

Questions and Answers About:

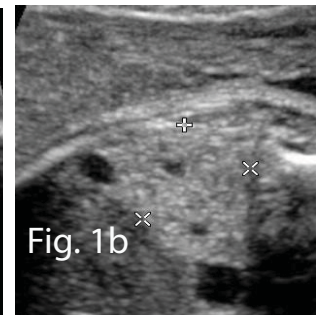
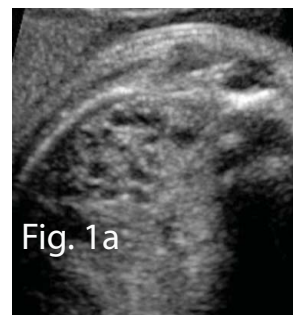
Congenital Cystic Adenomatoid Malformation (CCAM)

(More correctly known as congenital pulmonary airway malformation or CPAM)

CPAMS

Congenital pulmonary airway malformations or CPAMs are non-cancerous (non-malignant) growths of abnormally developed lung tissue occur within the normal tissue of the lung. They were previously congenital cystic adenomatoid malformations (CCAMs). We don't know the exact rate of occurrence, but we see 5-10 cases a year and estimate the frequency to be about 1-2/5000 live births. These abnormal lung masses may involve either the right or the left lung and usually replace much of one lobe (one of the separate sections) of the right or left lung. This abnormally formed lung tissue never functions properly and increases the risk of pneumonia or other lung infection. A CPAM can also cause pneumonia and other lung infections as a child gets older. Rarely, a CPAM can turn cancerous later in childhood. To prevent these potentially serious complications, CPAMs should be surgically removed before a baby's first birthday.

Other cystic (a cyst is a fluid-filled structure) masses can look much like a CPAM on prenatal ultrasound, chest X-ray, and rarely, even on chest CT scan. For example, a lung sequestration, another cystic lung mass of abnormally developed lung tissue, usually possesses a different blood supply from the rest of the lung and does not communicate



Figures 1a & 1 b. CCAM imaged by ultrasound in the second trimester. The CCAM lies toward the spine in the right lower lobe of the lung. The baby is laying with the spine at the top of the image. The large cysts make this a type I CCAM.

with the main breathing tubes (bronchi). Ultrasound cannot always tell the difference between CPAM and a lung sequestration. In fact, cystic lung masses often contain elements of both a CPAM and a lung sequestration. The treatment is the same for both problems, surgical removal of the abnormal lung tissue. Final diagnosis of CPAM vs. lung sequestration may not occur until the removed lung tissue is examined in the pathology lab and under a microscope.

How Is CPAM Diagnosed?

Your primary doctor usually diagnoses CPAM (and other cystic lung masses) by prenatal ultrasound (Figures 1a & 1b). A prenatal

CCAM continued...

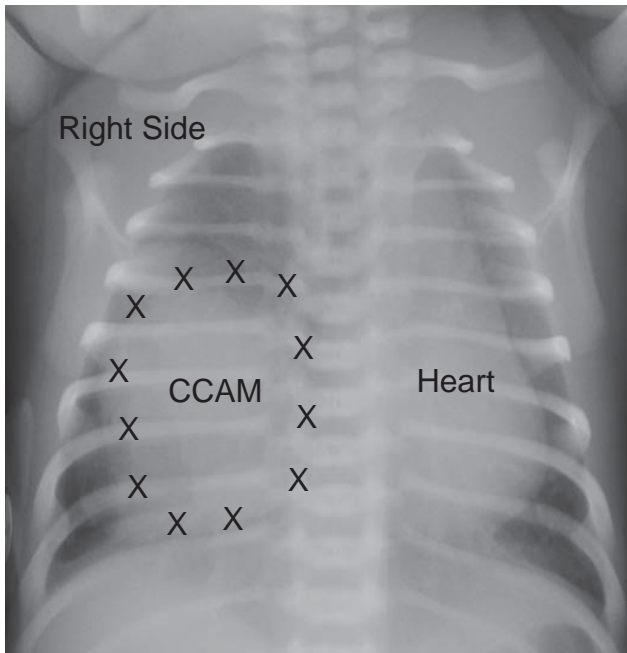


Figure 2. Chest X-ray of CPAM taken shortly after birth. The CCAM is on the infant's right side and is pushing the heart to the right side of the chest.

MRI may also be needed to assess cystic lung masses. Many women require several ultrasounds during pregnancy to monitor the size of the cystic mass and to watch for complications. After birth, your baby's doctor should assess your baby's breathing and obtain a chest X-ray (Figure 2). We have seen many children who were diagnosed before delivery with a CPAM, but after birth had normal breathing and a normal chest X-ray. This doesn't mean the CPAM has disappeared, just that it does not appear on a routine X-ray. We and others recommend that even with a normal chest X-ray immediately after birth, your baby should receive a CT scan of the chest to confirm the presence or absence of the lung mass. If left alone, most CPAMs will cause significant problems during childhood.

The CT scan also identifies the size and exact location of the CPAM or other cystic lung mass. The best time to do the CT scan is 12-16 weeks after birth. The CPAM (or other cystic lung mass), can always be seen on the chest CT scan.

CPAMs are categorized into four types based on the size of the cysts in the mass. Only the first two types are commonly seen:

- Type I (70% of cases) is characterized by one or several large cysts (>2 cm in diameter). These CPAMs seem to have the highest potential for becoming cancerous in later life.
- Type II (20% of cases) has smaller cysts (< 2 cm in diameter).
- Type III has very small cysts (but often appears solid on ultrasound or X-ray).
- Type IV is a combination of small and large cysts.

The exact histological type of CPAM is not usually important in determining the best treatment course. They should all be removed by 12 months after birth.

What Does This Mean for My Baby?

The outcome of babies with a CPAM (or other cystic lung mass) depends on:

1. Size of the mass. The larger the cystic lung mass, the less normal, functional lung tissue is available to the baby after birth. Small masses don't cause breathing problems. Even a large CPAM, as seen on early ultrasounds may shrink in size after about 25 weeks gestation. Occasionally, we have seen a large CPAM push the heart toward the other side of the chest in the second trimester, but then decrease in size, causing no problems at birth. The majority (>65%) of babies with cystic lung masses have no breathing problems at birth.

Large CPAMs can reduce the amount of normal lung tissue and even compress the major blood vessels leading to the heart. When this occurs, the fetus can develop a form of heart failure and accumulate extra fluid in the tissues (edema), chest cavity (hydrothorax), or abdominal cavity (ascites).

CCAM continued...

Physicians call this condition “hydrops.” Hydrops can lead to fetal death, or death soon after delivery. Babies with hydrops are candidates for fetal surgery, since they rarely improve before birth without intervention.

Large masses can also compress the esophagus. This prevents the fetus from swallowing amniotic fluid normally and leads to the accumulation of excess amniotic fluid around the baby (polyhydramnios). Polyhydramnios often leads to preterm delivery because it stretches the uterus, causing uterine contractions.

2. Pulmonary hypoplasia (undergrowth of the lungs). A large CPAM can interfere with the growth and development of the lungs resulting in smaller than normal lungs. When the lungs are small compared to the rest of the body, high blood pressure in the lungs often occurs as the heart tries to pump the normal amount of blood through lung blood vessels that are too small. This is similar to connecting a fire hose to a garden hose. The smaller diameter tube requires a much higher pressure to move the same amount of fluid—in this case, blood, over the same time. High blood pressure in the lungs is also called pulmonary hypertension. Small lungs and pulmonary hypertension can be life threatening and require admission to a NICU and specialized treatments, such as a ventilator.
3. Other birth defects. Occasionally, CPAMs may be associated with kidney cysts or other birth defects. This is unusual, but if it does occur, it complicates the infant’s course.

Will My Baby Survive?

Almost all babies with CPAMs survive and do well as long as there is no hydrops prior to birth. We have an overall survival rate of 95% for patients with CPAMs.

What Does This Mean for the Mother?

Most often, a CPAM just requires extra ultrasounds to assess the size and impact of the CPAM on the fetus. When a baby develops

hydrops (abnormal fluid collections in the skin, abdomen, or chest due to heart failure in the fetus) because of a large CPAM, the mother can also retain fluid and develop high blood pressure. This situation is called the Mirror Syndrome because the mother’s condition “mirrors” that of her fetus. Women can become quite ill with Mirror Syndrome. Although this condition is rare, it is another reason why the obstetrician asks for frequent follow-up visits and ultrasounds. Swelling of the placenta is often a warning sign that Mirror Syndrome is developing.

How Is a CPAM/Lung Mass Treated?

Prenatal treatment is rarely indicated and should be undertaken only when hydrops develops or appears imminent. Draining fluid from the cyst or from around the lungs may help in these cases. In a few centers, removal of the CPAM by fetal surgery has been undertaken, but remains an unproven therapy. In cases that are not treated with fetal surgery, administering steroid medication to the mothers can sometimes shrink the CPAM. This does not prevent the need for eventual surgery, but may improve survival in the newborn period.

CPAMs identified on prenatal ultrasound that do not disappear (by chest CT scan 12-16 weeks after birth) should be removed surgically. Approximately 20% of babies will have lung symptoms need surgery within the first week of life, and most will have surgery within the first year of life. The timing of surgery varies by the size of the mass and severity of respiratory symptoms in the baby. With a large CPAM, surgical removal usually occurs in the first few months after birth.

Typically, surgical removal of the CPAM or other mass occurs through a surgical incision on the side of the chest and open-chest surgery. More recently, surgeons have used a scope through a small incision (like laparoscopic surgery) depending on the size and location of the chest mass. The results of surgical removal of the CPAM have been very good.

CCAM continued....

Will My Baby Be in a NICU?

Babies with a CPAM and no symptoms can usually be admitted to the normal newborn nursery. Babies with symptoms or large CPAMs are admitted to a Level II (mild to moderate symptoms) or a Level III NICU (severe symptoms). In the NICU, babies are monitored for abnormal heart rate, respiratory rate, oxygen levels and other potential problems.

Will My Baby Need to Be on a Ventilator?

Approximately 35% of infants need a ventilator and extra oxygen from shortly after birth until after surgery. Another 10% will need oxygen, but not a ventilator during their hospitalization.

Why Did This Happen?

The cause of CPAMs and other cystic lung masses remains unknown. Other birth defects rarely occur in patients with cystic lung lesions, and there are no known genetic causes. Recurrence in subsequent pregnancies has not been reported. CPAMs and lung sequestrations usually are not connected to the main bronchial (breathing) tubes. Whether this is the cause or the effect of the abnormal lung development is not known.

Is There an Advantage to Preterm Delivery by Cesarean Section?

Parents often ask if there is any advantage to being born early or by cesarean section. As long as the baby is growing and developing appropriately there is no reason to deliver the baby early. However, if the baby has worsening hydrops or other complications, preterm delivery may be required. There is no advantage to cesarean delivery unless there is hydrops or other complication that prevents a safe vaginal delivery.

How Long Will My Baby Be in the Hospital?

Many babies diagnosed with CPAM do not show any symptoms at birth and can go home with the mother. These babies need no special evaluation or treatment in the newborn period, other than a chest X-ray. If the baby can go home with the mother, surgery to remove the

CPAM is usually scheduled for 3-4 months of age. This hospitalization lasts about 3-5 days.

Those babies who do require assistance with their breathing or who have masses so large that they cause hydrops will need to stay in the hospital until those problems resolve. Usually, this requires removal of the mass shortly after birth. If the lungs are small or there is pulmonary hypertension, the baby may also require other specialized treatments in addition to a ventilator. The average length of stay is 12 days, but can be shorter or much longer depending on how ill the baby is at birth. Babies with hydrops can be in the hospital for many weeks.

What is the Long-Term Outcome?

All children who have part of their lung removed have decreased lung function. However, for most children this decrease is minimal and can be seen only with sophisticated tests of lung function. From a practical standpoint, their lung function rarely limits their physical activity. Patients with large CPAMs, hydrops, or a requirement for extensive treatment in the newborn period have an increased risk of more serious long-term problems. These infants may have problems with growth, asthma, and increased respiratory infections during childhood. Each case is different. Your child's doctor will be able to make a better prediction after birth than he/she can prior to delivery.

If you have questions at any time, please contact us (612-813-6288 or 612-813-6295).