BEYOND THERAPY

Livingcas, Lifelong Connection

In 1995, after a routine amniocentesis, Roberto's medical team revealed that he had DiGeorge syndrome—a rare condition that can cause a variety of health issues. The team at Children's Hospital of Philadelphia (CHOP) recommended that Roberto be delivered by a scheduled cesarean section and be evaluated by their team of experts immediately after birth. Throughout Roberto's journey, the team at CHOP provided a high level of care and support, allowing Roberto to grow and thrive.

As a result of this experience, we have learned that the natural history and prognosis of fetal lung masses can vary widely. Early diagnosis, careful monitoring, and prompt intervention when necessary can improve outcomes for these patients. We have also learned that the role of the anesthesiologist in fetal surgery is crucial, as they must balance the need for aseptic technique with the need for effective analgesia and sedation. Our team at CHOP is committed to continued research and innovation in this field to improve outcomes for patients and families affected by fetal lung masses.

RECENT PUBLICATIONS

Management of prenatally diagnosed congenital cardiac malformations


Lung Lesion Experience at CHOP: 1991 to 2014

An RARE LEVEL OF EXPERTISE IN LUNG CARE


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

The Children's Hospital of Philadelphia

Lung lesions are extremely rare, serious diagnoses; so rare that only a handful of centers in the U.S. and worldwide have managed more than 20 cases. CHOP is one of these centers and has managed more than 120 cases of fetal lung lesions, including congenital cystic adenomatoid malformation (CCAM), diaphragmatic hernia, biventricular pulmonary sequestration (BPS), and other rare conditions. Our team at CHOP has developed expertise in this field and offers a comprehensive approach to the diagnosis and management of these challenging cases.

NEWS FROM ANESTHESIA

Anesthesia for fetal surgery.


The ability of a fetus to feel pain is a subject that will require continued research and investigation. Our team at CHOP is committed to advancing our understanding of pain in the fetus and developing effective strategies for pain management in fetal surgery.

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Advancements in imaging of lung lesions, particularly for prenatal diagnosis and treatment, are crucial for understanding and managing congenital abnormalities of the fetal chest. This review focuses on the imaging techniques used to diagnose, monitor, and manage lung lesions in utero, with a special emphasis on the integration of prenatal and postnatal imaging. 

**Image Examples:**
- Image 1: A fetal magnetic resonance imaging (MRI) scan of a fetal chest with a lesion, showing the anatomical details with high soft tissue contrast.
- Image 2: A prenatal ultrasound image of a fetus with respiratory distress syndrome, highlighting the lung fields and chest wall.
- Image 3: A postnatal CT scan of a child with congenital diaphragmatic hernia, demonstrating the diaphragmatic defect and its associated anomalies.

**Key Points:**
- **Types of imaging:** Ultrasound, CT, MRI, and MR angiography.
- **Indications for imaging:** Prenatal diagnosis, monitoring growth and development, and postnatal evaluation of treated lesions.
- **Techniques for image guidance:** Intravenous iodinated contrast material for angiographic studies.
- **Advantages:** High-resolution imaging, multiplanar reconstruction, and the ability to visualize complex anatomical structures.
- **Challenges:** Artifacts, motion, and the need for comprehensive imaging protocols.

**References:**
- Hoffman GM, Lee W, Adzick NS. Fetal rat lung GFP reporter gene. [Image 1576x52 to 1686x146]
Lung Lesions

The Children's Hospital of Philadelphia

Providing Anesthesia for Patients with Congenital Lung Lesions

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

The Children’s Hospital of Philadelphia

In 1995, after a rescue mission performed, Dr. Lori J. Howell, R.N., M.S., was called to Jonathan’s delivery table. His parents, Roberto and Felicia, had traveled to CHOP from New York to have him delivered. They feared for his life and realized they needed to act quickly to save Jonathan’s future. That day, Jonathan’s story became the first-ever fetal surgery at CHOP; it lasted just 1½ hours and led to his successful birth.

Jonathan, now 17 years old and in his senior year of high school, is determined to do everything possible to save his unborn baby. He would never have thought his life would turn out so different from what his parents experienced. The Children’s Hospital of Philadelphia (CHOP) provides him with a diverse array of care that can prevent serious problems for both fetus and mother. Thanks to the support of Dr. N. Scott Adzick and CHOP, Jonathan and his family have learned that there was something terribly wrong: the child Felicia was carrying was a boy. They also learned that there was something unusually large: Jonathan’s lungs were twice the size of normal lungs. Once the baby was born, Jonathan underwent surgery a few days to several weeks after birth. Most children diagnosed with lung lesions do not require fetal surgery. The Children’s Hospital of Philadelphia manages these care needs through a collaborative partnership with the Center for Fetal Diagnosis and Treatment (CFDT). The CFDT is dedicated to providing comprehensive, multidisciplinary care for children who come to our Center and undergo open fetal surgery. The CFDT provides care before birth and cares for them after birth. We provide comprehensive care from diagnosis to treatment to management, the full range of fetal therapy techniques, and the option of delivering in the obstetric and neonatal intensive care unit.

Anesthesia for patients with congenital lung lesions can be challenging because many cases require intensive care after birth. The Children’s Hospital of Philadelphia has had experience with the anesthesia and care of patients with lung lesions.

**The Anesthesiologist’s Role**

Anesthesiologists provide care to patients with congenital lung lesions before and after birth. Anesthesiologists are responsible for monitoring and maintaining a patient’s vital signs, such as heart rate, blood pressure, and oxygen levels, throughout the entire care process.

**The Diagnostic Process**

Before birth, anesthesiologists work closely with other specialists, such as obstetricians, neonatologists, and cardiologists, to evaluate and manage a patient’s condition. During the diagnostic process, anesthesiologists may perform procedures such as echocardiograms, cardiac catheterizations, and magnetic resonance imaging (MRI) to assess a patient’s health.

**The Surgical Procedure**

During the surgical procedure, anesthesiologists work closely with surgical teams to ensure a safe and successful operation. They monitor vital signs, administer medications, and adjust the patient’s condition as needed. Anesthesiologists also work with the surgical team to minimize pain and discomfort for the patient.

**Postoperative Care**

After the surgical procedure, anesthesiologists continue to monitor a patient’s vital signs and adjust medications as needed. They also work with other specialists to ensure a smooth transition to recovery.

**Conclusion**

Anesthesiologists play a critical role in the care of patients with congenital lung lesions. They work closely with other specialists to monitor and manage a patient’s condition throughout the entire care process, from diagnosis to treatment to management.

**References**


The high volume of patients we see from around the world with incredibly complex, rare conditions pushes us to advance the field of fetal surgery and current techniques in neonatal resuscitation. Mahboubi S.

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ADVANCES IN IMAGING

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

without hydrops or major lung compression, will likely be delivered vaginally or via cesarean section (for obstetric indications) and placement of an IV to deliver blood or medications. For fetuses with solid lesions with a CVR>1.6, maternal placement of an IV is imperative.

For fetuses that have macrocystic CCAMs and hydrops, a fetal ultrasound is used to assess the CCAM volume ratio (CVR) and plan the timing of delivery. A CVR of 1.6 or greater indicates a higher likelihood of hydrops developing. If the CVR is less than 1.6, the baby may be able to compensate for the lesion and may not develop hydrops. Now, in many cases, we are able to predict the outcome by measuring the CVR and correlating it with the size and growth of the mass.

The CCAM volume ratio (CVR) is calculated by dividing the volume of the CCAM by the volume of the fetal thorax.

To calculate the CVR, we use the following formula:

\[
\text{CVR} = \frac{\text{Volume of CCAM}}{\text{Volume of thorax}}
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For example, if the volume of the CCAM is 200 cm³ and the volume of the thorax is 1500 cm³, the CVR is 0.13.

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RECENT PUBLICATIONS

Management of prenatally diagnosed congenital diaphragmatic hernia

Semin Pediatr Surg.

Khalek N, Johnson MP.

Management of prenatally diagnosed congenital diaphragmatic hernia.

The Children’s Hospital of Philadelphia

IN UTERO

LUNG LESIONS

Anesthesiology.

Pediatr Anaesth.

Rakowski L.

J Obstet Gynecol Neonatal

Gregory CL, Wright J, Schwarz J, Rodriguez and her husband, Roberto, learned that they were pregnant with a baby that had a lung lesion. The lesion was discovered during a routine ultrasound at 18 weeks gestation.

They were referred to the Children’s Hospital of Philadelphia (CHOP) for further evaluation. Dr. Jack Rinehart, a neonatologist at CHOP, reviewed the ultrasound results and recommended that they proceed with a fetal surgery.

Dr. Rinehart explained that fetal surgery is a complex procedure that involves opening the uterus to remove a lesion from the lungs of the fetus. The procedure is only performed at a select number of hospitals worldwide, and it carries a significant risk.

The surgery was scheduled for 22 weeks gestation, and Dr. Rinehart’s team worked closely with the family to prepare them for the procedure.

During the surgery, the anesthesiologists carefully monitored the mother’s vital signs and administered appropriate medications to ensure her safety. The surgery was completed successfully, and the lesion was removed from the fetus.

After the surgery, the family was kept informed of the baby’s condition and received support from the medical team. They were encouraged to stay in touch with the hospital and to continue close monitoring of the baby’s health.

The baby was born weighing 6.5 pounds, with a birth weight that was consistent with their gestational age. The baby was placed on a ventilator to help with breathing, and the medical team worked diligently to ensure their safety.

It was the first-ever fetal surgery at CHOP; it lasted just 1½ hours and took place in the middle of gestation or near term, is a complex procedure that involves opening the uterus to remove a lesion from the lungs of the fetus. The procedure is only performed at a select number of hospitals worldwide, and it carries a significant risk.

The role of the anesthesiologist is to maintain patient safety and stability throughout the operation. Hypotension and maternal pulmonary edema are common complications that can occur during fetal surgery. The anesthesiologist must be prepared to manage these situations, an experienced anesthesia team cares for both the mother and neonate simultaneously.

Monitoring during fetal surgery is critical, as the anesthesia plan must be adapted to the unique needs of the mother and fetus. The anesthesiologist must be able to anticipate changes that may occur during the surgery and adjust the anesthesia plan accordingly.

Intraoperative blood administration and regional anesthesia techniques are often utilized during fetal surgery to minimize blood loss and reduce the risk of complications. The anesthesiologist must be skilled in these techniques to ensure the safety of both the mother and fetus.

Postoperative analgesia is also critical following fetal surgery. The anesthesiologist must be able to manage the mother’s pain and ensure a smooth recovery.

The Children’s Hospital of Philadelphia (CHOP) has managed the care of more than 1,000 pregnancies diagnosed with lung lesions — the largest volume of any center in the world. Through this experience, we have learned that the natural history and treatment outcomes for lung lesions can be complex and varied.

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In summary, fetal surgery for lung lesions is a complex and challenging procedure that requires a multidisciplinary approach. The Children’s Hospital of Philadelphia (CHOP) is uniquely suited to care for these patients and we welcome your feedback.

If you have any questions or would like more information about fetal surgery for lung lesions, please contact us at fetalsurgery.chop.edu/fetal-ed

Open Fetal Surgeries .........................................................24

Lung Lesion Experience at CHOP:

1995 to 2014

Table: Lung Lesion Experience at CHOP:

<table>
<thead>
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If you have any questions or would like more information about fetal surgery for lung lesions, please contact us at fetalsurgery.chop.edu/fetal-ed
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).