ea|tef

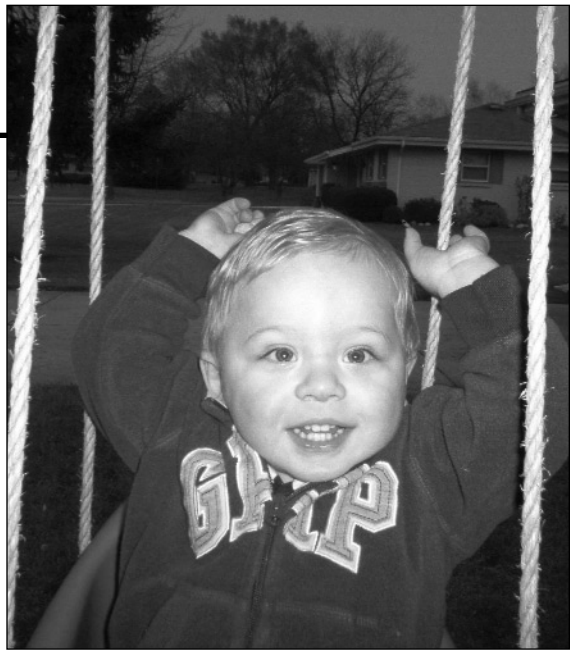
Child and Family Support Connection, Inc.

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

INSIDE

Colton's Story

By Debra Posanski



In April 2004 my husband and I discovered I was pregnant, again. There was no reason to believe this pregnancy would have the same outcome as the one before, which had ended in a very early miscarriage. Still, we were very nervous. About six weeks into the pregnancy, I began to bleed. Wearily, I went in for an ultrasound. Fortunately everything was fine. At 28 weeks, the baby was measuring large, so I went in for another ultrasound. Again, there was nothing to be concerned about. Thirty-six weeks came along and I developed preeclampsia and by thirty-eight weeks I was induced. My labor lasted about twelve hours, with the last two hours pushing.

Colton Michael was born on Jan. 3, 2005. He weighed in at 6 pounds 10 ounces and 18 1/2 inches long. When they handed him to me, I was so relieved that he was pink and breathing. I had not fully allowed myself to believe everything was going to be fine, but here he was, perfect in every way. They took him from me after about five minutes to clean him up. My husband Craig and my mother left to make phone calls.

As they returned, a pediatrician walked up and asked to speak with just the parents. The doctor told us he was called in to look at Colton because he had an unexplained rash all over his body. They didn't know what it was and wanted him to be admitted in the neonatal intensive care unit and put in quarantine. They could not risk any other babies coming in contact with him. We were definitely concerned, but how bad could a rash be?

After I was cleaned up, we stopped in the NICU on the way to our room. We were told to dress in protective gowns and gloves. Before we entered, the doctor told us more. He explained that when he was checking on Colton, he had stopped breathing. They had tried to suction him with different-sized catheters but the catheters couldn't get through. The doctor suspected Colton had tracheoesophageal fistula/esophageal atresia, in which the top portion of the esophagus ends in a blind pouch and the lower part attachs to the trachea. He said Colon needed to be transferred to Children's Hospital within a few hours. He described a surgery to repair the problem and told us that the recovery would take two to three months.

We finally walked in to see our little guy tucked away in the corner of the dark empty room. Craig and I stood nearby, unable to touch his little body. We prayed with him as tears were running down our faces. We were in complete shock. Everything seemed to be running in slow motion. Back in my room, we lay in bed next to each other silently sharing our anguish.

Three hours later, we revisited the NICU to see Colton off. He was in a transport incubator getting ready to leave. Craig and I huddled around him to pray, told him how much we loved him and stepped aside. We followed him out as far as we could go. Then, they turned left and we turned right. I was in such despair. This was not supposed to happen. My baby was leaving and I was not going with him. He was alone, and so were we.

(Continued on page 10)

Editor's Letter

It's about time this journal came out! The last issue of the journal was mailed two years ago. So, what's the problem? One thing is true: your editor has good intentions but seems to need someone to hold her feet to the fire in order to get the job done. Around my house, something always needs my attention before I get to my ea/tef duties.

But there's something else at work here. We are so much more connected via the internet these days that the journal is no longer as important as it was just nine years ago, when I first picked up a copy. The discussion forums on the ea/tef website serve to link and support parents and adults with ea/tef far beyond the occasional contacts that were possible a decade ago. I'm on the internet daily but I still get most of my information from newspapers. Of course, I'm 46. The reality is that new parents of ea/tef kids are more comfortable getting their information from the computer. The discussion forums at eatef.org are helping families in remarkable ways. Just a quick look at a couple of entries shows how users help each other. Dianna Q, for instance, posted an entry on Nov. 5 seeking advice about her son's possible pneumonia after an endoscopy. Lansima responded the next day, writing that it may not be pneumonia that is making her son sound so terrible. She wrote that her own son always sounded worse up to a month after a procedure, the result of trauma from insertion of the tube, not from pneumonia. That same day, another participant, SaraBeth, wrote that her own son had a probable case of pneumonia after a surgery and that it was treated with antibiotics. Later that day, Dianna Q wrote that she'd just gotten back from the pediatrician, who said that her son's lungs sounded clear. Just a few quick entries in a span of just over 24 hours, but it saved Dianna Q perhaps from rushing to the emergency room.

My only complaint about the discussion forums is that they offer loads and loads of personal information with very little hard fact. That's the reporter in me. But it seems ridiculous to wait around for a slow editor to put together a journal to get more factual information. The website needs to feature articles written in a timely fashion (note to self) that can be posted immediately.

I know there are many parents and ea/tef adults out there who are up to the challenge to helping put this together. If you are interested in writing for the website and have some experience with reporting, or if you are a web designer and want to help created a web-based journal, please contact me e.mcnamara6@verizon.net. It's time to move into the 21st century!

Best,
Elizabeth McNamara
December 2006

Smile!

Last year, we were at my brother-in-law's for a birthday party and they had a little baby pool set up for the little kids. The sun started to set, but my 3-year old was still in there, although shivering. His uncle said, "Justin! You'd better get out of there before you catch pneumonia, boy!" and started wrapping a beach towel around him. Justin replied, "I don't get pneumonia! Ethan does!" Ethan is Justin's little brother, who was born with type A ea and, no surprise, has had pneumonia loads of times.

Kim Mills
Jefferson, Maryland

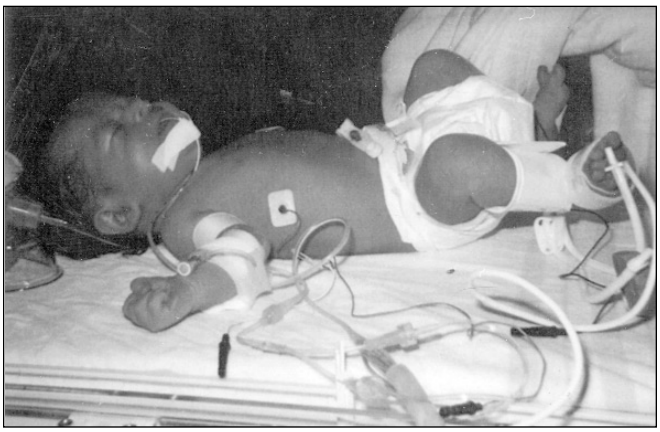
Valerie's Story

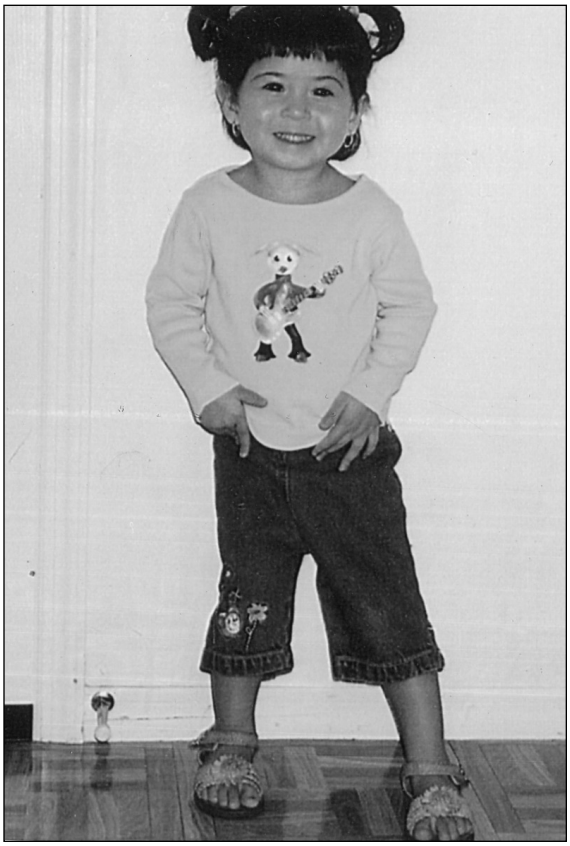
When my husband and I decided to have a baby, we never thought of all the pain we would have to endure. I had a healthy pregnancy without complications until the day of labor. My doctor decided to induce my labor a week early because my belly had grown *very* large. All was going well until my blood pressure started to go up. I was in labor for eleven hours and the baby was in position, but could not come out.

At this point, the doctor decided to perform a Cesarean. Valerie was born November 14, 2000. She was beautiful and had lots of hair. As they were cleaning Valerie, they noticed that when they suctioned her lungs, the lungs would fill up again and she would have trouble breathing. After repeating the procedure several times, they decided to entubate her (put her on a ventilator). They found that the breathing tube was not going through and that there was an obstruction. Valerie was taken for x-rays and they found what they suspected: ea/tef.

After that, Valerie was tested for other abnormalities. They found one extra rib and a right arching aorta. Of course, my husband and I were in shock with all this. We'd never heard of this condition before. The doctors explained to us about it, but it still was very scary thinking that there was a danger that our daughter might not survive.

Valerie was transferred to another hospital and was scheduled for surgery the next day. This was very hard for us, especially for me, because I had only seen Valerie for a few seconds after she was born. I had not held her or even touched her. I was terrified that she might die. Thanks to the almighty and powerful God, Valerie survived the surgery and it was successful. The pediatric surgeon, Dr. Mark Levy,





talked to my husband after the surgery, giving him the details. Valerie's esophagus was repaired without complications. She improved every day. It was great news, but we still had to go through a lot that we did not know about at that time.

It was two weeks after the surgery before I could hold Valerie. I was very nervous and was afraid I would hurt her. I cried many hours and could not believe that my little baby had been through such a serious operation. She was released one week later. We took her home with an apnea monitor for her heart rate and lungs. I was happy to have her home, but very scared. We had to take her back to the hospital in two weeks to start dilating her esophagus. The surgeon started them twice a week. After a month, they went to once a week, then once every two weeks, then once a month until she was six months old.

The doctors thought Valerie was doing so well, they wanted to wait two months before performing another dilation. The monitor was removed and she was breathing and swallowing

(Continued on page 6)

The cough that stops traffic

There are so many things to learn when you have a child born with ea/tef that certain aspects of the condition remain a little fuzzy for a while. You figure out the eating stuff quickly. And surgeries, you focus on those too. But you keep hearing one word that you can't quite keep in your head, the whole word anyway: *trachea-something*.

You start to focus a bit more when you notice that your adorable little cherub sounds like a walrus when she coughs. Or when, after surgery, the doctors can't get little Jimmy off the ventilator for a while. His airway keeps collapsing when they remove the breathing tube. That's when a lot of us take out a pen and paper and ask the doctor to spell out that word: *tracheomalacia*.

The word "malacia" (pronounced: muh-lay'-she-a) means softening.There are three places where a person's airway can be soft: the voice box, the windpipe, and the breathing tubes. A person may have softening in one, two or three parts of the airway. People with ea/tef most commonly have softening of the windpipe, or trachea, hence the name tracheomalacia. Soft airways do not hold their shape, and air does not flow through as well as it should.

With tracheomalacia, you may hear a high-pitched, crowlike noise when your child breathes in (stridor). The child may also wheeze, cough or choke. Severe tracheomalacia can also cause very short spells of no breathing (apnea). Your child may turn blue during these apnea spells.

Life with tracheomalacia has been described as what happens when you drink with a paper straw. After a while, the straw gets damp and no longer holds its shape. It collapes when you try to drink from it. Imagine your trachea as that wet straw. The worst part? The harder someone with tracheomalacia tries to breath, the more the "straw" collapses.

When children with soft airways are active, upset, eating or have colds, their breathing may be noisy. When they are calm and lying on their stomachs, their breathing may be quieter. Some babies may hyperextend their neck in order to adequately keep their airway open. As children grow, their airways get larger and firmer. In most children, soft airways are gone by two years of age. Some children may have noisy breathing until they are school age.

If children can breathe and are growing, their soft airways are not treated. They will grow out of it. If your child has trouble breathing because of soft airways, your child's doctor may ask you to see an ear, nose and throat doctor or a pediatric pulmonologist for treatment. The doctor may do a test called a *bronchoscopy* to look at your child's airways.

Thanks to Marsha (from the ea/tef website discussion forums), mother of Logan, for some of the information in the article, as well as for a couple of good referrels. These websites were also used: www.phoenixchildrenshospital.com and www.emedicine.com

The Cough

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

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

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

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

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

—Elizabeth McNamara

The Joys — and Perils — of Eating

Most ea/tef parents probably remember the first time their child took nutrition by mouth. With surgeries, leaks, delays, illness, it is sometimes months before we are able to put a nipple in our child's mouth. Even if it's only a few days, the wait is agonizing. Eating is such a basic part of life. So that first sip is momentous.

But the fact is, food for our ea/tef kids can be a double-edged sword. Some have little or no difficulty transitioning to oral nutrition after surgery, not bothered by motility issues and strictures. The rest of our kids, however, must learn how, what and when to eat so that food goes down and stays down.

Years ago now, my son's surgeon explained esophageal motility (movement) to us this way: "You and I can eat standing on our heads. James won't be able to do that." In other words, the coordinated, rhythmic muscle contractions (the "peristalsis") of a typical esophagus will provide the necessary pushing motion to move food into the stomach even if we are upside down. But people with ea/tef usually have terrible motility and no peristalsis. They must rely on gravitiy to bring food through the esophagus to the stomach.

HOW TO START

Judging from the discussion forums on our website, parents figure out pretty quickly what foods work and which ones to stay away from. Number one slider: smooth yogurt. My son James still eats loads of the stuff. And it can be mixed with all sorts of foods. Purees work well too, store bought or homemade.

Still, there comes a time when you want your child to experience biting something. Actually, speech and occupational therapists make clear the connection between eating and speech development. Turns out, we need to strengthen the muscles in our mouth and jaw to be able to speak. Luckily, there are choices out there for children who are just starting to move away from purees or for those children who are not able to tolerate many solid foods.

Puffed foods are a great way to help your child begin to bite and enjoy foods. Many people posting on the website sing the praises of Gerber Fruit and Veggie Puffs. For a less expensive and saltier experience, cheese puffs are a tasty (if often messy) alternative. Veggie Stix are yet one more type of puffed food that most ea/tef kids will be able to enjoy. Only slightly more challenging are Ritz-type and graham crackers, which provide a satifying crunch.

The actual weight of food can sometimes make a difference. James once choked on scrambled eggs. His doctor suggested that the eggs were too "light." Puffed foods are impossibly light, of course. The difference: puffs dissolve completely. Bits of cheese may be a better alternative to eggs. Cheese tends to kind of melt as it goes down, as long as the pieces aren't too big.

Breads tend to be challenging for our kids early on, unless they are the banana-bread variety, i.e. more like a

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Strictures

Strictures develop post surgery usually because scar tissue grows at the surgery site. Scar tissue, sadly, is inevitable, but not all ea/tef children are affected by it. Some have their esophagus repaired and never look back. Others need a few post-op dilations early on before being able to take the surgeon's phone number off the speed dial.

A few ea/tef children need to be dilated even years after surgery. My son is one of those. After the dozens of dilations he had early on, going in twice a year to be dilated seems pretty manageable, if not exactly fun.

How do you know when your child has a stricture? It can be immediate — whoops, she swallowed a cherry! After a few fruitless hours of trying to dislodge it, you know you have to call the surgeon. Alternatively, sometimes it happens over weeks and months, so it can be hard to pinpoint when Julia or Sam started having trouble with certain foods. And other things can cloud the picture. Colds and allergies can interfere with motility, coating the esophagus with thick mucus that makes it harder for the food to go down. You may think your child's esophagus isn't working very well, but when the cold passes, suddenly everything is sliding down again. Eventually, though, there can be the "aha" moment, when you realize that your child isn't sick and isn't eating like he used to. Time to call the surgeon.

Some doctors will want your child to have an upper g.i. study (i.e. a barium swallow) to have a look at the esophagus. This isn't a bad idea, but these studies don't always tell the whole story. Barium (a thick, chalky liquid visible on x-ray) is not regular food. Your child may have a stricture even though the upper g.i. study looks basically okay. You are in the best position to judge. Don't be afraid to question your child's doctor if you think there's a problem that's not being addressed.

(Continued from page 5)

cake. One rule: if it has yeast, stay away from it. Not because of the yeast, but because of the texture. Yeasted breads just don't dissolve very quickly. For that reason, for instance, pancakes are more popular at our house than French toast.

As it happens, my ea/tef child likes veggies more than my other children. People are amazed when I tell them that James loves to eat raw carrots. Yes, he can eat them. They just need to be sliced into thin sticks, chewed well and taken with lots of drinks in between bites.

Meats are usually the final frontier. They don't dissolve and they require studious chewing. Deli meats are a good start. Beware of hotdogs, which are the perfect size to get lodged in the esophagus. Again, chewing is the answer. Be patient and try to think outside the box when it comes to protein. For instance, tofu is a great alternative for ea/tef kids. It's slippery, breaks up easily and can be flavored in many different ways. Dried beans, mashed or cooked until very soft, are also worth trying.

SOME TIPS ON METHOD

There is nothing worse for a person with a lousy esophagus then to gobble down food. What you put in your mouth is only part of the equation. How fast you eat, how well you chew and, again, how much liquid you take with your meals can make all the difference. Having your child use a small spoon, even as he or she grows, can help by literally slowing the rate of eating. Small portions also help. Reassure your child that he or she can always have seconds (or thirds!) as long as the food is going down.

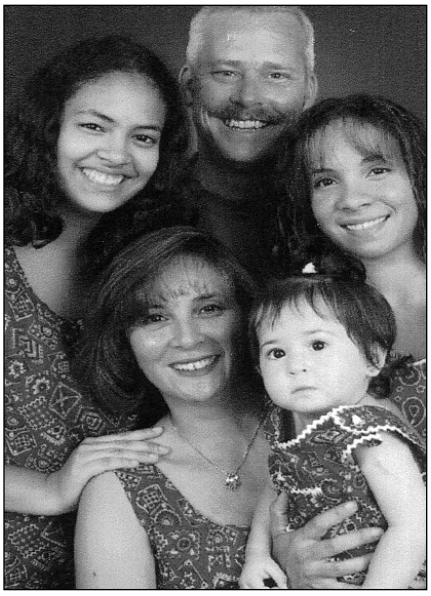
Over and over, parents stress the importance of having their ea/tef child wash down food between bites. Some parents develop a little song, others simply repeat the "wash it down" mantra. Teaching your child to drink between bites, to eat slowly, and to chew well will help them to become independent eaters.

If there is a question as to whether your child is chewing well enough, consult with an occupational therapist. Some children who spend time in the hospital and have had lots of tubes in their mouths become "orally defensive,"—i.e., they don't want to put anything in their mouths. Many hospitals now have "feeding teams" to work with you to help your child overcome his or her defensiveness and learn to chew and swallow. Eating is a wonderful thing and our ea/tef kids usually need every calorie. Try to make food and eating fun — you and your child will reap the rewards.

(Continued from page 3)

on her own. Valerie responded well to cereal and baby food, then small pieces of cookies and bread. We were very happy with her progress.

Valerie's motor skills were not totally developed for her age; therefore she started receiving physical therapy each week. She began to improve. When Valerie went for her dilation at 8 months old, Dr. Levy told us that her esophagus looked good, it needed very little stretching. He wanted Valerie to have an esophagram (a test that shows swallowing and food going down the esophagus) every two to three months. Well, a couple of tests were done and no more dilations were needed.



Valerie, nine months old, with her parents and sisters Veronica and Beatriz.

Today, Valerie is doing very well. We have two older daughters and they are so good to her. They love her very much. If it was not for them, for the love they have for her, we would not have been able to bear all the pain we went through. God gave me, through them, hope and strength to keep on going and to wait with faith.

Valerie is 6 now and she is a happy and healthy child. She is Daddy's little girl, Mommy's princess and a lovable sister. She is eating well and growing. We thank God for letting us have her, for teaching us that life is precious and that there is no wasted time when there is love and faith.

Bethzaida Arroyo-Gentry
Kissimmee, Florida

Smile!

My son, Nick, will sometimes do anything to get out of eating something. More than once, he's said he can't eat because he has a "broken esophagus." Nick, however, was not born with ea/tef — his baby sister, Alexandra, was. Needless to say, when I mention chocolate chip cookies for dessert, suddenly his "esophagus better now."

Jacqui Sept. 3 '05
eatef.org/forums

Mathew's Story

By Diana Wells

Mathew Robert Wells was born on August 17, 1996, at Evanston Hospital, in Evanston, Illinois, at 34 weeks. He weighed 4 pounds, 1 ounce, and had esophageal atresia type A (no fistula) and Down syndrome with no other complications.

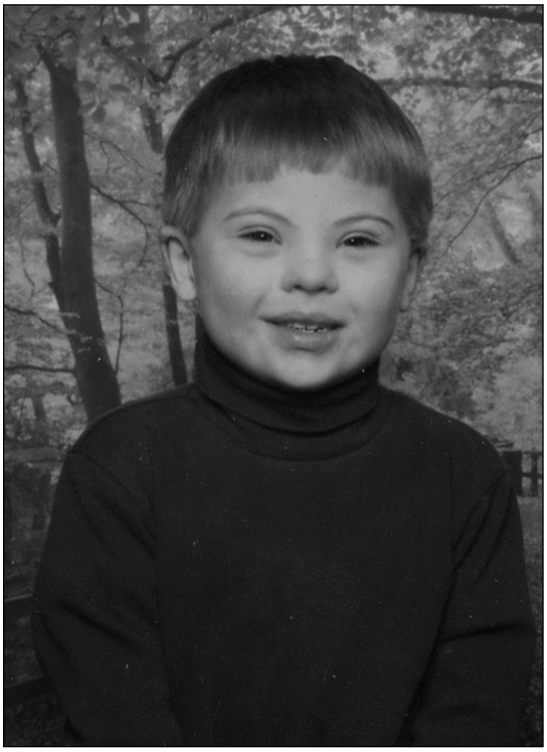
Matt had surgery to place a g-tube at two days old and came home in late September. He had surgery to connect his esophagus on Dec. 3 at Children's Memorial Hospital in Chicago, using only his esophagus and no colon. Because of post-surgery complications of staph and double pneumonia, his 10-day stay was extended to two and a half months — our own personal hell on earth. By the grace of God and help of the doctors at Children's, especially Dr. Juda Jona (our "guardian angel"), we made it through. There were six dilations after the surgery, the last one in May 1997. Matt's g-tube was removed in July 1997.

Matt was eligible for occupational, physical, speech and education services right away. Between the nurses that were assisting with Matt's care at home and his therapy services, we had a revolving door at our house until Matt turned 3.

We moved on to the early education phase of Matt's life. He attended Lakeland School in Walworth County in Elkhorn, Wisc., a school for children with special needs. Our own school district did not have an early education program. That first year, Matt went to school three days a week for half days. Each year, he went for longer, until he was enrolled in a regular kindergarten class with a full-time aide.

Today, Matt is in a regular fourth grade classroom with an aide. He has blossomed this year and plays soccer and baseball too. A couple of years ago, we noticed that Matt was having problems with reflux. He would lie down in bed, start coughing and food would come up. Sometimes, it would be brown colored and we realized there was blood. That's when we consulted Dr. Colin Rudolph, a gastrointerologist. After an esophagram, Dr. Rudolph put Matt on Previcid. Today Matt takes 30 mg a day in dissolveable pill form. We go back once a year.

Although we still pay attention when Matt eats, he has gotten better at handling his eating himself. He eats anything and everything. We just make sure there is fluid around when he eats. For years we were telling him, "chew, chew, chew" and "drink often." When something does get stuck, Matt usually projects it out, which sometimes causes school officials to thing he's got a stomach virus. It took a while but with the help of Dr. Rudolph's terrific nurse, Judy, it is now written into Matt's education plan that symptoms beyond just "throwing up" must be established before he needs to be sent home.



Mathew Wells

A couple of years ago, we were at a birthday party and another guest was a gentleman in his 70s who had had esophageal problems all his life. Pork sandwiches were served and this man wasn't chewing well enough and something got stuck while he was eating. Meanwhile, Matt was eating Goldfish crackers and not drinking, so he ran into problems too. Both this man and Matt had to adjust, with the man chewing more carefully and Matt eating more slowly and drinking more. Matt will learn what he has to do. Right now, we monitor his eating habits and remind him to "chew, chew, chew and drink."

Today, we are now just getting on with life, with a boy who has real dreams and the energy to grow up and be the best he can be.

Matthew lives with his parents Robert and Diana Wells in Genoa City, Wisconsin. Their phone number is (262) 279-2092.

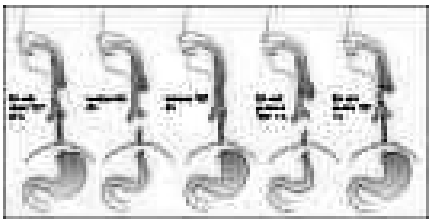
ea/tef: THE FACTS

Definition

Esophageal atresia (ea) is a birth defect (congenital anomaly) in which the esophagus, which connects the mouth to the stomach, is shortened and closed off (dead ended) at some point along its length. This defect almost always occurs in conjunction with tracheoesophageal fistula (tef), a condition in which the esophagus is improperly attached to the trachea, the "windpipe" that carries air into the lungs. It is believed that these defects occur around the fourth week of pregnancy when the digestive tract is forming. There is no known cause for the defects.

Description

Failure of an unborn child (fetus) to develop properly can result in birth defects. These defects typically involve organs whose function is either incidental or not necessary at all before birth, meaning that the defects will not be detected until the baby is born. The digestive tract is unnecessary for fetal growth, since all nutrition comes from the mother through the placenta and umbilical cord. During fetal development, the esophagus and trachea arise from the same original tissue, forming into two side-by-side passageways, the esophagus leading from the throat to the stomach and digestive tract, and the trachea leading from the larynx to the lungs and respiratory system. Normally, the two tubes form separately (differentiate); however, in the case of ea/tef, they do not differentiate, which results in various malformed configurations. There are five configurations:



• Type A (7.7% of cases): Esophageal atresia in which both segments of the esophagus end in blind pouches. Neither segment is attached to the trachea.

• Type B (0.8%): Esophageal atresia with tracheoesophageal fistula in which the upper segment of the esophagus forms a fistula to the trachea. The lower segment of the esophagus ends in a blind pouch. This condition is very rare.

• Type C (86.5%): Esophageal atresia with tracheoesophageal fistula, in which the upper segment of the esophagus ends in a blind pouch (EA) and the lower segment of the esophagus is attached to the trachea (TEF).

• Type D (0.7%): Esophageal atresia with tracheoesophageal fistula, in which both segments of the esophagus are attached to the trachea. This is the rarest form of EA/TEF.

• Type H (4.2%): Tracheoesophageal fistula in which there is no esophageal atresia because the esophagus is continuous to the stomach. Fistula is present between the esophagus and the trachea.

Normally, the esophagus moves food from the mouth to the stomach. When the esophagus ends in a pouch instead of emptying into the stomach, food, liquids, and saliva cannot pass through. The combination of ea with tef compromises digestion, nutrition, and respiration (breathing), creating a life-threatening condition that requires immediate medical attention. All babies with ea/tef require surgical repair to correct the condition and allow proper nutrition and swallowing.

Demographics

Esophageal atresia alone or with tracheoesophageal fistula (ea/tef) occurs in approximately one in 4,000 live births.

Causes and symptoms

The cause of esophageal atresia, like that of most birth defects, is unknown.

An infant born with ea/tef may initially appear to swallow normally. However, the first signs of ea/tef may be the presence of tiny, white, frothy bubbles of mucus in the infant's mouth and sometimes in the nose as well. When these bubbles are suctioned away, they reappear. This symptom occurs when the blind pouch begins to fill with mucus and saliva that would normally pass through the esophagus into the stomach. Instead these secretions back up into the mouth and nasal area, causing the baby to drool excessively. Although the infant may swallow normally, a rattling sound may be heard in the chest along with coughing and choking, especially when the infant tries to drink. Some infants, depending on the severity of the defect, may appear blue (cyanosis), a sign of insufficient oxygen in the circulatory system. The infant's abdomen may be swollen and firm (distended) because the abnormal trachea allows air to build up in the stomach, filling the abdominal space that holds the surrounding organs. Aspiration pneumonia, an infection of the respiratory system caused by inhalation of the contents of the digestive tract, may also develop.

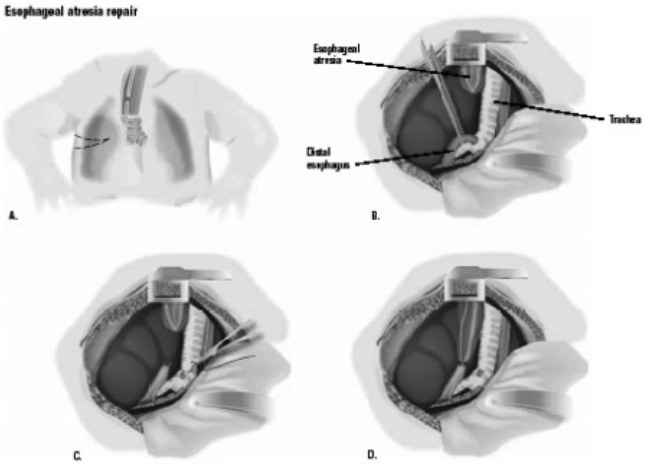
Diagnosis

When a physician suspects esophageal atresia after being presented with the typical symptoms, diagnosis usually begins with gently passing a catheter through the nose and into the esophagus. Esophageal atresia is indicated if the catheter stops at the blind pouch, indicating that it has hit an obstruction. If EA is present, the catheter will typically stop at 4 to 5 inches (10–12 cm) from the nostrils. Barium-enhanced x-ray examination may reveal a dilated esophageal pouch, made larger by the collection of amniotic fluid in the pouch.

During fetal development, the enlarged esophagus may also have pressed on and narrowed the trachea, a condition in the fetus that can contribute to fistula development. Air in the stomach may confirm the presence of fistula; gas in the large intestine rules out intestinal (duodenal) atresia. The physician will also perform a comprehensive physical examination, looking for other congenital anomalies that are known to accompany EA/TEF. Chest x rays may be taken to look for skeletal and cardiac abnormalities. Abdominal x rays may be taken as well to look for intestinal obstruction and abnormalities. An echocardiogram (ECG) may be performed to evaluate heart function and ultrasound of the kidneys performed to evaluate kidney function.

Treatment

Infants with ea, with or without tef, are unlikely to survive without surgery to reconnect the esophagus. The procedure is done as soon as possible; however, prematurity, the presence of other birth defects, or complications of aspiration pneumonia



To repair esophageal atresia, an opening is cut into the chest (A). The two parts of the existing esophagus are identified (B). The lower esophagus is detached from the trachea (C) and connected to the upper part of the esophagus (D).
Illustration by GGS Information Services

may delay surgery. Once diagnosed, the baby may be fed intravenously until surgery is performed. Mucus and saliva will also be continuously removed via a catheter. Healthy infants who have no complications, such as heart or lung problems or other types of intestinal malformations, can usually have surgery within the first 24 hours of life. Surgery techniques used to treat the five types of ea/tef defects are similar.

Surgery is conducted while the infant is under general anesthesia; a tube is placed through the mouth to continuously suction the esophageal pouch during the procedure. An intravenous line (IV tubing into the veins) is established to allow fluids to be administered as needed during surgery. Usually, the infant is placed on a ventilator, with a tube placed down the airway for at least the length of the surgery.

Typically, the surgeon makes an incision in the right chest wall between the ribs, allowing access to the esophagus and the trachea for repair of one or both as needed. If the gap between the two portions of the esophagus is short, the surgeon may join both ends of the esophagus (anastomosis). If the upper portion of the esophagus is short and a long gap exists between upper and lower portions, reconstructive surgery cannot be performed, and the infant must receive nutrition in some way to allow several months of growth. In this case, a gastrostomy (stomach tube) may be surgically placed directly into the stomach for feeding. In the most typical ea/tef repair, the fistula is first closed off, creating a separate airway. Then the blind esophageal pouch is opened and connected with suturing (stitching) to the other portion of the esophagus, creating a normal "food pipe" directly into the stomach. The esophagus is separated from the trachea if necessary.

If the two ends of the esophagus are too far apart to be reattached, surgury is put off to wait for growth of esophageal tissue, sometimes accompanied by stretching procedures. Surgery to connect the two ends of the esophagus can happen within three to four months of life. Or tissue from the large intestine is used to breech the gap between the two ends.

Nutritional concerns

If an infant is unable to nurse normally before surgery can be performed, nutrition is provided intravenously (parenteral) or directly through a tube into the stomach (gastronomy). After the surgery, infants should be able to swallow normally and resume nursing or feeding. However, a feeding tube (g-tube) may stay in place for months or years, depending on the child's nutritional needs.

Prognosis

Surgery to correct esophageal atresia is usually successful, with survival rates close to 100 percent in otherwise healthy infants after the condition is corrected. Postoperative complications may include difficulty swallowing, since the esophagus may not contract efficiently, strictures (scar tissue build up) at the surgery site, and gastrointestinal reflux, in which the acidic contents of stomach back up into the lower part of the esophagus, possibly causing ulcers.

Parental concerns

Despite a difficult beginning for infants with esophageal atresia with or without tef, parents can be reassured that the defect can usually be corrected with surgery, allowing normal digestion, nutrition, and breathing to take place in their child. Concerns about complications are well founded, including increased susceptibility to colds and infections, as well as the presence of chronic conditions. Ongoing medical care helps manage these conditions and maintain good health in children who have had ea/tef. Parents can seek advice about strengthening the child's immune system through appropriate nutrition and supplements.

(Continued from page 1)

By now it was about 7:30 in the morning. We proceeded back to the room to call our family. My heart broke as I listened to Craig weeping on the phone to his dad. When I called my mom, she tried to sound positive, telling me everything was going to be okay. I told her how long he was expected to be in the hospital and then she broke down crying. She knew nothing she could say would make this okay.

It felt as if an angel arrived to give me hope at a very disheartening time.

Craig was able to visit Colton later that morning and learned they planned to operate the next day. He handed me two pictures the nurse had taken for me. That day in the hospital without Colton was the loneliest day of my life. Most new mothers get to hold and feed their newborns. I had two pictures of him to remember what he looked like.

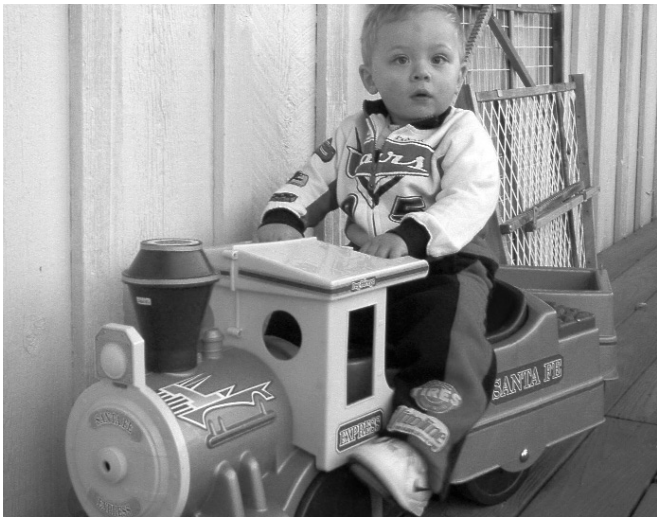
During a routine blood draw, a woman came in and asked where my baby was. I explained. Amazingly, she had been born with the same thing and was living a wonderful life. It felt as if an angel arrived to give me hope at a very disheartening time. When my doctor came in to see me, I told her that I absolutely would not miss my son's surgery. I checked out of the hospital 29 hours after giving birth. It was a very snowy drive to Children's Hospital, but nothing would keep me away.

In the neonatal intensive care unit, Craig and I were each able to hold Colton for a few minutes before they took him to surgery. Again, we huddled around his crib and prayed, then stepped aside. When they wheeled him away I felt such an overwhelming fear. I didn't even really know my baby yet and there was already a chance he could die. The only thing that gave me peace was to pray and know he was in good hands.

A few hours after Colton was taken away, a nurse told us the surgery was successful. When we went in to see him, he looked helpless. He was lying there with tubes, wires and monitors everywhere. He had I.V. lines in every limb, a breathing tube down his throat, a feeding tube down his nose, a catheter for his urine and a protective mask over his eyes for the jaundice lamp. He looked very large and swollen.

A few days after the surgery, his breathing tube was removed. We were walking down the long hallway on our way to see him and could hear this very loud gasping. As we grew closer we realized that horrific sound was coming from our baby! We walked in to see several doctors around Colton with panicked looks on their faces. I asked what was happening as I saw our baby lay there gasping. Nothing being done seemed to help. The image of my son's chest completely concave, breathless, will haunt me for the rest of my life. I started to panic and had to leave the room. After 45 minutes of complete agony, Colton was reintubated. The doctors explained that his esophagus was still very swollen from the surgery and in a few days they would try again.

One week had passed and Colton had an esophagram to check his repair. Thankfully everything was fine. It was time to try again to get Colton off the ventilator. This time I could not bear witness to his torment and asked the nurse to come and get us when it was over. Twenty minutes later, Colton's nurse came rushing out with tears in her eyes. Colton was breathing on his own and doing well. This time, he stayed off for 12 hours. He was working hard to breath and the doctors worried that he would tire out, so they reintubated him. They thought Colton could have diseased



lungs, a swollen esophagus, paralyzed vocal cords or a severed nerve from surgery. The ear/nose/throat doctor came in to scope his vocal cords and found no problem. The staff kept telling us he just needed more time.

At this point, Colton was pretty well healed from surgery and was on little or no medication. He was alert and the ventilator tube bothered him so he would tug at it, cry and then have an even worse gasping spell. Because the tube separates the vocal cords, he couldn't make sounds. The only way a nurse away from Colton's bed was aware he was crying would be for his monitor to go off because his heart rate would spike. This made it so difficult to go home at night because I kept imagining his frowning little face sobbing and no one there to comfort him.

A few days later he was extubated again. This time he was put on a cpap nose cannula, which delivers a constant strong flow of air into the airway to help keep it open. Another scope of his vocal cords was done and this time it showed that his vocal cords were paralyzed. Doctors didn't know if it was from a severed nerve or the ventilator tube being down his throat for so long. It's possible for a vocal cord to regain movement so they wanted to wait. We spent the next three weeks listening to Colton struggle for every breath. As he was healing and becoming more active, he would struggle to get the air he required. The nurses urged me to hold Colton, but it was a very big ordeal to get him situated and comfortable. Often, it would send him into a panic of not being able to catch his breath that could last for hours. Instead, I would usually just lay my head next to his in his crib so he could look at me or smell my presence.

Finally, the doctors suggested that maybe Colton needed

a tracheotomy. This news fell on us like a ton of bricks. Again I began telling myself that this was not supposed to happen. We were supposed to be working on feeding him by now. No one had said this might happen. We had spent so many days going downhill and now this, another surgery!

The nurse told us that having a trach would not be that bad. Colton would still be okay mentally and hit all of his normal milestones. He wouldn't be able to talk for a while so we would have to learn sign language and he would be sent home with lots of equipment, making him homebound. This was way too much for us to handle. My husband and I hit our breaking point. We found a place in the NICU that was private so we could fall into each other's arms and just cry.

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We thought about all of the things we would miss out on, like going to family gatherings together, going shopping or dining out. We wept at the notion of our son not being able to say "Mommy" or "Daddy," and we imagined him being made fun of in school for being different. We left that Saturday and went straight to our priest. We told him all that we were going through, that it felt as if everything was going from bad to worse. Why is this happening to us, we asked? He told us to keep having faith and to keep praying. He told us we could scream at God if we needed to, just to keep having faith.

Since Colton was preparing to have another surgery, we decided to have him baptized and to have the sacrament of the sick performed. A few days later, hours before Father Dick came, the ENT scoped Colton again. He found the same prognosis; Colton's vocal cords were paralyzed. Soon after his scope, we covered him with a beautiful white baptismal garment. The three of us surrounded Colton with our prayers. He was very content throughout the whole ceremony.

The same day, we scheduled a conference for the next Tuesday to sit down with everyone involved in Colton's care and make plans for his tracheotomy. Craig and I had still not come to terms with it, but we knew we needed to get Colton breathing on his own. We had no other choice. Days after the baptism, Colton got a little feisty and he pulled off his cpap nose cannula. The nurses did not realize it for close to an hour. In that time he had been breathing on his own with very little struggle. They decided to disconnect it and monitor how he did on his own. He was still stridorous, but not nearly as much as the days before.

Tuesday came and it was time to sit down with all of the doctors and nurses and discuss our plans. That morning he was scheduled for another scope so they would have validation for the next surgery. I received a phone call from Colton's nurse practitioner just as I was ready to walk out the door to the hospital. She began crying as she told me that something astonishing happened when they scoped Colton this morning: both of his vocal cords were working! She said that as soon as I come in this morning we could begin breastfeeding. I can't possibly express the excitement I felt as I called Craig at work to tell him. Finally things were improving.

At the hospital, Colton's nurse and my lactation consultant were ready to help me with his first feeding. They warned me that this is the hardest part because after babies lose their natural instinct to suck, swallow, and breath it is very difficult to teach them again. I understood that it may take months, but I didn't care. Colton was breathing. I could actually see all of his face for the first time since he was born. The curtains in his pod closed as I began to feed him. He latched on right away and began feeding. He had no problems. The nurse and the consultant were crying as they called in other nurses to come and see what was happening.

The conference to plan Colton's surgery ended up turning into a discharge meeting. They wanted to wait through the weekend to monitor if he would have any breathing difficulties. If everything went well, we could go home on Monday. The days went by and Colton would eat by breast or bottle without any tribulations. Many people would peek their head in to see if he was still there because they had become accustomed to hearing him all throughout the unit.

Monday came and we were ready to leave. That day was bittersweet because I had gotten to know everybody there. I especially felt bad for the babies who remained behind. The first days at home were very stressful for us, as any first-time parents would experience. Colton's reflux was pretty severe causing him to regurgitate through his nose. After two months we switched from breast milk to Enfamil A.R. and that helped a lot. He still remains on his reflux medication, but as he grows reflux is becoming less of an issue.

Colton is now almost two years old. He is very similar to children without his defect. He still does gag and vomit if food gets clogged in his esophagus, but this too is improving with age. He has been in the 75% to 90% range for height and weight consistently. He has also hit every milestone right on time.

This has made me so much stronger as a person and more patient as a mother.

What happened to Colton will affect me for my whole life. It has been almost two years and still there is not a day that passes that I don't tear up thinking about it. This has made me so much stronger as a person and more patient as a mother. The only thing that got me through this was faith and prayer. Colton's vocal cords were paralyzed hours before his baptism and immediately following he started to improve. Colton is a true miracle and his story has touched so many people's lives. This may not have been what I had planned, but I don't think I would change a thing.

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Illustrations With Pizzazz (and Moving Parts!)

We received an email from Catie Caponetto last year asking for feedback on a project she did while completing her master's degree in medical illustration at the University of Chicago at Illinois. She created an interactive illustration on ea/tef that can be accessed online by families, friends, anyone looking for information about the condition. She used a reference book from TOFS, the ea/tef support organization based in England. The illustrations are large and very easy to understand. In one, you can click on a bottle and the bottle will rise to the baby's mouth and you can watch the liquid as it is swallowed and goes through a normal esophagus into the stomach. In a section on diagnosis after birth, you can watch as a tube is threaded into the mouth and to the top of the esophagus, where it ends. This is a wonderful way to learn quickly just what is meant by ea/tef. It would be especially helpful for grandparents and others spending anxious hours at home during those early days. I'm not sure that Catie is still looking for feedback, but I highly recommend checking out the sight. http://www.caponettostudios.com/interactive/interactive3.html