Novel surgery at Packard Children's repairs boy's airway, voice box

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Noah Jackson no longer needs to rely on a special tube to breathe because of the innovative treatment he received at Packard Children's. Credit: Courtesy of the Jackson family

Noah Jackson was born without a voice. Because of a rare genetic disease, his airway was so narrow he couldn't cry at birth. In fact, he could scarcely breathe, and had surgery when he was 5 days old to implant a tracheostomy tube that let air pass through a hole in his throat. Cuddling their newborn, parents KC and Rebecca knew Noah's only hope for someday speaking and breathing normally lay in the possibility

that his voice box could be surgically reconstructed later on.

In the summer of 2010, when Noah was 18 months old, his surgeon at home in Fresno, Calif., referred him to Packard Children's world-class otolaryngology team, including Peter Koltai, MD, who is experienced at reconstruction of the voice box. But even Koltai's considerable surgical expertise provided no guarantees of success.

"This was a complete obstruction, as bad as it gets," said Koltai, remembering his early assessments of Noah's airway. "There was no opening at all to his voice box." After the "trach" tube was placed, Noah's tiny airway had scarred shut. When Koltai first evaluated Noah, no air came down from his nose or mouth to his lungs.

Noah also faced other difficulties. His rare genetic disease, Fraser Syndrome, causes structural anomalies in many parts of the body. Noah has only one eye, and was born with hand, foot and digestive-tract problems that required surgery in infancy. But the trach was Noah's biggest medical challenge. The tube dislodged about once a month, leaving Noah breathless and sparking anxiety for his family.

"Every time he played, we were constantly watching the trach," Rebecca said. If the tube came out when a caregiver's back was turned, Noah could not cry for help; he communicated only in sign language.

Koltai's attempt to free Noah from the trach and give him a voice was a multi-stage undertaking with an estimated 70 percent chance of success. By November 2010, Noah was ready for the reconstruction surgery. Koltai and his team opened Noah's voice box and removed the scar tissue that blocked Noah's airway.

"The scar came up to the bottom of the vocal cords, but we were able to dissect them free," Koltai said. The team then used two pieces of rib

cartilage from Noah's chest to enlarge the framework of the voice box. The new airway was supported with a stent inserted through the center; Noah would keep breathing through his trach until the airway was fully functional.

A month after the stent was removed, Noah had a check-up.

"The reconstruction had worked well below the level of the vocal cords," Koltai said, but problems remained. "Because the vocal cords had been involved in scarring, they had fused back down like a zipper, almost totally closed," he added, explaining that this not only prevented Noah from speaking, but also jeopardized his ability to breathe normally.

Fortunately, there was still a small opening. Over the next four months, the team repeatedly inserted a high pressure airway balloon of Koltai's own design that gradually re-opened the airway and allowed the vocal cords to heal in a normal configuration.

Gradually, Noah learned to breathe through his nose and mouth. "Having a trach, the air just kind of dumps in and oozes out," Rebecca said. Noah's speech and sign-language therapists helped him build lung power with toys such as pinwheels to blow. It was hard work.

In June 2011, Koltai re-examined Noah's airway. "I'll never forget it," Rebecca said. "Dr. Koltai came running out and said 'Do you want the good news or the good news? I'm going to take the trach out right now!'"

A few minutes later, with Noah seated on Rebecca's lap, Koltai undid the Velcro straps that held the trach in place. The little boy breathed: in and out, in and out. Noah seemed bewildered by the excitement of the adults around him.

The fact that he leads a renowned surgical team and has reconstructed

many children's airways didn't diminish Koltai's enjoyment of the moment. "It gives you goose bumps every time it works," he said.

Noah's life is now much like that of any 3-year-old. He can roughhouse with friends without risk of dislodging a trach tube. He can play in his room while his parents keep an ear out from around the corner. He attends preschool, and is hitting cognitive and developmental milestones on schedule. And he is talking. "His voice will probably always be on the quiet and raspy side, but he's understandable," Rebecca said. "That's huge."

For Rebecca and her husband, KC, it's a dramatic change from their son's early days when they focused on his medical care. Now they can think and dream about what lies ahead for him. If your infant has complex medical needs, said Rebecca, "it's scary to think about the future, so you just don't. You don't think, 'What's going to happen to my child when he's 16?'"

Noah, it turns out, has plenty to say about his future. He loves visiting the Monterey Bay Aquarium and recently told his parents that when he grows up he wants to work with fish.

It'll be a little while before Noah is ready for his career in marine biology. But in the meantime, he's happy to chat about his favorite species of sharks — or whales, or turtles, or jellyfish, or seahorses.

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