

CONGENITAL LUNG DISORDERS

What is a congenital problem?

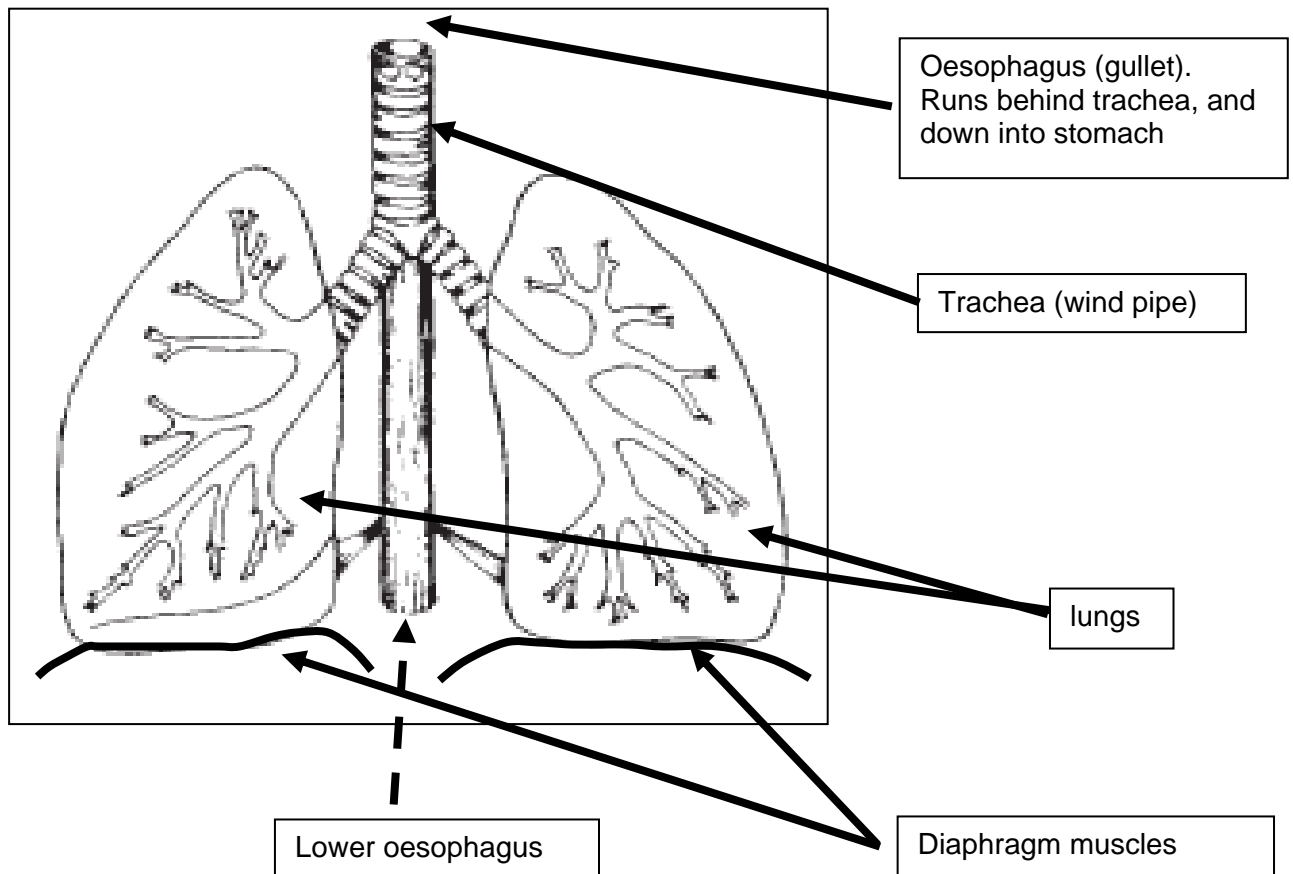
A congenital problem is one that a child has been born with. For lung problems, they usually refer to abnormalities of embryological development. They can be grouped into problems of airways, problems of the lung itself, and problems secondary to abnormalities in other parts of the body.

Basic anatomy

See diagram below

The trachea (windpipe) is in front of the oesophagus (gullet). When we breathe in, the air travels down the trachea to the left and right lungs. The trachea splits into the left bronchus and right bronchus which go to each lung.

The diaphragm is a sheet of muscle that sits under the lungs and separates the chest from the abdomen. It contracts during normal breathing.



What causes Congenital Lung Problems?

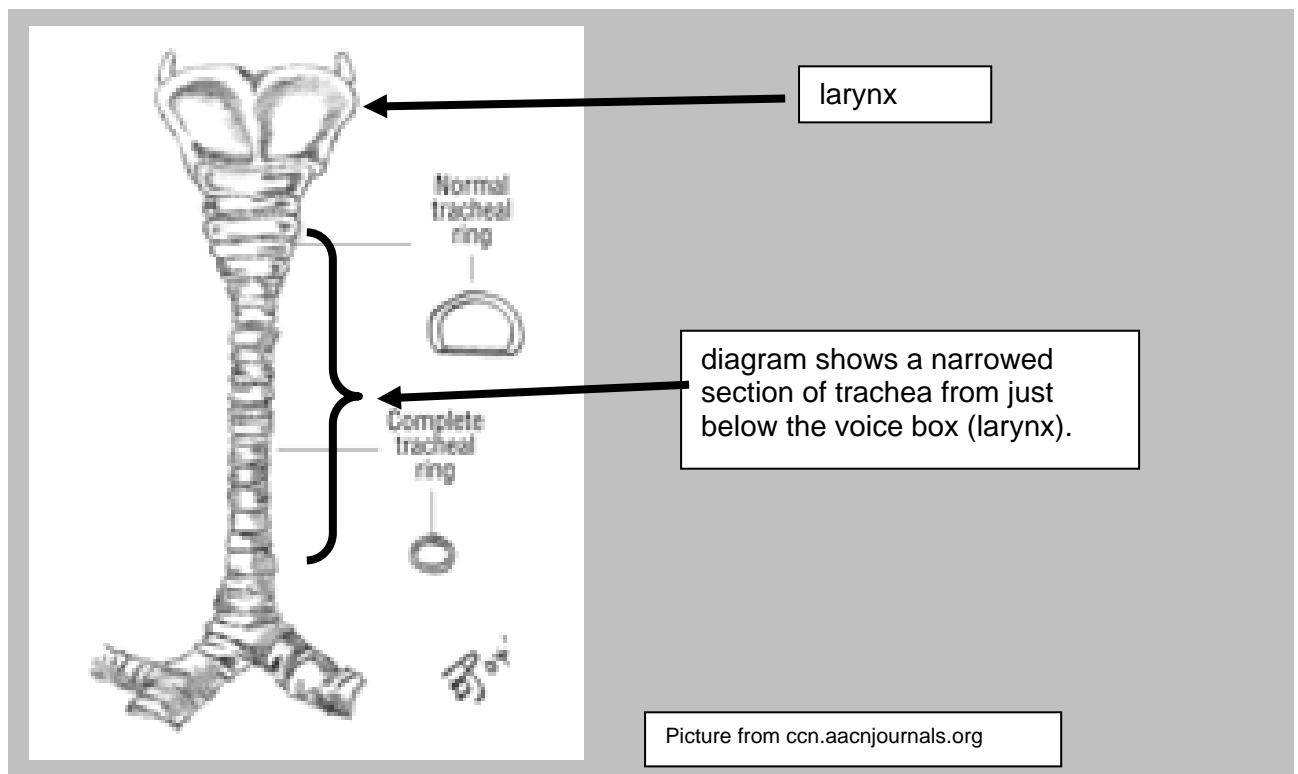
The exact cause is usually unknown. Occasionally the lung problem is associated with congenital abnormalities in other organs. Sometimes the combination of abnormalities suggests a chromosomal problem. Sometimes the cause is not known, but the combination of abnormalities has been seen before (usually called a 'syndrome'). However, it is not uncommon for the congenital abnormality to be isolated (i.e. not associated with any other problem). When this occurs the cause may never be known.

What types of problem can occur?

There are many different types of congenital lung problems. This leaflet will give you a basic idea of some of the problems that can occur. Your child's doctor will discuss your child's particular problem with you, explain how it is affecting your child, and what treatments can be offered to help.

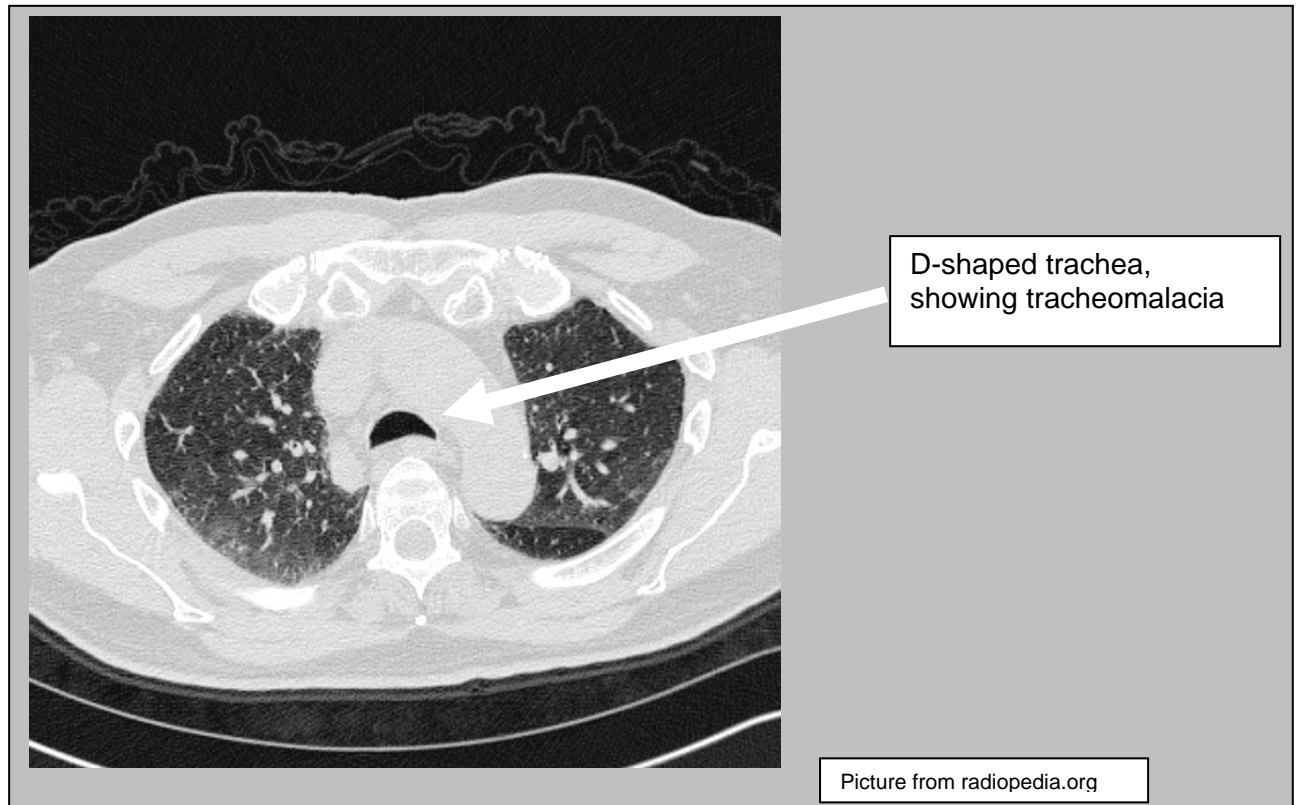
Tracheal Stenosis

This is where the trachea is narrower than it should be. This makes the work of breathing much harder, and even with simple chest infections (e.g. bronchiolitis) a child can become very unwell.



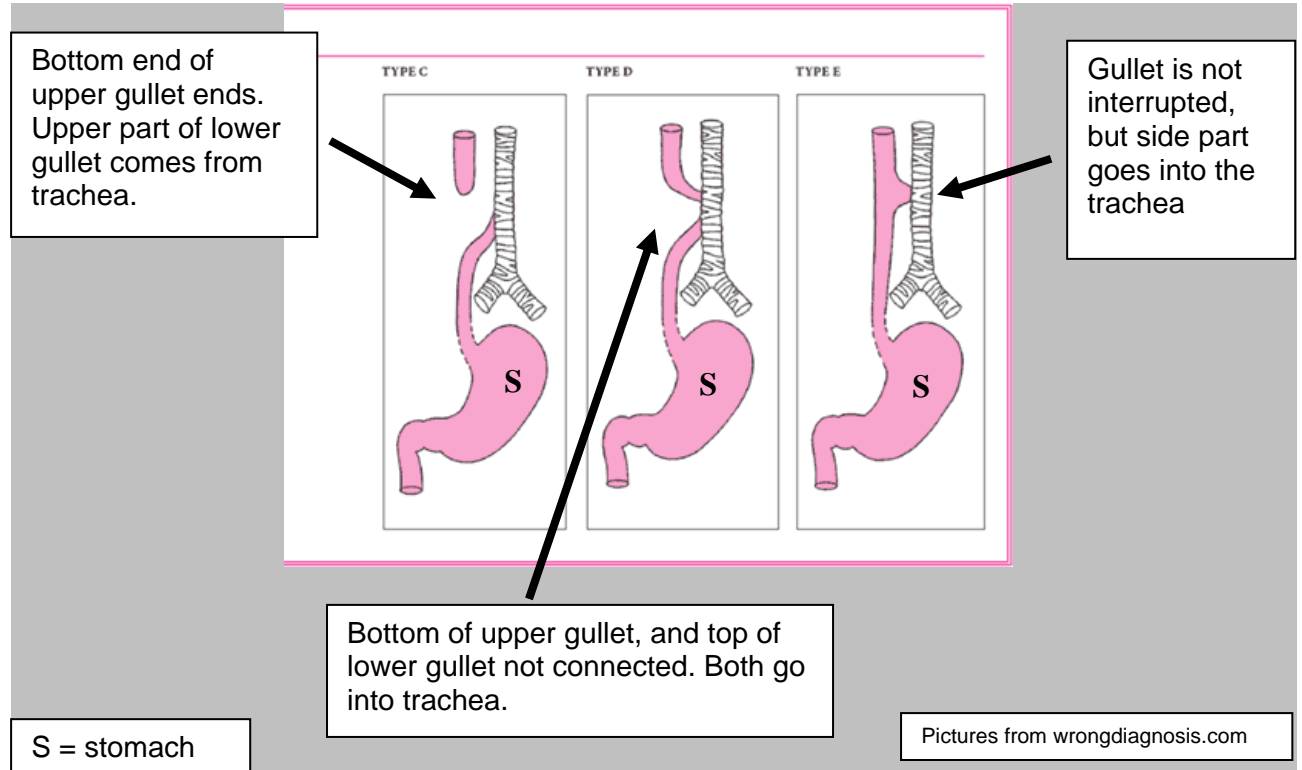
Tracheomalacia

Tracheomalacia refers to the trachea being softer than normal, and so when the child breathes out or coughs the calibre of the trachea gets smaller. The picture is of a CT of the chest showing an abnormal "D" or kidney shape to the trachea, as it collapses a bit. Normally the trachea is round.



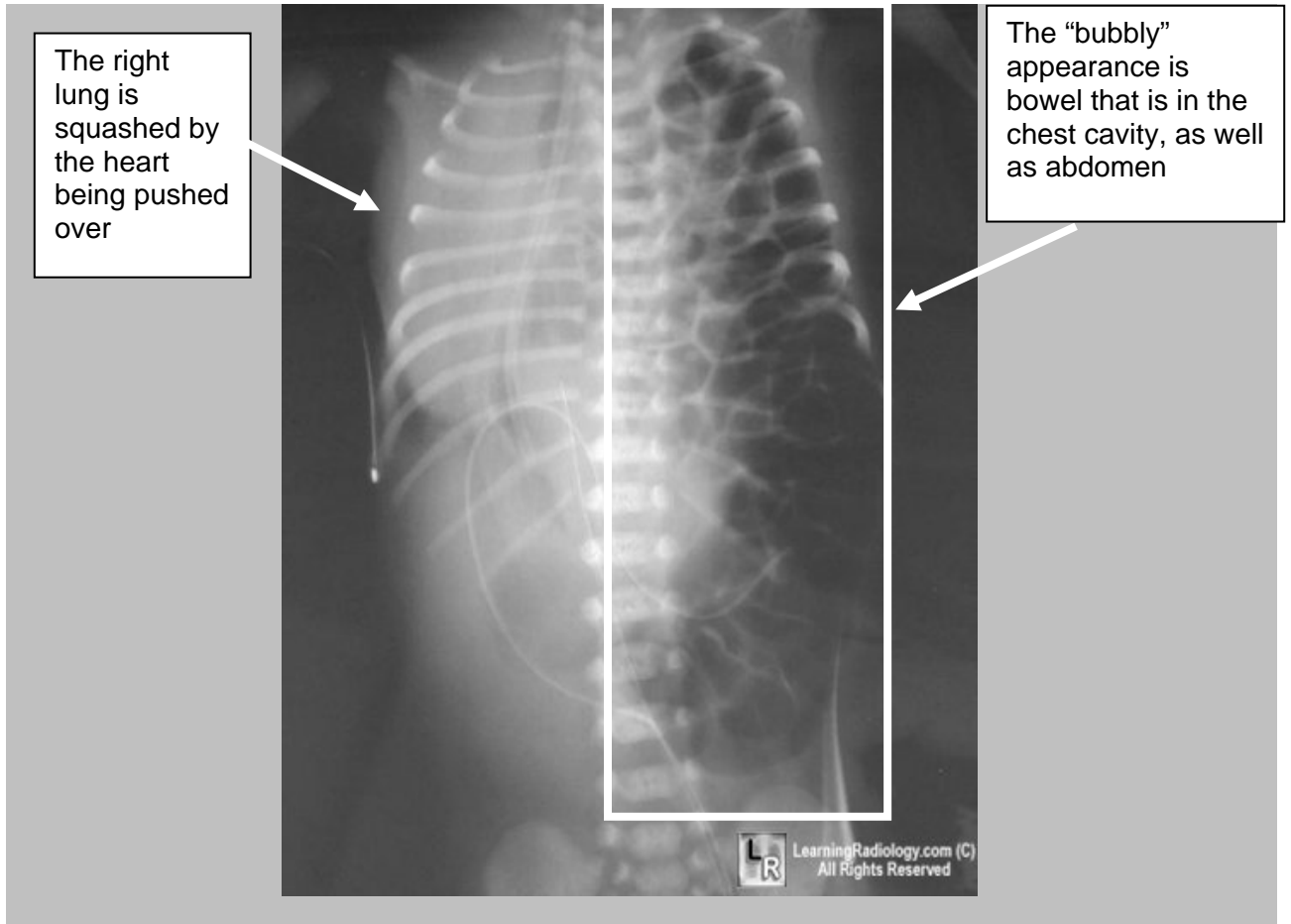
Tracheo-oesophageal fistula

A fistula is an abnormal connection between two organs. In a tracheo-oesophageal fistula, there is a connection between the oesophagus (gullet) and trachea (windpipe). There are different types, as shown in the diagram. The abnormal connection can be separated by an operation, but even afterwards the trachea is abnormal (tracheomalacia – see above)



Congenital diaphragmatic hernia

Sometimes the diaphragm muscle doesn't form properly, and the bowel is able to get into the chest. This then compresses the growing lungs, and can cause very severe problems with the way the lungs grow (pulmonary hypoplasia). Most children with a diaphragmatic hernia are seriously ill when they are first born, but given time and Intensive Care support they slowly improve. Very rarely the damage to lung growth is so severe the child cannot survive. The xray shows a large amount of bowel in the chest cavity.

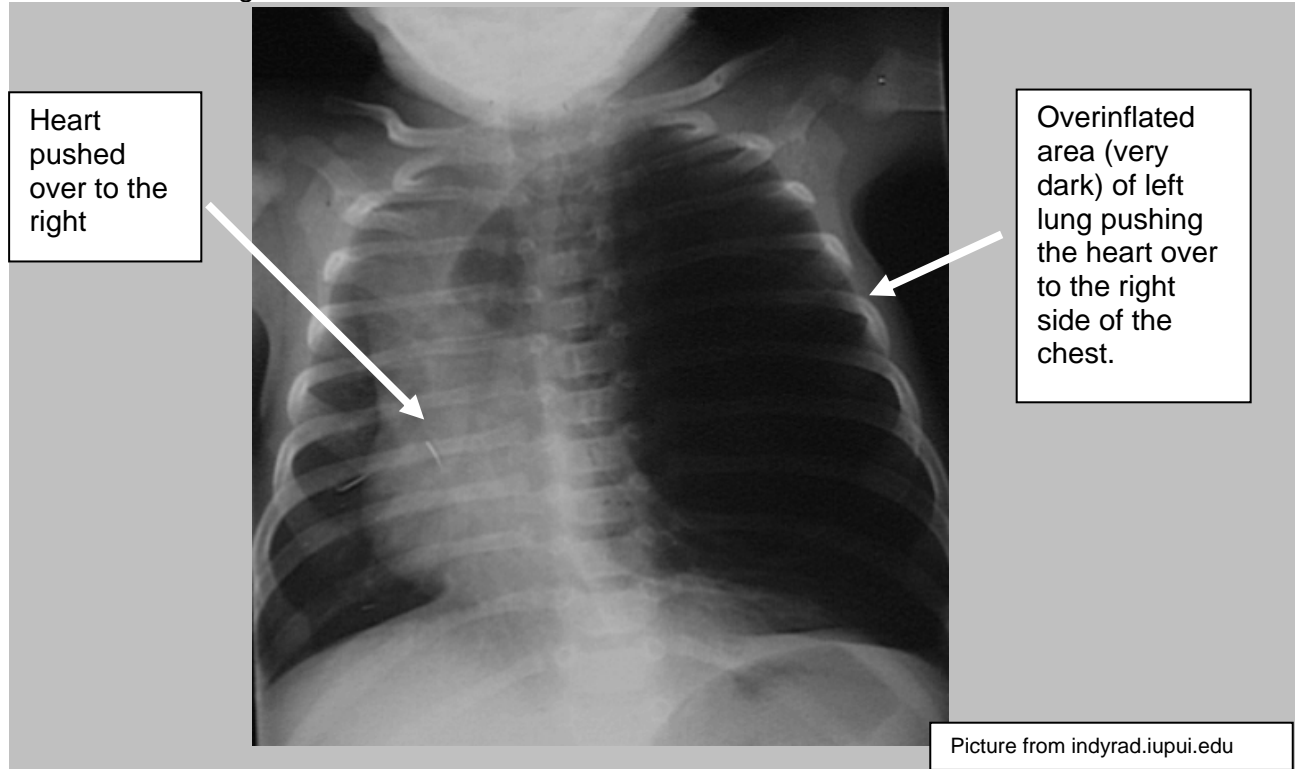


Picture from learningradiol.com

Congenital lobar emphysema

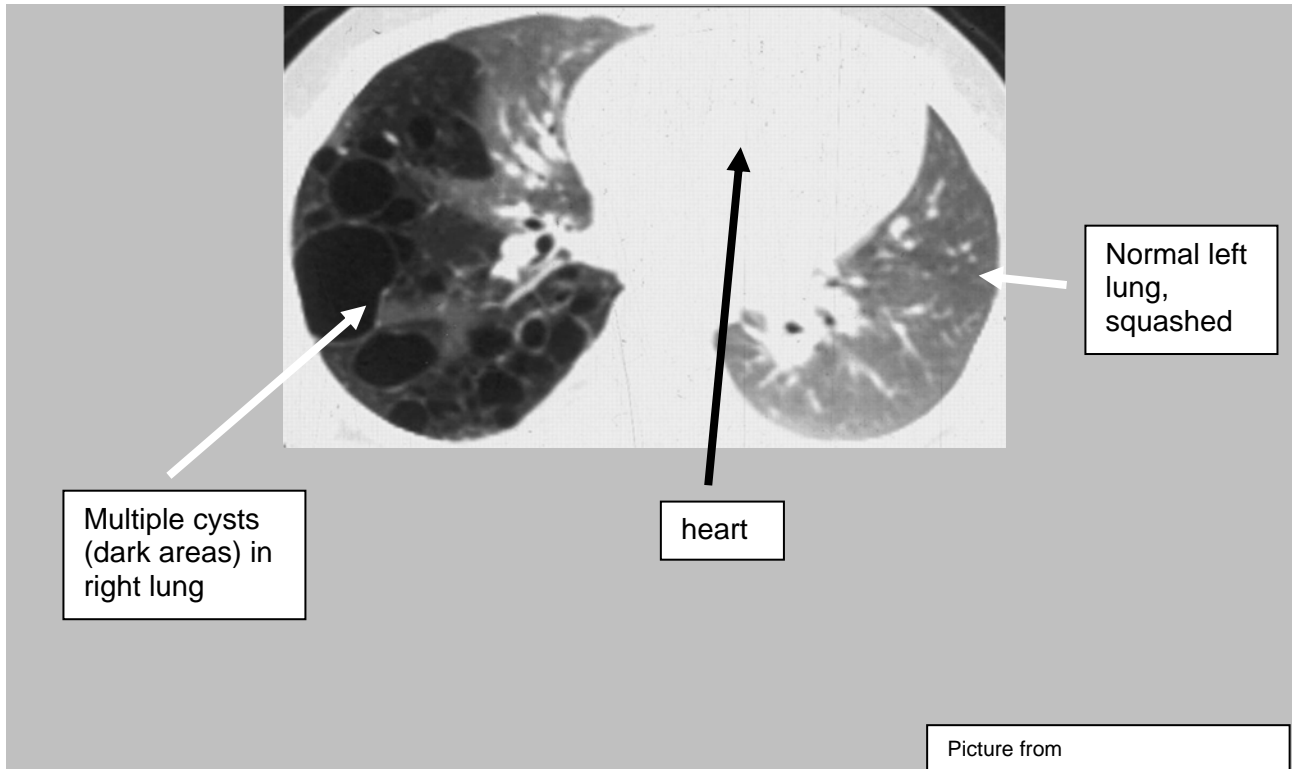
This is a problem where a portion of one lung forms abnormally so it is permanently overinflated and too large. It then compresses other parts of the lungs in the chest. It is usually removed by an operation, although the timing of when this is best done varies from child to child. Your child's doctors will be able to advise what is best for your child.

Overinflated L lung



Congenital Pulmonary Adenomatoid Malformation (CPAM)

This abnormality is a problem with the formation of lung tissue itself. Part of the lung grows as a solid collection of cysts and solid tissue. This usually needs removal. It is quite common for the problem not to present itself until the child is much older, or even into adulthood.



Can these problems be picked up early?

Some of these problems can be seen, or at least suspected, on antenatal scans done in pregnancy. However many of these problems may not be seen on antenatal scans. They may present early after the baby is born, but sometimes can go unrecognized for several months or even years.

How will they affect my child's health?

The main factor affecting how unwell a child will be how much these problems damage the normal growth and development of the lung.

How can these problems be treated?

Problems with the large airways can sometimes be operated on to correct them. However for problems with the lung tissue, there is no operation that can solve the problem and neither is there any medicine that will make the lung normal. The best way for a child to get better from a congenital lung problem is for them to grow as well as possible. However other things (e.g. oxygen therapy, tube feeding, physiotherapy, antibiotics etc) may help to make sure that the lungs have the best chance to grow. Some congenital lung problems do not get much better, even with the best nutrition. For some children, the problem is so severe that they are too fragile even to have the basic operation. Sadly, children who are this unwell often have a very short life expectancy

Where can I get more information?

The internet is a source of much information for all types of congenital lung problems. If you access anything from the web, you should always discuss it with your child's Respiratory Specialist, as they can put your child's condition into context of the information you have obtained off the internet. Some conditions have their own parent support group, either here or based in other countries.

This information sheet is produced to answer some of the questions parents ask about congenital lung disorders. It is not intended to replace discussion with your child's Respiratory Specialist and you are encouraged to discuss your child's condition with the specialists when you attend clinic.