ea|tef

Child and Family Support Connection, Inc.

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

INSIDE

Marlo's Story

In April 2000, at my 8-week prenatal visit, my ob-gyn gave me the news—twins, two babies growing in my stomach! My husband, Brent, and I were so very excited, but also a little nervous, because we knew how much work it was going to be. We were also concerned because of what had happened before…

In 1998, our son, Bryan, was born six weeks early, weighing only 3.8 pounds. On his third day of life, Bryan was diagnosed with a heart defect, a large VSD (ventricular septal defect). He developed congestive heart failure shortly after, and he was put on medicines to help him breathe and get his heart working more efficiently. We took Bryan home at 4 weeks, but by 10 weeks he was getting sicker, so we decided to take him to Texas Children's Hospital in Houston for surgery. Bryan sailed through the operation. He was fine, lots of tubes and wires and some scary times, but he did wonderfully, and we were back in Austin after one week. So Bryan, our little hero, prepared us for what was to come next.

I had a very healthy and happy pregnancy with the twins. I found out at 16 weeks that I was expecting not one, but two girls! I was so excited, and spent lots of time nesting and creating a pretty little twin nursery. I went on bed rest at seven months because I was getting so big and the twins weren't growing as fast as they needed to.

In early October, during a routine visit to the perinatologist (my ob gyn sent me to one because of the twin pregnancy and what had happened with Bryan), the doctor mentioned that there was some "extra fluid" in baby A's stomach. He wasn't really certain what that meant, and explained to my husband and me that it could be something or nothing—but that we'd have to wait until the birth to find out.

I had a C-section scheduled for October 20. I was 36 weeks pregnant. I had had a C-section with Bryan and with twins my doctor thought a C-section was the way to go. That morning, my water broke, so those babies were planning to be born that day anyway!

Audrey Grayce (Twin A) was born at 7:01 a.m. and Marlo Diane (Twin B) came 1 minute later. Audrey weighed 4.11 pounds and Marlo 4.1 pounds. In the neonatal intensive care unit, poor Audrey was put through a battery of tests since the perinatologist had thought she was the one with the excess fluid. They began the same tests on Marlo and found that they couldn't pass a tube down her throat. Further tests confirmed that Marlo had type C ea|tef. They told Brent what was happening and he came to explain to me the good news that Audrey was fine, and the bad news that Marlo was not. We were strong, though, because we knew she was in good hands.

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Editor's Letter

Again, too many months have passed since the last issue of the *ea/tef Journal* came out. I know what a lifeline this publication is for families of newly diagnosed babies. Luckily, these days families can visit our website and not only learn about the condition of ea/tef, but also make contact with other families through the discussion forums. For the many, many families of older ea/tef-ers out there who lived in almost total isolation with this difficult disease, this incredibly easy way to meet others with ea/tef must seem dreamlike.

Yet it can still take time for some people to find our organization. Last winter, my husband and I heard from grandparents of a 2-month-old ea|tef baby in Illinois (of all places—home base for this very organization!) who called our home in Rhode Island after someone had given them a copy of an article I'd written about our son James for the local newspaper. Perhaps they'd found it on the web, but no one had yet told them about ea/tef Child & Family Support Connection. Yikes!

So I charge each and every one of us with the job of helping new families find this organization. One way is to remind the pediatric surgeons and other doctors we know about the organization. Or remind nurses or hospital workers you may run into on a well visit or scheduled endoscopy.

It happens that a charge nurse from the local neonatal intensive care unit lives just a block away from us (pretty lucky, huh!). Occasionally, I ask if any new ea/tef babies have been born. She's bound by pretty strict privacy rules (especially now), but even if she doesn't tell me about a baby, she will be reminded that there is a way to help that new family. We can all help to get the message out.

This issue features two stories about traveling with ea/tef kids (page 3). I have to admit that after I'd written mine, I wondered if readers wouldn't think that we'd been reckless. The story highlights all the things that have gone wrong on our trips, but please understand that much more went right! Shari Fensterheim's charming story about her daughter's inopportune encounter with a cocktail cherry reminds us that our children are nothing if not resilient. Thank goodness!

As always we welcome your stories— travel stories, reflux stories, g-tube stories, illness stories, any stories. We hope to inaugurate an advice column, featuring suggestions from members on food, eating, choking, reflux, medicine delivery, hospital stays, you name it. If you have something you'd like to share, send me an email…please! Remember, the more you write, the less I have to! Happy summer to all. May your illnesses be brief and your chest x-rays be clear!

—Elizabeth McNamara
July 2004

In Coming Issues...

The next issue of the *Journal* will feature an article from a medical journal on how ea|tef children fare in the years after surgery. Our future topics include reflux, sugery options, and medicines. As always, your stories make the difference.

Seeing the World
(and an E.R. or two)

By Elizabeth F. McNamara

Rhode Island, my home state, is a very small place. You have to work pretty hard to take more than one hour driving across it at any particular spot. Still, after our son James was born with esophageal atresia and he took up residence at the local NICU, then PICU, any trip beyond the state's borders began to seem as unlikely and difficult to acheive as a trip to the moon.

A year after James was born, we did manage a family vacation—to an island off the Rhode Island coast. It was still part of the state, however, and that comforted us. It also made James's need to be dilated halfway through the vacation possible. He and I took the ferry to the mainland, a neighbor drove us up to the hospital and after the out-patient procedure we stayed one night at the nearby Ronald McDonald House, just in case. The next morning, we got a ride back to the ferry and we were vacationing again by lunchtime.

A few months later, we took our first big trip—to California, the land of my birth and where most of my family still lives. At this point, James still took most of his nutrition via g-tube, required monthly esophageal dilations, sometimes needed suctioning, needed a pulse oximeter for spot checks on his breathing, and oxygen via nasel cannula when his 0_2 saturation numbers dipped below 90 percent (usually just when he was sick).

Going to the island had meant alerting the local medical facility (a one-doctor establishment) about James and remembering to bring a spare g-tube kit (as well as all the day-to-day equipment James then required—feeding bags, I.V. sponges, medicines and syringes, etc.). We also had to check with the owner of the house we rented that the electrical system could handle charging batteries for James's feeding pump, his pulse oximeter and his portable suction (which we could plug into the car in an emergency). It could, we learned, as long as we only plugged in one charger at a time.

Planning for the California trip required most of that and more. First of all, there was the plane trip to think about. How would James handle it? What if he needed oxygen en route? I contacted the airline and was told that to have oxygen on board for James would cost $50 per flight "segment." (FAA regulations do not allow passengers to bring their own oxygen on board.) The likehood of a healthy James requiring oxygen was slim, but we opted to be safe and paid the extra $200 for the total of four flight segments.

What about once we arrived? If James got sick while we were there, we could need oxygen as well as a doctor or, dread the thought, even a pediatric surgeon. We arranged for an "E" tank of oxygen through a local medical supply company. My parents were also able to give me the name of a nearby pediatrician. I went to James's pediatric surgeon in Rhode Island for the name of a colleague in the Los Angeles area. He gave me the names of two—one at a large hospital near by and one at a pediatric hospital downtown.

James McNamara (long gap ea) sits on his brother Aidan's lap on a flight to California in 2002.

We were ready. Now, James just had to stay healthy. I listened to every cough, wondering if it was just a regular James cough or meant something more. But our departure day came and everyone was healthy. The flights were uneventful (so much so that I can't even recall them) and James seemed to take to California. During a trip to the beach one day, James did a face plant in the sand and came up with a mouthful of the stuff. I panicked for a moment and got as much sand out as I could, but James's esophagus seemed no worse for the grit.

We had a wonderful time. Many of my family members were seeing James for the first time and they were thrilled to meet the little boy who had already been through a lifetime of trauma. I even managed to relax.

Undoubtably, there are families who would choose to not to take such a trip with a medically involved child. My husband and I, however, have always loved to travel and our older children liked it too. We wanted to see relatives in California, of course, but we also wanted to retain a part of our life that we loved. We were willing to put up with the added work involved in traveling with our medically fragile little boy.

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James with his brother Aidan and sister Catherine on a double-decker bus in Dublin, Ireland, in 2000

"Is there a doctor on board?"

After the California success, more trips followed. Invariably, we stayed close to home during the winter months, since James's favorite place to visit that time of the year was the pediatric hospital's ICU. But come summer we planned big. When James was 2, we returned to California for a family reunion and trip to San Francisco.

James had had a second surgery on his esophagus two months before. We flew without realizing that scar tissue was building up around the surgery site and that he would soon need to be dilated. A few days into the trip, James started having difficulty swallowing his food. (My sister remarked, "He's fine, as long as you don't feed him!") By the end of the trip, even his morning secretions were causing him trouble. We'd had a terrific time, but I was glad when it was time to fly home.

For some reason that I will never understand, I checked the portable suction machine that we kept with us to help clear James's esophagus in times of trouble. A half hour into the second flight, as I was giving James some formula in a cup (he took about half his total nutrition by mouth at this point, the other half via g-tube), he started choking. The formula wasn't going down. And my husband and older children were several rows in front of us, with a flight attendent and beverage cart between us.

I signaled to another flight attendent and asked that a request be made for a doctor or nurse. Sure enough, there was a nurse on the flight and she came with me as I carried my blue-tinged boy to the back of the plane. I explained the situation and she had a simple solution: gag him with my finger. It hadn't occurred to me! Sure enough, after I did that a couple of times, James was more comfortable and his color improved. It was then that the flight attendent asked me if she should prepare the pilots for an emergency landing. Emergency landing! I thought. "No! He's okay," I said. We landed in Rhode Island without further incident. And I had a new tool in my arsenal—my finger.

Ireland or bust

The following summer we traveled to Ireland, a long-held dream. Again, I spoke to our pediatric surgeon to get the names of surgeons and hospitals in Ireland, just in case. And we arranged to have two E tanks of oxygen available for us at the airport there, also just in case. (James had had so many respiratory illnesses that winter that his lungs hadn't completely recovered.) My careful plans didn't quite work out—the oxygen wasn't delivered to the car rental agency and we had to ferret out the company in a nearby city to pick it up, and the electrical adapter I'd brought for the feeding pump and pulse oximeter didn't work so we had to track down a usable one.

But, as the Irish say, the trip was grand. For two weeks we

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Sarah

and the Cherry

by Shari Fensterheim

President's weekend 2003. We took a family vacation from sunny Phoenix, to Washington D.C., for a family event. We arrived, and then the snow arrived! We had fun with family and friends and more snow arrived! Okay, you get the picture. Yes, the Phoenix family of six left sunny, 75 degree weather to get snowed in—26 inches of the white stuff to be exact. With all roads closed, airports closed, and the hotel running low on pretty much everything, our daughter Sarah (born in 1994 with ea/tef) decided to add excitement to the vacation. Here's the blow by blow…

Saturday, 8 p.m., Sarah apparently sneaks a Maraschino cherry (we do not let her eat round fruits or veggies) from her "Shirley Temple" drink. Her cousins make her laugh and she swallows the cherry whole. She chooses not to tell us as she does not want to get in trouble. By midnight all roads are closed due to the snow.

Sunday, 10 a.m., Family brunch for the "out of towners" at the hotel. (None of the local family can join us as they are all snowed in.) Sarah mentions she is having difficulty with her eating. We think that she had some fruit at the brunch that may have gotten stuck. She begins the "Sarah routine" for getting food dislodged: water, water, and more water, followed by coughing up all food that is sitting on top of whatever is stuck. She works her way backwards until she gets to the suspected culprit. She can generally dislodge most stuck food within an hour. This time is different. Very different! Sarah would think she is "unclogged" and begin eating again. Food would pile back up and within an hour or so she has to repeat the entire process. This continues the rest of Sunday and into Monday.

Monday, 10 a.m., Sarah is still unable to pass any food or liquid without having to cough it back up. At this point, she admits she swallowed a whole cherry two days ago. She is very hungry and very thirsty. Once we know it is a cherry, we put our heads together and decide we need to alert the local hospital. Realizing the size of a cherry, we know it's unlikely that any of our home remedies will break it down enough to dislodge it.

The hotel staff becomes aware of our problem and joins the ranks of "how to get rid of a cherry without damaging a thin esophagus." By this point, we have been snowed in for more than 40 hours. Sarah continues to complain of hunger and thirst, yet manages to continue smiling.

We attempt to contact a few doctors recommended by a local family but are unsuccessful due to the President's Day holiday and road closures. Next, we call 911 for advice. They send paramedics to the hotel to assess Sarah. The paramedics feel that she should be seen at the local hospital but that hospital is diverting everyone to a farther hospital. The paramedics are not sure if they can get us there due to the road closures. The hotel manager offers to take us in his 4-wheel-drive jeep, but after much debate, the paramedics take Sarah, her dad and me by ambulance to a hospital. It isn't a fast trip—we speed along at 30 m.p.h., but the paramedics are awesome! They make Sarah feel comfortable and answer her many questions along the way (most of which have to do with local food… she's very hungry!)

Once we arrive at the hospital, the doctor tells Sarah that he will admit her, get an I.V. started, and that they should be able to do the surgery to clear her esophagus by the next day. Sarah looks up at the doctor with her big brown eyes and says, "Can you please do the surgery tonight? I am very hungry, the airport is closed and I can't get home to my surgeon." The doctor says he will see what he can do.

Sarah is in surgery about three hours later. The hospital sent volunteers with 4 wheel-drive vehicles to pick up the surgeon and the anesthesiologist at their homes (both were snowed in) just for Sarah! The procedure is successful and before long we are headed back to our hotel, driven by yet another volunteer.

What an amazing experience! Everyone who met Sarah wanted to help her. Not even a record snow storm could stop them!

The airports finally opened back up on Wednesday. We flew home safely and Sarah got to take her prize cherry back to Phoenix for "show and tell."

—sbfenster@yahoo.com

(Marlo's Story, continued from page 1)

Dr. Jeff Horwitz, a pediatric surgeon, came to my hospital room to talk about the surgery that Marlo would need that day. He explained everything about the operation, and I think he was surprised how calm we both were. After our experience with Bryan, we knew that a rough start could be overcome.

By 12 p.m. Marlo was in surgery. Beforehand, a NICU nurse brought her to my room so that I could hold her—God bless that woman! She knew what I didn't, that I would not be able to hold my new baby for close to five days! The operation lasted three hours. The gap in her esophagus was slight so the operation was relatively easy. After reading so many of the stories in this journal, we now know how important this was.

Marlo came through well and was extubated almost immediately post op. Her chest tubes were taken out on the fourth day post surgery and a barium swallow revealed no leaks at the surgery site! All the NICU nurses were amazed at her progress. We felt so blessed. She came home on one medicine, Zantac, for reflux.

Audrey was already home when Marlo came home at 11 days old. Our son Bryan had been so excited when Audrey came home. Then a few days later we came home with Marlo. A few days after that, I was surprised by how upset Bryan got when I was leaving to run an errand. It turned out he thought every time Mommy left she came back with another baby!

We were blessed to have a baby nurse who specialized in twins helping me care for the babies at home. Sherma noticed after Marlo's third cold in four weeks that she was coughing a little when she started to bottle feed. In addition, she wasn't gaining weight nearly as fast as her sister. Concerned, I brought her back to the hospital for another barium swallow. The swallow showed the barium leaking into her lungs. I immediately thought there was a leak somewhere ñ and that she would need another operation. The surgeon assured me that it was probably reflux, so she was put on Prevacid and a nebulizer for asthma.

On New Year's Day, Marlo was so sick that Sherma insisted we take her to the doctor before the office even opened. Glad we did. The doctor was extremely concerned with her condition and checked her O2 sats, which were a dismal 77 percent [normal is in the mid- to high-90 percent range]. Marlo and I went by ambulance back to Brackenridge Children's Hospital where the ER docs told me she had RSV (Respiratory Synctal Virus) and would be going directly to the PICU for treatment.

For the next 10 days, we struggled to get Marlo off oxygen. Finally, a barium swallow revealed what we knew all along. The fistula had returned, and every time Marlo drank anything, she was aspirating it into her little lungs. Immediately, she was placed on nasogastric feeding and a tube was passed through her nose and into her tummy.

NG feeding was rough but Brent was the master. He could get the tube into her nose and down to her tummy in seconds. But it just about killed him every time he had to do it. And at 4 months old, Marlo had a habit of yanking it out. We learning everything there was to know about French tubes, Tegaderm and automated feeding machines. Sometimes I think I can still hear that beeping in my sleep.

The worst part was never being completely sure if the tube was in the right place in her tummy. Even though we used the stethoscope method, the thought of making a mistake and filling her lungs with formula would get me weak in the knees every day.

As for sleep—there wasn't much of that to be had! Marlo had to be fed every three hours (at four and a half months old, she was still only 10 pounds). So between night feedings for the twins and getting the machine set up and running, it was a tough time.

Despite all this, Marlo was a smiling, sweet and unusually happy baby. With so many invasive procedures, her positive demeanor amazed us. We were also impressed that she only lagged behind her sister's development (rolling over, smiling, etc.) by a couple of weeks. Remarkably, Marlo sat up at 6 months and walked at 9 months!

I started researching recurrent fistulas and found that they were highly unusual. So unusual, in fact, that I had real concerns about having the repair done in Austin, since none of the local doctors had any real experience with a recurrent TEF. Thank goodness for my ea/tef newsletter, because there I found an article by a doctor named Juda Jona, a surgeon in Chicago who had several recurrent TEF repairs to his credit. I immediately contacted his office, and we had a great conversation about our options. Dr. Jona even offered to fly to Texas to do the surgery for us. Amazing.

At the same time, I had contacted Chuck Fraser's office at Texas Children's in Houston to see if he could recommend anyone in Houston. We were lucky to have Dr. Bob Bloss, probably one of the best known and experienced pediatric surgeons in the country, offer to do the procedure. He had done a few recurrent TEF repairs over the years and was confident he could fix Marlo's.

So Brent, Marlo and I headed down to Houston for the surgery. First they conducted a barium swallow test on Marlo. Her fistula was so hard to see that four different radiologists and Dr. Bloss were there in the room with us trying to figure out where it was. It was on the rear side of the trachea, so Dr. Bloss knew it was going to be tough. He asked if we wouldn't mind waiting until the next week for his "A" team of doctors to do the procedure. We didn't really have much to consider there. The operation was rescheduled.

The next week we returned to Houston. The operation lasted six hours. When Dr. Bloss finally showed up in the waiting room, we were completely frazzled. He took his scrub cap off and told us it was one of the hardest operations he'd ever

From left, Audrey, Bryan and Marlo Beesley far, far away from the hospital.

done. That was pretty amazing to us, considering he had operated on over 30,000 kids!

Marlo stayed in the level 2 NICU at Texas Children's for 10 days. Bryan came down to stay with us in the hotel room and Audrey (still just an infant) stayed behind in Austin. We were so happy to be putting this phase of our lives behind us and so ready to move on to easier times. Little did we know that things were going to get worse before they got better.

It turned out that after two months on an NG tube, Marlo didn't want to take a bottle. The occupational therapists at Texas Children's tried, I tried, Brent tried—we all did everything we could, but Marlo would take one sip from the bottle, start coughing and then spit the nipple out. I think the idea of swallowing the milk was just too unpleasant for her and we certainly understood. But I didn't want to put that NG tube back in, so we started trying every hour and a half to get her to drink. When she finally would take at least an ounce of formula, they let us take her home. But Dr. Bloss told me that Marlo would most likely wind up needing a g-tube and a Nissan fundoplication [an anti-reflux surgery]. Ugh!

At home, our nurse Sherma was determined to get Marlo to eat. It was grueling. Some days she'd take only 8 ounces!! I was terrified that she'd lose even more weight. She was almost 5 months old, and she only weighed 11 pounds. Truth be told, though, her perfectly healthy sister was only 13 pounds. I tried solid foods in bottle, solid foods watered down, rice cereal, everything! The occupational therapists were at our house three times a week. The nurses also would make home visits, bringing a scale with them—oh, how I dreaded that scale!

Finally, in the middle of March, an OT came with her supervisor. They sat us down and said they thought that Marlo needed a feeding tube in her stomach. Together with Dr. Bloss, they thought Marlo would only gain weight if she had the surgery. Dr. Bloss would perform a Nissan fundoplication on Marlo at the same time.

Brent really did not want this surgery for Marlo, but I was frustrated and anxious over Marlo's weight. We scheduled the surgery. The night before, I sat in my living room thinking about it and I just changed my mind. Brent and I discussed it, and we decided to postpone it ñ indefinitely. I don't know why or what made me change my mind, although I was praying a lot about it so I'm sure that helped.

It was the right choice. Slowly but surely, Marlo started to eat again. By early March, she was back up to 6 ounces at a feeding. She was still barely gaining weight, but at least the scale was moving in the right direction!

At the same time that her eating improved, Marlo started to develop loud, raspy breathing. The surgeon told me it was probably asthma from the reflux (which she was still having ñ and taking Prilosec for). I brought her to the pediatrician who took one look at her and told me something else was going on. She told me to set up an appointment with an ENT to have it checked out.

That night, April 6, Sherma was feeding Marlo while I cooked dinner. After Marlo finished eating she was sitting in her bouncy seat when she started fussing. I went to pick her up and she stopped crying but immediately starting turning blue. I yelled to Sherma who grabbed her from me, turned her over and slapped her between the shoulder blades. Marlo pinked up again for a moment and then turned blue again. I immediately called 911 ñ and asked Brent to take Bryan (now 2) out of the room. Sherma began rescue breathing while we waited for the paramedics. They arrived within seconds, got her breathing and rushed her to the ER.

After what seemed like an eternity, the doctors came and told me Marlo was fine. I went back to see her, and there she was, sitting up in her bed smiling. She was still doing that raspy breathing. They told me they needed to run tests.

The next day her pulmonologist, Dr. Bennie MacWilliams, did a bronchoscopy that revealed that Marlo had a huge piece of scar tissue on the inside of her trachea that had formed after the second TEF repair. The width of her trachea was around 6 mm, and the scar tissue took up about 4.5 mm, so when she refluxed, it completely blocked her airway.

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(Marlo's Story, continued from page 7)

They kept her in the PICU for two days and they called in an ENT, Dr. Patrick Connolly, to remove the scar tissue. Poor Marlo did fine during the procedure, but her little tongue swelled up afterward and she was miserable. Believe it or not, this was probably the most difficult time for us as parents. She was old enough to understand when Mommy and Daddy left her, and old enough to be frightened and uncomfortable. Our families and friends helped us out a lot but it was hard.

She was finally discharged on April 12, and we were so happy to bring her home. The first night she was home, she wailed for what seemed like hours. We thought she was disoriented. Brent's mom and I took turns rocking her for hours. I took her to the pediatrician for a quick check and she looked in her ear and found a horrible ear infection (courtesy of the PICU). That's why she was wailing!!

Marlo had a follow up brochoscopy a few weeks later and it revealed that the scar tissue had not grown back. Marlo was finally healthy.

The next winter she had a few colds, and then last winter only two. She is the happiest, smiliest, most easygoing kid you can imagine. She loves to cuddle with her precious blankie and keeps up just fine with her sister, Audrey, and her big brother Bryan. She loves animated movies and has learned her alphabet and loads of Spanish. She eats anything and everything you put in front of her—she's the best eater of all three children!

Marlo is totally caught up mentally and physically, although she is still quite thin for her age—32 pounds at almost 4 years old. Then again, her identical twin sister who has never been sick is exactly the same size!

You never really know why things like this happen, but Brent and I wouldn't trade one minute of it. Kids like ours, and yours, remind us how precious life is, and how blessed we are to get to be parents at all. It is hard but like so many things in this life that are hard, it is worth it.

When Marlo was in the PICU that last time in March, one of the pulmonologists took me aside and told me how much he admired how hard we worked to make sure Marlo got the best care. He told me that I had to be my child's advocate—and that she did well not only because of her strength, but ours too. It made us feel good and reminded us how important our job as parents was. Never be afraid to ask questions, to get that second opinion, to find someone more experienced, or just to ask for help when you really need it. And please don't hesitate to call me if you ever need someone to talk to. ea|tef

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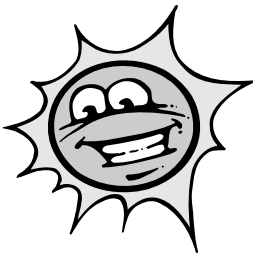
ea|tef

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!

(Seeing the World, continued from page 4)

traveled around Ireland, reveling in the rugged beauty of the country and learning a new appreciation for sheep. James did well up to the very last meal we shared in Ireland, a picnic on our way to the airport. He choked on some of his food and we used the portable suction machine for the first time in Ireland. At the airport, too, James had a choking incident, but then he drank some water and seemed all right.

On board the plane, we discovered that the oxygen tank we'd ordered wasn't stowed under our seats as it had been on the flight over. Getting it caused a brief delay, but we were glad once we were airborne, because the problem that had surfaced at the airport returned: James couldn't swallow any food. Even drinking was a problem. We used the oxygen intermittently during some of these "spells" and prayed for a speedy flight. Amazingly, none of the flight attendents even realized there was a problem. The plane was full and there were lots of children, including one in the seat behind us who threw up all over his mother. The staff had more than enough to keep them busy.

James was admitted to the hospital the day after we got back. Just hours before the surgeon was to perform a rush endoscopy, James coughed up a small patch of plastic wrapper and he was all better. It seems that during that last picnic lunch in Ireland, James had swallowed a bit of the plastic wrapping from the spoon we used for his yogurt. We got endless teasing for that one from hospital staff.

Leaking at 30,000 feet

James finally got to get rid of his g-tube when he was 4. It was a messy affair—the hole refused to close on its own so the surgeon ended up having to sew it closed. Shortly afterwards, we flew to St. Louis for a family wedding. James's skin around the g-tube site had been burned raw by the stomach acids that had been oozing out of the site before it was sewn shut. But now that it was closed and the skin was beginning to heal, I packed without bringing along any skin barrier creams or gauze bandages.

During the flight, I noticed that James's shirt was damp. Hmmm. I smelled the shirt. It smelled of vanilla, the flavor of the formula James had been drinking. Inspecting his belly, I saw a fissure where the hole had been sewn. There was a leak! I was furious and crazed, knowing how horribly uncomfortable James was going to get if the skin around the site was damaged further by leaking stomach acids. While changing planes in Chicago, I placed a frantic call to Neal, who was flying in that night, and gathered what supplies I could (not much) to protect James's skin until we arrived in St. Louis.

By the time we got to St. Louis, James was just starting to be uncomfortable. Neal, meanwhile, had worked a little phone magic. It turns out that the surgeon on call in Rhode Island had trained in St. Louis, at Washington University Hospital. She immediately called the hospital and placed the emergency room on alert for James. I still had to rent a car, drop off our older children with family and find the hospital with a now-miserable James in the back seat.

The hospital visit couldn't have gone smoother. The emergency room had valet parking, the first time I'd seen that, and they were indeed ready for James. We were seen within minutes and the surgical fellow who met with us was kind and very willing to help. After considering the options, however, we decided to hope that pressure bandages would staunch the flow of stomach contents for the weekend until we could get back home and have the leak repaired. Someone from the surgical team even called us the next morning at our hotel to see how James was doing. I was impressed and grateful.

The pressure bandages worked and James made it through the weekend with no real discomfort. The g-tube site was resewn when we got back home and that truly was the end of it. Trips since then—to Wyoming, to Arizona, to California, to Washington D.C.—have been wonderfully uneventful. We're not ready to take James to the ends of the earth, but we are still the traveling McNamaras and that suits us all. ea|tef

Lessons learned

1. Do your research. Find the names of pediatric hospitals and doctors in the places you are traveling and bring phone numbers with you. (Your own doctors are the best place to start for this information.)
2. Bring help if possible. When we went to Ireland, we realized that if something did happen with James, it would be helpful to have another adult along to stay with the older children. A family friend came along and was a wonderful extra pair of hands the whole trip. It worked out so well that she has been with us on subsequent trips as well.
3. Always carry a copy of your child's medical history. Not only will it supply important information, but it will signal to new medical personal that you are knowledgeable about your child's condition.
4. If your child has a g-tube, always carry an extra gastrostomy kit with you. If no one has taught you how to replace a g-tube, it's time you learned. It's very easy. We have all spent enough time in emergency rooms with these kids. Give yourself and your child a break from that torture.

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.

This Just In... off our own website!

Browsing through the Discussion Forums of our website, I found many interesting items, some of which are shared here. The story below is from Jeane, a 46-year-old ea|tef survivor. It's always good to read about ea|tef adults who are doing well.

I discovered this site today, and I just had to post my story. I was born in January 1958, with type C ea|tef, however it was five days before the problem was diagnosed. At that point I was rushed to the Medical College of Virginia where Dr. James Brooks and staff performed their miraculous surgical procedure. But after those five days of gastric juices spilling into my lungs, I had double pneumonia and my recovery was slow.

My post-op included 24-hour nursing care. Dr. Brooks knew my parents didn't have the money to continue such treatment, and my prognosis was poor. He suggested to my dad to consider discontinuing the 24-hour care, but my dad said, "No!"

After a month in the hospital, I came home to a mother who had never held me. She cringed every time she picked me up, because her hand would slide into the deep gash under my right shoulder. For her it was a constant reminder of the pain and suffering I had endured.

My parents were assured that I would spit up at least one feeding a day. However, to keep them guessing, I would never spit up the same feeding!

For the next 10 years I went for annual check ups and barium swallows/x-rays at MCV. After that, Dr. Brooks was satisfied and discharged me from his care. Aside from the check ups, I had a normal childhood. My recovery was much harder on my parents than it was on me!

I don't feel that I've gone through so much. I was a baby and I don't remember any pain or suffering. I have the physical scars that remind me, but I've had them all my life. They are a part of me and why I am alive. They are my "beauty marks"!

Today I'm living a wonderful life full of adventure. I snow ski, scuba dive, fly sail (glider) planes, and have traveled to a number of places throughout the world. Life is beautiful!

Several people on the website have written about how to pay the crazy expensive medical bills we all incur with our ea|tef kids. If your child is still in the hospital, the best help could be close at hand, through a hospital social worker. Here's more information from Michelle Kiger, a local ea|tef coordinator from Indiana, who responded to one person's request for help with insurance.

I realize the toll of medical bills can place a major stress on your family. Try to apply for any and all programs that you can—every state has different programs and guidelines so I cannot give you full details but definitely look into them.

The best place to start is with your local division of Family

(Continued on page 12)

Kids Speak Out

Living the ea|tef Life

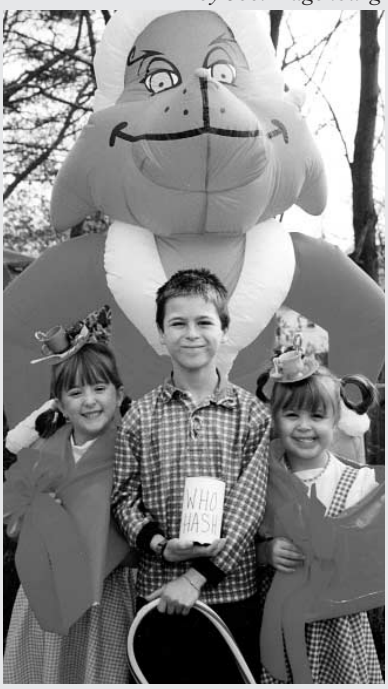
Hey Kids—We Want to Hear From You!

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.

Rotor Rooters, *reflux and regular life*

by Joel Hagenburg



Hi, I'm Joel. I'm 12 years old and I have esophageal atresia like some of the other kids I've read about in the ea|tef newsletter.

As a little kid it was rough, being in the hospital day in and day out. Now that I'm older, Hasbro Children's Hospital isn't a second home to me, it's just a place to visit. I go about every nine months for an endoscopy or "rotor rooter," as my mom calls it, and occasionally for a check-up.

When the newsletter arrived today, I was doing my nebulizer treatment, which I haven't done in a while. I got a little cold and that affected my asthma, which turned it into a big cold. I do some sports, but I'm kind of small so baseball is just the sport for me. Besides, I'm good at it. I have a hard time with reflux, and I also have trouble playing in the summer heat because my blood sugar goes haywire.

My doctors found out a couple of years ago that I have Pediatric Dumping Syndrome. Pediatric Dumping Syndrome means that my body can't process sugar right. What do you think the first study for it was? They fed me sugar and sugar and more sugar. I got really sick. Now, I never want milkshakes and candy bars again. I test my sugar level with a kit called One Touch Ultra. I'm not diabetic, but I have diabetes-like symptoms. I have to monitor my sugar and watch what I eat very closely.

My doctors say that I got the dumping syndrome from all of the surgeries I've had on my stomach. The last surgery, my second fundoplication, was about four years ago.

Usually I'm great. I've learned that I have to be careful when I'm eating because I'm prone to choking—meats have the most chance of getting stuck. During the fall and winter months I can get very phlegmy, and that's when the rotor rootering comes in, but my parents have helped me every step of the way, and I have great doctors too!

It's not so bad being an esophageal atresia kid. I guess it's all what you get used to!

Joel Hagenburg lives in Coventry, Rhode Island, with his twin sisters Nicole and Noel and his parents Bill and Donna.

(Surfing the Web, continued from page 10)
& Children. Definitely apply for Medicaid as a supplement to your primary insurance. To that end, make sure you are very detailed about her health history, and get statements from her doctors. The process can take a while, so you need to get started immediately. We did this for Noah and the coverage was retroactive back to his birth! This was a Godsend for us as they picked up most everything our insurance did not!

Medical condition, not income, is usually the qualifying factor for a state's Medicaid program. Some children may also qualify for SSI (supplemental security income), but this program is based on household income and many ea|tef families will not qualify. There may be other resources you may find, but you will need to do research. Try contacting your local state representative and explaining your situation. I am sure he or she would be glad to locate the programs for you!

Anyway, hope this helps you get started, but there is something else to keep in mind: Pay only what you can on the medical bills. As long as you are attempting to pay them —no matter how much you send—you cannot get in trouble. So, try not to get too stressed about it. If you are badgered about payments, tell them you are going to contact your state's Attorney General because you are attempting to pay the bills and they are harassing you! Take care!

Michelle Kiger, proud mom of Bethany (9), Chelsey (6), and Noah (age 4—"Type A" ea (5 cm), severe reflux, Nissen Fundo, severe tracheomalacia, tracheostomy removed 6-01, G-tube removed 9-01, and wants to be a cowboy!)

—michelle@eatef.org

Sharing our stories is much of what our organization is about. The journal has always done that, but now with the website there are more personal stories than ever before. The one below recounts those early moments we all have seared into our brains—diagnoses out of the blue, separations from our babies, surgeries.

I am a new person on this site and would like to tell you my story. My pregnancy started off fine until I was 10 weeks along and I started to lose a lot of water and started to bleed. This of course had to happen out in the middle of nowhere. I was lucky enough to have a friend with me who had a cell phone. We called an ambulance and I went for a ride.

At the hospital I was told I had placenta preevia and would lose my baby. Of course I was sad but was not going to give up hope. I was to take it easy (hard to do with three other kids around). Anyway, to make a long story short, I was on bedrest throughout the pregnancy and had several ultrasounds saying I was having a girl. During the last week of the pregnancy I went to the doctor and was told I was gaining too much weight—approximately five pounds a day—too much for my little 5'2" body (I'd started out weighing 105 pounds and ending up 150!). The doctor then told me that I had to be induced. So, on Jan. 22, 2002, I gave birth to a 7 lb., 7.7 oz. baby boy!

After all of the excitement of our son's birth, he was taken to the nursery, where they found out about his ea|tef. The nurses were trying to clear the mucus from his throat and the tubing would not go down. Meanwhile I was being wheeled into the surgery room to have my tubes tied when I was told the news about my son. After my surgery I was told that my

son was being flown to St. Joseph's Medical in Phoenix. I could not go with him due to my surgery.

To finish the story, my son spent seven days in the neonatal intensive care unit before coming home. I can't say that it has been a nice smooth ride for the first two years of his life (or mine). He has had three dilations since his surgery. I am hoping for the best from here on out, but I know there may be more troubles.

One mom on the website asked about barium swallows. Her child was scheduled to have one but it hadn't occurred to her until later that she didn't even know what the test involved. (How many times have we all walked away from a conversation with a doctor only to realize that we still had questions!) You'll find one of the answers to her query below, complete with a terrific web site suggestion.

Becky,

An upper GI study is when they have the patient swallow a barium solution that will show up on x-ray. They are able to visualize on x-ray what happens to food when the patient swallows it. They can follow the solution down the esophagus, into the stomach and small intestines. If there are any problems—like reflux—they will be able to see it happening. Our son had this done. I was able to be there with him and watch the little TV screen to see the barium go up and down his esophagus instead of into his stomach. I'll never forget it.

A cool site I found for you is: http://www.mayoclinic.com/. Click on "H" for Heartburn/GERD.

Check it out. Scroll down towards the bottom—there is a button to click on to actually SEE a barium swallow and hear an explanation of what is going on!

Hang in there. These bumps are disconcerting when you think you might actually be heading out of the woods. You are heading for the clearing… I promise! Before long everything will be fixed and working great.

— Glenda

Mom to healthy 17 yr. old TEF|EA, PDA, ASD, VSD, laryngeal cleft, severe tracheomalacia, massive reflux, post-tracheostomy, postop Nissen fundo, bilateral inguinal hernias, scoliosis.

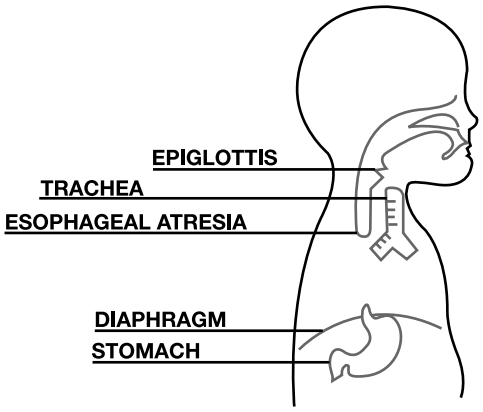
What is EA|TEF ?

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.

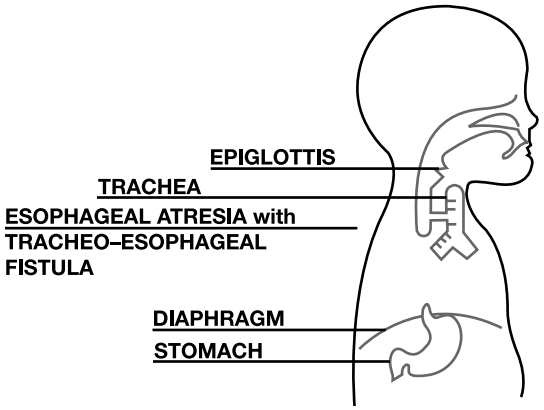
Second, tracheoesophageal fistula (TEF): The trachea (or windpipe) is the passageway through which the air we breath travels to the lungs. In TEF, there is a connection between the trachea and the esophagus, so that potentially either air can pass from the airway into the stomach, or food can pass from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

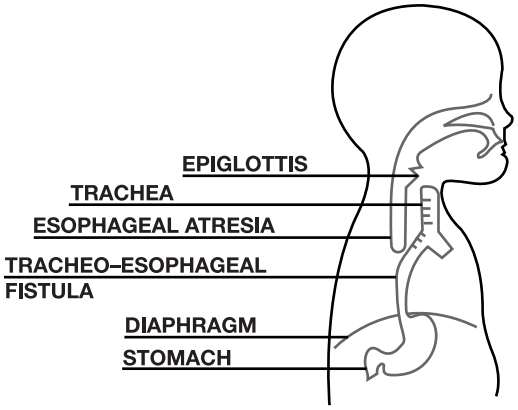
Below are diagrams of the five different types of EA|TEF.



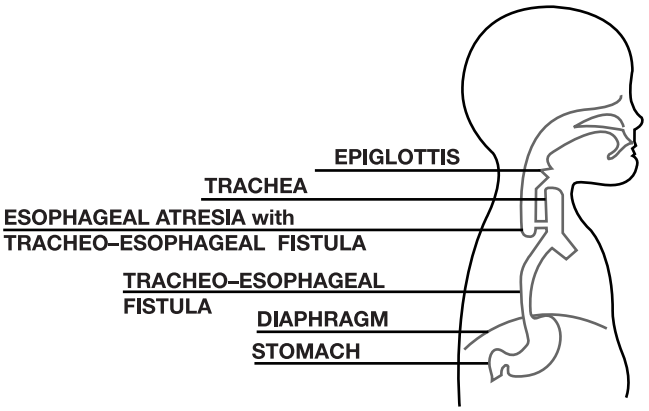
Type A Esophageal Atresia (7.7%) Both segments of the esophagus end in blind pouches. Neither segment of esophagus is attached to the trachea.



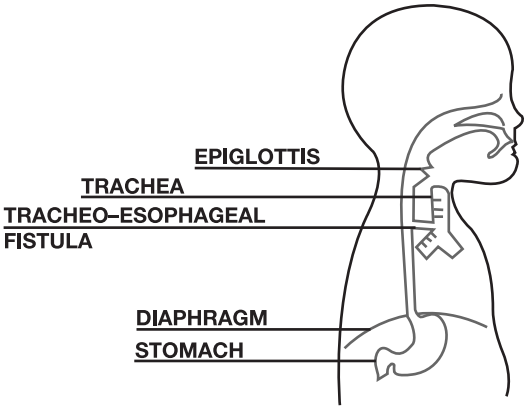
Type B Esophageal Atresia with Tracheo-esophageal Fistula (0.8%) The upper segment of the esophagus forms a fistula to the trachea (TEF). The lower segment of the esophagus ends in a blind pouch (EA). This is a very rare form of EA|TEF.



Type C Esophageal Atresia with Tracheo-esophageal 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 Tracheo-esophageal 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.

Reflux 101, Part 1

Gastroesophageal reflux disease (GERD) is a condition in which the acids from the stomach move backward from the stomach into the esophagus (an action called reflux). Reflux occurs if the muscular actions in the esophagus or other protective mechanisms fail. The hallmark symptoms of GERD are heartburn, a burning sensation in the chest and throat, and regurgitation: a sensation of acid backed up in the esophagus.

When a person swallows food, the esophagus moves it into the stomach through the action of peristalsis, wave-like muscle contractions. In the stomach, the starches, fats, and proteins are broken down by acid and various enzymes, notably hydrochloric acid and pepsin. The stomach has a thin layer of mucus that protects it from these fluids. If acid and enzymes back up into the esophagus, however, its lining offers only a weak defense. The esophagus is protected using specific muscles and other factors.

The most important structure protecting the esophagus may be the lower esophageal sphincter (LES). The LES is a band of muscle around the bottom of the esophagus where it meets the stomach. The LES opens after a person swallows to let food enter the stomach and then immediately closes to prevent regurgitation of the stomach contents, including gastric acid. The LES maintains this pressure barrier until food is swallowed again.

If the pressure barrier is insufficient to prevent regurgitation and acid backs-up (reflux), then peristaltic action of the esophagus serves as an additional defense mechanism and pushes the contents back down into the stomach.

Conditions Associated with GERD

Esophagitis: In most people, GERD symptoms are short-lived and occur infrequently. In about 20 percent of cases however, the condition becomes chronic. When the acid causes irritation or inflammation, the condition is called esophagitis. If the damage becomes extensive and injures the esophagus, the disorder is known as erosive esophagus.

Non-Erosive Esophageal Reflux Disease: Symptoms of gastroesophageal reflux disease can occur without any signs of inflammation or injury to the esophagus. This condition is tentatively referred to as non-erosive esophageal reflux disease (NERD). NERD rarely progresses to full-blown GERD. In NERD, patients have no signs of inflammation or erosion in the esophagus, but they experience certain symptoms of GERD, such as burning sensations behind the breastbone for at least three months.

Barret's Esophagus: In a small percentage of chronic patients, a serious form of GERD called Barrett's esophagus may eventually develop, in which erosion can lead to cancerous changes in the tissue lining of the esophagus.

Causes of Reflux

Malfunction of the Lower Esophageal Sphincter (LES) Muscles: The band of muscle tissue called lower esophageal sphincter (LES) is responsible for closing and opening the lower end of the esophagus and is essential for maintaining a pressure barrier against contents from the stomach. It is a complex area of smooth muscles and various hormones. If it weakens and loses tone, the LES cannot close up completely after food empties into the stomach. In such cases, acid from the stomach backs up into the esophagus. Dietary substances, drugs, and nervous system factors can weaken it and impair its function.

Impaired Stomach Function: A study showed that over half of GERD patients showed abnormal nerve or muscle function in the stomach. These abnormalities cause impaired motility, which is the inability of muscles to act spontaneously. The stomach muscles do not contract normally, which causes delays in stomach emptying, increasing the risk for acid back up.

Abnormalities in the Esophagus: Some studies suggest that most people with atypical GERD symptoms (such as hoarseness, chronic cough, or the feeling of having a lump in the throat) may have specific abnormalities in the esophagus.

Hiatal Hernia: The hiatus is a small hole in the diaphragm through which the esophagus passes into the stomach. It normally fits very snugly, but it may weaken and enlarge. When this happens, part of the stomach muscles may protrude into it producing a condition called hiatal hernia.

Genetic Factors: Studies suggest an inherited risk exists in many cases of GERD, possibly because of inherited muscular or structural problems in stomach or esophagus.

Asthma: At least half of asthmatic patients also have GERD. Some experts speculate that the coughing and sneezing accompanying asthmatic attacks cause changes in pressure in the chest that can trigger reflux. Certain asthmatic drugs that dilate the airways may relax the LES and contribute to GERD. On the other hand, GERD has been associated with a number of other upper respiratory problems and may be a cause of asthma, rather than a result.

Other Causes of GERD: Weakened peristaltic movement in the esophagus may contribute to GERD. If the mucous membrane is impaired, even a normal amount of acid can harm the esophagus. Pressure on the abdomen caused by obesity and also wearing tight clothing can contribute to acid backing up into the esophagus.

In the next issue, we will discuss treatment options for reflux. This information was taken from the Evanston Northwestern Healthcare website:

www.enh.org/WellConnected/articles.

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