was very much alone, and inconsolable. It was unbearable. All I wanted was a healthy baby, and once again this dream was lost.

The next day, Ethan was in the hands of Dr. Bodenstein. I will never forget my husband's face as they wheeled Ethan away, this tiny fragile baby almost covered by all the machinery keeping him alive. We were so low. Ethan was in surgery for six hours. We waited all day for word, and then finally it came. Ethan came through the surgery okay. It was a long gap, but Dr. Bodenstein was able to reconnect the two ends. Now we just had to wait to see if it held, if it would leak. We were given no assurances. There was nothing to do but wait and pray.

When I saw Ethan after surgery, he was a different baby than before. He was swollen and bruised. He was paralyzed so as not to disturb the stitches. He had a chest tube, IVs, and of course, a gastrostomy tube. Every time I looked at this 22-centimeter tube, I wanted to scream how unfair this all was. Where were all of his new bottles and pacifiers? His beautiful new bedding was replaced by a cold isolette. I really did not think that he or I would make it.

Days turned into three long weeks. I spent as much time as I could at the hospital. Now I understood why there were also big babies in the NICU with the preemies. Though he never

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(Ethan's Story, continued from page 7)

had to be resuscitated in front of me like Jacob had, Ethan came with his own set of problems. I had to learn how to feed him through the tube. I hated the tube. I wanted him to eat like a normal baby. I worked with the feeding therapists and with the doctors. I couldn't get used to it. I learned how to flush the tube, and to clean the site. He was still so sickly, and I was told that he could go home. I was shocked. I didn't want to take him home. He was a mess! How could I handle this? They say God only gives you what you can handle, and I forged ahead.

We came home on a beautiful May day. My friends had ordered a gigantic stork announcing Ethan's homecoming for my front lawn. I needed the laugh I had when I saw it. My husband and I had a wonderful support system in place, our friends and family stood by us through everything. I also had an RN with me for the first week. She was a godsend. Together, we had him taking all his formula from a bottle by the end of the week. He learned how to suck, swallow and breath at the same time. Every day he got stronger, and soon I had my regular baby nurse, Paulette, with me. She taught me how to treat him like a regular baby. She sang to him, and read to him. It was the first time she had ever seen a g-tube, but she wasn't scared of it. How could I be?

"Every day that Ethan wakes up and is not coughing is a great day."

A button soon replaced the tube. We needed to have it in place just in case he had a setback. It kept getting infected, and it clearly bothered him. I couldn't wait for the day it came out. Finally, the day came. We were so happy. Everything seemed normal now. Ethan continued to take medication for reflux—pepcid and reglan—and was developing normally. He was receiving physical therapy to keep him strong, and would soon begin feeding therapy.

Then he got his first cold. I was warned that ea|tef babies were prone to respiratory infections, but I was unprepared for what followed. Ethan got a little cold in November. We treated it as we had with our other children, with over-the-counter medications. There was one difference though; we had to give him oral steroids. I was familiar with them because Zachary had had bronchiolitis several times. Orapred really helped open Ethan's airway and stopped his wheezing, and the cold passed in a week.

Then he got sick again. This time it was worse. We tried the same remedies, but this time Ethan was so congested and full of mucus that he couldn't keep anything down. We had to take him to the doctor's office to get shots of the steroid Decadron to keep his airways open. It does the same job as the Orapred, only it's not a battle to keep it down. All the time he was sick, he kept coughing. It went on for two long weeks. We didn't sleep at all during that time; I was too worried that he would stop breathing. Between the wheezing, the vomiting and the constant coughing, there was no way we could shut our eyes. I remember putting him in the car one morning at 2 a.m. and just driving around. I remember my mother-in-law sitting outside with him for two hours in 20 degree weather because the cold air seemed to help. I remember my pediatrician opening his office for me early on a Sunday morning because I was really losing it. I was nebulizing him every two hours with assorted medications—Albuterol, Atrovent, Pulmacort, to name a few. I really didn't think there was anything more a hospital could do for him, except give me a break, so we opted to care for him at home. He finally got well. But since then every time he gets a cold, I am a nervous wreck. I never know how severe it will be.

I have become a total germ freak. My big boys have to wash their hands the second they get off the bus. If one of them has even a sniffle, they know not to go near Ethan, or to touch any of his toys because Mommy will flip. When I take Ethan to the supermarket, the first aisle we go to is the cleaning supplies one, so that I can spray the shopping cart with Lysol, before I put him into it. I have him in one gym class, and every time I go, I can't wait till it's over so that I can pour Purel all over his hands. We've decided not to enroll him in a nursery program this year for obvious reasons.

When Ethan is well, I can deal with his choking episodes and all of his funny noises. But when he is sick it is so difficult. All of his regular issues become exacerbated. And people can be so insensitive. I could be in the bagel store, and someone will ask me, should I really have such a sick baby out? They tell me stories about their child's bout with croup. Croup is nothing! I want to scream at them, He's not contagious! Mind your own business! What, you've never heard a baby with tracheaomalacia breathe before?

Ethan had to go to the emergency room two weeks ago. He had a cold. As usual, a cold is never just a cold for him. I didn't like the way he sounded, it was different than usual. So, we made our first trip to the ER. We went to NYU of course. He got a shot of Decadron, a chest x-ray, and we were on our way. I consider us very lucky that this was our first visit there in almost two years.

Ethan is a wonderful little boy. I love him more than anything. Every day that he wakes up and is not coughing is a great day. Every day that he eats a yogurt and a slice of cheese without choking is another one. When I take him out of the bath and see his scars, I see a miracle, and I can't stop kissing him. He just drew with magic marker all over the basement walls. Oh well, I hope they are washable. When I watch him chase his Grandpa, or blow kisses to his Grandma, I feel pure joy. When I watch him try to play Nintendo or basketball with his brothers, I know that I am blessed.

Ethan has come so far. They tell me that when he is around five, all this will seem like a dream. I can't wait.

Brian's Story

by Kathleen and Robert Serow

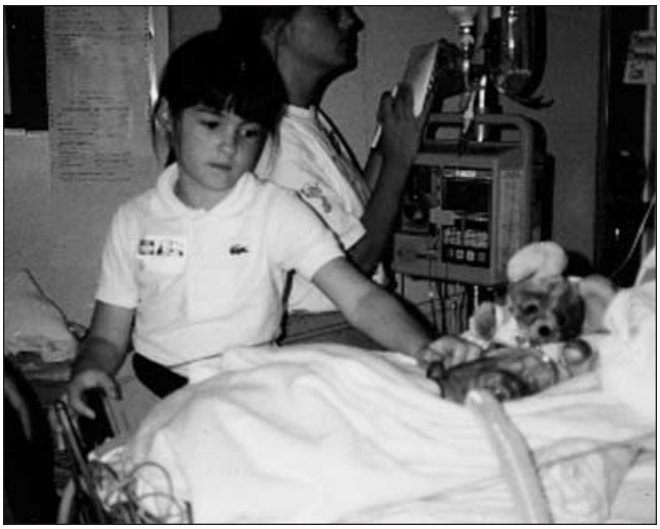
Today Brian is 13, in the eighth grade and plays a mean game of tennis, soccer, and basketball.

Our son, Brian, came into the world on August 23, 1990, giving our 6-year-old daughter, Megan, a little brother. Within hours, our lives were turned upside down and we began an emotional roller coaster ride that remains bumpy even now, but is without the 360 degree loops we came to accept as normal.

Brian was diagnosed with ea|tef. In hindsight, we realize now that Brian was lucky compared to some of the children we've met or read about. But at the time of course it was a shock.

When Brian was 2 days old—he was not diagnosed immediately but that's another story—he was transferred to the University of North Carolina Children's Hospital in Chapel Hill, 45 minutes from our home in Raleigh. We didn't have the wisdom and experience of the ea|tef support group so our information came from medical textbooks and medical personnel. Brian is still cared for by the compassionate and skilled doctors at UNC Children's.

When Brian was 2, our daughter was hospitalized at a local hospital and in chatting with the pediatric nurses at the hospital, we learned about an extraordinarily gifted



pediatrician, Dr. Steve Kubicki. Having a guiding pediatrician was a true blessing since it seemed that much of Brian's treatment has not been textbook. Dr. Kubicki has always gone beyond our expectations to help Brian. As Brian grew, his problems were easier to deal with but persistent reflux, constant respiratory infections, reactive airway disease, and an esophagus that would not heal despite many different medications led Brian to a fundoplication on Election Day, 2000.

Brian's gastroenterologist, Dr. Marc Rhodes, who has since moved to New Orleans, suggested that Brian have a fundoplication several years earlier. Eventually, our own research and consultations with Dr. Duncan Phillips, a pediatric surgeon at UNC Hospital, convinced us that surgery was necessary.

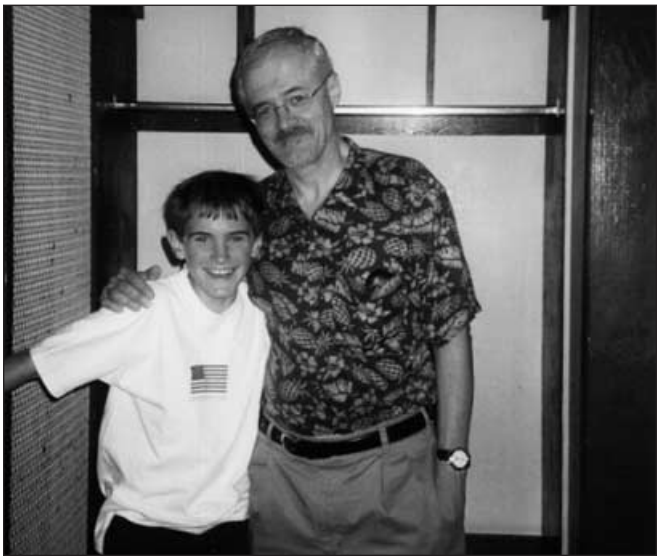
Following the surgery, respiratory infections and incidents of reactive airway disease decreased significantly but we were uncertain whether the esophagus had healed. Brian hadn't ever complained about reflux pain so we couldn't go by that. Two biopsies after the surgery showed that Brian's esophagus hadn't healed, but at least Brian's overall health had improved and a pH probe indicated he had no reflux. But we worried that the erosion of the esophagus over time could have disastrous consequences.

(Continued on page 10)

(Brian's Story, continued from page 9)

Last year, Dr. Rhodes contacted us and told us of a study that used Flovent (swallowed, not inhaled) to heal the esophagus of children like Brian so we tried it and the biopsy in October 2002 was normal. We've since learned the healing may not be long term, but we remain hopeful.

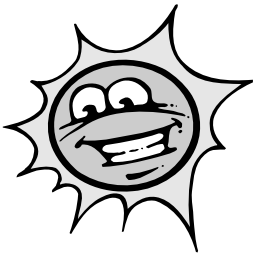
The only negative aspect of the surgery is that Brian cannot vomit, and although that hasn't been a huge problem, it can be uncomfortable at times. And it took Brian a little while to get used to the feel of the fundoplication, which caused some anxious moments after the surgery. The scars are small—barely noticeable alongside his large tef scar.



Today Brian is 13, in the eighth grade and plays a mean game of tennis, soccer, and basketball. He is already looking forward to going away to college. We now think that is possible. Oh, and Megan is now at the University of North Carolina—we definitely know the way there!

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 families 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.

Want to Help?

Become a Local Support Group Coordinator

Some members have emailed or written to say that they'd like to become more involved. One way is to become a local support group coordinator. As you can see on Page 20, there are just a handful of local coordinators, covering only pockets of the United States. Local coordinators provide a link to families of children who've been diagnosed with ea|tef. Families who contact the national office are referred to a local coordinator when one is available.

It's extremely comforting for new ea|tef families to be able to talk to others who know area doctors and hospitals. Some coordinators have organized get-togethers, others stick to phone support, while still others make hospital visits. It's a great way to help new families. If you are interested, contact Bruce Davis at eatef@msn.com.

Need Help?

Contact Your Local Coordinator

Sometimes local coordinators are interested in hearing from area ea|tef families, but have only limited success reaching them. Especially these days, with tightened privacy regulations, it is important for families needing support not to wait for someone to contact you. If you have a local coordinator and are interested in meeting with or talking to other ea|tef families in your area, give your coordinator a call. The contact information is on the back page of this journal. "It is extremely frustrating when you want to do something to reach out to families, but have no way of contacting them," says Michelle Kiger, who with her husband, Jeff, is a coordinator in Indiana. Don't be shy—contact your local coordinator!

Rx Update: Proton Pump Inhibitors

What are proton pump inhibitors, anyway?

Chances are, your child is has been on one or is on one now, since PPI's are a primary weapon in the arsenol against reflux and esophagitis. Here's how they work: Your stomach produces acid to help break down food so it is easier to digest. In certain circumstances, this acid can irritate the lining of your stomach, esophagus and duodenum (the top end of your small intestine), causing indigestion and even ulceration and bleeding. The proton pump inhibitors work by blocking the production of stomach acid. They do this by inhibiting (shutting down) a system in the stomach known as the proton pump.

As with Prilosec and all of the PPIs, the granules are enteric-coated and delayed release. Absorption begins only after the granules have passed through the stomach. So, the capsule should be swallowed without chewing, but the granules can be emptied into juice or applesauce, for instance. The drug needs to remain intact until it reaches the stomach.

Nexium versus Prilosec

So, your child's been on Prilosec for three years and it seems to be doing the job, but you are hearing a lot about this NEW proton pump inhibitor, Nexium. What's new about it? Specifically, there's a change in its chemical structure: Nexium (esomeprazole) contains just the S-isomer of omeprazole. Prilosec contains both R- and S-isomers.

According to the American Gastroenterological Association, "a review of randomized clinical trials comparing two or more proton pump inhibitors found that there are minimal clinical differences between the products. Omeprazole (Prilosec), Iansoprazole (Prevacid), pantoprazole (Protonix), and rabeprazole (Aciphex) all produce comparable rates of healing and remission of sores in the esophagus caused by reflux. Time to healing for esomeprazole (Nexium) may be shorter than the other PPIs; however, the clinical significance of this is not substantiated."

Should your child be on Nexium instead?

Obviously, this is a question for your child's doctor. If Prilosec (or one of the other PPIs) is working well for your child, there doesn't look to be any reason to switch. Coincidentally, Prilosec's manufacturer, Astro Zenica, is also the maker of Nexium. And Prilosec has just gone generic. For more on that, read on.

Prilosec Generic

In recent months, a generic form of prescription Prilosec has come on the market. The cost difference at this point is not huge. Prilosec costs about $4 a capsule; generic omeprazole is about $3.50/capsule. But, says Gayla Waller, manager of clinical services, CVS/pharmacy, the generic price should drop to about $2/capsule within a year.

Unlike purple Prilosec, the generic version now available is a dusty yellow color and the granules contained inside are significantly larger.

What about the liquid omeprazole formula (especially helpful for g-tube dependant children) some ea|tef children have been given during hospital stays? According to Waller, the manufacturer does not make a liquid form. Rather, hospital pharmacists may make a liquid by combining the capsule contents with sodium bicarbonate. This is called "compounding" and some retail pharmacists do this too. Your best bet is to contact your pharmacist and ask for a referral if he/she is unable to make it up for you.

Over-the-Counter Prilosec

In June, the U.S. Food and Drug Administration approved a form of Prilosec (omeprazole) for over-the-counter sales. Prilosec OTC will be available in 20 mg delayed-release tablet form. The same amount of the active drug will be available in both the OTC and prescription forms of Prilosec, according to Gayla Waller of CVS. The difference will be in the dosage recommendation. Prilosec OTC should be taken once a day, every day for 14 days; it is recommended for people who experience frequent heartburn.

Prescription Prilosec, first approved by the FDA in 1989, will remain available as a treatment for gastroesophageal reflux disease (GERD), inflammation of the esophagus (esophagitis) and ulcers.

Gayla Waller predicts that Prilosec OTC will be available in the next month or two. There is no cost information yet available, but generic versions of the medication will not be available for three years. *—E.F.M*

Siblings

by Elizabeth F. McNamara

The revelation came over dinner at the hospital cafeteria. Our son James, the youngest, was in the hospital again (pneumonia that time, I think) and I'd taken our two older children to get something to eat. During the meal, I played the role of journalist, asking both Catherine and Aidan what it was like to have a brother in the hospital.

"I don't like it—you're never home," my daughter said. Never home? In the weeks since James had been admitted, I'd taken great pains to spend time with the older children every day. If I slept at home, I'd see them before school then visit with James at the hospital until midafternoon and go home in time for dinner and homework. My husband would go to the hospital after work. By the time he got home, I was ready to return to the hospital. It was hectic but as long as James was stable, I felt I could split my time between hospital and home.

But I learned that night in the cafeteria that children see things differently than parents. There was no use arguing the point with the two of them, no matter how valid my case. At a time when my older children were feeling more anxious than usual—after all, their beloved baby brother was sick enough to be hospitalized—I was less available to them.

What's a parent to do? As it happens, at our local children's hospital and many hospitals around the country, help is at hand. Programs developed for children who have a brother or sister with special needs are increasingly available. At Hasbro Children's Hospital in Providence, R.I., the Sib Group is run by Debra J. Lobato, Ph.d.

"We try to bring together kids of similar ages whose brother or sister have a developmental or medical illness," says Dr. Lobato. "It's not illness specific. It gets the children to see that there are other children with brothers or sisters who have special needs."

How much should your other children know about their sibling's illness or disability? "We've found that shielding children tends to backfire and creates more anxiety," says Dr. Lobato. "Talking plainly and honestly to siblings to explain the diagnosis and where it may have come from helps to allay their fears."

According to Dr. Lobato, it's not so much what you say, but how you say it. "That's especially true with young children. They can't understand a lot of technical information but they respond very acutely to the emotion or lack of emotion in the delivery," she says.

Give children the opportunity to talk to children who have had similar experiences, says Dr. Lobato, whether those experiences are positive or negative. Sometimes children won't talk honestly with their parents about what they are feeling because they don't want to add to their parents' worries. Sibling groups provide a safe place for them to talk.

An important facet of the sibling group is celebrating the children's strengths and accomplishments in areas that have nothing to do with their brothers or sisters. In Dr. Lobato's program, for one session there is a moratorium on talking about their ill brother or sister. The focus is just on them.

If your ea|tef child has an extended hospital stay, Dr. Lobato recommends creating a family log book that parents and children could use to communicate. For instance, if Mom is staying over at the hospital at night and daughter Jane wants to talk to her about something at bedtime, Jane could write it down for Mom to read when she comes home. Or Dad could write something for the kids if he's not going to be home in time to see them before bedtime. Updates from the hospital could also be added.

How do you deal with your well child when he or she misbehaves during a brother or sister's illness? According to Dr. Lobato, it's important to deal with the behavior when it happens, using your normal disciplinary techniques. But afterward, return to the child and listen to them. "Try to see things from their perspective," she says.

Most of us know that kids with ea|tef have extraordinarily loud coughs. How do we deal with siblings who are embarrassed by their ea|tef brother or sister? Try not to get mad at the sibling for being embarrassed, urges Dr. Lobato. "It's normal and not bad for kids to be embarrassed," she says. "If they turn around and get angry at the child, that would be a warning sign."

The fact is, everyone in a family experiences one member's illness. Making the best of a difficult situation is often the most we can hope for. According to research, at least half of the siblings come out of these experiences saying that they were blessed or enriched by having a brother or sister with a disability, says Dr. Lobato. "We are trying to improve those numbers."

Hey Kids—We Want to Hear From You!

Were you born with ea|tef? Or do you have a brother or sister who was born with ea|tef? We're looking for stories from the younger set about their experiences. What does it feel like when you're eating and something gets stuck? Or, if you're a sibling, how does it feel when your brother or sister has to go to the hospital? Do you sometimes feel jealous because your sick sibling needs a lot of attention? Or, for you ea|tefers, are you jealous of your healthy brother or sister? What are some of the good things about being in an ea|tef family? Any funny stories?

Send us your thoughts, long or short, and we'll try to put them in coming journals. And, parents, that doesn't mean that you are off the hook. We still want to hear from you too.

My Little Brother with the Big Cough

by Catherine McNamara

To some, esophageal atresia is just a medical condition. To others, it's a way of life. I'm the second type. My 6-year-old brother, James, was born with esophageal atresia and Down syndrome. He's always had a huge, hacking cough. Whenever we're out in public with him and he coughs, people stop what they're doing and stare. Sometimes people will whisper things like, "Oh, that poor child!" or "What are they doing, bringing such a sick child outside?" I guess it's not their fault. I mean, that's what people do— they stare. I guess I would too, if it were someone else with James and not me.

Nevertheless, it gets on your nerves after a while. Sometimes I wish I could just say, "What are you staring at? It's just a birth defect!" I remember one time on a plane, a woman behind us commented on James's cough and I turned around to face her and said, "It's a birth defect," in a very matter-of-fact voice. (Keep in mind, I was a great deal younger then.)

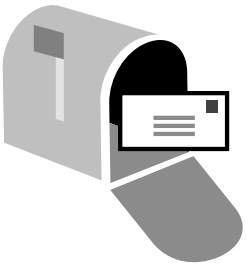
Another time, on the message for our answering machine, you could hear James coughing in the background. When my mom's friend from out of state left a message, she said, "I didn't know you got a dog," referring of course to James! Eventually you get used to it, and just ignore the stares and sometimes even get a good laugh out of how alarmed people get.

James chokes a lot too. When I think about it, I'm amazed at how routine it's become to me. Whenever James chokes, no one panics or goes into some sort of hyperactive mode. James has learned not to panic too. Sometimes my mom has to help James cough up the food that is giving him trouble. Other times he can do it on his own. Although if people are with us when he chokes, they're all like, "What's happening? What's going on? Is he okay?" But if they spend enough time with us, they get more used to it too.

With James, there are many ups and downs, but I prefer to look on the bright side of all of it. So, it turns out that maybe esophageal atresia isn't such a bad thing after all.

Catherine McNamara, 12, lives in Rhode Island with her parents and two brothers. She is in the eighth grade, plays piano and clarinet, likes to swim and reads a lot.

Mail Bag



Your "The Cough" poem in the Fall 2002 newsletter is right on the money! I can't tell you how many times my wife, Leslie, and I have heard: "Oh, poor little thing, my daughter was that sick a few weeks ago…" Or, "Isn't it horrible when they get croup?" The best was when we finally took Kristen and her older brother (almost 3 years old) out to dinner one night. Two little old ladies walked up and did the "coochie coos" to her and she let out one of her biggest "honks," as we've taken to calling them. Well, they both jumped back and screamed, "Oh gracious!" I thought they were just going to drop right then and there. I had to laugh.

Kristen just turned 8 months old last week and, thankfully is developing normally. We've just got that cough to remind us of what she's been through and what we need to look out for.

Thanks for the poem and for all your work with ea|tef. It really helped us out a lot when Kristen was born.

Alan Kohler
EMAIL: aklk@erols.com

We were wondering if you might know of some parents who may have had a similar situation to ours who we could contact...

We were wondering if you might know of some parents who may have had a similar situation to ours who we could contact (or perhaps parents with children who have reflux issues).

James Reed Blake was born on Oct. 31, 1998. He has a condition known as Pierre Robin sequence. He had several surgeries in his first year of life which included TE fistula surgery—he was born with his trachea and esophagus joined together and had them separated with surgery the day after he was born in order to survive. Without this, anything he consumed would go into his airway.

About 9 months later he started having a problem with swallowing milk. After some testing we found out a severe stricture had developed in his esophagus where it had been separated from his trachea preventing almost anything from passing down into his stomach. His surgeon recommended a "Nissen" fundoplication to stop the reflux which was causing the stricture to develop. A G-tube and esophageal dilations were also recommended.

After these procedures were done, James was relatively okay except for very slow weight gain. He has had other feeding issues because of a cleft palette inside his mouth (which was also surgically repaired). Because of his stricture, he has never been able to eat any solid food. Just baby food, pureed foods and Pediasure.

After a recent test to check his esophagus to see if any more can be done so that he might eat solid food, we have been told by his surgeon that his reflux has returned (that the Nisssen was supposed to have fixed). The surgeon says the reflux is aggravating his stricture and medication will not help in his case. He is recommending doing another Nissan Fundiplication surgery and a balloon technique to re-stretch his esophagus. The esophagus is narrowing, and in a matter of months he will possibly not be able to swallow anything.

We don't really want our son to face another major surgery and are hoping for other options. Can you give us some other ideas or techniques that we could look into, especially for managing severe reflux? Also is there anyone who might have some similar experiences with this that you know of? Thank you.

Ken and Terrie Blake
2465 Amanda Lane, Sevierville, TN 37876
Phone & fax # (865) 428-0799, ktjblake@yahoo.com

I wanted to contact you because we have a 12-month-old baby girl who had esophageal atresia and a VSD [a heart defect]. I just heard about you and am interested in being able to talk or email other families who have been through this also. I feel pretty alone. We had never heard of this birth defect and no one can really relate. I would love to be contacted by someone and I can give you more info about our baby.

Gracie is on a feeding pump during the night. She has only needed three dilitations so far and she had done well with them. She is still not a great eater during the day but little by little she is getting better. Her gross motor skills are a little behind other babies her age but she is getting some therapy and she has been improving and catching up. She is still pretty tiny.

Gracie is the youngest of seven children in our family and she seems to have a pretty strong personality. She has been such a wonderful blessing to our family. I would just love to get more information about babies like her.

Carmen Silveira
670 Santa Ana Circle, Santa Rosa, CA 95404
(707) 579-4901, JCRKKBM321@AOL.COM

My name is Billie and I have a wonderful 15-month-old son name Zach. He is perfect. That wasn't always the case. Zach was born full term after an incredibly easy labor. He looked fine. However, thankfully, the nurse who was trying to suction his throat sensed something just wasn't right. After a few minutes and about 10 new doctors and nurses visiting my room, I was told that further tests would need to be done, but that it was likely Zach had an esophageal atresia. My husband and I were able to hold him for a few short minutes before he was whisked away to the NICU.

Tests later showed that Zach had ea|tef. Luckily, he did not have any other problems. Zach had surgery when he was 2

days old to repair his esophagus and spent the next week recovering in the NICU. He was a giant among the other babies and had his own idea about things. In fact, when he was tired of having a tube down his nose, he just pulled it out. Later, just before a procedure, he decided he didn't want his chest tube in anymore, so he pulled that out, too. He kept the doctors on their toes.

Zach spent a total of 11 days in the NICU recovering and then returned to the hospital four times to have his throat dilated at the surgery site so that scar tissue would not tighten too much.

A few months ago, Zach visited his surgeon. His esophagus is just a little smaller than normal at the surgery site. Other than that, and a strange sounding cough every once and a while, he is a completely normal 15-month-old. In fact, although I don't think I'd ever want to go through the first few months again, I think this experience has shaped Zach's personality. He is the most easy-going, happy child I have ever seen. In fact, both family and friends have commented on what a happy child he is.

I wanted to send you this so that my story could be passed on to any new parents facing the same situation my husband and I faced. A lot of information I received when Zach was first born was very scary. Most stories talked of months in the hospital. I know we were very lucky that Zach did not have other complications and every case is different, but I want to relay this true success story on to parents looking for information. I hope my story can help others facing a similar diagnosis.

Billie Bellamy
Orlando, Florida
EMAIL: bbellamy@tampabay.rr.com

My husband and I have recently welcomed a new baby boy on June 22. Jonah was born with ea|tef and has been home for three weeks following successful repair.

I would very much like to become a member of your organization and learn more about what to expect in the future and to allay the fears that are surfacing with day-to-day life. Jonah has a tendency to become cyanotic often when he cries and I have been trying to learn more about this side effect but haven't been too successful. I realized then that I needed to join a support group and communicate with other parents about this and other issues as they come up.

I look forward to your reply.

Shannon Tambolleo
shannon103000@cs.com

I live in Australia and I have a support group here for VATER syndrome and ea|tef that I wanted to tell your readers about. We offer support to those who have someone in their family with VATER syndrome and/or ea|tef. We send out monthly newsletters and would like to organize regular get togethers. We also want to try to organize yearly conferences to be held addressing VATER and ea|tef issues and to raise money for research. If you would like more information about our group or would like to join us, please feel free to contact me.

Julie Clee
390 Union Road, Lavington, NSW 2641, Australia
julieclee@bigpond.com

Hello! Just wondering if there are any local groups in New Jersey. We live in Monmouth County. We have a healthy 4-year-old daughter, Francesca, and a 10-month-old daughter, Gabrielle, who had a successful tef repair at birth. We would like to learn more about any future concerns with the repair and how to prepare baby sitters (we have not gone out since Gabrielle was born). We would also like to share our success story with new parents or assist in any other ways in which we can to be helpful to others in similar situations. Thank you!

Robert & Dagny Fiore, New Jersey

Unfortunately, there are no local ea|tef Child & Family Support Connection coordinators in New Jersey right now. But it doesn't have to stay that way! Perhaps you or someone else in the area would be interested in becoming a local coordinator. See the "Want to Help" box on page 10 for more information about local coordinators. —The Eds.

I'm wondering if you could assist me with some information. I underwent surgery for esophageal atresia 31 years ago at Yale New Haven Hospital. I believe that I was a very early case study for this birth defect. They constructed an esophagus from my colon tissues and to date I have had no problems with my health other than a slight case of reflux, which is common.

I'm looking to see if this defect has been classified as a genetic disease or is simply an unknown birth defect. I am at the age of planning to have children in the near future and would like to know. Any help you can provide would be wonderful.

Elizabeth McMahon

You should be evaluated by a pediatric/adult geneticist to find out whether the ea|tef occurred as part of a syndrome. Even if you were evaluated as a child, you should do so again, since a lot has changed. Also, you should be very clear when scheduling the evaluation that you want to know whether your ea|tef was syndromic in preparation for pregnancy, so that your time is well spent and testing is kept to a relevent minimum. —The Eds.

Send up your questions, comments or thoughts via email or U.S. post (information on back page). Please include your full name and the city and state in which you live.

Surfing the Web

EA|TEF DISCUSSION FORUMS NOW AVAILABLE

We are happy to announce our new web site discussion forums. A forum is simply a category of discussion topics. Click on the forum name to bring up a list of topics for discussion. You can post messages under an existing topic or start a new topic.

Log on to http://www.eatef.org/forums/ to access by registering a username and password by clicking the "Register" link. We hope you find this a useful new addition to our site.

WEB SITE GETS NEW FACE

We have a new member of the ea|tef Child & Family Support Connection team. Matt Parker has taken over responsibility for our web site. He's a software engineer and, in keeping with the rest of us volunteers, a member of the ea|tef "club." His son, Chase, who is 2½, was born with ea|tef. "Chase is doing great. I think we are lucky in that it's been a relatively mild case," he says. Matt and his wife Tracy live in Colorado and have two other children, Christopher, 4, and Jillian, 6 weeks!

NEW EMAIL FOR THIS JOURNAL

In an effort to streamline things a bit, Matt has given the journal its own email address: newsletter@eatef.org. So if you have something for the journal, a letter, a story, or a story idea, you can route it directly to us. But don't worry, items sent to eatef@msn.com will find their way to us too.

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.

UNIVERSITY OF MINNESOTA DELIVERS

Doing a basic "ea|tef" search using Google, we found a very informative web site created by the University of Minnesota Medical School. The site— www1.umn.edu/eatef/ —contains diagrams of the different ea|tef types as well as information about different types of repairs, while advocating primary end-to-end esophageal repairs.

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.*

Ototoxicity

by Heather Fowlie

Many children born with ea|tef spend significant periods of time in hospital. As parents and caregivers we know that being in the hospital can have affects that are not directly related to ea|tef. One such effect which is not commonly discussed is ototoxicity, or the poisoning of the inner ear by certain common drugs. Hospitalization, especially long term hospitalization, often involves exposure to many kinds of drugs for many purposes. Some drugs, while necessary for fighting deadly infections, can unfortunately be very toxic to the ears.

The U.S. Food and Drug Administration does not require either testing of inner ear function or an examination of the inner ear structures when determining the safety of a drug before it is released onto the market. This is one reason it is almost impossible to say with confidence how many or which drugs cause ototoxicity and how many or which people are affected by it.

Problems with a particular drug are usually only discovered after enough people have suffered the consequences and when physicians or other health care professionals can see the likely connection between the symptoms/problems and a drug. This was the case with aspirin and quinine centuries ago, with the antibiotic streptomycin in the 1940s, and more recently with some anti-cancer drugs. Since then, scientific studies have shown that these drugs cause ototoxicity in animals and people. Other, newer drugs, have seemed to be ototoxic as well, but solid scientific proof is often lacking. It can take years to determine scientifically if a drug causes ear poisoning.

Inner ear poisoning occurs in many ways and depends upon the drug involved and many other factors, including heredity. It can be temporary or permanent, affect hearing alone, balance alone, or hearing and balance together. Some drugs mostly have a temporary affect, others can have permanent effects, and some can have either temporary or permanent problems in different people.

Unfortunately, many chemicals have ototoxic potential. These chemicals include over-the-counter drugs, prescription medications and environmental chemicals. The information below includes some substances thought to cause ototoxicity. It can't be complete because so little research has been done to determine the ototoxic potential of drugs in general. This list has also been edited to include only those drugs most likely to be given to children in hospital. Much more complete lists of drugs and other chemicals are available from many sources on the internet. Some of these sites are listed at the end of this article. **If you or your child is receiving any of these drugs it is important not to stop taking them.** Talk to your doctor or other members of your healthcare team to determine if the benefit is likely to outweigh the risk.

Loop Diuretics (a specific family of "water pills")

The family of loop diuretics is known occasionally to cause temporary ototoxicity in some people. These drugs cause ringing in the ears or decreased hearing that reverses when the drug is stopped. Note: If loop diuretics are given at the same time as a member of the ototoxic aminoglycoside family of antibiotics, the chance of permanent ototoxicity is thought to be higher.

The loop diuretics include:

- bumetanide (Bumex)
- ethacrynic acid (Edecrin)
- furosemide (Lasix)
- torsemide (Demadex)

Aminoglycoside Antibiotics

All members of the aminoglycoside antibiotic family are well known for their potential to cause permanent ototoxicity if they enter the inner ear. Some of these drugs are more likely to cause hearing loss, and others are more likely to cause loss of balance. These drugs enter the blood stream in largest amounts when given intravenously (by IV) and in the least amounts by pill.

Members of the aminoglycoside family include:

- Amikacin (Amikin)
- Gentamicin (Garamycin)
- Kanamycin (Kantrex)
- Neomycin (Mycifradin)
- Netilmicin (Netromycin)
- Paromomycin (Humatin)
- Streptomycin
- Tobramycin (TOBI Solution, TobraDex, Nebcin)

Vancomycin (Vancocin) is another antibiotic known to cause "significant" ototoxicity. Given intravenously, it's the main alternative to penicillins for serious infections in patients who are or are thought to be allergic to penicillins. It's sometimes administered along with an aminoglycoside for certain conditions.

If your child has received large doses or many doses of these drugs, you should have their hearing tested on a regular basis, even if you have not seen indications of hearing loss. Some inner ear hearing loss can get worse over time and may not be detected for months or years after receiving ototoxic drugs. Even mild or moderate hearing loss, while it doesn't seem to affect daily living, can affect a child's ability to effectively learn and communicate.

References:

www.vestibular.org/ototox.html
www.personal.umich.edu/~mshlafer/ototox2.html
www.ifhoh.org/Epstein.htm

The ea|tef Child at School

For those of you who might be sending your ea|tef child off to school for the first time this fall, it can be a very frightening prospect. For five years you have guarded your child from choking hazards, you have watched him closely at mealtimes in case something gets stuck and you have diligently instructed family and friends never to feed your child anything without your permission. Now, you are facing kindergarten where you won't be able to supervise his or her every move.

Here are some basic things you will want the teacher and other caregivers to know:

- Children born with ea|tef have a characteristic "TEF cough" that is caused by weakness in the trachea (windpipe). This coughing, by itself, does not mean the child is ill.
- Children born with ea|tef may be more susceptible to respiratory infections. Hand washing is very important and should be encouraged with all children in the class.
- This child may have gastroesophageal (G.E.) reflux which causes stomach contents to flow back into the esophagus.
- The child may need to take medication during the day.

You will also want to make the teacher/caregivers aware that because your ea|tef child has an esophagus that doesn't work the way it should, food can easily get stuck on the way down. Choking episodes can look very frightening to people who have never seen one before. Let your child's teacher know what your usual procedure is. They need to know if your child is usually able to cough up lodged food or if a drink of water can help wash it down.

Children in elementary school may be rushed during the lunch period. It is very important that all lunchroom supervisors are told the following things about ea|tef children:

- The child should never be rushed while eating or drinking. Rushing him could lead to a choking episode.
- The child will need to eat slower than other children since smaller bites and more chewing are necessary.
- The child should always be upright during and after eating. This will help prevent food from accidentally entering the lungs and causing a lung infection.
- The child may only be able to eat small portions of food at a time and may need to be given other opportunities to snack.
- The child will need to drink more than other children to help food go down the esophagus. Do not limit the amount of fluid this child takes.

Of course the most important thing is for you, the parents or guardians of the child, to be in constant communication with your child's school. Be sure they have easy access to phone numbers to reach you in an emergency. If at all possible, identify a specific person or people in the school whom you can train to do things in a way you and your child are comfortable with. This includes identifying when they need to call you or call emergency medical services.

Open communication will make you more comfortable, and will make the school personnel more comfortable, knowing they have all the information necessary to keep your child safe and happy in school.

Adapted from the pamphlet *ea|tef: A Guide for Teachers*, available from the ea|tef Child and Family Support Connection.

(Madison's Story, continued from page 6)

Things went well over the next two years, her feeding going better and better, even though she was small (and still is) for her age. The spring she turned three, we had noticed one of her eyes would cross occasionally, so after a eye doctor visit, she was in glasses and wearing a patch over one of her eyes. Later that summer after a routine checkup with her surgeon, he noticed a umbilical hernia. Dr. Holcomb also told us he was transferring to Children's Mercy Hospital in Kansas City. This thought was terrifying to us, losing him, and yet another surgery too? There's no way you can move and leave us, I remember thinking. I asked if he would do the surgery before he left, because he knew Madi better than anyone. On August 3, 1999, Madi had her umbilical hernia repaired. Again, she did well. The night she got home, she wanted to go to the ballpark and get some french fries. Amazing, huh?

So by now everything is rocking along pretty well, until… yes, until the winter of 2001. I started noticing Madison's back after getting her out of the bath, that her left shoulder blade protruded outward. I asked my husband and mother if they noticed it, and they did, so after a few weeks, I decided to have her pediatrician look at it. I had already done some research on the Internet and decided Madi must have scoliosis. The pediatrician confirmed this and we were on our way back to Vanderbilt by April 2002.

Dr. Gregory Menico, a pediatric orthopedist, confirmed our fears. After scheduling a MRI to rule out any tumors, Madi began going every three months for checkups. By that November, it appeared Madi's curve was getting worse. It had started out of 24 degrees; now in 6 months it had grown to 32 degrees. Madison is now in a Boston Back Brace, wearing it 16 hours a day, every day. She is supposed to wear it until she stops growing, somewhere between 14 and 16 years old. Luckily, Madi is doing good with the brace, she thinks it's making her taller.

We have had some problems develop since wearing the brace. Madi's reflux problem, once the brace was tightened up more, became a major issue. Madi lost 3 pounds. She stayed out of the brace to gain weight back, and she also had another esophagram to see if she had any other problems going on. Everything appeared as normal as it can be for her. They think the brace was just too tight. Now we're in a dilemma as to know which route to take. Wear the brace loosely and hope it helps some… or go without it and know that her chances of having to have back surgery will increase more. It's disheartening for us to not know which route will be best for her. Right now she's just wearing it loosely and we're hoping it helps some, happy that she can eat and not have reflux so bad.

Madison has come a long way in her 7 years. She still has things she has to deal with, but I'm certain that she will be able to handle anything that life deals her. Her true love is horses. She had rather be outside petting and feeding our horses then to eat. She is not only tough and cute as a button, she is our little miracle, a gift from God. If parents want to contact us, my email is mellyorrick@hotmail.com.

Other Organizations:

ABOUT KIDS GI Disorders
158 Pleasant Street, N. Andover, MA 01845-2797
Phone: (978) 685-4477 or 1-800- 394-2747
Fax: (508) 685-4488

Birth Defect Research for Children INC.
930 Woodcock Road, Suite 225, Orlando, FL 32803
(407) 895-0802

March of Dimes Birth Defect Foundation
1275 Mamaroneck Ave., White Plains, NY 10605
(888) 663-4637

MUMS-Mothers United for Moral Support -National Parent to Parent Network
150 Custer Ct., Green Bay, WI 54301-1243
Contact: Julie Gordon (877) 336-5333

National Fathers Network
16120 NE 8th St., Bellevue, WA 98008-3937
(425) 747-4004 ext.218

NORD Inc.(r)-National Organization for Rare Disorders
P.O. Box 1968, Danbury, CT 06813-1968
(800) 999-6673 (voicemail only) Fax: 203.798.2291

The Oley Foundation
214 Hun Memorial, A-28
Albany Medical Center, Albany, NY 12208-3478
(518) 262-5079 or (800) 776-OLEY

Pediatric/Adolescent Gastroesophageal Reflux Association
Main Office
P.O. Box 1153, Germantown, MD 20875-1153
(301) 601-9541
West Coast Office
2329 Fallbrook Place, Escondido, CA 92027
(760) 747-5001

The Sibling Support Project
Children's Hospital and Medical Center
P.O. Box 5371, CL-09, Seattle, WA 98105
(206) 527-5705

United Ostomy Association, Inc. (UOA) - Parents of Ostomy Children (POC)
19772 MacArthur Blvd., suite 200, Irvine, C.A. 92612-2405
(800) 826-0826

The VATER Connection
1722 Yucca Lane, Emporia, KS 66801
Contact: Angie Schreiber

Medical Advisory Board

Dr. Keith W. Ashcraft
Surgeon in Chief
Children's Mercy Hospital
Kansas City, Missouri

Diane Kane, R.N.
Pediatric Surgery Nurse

Dr. Preston Black
Pediatric Surgeon
Loyola University Medical Center
Chicago, Illinois

Becky Pruitt, L.C.S.W.
Ethics Program
St. John's Regional Health
Springfield, Missouri

Dr. Grant Geissler
Pediatric Surgeon
Chicago, Illinois

Dr. Jayant Radhakrishnan
Pediatric Surgeon
Chicago, Illinois

Dr. T.S. Gunasekaran
Pediatric Gastroenterology
Director, Pediatric Endoscopy and
Motility Services
Lutheran General Children's Hospital
Park Ridge, Illinois

Donna Schwartz, R.N.
Children's Memorial Hospital
Chicago, Illinois

Dr. Lauren Holinger
Head, Pediatric Otolaryngology &
Dept. of Communicative Disorders
Children's Memorial Hospital
Chicago, Illinois

ea|tef Child and Family Support Connection, Inc.

Phone: (312) 987-9085 Fax: (312) 987-9086
email address: eatef@msn.com
111 West Jackson Boulevard
Suite 1145
Chicago, IL 60604-3502

LOCAL SUPPORT GROUPS

CALIFORNIA — Northern
Coordinator: Joan Crook
Moraga, CA
jcrook2@aol.com

SOUTHEASTERN NEW ENGLAND
Coordinator: Elizabeth McNamara
East Greenwich, RI
emcnamara@eatef.org

INDIANA
Coordinators: Jeff & Michelle Kiger
Greencastle, IN
jkiger@eatef.org
mkiger@eatef.org
(765) 522-3586

OHIO - Cincinatti Area
Coordinators: Martha & Mark Reed
Cincinatti, OH
(513) 921-1676

MICHIGAN
Coordinator: Angel Storey
Allegan, MI
(616) 673-5582

NORTHERN NEW ENGLAND
Coordinators: Dee & George Willant
Newmarket, NH
(603) 659-7312

OHIO - Cleveland Area
Coordinator: Sherm Langan
Hinkley, OH
(330) 225-8048