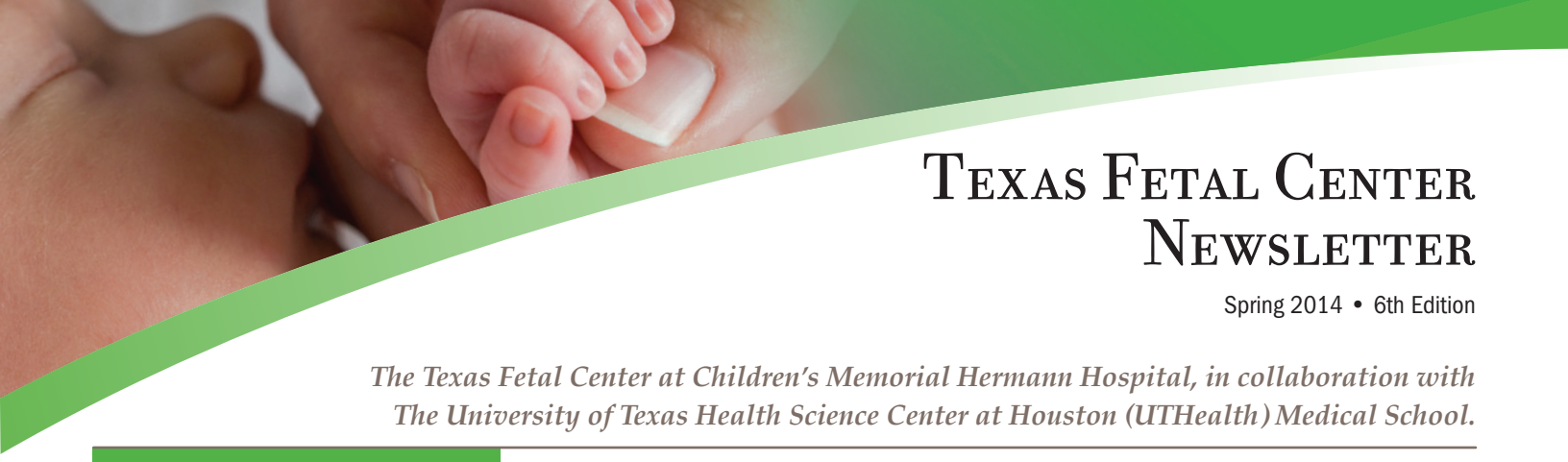

Texas Fetal Center Newsletter

Spring 2014 • 6th Edition

The Texas Fetal Center at Children's Memorial Hermann Hospital, in collaboration with The University of Texas Health Science Center at Houston (UTHealth) Medical School.

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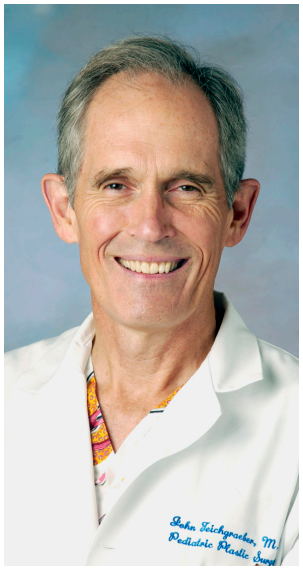
Ways to Give to the Texas Fetal Center

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

For more information on how to support the Texas Fetal Center, visit texasfetalcenter.org/donate.

Facebook.com/TexasFetalCenter

Texas Cleft-Craniofacial Program



As one of the more common birth defects, it is estimated that clefts of the lip, palate or both occur in approximately one in 700 births. Typically, the lip segments of a fetus join together during the seventh week of pregnancy and the palate closes by the 11th week. When this process does not occur, for either genetic or environmental reasons, the result is a cleft lip and/or palate.

According to John Teichgraeber, M.D., FACS, professor and director, division of Pediatric Plastic and Craniofacial Surgery, and leader of the Texas Cleft-Craniofacial Program, cleft lip and/or palate cases can vary widely in severity. Although they are highly treatable, many children will require multiple procedures throughout their childhood to ensure a normal level of oral function and appearance.

"The emotional effects surrounding a cleft diagnosis can be overwhelming, which is why early detection, education and access to appropriate resources are essential in ensuring the best outcomes for both the baby and family," said Dr. Teichgraeber. "It's important for families to find a program with the breadth and depth to provide not only surgical expertise, but psychological support as well."

The Texas Cleft-Craniofacial Program is one of the largest programs of its kind in the region. The multidisciplinary team includes a number of specialists, including affiliated plastic and oral surgeons, pediatric dentists, dietitians, orthodontists, geneticists, speech therapists, otolaryngologists and audiologists, among others. Together they have successfully treated patients across the spectrum of clefts, from those with small notches in the lip to those with more advanced cases involving large openings bilaterally of both the lip and the hard and soft palates. "There are multiple variations, but we see mostly cleft lip and palate," said Dr. Teichgraeber, adding that last year alone they treated 40 new cases of cleft lip and 50 new cleft palates.

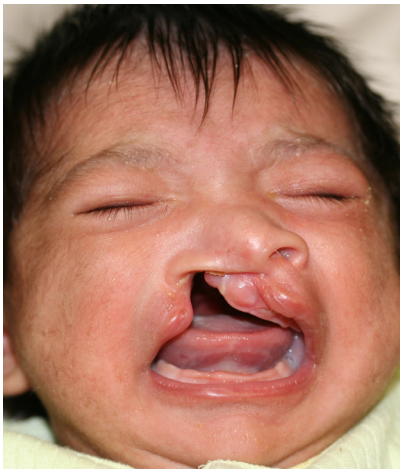
continued

Diagnosis and Treatment

Cleft lip and palate are often diagnosed in utero by maternal-fetal medicine specialists, who also perform additional tests to evaluate for other associated anomalies. In most cases, management of pregnancy is routine and the Center works closely with the obstetrician to determine the patient's delivery plan. There are no fetal treatments for clefts, therefore coordination of care for the baby after birth is recommended. Babies undergo surgery during the first year of life, typically in two stages. The lip is repaired between 3 and 6 months of age and the palate between 10 months and 1 year. In later childhood, many will undergo upper jaw surgery with bone grafting and revision of both the lip and nose, usually somewhere between the ages of 7 and 10 years old.

Dr. Teichgraeber credits early detection in utero as one of the most critical advances in terms of helping parents cope with a cleft lip and/or palate diagnosis, and he says the team sees approximately 60 percent of cases prenatally. "A baby's smile is communication, and the emotional impact of this diagnosis is upsetting. For parents to have the ability to learn about what this diagnosis means early on gives them a head start. They are better educated and more prepared to see past the small deformity."

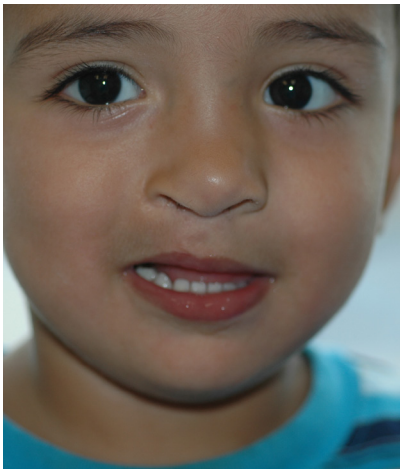
Below are progression photos of a Texas Cleft-Craniofacial Program patient ranging from 1 month to 8 years old.



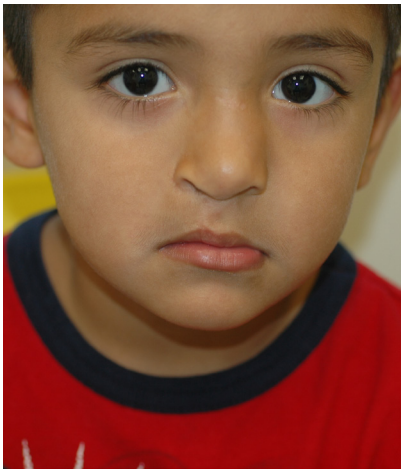
1 month old, before Pre-nasoalveolar molding (PNAM) treatment

After completed PNAM treatment

1 week after surgical lip repair



1 year old



4 years old

8 years old

Patient Story: Cleft Lip and Palate Program Brings New Smiles to Life

Daniella Coca's son Anthony was born last November with both a cleft lip and palate. Coca was five months pregnant when she received the diagnosis after a 3-D ultrasound, which afforded her and Anthony's father several months to meet John Teichgraeber, M.D., and his team, determine a plan of action and better prepare themselves emotionally for the journey ahead. According to Coca, the team's clinical coordinators in Dr. Teichgraeber's office played an integral role in helping her cope. "I still keep in close contact with both ladies. They were so understanding, responsive and supportive. They helped me better understand what to expect during the first year of Anthony's life and provided encouragement along the way. They were like my best friends and therapists all rolled in one."

Anthony had surgery at Children's Memorial Hermann Hospital on February 23, undergoing a simultaneous lip and nose repair by Dr. Teichgraeber, ear tube placement by pediatric otolaryngologist Sancak Yuksel, M.D., and perineal anoplasty for a secondary medical condition unrelated to the clefts by Charles Cox, M.D., director of the department of Pediatric Surgery. "I was overwhelmed when I first saw Anthony's face. My emotions were everywhere," said Coca. "I was absolutely shocked at how different he looked and what a great job Dr. Teichgraeber did. Anthony looked like a different baby. If you didn't know he had a cleft lip and palate, you would not be able to tell."

Today, Anthony's prognosis is good, and his hearing has greatly improved. Coca is confident in his ability to bounce back from his future surgeries, with the cleft palate repair scheduled to occur this summer. "He is such a strong baby, and he recovers so quickly," said Coca. "It has been really hard, but ultimately, I know that I am very blessed."

For more information about the Texas Cleft-Craniofacial Program, call 832.325.7234.

Anthony Coca at three months old, prior to surgical repair.

Anthony at three-weeks post-surgery.

CAPS Program Supports Families Through Continued Growth

The Chronic and Palliative Services team in Marnie's Garden Room, a private room where families can spend final moments with their loved one.

Nearly 10 years ago, a donation from the Dr. Marnie Rose Foundation provided funds for a part-time palliative care nurse at Children's Memorial Hermann Hospital, marking the beginning of what would become one of the most respected pediatric palliative care programs in the region. Today, the Chronic and Palliative Services (CAPS) Program at Children's Memorial Hermann Hospital has grown into a full-fledged multidisciplinary program, consisting of a dedicated team of affiliated neonatologists, nurse practitioners, nurses, social workers, child life specialists and chaplains. The CAPS Program is part of the continuum of care for patients and families at the Texas Fetal Center that have been prenatally diagnosed with a life-threatening or life-limiting condition.

Patrick Jones, M.D., M.A., assistant professor of pediatrics, division of Neonatal-Perinatal Medicine, together with Suzanne Lopez, M.D., FAAP, associate professor of pediatrics, division of Neonatal-Perinatal Medicine at UTHealth Medical School serve as clinical leaders of the CAPS program at Children's Memorial Hermann Hospital. "We recognize that the families we consult with need services above and beyond what a typical pregnancy may require," said Dr. Jones, "Our focus is to help them process difficult information, and to better understand how their baby's particular diagnosis may affect their child and their family. Many of these patients require admission to the neonatal intensive care unit, and it's helpful for the families to see familiar faces and have a network to rely on for answers to difficult questions, from diagnosis to discharge." Dr. Jones lauds the importance of the team's child life specialists, who help families with the important task of discussing and explaining the diagnosis with the patient's siblings, including what they can expect during the first visit to meet their younger brother or sister.

Many of the conditions these families face include congenital heart disease, diaphragmatic hernias, skeletal dysplasias and various genetic syndromes. "Our approach is one of honest communication about the facts each family is facing, while maintaining hope and providing a positive environment for the family," said Dr. Lopez. When a family is provided with a prenatal diagnosis of a life-threatening condition and the child is unlikely to survive long after delivery, the CAPS team works with the family to create memories, which is an important part of the grieving process, and does its best to meet whatever goals the family has for the time they have with their child. Marnie's Garden Room, located within Children's Memorial Hermann Hospital, was established to support the team's goal to provide families with a comfortable, private and serene place to spend final moments with their loved one.

A Comprehensive and Unified Approach

In 2012, the team traveled to Akron, Ohio, to participate in an operational training and mentorship program provided by the Center to Advance Palliative Care (CAPC), with nationally renowned palliative care expert, Sarah Friebert, M.D. Every year, CAPC brings palliative care teams from all over the world to interface with experts from some of the country's best programs during three days of intensive training. "Dr. Friebert's expertise led us to create a plan to further develop our program, and we have continued to benefit from her mentoring as we have put this plan into action," said Dr. Jones. "The work implemented almost 10 years sago by both Dr. Lopez and Shih-Ning Liaw, M.D., a pediatrician formerly with UTHealth Medical School, has provided a solid clinical foundation for the program." Dr. Jones, collaboratively working with Caryn Douma, RN, director of the Patient Care, Chronic and Palliative Services Program at Children's Memorial Hermann Hospital, is continuing to focus on improving the implementation and growth of the program in order to reach more patients in our community.

Pediatric palliative care is a relatively new field, and part of what sets the CAPS program is the complex patient population at Children's Memorial Hermann Hospital, where patients are treated across multiple service lines and include those diagnosed with fetal anomalies or high-risk pregnancy conditions, NICU patients and complex pediatric patients. The team also cares for women, post-delivery, who require additional treatment in the adult ICU or at Memorial Hermann Heart & Vascular Institute-Texas Medical Center.

The CAPS program is a coordinated effort between Children's Memorial Hermann Hospital and UTHealth Medical School, and works collaboratively with the clinical team at the Texas Fetal Center. Dr. Jones and other members of the Program regularly attend the Texas Fetal Center's weekly multidisciplinary meeting where patients' customized plans of care are discussed with physicians of all pediatric specialties. "It has been a great relationship, made possible by the leaders of the Center recognizing the importance of our services to their patients and families," said Dr. Jones.

Research and Growth

CAPS team members recognize that research and quality improvement are important to the future of palliative care. Through the collaboration with the Texas Fetal Center, the team is actively challenging its own ideas and conducting important research in order to provide the best possible treatments to patients. "Palliative care is a young field in pediatrics, and now that programs are being established across the country, it's time to take that next step by conducting studies that will help us better serve patients and their families," said Dr. Jones. For example, Amanda Hutchens, M.D., a fellow in neonatal-perinatal medicine at UTHealth Medical School, is working with the CAPS team to determine the prevalence of depression and post-traumatic stress disorder in parents at six and 12 months postpartum. Information from this study will be used to develop new tools to help support families as they focus on providing their child with the best possible care.

Looking to the future, Dr. Lopez anticipates further growth as interest in pediatric palliative care intensifies in the physician community. "We have added two additional pediatric nurse practitioners and a licensed psychologist to our team," said Dr. Lopez, who also noted that many residents who rotate with the team are beginning to express interest in the field. "Our program is very robust, and it's providing an important service to the medical community by providing these future physicians with a glimpse at the importance of pediatric palliative care."

The Chronic and Palliative Services (CAPS) team at Children's Memorial Hermann Hospital works hand-in-hand with the Texas Fetal Center. The CAPS program is centered on compassion, and helps:

- Assist parents in creating a birth plan that is consistent with their hopes, goals, wishes and values
- Explore treatment pathways and facilitate medical decision-making
- Offer emotional support for the entire family
- Provide resources for coping strategies in the struggle of uncertainty
- Continuity of care from diagnosis to discharge

For more information about Chronic and Palliative Services at Children's Memorial Hermann Hospital, email texasfetalcenter@memorialhermann.org.

Texas Fetal Center: Research

Parental Decision-Making for an Unborn Child with Spina Bifida

Principal Investigator: Lynnette J. Mazur, M.D., M.P.H.

Co-Investigators: Anthony Johnson, D.O., Kenneth Moise Jr., M.D., KuoJen Tsao, M.D., Co-Directors, Texas Fetal Center; Karen Moise, RN, Nurse Coordinator, Texas Fetal Center; Rebecca Carter, CGS, Genetic Counselor, and Sangbum Choi, Ph.D., Statistician, The University of Texas School of Public Health

Lynette Mazur, M.D., pediatric spina bifida consultant affilliated with the Texas Fetal Center, along with members of the Texas Fetal Center team, was successful in obtaining a $10,000 grant from The Program of Interprofessional Ethics at UTHealth Medical School to look into various aspects of decision-making faced by parents of an unborn child with spina bifida, including frequency of pursuing or not pursuing surgical repair and factors involved in decision-making.

Spina bifida is a defect of the spine that affects a child's cognitive development, bladder and bowel continence, and ability to walk, and fetal repair of the defect has been shown to markedly improve a child's quality of life in these areas. Since the first repair at Children's Memorial Hermann Hospital in March, 2011, many families have traveled from across the country and around the world to the Texas Fetal Center to be evaluated by the team to determine if they are a candidate for fetal repair. All families are provided with objective information about the condition and the treatment options, better equipping each family to reach its own decision.

To better understand what factors are important to parents when deciding between pre- or post-natal surgery or a termination of the pregnancy, the parents of all patients who have already been evaluated at the Center will be mailed a questionnaire to determine:

1. The frequency of parents who pursued fetal surgery, postnatal surgery, or termination of the pregnancy after presenting for consultation at the Texas Fetal Center

2. The importance of factors involved in the decision-making process, such as:
   - Parental age
   - Education level
   - Employment
   - Perception of the severity of the disability
   - Distance from subspecialty care such as neurosurgery, orthopedics or urology
   - Financial and insurance concerns
   - Religious affiliation
   - Number of children in the family

Their responses will rate each of these factors as unimportant, neutral or most important, and the answers for each group of parents will be compared. The questionnaire will also determine if parents would make the same decision again.

For more information about this study, email texasfetalcenter@memorialhermann.org or call 832.325.7288.

Connect With Us – Image Gateway Program

Access, store and share medical data through Image Gateway

Providing exceptional patient care begins with having immediate access to the most up-to-date clinical data for patients referred to the Texas Fetal Center. Referring physicians are now offered the ability to easily connect with the Center's affiliated team and provide important images such as ultrasound, fetal ECHO and MRI, prior to patient referral or when a second opinion is requested.

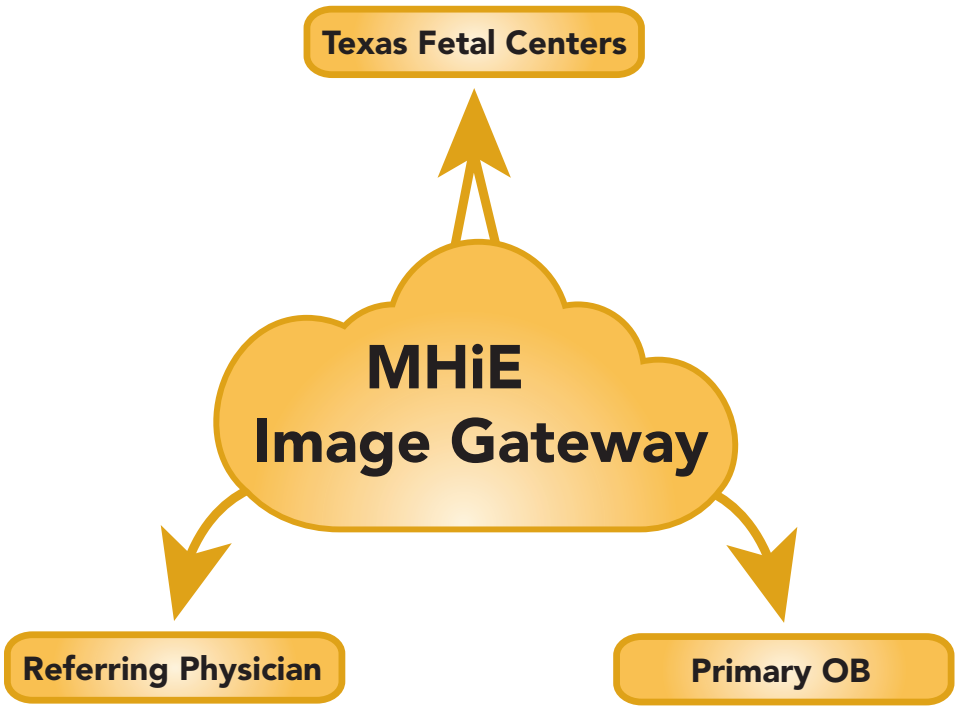
Memorial Hermann Information Exchange (MHiE) Image Gateway is a secure, encrypted and HIPAA-compliant cloud-based network utilized to access, store and share medical data between the referring physician and the Center's affiliated team. By simply creating a username and password, and providing the Texas Fetal Center's share code (available online in the instruction manual), physicians can easily share images and schedule a collaborative meeting to discuss the most current clinical information.

The Center's team recognizes the importance of open communication between the patient, the referring physician and the team. The implementation of the MHiE Image Gateway aims to enhance the Center's services by improving communication and streamlining patient care. Advantages include:

- Collaborative care, including quick and easy transfer of patient's medical data
- Secure, HIPPA-compliant cloud storage
- Unlimited cloud storage
- Synergetic resource for research studies
- Complimentary service

System requirements for Image Gateway include an HTML5-compatible browser, the installation of Java 7 or higher for full functionality, and Internet access. For more information and to sign up for MHiE Image Gateway, visit texasfetalcenter.org/ImageGateway. If you have questions about the program, email texasfetalcenter@memorialhermann.org.

How it works:



Fascinating Fetal Finds: Fetal Neck Teratoma

KuoJen Tsao, M.D., surgically removing a large fetal teratoma.

A large fetal neck teratoma is a very rare malformation. While the exact cause is unknown, these masses are composed of several types of embryonic tissue and appear on ultrasound and fetal MRI as large masses containing both cystic and solid components. The masses can cause complications during pregnancy as the mass displaces the normal structures of the neck. It can interfere with the fetus' ability to swallow by compressing and obstructing the throat and esophagus. This results in polyhydramnios, increased amniotic fluid volume, and can be associated with higher risks of preterm labor, preterm rupture of the membranes and placental abruptions. These masses can also distort and compress the structures of the upper airway. At the time of delivery, this can make the establishment of an adequate airway in the newborn very difficult.

A 21-year-old woman was transported to Children's Memorial Hermann Hospital at 28 weeks and 1 day gestation. Her pregnancy had been complicated recently by the diagnosis of a large fetal neck mass and polyhydramnios, with an amniotic fluid index (AFI) of 34 cm. The patient began to experience uterine contractions earlier that day resulting in the request for transfer. An ultrasound evaluation performed by the Texas Fetal Center at the time of admission confirmed the presence of a large mass consisting of cystic and solid components originating from the neck of the fetus. The overall size of the mass was calculated from 3-D reconstruction at 346 cc and was more left-sided, but it also extended across the anterior aspect of the neck to the right side. The fetal neck was extended and tilted to the right by the mass. There was no apparent invasion of the oropharynx. The stomach could not be visualized and was thought to be secondary to esophageal compression by the mass. The cervical length measured 3.4 cm.

Initial management was directed toward the threatened preterm labor. A fetal echocardiogram and ultrafast fetal MRI were obtained. The ECHO showed normal cardiac structure and function and the MRI confirmed the ultrasound findings (see Image 1, next page). The trachea was visible below the mass and was a normal caliber, suggesting there was no obstruction of the airway by the mass. The case was reviewed at the Texas Fetal Center's weekly multidisciplinary rounds and it was determined that the best course of management would be delivery by an ex-utero intrapartum treatment (EXIT) procedure. The family was extensively counseled and agreed with the management plan. The threatened preterm labor resolved and the mother remained in the hospital; the surgical team was on call, in case a complication developed.

One week later, re-evaluation on ultrasound showed the mass had enlarged to 861 cc. The polyhydramnios had increased with the AFI now measuring 44 cm. Evaluation of the cervix showed the cervix now measured only 3 mm, with bulging of the lower uterine segment. It was decided to perform the EXIT procedure despite the gestational age of 29 weeks 4 days.

A successful EXIT procedure was performed. It was possible to intubate the trachea from the oropharynx in spite of the distorted anatomy. The fetus was on placental support for 45 minutes. Once the airway was stabilized and the fetus well oxygenated, the umbilical cord was cut and the newborn was transferred to the NICU for stabilization and surgical resection of the tumor. The maternal recovery was uncomplicated.

EXIT PROCEDURE

Early diagnosis of a fetal neck mass is essential to the development of a strategic plan to optimize the delivery and postnatal management of the newborn. The EXIT procedure is the foundation of that strategy and has been shown to be an effective method for delivery of patients with potential airway obstruction at birth. The fetus is partially delivered but maintained on placental circulation to provide ongoing oxygenation and support. This allows access to the fetal airway in an elective, controlled and secure manner.

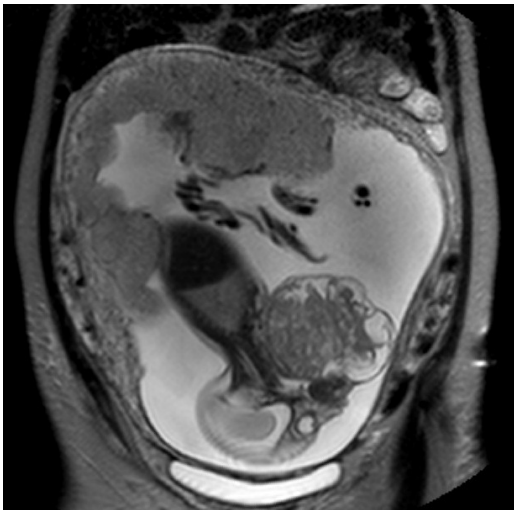


Image 1: MRI image confirming the diagnosis of a fetal neck teratoma.

Once an airway is secured, the rest of the fetus is delivered, the umbilical cord is clamped and the newborn transferred to the neonatal team for stabilization. While simple in concept, the success of an EXIT procedure relies on extensive preoperative evaluation to determine feasibility, careful planning and the availability of a highly skilled multidisciplinary team.

The Texas Fetal Center is one of the few centers in the world where such expertise exists. The EXIT procedure is a well-organized, multidisciplinary treatment plan; it is the optimal treatment plan for a number of other conditions where problems are anticipated with the baby's ability to breathe at the time of delivery.

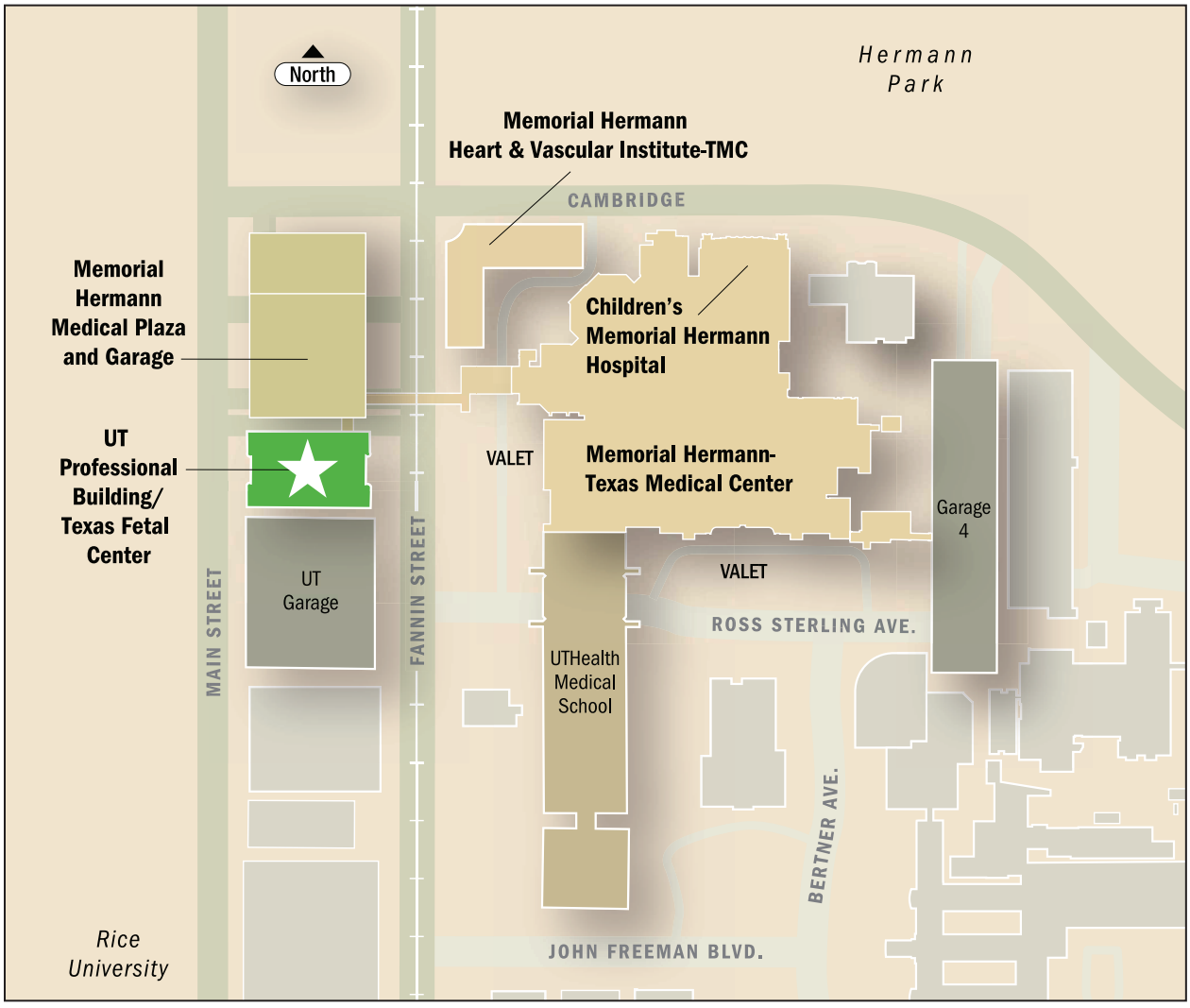
The EXIT procedure is not without risk for both mother and newborn. The level of general anaesthesia required to achieve uterine relaxation contributes to risks for increased bleeding. Postnatal care of the newborn can involve prolonged hospitalization. In addition to resection of the tumor, many newborns have underlying pulmonary hypoplasia from the neck mass distorting normal lung development. Many are born prematurely and thyroid/parathyroid gland dysfunction is common following resection of the tumor.

The Texas Fetal Center was the first in Texas to perform the EXIT procedure, which is utilized when doctors are concerned that the baby may not be able to breathe adequately on its own following delivery. This can be for several reasons, including problems with the lungs or masses of the neck or chest that may be compressing the airway. Such indications may include, but are not limited to:

- Large fetal neck masses
- Congenital diaphragmatic hernias
- Congenital high-airway obstructions
- Lung masses
- Chest or mediastinal tumors

The early diagnosis and multidisciplinary management of patients in a tertiary care facility is important to the delivery plan. For more information, contact us at 832.325.7288 or texasfetalcenter@memorialhermann.org

Contacts



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