

# THE FETAL CENTER NEWSLETTER

Winter 2015 • 8th 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

Help us continue to provide the highest level of specialized care to both mothers and babies and support the groundbreaking work of The Fetal Center.

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## Research Update: Toward a Safe Method of Minimally Invasive Myelomeningocele Repair



Since the 1930s, the first step in the treatment of newborns with myelomeningocele has been to surgically close the incompletely developed portion of the spinal cord within a few days of birth. In 2011, the Management of Myelomeningocele Study (MOMS), sponsored by the National Institutes of

Health (NIH), found that infants who undergo surgery in utero had a decreased need for ventriculoperitoneal shunting and improved motor function when compared to the standard-of-care procedure performed after birth.<sup>1</sup> After the publication of study results in March of that year, a team affiliated with The Fetal Center at Children's Memorial Hermann Hospital was among the first in the country to perform in-utero repair – with excellent outcomes. Today, researchers at The Fetal Center and UTHealth Medical School are engaged in laboratory studies aimed at developing an approach to minimally invasive spina bifida repair that is safe for both mother and child.

“Surgeons in several countries have performed cases of minimally invasive repair, many of which have been announced in the public sector before publication in peer-reviewed journals,” says Kenneth J. Moise Jr., M.D., co-director of The Fetal Center and a professor in the division of Maternal-Fetal Medicine and department of Pediatric Surgery at UTHealth Medical School. “Our concern with minimally invasive spina bifida repair is that fetal surgeons are using techniques and materials that have not been subjected to rigorous scientific testing. We believe strongly in the bench-to-bedside process – repeated studies in animal models, followed by human pilot trials, then randomized clinical trials, analysis of data and peer review before and after publication. Based on the results of our research, we're optimistic about the future of minimally invasive repair, but until we can show data to prove that this new approach is better, open surgical repair for spina bifida will remain the gold standard. If I were to perform a minimally invasive spina bifida repair on a fetus today, based on the data available, I'd have to tell the mother that yes, this is definitely better for you, but I can't predict your child's outcome.”

Investigators at Vanderbilt University were the first to attempt in-utero repair of myelomeningocele (MMC) in human fetuses using endoscopy. The surgeons used a maternal skin graft in an attempt to cover the defect using laparoscopic instrumentation inserted into the uterus. When the procedure resulted in high perinatal mortality, no further attempts at minimally invasive repair were made, and a new approach using an open hysterotomy was developed. After three fetuses underwent standard surgical repair and only one of the three required a ventriculoperitoneal shunt, the researchers concluded that open fetal surgery for spina bifida repair was feasible.

*continued*

## Fetal Spina Bifida Update

After the success of these cases and 79 others, four U.S. centers began offering fetal repair of myelomeningocele. In January 1999, investigators at one of the centers – the University of California, San Francisco (UCSF) – applied to the National Institutes of Health for a single-center study of in-utero MMC repair. A consensus conference on fetal myelomeningocele repair, hosted by the National Institute of Child Health and Human Development, led to a recommendation for a multicenter randomized trial.

Physicians affiliated with The Fetal Center at Children’s Memorial Hermann Hospital were moving forward aggressively with a fetal surgery program for spina bifida when the MOMS trial was announced by the NIH in early 2003. When three centers – UCSF, Vanderbilt University Medical Center and Children’s Hospital of Philadelphia – were chosen for the trial, The Fetal Center complied by stopping its program to avoid creating a “back door” that could affect enrollment in the trial. After the MOMS trial showed decreased need for shunting at 12 months and ambulation without assistance by age 2 1/2, open fetal spina bifida repair was adopted as the gold standard by which outcomes from all newer techniques would be measured.

Thomas Kohl, M.D., of the German Center for Fetal Surgery and Minimally Invasive Therapy at the University of Giessen-Marburg in Giessen, Germany, is among the few fetal surgeons who have published the results of their attempts at minimally invasive MMC repair. In a two-part article published in *Ultrasound in Obstetrics and Gynecology* in 2014,<sup>2,3</sup> he and other authors reported on 51 cases in which three trocars were inserted percutaneously into the amniotic cavity using ultrasound guidance. After insertion of the trocars, the amniotic fluid was partially evacuated and replaced with carbon dioxide to allow for improved fetal visualization and manipulation. Following the procedure, the gas was removed and the amniotic cavity refilled with warmed crystalline solution.

“One of our concerns about this procedure is that the effect of carbon dioxide on the fetus remains unknown,” Dr. Moise says. “We know that the fetus lacks an enzyme that allows it to process carbon dioxide. In a sheep model, fetuses subjected to CO<sup>2</sup> became acidotic.<sup>4</sup> We also haven’t adequately studied the effects of the gas on fetal membranes. Will it damage their integrity? We simply don’t know.”

There’s also the matter of the patch used to repair the defect. In research led by UTHealth Medical School maternal-fetal medicine specialist Ramesha Papanna, M.D., and presented at the 33rd Annual Meeting of the International Fetal Medicine and Surgery Society in Chatham, Massachusetts, Dr. Moise and his collaborators analyzed two materials that could be employed in a patch technique for minimally invasive fetal MMC repair.<sup>5</sup> Using a sheep model, they compared a biocellose patch (Dermafill®) attached by an underwater adhesive and a human umbilical cord graft (Amniogard®) secured to the defect with a continuous suture. When necropsy showed that the Amniogard patch remained in place in all fetuses, they concluded that the umbilical cord graft showed promise as an optimal material for minimally invasive correction of fetal MMC.

“This is a first step toward a safe minimally invasive repair. Our next step will be to apply the patch using a fetoscope in the sheep model,” Dr. Moise says. “When we’re comfortable with our results, we’ll move to a human pilot study.”

Pediatric neurosurgeon Stephen Fletcher, D.O., an associate professor in the department of Pediatric Surgery with 31 years of experience performing postnatal spina bifida repair, believes the human umbilical cord graft may also be a suitable patch material for neonatal repair. “We currently use bovine pericardium lining for the patch in postnatal repair,” he says. “We’re discovering that Amniogard is a tremendous patch that almost replicates the normal anatomy.”

The researchers are working with Russell Stewart, Ph.D., of the University of Utah department of Bioengineering to develop a waterproof patch sealant derived from the sandcastle worm. “We think we’ll be able to use the sealant while operating inside the amniotic sac, eliminating the need to evacuate the amniotic fluid and replace it with carbon dioxide,” Dr. Moise says. “This is an exciting development for the future, but we’re not there yet. We need more data to ensure its safety. Our upcoming series of sheep studies has never been done before; we’ll place the patch fetoscopically and deliver the lambs to determine if they can walk.”

The researchers’ long-term goal is to reduce the risks associated with open spina bifida repair. “We know open repair presents all the risks to the mother involved with a uterine incision, including preterm delivery,” Dr. Moise says. “Although no data equivalent to the MOMS trial outcomes has been presented showing that the minimally invasive approach produces equivalent results, in my heart I believe it will. The challenge lies in how to get there. We’re pushing the envelope but doing it in a scientific fashion. I’m as cautious as I was when we first started doing open fetal myelomeningocele repair. We’re still working on the next chapter in the story.”

<sup>1</sup>Adzick NS, Thom EA, Spong CY, Brock JW III, Burrows PK, Johnson MP, Howell LJ, Farrell JA, Dabrowiak ME, Sutton LN, Gupta N, Tulipan NB, D’Alton ME and Farmer DL for the MOMS Investigators. A Randomized Trial of Prenatal versus Postnatal Repair of Myelomeningocele. *N Engl J Med.* 2011 Mar 17;364:993-1004.

## Fetal Spina Bifida Update

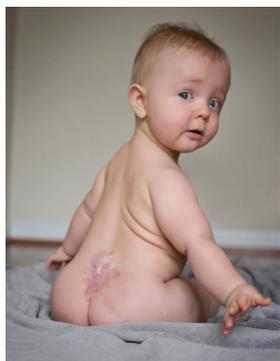
<sup>2</sup>Kohl T. Percutaneous minimally invasive fetoscopic surgery for spina bifida aperta. Part I: surgical technique and perioperative outcome. *Ultrasound Obstet Gynecol.* 2014;44:515-24.

<sup>3</sup>Degenhardt J, Schürg R, Winarno A, Oehmke F, Khaleeva A, Kawecki A, Enzensberger C, Tinneberg H-R, Faas D, Ehrhardt H, Axt-Fliedner R, Kohl T. Percutaneous minimal-access fetoscopic surgery for spina bifida aperta. Part II: maternal management and outcome. *Ultrasound Obstet Gynecol.* 2014;44:525-31.

<sup>4</sup>Gratacos E, Wu J, Devlieger R, Van de Velde M, Deprest JA. Effects of amniodistention with carbon dioxide on fetal acid-base status during fetoscopic surgery in a sheep model. *Surg Endosc.* 2001;15:368-72.

<sup>5</sup>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. Presented at the 33rd Annual Meeting of the International Fetal Medicine and Surgery Society, Chatham, Massachusetts, Sept. 2014.

### Beyond the MOMS Trial: Four Years of Successful Outcomes in Fetal Repair of Myelomeningocele



[Left to right] Duke and Faith, both had prenatal surgery performed at The Fetal Center to repair their spina bifida defect.

On March 17, 2011, the *New England Journal of Medicine* published the results of the National Institutes of Health-sponsored Management of Myelomeningocele Study (MOMS) comparing open in-utero repair of myelomeningocele to the traditional postnatal repair.<sup>1</sup> When the study found that fetuses treated in utero had improved motor function and a decreased need for ventriculoperitoneal shunting, pediatric surgeon KuoJen Tsao, M.D., and his colleagues at The Fetal Center at Children's Memorial Hermann Hospital began screening patients for in-utero repair of the defect. Among the families referred to the Center were Dallas residents Ivan and Colette Hagler. Based on MOMS trial protocols, Dr. Tsao determined that Hagler was an ideal candidate for the procedure, and her daughter Faith became the first patient in Texas to undergo in-utero repair of myelomeningocele (MMC).

Today, Dr. Tsao, who is co-director of The Fetal Center, calls Faith Hagler his "poster child" for the procedure. Delivered on July 4, 2012, Faith has met all of her developmental milestones. "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."

The four-year-old program at The Fetal Center continues to produce good outcomes with patients chosen based on criteria established in the MOMS trial. "We've been very strict in maintaining the same selection criteria," says Dr. Tsao, an associate professor in the department of Pediatric Surgery and department of Obstetrics, Gynecology and Reproductive Sciences. "As a member of the North American Fetal Treatment Network (NAFTNet), we've vowed to maintain our adherence to the best practices they've established. If any fetal spina bifida surgery varies from the MOMS criteria, it should be done under a research protocol."

The selection process for in-utero MMC repair is based on a strict prenatal care algorithm (see page 5) that emphasizes education. "Patients come to us with one of two mindsets," Dr. Tsao says. "Many have been given a diagnosis of spina bifida and have done their homework and want to know if they qualify for fetal surgery. Other families know very little about the defect and are overwhelmed by the diagnosis. They want to learn about the condition and fetal surgery, but the last thing they want to do is wade through websites with detailed medical descriptions. Because we view it as our obligation to teach families about the condition itself – not just fetal surgery – we approach both types of patients in the same way, preparing them to make an informed decision about their treatment options. We've made an impact on lives through fetal surgery but we feel our major impact has been in the area of spina bifida education as a consequence of the availability of surgery."

As part of the education process, each patient referred to The Fetal Center, regardless of whether she and her baby qualify for fetal MMC repair, undergoes extensive counseling with experts in maternal-fetal surgery, fetal surgery and spina bifida. Over the span of two days, the family also meets with a pediatric surgeon, a pediatric neurosurgeon, a genetic counselor, a pediatric expert in long-term spina bifida outcomes, a neonatologist, an anesthesiologist, a social worker and a representative of Child Life services.

## Fetal Spina Bifida Update

Pediatric spina bifida specialist Lynette Mazur, M.D., professor of pediatrics at UTHealth Medical School, is a key member of the multidisciplinary team that provides long-term care for children with myelomeningocele. An expert with more than 10 years of experience, she uses a specially designed doll to help parents understand the defect and complications their child may have based on the location of the lesion.

“Dr. Mazur’s consultation is incredibly important to the process because it’s here that parents can find answers to their foremost question: ‘What will happen to my baby after birth?’” Dr. Tsao says. “Although it’s never a completely clear-cut answer, she is able to provide information related to possible complications and what parents can expect in terms of the care their child will need throughout life. Some parents who come to us for fetal surgery change their minds, even if they qualify, and choose postnatal repair after their consultation because of the education provided.”

Candidates are accepted for open fetal MMC surgery based on the criteria set forth by the MOMS trial. The risks and benefits to both mother and baby are considered in the process. Specific factors that exclude the fetus from surgery are variants of spina bifida not considered open neural tube defects, the presence of a significant fetal condition not related to myelomeningocele, kyphosis, multiple pregnancy, significant clubbing of the legs or evidence of existing paralysis. Maternal exclusion criteria include medical or personal reasons for withholding surgery or anesthesia, morbid obesity, previous or planned incision on the cervix or a documented history of a weak cervix, a cervix of less than 20 millimeters by vaginal ultrasound, preterm labor in the current pregnancy, a history of spontaneous preterm delivery in previous pregnancies, bleeding or placental abruption, red cell or platelet alloimmunization, insulin-dependent diabetes prior to pregnancy, abnormal anatomy of the uterus such as uterine fibroids, infection with HIV or hepatitis B or C, inability to adopt a lifestyle change of restrictive activity during the remaining portion of the pregnancy, and inability to travel to The Fetal Center or to comply with follow-up care requirements.

Early diagnosis of spina bifida is important. “Because we have a small window for fetal MMC repair, we like to see mothers at 22 or 23 weeks gestation, which allows them time to complete the consultation and go home and consider their decision,” Dr. Tsao says. “We don’t perform fetal repair past 26 weeks. Once a mother decides on fetal surgery, it’s a lifestyle change. If the family is from out of town, they’re in our city for two to three weeks after a five-day hospital stay. If there’s any complication, they may stay in the hospital for the duration of the pregnancy. In addition to the Greater Houston area, our patients come from across Texas and surrounding states. Recently, we’ve had patients from as far away as Hawaii, North Dakota and the East Coast.”

Dr. Tsao also credits the pediatric neurosurgery team for the growth of The Fetal Center’s spina bifida program. Stephen Fletcher, D.O., has been doing postnatal MMC repair at Children’s Memorial Hermann Hospital for the past 31 years and performed his first fetal repair on Faith Hagler in 2011. “Faith nearly gave me a heart attack preparing for and performing that first fetal surgery, and now look at her,” Dr. Fletcher says. “She’s doing great and didn’t need a shunt. She’s the phenomenon we hoped for when we started the fetal spina bifida surgery program.”

Two new faculty members have joined the pediatric neurosurgery team in the last two years. David I. Sandberg, M.D., FAANS, FACS, FAAP, an expert in minimally invasive endoscopic approaches to hydrocephalus, is director of pediatric neurosurgery at Children’s Memorial Hermann Hospital and an associate professor in the Vivian L. Smith Department of Neurosurgery and the department of Pediatric Surgery at UTHealth Medical School. Dr. Sandberg joined Children’s Memorial Hermann Hospital and UTHealth Medical School from Miami Children’s Hospital and the University of Miami Miller School of Medicine in 2014.

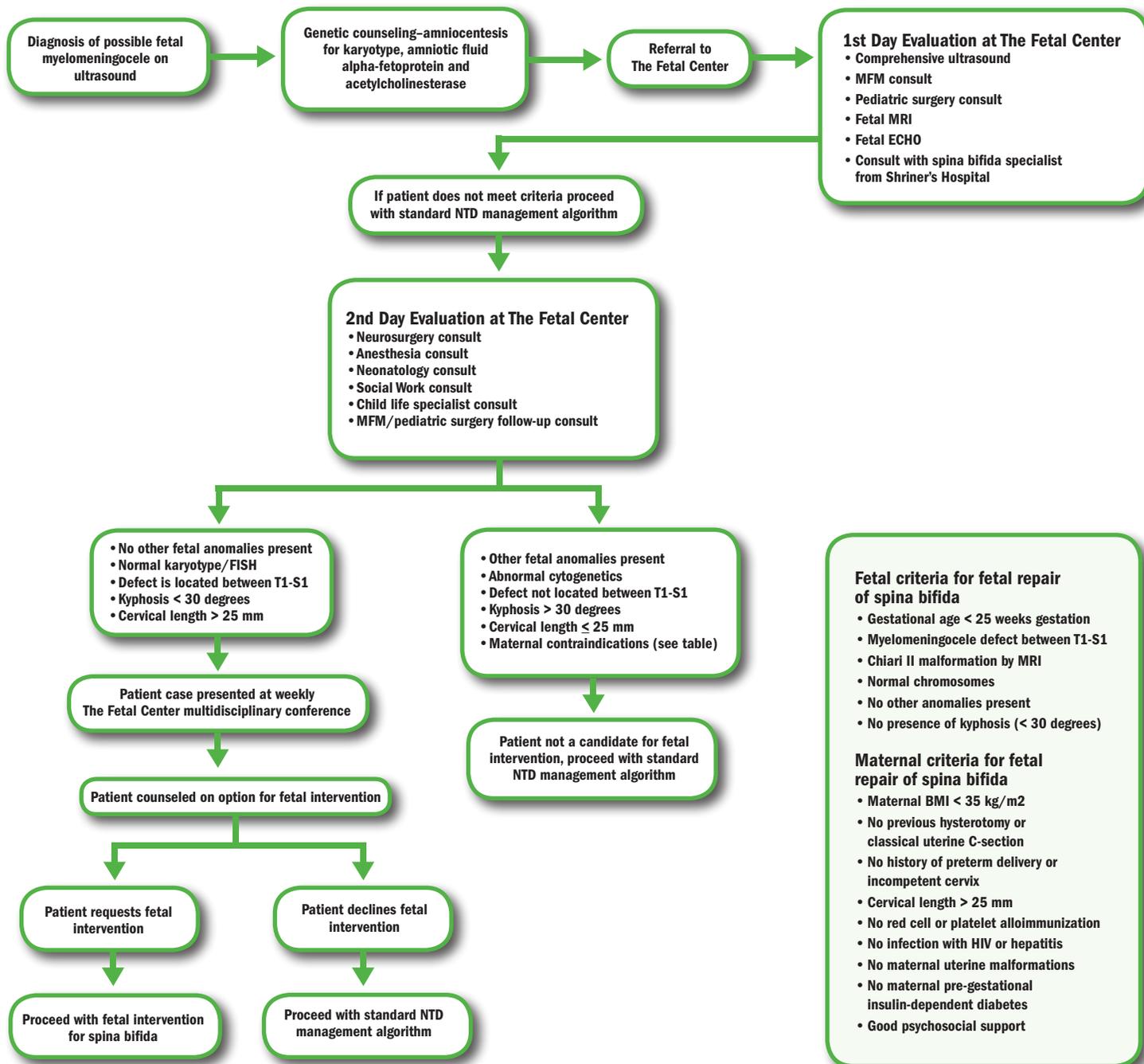
Manish Shah, M.D., who is fellowship trained in pediatric neurosurgery with special expertise in pediatric epilepsy, craniofacial surgery and craniocervical spine surgery, was recruited from Washington University in St. Louis after completing his fellowship at St. Louis Children’s Hospital under world-renowned pediatric neurosurgeon Tae Sung Park, M.D.

“Although a diagnosis of spina bifida is very difficult for parents, in the matter of decision-making we have yet to meet a mom who won’t say ‘Do what you need to do for my baby,’” Dr. Tsao says. “We always advise her to think about herself and her future pregnancies. The mother is our first patient and her fetus is the second, but they’re both our patients. We consider them equally important.”

<sup>1</sup>Adzick NS, Thom EA, Spong CY, Brock JW III, Burrows PK, Johnson MP, Howell LJ, Farrell JA, Dabrowiak ME, Sutton LN, Gupta N, Tulipan NB, D’Alton ME and Farmer DL for the MOMS Investigators. A Randomized Trial of Prenatal versus Postnatal Repair of Myelomeningocele. *N Engl J Med.* 2011 Mar 17;364:993-1004.

# Patient Care Resources

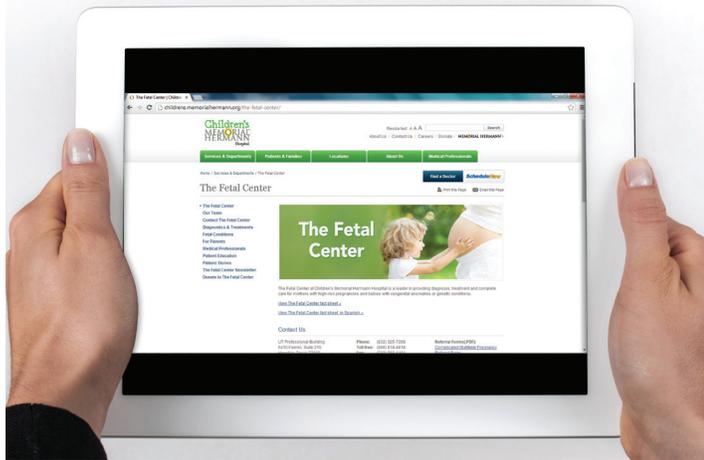
## The Fetal Center Fetal Intervention for Spina Bifida Prenatal Care Algorithm



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.

## Patient Care Resources

### Educational Video Empowers Patients Prenatally Diagnosed with Spina Bifida



The Fetal Center is pleased to announce the new fetal surgery for spina bifida patient educational video. The video serves as a guide for patients and their families faced with the prenatal diagnosis of spina bifida as they explore their treatment options. The animated feature takes patients through the decision-making process by providing information on the etiology and natural history of spina bifida as well as the pathophysiology leading to the many complications associated with the condition. In addition, an extensive breakdown of the MOMS (Management of Myelomeningocele Study) is provided in a patient-friendly format in order to understand the key principles and outcomes associated with the randomized trial. Families are provided with a step-by-step guide through the comprehensive, multidisciplinary evaluation and consultation process followed by a detailed look at the fetal surgery patient experience.

“Often, when we meet patients and their families for the first time, they have a limited knowledge base of spina bifida,” said KuoJen Tsao M.D., co-director of The Fetal Center, “We believe that it is our responsibility to educate our patients about spina bifida first, and fetal surgery second. Patients cannot make a fully informed decision about their treatment options if they don’t have the fundamental knowledge of their condition.”

The video is intended to be utilized as an adjunct to The Fetal Center’s two-day consultation by providing objective information about spina bifida and fetal surgery. Patients are asked to view the online video prior to their initial consultation at The Fetal Center. By reducing information gaps and proactively addressing preconceived notions, we anticipate patients will begin with a better knowledge base and be able to ask more in-depth questions when meeting with the affiliated spina bifida fetal surgery team at The Fetal Center.

To view the educational video, please visit [childrensmemorialhermann.org/spina-bifida](http://childrensmemorialhermann.org/spina-bifida).

### The Fetal Center’s Patient Resource Fund

Families with a prenatal diagnosis of a fetal anomaly or genetic abnormality are faced with tremendous physical and emotional challenges. At a time when patients should focus on treatment and healing, some are challenged by the need to travel to the Center due to financial limitations. To ease the burden, The Fetal Center has established the Patient Resource Fund, available to patients in need of assistance for travel and accommodations during their treatment at The Fetal Center.

For more information about eligibility and assistance, contact us at 832.325.7288. It is our mission to provide every family with compassionate care and provide the best treatment options available to them.

For more information on how to support The Fetal Center’s Patient Resource Fund, contact the Memorial Hermann Foundation at 713.242.4409.

## Patient Story

### Patient Education Empowers Family



Sophia with her older sisters.

Sophia, prenatally diagnosed with spina bifida, is called the family's "miracle baby" because she is developing and doing everything her family was afraid she would never be able to do.

A maternal-fetal medicine specialist in Memphis, Tenn., where the family lives, referred Rebecca and her husband, Jeremy, to The Fetal Center at Children's Memorial Hermann Hospital in January 2013.

Rebecca and Jeremy knew they had a lot to learn about the diagnosis of open neural tube defect, or myelomeningocele. Also known as spina bifida, this major birth defect occurs when a fetal spine fails to fully form during early pregnancy. It can develop anywhere along the spine if the neural tube does not close all the way. One of the most common birth defects in the United States, spina bifida occurs in approximately one in every 1,500 live births.

"Coming to The Fetal Center was the best decision we made," she explains. "I cannot overestimate the value of the compassion and patient education we received."

### Compassionate, multidisciplinary care makes difference

Patient education is at the core of the consultations provided to patients at The Fetal Center. With affiliated physicians from the faculty of UTHealth Medical School, the Fetal Center is dedicated to providing comprehensive, multidisciplinary consultations to educate families about their fetal diagnosis. Consultations provide the family with indications based on the exact location of their child's spina bifida lesion and an assessment on the severity of the disease. In addition, families are provided information on available treatment options so they can make the best decisions for their unique situation. Even before the initial consultation, educational materials, including an online video, are available to families.

When a patient is referred to The Fetal Center, the affiliated team carefully evaluates that patient as a potential candidate for open fetal repair. All patients, regardless of whether they qualify for fetal surgery, undergo extensive multi-day counseling with the multidisciplinary experts in fetal surgery and spina bifida. During a two-day period, patients meet with the entire affiliated fetal spina bifida team, including specialists in maternal-fetal medicine, fetal surgery, pediatric neurosurgery, long-term spina bifida outcomes, neonatology, anesthesiology, social work and Child Life services.

"When I arrived at The Fetal Center, they gave me a book where I could make notes during our discussions with all of the team members," Rebecca says. "I still have it and, believe it or not, I still read it."

Rebecca appreciates the personal attention they had during their three days at the Fetal Center. "Each team member took so much time with us and we were made to feel that there was no such thing as a dumb question." Rebecca and Jeremy learned about treatment options, what to expect throughout their child's lifetime, and the impact the diagnosis may have on other children in the family.

Rebecca praises Jessica Weir, RN, clinical nurse coordinator for the spina bifida program. "She was our go-to person at The Fetal Center and she made our life so easy while we were there. I never felt like we were alone in this process. Jessica and the team at The Fetal Center provided so much support to us. I'm not sure we would have gotten through this somewhere else. We were not just patients; I believe they truly wanted what is best for us." Rebecca was identified as a candidate for open fetal surgery but after all of the evaluations, consultations and education provided she and her husband felt most comfortable with their decision to wait and have the surgery performed after Sophia was born. "I was not only very prepared but I had peace of mind about my decision," says Rebecca.

## Patient Story

“When a patient comes to The Fetal Center, our first priority is to educate the family about the condition and its severity,” says Kuojen Tsao M.D., co-director of The Fetal Center. “If they decide that post-natal repair, instead of fetal surgery, is the best option for their family because they have a clear understanding of what life will be like raising a child with spina bifida, then we’ve done our job.”

Sophia was delivered by a C-section on April 15, 2013, “when I was 38 weeks and one day pregnant,” notes Rebecca. “The spina bifida repair surgery was performed on Sophia the next day and we were amazed that 14 days later we could take our darling little girl home to Memphis.”

“Sophia has exceeded everyone’s expectations, going above and beyond what was expected,” adds Rebecca. “We just start laughing. She laughs at herself all the time and then she claps for herself because she thinks she is funny, too!” Sophia is walking, both with and without a lightweight ankle brace support. “The process is life-changing but it is good to know that spina bifida can be managed – with the right knowledge and professional guidance,” she adds.

### Double the Birthday Celebrations for Spina Bifida Patient



Duke celebrating his 1st birthday.

Kelly received another wonderful birthday present this year. She shares her birthday with her precious son, Duke, and the day he was born was a special gift. “A year later we couldn’t be more thankful at the joy he brings and he is a true blessing,” says Kelly, who looks forward to many more double celebrations.

Kelly and her husband, Chad, count themselves very lucky. The diagnosis of an open neural tube defect, or myelomeningocele, led them on a journey of education, referral, consultations and, ultimately, to open fetal surgery for spina bifida repair.

Spina bifida – a major birth defect of the spine – occurs when a fetal spine fails to fully form during early pregnancy and is associated with many problems that persist throughout life. Spina bifida occurs in approximately one in every 1,500 live births and is one of the most common birth defects in the United States.

When Kelly became pregnant, the family was living in Hawaii where Chad was stationed in the U.S. Army. After the diagnosis, the couple researched options and learned as much as they could. In the process, they met Benjamin Kase M.D., former maternal-fetal medicine fellow at UTHealth Medical School, who had recently moved to Hawaii. Dr. Kase consulted with the family and suggested they contact the specialists at The Fetal Center at Children’s Memorial Hermann Hospital.

“The first person we met when we arrived at The Fetal Center was a sonographer affiliated with the team and she became our hero,” says Kelly. “She performed our first ultrasound there and her professionalism and upbeat spirit gave us instant hope. She had a tremendous positive impact on us.”

The Fetal Center, along with affiliated physicians from the faculty of UTHealth Medical School, is committed to educating parents with detailed information about their unborn child’s diagnosis. The first priority is to make an accurate diagnosis of the condition and assess the degree of severity. Patients are informed about the exact location of the spina bifida lesion and its associated consequences as well as available treatment options, empowering them with the information they need to make the best decision for their baby and family. To educate families about their fetal diagnosis, the specialized team provides comprehensive, multidisciplinary consultations, enhanced by helpful educational materials.

When a patient, such as Kelly, is referred to The Fetal Center, the team carefully evaluates her as a pregnancy with spina bifida and then as a potential candidate for open fetal repair. All patients have the benefit of extensive two-day counseling with multidisciplinary experts in fetal surgery and spina bifida. The team includes specialists in maternal-fetal medicine, pediatric surgery, pediatric neurosurgery, long-term spina bifida outcomes, neonatology, anesthesiology, social work and Child Life services.

## Patient Story

Based on the MOMS clinical trial, funded by the National Institutes of Health, open fetal surgery for spina bifida is now an alternative treatment option for some patients and families. In 2012, surgeons at Children's Memorial Hermann Hospital were the first in Texas to conduct open fetal surgery for spina bifida repair.

The Fetal Center now has outcomes of the first group of patients to reach the milestone of their first birthday. "We are pleased that our outcomes closely match those of the MOMS clinical trial," adds Michael Bebbington, M.D., director of prenatal imaging and fetal diagnosis at UTHealth Medical School. Infants born with spina bifida are at risk for a range of disorders, such as hydrocephalus, which may require a shunt to relieve pressure inside the skull. Being able to avoid shunt placement by 1 year of age was one of the benefits shown by the MOMS trial.

After Kelly was evaluated by The Fetal Center's affiliated multidisciplinary team, the lead fetal surgeon on her case, Dr. Bebbington, "assured us that I was a good candidate and his confidence and knowledge were reassuring," she notes.

Kelly underwent surgery in August 2013 and she describes her open fetal surgery as nothing short of a miracle. She stayed in Houston on modified bed rest until Duke was born via C-section when Kelly was 37 ½ weeks pregnant. Duke's arrival on November 13, 2013, ensured her birthday was extra special.

The parents were overjoyed to bring baby Duke Alexander home a few days before Thanksgiving. "We are forever grateful to Dr. Bebbington and the whole team at The Fetal Center. Because of everyone there, our Duke is defeating all the odds."

"The value of the surgery is huge," adds Duke's father, Chad, "and the benefits for Duke are tangible. Every week we see progress with his brain development and his competency levels," explains Chad. "Duke is doing great, all the signs point in the right direction and the doctors tell us he will not need a shunt." Today, Duke meets weekly with a physical therapist and is making progress in sitting up for longer periods of time and strengthening his leg muscles.

Kelly and Chad's humility and strength shine: "Never be ashamed of a scar. It simply means you were stronger than whatever tried to hurt you."

## EVENTS



### THE FETAL CENTER'S FRIENDS AND FAMILY REUNION

**Saturday, April 11, 2015**

Houston Downtown Aquarium | 410 Bagby Street, Houston, TX 77002

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](mailto:thefetalcenter@childrensmemorialhermann.org)

## News of Note

### Helena Gardiner, M.D., Ph.D., Honored with The Outstanding Research Award in Pediatric Cardiology



The Council on Cardiovascular Disease in the Young presented The Outstanding Research Award to Helena Gardiner, M.D., Ph.D., co-director of the Fetal Cardiology Program at The Fetal Center. This award recognizes innovative and outstanding research that results in fundamental insights in the pathophysiology, etiology, treatment or outcome of cardiovascular disease into young people.

Dr. Gardiner was presented the award as senior author at the American Heart Association (AHA) Scientific Sessions 2014 in Chicago, on behalf of the Fetal Working Group of the European Pediatric Cardiology Society (AEPC), following her abstract entitled, “Does fetal aortic valvuloplasty alter the natural history of aortic stenosis?”

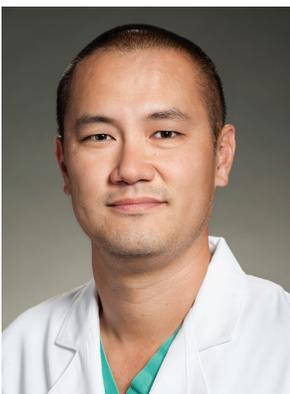
The study enrolled 214 fetuses diagnosed with aortic stenosis at medical centers in 13 European countries. Gardiner and colleagues considered fetal aortic valvuloplasty in 70 of the 214 fetuses, and the procedure was successful in 59 of the 67 accepted for therapy. The researchers analyzed the data by matching the live-born fetuses with aortic stenosis treated by aortic valvuloplasty with natural history controls as closely as possible at 23 weeks gestation for the size of left-sided heart structures and blood flow patterns in the aortic arch.

Survival was similar for both cohorts at 30 days, one year, and four years of follow up. Serial left-sided growth was similar in both cohorts. There was no significant difference in outcome between the cohorts; a similar proportion of survivors had a biventricular circulation in the valvuloplasty-treated cohort and the natural history cohort. Funnel plots showed more survivors with a biventricular circulation among fetuses treated in centers with experience of performing valvuloplasty, while in centers with limited surgical options, more fetuses were converted from biventricular circulation to univentricular circulation, with increased morbidity and mortality.

“The long-term clinical significance of our findings is that fetal valvuloplasty can make a difference for some but not all fetuses with aortic stenosis. It gives these fetuses a chance to have a life with a two-ventricle circulation after they are born, instead of the restrictions associated with a univentricular circulation following the Norwood procedure. We know that the Norwood procedure will result in, at best, a 60 percent five-year survival and long-term functional issues,” says Dr. Gardiner.

The researchers concluded that fetal valvuloplasty treatment for fetuses with aortic stenosis should be concentrated in experienced centers with expert fetal and surgical facilities to improve survival and reduce morbidity.

### KuoJen Tsao, M.D., Appointed to NAFTNet Executive Committee



KuoJen Tsao, M.D., co-director of The Fetal Center, was recently appointed to the executive committee of the North American Fetal Therapy Network (NAFTNet). He joins fellow co-director at The Fetal Center, Anthony Johnson, D.O., on the executive board.

NAFTNet is a voluntary association of 24 medical centers in the U.S. and Canada that perform advanced in-utero fetal therapeutic procedures, representing a variety of specialties, including maternal-fetal medicine, pediatric surgery, neonatology, pediatric cardiology and fetal ultrasound, and practicing in both academic and community-based organizations. NAFTNet was founded by a governing principle that continues to fuel the program: to provide an umbrella organization to assist the various medical centers that practice fetal medicine, promote cooperation between these centers and foster research in fetal therapy.

With the addition of Dr. Tsao to the executive committee, The Fetal Center continues to remain focused on the development of therapeutic prenatal interventions to maternal and fetal outcomes in high-risk pregnancies affected by fetal disease.

## News of Note

### Growth

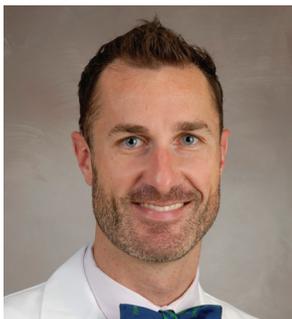


#### **Manish N. Shah, M.D.**

Dr. Shah is a fellowship-trained pediatric neurosurgeon with special expertise in spina bifida, tethered cord syndrome, pediatric epilepsy, and craniofacial and craniocervical spine surgery. He serves as assistant professor in the division of Pediatric Neurosurgery at UTHealth Medical School. He is also an expert in the surgical management of spasticity and dystonia in children, and performs selective dorsal rhizotomies, baclofen pump placements and advanced deep brain stimulation techniques.

Dr. Shah was recruited from Washington University in St. Louis, after completing his fellowship training at St. Louis Children's Hospital under the direction of world-renowned pediatric neurosurgeon Tae Sung Park, M.D. He earned his undergraduate degree in physics from Princeton University and his medical degree at Vanderbilt University in Nashville, followed by a residency in neurosurgery at Washington University. He is the recipient of numerous awards and was elected to Sigma Xi Scientific Research Honor Society in 2002 and Alpha Omega Alpha Medical Honor Society in 2005.

Dr. Shah is an author of research articles published in *Proceedings of the National Academy of Sciences*, *Journal of Neurosurgery*, *Neurosurgery*, *Neurosurgical Focus* and *Journal of Neurosurgery: Pediatrics*, as well as a book chapter entitled "Congenital and Acquired Abnormalities of the Thoracic and Lumbar Spine," published in *Youmans: Neurological Surgery*. He is a member of the American Association of Neurological Surgeons and the American Association of Neurological Surgeons/Congress of Neurological Surgeons' Joint Cerebrovascular Section and Joint Pediatrics Section.



#### **Matthew R. Greives, M.D.**

Dr. Greives is a fellowship-trained craniofacial and pediatric plastic surgeon specializing in patients with cleft lip and palate, craniosynostosis, craniofacial distraction, vascular anomalies, and ear reconstruction. He is currently an assistant professor in the division of Pediatric Plastic Surgery at UTHealth Medical School.

Dr. Greives earned his medical degree at the New York University School of Medicine in New York City, graduating with honors. He received his residency training in plastic and reconstructive surgery at the University of Chicago Medical Center, which included a clinical elective learning the intricacies of ear reconstruction from Dr. Francoise Firmin of Paris, France. He completed a fellowship in craniofacial and pediatric plastic surgery at the Children's Hospital of Pittsburgh/University of Pittsburgh Medical Center.

Dr. Greives has authored numerous articles on vascular biology, wound healing, and cleft and craniofacial surgery. He has traveled with multiple cleft and craniofacial surgical mission trips to Peru, Chile and the Dominican Republic, and was named as an Albert Ellis Scholar for international research. His professional recognitions include the Huggins Surgical Research Symposium Best Clinical Presentation and the ASPS Senior Resident Conference Best Microsurgery Paper. He is currently a member of the American Society of Craniofacial Surgery, American Association of Pediatric Plastic Surgeons, American Society of Maxillofacial Surgeons, the American Cleft Palate-Craniofacial Association, and the American Society of Plastic Surgeons.



#### **Monica McGuire, RN**

*Clinical Nurse Coordinator*

Monica McGuire, RN, joined The Fetal Center in September 2014 as clinical nurse coordinator. She has more than 10 years of nursing experience in pediatrics, bedside nursing and case management. She is a certified pediatric nurse (CPN) and is a fellow in the American Academy of Case Management (FAACM). Last year, she was a finalist for the Nurses as Collaborators, part of The Nurses in Excellence Awards at Children's Memorial Hermann Hospital.

## Genetic Counseling Corner

### Noninvasive Prenatal Testing: Understanding the Importance of Proper Interpretation and Genetic Counseling

Noninvasive prenatal testing (NIPT), also referred to as noninvasive prenatal screening (NIPS), has drastically changed the landscape of prenatal testing and offers tremendous potential as a screening tool for fetal aneuploidy. This option first became available through one laboratory in the U.S. in late 2011 as a screen for trisomy 21. Since then, this type of screening has rapidly expanded and is now available through numerous laboratories across the country for multiple chromosomal abnormalities. While most are in agreement that NIPT has advantages over the more traditional serum screening, there is much to consider with regard to interpretation of results and counseling families both before and after testing.

Studies conclude NIPT has increased sensitivity, specificity and positive predictive value over both first trimester screening and second trimester serum screening; however, it is imperative that practitioners and patients alike recognize NIPT is a screening tool and is not diagnostic. NIPT has been advertised and extensively marketed as a highly accurate test and is frequently described as being over 99 percent accurate. While the sensitivity and specificity of NIPT are frequently greater than 99 percent, the positive predictive value is almost always less than 99 percent. Counseling patients about the meaning of NIPT results can be complicated; therefore, clear explanations of sensitivity, specificity and positive predictive value are essential.

**Sensitivity**—the proportion of affected pregnancies that will screen positive.

“What are the chances this test will detect Down syndrome if it is present?”

**Specificity**—the proportion of unaffected pregnancies that will screen negative.

“What are the chances I will get a reassuring result if my pregnancy does not have Down syndrome?”

**Positive Predictive Value**—the proportion of positive test results that are true positives.

“What are the chances that my positive NIPT result actually means my pregnancy has Down syndrome?”

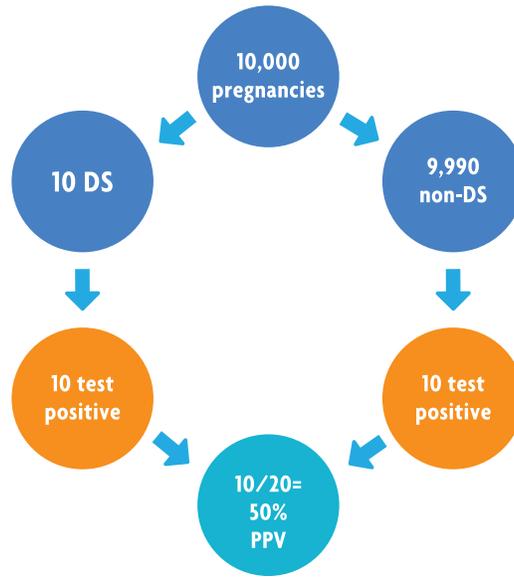
When interpreting abnormal results and counseling patients about the meaning of such results, positive predictive value (PPV) is perhaps the most meaningful number for patients to understand. PPV takes into account not only sensitivity and specificity of the screening test, but also the prevalence of the condition. Prevalence depends on many factors, such as the genetic condition, maternal age, gestational age and ultrasound results. As the use of NIPT expands to low-risk populations and less prevalent chromosome conditions, the impact of prevalence on PPV will become more evident. Therefore, while performance of NIPT is impressive in both high- and low-risk populations, counseling about abnormal results will be different for each individual patient based on their a priori risk for the condition.

For example, interpretation of abnormal NIPT results for Down syndrome differs for a 40-year-old woman versus a 20-year-old woman. The PPV for the 40-year-old woman is higher (91 percent) than the PPV for the 20-year-old woman (50 percent). Given the lower prevalence of Down syndrome in younger women, the PPV in this population is lower, resulting in a higher number of false positive results in this population. Therefore, the genetic counseling for these two patients is different despite them both having a “positive” NIPT for Down syndrome. See figures on the following page.

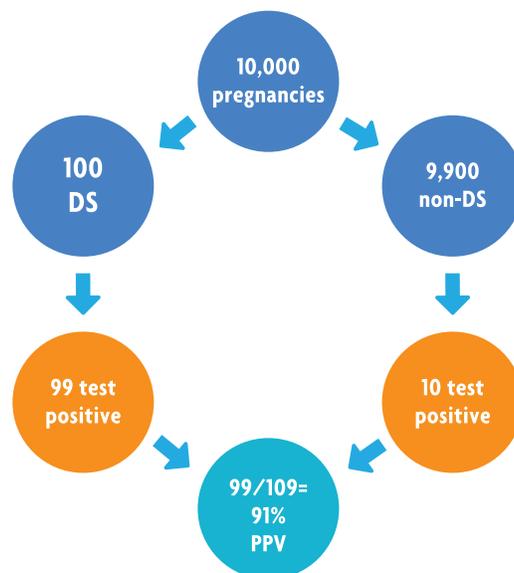
The American Congress of Obstetricians and Gynecologists (ACOG) recommends all women be offered screening for aneuploidy during pregnancy. Screening with increased sensitivity, specificity and PPV is ideal; however, it is important to note for some families, screening is not desired. Pre-test counseling is essential for patients to understand the benefits and limitations of their screening options and aids in informed decision-making and anticipatory guidance regarding follow-up recommendations. Additionally, genetic counseling for detailed interpretation of results and options for diagnostic testing is recommended for all women with abnormal screening results.

For more information or to connect with a genetic counselor, email [thefetalcenter@memorialhermann.org](mailto:thefetalcenter@memorialhermann.org).

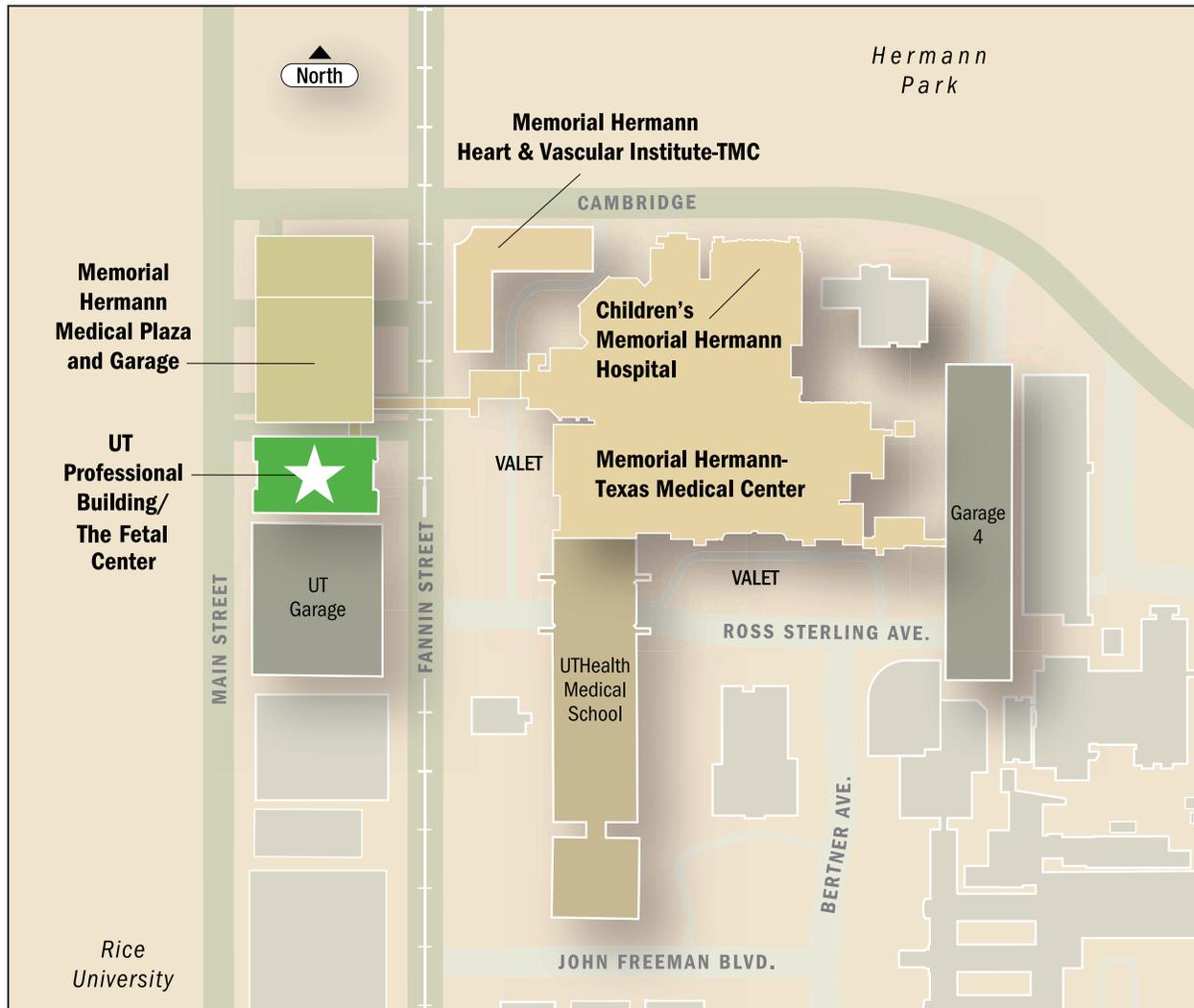
## NIPT Performance: Down syndrome, 20-year-old



## NIPT Performance: Down syndrome, 40-year-old



# Contacts



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