IN UTERO insights

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The Children's Hospital *of* Philadelphia®

CENTER FOR FETAL DIAGNOSIS & TREATMENT

Dear Colleague,

Lung lesions are extremely rare, serious diagnoses; so rare that most obstetricians may see only one case in their entire career. Our Center for Fetal Diagnosis and Treatment has seen the largest volume of these patients of any center in the world and offers an unmatched level of experience diagnosing, managing and treating these conditions.

Lung lesions are unpredictable and have a broad spectrum of presentations, from a very small mass to a very large growth that can cause significant problems for both fetus and mother. Our team understands and can anticipate challenges that may occur during these pregnancies, and has the skill and experience to safely and quickly respond.

We work in concert with referring physicians to educate families about the condition and take them from the devastation of diagnosis to a clear plan for management before birth and care after birth.[1] We provide comprehensive care, including a one-day evaluation and thorough prenatal management, the full range of fetal therapy techniques, surgical and anesthesia expertise, the option of delivering within our hospital when medically appropriate, and long-term clinical follow-up.

This issue of *In Utero Insights* offers a detailed look at the care and resources we offer families whose pregnancies are diagnosed with lung lesions — as well as our research into improved therapeutic interventions. We value the opportunity to partner with you in providing the highest level of care possible for these patients and we welcome your feedback.

Sincerely,

N. Scott Adzick, M.D., M.M.M.
Surgeon-in-Chief
Director, Center for Fetal Diagnosis and Treatment

Lori J. Howell, R.N., M.S.
Executive Director
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[1] Breathing Easier: Fetal Lung Anomalies Video
www.chop.edu/video/breathing-easier-fetal-lung-lesions/home.html

A RARE LEVEL OF EXPERTISE IN THE DIAGNOSIS AND CARE OF LUNG LESIONS

The Center for Fetal Diagnosis and Treatment at The Children's Hospital of Philadelphia (CHOP) has managed the care of more than 1,600 pregnancies prenatally diagnosed with lung lesions — the largest volume of any fetal center in the world. Through this experience, we have learned that the natural history and clinical spectrum of these lesions is extremely variable and requires meticulous prenatal diagnosis. Advances in imaging have permitted definition of the natural history of fetal lung lesions, determination of the pathophysiologic features affecting fetal and neonatal outcome, and formulation of effective management strategies as early as possible.

The differential diagnosis of a fetal lung mass includes congenital cystic adenomatoid malformation (CCAM), bronchopulmonary sequestration (BPS), hybrid lesions (CCAM and BPS elements), as well as other conditions such as bronchial atresia, bronchogenic cyst, esophageal duplication cyst, lobar emphysema and congenital high airway obstruction syndrome (CHAOS). These intrathoracic lesions can be confused with one another, but have distinct differences that require evaluation by a skilled team. For example, a right microcystic CCAM can appear to be a right congenital diaphragmatic hernia, while a BPS with a systemic arterial feeding vessel may also present as a solid chest mass, but cysts on ultrasound would make it more consistent with a hybrid lesion.

At CHOP, the experienced multidisciplinary team involved in diagnosis and care of lung lesions — including high-risk obstetricians, pediatric surgeons, advanced practice nurses, radiologists, a coordinator, genetic counselors, social workers, anesthesiologists, neonatologists and cardiologists — is centrally located under one roof and is here to help 24/7/365. Families who come to our Center undergo a one-day comprehensive

continued >

Lung Lesion Experience at CHOP: 1995 to 2014

Referrals	>1,600
Postnatal Surgical Care of Lung Lesion Patients	>1,200
Open Fetal Surgeries	24
EXIT Deliveries	31
Thoracoamniotic Shunts	95

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A RARE LEVEL OF EXPERTISE IN THE DIAGNOSIS AND CARE OF LUNG LESIONS

evaluation that includes Level II ultrasound using color flow Doppler, fetal echocardiogram and ultrafast fetal MRI — a technique we helped develop — which can be very helpful in ruling out alternate diagnoses such as bronchial atresia, bronchogenic cyst or multilobar involvement.

Prenatal management and the overall prognosis of a lung lesion depends on the size and growth of the mass. More than 15 years ago, our imaging specialists developed the CCAM volume ratio (CVR) as a prognostic tool to measure the size of lung lesions before birth. This ratio helps determine the frequency of sonographic evaluation required over the duration of the pregnancy. Historically, we did not have a CVR to reassure families that their baby would not develop hydrops. Now, in many cases, we are able to predict the likelihood of hydrops developing.

Most lesions are followed by ultrasound and treated electively after birth. Esophageal compression by a large thoracic mass can interfere with fetal swallowing of amniotic fluid and result in polyhydramnios and potential preterm delivery. If a lesion continues to enlarge, further mediastinal shift occurs resulting in compression of the heart and vena cava which may lead to hydrops. Maternal mirror syndrome, a preeclamptic state where the mother mirrors the condition of her sick fetus, can occur in this scenario and requires careful maternal observation. When maternal mirror syndrome occurs, the only therapeutic alternative is delivery of the fetus, and invariably the sick, hydropic premature neonate dies.

For fetuses that have macrocystic CCAMs and hydrops, a fetal thoracoamniotic shunt can be employed to decompress the cyst fluid into the amniotic space and decrease the mass effect in the chest. For fetuses with solid lesions with a CVR>1.6, maternal betamethasone can abrogate lesion growth. If betamethasone fails to stop the lesion's growth and hydrops develops or persists, fetal resection is an option offered to families.

Our pediatric surgeons developed the open fetal surgical technique used to resect lung lesions. Intraoperative management of the mother and fetus is intensive. Some of the critical elements needed to carry out this procedure are sterile intraoperative ultrasound to determine the placental and fetal position; real-time fetal echocardiography, provided by fetal cardiologists from our Fetal Heart Program, to determine cardiac function, aortic reversal of flow, ductal constriction, tricuspid regurgitation and decrease in fetal heart rate; employment of a miniaturized fetal pulse oximeter; and placement of an IV to deliver blood or medications.

Over the course of the pregnancy, our team works with referring physicians to determine the delivery location to ensure the best outcome for mother and baby. Babies with small lesions can be delivered vaginally or via cesarean section (for obstetric indications) at a hospital close to home. Those with a mediastinal shift, but without hydrops or major lung compression, will likely be symptomatic and should be delivered by planned cesarean section at a quaternary center such as our Garbose Family Special Delivery Unit. This unit was specifically created to allow immediate access to the baby by necessary subspecialists and avoid transport and separation of mom from her newborn. Two operating rooms separated by a neonatal resuscitation room provide access to the baby for immediate resection of the lung lesion if necessary.

For those fetuses late in gestation with a large mediastinal shift and lung compression, we anticipate difficulties with neonatal ventilation and recommend ex utero intrapartum therapy (EXIT) delivery. The EXIT ensures uteroplacental gas exchange and fetal hemodynamic stability, converting a possible catastrophic emergency into a controlled, planned operation for mother and baby. We have performed more than 100 EXIT procedures for a variety of indications, by far the largest experience in the world. The procedure requires a multidisciplinary team; deep general anesthesia for good uterine relaxation; supplemental fetal anesthesia by intramuscular vecuronium and fentanyl; and sterile intraoperative sonography.

Babies born asymptomatic are seen at 1 to 2 months of age by the pediatric surgeon. Typically a CT scan with IV contrast and the low-dose pediatric radiation protocol pioneered at CHOP is performed and these images are compared to the prenatal ultrasound and ultrafast fetal MRI. We offer postnatal resections by either a short muscle-sparing thoracotomy or thoracoscopy depending upon the presentation. The highly specialized neonatal anesthetic management is detailed in the "News from Anesthesia" article in this newsletter. We recommend elective resection between 1 to 3 months of age due to risks of infection and malignant degeneration. Infants are usually discharged within 48 hours of the procedure, and follow-up occurs one month after surgery and then six months to a year later.

After the lung lesion is removed — whether before or after birth — the remaining normal lung tissue shows remarkable compensatory growth to fill the chest in children until about age 8. Early resection maximizes this compensatory lung growth and long-term follow-up has shown no residual respiratory problems as a result. Results with postnatal resection are excellent and parents should be reassured. We have come a long way in our understanding of lung lesions, but there is much more work to do. The pathophysiology is complex and our Center for Fetal Research laboratory is focused on furthering our knowledge of these rare conditions.

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"The high volume of patients we see from around the world with incredibly complex, rare conditions such as these makes all the difference in achieving favorable outcomes."

— N. Scott Adzick, M.D., M.M.M., Director, Center for Fetal Diagnosis and Treatment

BEYOND THERAPY

Lifesaving Care, Lifelong Connection

In 1995, after a routine pregnancy ultrasound, Felicia Rodriguez and her husband, Roberto, learned that the child Felicia was carrying was a boy. They also learned that there was something terribly wrong: a large mass was growing in the fetus' left lung.

Determined to do everything possible to save their unborn child, Felicia and Roberto traveled from their West Palm Beach, Florida home to the newly opened Center for Fetal Diagnosis and Treatment, at the time one of only two programs worldwide capable of treating birth defects *in utero*. An extensive evaluation at the Center consisting of a level II ultrasound using power Doppler, ultrafast MRI and fetal echocardiogram showed a congenital cystic adenomatoid malformation (CCAM) so large it was filling virtually the entire chest cavity and squeezing against the fetal heart, causing fetal hydrops. After discussing the diagnosis with the family and counseling them about the risks of fetal surgery, the Center's team — led by N. Scott Adzick, M.D., M.M.M., a pioneer in the field — performed a prenatal operation to remove the lesion from the left lung at 22 weeks gestation.

It was the first-ever fetal surgery at CHOP; it lasted just 1½ hours and was a success. Thirteen weeks later, Roberto Rodriguez Jr. was born weighing 6.5 pounds, with a healthy set of lungs. Roberto is now a 17-year-old in his senior year of high school. He plans to go to college to study physical therapy and to play baseball, his favorite sport, which he's played at a competitive level for most of his childhood. His lifelong dream is to one day play professional ball.

"We feel truly blessed for the CHOP program, its staff and the great miracle we received for our family," Felicia says. "We feel as if we could never give back nearly as great as what we were given."

Since that first surgery on Roberto, Dr. Adzick and CHOP have become world leaders in the management of birth defects and fetal surgery. More than 15,000 families from all 50 states and more than 56 countries facing the impact of a prenatally diagnosed birth defect have found support and clinical expertise at the Center. And though the Rodriguez family returned home after Roberto's birth, their relationship with the Center had just begun.

They have returned to Philadelphia nearly every year since to attend the Center's annual Fetal Family Reunion. The reunion is a special gathering for former CHOP patients who either underwent fetal surgery to treat conditions before birth, or needed specialized care before or surgery immediately after birth. Families who have gone through similar struggles can interact with each other and reconnect with their physicians, surgeons and nurses. Roberto and his family have seen the reunion grow from 10 families in 1996 to more than 1,400 people from nearly 20 states this past year.

"It's phenomenal to come back and see all of the kids running around, the result of all the good things CHOP is doing," Felicia says. "Now you can barely fit everybody in one photo frame."

Recognizing that many families travel a great distance to CHOP for care and that making the trip for the reunion is a struggle, the Center organized its first-ever Florida Fetal Family Reunion this past spring. Former CHOP patients from across Florida gathered at the spring training home of the Philadelphia Phillies in Clearwater, Florida.

Roberto and his family were in attendance; the budding baseball star even threw out the first pitch. He wears his fetal surgery scar as a badge of honor and feels a debt of gratitude to his medical family at CHOP, which he repays every year by serving as a positive role model at the reunion for the children who have come after him.

"I feel like I was brought into this world to do something big and not just be mediocre," says Roberto. "[I] feel like I'm going to go far in life."

Roberto, the Center's first fetal surgery patient, pictured in the circular photo above during his fetal surgery and here playing baseball, his favorite sport. He and his family have been able to keep in touch with the team that cared for him more than 17 years ago through the Center's annual Fetal Family Reunion.

EDUCATIONAL RESOURCES

Free Online CME Now Available: Management of Fetal Lung Lesions

This online CME activity will provide participants a foundation and framework for confidently identifying prenatal lung anomalies. Participants will learn evidence-based diagnostic techniques, current options and criteria for fetal intervention, considerations for appropriate delivery planning, and guidelines for neonatal and pediatric surgical management. Clinicians will acquire enhanced skill in diagnosing, counseling and managing pregnancies complicated by these anomalies and caring for neonates with these defects.

The Children's Hospital of Philadelphia designates this enduring material for a maximum 1.5 *AMA PRA Category 1 Credit(s)™*.

To take this course, go to: **fetalsurgery.chop.edu/fetal-ed**

RECENT PUBLICATIONS

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Current imaging of prenatally diagnosed congenital lung lesions. Epelman M, Kreiger PA, Servaes S, Victoria T, Hellinger JC. *Semin Ultrasound CT MR.* 2010;31(2):141-157.

Comprehensive Evaluation of Lung Lesions, Before and After Birth

By Beverly G. Coleman, M.D., F.A.C.R., attending radiologist, and Ann M. Johnson, M.D., pediatric radiologist

At the Center for Fetal Diagnosis and Treatment, we believe one of the most important elements of care is careful diagnostic testing and interpretation of results, both prenatally and postnatally. At their first visit with us, women with a suspected lung lesion undergo a day of testing using the most advanced diagnostic equipment to evaluate the fetal chest. The expertise of our imaging team frequently leads to additional findings and may significantly alter the initial diagnosis.

Fetal Ultrasound

A sonographer and a radiologist, both highly specialized in diagnosing fetal anomalies, use state-of-the-art ultrasound machines and high-frequency probes, as well as 3-D and 4-D techniques if needed, to visualize a baby's condition. The examination is held in a private room with a TV screen overhead so the family can watch the ultrasound in real time. Our team thoroughly examines the fetus from head to toe, including fine details such as the ears, corpus callosum and the conus medullaris. We have developed specific ultrasound protocols for each suspected anomaly and typically devote up to 1½ hours or more for each initial scan.

Fetal ultrasound imaging protocol

- The highest frequency transducer possible (9-12 MHz) to show the most detailed resolution
- Identification of the size and type of lesion with volumetric measurements
- Localization and assessment of lobar involvement based on the lesion location
- Color and spectral Doppler views of the blood vessels supplying and draining the lesion
- Measurement of the largest macrocysts, if applicable
- Calculation of the CVR (CCAM volume ratio), if applicable
- Evaluation for evidence of a dilated bronchus or fluid-filled trachea, if applicable
- Identification of ipsilateral and contralateral uninvolved normal lung parenchyma
- Subjective determination of the degree of mediastinal shift as mild, moderate or severe, with mild representing cardiac displacement just past the midline and severe representing cardiac displacement to the contralateral chest wall
- Evaluation of the fetus for signs of hydrops, including scalp/skin edema, polyhydramnios, ascites, pleural effusions, pericardial effusions, placental thickening or Doppler changes

Fetal MRI

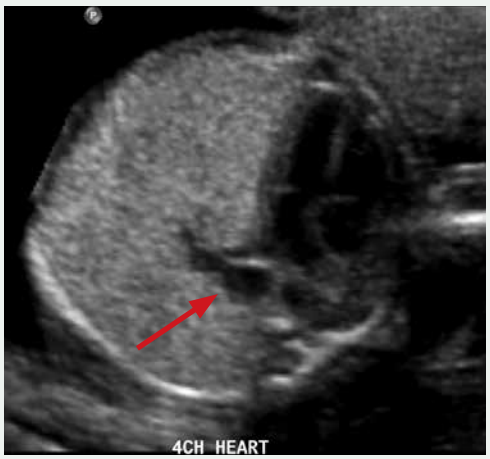
Fetal magnetic resonance imaging (MRI) provides excellent soft tissue resolution and proportionate anatomic detail of the anomaly, and often provides important information about the patency, caliber and position of the fetal airway. This information is critical in counseling and management decisions.

Fetal MRI imaging protocol

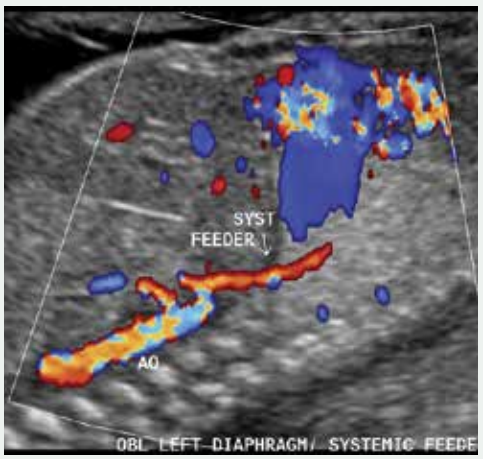
- Performed on 1.5 or 3 Tesla magnet systems
- Patient positioned in supine or decubitus position with coil overlying the abdomen
- No sedation or contrast material used
- 30 to 40 minutes average imaging time
- Localization and characterization of the lung lesion and the central airway to complement ultrasound evaluation
- Volumetric assessment of normal lung and lesion
- Evaluation for additional fetal or maternal abnormalities

Postnatal CT

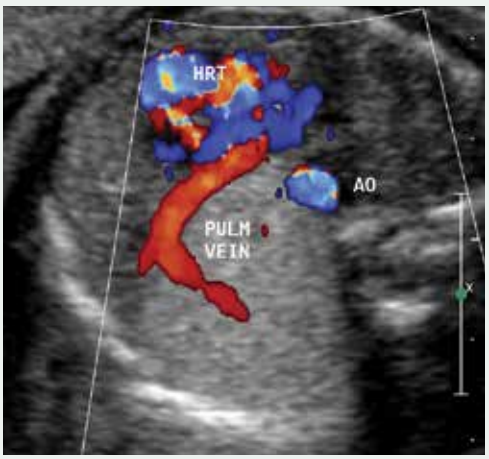
Postnatal computed tomography angiography (CTA) within the first two months of life provides information essential for diagnostic confirmation and surgical planning. This technique allows multiplanar reconstructions of the airway, lung parenchyma and vasculature. CT images are compared with prenatal studies. Studies performed at CHOP also allow for



Transverse view of the chest demonstrating a dilated fluid-filled bronchus in a case of bronchial atresia that was referred as a CCAM.



Sagittal view of a hybrid lesion, demonstrating a systemic feeding vessel from the celiac axis below the diaphragm.



Transverse view of the same hybrid lesion, demonstrating the pulmonary draining vein.

additional imaging post processing that provides important information in some cases. The CHOP nursing and sedation staff is highly attuned to the unique imaging and sedation needs of pediatric patients. Our experienced pediatric radiologists perform CTA adhering to ALARA (As Low As Reasonably Achievable) principles for radiation exposure. CTA protocols have been designed with attention to radiosensitivity, low body weight index, faster heart rates, smaller cardiovascular structures and increased patient motion. The administration of intravenous iodinated contrast material helps to define vascular anatomy.

Types of anomalies in the fetal chest we evaluate include:

- CCAM (congenital cystic adenomatoid malformation)
- BPS (bronchopulmonary sequestration)
- Hybrid lung lesion
- Bronchial atresia
- Foregut duplication cyst
- Pulmonary agenesis

Complex Referred Cases Made Clear by Our Imaging Team

- Hybrid lesion misconstrued as a simple CCAM
- CCAM misidentified as a CDH or CDH mistaken for CCAM because of a herniated gallbladder
- Extralobar BPS in the abdomen thought to represent an adrenal mass
- Bronchogenic cysts mistaken for bronchial atresia or CCAM
- Bronchial atresia misconstrued as a CCAM or BPS
- Pulmonary agenesis referred as a CCAM or CDH
- CHAOS referred as a bilateral CCAM

RESEARCH UPDATE

Increasing Understanding of the Genetic and Molecular Mechanisms Responsible for CCAM

By Marcus Davey, Ph.D., and William H. Peranteau, M.D.

At the Center for Fetal Research, the research extension of CHOP's Center for Fetal Diagnosis and Treatment, an area of great interest involves genetic manipulation of lung development via viral-vector gene therapy. We recently developed an efficient method for gene transfer to fetal lung interstitial cells during the pseudoglandular stage of lung development by direct ultrasound-guided intrapulmonary injections of viral vector (see images below). Using this technique, we investigated whether focal expression of Fibroblast Growth Factor-10 (FGF10), an epithelial growth factor pertinent to fetal lung growth, would initiate epithelial proliferation.

An adenovirus incorporating green fluorescent protein (GFP; reporter gene) and FGF10 was injected into fetal rat lungs at either 15.5 (pseudoglandular) or 18.5 (canalicular) days of gestation (term~21 days). We found that overexpression of FGF10 restricted to the proximal tracheobronchial tree during the pseudoglandular phase resulted in large cysts lined by tall columnar epithelium comprised primarily of Clara cells with a paucity of Type II pneumocytes, resembling bronchiolar type epithelium.

In contrast, FGF10 overexpression in the distal lung parenchyma during the canalicular phase resulted in small cysts lined by cuboidal epithelial cells comprised of primarily Type II pneumocytes resembling acinar epithelial differentiation. The cystic malformations induced by FGF10 overexpression appear to closely recapitulate the morphology and histology of the spectrum of human CCAM.

The development of this animal model moves us closer to understanding the genetic and molecular mechanisms responsible for the etiology of CCAMs in human fetuses. Additionally, it provides a platform upon which to explore therapeutic interventions which are currently clinically utilized such as maternal steroids or those which are potential therapeutic targets for the future such as gene therapy. With this armamentarium and continued research, we hope to enhance the care we provide to the many families carrying a fetus with this diagnosis.

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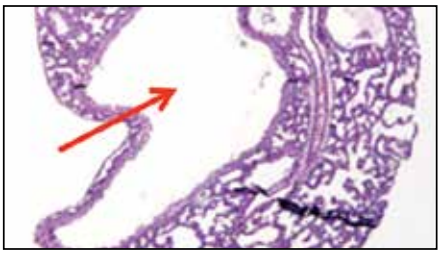
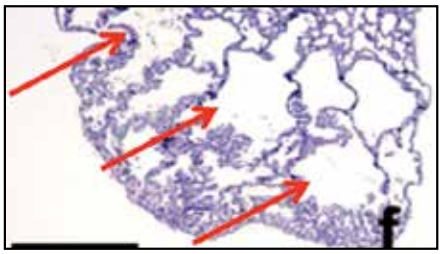
Histology of CCAM

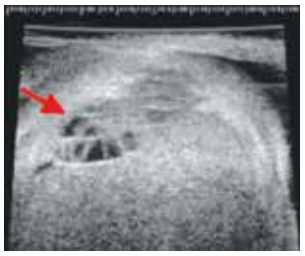


Macrocystic CCAM

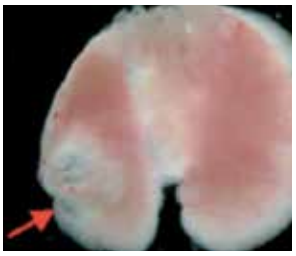


Microcystic CCAM

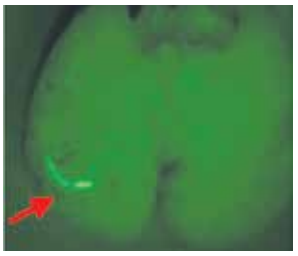
FGF10 overexpression produces CCAM-like malformation in fetal rat lung



Lung ultrasound



Fetal rat lung



GFP reporter gene

Providing Anesthesia for Patients with Congenital Lung Lesions

By Kha Tran, M.D., Director, Fetal Anesthesia Services

The role of the anesthesiologist is to maintain patient safety and comfort while facilitating the planned surgical procedure. Many puzzle pieces interact and influence the anesthetic plan, or "prescription." A small selection of items which become the puzzle pieces include the age and weight of the patient; the surgeon and their surgical approach (e.g., open thoracotomy vs. thoracoscopy); family history of anesthetic issues (such as malignant hyperthermia); the physiologic compromise caused by the lung lesion itself; and the physiologic derangement that occurs during the surgery itself. The anesthesiologist must fit many pieces together to come up with a sound anesthetic prescription, which includes management of the patient's airway, ventilation strategies to avoid further lung injury, fluid and possible blood administration, and postoperative analgesia.

Depending on the degree of compromise to the developing fetus or child, surgical interventions may occur at various times. Some cases may require intervention while the fetus is still developing. In these situations, an experienced anesthesia team cares for both the mother and the fetus simultaneously. Minimally invasive procedures may require light maternal sedation, but open fetal surgery, which may take place in the middle of gestation or near term, is a complex undertaking.[1]

Anesthesia for open mid-gestation procedures or near-term EXIT procedures involves deep general anesthesia for the mother. The anesthesia for open fetal surgery is about twice as deep as anesthesia for normal surgery. This serves to anesthetize the mother, to anesthetize the fetus, and to ensure adequate uterine relaxation for the operation. Hypotension and maternal pulmonary edema are known complications of these procedures. The anesthesiologist must balance these competing needs with meticulous fluid and blood pressure management, while preparing for possible (albeit quite rare) rapid maternal blood loss, and providing post-operative maternal analgesia with an epidural. During these cases, the fetus is also carefully monitored and anesthetized, and the fetus may receive fluids, blood and other medications for anesthesia and resuscitation.

The ability of a fetus to feel pain is a subject that will require continued study. What is clear, however, is that a fetus will demonstrate a stress response to surgical stimulation, stress responses are associated with poorer outcomes in premature infants, and these stress responses are attenuated with opioids. Our current strategy with opioid management in fetal surgery results in blood concentrations of opioids that are comparable to those of neonates undergoing major surgery in the postnatal period.[2]

Most children diagnosed with lung lesions do not require fetal intervention or immediate surgery at the time of delivery. These children will undergo surgery a few days to several weeks after birth.

The anesthesiologist must be comfortable taking care of neonates and small babies, with all of their attendant concerns. Some issues include peaceful induction of anesthesia, skillful intubation, careful glucose control, and familiarity with still maturing organ systems. Some surgical techniques require advanced management of ventilation, such as ventilating only one lung while the diseased lung is intentionally collapsed and made still to optimize surgical exposure.[3] The vast majority of patients are extubated in the operating room and recover in the neonatal intensive care unit for two to three days.

Postoperative analgesia may be quite a challenge for these patients. A comfortable child will have a better recovery, and a comfortable child will also help allay family anxiety. After thoracotomy, pain is often managed with a catheter, which is introduced into the epidural space via the sacrococcygeal ligament. The epidural catheter tip is advanced up to the thoracic vertebrae to cover the dermatomes involved in the incision. The epidural medications are chosen to maximize analgesia while minimizing side effects.[4] A special team of doctors and advanced practice nurses follows children with epidurals to come up with patient-specific plans to manage their pain and to help transition these children to intravenous and oral analgesics.

Surgical care for these patients is challenging as there are many variables that can change at any time: tumors may suddenly increase in size in the prenatal period, bradycardia may occur during an open fetal surgery, hypoxia may occur during a postnatal pulmonary lobectomy, or pain relief may become inadequate in the intensive care unit. The anesthesia team at CHOP is uniquely suited to care for these patients in all these stages. The surgical volume at CHOP is extraordinarily high by any standard, and high volume often makes for safer, more efficient care systems. To concentrate the anesthetic expertise, subsets of anesthesiologists within the department focus their care on certain patient populations (e.g., cardiac, neurosurgical, fetal, thoracic, craniofacial and orthopedic). This strategy further leverages the anesthesiologist's knowledge base and technical skills. We understand and can anticipate the changes that may occur, we know how these changes will impact the anesthesia plan, and we have the skills to safely and quickly respond.

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CASE STUDY: Thoracoscopic Lobectomy for Asymptomatic CCAM

By Alan W. Flake, M.D., Director, Center for Fetal Research

Congenital cystic adenomatoid malformation (CCAM) is a benign malformation of the lung that is usually diagnosed during the mid-gestational screening ultrasound as a "bright" lesion in the lung tissue that may or may not contain visible cysts. During prenatal life, CCAMs may grow rapidly until 25 to 28 weeks gestation, when they usually plateau in size, and then usually decrease in size relative to the fetus during the remainder of gestation.

Although a very large CCAM may cause heart failure (hydrops) in the fetus and require fetal intervention, the majority of CCAMs are asymptomatic at birth. In postnatal life, CCAMs usually present with infection, typically as difficult-to-treat pneumonias that are resistant to antibiotic treatment. They may also rupture a cyst, and present with lung collapse (pneumothorax). Finally, CCAMs have been documented to turn into malignant tumors over time. For these reasons, we recommend early resection of even asymptomatic CCAMs, typically prior to 2 months of age.

A 32-year-old woman was referred to CHOP at 23 weeks gestation for a cystic lung mass detected on routine screening obstetrical ultrasound examination at 21 weeks gestation. An ultrasound and MRI (figure 1) confirmed the presence of a right-sided, multicystic lung mass, with compression of the fetal heart to the left side and no other anomalies. She was followed by serial ultrasound assessment for the remainder of her pregnancy, during which the mass decreased in size relative to the fetus and the heart returned to a normal position. She was counseled that the infant would be asymptomatic at birth, and it was recommended that she deliver at a hospital close to home and that she return to CHOP for postnatal imaging of the lesion and treatment recommendations after 1 month of age.

As anticipated, the infant was asymptomatic after a normal vaginal delivery and continued to thrive at home until he returned to CHOP at 5 weeks of age. A CT angiogram study at that time confirmed the persistence of a right lower lobe multicystic CCAM (figure 2), and it was recommended that the baby undergo a thoracoscopic right lower lobectomy. At 6 weeks of age, the baby was taken to the operating room and underwent successful resection of the lesion using thoracoscopic technique (figure 3).

Thoracoscopic lobectomy in infants is an advanced, minimally invasive procedure that is performed through three 5mm trocars passed through the chest wall. High definition visualization and magnification is provided by a 4mm diameter thoracoscope and the dissection and ligation of the blood vessels and bronchus (airway) to the lobe is accomplished using small instruments passed through the two instrument ports. Thoracoscopic surgery offers the advantages of less pain and a potentially better long-term functional and cosmetic result. The infant was taken to our Harriet and Ronald Lassin Newborn/Infant Intensive Care Unit after surgery and was discharged home in normal condition on the second postoperative day.

At one year of follow-up, he has healed with minimal scar formation and is developing normally (figure 4).

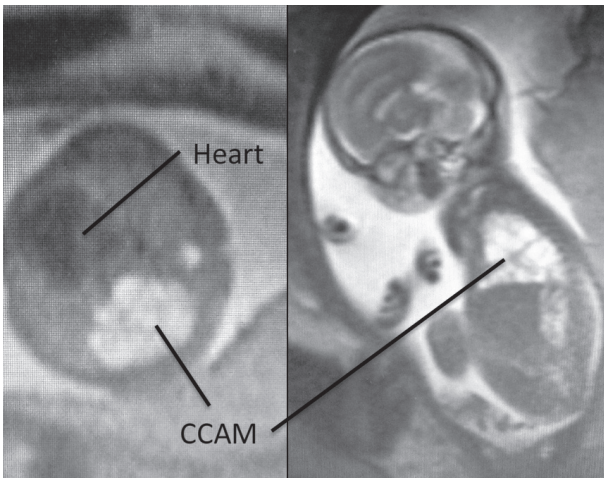

Figure 1. Fetal MRI images at 23 weeks gestation demonstrating a bright cystic lesion in the right lung.

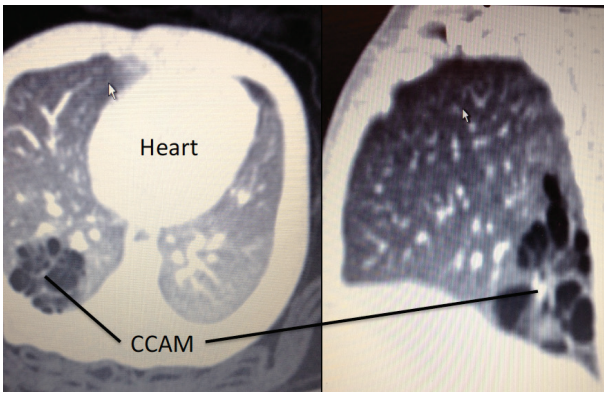

Figure 2. Postnatal CT scan images at 5 weeks of age demonstrating a decrease in relative size but persistence of the multicystic right lower lobe CCAM.

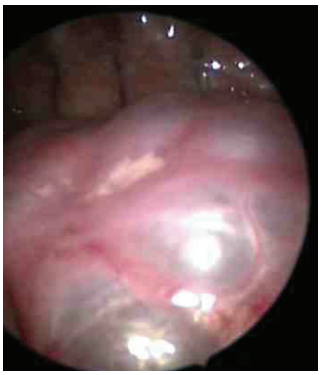

Figure 3. Thoracoscopic view of the right lower lobe CCAM.

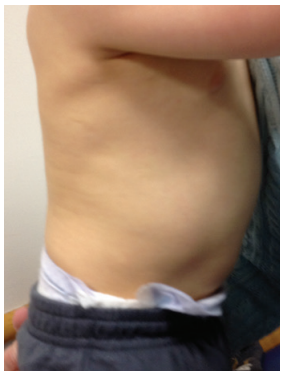

Figure 4. Appearance of the chest one year after thoracoscopic lobectomy.