

TESTING THEIR METAL

Titanium rib procedure gives patients room to breathe



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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BRACES FOR THE CHEST



The X-ray at near left shows Zoe Lambert's ribs and spine before titanium rib surgery. At far left is Zoe's chest with the titanium implants.

Images: Courtesy of John Emans, MD



Photo: Mark Ostow

The titanium ribs have begun to straighten Zoe's spine and have allowed her lungs room to grow.

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 Camp-

bell 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

