

Straightening *small necks*

**New Approaches to Treating Congenital Torticollis
Offer Excellent Results With Minimal Risk of Complications**

When Steven and Debbie Klein's twin girls, Jessica and Kaitlyn, were born in January 2006, their pediatrician warned them that Jessica might have a problem worth watching. Crowded with her sister in the womb, Jessica's head had been placed at an awkward angle, causing trauma to the neck muscles. While Jessica looked fine to her parents at first, as she grew, it became noticeable that her head was drawn to one side. So when Jessica was 4 months old, her parents decided it was time to seek help.

■ Their search took them through four months of physical therapy and, when that was not as successful as they had hoped, they consulted with a neurosurgeon who then referred Jessica to craniofacial surgeon Fernando Burstein, M.D., at the Children's Healthcare of Atlanta Center for Craniofacial Disorders. ■ After a review of her medical records and an evaluation, Dr. Burstein confirmed Jessica was an appropriate candidate for a minimally invasive surgical procedure he had developed and that has helped many children like Jessica.

The Kleins agreed to the procedure. By that time, Jessica's problem was quite obvious. "At the back of her neck, the muscles and tendons were very pronounced because she was constantly turning her head to one side," said Dr. Burstein. If not corrected, the problem could have led to irreparable changes in her appearance as one side of her face developed larger than the other. "A facial deformity was a major concern for us," Debbie said.

A common problem with serious consequences

Jessica, who is now a healthy toddler with little evidence of the problem that once concerned her physician and parents, is one of the estimated 45,000 babies born in the U.S. each year with congenital torticollis, a condition in which the neck bends or twists to one side.

The problem occurs when trauma to the sternocleidomastoid muscle in the womb or during birth leads to scarring. Scarring causes the muscle to tighten and shorten, which in turn pulls one side of the head toward the shoulder as the chin tilts to the opposite side.

In about 20 percent of cases, torticollis is accompanied by plagiocephaly. Neither problem may be evident at birth, said Dr. Burstein. "The trauma to the neck muscle may not become fibrosed to the point where it affects the way the neck moves until the child is a few months old," he said. In some children, the presence of a pseudotumor (a lump in the neck caused by bleeding into the neck muscle) at birth can lead to scarring which results in torticollis later.

Regardless of when torticollis becomes apparent, prompt treatment is important. If not resolved, torticollis can cause severe limitations of neck position and, over time, the effect of gravity can lead to facial asymmetry and deformity.

As growth progresses, the jaw and bite may be affected requiring complex orthodontic and orthognatic surgical correction. In rare cases, combined craniofacial and neurosurgical intervention may be necessary to correct head and orbital deformities.

For the majority of children, a program of physical therapy to gradually restore muscle length—and if plagiocephaly is present, the use of a molding helmet to position and straighten the head—resolves the problem over the course

of several months. When therapy is difficult or does not produce the expected results, some physicians, including Michael Johnston, M.D., Pediatrician in the Children's Comprehensive Inpatient Rehabilitation Unit and Clinical Assistant Professor of Rehabilitation Medicine at Emory University School of Medicine use injections of BOTOX® (botulinum toxin) into the tightened muscles to facilitate the process.

"We are finding that BOTOX injections can be very helpful for children who have gone through some therapy and have not gotten completely better in the time that a lot of patients do," said Dr. Johnston, who has been using the injections for torticollis patients for about six months. "At that point, we say, 'Let's try something short of surgery; let's try to

relax some of these muscles with BOTOX and then be more aggressive with their therapy, and see if we can get some better results.'"

In fact, in a retrospective case series of 27 children published in the *American Journal of Physical Medicine & Rehabilitation*, 20 of the 27 had improved cervical rotation or head tilt following BOTOX injections. All of the children had congenital muscular torticollis that had failed to respond to a traditional program of physical therapy.

A safe surgical procedure

When conservative therapies fail to bring full range of motion to the neck, or if the child is having

significant facial asymmetry, surgical release of the sternocleidomastoid is recommended.

Not long ago, release of the sternocleidomastoid required open surgery.

The surgery not only left a visible scar, but also carried the risk of recurrent muscle band formation and the loss of neck contour, possibly due to incomplete initial release and denervation of the muscle, said Dr. Burstein.

The newer surgery developed by Dr. Burstein is a less invasive endoscopic procedure performed through a small incision behind the child's ear at the hairline.

Because of the size and location of the incision, the procedure leaves only a barely visible scar, said Dr. Burstein. The endoscopic approach also avoids many other problems of



Jessica pre-treatment 2006



Jessica post-treatment 2007

"All of our patients have achieved satisfactory neck range of motion."

Dr. Burstein

open surgery by allowing the surgeon to view the operative field directly, with magnification, ensuring precise muscle fiber transection and dissection of the intramuscular branches of the spinal accessory nerve, which conveys motor impulses to the pharynx and muscles of the upper thorax, back and shoulders.

For the Klein family, other benefits of the endoscopic approach include short surgery time—Jessica was in and out of the operating room in less than an hour—minimal discomfort and short recovery time. "Jessica wore a soft cervical collar for about a week afterwards and continued physical therapy for about seven weeks after that," Debbie said. "She really had minor discomfort. She really was not upset about it, even with wearing her collar. She did not look like she was in any kind of pain, except maybe the second day. The first day she was still on anesthesia, but the second day she was uncomfortable. After the third day, we were able to stop acetaminophen."

Excellent outcomes with endoscopy

Six months after her surgery, Jessica has had no complications. Neither have the vast majority of Dr. Burstein's other patients. In a 2003 review of 85 cases published in the journal *Plastic and Reconstructive Surgery*, only one patient experienced a complication of endoscopic treatment for torticollis. A 14-year-old girl required a small skin incision to ligate a branch of the external jugular vein.

"All of our patients have achieved satisfactory neck range of motion," said Dr. Burstein, who treats patients from across the U.S. and from as far away as Japan. "There have been no injuries to the spinal accessory nerve. None of the patients who have been treated with our protocol have required surgery of the jaws or craniofacial skeleton. All have achieved good facial and cranial symmetry with no gross occlusal abnormalities."

Aside from the one adolescent, all of the patients who have undergone the endoscopic procedure have been under 2½ years old, with the average age being around 12 months. "The surgery only works in children under 2," said Dr. Burstein. "When you get older there is less growth potential for your face. When there is less growth potential, you can release the muscle and the neck will straighten out, but the face may not necessarily straighten out. That's why it is critical to be diagnosed and treated early."

Although Dr. Burstein has been performing the procedure for more than a decade, just recently he has begun the new practice of referring patients presurgically to the pediatric physiatrist for BOTOX injections into the other muscles of the neck. This relaxes the additional muscles stressed by the torticollis as the patient recovers. "That helps them recover better," said Dr. Johnston. "It helps pain. It helps them to stretch more easily. The big thing if their neck is tight is trying to get it to just stretch out better and get back to normal range of motion."

For little Jessica Klein, limited range of motion is no longer a problem. "She is able to turn her head both directions easily whenever she wants. She looks easily both ways and uses both hands. The back of her neck is completely evened out at this point. She is a well-balanced kid," said Debbie. Although Jessica's head still has a little tilt and may well have a slight tilt for the rest of her life, her parents are not worried.

Looking at pictures of her little girl from before the surgery, Debbie said "Wow, look at the difference. That, more than anything else, tells me we did the right thing." ©

Signs of Congenital Torticollis

- Tilting of the head to one side, which may be evident at birth or become evident within a few months after birth. In 75 percent of cases, the right side is involved.
- Turning of chin toward the side opposite the tilt
- Limited range of motion in the neck
- A firm, 1-to-2 centimeter mass (pseudotumor) in the sternocleidomastoid muscle

Treatment for Congenital Torticollis

- Physical therapy to help lengthen the shortened sternocleidomastoid muscle
- BOTOX injections to relax the muscle and facilitate therapy
- Surgical release of the affected sternocleidomastoid muscle

A Related Problem: Checking Infants for Developmental Hip Dysplasia

As many as one in five children with muscular torticollis (compared to just three to four per 1,000 children in the general population) also have developmental hip dysplasia, a condition of abnormal development of the hip, resulting in instability and potential dislocation.

Early detection of dysplasia is important to limit malformation of the joint, which can ultimately lead to problems including leg-length discrepancies, gait irregularities and osteoarthritis.

It is recommended that children with congenital torticollis have an ultrasound of the hip in the first four to six weeks of life to rule out dysplasia.

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