Congenital Pulmonary Airway Malformation (CPAM)

also known as

Congenital Cystic Adenomatoid Malformation (CCAM)

What is a CPAM?

A congenital pulmonary airway malformation (CPAM) is an abnormally formed piece of lung made up of closed sacs (cysts) that will never function as normal lung tissue . A CPAM usually involves only one lobe of a lung. We do not know what causes CPAM, but CPAM occurs in about 1 in 10,000 fetuses. CPAM was previously called congenital cystic adenomatoid malformation (CCAM).

On ultrasound examination a CPAM may appear macrocystic, microcystic, or a mixture of the two. A macrocystic CPAM has one or more large cysts (>= 5mm) that appear as empty spaces or holes in the lung . A microcystic CPAM has very small cysts (< 5mm) giving it the appearance of a solid well-defined mass that is brighter (more white) than the surrounding normal lung.

Sometimes, a CPAM may be seen with heart abnormalities or other birth defects. This is not common, but if it does occur, a test called an amniocentesis may sometimes be offered for further evaluation.

Effect on Pregnancy

- Most babies with CPAMs do well., and most CPAMs are expected to remain the same size or shrink.
- Sometimes pressure from a CPAM compresses the baby's throat and prevents the baby from swallowing amniotic fluid. This allows the level of fluid around the baby to get very high, a condition called polyhydramnios.
- If the CPAM grows large enough to put pressure on the baby's heart and blood vessels, it can lead to heart failure causing fluid to collect in the skin (edema) , the abdomen (ascites) , and chest (effusions). This condition is called fetal hydrops. Hydrops in the baby can cause fetal death, and in extreme cases cause the mother to develop edema and high blood pressue a situation where the mother's condition "mirrors" the baby's condition
- Once in awhile CPAMs may grow large enough to prevent the surrounding lungs from developing l(lung hypoplasia) .

Management

Your baby will be evaluated frequently between 20 and 28 weeks for growth of the CPAM, because CPAMS grow most quickly during that time. During your ultrasound examinations the size of the CPAM will be compared to the size of your baby's head to calculate a number called the Congenital Pulmonary Airway Malformation Volume Ratio (CVR) . The value of the CVR tells your doctor how likely your baby is to develop fetal hydrops . If the CVR remains less than 1.0 after 28 weeks, then the ultrasound examinations can be performed every three to four weeks until delivery.

A CVR greater than 1.6 suggests the risk of hydrops developing is high. Baby's with a CVR > 1.6 are seen two to three times per week. You may be given:

Betamethasone 12 mg intramuscularly every 24 hours for 2 doses to slow the growth of the CPAM,

Babies less than 30 weeks with signs of possible hydrops such as fluid in abdomen, around lungs, or heart, or skin edema may be referred for fetal surgery to either drain the cyst (s) or to remove the mass. However, if the baby is old enough it may be considered for early delivery and removal of the CPAM at birth using the ex utero intrapartum treatment (EXIT) procedure . During the EXIT procedure the baby is partially or completely delivered by cesarean section. However, the umbilical cord is not clamped and the placenta remains in place. The baby continues to exchange gases through the placenta while the surgeon removes the mass and secures the baby's airway before the baby is delivered.

Delivery

Except in cases where the baby's or mother's health is threatened early delivery is not usually recommended.

The baby should be delivered at a hospital with a neonatal intensive care unit and pediatric surgery services available. If there is a high chance that the other lung in the baby is very underdeveloped, then delivery at a center that can provide Extracorporeal Membrane Oxygenation (ECMO) is also recommended.

After Delivery

If your baby has symptoms due to the CPAM after delivery, the CPAM will be removed before your baby leaves the hospital. Babies who do not have any symptoms at birth can go home with the mother, after they have been evaluated.

Many surgeons recommend a CPAM be removed by one year of age to prevent infection or possible cancer developing in the CPAM later in life.