

Case Study:

Pygopagus Conjoined Twins

Le Bonheur Separates Conjoined Twins

urgeons at Le Bonheur Children's Hospital successfully separated conjoined twins, Joshua and Jacob Spates, on Monday, Aug. 29. The twins were joined at the lumbosacral spine and shared a rectum, muscle and nerves. The 13-hour surgery featured a 35-member team and four surgical specialties — general surgery, neurosurgery, orthopaedic surgery and plastic surgery.



Joined at the lumbosacral spine, conjoined twins Joshua and Jacob Spates begin their first week of life in Le Bonheur Children's Neonatal Intensive Care Unit. The boys spent seven months in the NICU before being separated on Aug. 29.

The first of the twins, Joshua, went home on Sept. 20, 23 days after separation from his brother, Jacob. Jacob remains at Le Bonheur awaiting repair of a double outlet right ventricular heart defect.

Born via Caesarian section on Jan. 24, 2011, the babies were pygopagus twins, joined back to back at the pelvis and lower spine. Each had separate hearts, heads and limbs.

Pygopagus represents 15 percent of conjoined twins.

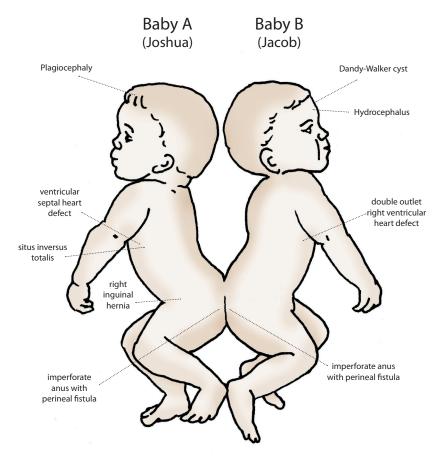
Diagnosis and Prenatal Care, Le Bonheur Fetal Center

The Spates twins were diagnosed prenatally via ultrasound at 25 weeks gestation. Their mother, Adrienne Spates, was referred to Le Bonheur's Fetal Center in November 2011. Spates and her boys were followed by Fetal Center Medical Director Giancarlo Mari, MD, who coordinated care with various subspecialists over the duration of the pregnancy.



Le Bonheur Fetal Center Medical Director Giancarlo Mari, MD, performs a fetal ultrasound on 27-year-old Adrienne Spates. Spates was followed in the Fetal Center for eight weeks, after learning she was pregnant with conjoined twins.

Pygopagus Conjoined Twins



Joined at the lumbosacral spine shared a rectum, muscle and nerves



From left, Plastic Surgeon Robert Wallace, MD, Neurosurgeon Rick Boop, MD, Craniofacial and Pediatric Plastic Surgical Fellow Ben Gbulie, MD, and Neurosurgeon Michael Muhlbauer, MD, evaluate the Spates twins during surgery. Jacob and Joshua Spates were separated almost 10 hours into their separation surgery on Aug. 29.

Fetal ultrasound, with three-dimensional software, and fetal MRI — coordinated by the Department of Radiology — were used to help diagnose the twins in utero.

The boys were transferred to Le Bonheur Children's Hospital seven hours after Mari delivered them via Caesarean section on Jan. 24.

Baby A, or Joshua, was diagnosed with complications of situs inversus totalis, calcifications in the spleen and a heart defect. Baby B, or Jacob, was diagnosed with Dandy-Walker syndrome, hydrocephalus, two-vessel umbilical cord and a heart defect.

About Conjoined **Twins**

- Conjoined twins are rare and occur in roughly one in every 100,000 pregnancies.
- 40 to 60 percent of conjoined twins are stillborn, and many die within the first few days of birth.
- About 70 percent of conjoined twins are female.
- About 15 percent of conjoined twins are pygopagus and joined back to back at the pelvis and lower spine, each with separate hearts, heads and limbs.

About Conjoined Twins in Memphis

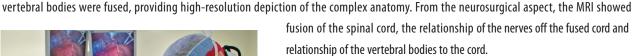
- Six documented conjoined twins in Memphis history.
- Last separated case occurred in 1970s.
- Previous separations performed in Memphis resulted in the death of at least one of the twins.

Growth and Surgery Preparation, Le Bonheur Neonatal Intensive Care Unit

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Two days after birth, pediatric surgeons performed a colostomy and inserted a gastrostomy tube to assist with nutrition and waste elimination. The twins spent the next seven months in Le Bonheur's Neonatal Intensive Care Unit, where they grew from 2.92 kilograms to 13.53 kilograms. Ongoing therapy and treatment from a multidisciplinary team of subspecialists allowed them to grow and develop until their surgical separation.

Meanwhile, surgeons used the time to plan for separation of the boys, largely relying on Radiology's 320-slice CT Scan and MRI technology to understand their connection. The hospital's 320-slice CT Scanner provided software that helped physicians see where the



Conjoined twins Joshua, left, and Jacob, right, sit in a swing in their NICU

room during a therapy session. The swing allowed the 4-month-old boys

to get out of their bed and play during their long stay at Le Bonheur.

On July 26, Plastic Surgeon Robert Wallace, MD, implanted tissue expanders to prepare for the separation. Those expanders were injected with saline six subsequent times prior to separation.



Plastic Surgeon Robert Wallace, MD, left, and Craniofacial and Pediatric Plastic Surgical Fellow, Ben Gbulie, MD, close the skin on each twin's back. Shortly after separation, surgeons quickly closed the skin on each boy so Joshua could be transferred to another operating room.

Future Prognosis

Baby A (Joshua) has since gone home. Baby B (Jacob) remains at Le Bonheur, awaiting surgery to repair his double outlet right ventricular heart defect. Both have ostomies and will be followed long term in Le Bonheur's Spina Bifida Clinic.

Joshua is expected to function like a child with spina bifida involving a lower lumbar spinal cord and should be able to walk with braces, said Orthopaedic Surgeon William Warner, MD. Warner hopes Jacob will be able to walk with braces as well.

Separation Surgery

The Spates twins were separated on Aug. 29, 2011, in a 13-hour surgery where surgeons separated the spinal column, spinal cord and muscles and completed gastrointestinal repairs. The surgical team included 35 members, including surgeons, anesthesiologists, certified registered nurse anesthetists, surgical technicians, registered nurses and a radiological technician.

Each twin was assigned a surgical team for each specialty — general surgery, neurosurgery, orthopaedics and plastic surgery. An anesthesiologist and certified nurse anesthetist were also assigned to each twin. The boys were separated a little more than 10 hours into the surgery and then moved to separate operating rooms.



Flat television screens in Le Bonheur's operating rooms allow the entire surgical team to see a surgeon's microscopic view. The technology served as a key teaching tool for this complex separation surgery



Surgical Timeline: Separation of Conjoined Twins Joshua and Jacob Spates

Aug. 29, 2011

7:07 - 9:27 a.m.

Anesthesia team begins induction of anesthesia and inserts intravenous lines

9:45 - 10:04 a.m.

10:11 - 11:08 a.m.

11:11 – 11:38 a.m. Plastic surgery team elevates the skin flap on Jacob's leg

Neurosurgery team places ventricular access device into Jacob's head Team prepares and applies sterile drapes to both babies to ensure a sterile field

and elevates the second skin flap on Joshua's leg Neurosurgery and orthopaedics teams access the 1:32 – 3:41 p.m. conjoined spinal canal and separate the bony elements of the spine and spinal column 3:42 - 5:22 p.m.

12:41 – 1:23 p.m.

1:27 – 2 p.m.

General surgery completes the colon separation and prepares Joshua to be moved to Operating Room 6

Plastic surgery team removes both tissue expanders

Babies are flipped from side to side

Joshua is moved to Operating Room 6 5:02 p.m.

11:40 a.m. – 12:10 p.m. General surgery team begins preparation for separating the colon

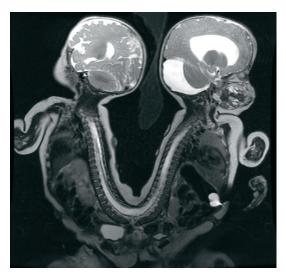
Diagnostic images, radiologists give surgeons a leg up in planning

 $\overline{}$ rom the moment the Spates twins were diagnosed as conjoined, diagnostic imaging became a key tool for their care. Physicians relied on ultrasound, MRI and CT scans, plain film and fluoroscopy and an experienced team of radiologists to develop a plan to separate the boys and diagnose their problems.

Prenatally, fetal ultrasound with 3D software and fetal MRI confirmed the fusion of the twins' vertebral column and soft tissue of posterior back area, along with showing other congenital defects for each fetus.

Once the boys were born, MRI imaging and 320-slice CT scans were used to show the fusion of the spinal cord and vertebral bodies. The boys received both MR and CT scans in July — about six weeks before separation to help surgeons plan procedures.

Pre-operative, 320-slice CT scans allowed for accurate 3D computer reconstruction of vertebral bodies. Orthopaedic surgeons used the CT scans to define the complex bony connections between the twins' pelvis



MR images taken of Joshua, left, and Jacob, right, show where the boys were conjoined, in the lower sacral spine. Radiologists used fetal ultrasound and fetal MRI to learn more about them before they were born. After birth, 3D CT scans and MRI technology helped further characterize their conditions.

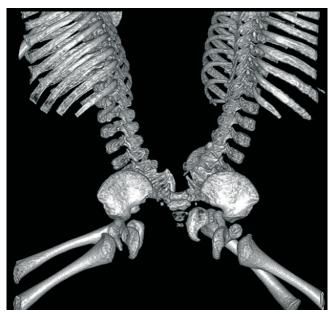
and shared sacrum — and found no surprises during the actual surgery. The additional information enabled surgeons to move quickly through the orthopaedic portion of the separation.

MR scans clearly showed fusion of the cord, the relationship of nerves off the fused cord and relationship of the vertebral bodies to the cord.

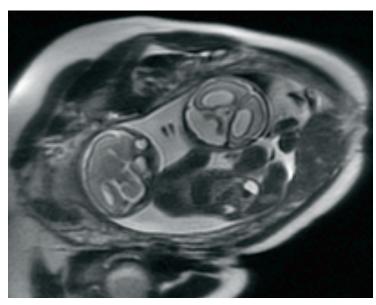
"Neuroradiologist Asim Choudhri, MD, created some beautiful spinal cord tractography on the case showing the crossing nerve fibers," said Rick Boop, MD, chief of Neurosurgery at Le Bonheur Children's. "It helped us tell mom what to expect neurologically."

Le Bonheur Chief of Radiology Harris Cohen, MD, credits both the technology and experienced Maternal Fetal Medicine and Radiology clinical images with helping plan for the surgery.

"Our team of pediatric radiologists have varying levels of broad and subspecialty experience. They are able to use the great technological tools and software programs to get the most from our images," Cohen said. "I can't emphasize enough how our experience, ability to think outside the box and willingness to consult with each other helps put things together for best patient care and diagnosis."



A 3D reconstruction of 320-slice CT scan shows how vertebrae for the Spates twins are conjoined at 6 months of age.



A fetal MRI before they were born shows the Spates twins and where they are conjoined.



Operating Room 7 Timeline

5:33 – 5:50 p.m.

Team prepares and re-drapes Jacob to maintain the sterile field

5:54 – 6 p.m.

Jacob's surgery resumes; neurosurgery and orthopaedic teams repair the myelomeningocele and close Jacob's back and spine

6 – 7:15 p.m.

7:17 – 8:15 p.m.

Jacob's anus Plastic surgery team advances expanded skin and rotates flap to cover vital structures

General surgery team repairs



Operating Room 6 Timeline

5:40 – 5:45 p.m. Team prepares and re-drapes Joshua to maintain

sterile field

5:47 – 6:38 p.m.

Joshua's surgery resumes; neurosurgery and orthopaedic teams repair the myelomeningocele and close Joshua's

back and spine

6:38 – 6:52 p.m. General surgery team repairs Joshua's anus

6:54 – 8:02 p.m. Plastic surgery team advances expanded skin and rotates

flap to cover vital structures

Both babies are moved to the Pediatric Intensive Care Unit 8:16 p.m.



Jim Beaty, MD Role: Orthopaedic surgeon

Orthopaedic surgeon, Campbell Clinic Orthopaedics and Le Bonheur Children's Hospital; Professor of Department of Orthopaedic Surgery, The University of Tennessee Health Science Center, (UTHSC)



Max Langham, MD Role: Team leader, lead general surgeon, leader of planning and preparation efforts

Medical director of Pediatric Surgery, Le Bonheur Children's Hospital; Chief of Pediatric Surgery and director of the Residency Program, UTHSC



Rick Boop, MD Role: Neurosurgeon

Chief of Pediatric Neurosurgery, Le Bonheur Children's Hospital; Professor and J.T. Robertson Chairman, Department of Neurosurgery, UTHSC and St. Jude Children's Research Hospital; Neurosurgeon, Semmes-Murphey Neurologic and Spine Institute



Giancarlo Mari, MD Role: Prenatal care and obstetrics team leader

Medical director of the Fetal Center, Le Bonheur Children's Hospital; Director of High-Risk Obstetrics Center of Excellence, The Regional Medical Center; Professor and chairman of the Department of Obstetrics and Gynecology, UTHSC



Asim F. Choudhri, MD Role: Neuroradiologist

Neuroradiologist, Le Bonheur Children's Hospital; Assistant Professor of Radiology and Neurosurgery, UTHSC



Michael Muhlbauer, MD Role: Lead neurosurgeon, spinal cord separation

Neurosurgeon, Semmes-Murphey Neurologic and Spine Institute and Le Bonheur Children's Hospital; Assistant professor, UTHSC



Harris L. Cohen, MD Role: Radiologist

Medical director of Radiology, Le Bonheur Children's Hospital; Professor of Radiology, Pediatrics and Obstetrics and Gynecology, UTHSC; Chairman of Radiology, UTHSC;



B. Rao Paidipalli, MD Role: Anesthesiologist

Operating Room Patient Flow director, Le Bonheur Children's Hospital; Assistant professor for the Department of Anesthesiology, UTHSC



Ramasubbareddy Dhanireddy, MD Role: Neonatologist

Medical director of Neonatology, Le Bonheur Children's Hospital; Chief of Neonatology, UTHSC



Marilyn Robinson, MD Role: Neonatologist

Neonatologist, Le Bonheur Children's Hospital and The Sheldon Korones Newborn Center; Assistant professor, UTHSC



James "Trey" Eubanks, MD Role: General surgeon

Medical director of Trauma Services, Le Bonheur Children's Hospital; Assistant professor, UTHSC



Joel Saltzman, MD Role: Anesthesiologist

Medical director of Pediatric Anesthesiology, Le Bonheur Children's Hospital; Assistant professor of Pediatrics and Anesthesiology, UTHSC



Eunice Huang, MD Role: General surgeon

General pediatric surgeon, Le Bonheur Children's Hospital; Assistant professor, UTHSC

Derek Kelly, MD

Clinic, Le Bonheur Children's Hospital;

Assistant professor for the Department

of Orthopaedic Surgery, UTHSC



Jeff Sawyer, MD

Orthopaedic surgeon, Campbell Clinic Orthopaedics and Le Bonheur Children's Hospital; Director of the Pediatric Orthopaedic Fellowship Program, University of Tennessee/Campbell Clinic Department of Orthopaedic Surgery; Associate professor for the Department



Robert Wallace, MD

THE SURGICAL TEAM

twins Joshua and Jacob Spates was

required multiple pediatric surgical

orthopaedic surgery, plastic surgery

and radiology. Each team member

was assigned to a twin and specialty

The 35-member pediatric surgical

The separation of conjoined

the most complex in Le Bonheur

specialties, including anesthesia,

general surgery, neurosurgery,

throughout the procedure.

6 general surgeons

4 neurosurgeons

3 plastic surgeons

2 anesthesiologists

anesthetists

7 registered nurses

organizer

5 surgical technicians

1 radiological technician

1 sterile processing department

4 orthopaedic surgeons

2 certified registered nurse

team included:

history. The 13-hour surgery

Role: Lead plastic surgeon, insertion and removal of skin expanders, incision design

Program director for the Craniofacial and Pediatric Plastic Surgery fellowship at Le Bonheur Children's Hospital; Professor and chairman of the Department of Plastic Surgery, UTHSC; Chief of Plastic Surgery, St. Jude Children's Research Hospital



William Warner, MD

Role: Lead orthopaedic surgeon, spinal column separation

Orthopaedic surgeon, Campbell Clinic Orthopaedics and Le Bonheur Children's Hospital; Professor, department of Orthopaedic Surgery, UTHSC; Chief of Orthopaedics, St. Jude Children's Research Hospital; Chief of Orthopaedics, Mississippi Crippled Children's Services



Role: Orthopaedic surgeon

of Orthopaedic Surgery, UTHSC



Mark Williams, MD, FAAP, **FACS**

Role: Urologist

Associate chief of staff-elect, Le Bonheur Children's Hospital; Chief of Urology, Le Bonheur Children's Hospital and St. Jude Children's Research Hospital; Assistant professor, chief of Pediatric Urology and program director for the Pediatric Urology Fellowship Program, UTHSC

OTHER SURGICAL TEAM MEMBERS

Fridae Hammons, RN, PCC Trauma and Orthopaedics Role: Operating Room Team Leader, Circulator

Nikki Freeman, RN Role: Command Center Facilitator

Surgical Fellows Sonia Alvarez, MD,

Plastic Surgery
Berkeley Bate, MD,
Neurosurgery
Uzoma "Ben" Gbulie, MD,

Plastic Surgery
Jimmy Green, MD,
General Surgery
Jeremy Rush, MD,
Orthopaedics

Thomas Sims, MD, General Surgery Scott Wate, MD, Neurosurgery
Regan Williams, MD,
General Surgery

Circulators

Ben Cornelius, RN

Dawn Cunningham, RN General Surgery, Plastic Surgery Chad Owens, RN

Neurosurgery
Cheryl Perkins, RN,
Orthopaedics
Casey Turner, RN Neurosurgery

Surgical Technicians

Wilhelmenia Dill, CST Valerie Guy, CST Plastic Surgery Jacob Howell, CST Neurosurgery Ken Taylor, CST Neurosurgery, Orthopaedics, General Surgery, Plastic Surgery Torya Woods, CST General Surgery

Certified Registered Nurse Anesthetists Brian Cain, CRNA

Radiographer Patrick Martin

Sterile Processing Department Joyce Mitchell, ST

Equipment, In-room Camera Brenda Bryan, OR Biomed