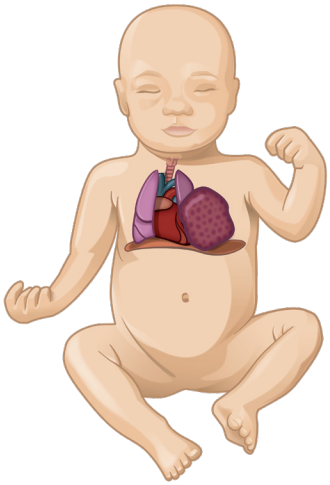
Congenital Pulmonary Airway Malformation

What is Congenital Pulmonary Airway Malformation (CPAM)?

A CPAM (formerly referred to as a CCAM or Congenital Cystic Adenomatoid Malformation) is a cystic mass which forms in the lung tissue of a fetus. The mass is usually located in one lung, and it does not function as normal lung tissue. The cause of a CPAM is unknown, and it is not related to anything the mother did or did not do during the pregnancy. The CPAM is made of somewhat immature, fast-growing lung cells. The size of the CPAM is often such that a normal lung cannot grow well. It may also cause the heart to shift to the opposite side of the chest or push downward on the baby's diaphragm.

How is CPAM diagnosed?

CPAMs are detected during a routine prenatal ultrasound. At the SSM Health Cardinal Glennon St. Louis Fetal Care Institute the diagnosis can be confirmed with a fetal magnetic resonance imaging (MRI) exam which will also document the size and location of the CPAM. Another test that may be recommended is a fetal echocardiogram (echo). This is an ultrasound of the heart performed by a pediatric cardiologist. This test is recommended to rule out structural heart defects and assess heart function.



Baby With A Congenital Pulmonary Airway Malformation (CPAM)

A Cardinal Glennon St. Louis Fetal Care Institute nurse is available 24 hours a day, seven days a week to discuss referrals with physicians and potential families by calling 314-268-4037, option 2.

Phone	**314-268-4037, option 2**
Toll-free	**1-877-SSM-FETL (776-3385)**
Web	**stlouisfetalcare.com**
Email	**fetalcare@ssmhealth.com**
Facebook	**facebook.com/fetaldocs**

SSM Health Cardinal Glennon
St. Louis Fetal Care Institute®

Congenital Pulmonary Airway Malformation (CPAM)

How does CPAM affect my baby?

The vast majority of babies do well and have normal development and lung function. During the pregnancy, the cyst often grows with the fetus and appears quite large; however, the growth usually starts to slow down in the second trimester. Since the fetus and the remaining normal lung continue to grow rapidly, the CPAM appears to shrink over the pregnancy. Sometimes, the CPAM becomes very small and even undetectable by ultrasound before birth. It is always there, but studies will have to be done after birth to find the CPAM. In all these cases, the outlook for a normal life is excellent.

In a small number of fetuses, the CPAM may grow so rapidly that it becomes life-threatening before birth. This usually happens between 18 and 26 weeks gestation. The large size of the CPAM causes compression of the heart and heart failure. Compression of the lungs can also cause the lungs to be too small for survival. In these cases, either fetal surgery or early delivery may need to be performed depending on how far the pregnancy has progressed.

How is CPAM managed during pregnancy?

During the initial ultrasound, we will measure the volume of the CPAM relative to the size of the fetus. This ratio is called the CPAM volume ratio, or CVR. For every fetus with a CPAM, we start by measuring the CVR every week. By comparing the CVR measurements, we can determine how fast the CPAM is growing and whether it will become life-threatening. If the CVR remains small (less than 1.0) after 28 weeks, then the ultrasound examinations can be performed every three to four weeks until delivery.

If the CPAM pushes the heart out of the normal position, then a second MRI may be performed around 32-34 weeks of pregnancy. The lung volume will be calculated and the information will be used to determine whether your baby should be delivered at SSM Health St. Mary's Hospital - St. Louis or SSM Health Cardinal Glennon Children's Hospital, both of which offer advanced breathing machines and additional therapies for critically ill newborns.

We understand that Congenital Pulmonary Airway Malformation (CPAM) can be a scary diagnosis. That's why we're available to help 24 hours a day, 7 days a week. For more information or to schedule an appointment, call us at 314-268-4037 or toll free at 877-SSM-FETL (877-776-3385).

While we can't change the diagnosis, we can provide you expert care and support, helping your baby get the most out of treatment and life.
