Texas Children’s Hospital surgical team collaborates to separate conjoined twin girls

On February 17, a multidisciplinary team of 12 surgeons from seven specialties, six anesthesiologists, eight nurses and support staff performed one of the first successful separation surgeries for thoraco-omphalo-ischiopagus twins who share a chest wall, lungs, pericardial sac (the lining of the heart), diaphragm, liver, intestines, colon and pelvis.

The surgery lasted about 26 hours, with the actual separation occurring 18 hours in. The team, led by Dr. Darrell Cass, spent nearly 10 months planning and preparing for the surgery, which involved multiple disciplines including pediatric surgery, plastic surgery, cardiovascular surgery, urology, orthopedic surgery, pediatric gynecology and liver transplant surgery.

**Before Birth**

The Mata family was referred to Texas Children’s Hospital by their hometown maternal fetal medicine specialist in Lubbock when Elyssè Mata was 23 weeks pregnant. They met with Cass – pediatric surgeon, co-director of Texas Children’s Fetal Center™ and associate professor of surgery, pediatrics and obstetrics and gynecology at Baylor College of Medicine – who reviewed the results of a comprehensive evaluation performed by the Fetal Center which included a fetal echocardiogram, ultrasound and fetal MRI scan. The tests showed the chest walls, abdomens and pelvises were connected. Although around 75 percent of babies diagnosed as conjoined will die before birth or directly after, Cass believed a surgical separation could be successful.

Cass and Dr. Michael A. Belfort, obstetrician and gynecologist-in-chief of Texas Children’s Pavilion for Women, counseled the family about what to expect with the pregnancy and delivery. With a

A message from the surgeon-in-chief

Dear colleagues,

The Department of Surgery at Texas Children’s Hospital was recently involved in a groundbreaking surgery. The Mata family entrusted us with the surgical separation of their daughters who were born joined from chest to pelvis.

A soon as the Mata’s found out their diagnosis, they worked with our surgeons and maternal-fetal specialists to develop a plan that would give the girls the best lives possible. After a marathon surgery, Knatalye Hope and Adeline Faith experienced a remarkable recovery and are now home.

In this issue of Inside Surgery, you will read details about the planning and preparation that took place to make this separation surgery a success.

We also continue to break ground in pediatric robotic surgery. Texas Children’s Hospital has the highest volume of pediatric robotic surgeries in the U.S. Here we present a case of how the technology was used in complex bladder reconstruction.

As we continually seek to improve our diagnostic capabilities, the Division of Ophthalmology recently added Optical Coherence Tomography equipment to its clinic and surgical areas. This greatly enhances our ability to diagnose retinal and optic nerve issues earlier and more accurately.

Thank you once again for your continued interest in the Department of Surgery at Texas Children’s Hospital.

Respectfully,

Charles D. Fraser, Jr., M.D.
Surgeon-in-Chief
short cervix, the mother was at risk for early delivery and was prescribed hospital bedrest at 28 weeks. Due to the bulky anatomy of the twins, a larger than average C-section was planned.

**Post-Birth Planning with Neonatologists**
The twin girls, Knatalye Hope and Adeline Faith, were born at 31 weeks with a combined weight of 7 pounds, 9 ounces. The girls developed a few minor complications, including hydronephrosis and breathing insufficiency, as the result of premature lungs. Their lungs struggled with the weight of their combined bodies. Their diaphragm muscles were uncoordinated because they were connected, yet had two brains controlling them. Their livers were gelatinous as a result of prematurity and wouldn’t respond well to surgery, being less likely to seal once cut.

Texas Children’s surgical team decided to delay the separation surgery, to allow the twins’ lungs and other organs time to develop and be in the best condition possible to undergo a complex operation. This would minimize their time on the artificial ventilator post-surgery, reducing secondary injury and scarring. The twins were cared for by a team of specialists in Texas Children’s Level IV neonatal intensive care unit, where they thrived.

**Pre-Separation Preparation**
The multidisciplinary team that was assembled for the separation surgery represented different specialties and areas of expertise. Cass submitted a separation surgery proposal for review, which was revised several times with input from the team. In addition, the team reviewed imaging, mapped out their approach to the surgery and discussed various procedures.

Operating rooms are typically set up to service one patient at a time, so preparations were made to include ample equipment for two, including a computer line and additional gas lines. During surgery, each patient would be monitored individually, and each of the twins would be color coded – one red, one green – to ensure that each patient received the appropriate anesthesia and medications for surgery.

At eight months, the twins underwent a five-hour surgery led by Dr. Larry Hollier, chief of Plastic Surgery, and Dr. Ed Buchanan to place tissue expanders into their chest and abdomen area. This surgery required a recovery time of eight weeks, during which additional fluid was added to the balloon-like expanders, allowing the skin to be stretched gradually.

This surgery acted as a test run for the separation and provided an opportunity to inspect the groin area and perineum to identify their gynecological anatomy. To assist in post-operative healing and to alleviate pressure on the incisions, a swing was custom-made for the twins, allowing them to sit upright and produce the movement necessary for development.

Meanwhile, Texas Children’s Chief of Radiology Research and Cardiac Imaging, Dr. Rajesh Krishnamurthy, designed a 3D model of the twins’ anatomy. The detailed model, which included a detachable, transparent liver, aided in planning the pelvic portion of the separation surgery.

“We had to make decisions about which bladder goes to which girl, as well as which uterus and ovaries,” Cass said. “That might have been the most complex aspect of the operation.”

Around 10 days prior to separation, the entire multidisciplinary team went through two walk-throughs of the surgery, led by Texas Children’s Simulation Center. They used two mannequins that matched the twins’ weight exactly – around 40 pounds – to simulate the separation. The simulation allowed the team to determine how to best position the twins on the operating table to permit placement of central venous access and how to conduct a cardiac resuscitation should the twins suffer a cardiorespiratory arrest. As a result of the simulation, around 10 issues were identified and revised in the planning stage, which saved time and optimized safety during surgery itself.

**Surgery/Post-Surgery**
At 10 months old, the twins finally underwent a separation surgery. It took nearly 18 hours to separate them, then five more hours of work on Knatalye and eight more on Adeline. The surgeons worked from the side first, and then from top down, using maneuvers partly rehearsed in simulation exercises.

“I found the simulation exercises to be invaluable,” said Cass.

Immediately following surgery, each twin showed signs of significant gastroesophageal reflux, vomiting and oral aversion (a condition common to children hospitalized immediately after birth). Both twins required additional operations to remove external fixator devices and uteral stents, to place feeding tubes and to minimize reflux.

Knatalye was discharged just two months after separation. Adeline, who had more baseline lung problems and needed a tracheostomy, was discharged in June.

“Overall, their recovery has been remarkable,” said Dr. Charles D. Fraser, Jr., Texas Children’s Hospital surgeon-in-chief. “Texas Children’s is one of only a handful of places that can provide the highly-specialized, multidisciplinary care coordination needed to ensure the best possible outcome for a case such as this.”

![A multidisciplinary team participated in planning discussions.](image)

![A 3D model of the conjoined twins’ anatomy, which included a detachable transparent liver, aided in planning the pelvic portion of the surgery.](image)
Urology offers robot-assisted appendicovesicostomy

In January, Dr. Patricio Gargollo, director of the Complex Urogenital Reconstruction Program, completed a four-hour pediatric robot-assisted bladder neck reconstruction and appendicovesicostomy on a 15-year old female who struggled with incontinence as a result of spina bifida. She regained full continence as a result.

Typically, appendicovesicostomies are used to treat children ages 7 and up who struggle with neurogenic bladder and sphincteric incompetence, often due to nerve injuries such as spinal cord trauma, multiple sclerosis, brain tumors and spina bifida. Several urologists at Texas Children’s perform open appendicovesicostomies, but Gargollo is the only one at Texas Children’s – and one of only a few physicians in the U.S. – to use a robot for the procedure.

For complex reconstructive surgeries involving the bladder neck, Gargollo prefers using the da Vinci Surgical System robot to doing the surgery traditionally for several reasons:

1. Small spaces are easier to reach and visibility is magnified.
2. Scarring is reduced, exchanging a large midline incision for three to four incisions less than 10 mm each.
3. Duration of hospital stay is shortened by one to two weeks.

“You really see a difference in recovery using the robot for this procedure,” Gargollo said. “For me, that’s the most impressive part.”

Texas Children’s Hospital is nationally ranked in all ten subspecialties in the U.S. News & World Report’s list of America’s Best Children’s Hospitals and was one of only 12 hospitals to make the 2015 Honor Roll.

Three surgical sub-specialties ranked in the top 5 by U.S. News & World Report:

#2 Congenital Heart Surgery
#2 Neurology and Neurosurgery
#3 Urology

OCT technology improves diagnostic capabilities in Ophthalmology

In children, early signs of vision loss or problems due to retina and/or optic nerve dysfunction can be difficult to detect and diagnose. Optical Coherence Tomography (OCT) allows pediatric ophthalmologists to evaluate for retinal diseases, uveitis and optic nerve disorders. This technology has been used for more than a decade in adults but is relatively new in pediatric ophthalmology.

“The OCT is similar to ultrasound but uses light waves instead of sound waves to create high resolution images of the retina and optic nerve,” said pediatric neuro-ophtalmologist Dr. Veeral Shah. “It allows us to detect changes in retina and optic nerve structure that may reflect loss and damage of visual function that might not be visible with an ocular examination.”

Texas Children’s purchased two OCT machines. One is used daily in the Ophthalmology Clinic. The other OCT machine has a hand-held design that can be used on sedated children or in the operating room to inform intraoperative decisions.

“It is a great ancillary modality to make clinical decisions,” Shah said. “It’s especially helpful when evaluating young children who are preverbal, nonverbal or unable to recognize vision loss.”
**Announcements**

**NEW FACULTY SURGEONS**
John P. Dormans, M.D. – Orthopedics, Chief
Lauren C. Kane, M.D. – Congenital Heart Surgery
Sundee G. Keswani, M.D. – Pediatric Surgery
Shraddha S. Mukerji, M.D. – Otolaryngology
Duong D. Tu, M.D. – Urology

**SURGICAL RESEARCH DAY**
Almost 300 clinicians in the Department of Surgery attended the sixth annual Edmond T. Gonzales, Jr. Surgical Research Day at Texas Children’s Hospital. The event provided a forum for researchers to highlight their advancements in pediatric surgery.

“I really look forward to this day every year where we get to highlight some of the important research being conducted in the department,” said Dr. Charles D. Fraser, Jr., surgeon-in-chief at Texas Children’s Hospital. “Creating new knowledge is central to our core values as academic surgeons at Texas Children’s Hospital.”

The program featured seven oral presentations and 89 poster presentations spotlighting the academic efforts of the department. In addition, lectures were given on surgical issues and awards were presented to honor faculty and research presentations.

**PEDIATRIC PROVIDER CONFERENCE**
The Cutting Edge of Pediatrics conference sponsored by the Texas Children’s Department of Surgery was held Saturday, Jan. 24, providing 138 Houston-area pediatricians, family practice physicians and advanced practice providers an opportunity to learn about common pediatric surgical conditions from surgeons at Texas Children’s.

This was the department’s first educational conference for local pediatric providers.

The Department of Surgery would like to thank the speakers and the planning committee for their involvement in The Cutting Edge of Pediatrics conference. Speakers included Dr. Dean Andropoulos, Dr. Jennifer Bercaw-Pratt, Dr. David Coats, Dr. Nicolette Janzen, Dr. Edward Lee, Dr. Mark Mazziotti, Dr. Julina Ongkasuwan, Dr. Scott Rosenfeld and Dr. Veeral Shah. The planning committee included Dr. Anita Jimenez-Belinoski and Dr. Stanley Spinner from Texas Children’s Pediatrics, Dr. Carol Green from Kelsey-Seybold Pediatrics, Dr. Ellis Arjmand, Ryan Krasnosky, MPAS, PA-C, Dr. Megan May and Dr. Allen Milewicz from Texas Children’s Hospital.