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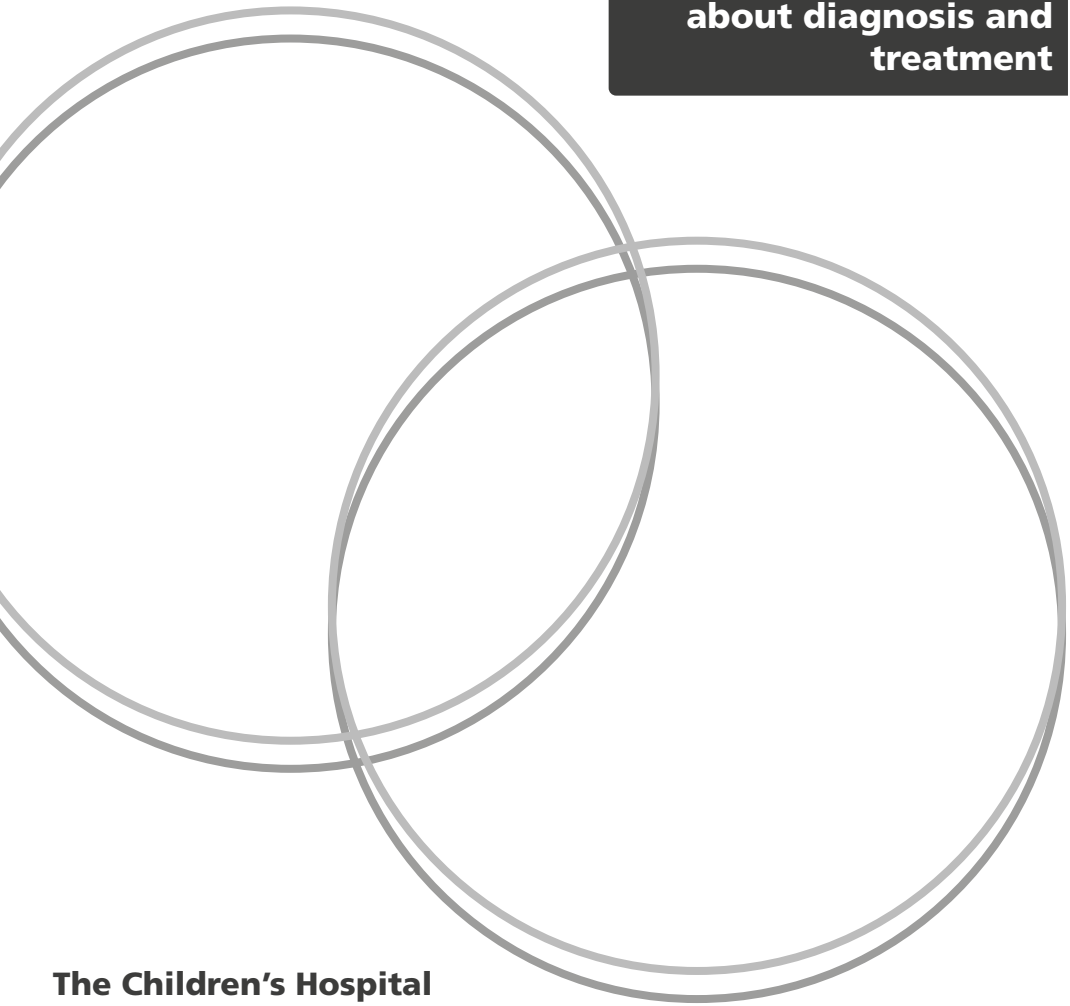
Cystic Adenomatoid Malformation of the Lung (CCAM)

Information for parents about diagnosis and treatment



The Children's Hospital

What is CCAM?

Cystic Adenomatoid Malformation of the Lung (CCAM) is a very rare condition in which part of a baby's lung does not develop normally. The affected part of the lung overgrows and forms cysts filled with fluid. This tissue does not function as normal lung tissue.

How is it diagnosed?

Babies with CCAM are usually detected during routine pre-natal ultrasound scans.

Sometimes this condition is not diagnosed until after the baby is born. These babies often need X-rays and CT scans (a special kind of X-ray scan) to make the diagnosis.

What symptoms does it cause?

Many babies are diagnosed before birth and this does not affect the course of the mother's pregnancy.

After the malformation has been diagnosed pre-natally, it may remain the same or it may increase in size.

Sometimes a condition called "polyhydraminos" develops. This means that there is too much amniotic fluid surrounding the baby which can increase the chance of the baby being born prematurely.

Rarely the CCAM can grow so big that it affects the working of the baby's heart. This may endanger the life of the baby and sometimes that of the mother.

Most babies have no problems after birth and can go home normally. However these babies can go on to develop problems with breathing or repeated chest infections later in life. Children with CCAM may also develop other lung lesions later in life such as tumours. However, this is rare.

For these reasons even babies healthy at birth still need a special scan (CT scan) at 1 month of age, followed by surgery around 6 months of age.

Some babies with CCAM (around 10%) are born with difficulty in breathing and need oxygen or even help with breathing using a ventilator. Babies with severe breathing problems after birth may need an operation shortly afterwards.

Apart from difficulties with breathing babies with CCAM do not tend to have problems with any other organs.

What causes CCAM?

There is no known genetic or environmental cause for CCAM.

What happens after it is diagnosed?

After CCAM is diagnosed a series of ultra-sound scans are performed throughout pregnancy to watch the condition develop. Special scans of the heart and kidneys are also carried out to check that the baby is healthy.

The rest of the pregnancy is followed-up by a number of specialist doctors. These doctors will help make sure both mother and baby remain healthy during pregnancy and make plans for safe delivery.

The place of delivery depends on the size of malformation and the condition of the baby. Most deliveries can take place at a local hospital. However, if pre-natal scans indicate that the baby is at risk of problems at birth, then delivery is planned to take place at a specialist unit. This means you may need to be looked after in hospital some distance from home.

Once the baby is born it may have be given oxygen or need to use a ventilator. This helps the baby to breathe. If the baby does not need help with breathing it can go home but will need regular follow-up with specialist doctors.

After going home your baby will need to be seen as an outpatient at a special centre. This involves a further scan at 1 month of age, followed by a clinic appointment at 2 months and possible further treatment at 6 months.

How is CCAM treated?

There are two schools of thought on how to manage CCAM.

Most surgeons (around 90% world-wide) will offer surgical treatment to remove the abnormal tissue.

However, some surgeons choose to follow these babies up with yearly CT scans, and only operate if the CCAM causes serious symptoms.

Surgery usually occurs in baby's first 3 to 6 months of life, unless the baby is very unwell. This time is chosen so that the baby is big enough to have an operation but before any long term problems with the lung occur.

What happens before the operation?

Your baby will need to come into the Children's Hospital on the day of the surgery. A number of tests will be performed to check the baby is well enough for surgery.

The surgeons will ask for your consent for the operation to go ahead. If you are unsure about anything, or have any questions you should ask the doctor before signing the consent form.

What happens during the operation?

The operation is done under a "general anaesthetic". This means that the baby will be asleep throughout the operation. You can stay with your child before and after the operation, and one parent may stay in the anaesthetic room until (s)he is asleep.

The operation takes around 2 hours and involves removing the abnormal lung tissue.

It is usually done using a keyhole technique with a small cut in the side of chest wall and using a type of telescope (known as a "VATS" procedure).

Sometimes this is not possible and an open operation is carried out instead. In this case the surgeon will need to make a larger cut in the chest wall.

What are the risks?

As with all operations there is a risk of **bleeding** and **infection** and **air leaks**.

Sometimes the operation can cause part of the lung to **collapse** or cause air to get into the chest (a **pneumothorax**). This might mean the baby needs extra help with breathing after the operation but both conditions usually resolve with treatment.

Very rarely a larger **part of the lung** or even a **whole lung** may need to be **removed**. Although this is a serious complication many children can survive with just one healthy lung.

Your doctor will discuss these risks with you in more detail.

There are also risks associated with the general anaesthetic. The anaesthetist will be available to discuss these with you before the operation.

What happens after the operation?

Most babies will come back to the ward with a chest drain. Rarely babies need intensive care and further support.

Please see the leaflet "Chest Drains – Information for Parents" for more information on this, or ask your doctor if you have any questions.

It takes a little while for the baby to wake up from the anaesthetic. Before the baby wakes up (s)he will be given pain relieving medicines so will not be in pain.

The baby can re-start feeding soon after surgery, and is usually well enough to go home in about 5 days.

Wound care

The wound will need to be kept clean and dry for 2 days so the baby cannot be bathed in that time.

The stitches are dissolvable so will disappear with time. The "steri-strips" used over the wound site will loosen and fall off by themselves. If they do not, you can soak them off with water after 5 days.

Follow-up

The paediatric surgery team will continue to see the baby after (s)he is discharged from the hospital. The first appointment will usually be about one month after the operation.

The future

Babies who have CCAM removed usually do well afterwards. The lungs continue to grow until the child is 8 years old so this leaves plenty of time for the development of normal healthy lungs.

However, if you notice signs of any of the following:

- difficulty breathing – this includes noisy breathing or fast breathing, flaring of the nostrils or the baby looking pale or blue
- repeated chest infections
- problem with feeding
- poor weight gain

please tell your doctor or GP.

You should avoid air travel for 1 month after the operation and not smoke around children.

Otherwise you should treat their child normally including normal schooling, socialising and childhood immunisations. You can also be reassured that future pregnancies are no more likely to develop CCAM because of one affected child.

How to contact us

If you have any questions or concerns, please telephone:

Tom's Ward

Telephone: 01865 234 111 or 01865 234 110

John Radcliffe Hospital Switchboard

Telephone: 0300 304 7777

Paediatric Surgery Secretaries

Telephone: 01865 234 197

Further information

If you would like an interpreter, please speak to the department where you are being seen.

Please also tell them if you would like this information in another format, such as:

- Easy Read
- large print
- braille
- audio
- electronic
- another language.

We have tried to make the information in this leaflet meet your needs. If it does not meet your individual needs or situation, please speak to your healthcare team. They are happy to help.

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June 2024
Review: June 2027
Oxford University Hospitals NHS Foundation Trust
www.ouh.nhs.uk/information

Leaflet reference number: OMI 65492