

## **Congenital Cystic Adenomatoid Malformation of the Lungs (CCAM)**

### **What is Congenital Cystic Adenomatoid Malformation of the Lungs?**

The bronchioles are very small breathing channels found in the lungs. There are about 70,000 within each lung.

Congenital Cystic Adenomatoid Malformation (CCAM) is a type of benign tumor made up of an overgrowth of bronchioles. Benign tumors cannot spread to other parts of the body and remain in the same area. It is not a cancerous tumor. The area of the lung affected, does not work like normal lung tissue.

CCAM is divided into 3 major types. Type I (macrocytic) is the most common and is usually made up of 1 – 4 cysts measuring 2-10 cm in size. Type II (mixed) cysts are smaller in size, less than 2 cm and are larger in number. Type III (microcystic) is the least common and is made up of very small cysts and appears solid on ultrasound.

CCAM can often be identified during a prenatal ultrasound at 18-20 weeks of pregnancy, but some cases are not found until later in pregnancy or after birth.

### **What are the causes of CCAM?**

The cause of CCAM is unknown. It appears to be a random event that can happen to anyone.

### **What does this mean?**

Congenital Cystic Adenomatoid Malformations can vary in size and can change in size throughout pregnancy. Some may grow with the baby and appear quite large. It may even push the baby's heart out of the usual position. Some may remain the same size as the baby continues to grow and the CCAM becomes relatively small. Some may even shrink or disappear before birth. In all these cases, the outlook for a normal life is excellent.

Rarely, a CCAM grows very large. This large CCAM can put extra pressure on the baby's esophagus (the tube between the mouth and stomach), lungs or heart. Pressure on the esophagus that can prevent the baby from swallowing the amniotic fluid (the water around the baby) as it normally does. This results in a build up of fluid around the baby called polyhydramnios.

With a very large CCAM, the lungs may not have enough space to develop properly. This is called pulmonary hypoplasia. This can be a life threatening illness as it can cause breathing problems for the baby after birth.

A large CCAM can also push the baby's heart so far out of its normal position that it doesn't have enough room to work properly. This can cause heart failure and fetal hydrops can develop. Fetal hydrops is an abnormal collection of fluid in the baby's skin, chest or belly. Fetal surgery (removing the CCAM while the baby is still in the mother's uterus) may be needed for a baby with hydrops. This is done at a specialized hospital. Babies with this condition are very sick and may not survive. Mothers of babies who have hydrops are at risk of developing high blood pressure and need close monitoring. A large CCAM that is fluid filled may be able to be drained or emptied with a needle or special catheter called a shunt.

It is important to remember that most babies do not require any procedures during pregnancy.

### **What other tests should we consider?**

Other tests may include a detailed ultrasound and a fetal heart ultrasound (echocardiogram). The detailed ultrasound is used to assess the baby's other organs. A referral to a paediatric surgeon (specialist in surgery of children) will be made to discuss with you the type of surgery the baby may need after birth.

Ongoing ultrasounds are used to monitor the growth of the CCAM and to look for early signs of hydrops or polyhydramnios.

### **What will happen around the time of the baby's birth?**

Babies with CCAM may need to be born at a hospital with a neonatologist (specialist in newborn babies) and a pediatric surgeon (specialist in surgery of children). Babies with CCAM may require care in a neonatal intensive care unit. Some babies may not require this increased level of care and will be able to deliver at your local hospital. Your doctor will be able to tell you where your baby needs to be delivered.

### **What does this mean for my baby's future?**

Most babies do very well after surgery and continue to develop normally. The long-term outcome for babies with CCAM is excellent. These children appear to grow as well as other healthy children and do not experience an increase in lung infections.

### **What do we do now?**

You will meet with an obstetrician that specializes in high-risk obstetrics. The doctor will discuss CCAM with you in further detail.

## **Where can I get more information?**

The Children's Hospital of Philadelphia  
Center for Fetal Diagnosis and Treatment: Congenital Cystic Adenomatoid Malformation (CCAM)

<http://www.chop.edu/service/fetal-diagnosis-and-treatment/fetal-diagnoses/bronchopulmonary-sequestration-and-congenital-cystic-adenomatoid-malformation-ccam.html>

University of California

The Fetal Treatment Center: Congenital Cystic Adenomatoid Malformation

<http://fetus.ucsfmedicalcenter.org/ccam/>

Cincinnati Children's

Fetal Care Center of Cincinnati – CCAM/CPAM Overview

<http://www.cincinnatichildrens.org/service/f/fetal-care/conditions/ccam/default/>