Congenital Pulmonary Airway Malformation

Congenital pulmonary airway malformation (CPAM) is a rare abnormality of lung development. It is found either in unborn babies or in young babies. The name has recently changed from congenital cystic adenomatoid malformation (CCAM). It is increasingly detected by the routine ultrasound scan during pregnancy. Congenital means present from birth. Pulmonary means related to the lungs.

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 may have a poor outcome.

What is congenital pulmonary airway malformation in babies?

Congenital pulmonary airway malformation is an abnormal development of lung tissue during the pregnancy. It is not known why this happens.

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

CPAM types

CPAM has five different types defined by the size of the cyst. Type I is the most common type and occurs in about 7-8 cases out of 10. The cysts are between 2-10 cm in diameter. This usually has a good outcome.

Type III is caused by multiple small cysts that are smaller than 5 mm in diameter. It involves the whole lobe of the lung. It happens in approximately 1 case in every 10 and has a worse outcome.

Type 0 is the rarest and has the worst outcome. The cysts stop the development of the lungs completely.

How common is congenital pulmonary airway malformation?

CPAM is rare but the exact frequency is not known. Data from large population registries suggest that congenital lung cysts occur in about 1 in every 11,000 to 35,000 live births.

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 CPAMs which may previously have not been diagnosed.

What problems does a CPAM/CCAM cause?

CPAM is often diagnosed before birth, by a routine ultrasound scan. It may be associated with excessive fluid around the baby during the pregnancy. This is called polyhydramnios.

After birth, most of those affected develop symptoms soon after birth. The usual problem is breathing difficulty. Sometimes it 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.

How is congenital pulmonary airway malformation diagnosed?

CPAM is often diagnosed by ultrasound scan during the pregnancy.

Investigations used to diagnose and assess the severity include a chest X-ray, a computerised tomography (CT) scan and a magnetic resonance imaging (MRI) scan of the chest.

Babies diagnosed as having CPAM should have investigations to look for other possible associated abnormalities. The investigations include ultrasound scans of the brain and kidneys and a scan of the heart (an echocardiogram, or echo).

A sample of blood, or fluid from the womb (amniotic fluid), may also be sent to look for an associated chromosome abnormality but these are uncommon.
What is the treatment for congenital pulmonary airway malformation?

It is now known that some CPAMs 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 and may be needed before the birth. The operation removes the cyst or the part of the lung affected. Surgery before birth may be considered for large CPAMs or if it has become associated with other problems for the baby. The surgery allows the lung to then develop properly and children tend to do well after surgery.

An alternative to surgery before the birth is draining cyst fluid through the chest wall. This is called thoracocentesis. Unfortunately 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.

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 (chest infections) and air that is trapped next to a lung (pneumothorax).

There may be an increased risk of cancer developing as a result of congenital pulmonary airway malformation but it is not yet known exactly how common this is.

What is the outlook for congenital pulmonary airway malformation?

Smaller CPAMs, especially those with larger cysts, often have an excellent outcome and don’t need any treatment.

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

Surgical removal usually leads to full recovery. It is not yet known whether complete surgical removal removes any risk of a cancer developing.

Death may occur in affected babies who present soon after birth. Risk factors for a worse outcome include associated abnormalities, microcystic CPAM and larger lesions.

Further reading & references


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