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HIGHLIGHTS:

Tethered cord is a congenital malformation of the spinal cord affecting infants and children. Left untreated, it can result in pain, incontinence and even paraplegia. Although some cases are fairly easy to diagnose, many remain undetected until symptoms progress to a serious stage. Ezra Kahn, age 4-1/2 years, had undergone more than 10 operations before he was brought to Cedars-Sinai where he was diagnosed and treated by a team of multi-disciplinary pediatric surgical specialists. Now approaching 5 years of age, the active youngster takes swimming lessons, plays basketball and is living life to the fullest.

PEDIATRIC NEUROSURGERY TO RELEASE HIS TETHERED SPINAL CORD HELPS YOUNGSTER LOOK FORWARD TO A NORMAL, ACTIVE LIFE

LOS ANGELES (Jan. 2, 2003) – Although pregnancy and delivery had progressed normally and no problems had been expected, Ezra Kahn developed trouble breathing and taking in fluids just after his birth nearly five years ago.

Doctors discovered that Ezra's esophagus was too narrow to allow fluids to pass through and he had an anorectal malformation called an imperforate anus: There was no opening at the end of the digestive tract. Surgeons immediately inserted a feeding tube and performed a colostomy – the first of many surgeries Ezra would undergo in the first few years of his life.

Ezra was found to have several birth defects that often are seen together. The association of these defects is sometimes referred to as VATER syndrome, an acronym that includes vertebral defects, anal malformations, tracheo-esophageal fistula, esophageal atresia, renal defects and radial bone dysplasia. Other body systems may also be affected, resulting in variations of the acronym.

Ezra's mother became an active member of the VATER Association's "pull-thru network," an organization that provides information and support through a newsletter and Web site. Several years ago, another mother asked if Ezra had been checked for a condition called a tethered spinal cord. In normal childhood development, the lower part of the spinal cord grows at a different rate than the vertebral column and remains free of the column. However, if the cord is attached (or tethered), as it was in Ezra's case, it may be stretched and damaged, causing neurological injury.

"I had heard of tethered cord but it was my understanding that most kids were symptomatic – you'd notice something wrong with their gait or their spine or they would have discomfort. My son never had any of these symptoms. He was extremely athletic, always bouncing around," said Annabelle Gurwich, Ezra's mother.

To the family's shock, an MRI showed that Ezra did have a tethered cord. They were referred to Moise Danielpour, M.D., director of pediatric neurosurgery at Cedars-Sinai's Maxine Dunitz Neurosurgical Institute.

"This can be caused by any of several developmental malformations causing the last part of the spinal cord to be abnormal. The spinal cord becomes tethered, pulled down tightly to the bottom of the spinal canal," said Dr. Danielpour, adding that the condition can cause a variety of symptoms that may change or grow more debilitating over time.

Leg and back pain – more pronounced than that associated with childhood "growing pains" – is a common consequence, as are leg weakness and bowel and bladder incontinence. Abnormalities on a child's lower back – a fatty lump, dimple or hairy patch – are common signs that sometimes lead to a diagnosis.

"Some kids don't have these signs on their back but they have incontinence that continues or gets worse as they grow older," according to Dr. Danielpour. "Because there are many potential underlying causes of incontinence, special tests can be done to pinpoint the problem. If neurological injury has resulted in a hyperactive, or so called irritable bladder, we look for a tethered spinal cord.

Andrew L. Freedman, M.D., director of pediatric urology at Cedars-Sinai's Endourology Institute, specializes in diagnosing causes of incontinence.

"We recently treated a 9-year-old who was having four or five episodes of incontinence a day," said Dr. Danielpour. "He had been dismissed from several schools because they thought his incontinence was a behavior issue – they thought he could stop if he really wanted to. Dr. Freedman saw him and did the bladder studies, which suggested that the cause was a neurogenic bladder. We performed surgery to release the spinal cord and now he has improved dramatically. He says he can feel it when he has to go to the bathroom now."

"If a child doesn't grow out of bed-wetting and incontinence, if the problem is associated with back and leg pain or toe-walking, if there are indications that the nervous system is involved, or if there are cutaneous signs on the back, there's a good chance the child has a tethered cord," Dr. Danielpour said. "It can be very difficult to diagnose unless the child is seen by a physician who is trained to recognize the signs and symptoms."

The existence of scoliosis or other spinal and orthopedic malformations increases risk of having a tethered cord. The challenge for parents and physicians is in detecting "occult malformations" – those that do not have visible signs or symptoms. "If we have reason to suspect a tethered cord, we can order an MRI. We've become more and more aware of the incidence of tethered cords since the advent of MRIs," according to Dr. Danielpour.

TV personality Annabelle Gurwitz and her husband, comedy writer Jeff Kahn, thought the idea of having Ezra tested was absurd. "My husband and I debated it. We were actually scheduled for an MRI for my son's kidney so we asked if while they were doing that MRI they would check for tethered cord also," recalled Annabelle. "None of us thought he had it. He didn't seem to have any of the real symptoms. He has a perfectly formed tush. He doesn't have the classic signs of it. We're just so grateful that we had the test and that we had the surgery so that there was no time for the condition to degenerate."

Left untreated, a tethered cord can lead to serious problems, including paraplegia, according to Dr. Danielpour, who said surgery to correct a tethered cord can range from one to eight hours, depending on the child's circumstances. Ezra's operation took five hours. It was one of maybe 10 or 20 surgeries – his mom lost count

– that Ezra underwent during the first 3 ½ years of his life to treat the various problems associated with his syndrome.

Now approaching his fifth birthday, Ezra plays basketball and baseball. He takes swimming lessons and goes to school every day. “He’s just like your average kid with a bunch of really interesting-looking scars,” Annabelle said. “Each kid is different with these anomalies. There are more extreme cases that require more reconstruction. For some kids, the prognosis is better than others. I think my son is pretty lucky.”

Cedars-Sinai is one of the largest nonprofit academic medical centers in the Western United States. For the fifth straight two-year period, Cedars-Sinai has been named Southern California's gold standard in health care in an independent survey. It is internationally renowned for its diagnostic and treatment capabilities and its broad spectrum of programs and services, as well as breakthrough biomedical research and superlative medical education. Named one of the 100 "Most Wired" hospitals in health care in 2001, the Medical Center ranks among the top 10 non-university hospitals in the nation for its research activities.

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