Congenital Cystic Adenomatoid Malformation

Congenital cystic adenomatoid malformation is a rare abnormality of lung development in unborn babies. Congenital cystic adenomatoid malformation is increasingly detected by the routine ultrasound scan during pregnancy. Congenital means present at birth.

The severity of the abnormality is very variable. Some lesions can shrink or even disappear without treatment. Some lesions cause the baby to have breathing problems very soon after birth. Some lesions cause severe problems for the baby and have a poor outcome.

What is congenital cystic adenomatoid malformation (CCAM)?

A CCAM is a non-cancerous mass of abnormal lung tissue, usually located in one part (lobe) of a lung. The abnormal lung tissue may include solid areas as well as fluid-filled sacs (cysts).

CCAM is an abnormal development of lung tissue during the pregnancy. It is not known why this happens.

CCAM can be caused by multiple large cysts (called macrocystic CCAM) with cysts larger than 5 mm in diameter. This is the most common type of CCAM and usually has a good outcome.

CCAM may also be caused by multiple small cysts (called microcystic CCAM) with cysts smaller than 5 mm in diameter. Microcystic CCAM may be associated with congenital abnormalities such as abnormal development of the kidneys. Microcystic CCAM tends to have a worse outcome than macrocystic CCAM.

How common is congenital cystic adenomatoid malformation (CCAM)?

CCAM is rare but the exact frequency is not known. The use of routine ultrasound scans in pregnancy has meant that many affected babies are diagnosed before birth. Improved scan techniques have also meant that some babies are found to have CCAMs which may previously have not been diagnosed.

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

CCAM is often diagnosed before birth, by a routine ultrasound scan. CCAM may be associated with excessive fluid around the baby during the pregnancy (polyhydramnios).

After birth, most of those affected develop symptoms soon after birth with breathing difficulty. Sometimes CCAM is diagnosed by accident in later life. Recurrent chest infections may be a feature later in life. An affected child may also have poor growth and weight gain (failure to thrive).

How is congenital cystic adenomatoid malformation diagnosed (CCAM)?

CCAM is often diagnosed by ultrasound scan during the pregnancy.
Investigations used to diagnose CCAM and assess the severity of CCAM include a chest X-ray, a CT scan and an MRI scan of the chest.

Babies diagnosed as having CCAM should have investigations to look for other possible associated abnormalities, including ultrasound scans of the brain and kidneys and a scan of the heart (echocardiogram).

A sample of blood, or fluid from the womb (amniotic fluid), is also sent to look for an associated chromosome abnormality but this is uncommon.

What is the treatment for congenital cystic adenomatoid malformation (CCAM)?

It is now known that some CCAMs become smaller or even disappear with time. Therefore, treatment is not always necessary.

Oxygen or even artificial ventilation may be required for babies who develop breathing difficulties after birth.

Surgery is the main treatment for CCAM and may be needed before the birth. Surgery before birth may be considered for large CCAMs or if the CCAM has become associated with other problems for the baby.

An alternative to surgery before the birth is draining cyst fluid through the chest wall (thoracocentesis) but the fluid usually quickly returns after the procedure. A large fluid-filled cyst may be treated with a shunt (a thoracoamniotic shunt) that continually drains fluid from the CCAM.

Are there any complications?

Large lesions may be associated with the development of other developmental abnormalities such as abnormal development of the kidneys. The other main complication is abnormal lung development in the womb. This causes breathing difficulties after the birth.

Other possible complications include premature birth, recurrent pneumonia and air that is trapped next to a lung (pneumothorax).

There may be an increased risk of cancer developing as a result of congenital cystic adenomatoid malformation (CCAM) but it is not yet known exactly how common this is.

What is the outlook (prognosis) for congenital cystic adenomatoid malformation (CCAM)?

Smaller CCAMs, especially macrocystic CCAMs, often have an excellent outcome and don't need any treatment.

Shrinkage and even disappearance of a CCAM before birth may occur in a few affected babies.

Surgical removal of the CCAM usually leads to full recovery. It is not known whether or not complete surgical removal completely removes the risk of a cancer developing.

Death may occur in up to 30% of all affected babies who present soon after birth. Risk factors for a poor outcome include other associated abnormalities, microcystic CCAM and a large lesion.

Further reading & references

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