When you meet 4-year-old Zoe Lambert, the first thing you notice is her left shoulder. It sits higher than the right one and makes it look as though she got halfway through a child's shrug of "I don't know" before giving up.

If you knew the health problems she's had in her short life, however, and realized that a metal rib in her chest allows her to bend at the waist and wave at you through her legs with an upside-down smile, her raised left shoulder would take on greater significance. Instead of an indifferent "I don't know," you might actually see the result of her severe birth defects as expressing a defiant, "What else have you got?"

Zoe's mother Arlene had a complication-free pregnancy, and none of her seven ultrasounds revealed anything abnormal. But when Zoe was delivered at a hospital in their small hometown of Plymouth, Pa., it took her grandmother, Johanna, only one look to realize that all the prenatal tests had missed something very significant. Johanna says Zoe looked pasty and had a webbed neck, and scans done in the hour after her birth revealed that, in Johanna's words, "the hospital had never seen anything like Zoe before." Less than 24 hours later, she was flown to a hospital in Hershey, Pa., where she was diagnosed with VACTERL syndrome, an extremely rare collection of birth defects. When combined, the worst of the defects was likely to cause Zoe's rib cage to collapse in on itself, a situation that would prevent her lungs from growing and would most likely end up suffocating her.

Alphabet soup

VACTERL is an acronym for vertebral, anal, cardiac, tracheal, esophageal, radial (lower arm bone) and/or renal (kidney) and limb—making Zoe's problems sound more like the index of a medical textbook than the problems of a single patient. She had atrial and ventricular septal defects (holes between the upper and lower chambers of her heart, which have since closed on their own), has only one kidney, and her intestines are twisted.

But the "V" for vertebral was the most troubling letter in the acronym for little Zoe. She was missing the three vertebrae containing the nerves that provide bowel and bladder control; two additional vertebrae in her neck were fused; she had severe scoliosis (curvature of the spine); three hemivertebrae (or missing pieces of spine); and several ribs that were fused, plus one rib that hadn't formed at all.

From the day she was born, Zoe needed extensive medical treatment. When she was 6 months old, some of her vertebrae were surgically fused in an effort to lock her spine in place and stop the progress of her scoliosis. By the time she turned 2, however, it was obvious that the spinal fusion had failed and that the progress of Zoe's curvature was impairing her lung capacity and causing heart problems. The situation was quickly becoming life threatening.

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A doctor in Hershey told Arlene that her daughter's only hope was an investigational procedure known as a titanium rib implant. “When I heard the word ‘investigational,’ I was upset, but I realized this was the only thing that would help Zoe,” says Arlene.

The Lamberts did some research on the Internet and found that the surgery had been developed by Robert Campbell, MD, an orthopaedic surgeon at Christus Santa Rosa Children’s Hospital in Texas. Instead of traveling to and from Texas, they decided to come to Children’s Hospital Boston, where John Emans, MD, chief of the Division of Spinal Surgery, and his colleagues had been hand-selected by Campbell as the second team in the world to perform the innovative surgery.

**Braces for the chest**

The idea behind the titanium rib procedure is deceptively simple: it performs the same basic function on a patient’s chest and ribs that braces perform on teeth. The patient’s fused ribs are first separated, and then a lightweight titanium rod is attached vertically to the ribs and/or spine to act as a stabilizer and keep the bones in place. The rod is lengthened regularly, expanding very much like a curtain rod, to keep the fused ribs apart, enlarge the chest, keep the spine more aligned.

The titanium ribs have begun to straighten Zoe’s spine and have allowed her lungs room to grow.
and allow the chest and lungs to continue to grow.

With the help of a multi-disciplinary team that includes specialists from General Surgery and Pulmonology, Emans has performed more than a dozen initial titanium rib operations and many subsequent lengthenings. These experiences, combined with those of Campbell's team, have shown that the surgery works well for major chest and rib abnormalities such as severe congenital rib fusions and scoliosis. And unlike spine fusions, a common procedure for people with scoliosis, the titanium rib procedure allows for more growth of the chest and spine. "For children who have small chests as a result of these problems, this can be a life-saving procedure," says Emans. "And instead of having the spine of a 2-year-old's whole life as a result of fusion surgery, the titanium rib allows them to grow more than they otherwise would."

What surgeons don't know at this point is how their patients will fare in the long run. "It will take a number of years — maybe as many as 15 — to see how these children do," says Emans. "Many of them were under the age of 2 at the time of the procedure, so we'll need to wait until they have stopped growing to fully evaluate the results."

Campbell, who has performed the operation on more than 130 children, feels that surgeons have only touched the surface of the titanium rib procedure's potential. "It works well for children with VACTERL, and is a superb way to replace ribs, especially in kids with cancer of the chest wall," he says. "But I think we're going to be surprised by how many uses this surgery will have in the future. For the first time, we're able to treat three-dimensional problems with three-dimensional thinking and techniques."

Just as Campbell advised Emans on his titanium rib procedures, he sees Emans performing the same function for surgeons at other qualified hospitals throughout the country. "Dr. Emans will serve as a mentor so other programs can begin performing this surgery," says Campbell. "He is a superb teacher and never ceases to amaze me with his ingenuity."

The pair are currently recruiting patients for an FDA-mandated clinical trial that is expected to come to an end later this spring, and Emans hopes that the surgery will be generally available at qualified centers around the country by the end of the year.

**Straightening the S**

Living within a few miles of a qualified center has been a blessing for Susan Wilson and her daughter, Samantha. Shortly after Samantha's birth, Timothy Hresko, MD, assistant in Orthopaedic Surgery at Children's, found that she was suffering symptoms of respiratory distress and quickly realized that her chest and lungs had stopped growing. He diagnosed her with congenital kyphosis, an abnormal, backward curvature of the spine. This was compounded by the fact that she also had short ribs, some of which were fused while others had large spaces between them.

In 2000, Emans performed titanium rib surgery on both the right and left sides of Samantha's chest. Since then, Susan has noticed a vast improvement in her daughter's breathing and is amazed how different Samantha looks. "Before the operation she was shaped like an 'S' and doctors said she would never sit straight or walk well," she says. "But now she stands straight and does everything a typical 3-year-old can do."

Because of the other physical problems related to her VACTERL, Zoe Lambert is not yet able to do all of the same things as other 4-year-olds. She will need intensive rehabilitation to learn how to control her bowels and bladder, and the her twisted intestines continue to cause problems that have led her to be rushed to many hospitals, including Children's, for emergency treatment. But Emans' work has taken what were the most dangerous of her problems—those of her spine and ribs — and moved the "V" for vertebral to the bottom of a long list of concerns.

Zoe now tests near the normal range on physical therapy exams and is able to climb on and off chairs—

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Dr. John Emans' team was the second in the world to perform the titanium rib procedure.
something she couldn’t do before surgery. It is the simple things, though, that have made the biggest difference in Zoe’s life and the lives of her mother and grandparents. Every year for more than a decade, the Lamberts have gone to the beach for summer vacation, and last summer, Zoe, who has always loved her trips to the ocean, was finally able to get in the water and play with all the other kids. Zoe’s mother put her in bathing suits and little bikinis, the outline of the titanium rod showing faintly through the skin of her small body. Asked to describe her feelings at seeing her daughter grow up and be able to enjoy things like playing at the beach, Arlene states it simply: “She’s here. And if it wasn’t for Dr. Emans and the surgery, she may not have been.”

—Matthew Cyr

T is for tutor

A tutor a day can’t keep the doctor away, but for patients at Children’s Hospital Boston, a visit from the teacher certainly makes their hospital stay seem a little more normal.

While their classmates look forward to summer vacation, hundreds of kids in hospital beds would love to return to the routine of school. At Children’s, the Child Life Services Department and a company called Education, Inc., provide that opportunity.

For most of her 37 years at the hospital, Myra Fox, director of Child Life Services, has been doing her part to make sure sick students get an education in the hospital. She has hired teachers, fought legal battles and challenged the city of Boston to pay for the education of hospitalized children. Yet she was always met with roadblocks, and finally decided that enough was enough.

“The city dictates how many hours teachers can spend with student patients, and the teachers often had problems getting paid,” says Fox. “I was getting complaints from the teachers who came here, from the nurses, social workers and Child Life specialists on the floors, and from the students themselves. I thought to myself, ‘There has to be someone doing hospital tutoring.’”

She contacted Ken Munies, a former Brockton, Mass., teacher who began tutoring on the side after the parent of a sick child asked him for help. “I started tutoring more and more often and it wasn’t long before I realized I enjoyed the tutoring more than the classroom,” he says. “So I left the school system and started Education, Inc.”

What began as a two-teacher operation in 1995 has grown into a company with nearly 300 full-time tutors who provide 1,200 hours of education per week to students throughout Massachusetts, New Hampshire and Maine. Of the tutors, three work full-time at Children’s so they can teach patients on short notice.

Munies says tutors will tailor the program to each student’s needs. “No matter how long a child is out of school, our goal is that he or she will return at the same level as their classmates.” In order to be eligible for tutoring, a student must be absent from school at least 14 days of the school year.

Helping students keep up with their classmates may seem impossible given the fact that schools only pay for seven to 10 hours of teaching time each week for students receiving outside tutoring, but Munies says teachers accomplish their goal by using the same or similar materials as their fellow students. “Instead of repetition, which is how most learning is accomplished, we work one-on-one with the child. We use existing materials and are able to cover subjects in greater depth than is possible in a classroom with 20 children. For example, we will have them study a Shakespearean sonnet rather than just memorize it so they understand the meaning and the nuance.”

The system seems to be working for everyone involved. Pam Clarkson, the tutoring supervisor at Children’s, says this is the best teaching job she has ever had. “I feel like I’m able to give individual attention and a quality education to each child.”

The hospital is also pleased. The tutors teach only subjects in which they are skilled, and all teachers who come to Children’s go through training and follow the hospital’s policies and procedures. In addition, Education, Inc. provides the supplies, hires and pays the tutors and charges the school district where the child is from for the cost of the patient’s education.

Susan Shaw, RN, MS, director of Clinical Operations, has seen the value of having a company dedicated strictly to the education of patients. “One of the things that differentiates us at Children’s is our philosophy of caring for the whole child. Education is a huge part of their lives,” she says. “This program is a tangible example of how we treat them as children first and don’t define them by their illness.”