

Congenital Cystic Adenomatoid Malformation (CCAM)

What is congenital cystic adenomatoid malformation (CCAM)?

CCAM of the lung is a relatively rare abnormality that appears on an ultrasound as a solid or cystic mass (tumor) of pulmonary tissue that has an abundance of bronchial structures (breathing channels).

What causes congenital cystic adenomatoid malformation (CCAM)?

The cause of CCAM is unknown.

One theory holds that it is the result of bronchial structures failing to mature properly at approximately the fifth or sixth week of gestation, about the time the lungs begin to form.

Another theory holds that CCAM is due to an abnormal growth pattern of lung tissue due to bronchial obstruction. There is no known genetic cause for CCAM, and no cases of recurrence in a sibling or offspring have been reported.

What is the incidence of congenital cystic adenomatoid malformation (CCAM)?

Although CCAM is increasingly being diagnosed in utero, the true incidence of this lesion is not known. A CCAM lesion is slightly more common in males than females.

What are the different types of congenital cystic adenomatoid malformation (CCAM)?

Several classification systems have been used to describe the sonographic appearance of these malformations. Among the most commonly used is an adaptation of Stocker's histologic classification.

Type I

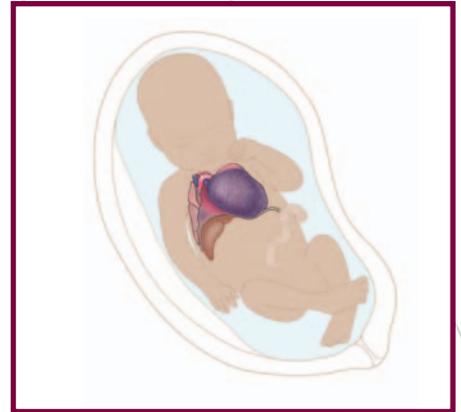
Approximately 50 percent of postnatal CCAM cases. The lesions consist of single or multiple cysts that are usually large in size (approximately three to 10 cm in diameter) and few in number (one to four).

Prognosis: In general, typically favorable, but dominant cysts (cysts comprising 1/3 of the volume) can grow rapidly and unpredictably.

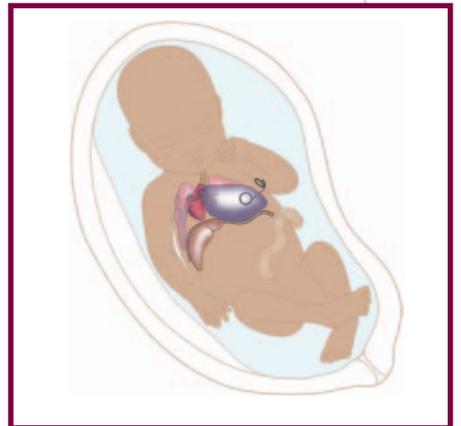
Type II

Approximately 40 percent of postnatal cases. These lesions have cysts that are smaller in size (less than one cm in diameter) and larger in number.

Prognosis: In general, a higher frequency of associated congenital anomalies.



Above is an illustration of CCAM of the lung.



Above is an illustration of CCAM shunt.

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Type III

Only 10 percent of postnatal cases. The lesions are composed of very small cysts and appear solid sonographically. These malformations can grow exponentially, between 20 and 26 weeks gestation. The large size can cause mediastinal shift (a change in the space in the chest between the lungs).

The large masses compress the lungs and depress the diaphragm, impairing the flow of blood to the heart. This can cause pulmonary hypoplasia (arrested or underdeveloped lungs) and hydrops (in utero heart failure).

Prognosis: In general, poor when associated with hydrops and when associated with pulmonary hypoplasia leading to respiratory problems in the newborn.

How is congenital cystic adenomatoid malformation (CCAM) diagnosed?

CCAM can be detected by a detailed ultrasound using color flow Doppler and by ultrafast fetal MRI to confirm the diagnosis and document size and location of the CCAM.

A fetal echocardiogram can rule out any associated structural heart abnormalities and assess the effect of the CCAM on the heart.

CVR (CAM Volume Ratio)

The best prognostic indicator we have is the CVR measurement. The CAM volume is a calculate volume based on ultrasound measurements obtained in three dimensions of the mass at presentation. The volume is then divided by the head circumference so the CAM volume is corrected for gestational age.

If the CVR is < 1.6 , this is a CCAM with a favorable prognosis. The risk of developing hydrops is less than two percent in these cases. The only exceptions are malformations with a “dominant cyst.” Dominant cysts are those that comprise greater than one-third of the entire volume of the CCAM. These lesions can enlarge acutely, do not follow a predictable pattern of growth and must be followed closely.

Those congenital cystic adenomatoid malformation with a CVR > 1.6 are at high risk for the development of hydrops and fetal demise, which occurs in up to 80 percent of cases in this category. Such malformations should be followed with twice-weekly ultrasound scans so that fetal surgery can be undertaken at the earliest signs of hydrops.

All congenital cystic adenomatoid malformation should be followed weekly for measurement of CAM volume and CVR until the growth of CCAM reaches a plateau. The natural history of CCAM is near exponential growth, from 20 weeks gestation until the plateau is reached.

The average gestational age at which the growth plateau is reached is 26 weeks. A fetus does not typically get into trouble from hydrops once the plateau has been reached. From that point on, there is usually a slow decrease in the size of the CCAM, as the baby grows around it.



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How is pregnancy managed when congenital cystic adenomatoid malformation (CCAM) are detected?

CCAM should be followed at least weekly with ultrasound scans for measurement of CAM volume. The CAM volume has been shown to be predictive of outcome in fetuses with these malformations.

At least weekly ultrasound scans also are recommended to detect signs of hydrops (in utero heart failure) caused by the CCAM. The development of hydrops in a fetus with a CCAM is usually an indication for fetal surgery.

Another sign of hydrops is placental swelling. If placental swelling develops as a result of the CCAM, the mother is at risk to develop the “mirror syndrome,” an unusual condition in which the mother’s health “mirrors” that of the sick fetus by developing swelling and high blood pressure.

This condition calls for immediate delivery of the fetus because “mirror syndrome” is potentially life threatening for the mother.

In contrast, low-risk CCAM with CVRs < 1.6 who reach a growth plateau can often be delivered without risk of respiratory compromise following delivery. All high-risk fetuses with CVRs > 1.6 should be delivered at a tertiary-care hospital, preferably with Extracorporeal Membrane Oxygenation (ECMO) (heart/lung machine) capability, and where a skilled surgical staff is on hand for resuscitation of the newborn and emergency removal of the mass.

The hospital should also have an intensive care nursery and staff trained to care for a newborn with potentially severe pulmonary hypoplasia. In rare cases, the size of the CCAM may preclude ventilation after the baby is born or support by ECMO because of shift of the mediastinum.

In these cases, an EXIT (ex utero intrapartum treatment) procedure may be performed to resect the CCAM while on placental support. Another consideration is multicystic CCAM, which may acutely enlarge after delivery due to “air trapping,” necessitating an emergency thoracotomy for CCAM removal.

What are the fetal interventions for congenital cystic adenomatoid malformation (CCAM)?

The indications for fetal intervention depend on the type of lesion, whether solid or cystic, and the development of hydrops. In malformations with a dominant cyst, the development of hydrops indicates the need for a thoracentesis or thoracoamniotic shunt to decompress the fluid within the cysts. In the case of solid lesions, the development of hydrops manifests by ascites, pleural or pericardial effusions, skin or scalp edema or placentomegaly.

In some cases, maternal steroids appear to arrest the growth of these lesions. This is sometimes a useful temporizing measure. In patients with large, solid CCAM with associated hydrops, open fetal surgery is indicated. Without intervention, CCAM associated with hydrops is almost always fatal.

In experience with 26 cases of open fetal surgery with hydropic CCAM, the survival has been 61 percent. The outcomes for fetal surgery for CCAM have been improved by careful observation and by intervention at the earliest signs of hydrops.



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In addition, the use of intraoperative echocardiographic fetal monitoring and thoracoabdominal surgical exposure have helped improve outcomes in this surgery. The survival in these cases has increased to 78 percent. Perhaps the best way to improve outcomes in high risk CCAM is through close surveillance to identify early signs of hydrops. Too often, patients are referred late in their course presenting with advanced hydrops.

Cases in which CCAM are large but have not resulted in hydrops may require resection during an EXIT procedure.

In some cases, the CCAM is so sufficiently large that the mediastinum is shifted and the lung compressed. Under these conditions, the lungs may not be able to be ventilated. Removing the CCAM during an EXIT procedure allows the lungs to function.

What are treatment options for the newborn?

Newborns with CCAM should be evaluated in the nursery to confirm the prenatal diagnosis and to exclude other health conditions.

A chest CT scan will confirm the diagnosis and define the anatomy. Complete resection of the CCAM is the preferred treatment. The reasons for recommending resection for CCAM, which are otherwise asymptomatic, are infection and neoplastic transformation.

All malformations have a connection with the tracheobronchial tree, which may predispose to bacterial contamination. Malformations can undergo malignant transformation. The youngest case reported was a patient only 13 months of age.

While some recommend observation for asymptomatic CCAM, we routinely recommend they be resected. These malformations remain a health concern for as long as they are present.

Unfortunately, we do not know how to identify which malformations will become infected or develop a tumor before the problem actually occurs.

What is the long-term outcome with congenital cystic adenomatoid malformation (CCAM)?

The long-term outcome of infants with CCAM following resection is excellent. However, if the mass is not fully removed, the child can be at risk for complications, such as air trapped in the residual CCAM, which causes gradual enlargement over time, infection and other problems.

Once the mass is removed, the remaining lung tissue will undergo compensatory lung growth to make up for the lung tissue lost to resection. These children live life without any apparent limitations on their activities and with no increased risk of respiratory infections.

Children who survived open fetal surgery for CCAMs associated with hydrops are still doing well from one to eight years after their procedure.

Contact the Fetal Care Center of Cincinnati

For more information, please call 1-888-FETAL59
or email us at info@fetalcarecenter.org.

References

For a detailed reference list, visit our web site at www.fetalcarecenter.org.

