The Fetal Center Newsletter

Fall 2014 • 7th Edition

The Fetal Center at Children's Memorial Hermann Hospital, in collaboration with UTHealth Medical School.

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Ways to Give to The Fetal Center

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Superior Pre- and Postnatal Care to Treat Congenital Heart Defects Leads to Rapid Growth

Gurur Biliciler-Dentkas, M.D. and Helena Gardiner, M.D., Ph.D., co-directors of the Fetal Cardiology Program.

In April 2014, after a review of relevant literature, the American Heart Association (AHA) released a scientific statement on the diagnosis and treatment of fetal cardiac disease. The AHA writing group, comprised of 18 physicians and one nurse, provided recommendations related to the specifics of fetal diagnosis and described the components of fetal echocardiography in detail.

"This is the first comprehensive statement on fetal heart disease made by the American Heart Association," says Gurur Biliciler-Denktas M.D., co-director of the Fetal Cardiology Program at The Fetal Center at Children's Memorial Hermann Hospital and an associate professor of pediatric cardiology at UTHealth Medical School. "It provides detailed information about modalities for fetal cardiac assessment, protocols for screening and referral, experience required for performing and interpreting studies and available fetal therapies."

The AHA statement's release coincides with a period of rapid growth for The Fetal Center, as well as the divisions of Pediatric Cardiology and Pediatric Cardiovascular Surgery at UTHealth Medical School. "Our volumes at The Fetal Center have increased and will continue to grow as more physicians develop an understanding of the importance of fetal cardiac screening, and because they have confidence in our diagnostic and surgical outcomes," Dr. Denktas says.

Once an expectant mother is referred to The Fetal Center to confirm the diagnosis of a congenital heart defect, she has access to a fetal cardiology team that consists of three affiliated physicians. Two of them – Dr. Denktas and Monesha Gupta, M.D. – also see patients after birth, ensuring continuity of care. Since 2008, Dr. Denktas has extended that continuity into adulthood through an adult congenital heart disease clinic for patients age 16 and older.

"Screening for congenital heart disease has generated a lot of interest in the last few years because of poor detection rate, with only one in four babies with major congenital defects discovered in ultrasound scans," says Helena Gardiner, M.D., Ph.D., who specializes in fetal echocardiography and is co-director of the Fetal Cardiology Program at The Fetal Center. "The diagnosis of cardiac disease in the fetus is generally made with ultrasound, but obstetricians were not ordering enough views of the heart. The International Society of Ultrasound in Obstetrics and Gynecology revised their guidelines a couple of years ago to include five transverse views of the heart, a proven and practical protocol. These guidelines were created to raise the bar and push providers around the world to the next level."

continued

Since her arrival at The Fetal Center in July 2013, Dr. Gardiner has established the Congenital Heart Defect Screening Program, which works to improve early detection and care by educating and training health professionals, campaigning for improvement and raising awareness of the signs of undetected heart defects in the community. Through the program Dr. Gardiner trains sonographers to perform the five transverse views to improve their skill, confidence and their success in the detection of heart defects before birth.

"We've taken our skilled technicians to referring maternal-fetal medicine specialists and obstetricians and trained their staff onsite in fetal heart screening," says Dr. Gardiner, a professor at UTHealth Medical School with dual appointments in the department of Obstetrics, Gynecology and Reproductive Sciences and the department of Pediatric Cardiology. "We give them practical hands-on training and show examples of heart defects to encourage them to use the five transverse-view approach. As a result we're seeing more referrals of major congenital heart defects early in gestation from these offices.

When babies with major heart conditions are diagnosed before birth, health professionals can plan for their care effectively, supporting and preparing parents through the pregnancy to ensure that the baby is born in a tertiary care facility with access to experts in fetal medicine and fetal cardiology, as well as multidisciplinary specialists. At The Fetal Center and Children's Memorial Hermann Hospital, care of mother and baby is a team effort based on an integrated care pathway.

The expert multidisciplinary team at The Fetal Center has contributed to the growth of its referral network and an increase in patient volumes. Referrals to the fetal cardiology program for major heart defects have more than doubled in the last 12 months. "The overall increase in fetal cardiology volumes has led to an increase in the number of pediatric cardiovascular surgeries we perform," says William Douglas, M.D., chief of pediatric cardiovascular surgery and director of the Children's Heart Institute at Children's Memorial Hermann Hospital. An associate professor and director of the division of Pediatric Cardiovascular Surgery at UTHealth Medical School, Dr. Douglas is actively recruiting a third pediatric cardiothoracic surgeon in response to the new demand.

"Our volumes are now growing at the rate of 10 percent annually," he says. "We're actively involved with The Fetal Center – I see one or two moms each week at the Center whose babies have been diagnosed with congenital heart disease. While most babies do not require emergent surgery, these mothers have the advantage of knowing in advance so they can prepare themselves mentally and emotionally. It's very difficult on mothers who learn immediately after delivery that their baby has a heart defect and will face surgery in the next few days. Our operative team is prepared to do surgery on relatively short notice, but we're grateful that more and more defects are diagnosed before birth."

With a new director of the division of Pediatric Cardiology in place at UTHealth Medical School, the fetal and pediatric cardiology programs are poised for future growth. John P. Breinholt, M.D., was named director of the division and chief of pediatric cardiology at Children's Memorial Hermann Hospital in June.

His collaboration with Dr. Douglas extends well beyond his short time at Children's Memorial Hermann Hospital, having worked with Dr. Douglas for the last eight years during annual medical mission trips to Bolivia. "When the position became available, Dr. Douglas encouraged me to consider it, then introduced me to Eric Eichenwald, M.D., chair of the department of Pediatrics. I was impressed with his leadership and the leadership from the other associated divisions and departments. I was excited about the opportunity to contribute to the growth of this program," said Dr. Brienholt.

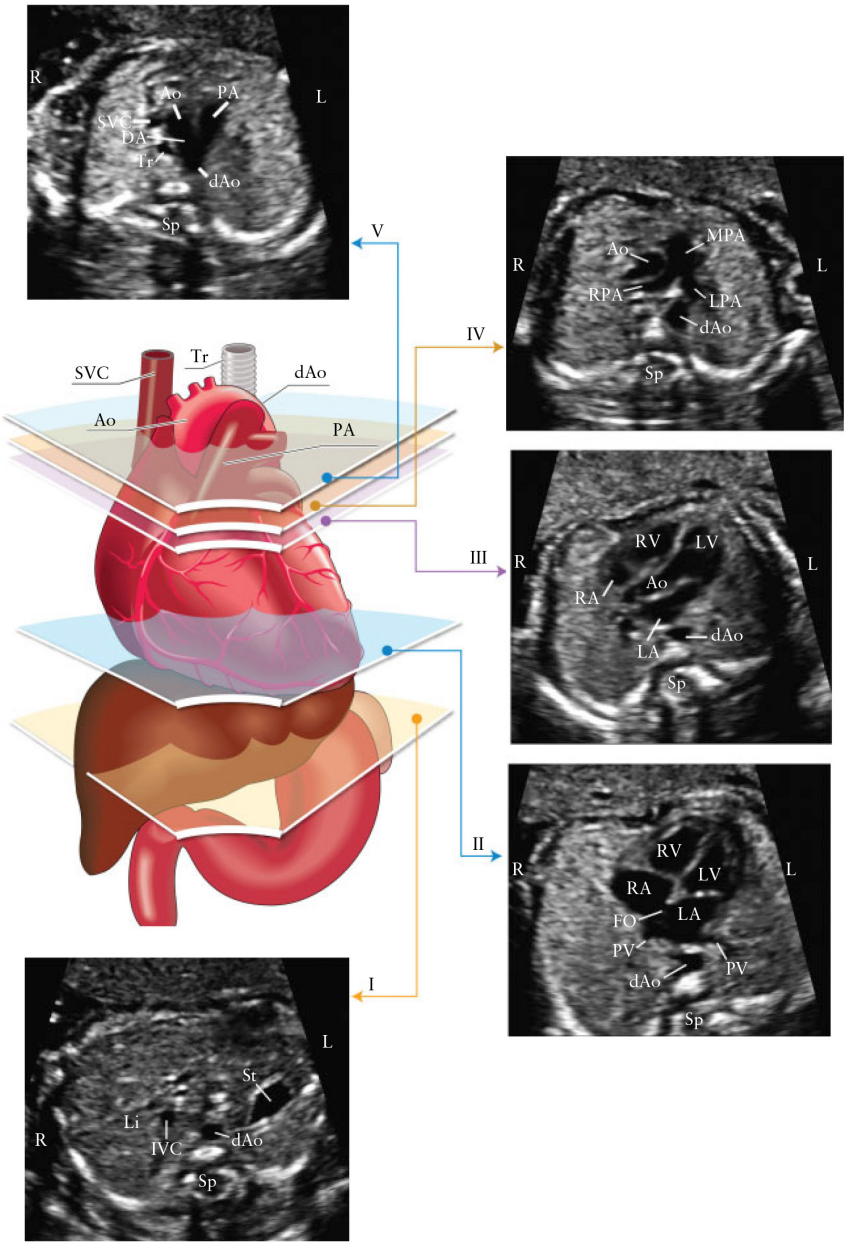
An interventional cardiologist whose clinical interests include cardiac catheterization, implementing new devices in the nonsurgical treatment of heart defects and hybrid surgical/catheter procedures, Dr. Breinholt has a bold vision for Children's Memorial Hermann Hospital and the division of Pediatric Cardiology. "Within the next year, I'd like to see us introduce pulmonary valve implants and expand the number of hybrid procedures cardiologists and cardiovascular surgeons perform together. We want to further integrate our services and develop practice plans to ensure that the care we provide is totally seamless. We meet weekly with The Fetal Center staff to discuss strategy and to ensure the family is taken care of from prenatal diagnosis through pregnancy and delivery and through adulthood."

To learn more about the fetal cardiology program, visit childrensmemorialhermann.org/fetalcardiology

Five Transverse View of the Fetal Heart

In 2013, The International Society of Ultrasound in Obstetrics and Gynecology (ISUOG) published updated practice guidelines for screening of the fetal heart, suggesting early detection rates can be optimized by performing a thorough examination of the fetal heart by complementing the four-chamber view with the outflow tract views in the cardiac screen, thus totalling five transverse views. This recommendation supports the educational outreach and sonographer training program led by Helena Gardiner, M.D., Ph.D.

UNDERSTANDING THE FIVE TRANSVERSE VIEWS:



The image is representative of the five axial views for optimal fetal heart screening. The color image shows the trachea (Tr), heart and great vessels, liver and stomach, with the five planes of insonation indicated by polygons corresponding to the gray-scale images, as indicated.

I. Most caudal plane, showing the fetal stomach (St), cross-section of the descending aorta (dAo), spine (Sp) and liver (Li).

II. Four-chamber view of the fetal heart, showing the right and left ventricles (RV, LV) and atria (RA, LA), foramen ovale (FO) and pulmonary veins (PV) to the right and left of the dAo.

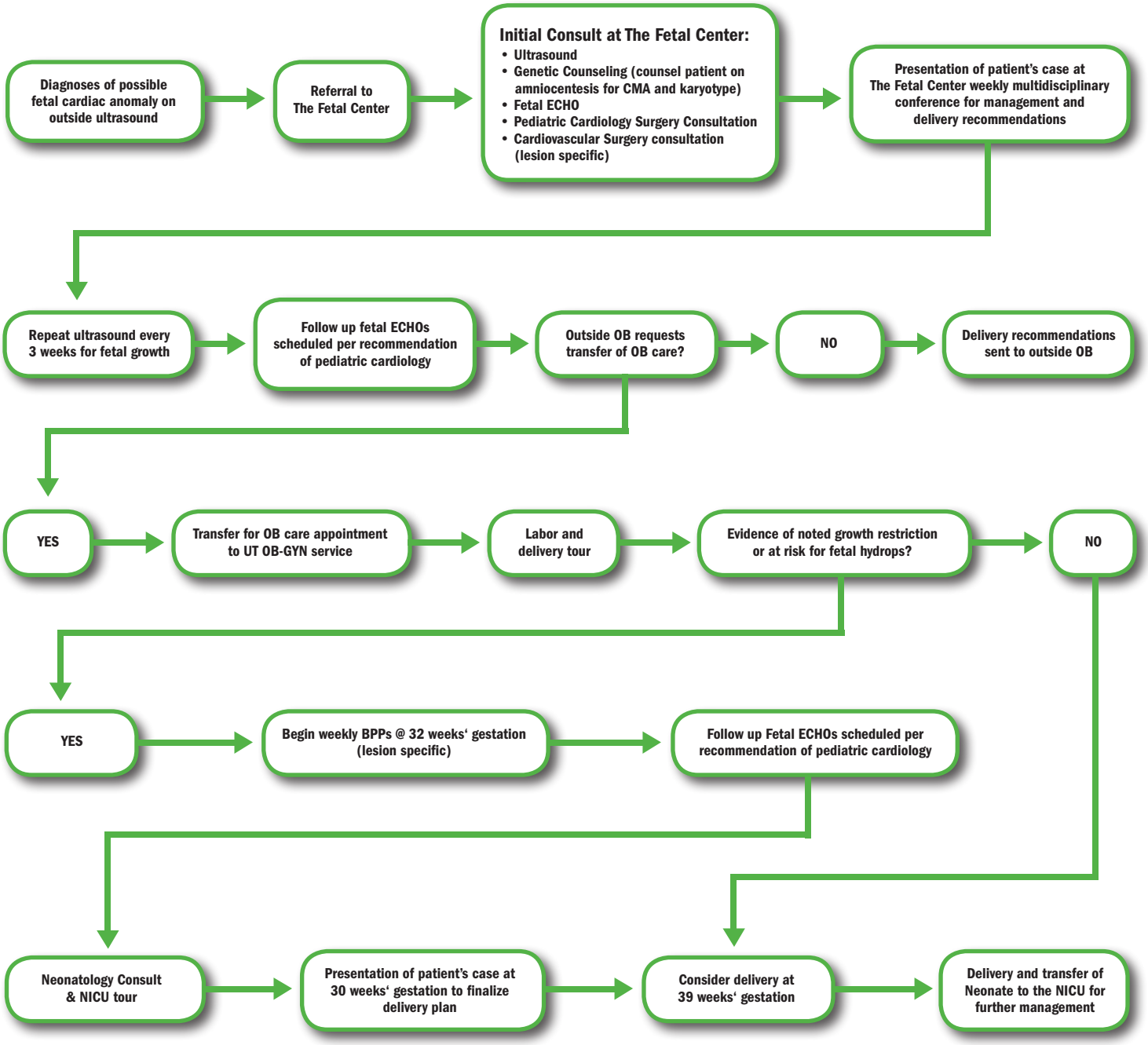
III. Left ventricular outflow tract view, showing the aortic root (Ao), LV, RV, LA and RA and a cross-section of the dAo.

IV. Slightly more cephalad view (right ventricular outflow tract view) showing the main pulmonary artery (MPA) and the bifurcation into the right (RPA) and left (LPA) pulmonary arteries and cross-sections of the ascending aorta (Ao) and dAo aorta.

V. Three vessels and trachea view showing the superior vena cava (SVC), pulmonary artery (PA), ductus arteriosus (DA), transverse aortic arch (from proximal Ao to dAo) and trachea (Tr). IVC, inferior vena cava; L, left; R, right. Modified with permission from Yagel et al.

Image provided by the International Society of Ultrasound in Obstetrics and Gynecology. Yagel S, Cohen SM, Achiron R. Examination of the fetal heart by five short-axis views: a proposed screening method for comprehensive cardiac evaluation. Ultrasound Obstet Gynecol 2001; 17: 367–369

Cardiac Anomaly Patient Algorithm



This algorithm was developed to promote consistency in the general care of patients. This algorithm is flexible and is not intended to set out a standard of care. Professional judgment must always be exercised in order to meet the specific requirements of individual patients. This algorithm does not preclude the use of professional judgment in a specific situation. This algorithm is the intellectual property of Memorial Hermann Health System and cannot be reproduced in whole or in part without expressed written permission.

Approved 09/02/2014

Patient Story

After a Fetal Diagnosis of Cor Triatriatum, a Successful Outcome

Gemma and baby Faustina

When Karolina Adam, M.D., found excessive fluid behind the neck of Gemma Nguyen's baby during her 10-week nuchal translucency screening, she suspected Down syndrome or a heart defect. After ruling out Down syndrome, the Houston-area maternal-fetal medicine specialist referred her patient to Helena Gardiner, M.D., Ph.D., for a closer examination of the fetal heart.

A world-renowned fetal cardiologist who specializes in fetal echocardiography, Dr. Gardiner is co-director of the Fetal Cardiology Program at The Fetal Center at Children's Memorial Hermann Hospital, and a professor at UTHealth Medical School with dual appointments in the department of Obstetrics, Gynecology and Reproductive Sciences and the department of Pediatric Cardiology. She first saw Nguyen when the 34-year-old was 24 weeks and four days into her pregnancy.

"On the first fetal echocardiogram, we saw a structure in the left atrium. I thought it was an enlarged coronary sinus but could not detect any abnormal venous structures entering it, so we couldn't reach a conclusive diagnosis," Dr. Gardiner says. "When we did a second echocardiogram at 34 weeks, we could see the baby had cor triatriatum, a rare congenital anomaly in which a fibromuscular membrane divides an atrium – usually the left – in two. It's important to detect this before birth because the membrane can block the return of red blood from the heart to the lungs once the baby begins to breathe. For that reason, we discussed with Dr. Adam how best to manage the delivery. In cases like this we want to be prepared for anything."

Dr. Gardiner arranged for Nguyen to meet the pediatric cardiovascular team, including pediatric cardiothoracic surgeon William Douglas, M.D., chief of pediatric cardiovascular surgery and director of the Children's Heart Institute at Children's Memorial Hermann Hospital. Dr. Douglas is an associate professor and director of the division of Pediatric Cardiovascular Surgery at UTHealth Medical School. "At that point I was worried and anxious about everything," Nguyen says. "I asked my family and friends to pray for my daughter and to give us a team of doctors who knew how to care for her. I received great support from my family, friends, doctors and the entire team at The Fetal Center – they were a wonderful help to me and gave me hope and strength. They made sure I understood what to expect and assured me that they would do their very best to take care of my daughter after her birth. Dr. Douglas was very optimistic about the case. If the heart doesn't function well, he said, she will have an open heart procedure to remove the extra tissue in her atrium. It's a rare defect but a doable procedure, they all assured me, and I had confidence in them."

Faustina Thao-Nhien Nguyen was born on April 18, 2014, at 1:53 a.m. On the morning of the delivery, a team that included her maternal-fetal medicine specialist, pediatric cardiology, pediatric anesthesiology and pediatric cardiovascular surgery was awaiting her arrival at Children's Memorial Hermann Hospital.

"Dr. Adam delivered Gemma at Children's Memorial Hermann Hospital, which is not her usual practice, to allow access to immediate cardiovascular surgery should it be necessary in the first few hours after birth," Dr. Gardiner says. "Luckily, Faustina came out crying and screaming. The membrane was still there, but it had a nice big hole in it, which was seen before birth. Sometimes after the child is born, we find the hole is smaller than expected and there is more blood passing through it, which causes an obstruction to flow into the left-sided pump. So this was a great outcome and a happy discovery for the team."

Faustina spent the night in the Neonatal Intensive Care Unit and was sent home with her mother a few days later. She is now being followed by the UTHealth Pediatric Cardiology team. Nguyen is thankful her baby is healthy and may not need surgery for the rest of her life. "I believed a miracle could happen," she says. "Faustina's heart is still carefully being monitored by her pediatric cardiologist but she doesn't have to go back as often anymore. First every four months, then six months, then only once a year."

Her story demonstrates how early detection, prenatal diagnosis, pregnancy management and coordination of delivery at a tertiary care facility like Children's Memorial Hermann Hospital can result in a good outcome for babies with a potentially life threatening cardiac condition. The multidisciplinary teams were able to monitor the baby throughout the pregnancy and develop a customized delivery plan with specialists ready to intervene and treat the baby as needed.

Patient Story

Faith at 3 years old.

Texas' First Fetal Spina Bifida Repair Patient Turns 3

Picture the bounciest, bubbliest 3-year-old you can imagine. Her songs and dance moves come straight from Beyoncé. She loves bright colors and runs to the mirror to check every outfit. She has the attitude of an actress, the energy of an entertainer and the million-dollar smile of a model. And she is a child who was prenatally diagnosed with spina bifida, whose future at one time looked more filled with shunts, catheters and leg braces than mirrors and dancing to music videos.

Faith's medical team at The Fetal Center at Children's Memorial Hermann Hospital would become the first in Texas to perform an in utero surgery for spina bifida repair. A complex and sometimes permanently disabling birth defect that affects about one in every 1,500 pregnancies, spina bifida involves incomplete development of a portion of the spinal cord and associated nerves, as well as the surrounding spinal bones and overlying muscle. Infants born with spina bifida are at risk for a range of disorders, including hydrocephalus, which may require a shunt to relieve pressure inside the skull. They are prone to life-threatening infections and may suffer loss of sensation or paralysis, difficulty walking and other problems that lead to lifelong disability.

In 2011, fetal surgeon KuoJen Tsao, M.D., and pediatric neurosurgeon, Stephen Fletcher, D.O., were following the results of the landmark study (Management of Myelomeningocele Study, or MOMS Trial) that found if a fetus undergoes surgery in utero to repair the spina bifida defect, serious complications could be reversed or lessened when compared to infants repaired after birth. The study found that fetal surgery decreased the need for shunting at 12 months, and nearly half were able to walk without crutches by 2 1/2 years.

Dr. Tsao and his maternal - fetal medicine colleagues were introduced to Faith's parents, Ivan and Colette Hagler of Dallas, when she was referred to The Fetal Center following a prenatal diagnosis of myelomeningocele at 20 weeks gestation. Colette underwent two full days of comprehensive evaluation by a multidisciplinary specialty team. After close examination, keeping strictly aligned to the MOMs Trial protocols, the team determined that Colette and Faith were ideal candidates for the procedure.

Following the surgical procedure, Colette remained at Children's Memorial Hermann Hospital for the remainder of her pregnancy and delivered Faith on the Fourth of July – a date that Colette describes as "symbolic" of the independence her daughter has discovered since her birth. The Hagler family travels to Houston regularly for Faith's follow-up treatment, most recently orthopedic surgery to repair club feet and low arches. Her first year was difficult, her mom recalls, "Faith had several bouts of a respiratory virus common among preemies. We knew there was no guarantee when we chose to undergo the fetal surgery. When her medical team determined that shunts or catheterization were unnecessary, we began to see the benefits that the clinical trials indicated."

At 11 months, Faith began crawling and at 21 months she was taking her first steps. It was a memorable moment, to say the least. Ivan confesses that daily he is in awe of the progress his daughter makes. "We thank God for blessing our little girl with the ability to walk."

According to Dr. Tsao, Faith has demonstrated exactly what the MOMS Trial showed the in utero surgery was designed to do. Faith has met all the milestones for her age: she knows her name and other facts about herself, answers questions and speaks in five- to six-word sentences. "Cognitively and linguistically, she's right on board," he says. "Much of her follow-up medical care now focuses on keeping an eye on things through her growth and development."

Dr. Tsao gives a lot of credit to Faith's parents. "When I look at her, I see their sacrifice and commitment. They've been totally committed to caring for a spina bifida child, which presents lots of challenges. They're a great family and they've been a resource for other families who are undergoing or have undergone fetal surgery."

Like the independence implicit in her birth date, there's providence in Faith's name as well. "When I see her," her mother says, "I see faith – the substance of things hoped for – in the amazing things she has been able to do in the past three years."

Patient Story

Coming Home: Moving Back to Houston For Our Twins

Dr. Anthony Johnson with Penelope and Charlotte at their baptism

Already parents of two, Ben and Ashley Agee knew this third pregnancy was going to be a little more complicated – but they didn't yet realize it would mean a move back to Texas. They were in the process of relocating their family from Houston to Jackson Hole, Wyoming, when Ashley learned she was pregnant. A week later they found out they were having twins.

During a routine checkup at 23 weeks, the couple discovered their identical twins were experiencing twin-to-twin transfusion syndrome (TTTS), a life-threatening condition that requires specialized care and intervention.

"There aren't any specialists in Jackson Hole. There are doctors that come every two weeks from Salt Lake City, Utah, to see cases like mine," says Ashley. "We didn't have time to wait. We hopped on a flight the night we were diagnosed and met with our fetal medicine specialist a few hours later." That flight took Ben and Ashley back to Houston to meet with Kenneth Moise, M.D., and Anthony Johnson, D.O., both co-directors of The Fetal Center at Children's Memorial Hermann Hospital and professors in the division of Maternal-Fetal Medicine and department of Pediatric Surgery at UTHealth Medical School.

Dr. Johnson, Dr. Moise and the fetal intervention specialty team successfully performed a fetoscopic laser ablation on the twins, which means using a special laser to seal off the blood vessels connecting them. "In general, the twins are going to do OK with careful monitoring," says Dr. Moise. "Survival rates today in intensive care units are very good, should the babies be born prematurely. At Children's Memorial Hermann Hospital, we have the full scale of expertise both on the fetal and the maternal side, and we have one of only two Level IV neonatal intensive care units in this region of Texas that can deal with all the complications of twins." In October, nearly 35 weeks into her pregnancy, Ashley delivered Penelope and Charlotte, who are now healthy and happy. The experience was life changing in more ways than one. "After this scare, sophisticated medical care is high on our priority list," says Ashley.

The Agee family is incredibly grateful for their experience with The Fetal Center and this year provided the center with a generous contribution. "The Fetal Center team was comforting. It felt like they were an extension of our family," says Ashley. "Daily, we are reminded of the life-changing work they do."

This May, the family's story was featured in the premiere Kids' Health Annual edition of Houstonia magazine, a parenting survival guide created in collaboration with Children Memorial Hermann Hospital to educate parents and caregivers with medical tips from physicians, facts about medical myths and patient stories. Ben and Ashley shared their story with the community and were honored guests at a private reception in May, held to celebrate stories such as the Agee family's as well as the affiliated physicians, nurses and staff who treat patients at Children's Memorial Hermann Hospital with the highest quality of care.

The entire care team would like to thank the Agee family for their generous financial gift to the Memorial Hermann Foundation to honor The Fetal Center at Children's Memorial Hermann Hospital. Their support will continue to drive forward the mission of advancing the field of maternal-fetal medicine through innovative research, physician outreach and patient education.

For more information on how you can support The Fetal Center, please call the Memorial Hermann Foundation at 713.242.4409 or email deborah.lackey@memorialhermann.org.

News of Note

Chief of Pediatric Cardiology Named

John P. Brienholt, M.D., was named chief of pediatric cardiology at Children's Memorial Hermann Hospital and director of pediatric cardiology at UTHealth Medical School. Dr. Breinholt was recruited from Indiana University School of Medicine, where he was an associate professor of pediatrics, director of the Pediatric Cardiology Fellowship Program and director of education for the division of Pediatric Cardiology.

The position marks a return to familiar territory for Dr. Breinholt. After graduating from the University of Utah School of Medicine, he completed his pediatric residency at Baylor College of Medicine and Texas Children's Hospital. After completing his fellowship in pediatric cardiology and an advanced fellowship in cardiac catheterization from the same institution, Dr. Brienholt was recruited to Indiana.

Dr. Brienholt is an interventional cardiologist whose clinical interests include cardiac catheterization, novel device therapies for nonsurgical treatment of heart defects, cardiac transplantation, heart failure and hybrid surgical/catheter procedures. He has a bold vision for Children's Memorial Hermann Hospital and the division of Pediatric Cardiology. Within the next year, he plans to introduce pulmonary valve implants and expand the number of hybrid procedures cardiologists and cardiovascular surgeons perform together. Through further integrated services and developed practice plans, he plans to ensure that the care provided to pediatric cardiology patients is efficient and seamless.

Appointed as Associate Editor of *Ultrasound in Obstetrics and Gynecology*

Helena Gardiner, M.D., Ph.D., co-director, Fetal Cardiology Program at The Fetal Center at Children's Memorial Hermann Hospital has been appointed as an associate editor of *Ultrasound in Obstetrics & Gynecology,* referred to as the White Journal, an official publication of the International Society of Ultrasound in Obstetrics and Gynecology (ISUOG). For more than 10 years, Dr. Gardiner has worked closely with ISUOG as a reviewer of abstracts for the annual meeting and reviewer of manuscripts submitted to the White Journal. In this advanced role, she will oversee the peer review process and joins a team of international physician leaders in ultrasound who will work to determine Journal strategy.

New Staff

ShaJuana Cassimire
Clinic Coordinator
The Fetal Center

Noemi Boring
Research Coordinator
The Fetal Center

Yisel Morales
Research Coordinator
The Fetal Center

Ana Rangel-Duenez
Clinic Coordinator
The Fetal Center

Events

Avery Grace Kargel Golf Tournament, benefiting The Fetal Center

For a second year in a row, The Fetal Center at Children's Memorial Hermann Hospital was selected to be the sole beneficiary of the 9th Annual Avery Grace Kargel Memorial Golf Tournament. Last year, the golf tournament raised more than $15,000 for The Fetal Center.

The Avery Grace Kargel Memorial Golf Tournament is an annual community charity event founded by Heather and Jason Kargel in honor of their late daughter, Avery Grace, who passed away in 2006 as a newborn. The memorial golf tournament was established that same year to raise money and support palliative care programs in the Greater Houston area.

A part of the multidisciplinary team at The Fetal Center, the Chronic and Palliative Services (CAPS) program is a fully integrated palliative care program with dedicated specialties including neonatology, nursing, social work, Child Life specialists and chaplaincy services. The team aims to provide families with honest communication and a comfortable and positive environment to create lasting memories and meet the goals each family has for their final moments with their child. The CAPS Program is part of the continuum of care for patients and families at The Fetal Center that have been diagnosed with a life-threatening or life-limiting condition.

This year's tournament, benefiting The Fetal Center, will be held on Saturday, December 6. 2014. For more information on how to participate in the tournament or to learn about ways to give to The Fetal Center, email us at thefetalcenter@memorialhermann.org.

Save The Date

5TH ANNUAL TEXAS TWO STEP CONFERENCE
NAVIGATING CHANGES IN OB/GYN: ARE YOU ON BOARD?

January 9 – 10, 2015
Hotel Zaza, Houston

The two-day conference will focus on healthcare policy changes impacting all clinical teams in obstetrics and gynecology.

For more information email childrens.events@memorialhermann.org

THE FETAL CENTER'S FRIENDS AND FAMILY REUNION

Saturday, March 7, 2015
Houston Downtown Aquarium

Join us at our first friends and family reunion!

For more information and to provide us with your current mailing address, email thefetalcenter@childrensmemorialhermann.org

Research And Education

EDUCATION

- **Pedro Argoti, M.D.,** completed his fetal intervention fellowship with The Fetal Center and began his fellowship in genetics and maternal-fetal medicine at Johns Hopkins University School of Medicine. Dr. Argoti was the Center's first fetal intervention fellow.
- **Roopali Donepudi, M.D.,** began her fetal intervention fellowship at The Fetal Center at Children's Memorial Hermann Hospital in July. She joins The Fetal Center following completion of her maternal-fetal medicine fellowship at Albert Einstein College of Medicine.
- **Saul Snowise, M.D.,** a second-year maternal-fetal medicine fellow at UTHealth Medical School, joined The Fetal Center at Children's Memorial Hermann Hospital to concentrate on research and clinical training in fetal intervention for the next two years of his fellowship training.

CLINICAL TRIALS

- Ramesha Papanna, M.D., M.P.H., continues to investigate the causes of preterm labor and preterm premature rupture of membranes in twin-twin transfusion syndrome. Basic science investigations are under way to study changes in the fetal membranes and amniotic fluid that occur in TTTS that may contribute to preterm delivery.
- Dr. Papanna, in collaboration with Kenneth Moise Jr., M.D., is continuing an investigation in a swine model to address the sealing of the site of fetoscopic entry at the time of fetal intervention. This work is funded by an RO1 grant from the National Institutes of Health in collaboration with Russell Stewart, Ph.D., of the University of Utah.
- Dr. Papanna and Dr. Moise, in collaboration with Stephen Fletcher, D.O., in the division of Pediatric Neurosurgery at UTHealth Medical School, continue to investigate methods in the development of a minimally invasive approach to in utero repair of myelomeningocele. In collaboration with Scheffer Tseng, M.D., Ph.D., of the Ocular Surface Center, P.A., preliminary results in an ovine model for MMC indicate that the processed outer covering of human umbilical cord has substantial regenerative properties as a patch for in utero repair. Additional experiments are planned for spring 2015.
- Dr. Moise, in collaboration with Judith Smith, Pharm.D., is studying the transplacental passage of remifentanyl in patients undergoing intratuterine transfusion.
- Saul Snowise, M.D., visited Children's Hospital of Philadelphia to learn the methodology of a rat model for fetal myelomeningocele (MMC). This fall, experiments are planned at UTHealth Medical School to study various patch materials that can be utilized for the minimally invasive repair of fetal MMC.
- Dr. Snowise has initiated a prospective trial on fetal thigh volume determined by ultrasound in the prediction of subsequent ambulation in fetuses affected with spina bifida.
- Helena Gardiner, M.D., Ph.D., has secured specialized software for the study of myocardial stress forces in an effort to detect early cardiac changes in monochorionic twins that will eventually develop twin-twin transfusion. Human IRB approval is pending for a prospective trial.

PUBLICATIONS

The *American Journal of Perinatology* will publish a monograph of articles sponsored by The Fetal Center at Children's Memorial Hermann Hospital and the Department of Obstetrics, Gynecology and Reproductive Sciences at UTHealth Medical School. The monograph is a summarization of articles based on presentations by invited speakers at the Complicated Monochorionic Twins workshop, hosted by The Fetal Center in 2013. Kenneth Moise Jr., M.D., serves as the guest editor for the special edition.

PRESENTATIONS AT NATIONAL AND INTERNATIONAL MEETINGS

Several physicians affiliated with The Fetal Center at Children's Memorial Hermann Hospital presented abstracts at the 24th World Congress on Ultrasound in Obstetrics and Gynecology in mid-September, 2014, in Barcelona, Spain.

- **Gardiner HM, Boudreaux D, Moise KJ, Johnson A, Papanna R, Bebbington M.** Are ductus venosus time intervals a practical method to verify early twin-twin transfusion syndrome in monochorionic pregnancies?
- **Gardiner HM**, Kovacevic A, Mellander M, Tulzer G, Roughton M, Öhman A, Dangel JH, Herberg U. Does fetal aortic valvuloplasty alter outcomes in aortic valve stenosis? Results of a retrospective European multinational multicentre study.

- **Gardiner HM**, Kovacevic A, Mellander M, Öhman A, Roughton M, Dangel JH, Mair R, Magee A, Ghez O, Tulzer G, Fetal aortic valvuloplasty: investigating institutional bias in multicentre contributions to studies.
- **Bebbington MW, Moise KJ, Johnson AJ, Papanna R, Tsao K.** Laparoscopic-assisted selective laser photocoagulation therapy (LA-SLPT) for twin-to-twin transfusion syndrome (TTTS).
- **Bebbington MW, Moise KJ, Johnson AJ, Tsao K.** Open fetal surgery for myelomeningocele: outcome in a post MOMs trial cohort.

Members of The Fetal Center presented abstracts at the 33rd International Fetal Medicine and Surgery Society held in Cape Cod in mid-September, 2014.

- Putnam LR, **Johnson A, Moise KJ, Bebbington MW, Papanna R, Garcia EI, Weir JW, Tsao K.** *Generalizability of a randomized-controlled trial: Can we maintain equipoise outside of the Management of Myelomeningocele Study (MOMS) trial?*
- **Gardiner, HM, Boudreaux D, Moise KJ, Johnson A, Papanna R, Bebbington M.** *Can ductus venosus time intervals identify early twin-twin transfusion syndrome in monochorionic pregnancies?*
- **Gardiner HM**, Kovacevic A, Mellander M, Tulzer G, Roughton M, Öhman A, Dangel JH, Herberg U. *Do current fetal aortic valvuloplasty selection criteria predict natural history outcomes? Results from a retrospective European multinational multicentre study.*
- Mann LK, Khan A, Jagannath C, Taegtmeyer, **Moise KJ, Johnson A, Papanna R.** *Amniodilution/amnioinfusion induced autophagy and cell death in amniotic epithelial cells (AEC): A novel mechanism for preterm premature rupture of membranes after fetal surgery.*
- **Papanna R**, Mann LK, Baschat AR, **Bebbington MW, Snowise S**, Khalek N, **Johnson A, Moise KJ**. *Preoperative cervical length below the 20th percentile is an independent indicator of spontaneous preterm birth after fetoscopic laser surgery for twin-twin transfusion syndrome.*
- **Papanna R**, Mann LK, **Bebbington MW**, Khalek N, **Johnson A, Moise KJ**, Baschat A. *Preterm delivery after fetoscopic laser surgery for twin-twin transfusion syndrome: Etiology and risk factors.*
- **Moise KJ, Papanna R**, Mann LM, **Fletcher S, Tsao K**, Argoti P, **Snowise S**, Schniederjan R, Bhattacharjee M, Stewart RJ, Tseng SCG. *Evaluation of different patch materials for in-utero repair of myelomeningocele.*
- **Snowise S**, Argoti P, **Moise K, Johnson A, Bebbington M, Papanna R**. *Fractional limb volume (FLV) of the fetal thigh for intrauterine assessment of neurological deficit of the lower extremities in fetuses affected by spina bifida.*

Members of The Fetal Center have submitted six abstracts for consideration for presentation at the 35th annual meeting of the Society for Maternal-Fetal Medicine scheduled for February 2015 in San Diego.

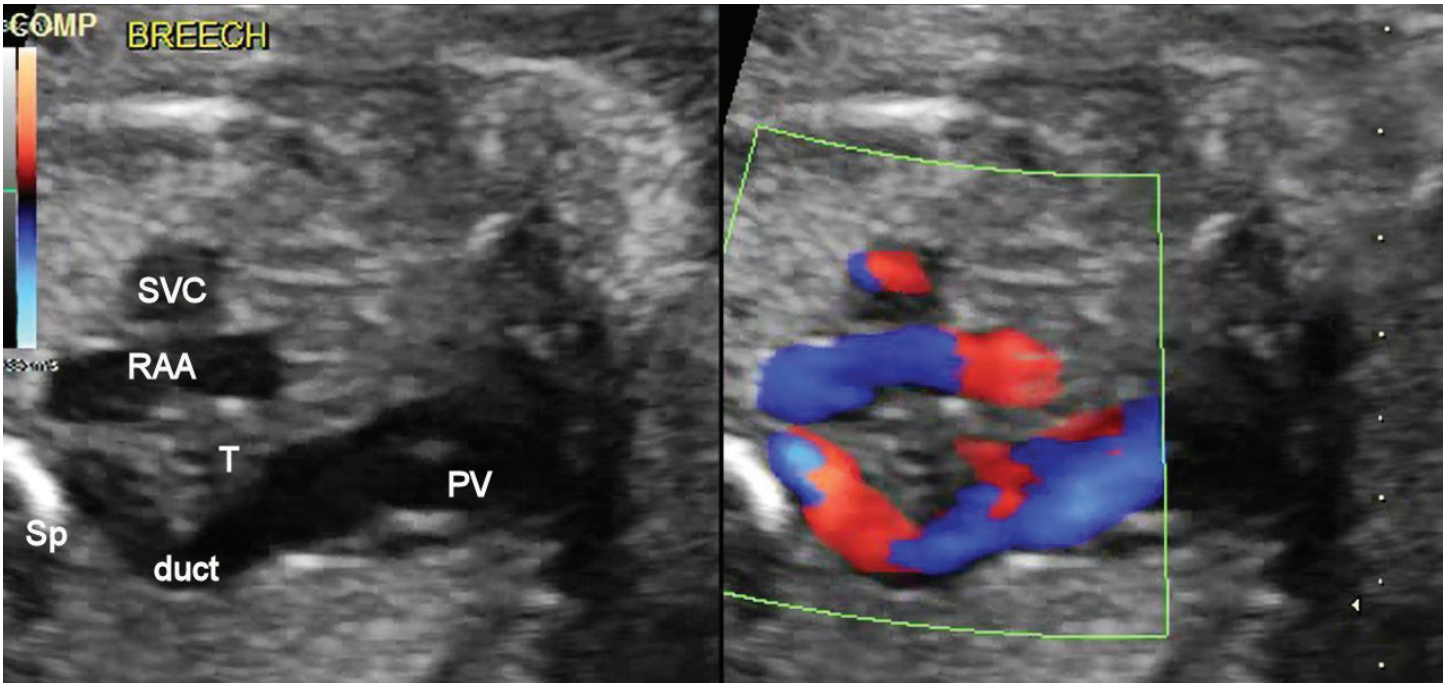
Helena Gardiner, M.D., Ph.D., will be presenting an abstract at the annual meeting of the American Heart Association scheduled for November in Chicago. She will present on behalf of the Association for European Paediatric Cardiology fetal working group. *Does fetal aortic valvuloplasty alter the natural history of aortic valve stenosis?*

INVITATIONS

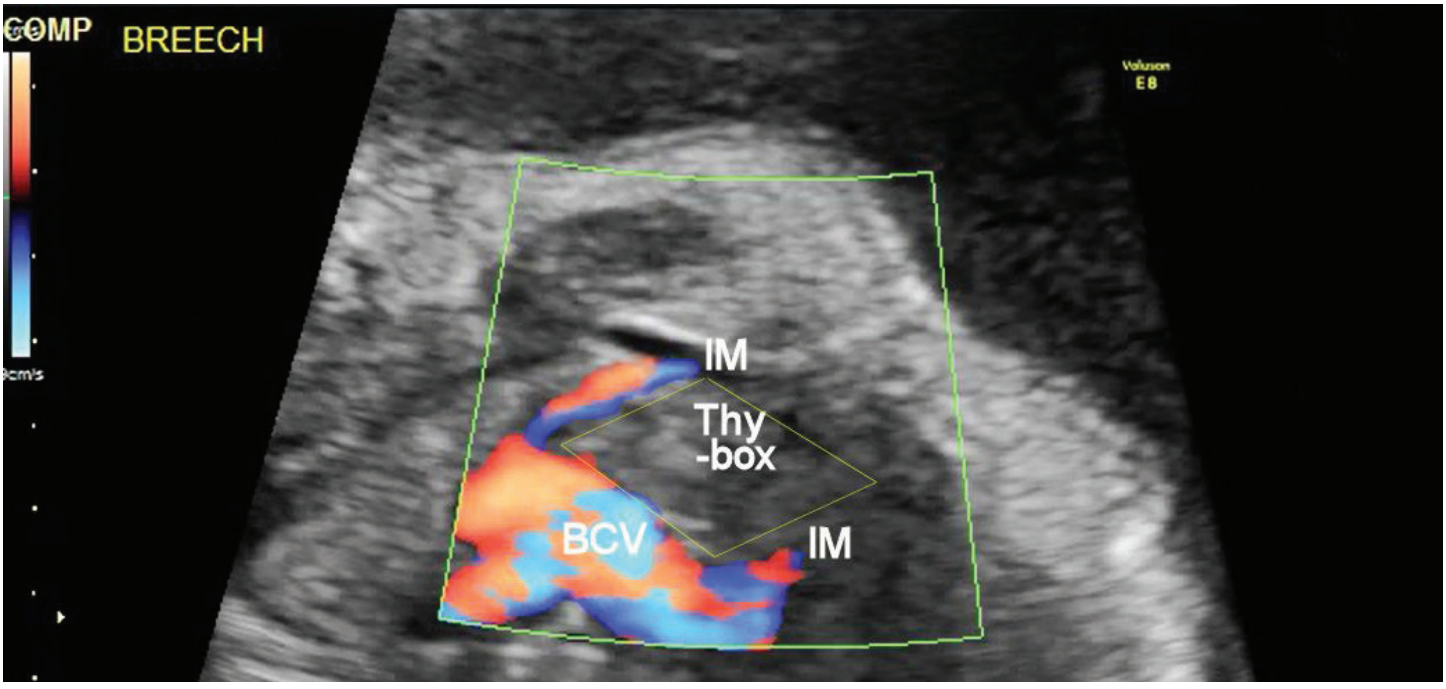
- The Fetal Center at Children's Memorial Hermann Hospital was well represented at the 24th World Congress of Ultrasound in Obstetrics and Gynecology (ISUOG) in September, 2014, in Barcelona, Spain.
  - **Anthony Johnson, D.O.,** chaired the session on ISUOG Outreach: Supporting ultrasound training in the field. He will also chair a session about Multiple Gestations.
  - **Michael Bebbington, M.D., M.H.Sc.,** chaired the oral communication and oral poster presentation sessions on Fetal Therapy. He also served as a content expert in a Meet the Professor session at the ISUOG annual meeting.
  - **Helena Gardiner, M.D., Ph.D.,** chaired the oral communication session on fetal cardiac function and the oral poster session on cardiac function in the assessment of fetal outcome. In addition, she chaired a workshop on cardiac function and will host a Meet the Professor session.
- **Helena Gardiner, M.D., Ph.D.,** is an invited speaker at the 10th Annual Symposium, Advances in Perinatal Cardiology, scheduled for October in St. Petersburg, Florida.
- **Dr. Gardiner, M.D., Ph.D.,** was invited to be the section editor on cardiology for *Ultrasound in Obstetrics and Gynecology.*

Fascinating Fetal Find

A patient was referred to The Fetal Center at Children's Memorial Hermann Hospital for a fetal echocardiogram because of a maternal history of mitral valve prolapse in addition to a family member born with congenital heart disease who died as an infant. The echo confirmed normal abdominal situs and cardiac connections but scanning at the level of the three vessel and tracheal view demonstrated a right-sided aortic arch with an aberrant left subclavian artery and a left-sided arterial duct.



Echo One: The vascular ring is formed by the right-sided aortic arch (RAA) and left-sided ductal arch surrounding the trachea (T). The pulmonary valve (PV), superior vena cava (SVC), and spine (SP) are also identified and used as reference points.



Echo Two: Shown with the aid of color doppler, the internal mammary vessels (IM) at the level of the brachiocephalic vein (BCV) delineating the fetal thymus. This is known as the "thy-box."

These images show the vascular ring formed by the right-sided aortic arch (RAA) and left-sided ductal arch surrounding the trachea (T). There was no prenatal compression of either the trachea or esophagus. We identified the internal mammary vessels (IM) at the level of the brachiocephalic vein (BCV) delineating the fetal thymus. This is known as the "thy-box."

A right-sided aortic arch (RAA) occurs when part of the embryological left arch regresses leaving the right fourth arch or right dorsal aorta. It can be a normal variant and may occur in isolation. More commonly it is associated with cardiac malformations such as tetralogy of Fallot. The most common chromosomal abnormality associated with a right-sided aortic arch is 22q11.2 deletion, or Di George syndrome, reported in 8 percent of fetuses with isolated RAA. The thymus may be small in these babies and thus the demonstration of the 'thy-box' and a normal-sized thymus may lower the risk of a genetic etiology.

Normally both the aortic and ductal arches lie to the left of the trachea. When the ductus arteriosus closes a few days after delivery it leaves a fibrous cord behind. Usually this does not cause any problems; however, when the aortic arch and ductal arch are on different sides of the trachea and esophagus, as in this case, the fibrous cord may cause compression of the trachea and esophagus, resulting in the development of symptoms such as noisy breathing or difficulty swallowing. This can be difficult to diagnose and sometimes is not recognized until a child has had difficulty swallowing solids for several years. In addition, compression may be caused by an aberrant left subclavian artery arising from the descending aorta coursing behind the trachea and esophagus. This is most commonly associated with an isolated RAA and normal cardiac anatomy.

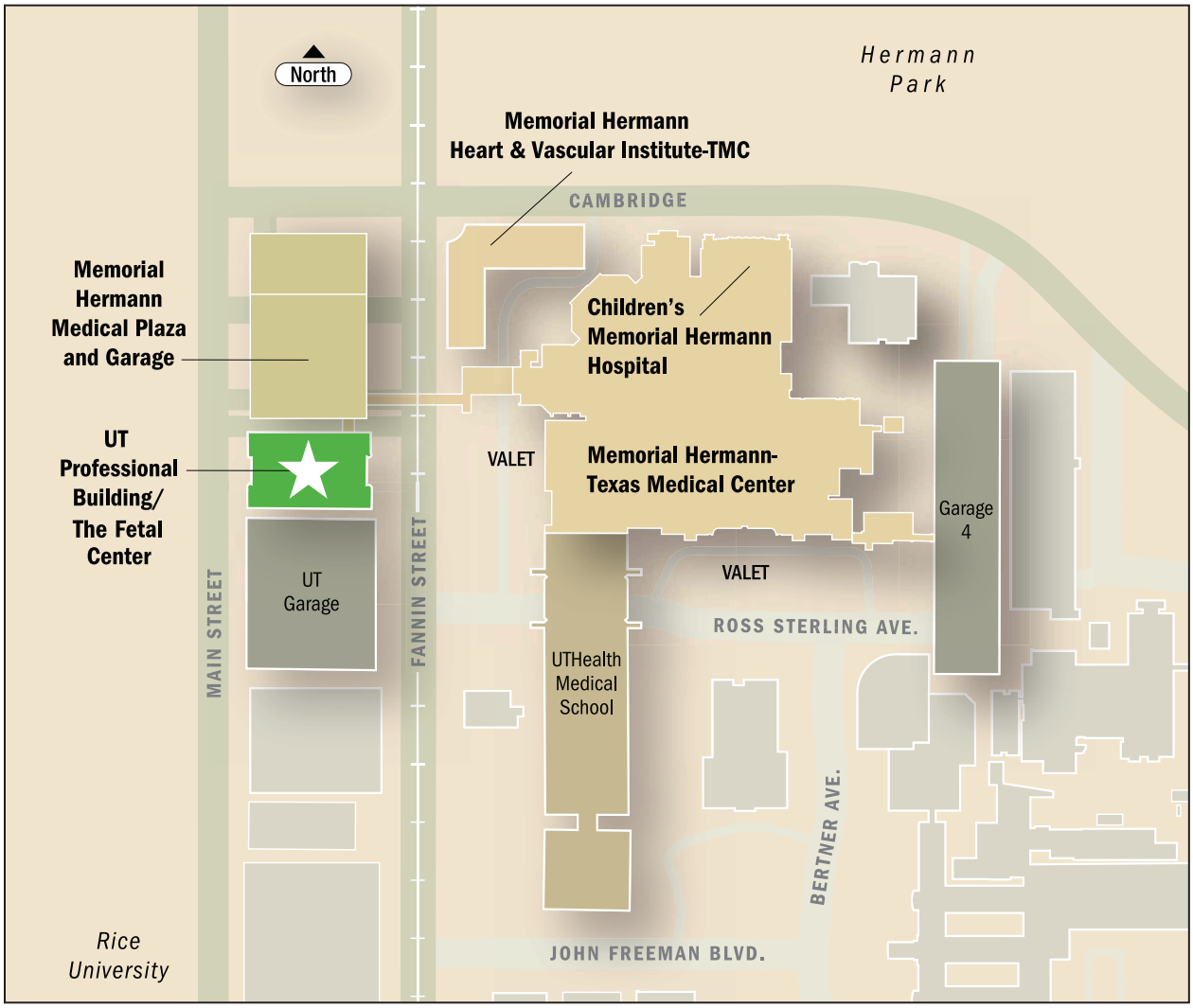
The newborn was clinically stable after delivery without any acute events including stridor or respiratory distress or any feeding issues. A repeat echocardiogram after birth confirmed the prenatal diagnosis and a cardiac MRI was performed when she was 4 days old and confirmed there was a right-sided aortic arch with a four-vessel branching pattern including an aberrant left subclavian artery. The aberrant left subclavian artery arose from the distal arch, coursing posterior to the esophagus and trachea, with mild narrowing of the trachea just above the carina. There was a diverticulum of Kommerell and thyromegaly without a focal lesion. The baby was discharged home and remains well at follow-up at 9 months. The parents understand that surgical intervention may be required in the future if she starts to develop symptoms of tracheal or esophageal compression.

The Fetal Center at Children's Memorial Hermann Hospital, through the Congenital Heart Defect Screening Program, is advocating and training physicians and sonographers to routinely evaluate the three vessel and tracheal view as part of the second trimester anatomy review. This will allow for improved identification of fetuses at risk for developing a vascular ring. Prenatally we counsel parents and pediatricians about the risks associated with a vascular ring, thus avoiding a delay in diagnosis and associated morbidity for the child. The diagnosis of a vascular ring is best confirmed after delivery by MRI and most children will become symptomatic and require surveillance.

At Children's Memorial Hermann Hospital, Michael Hines, M.D., professor of pediatric cardiovascular surgery at UTHealth Medical School, performs thoracoscopic division of the vascular ring for patients requiring surgical intervention. This minimally invasive approach releases the tracheal and/or esophageal compression and avoids the need for more complicated surgery. The benefits are immediately obvious to the family.

Visit childrensmemorialhermann.org/thefetalcenter for more information or to request a second opinion.

Contacts



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