

Lung Lesions

Congenital Cystic Adenomatoid Malformation (CCAM) and Congenital Lobar Emphysema and Pulmonary Sequestration

What are Congenital Cystic Adenomatoid Malformation (CCAM), Pulmonary Sequestration and Congenital Lobar Emphysema?

These fetal conditions are seen by the prenatal ultrasound and they are masses within the fetal lung and chest. They are considered to be within a broad category of lung abnormalities, called congenital pulmonary airway malformations (CPAM).

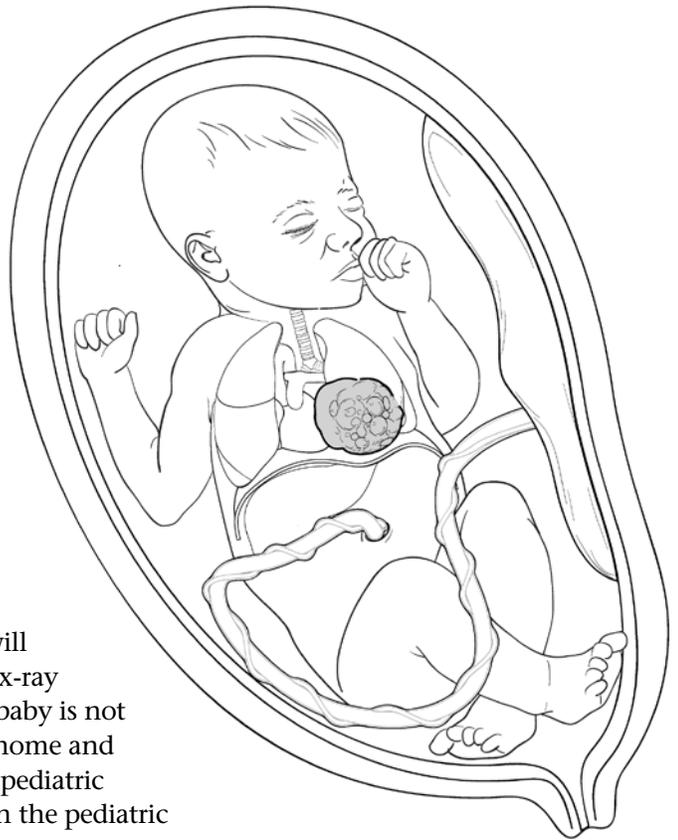
Typically, the prenatal ultrasound around 16 to 22 weeks by the obstetrician has found a mass located in your unborn baby's chest. The mass may be fluid filled or a solid mass. Color flow doppler ultrasound may be used to more accurately determine the characteristics of the mass. Sometimes a fetal MRI is done to confirm the diagnosis. Approximately 10-20 percent of lung lesion cases will shrink in size while the mother is pregnant. Sometimes a small lung mass can go undiscovered until later in childhood and it can cause an infection, such as pneumonia.

Causes:

The exact cause of congenital pulmonary airway malformations is unknown. The mass in the chest can cause poor lung growth (hypoplasia), and heart and breathing problems at birth. The survival rate is approximately 90 percent, as long as the fetus does not develop hydrops. Hydrops is retention of fluid in the body causing severe complications. The high risk obstetrician will closely monitor the fetus with frequent ultrasounds for any signs of hydrops and other complications. There is not an increased association of chromosomal anomalies with this condition.

What happens after your baby is born?

Following birth, the baby will be closely monitored by a neonatologist (a doctor who specializes in newborns). The neonatologist will make sure the baby is breathing properly. An x-ray will be done to check the baby's lungs. If the baby is not having breathing problems, the baby can go home and follow up later at a few weeks of age with the pediatric surgeon. It is very important to follow up with the pediatric



surgeon during this time. Typically a CT scan will be done around one month of age. The pediatric surgeon will decide when surgery is appropriate. It is important that the mass be surgically removed by a pediatric surgeon because of the risks of infection during later childhood and the potential for cancer in the lung lesion later in life if it is not removed. (See Treatment of Lung Lesions)

If the baby is having breathing problems at birth a ventilator will be needed to help support your baby's lungs. Your baby will then be transferred to Nationwide Children's Hospital neonatal intensive care unit (NICU) for a complete evaluation.

An intravenous line (IV) will be started in your baby's arm, hand, foot or scalp. The IV provides fluids and antibiotics to prevent dehydration and infection. A special intravenous line called an umbilical (belly button) catheter will be started. An umbilical catheter is a long plastic tube that is placed in the umbilical artery and umbilical vein. The catheter provides IV fluids and medications and monitors your baby's blood pressure. This special catheter typically only stays in place for up to two weeks. A tube will be put into your baby's nose or mouth. It will be either an NG tube (nasogastric) or an OG tube (orogastric). This tube goes into your baby's stomach and helps to keep the stomach empty.

Treatment of lung lesions:

The mass will need to be surgically removed by a pediatric surgeon. The timing of the surgery depends on how the baby is breathing at birth. If the baby is having breathing difficulties, the surgery will occur shortly after birth and before hospital discharge. If the baby is having no breathing difficulties, the surgery may happen later in infancy and the baby can be discharged from the hospital at the same time as the mother.

The pediatric surgeon will surgically remove the mass that has been located in your infant's lung. The surgery removes only the portion of the lung that is involved with the mass and aims to preserve the entire normal lung.

If the lesion is moderate to large in size and the baby's lungs are not working effectively, the baby may require special treatment called extracorporeal membrane oxygenation (ECMO), which is a specialized procedure that supports the lungs and the heart. During the prenatal consultation, the pediatric surgeon and neonatologist will talk to you about ECMO.

The baby will not be able drink formula or breast milk until after surgery. A special IV called a central venous catheter (CVL) will be used to provide IV nutrition and give medicines. It will stay in place until IV nutrition is not needed anymore. The catheter will provide total parenteral nutrition (TPN) which has all of the calories and nourishment needed for your baby to grow.

What happens after surgery?

After surgery, your baby will continue to be on a ventilator to help him or her breathe. The ventilator will be removed when your baby has recovered from surgery and the lungs are stable, which may take days or weeks. The baby may have a chest tube after surgery. This tube helps drain fluid from the incision. The chest tube will stay in place for a couple of days. Babies recovering from surgery are very sensitive to noise and pain. The babies may be easily agitated. The medical staff will give medicines to help reduce any pain or discomfort your baby may be having.

Feedings of formula or breast milk will be started in small amounts and increased gradually over time. When the baby is taking enough formula or breast milk to grow and gain weight, the IV will be removed.

The length of hospital stay depends on how sick the baby is and how well the lungs are working.

Frequently Asked Questions

What is the survival rate?

Survival rate is as high as 90 percent when fetal hydrops is not discovered. Hydrops is retention of fluid in the body causing severe complications.

Will my baby have normal lungs?

Nearly all babies have normal lung function after surgery. This means that they will be able to play sports and do all typical childhood activities without any restrictions. If the baby had ECMO treatment, there is the possibility that oxygen supplementation may be necessary in the postoperative period. Some babies who had ECMO need to receive oxygen at home for a period of several weeks or months.

Will my baby have asthma?

Asthma is a common condition of infancy and childhood that affects many otherwise healthy children. Having surgery for CPAM does not increase the chance of asthma in your child.

When can my baby go home?

Your baby can go home when he or she is taking enough formula or breast milk to grow and gain weight.

How long will my baby have to stay in the hospital?

The average length of hospital stay is two to three weeks to months. Some babies may have complications and need to stay longer.

What other tests are done on my baby?

The doctor is going to schedule an echocardiogram (scan) of the heart, kidneys and head. Also, your baby will have blood drawn to monitor lab values. The medical staff will talk with you regarding the results.

Is it okay to have a vaginal delivery?

It depends on the size of the lung mass. Your obstetrician will talk to you about what is the safest way to deliver your baby.

Where do I need to deliver my baby?

Your high risk doctor will let you know the Columbus hospital where your delivery will be scheduled.

I live out of town. Where can I stay?

Nationwide Children's Hospital has a wonderful Ronald McDonald's House where families who live out of town may stay while their child is in the hospital. Ask the Nationwide Children's Hospital program coordinator if this can be arranged for you.

How will all of this get coordinated?

Nationwide Children's Hospital has a program coordinator to help with these arrangements. You may contact Becky Corbitt, RN, BSN, at (614) 722-6520 any time during your pregnancy if you have questions or concerns.

When can I hold my baby?

This is a difficult time for most parents. Talk with the doctor or nurse to see when you can hold your baby.

How can I get involved with my baby's care?

The health care team at Nationwide Children's Hospital encourages family centered care. If you want to speak with any members of the team, please ask. The staff is here for you and we want to help you to be involved with daily care of your baby.

Where do I follow up once my baby is discharged from the hospital?

You will want to find a local pediatrician to take care of all the routine checkups, immunizations and doctor's visits. The program coordinator can help if needed. A pediatric surgeon will follow up regarding your baby's surgical needs.