Echogenic lungs

Patient Information Series – What you should know, what you should ask.

What are echogenic lungs?

The term “echogenic lungs” refers to an appearance of the fetal lungs that appear brighter than usual on prenatal ultrasound.

What causes this appearance on bright or echogenic lungs?

This appearance on prenatal ultrasound is commonly caused by one of three fetal lung conditions: 1) a condition called cystic adenomatoid malformation of the lung (CCAM) /CPAM; 2) another lung lesion caused “Pulmonary sequestration “, or 3) a third condition called “bronchial atresia.” The former condition is a developmental condition that results in an overgrowth of the air sacs whereas the second condition is the result of an extra portion of lung tissue developing in an unusual place, either within the chest or below the diaphragm. The third condition, bronchial atresia, is caused by obstruction of the upper airways or lack of development of the air passages. In all cases the abnormal lung tissue will appear unusually bright on ultrasound and sometimes appears cystic i.e. displaying multiple small fluid-filled spaces in the lung or larger cystic spaces.

How can these conditions be differentiated?

These two conditions can appear very similar on ultrasound. After making a diagnosis of a bright lung lesion on ultrasound your doctor will attempt to discriminate these lesions by looking at the blood supply to the lung lesions by a using an ultrasound technology called colour Doppler. The origin of the supplying blood vessel, can provide clues as to the nature of the lesion. i.e. blood supply from a tributary of the main artery leaving the lung (pulmonary artery) will suggest a CPAM/CAM or possibly bronchial atresia whereas blood supply from the main artery leaving the heart and supplying the rest of the body (aorta) will suggest a pulmonary sequestration. Evaluation of these lesions requires a detailed ultrasound by an experienced provider and often takes place in specialized high risk perinatal centres.

The third condition, bronchial atresia or congenital high airway obstruction is very rare and is discussed separately in another patient information document.

How do these lung conditions happen?

CPAM is thought to develop as a result of overgrowth of some of the lung smallest airways, called terminal bronchioles, which in turn can develop cystic enlargement. Conversely pulmonary sequestration can develop due to unusual blood supply developing from vessels that usually supply the rest of the body, not the lungs. Finally, bronchial atresia can develop following obstruction of the lungs resulting in trapping of fluid in the lung and enlargement of the trapped airways.

These lung problems are fairly rare lesions, seen in about 1/3000 pregnancies.

Should I have any special testing?

If the lung lesion is the only problem found in the fetus, genetic testing for chromosomal abnormalities is unlikely to yield a significant result. If lung lesions are found in association with other structural differences detected on ultrasound, the risk of chromosomal abnormalities rises dramatically and genetic testing will be recommended.

Your doctor may suggest a special fetal surgical procedure called bronchoscopy to evaluate the condition of the airway before birth in order to plan for delivery and anticipate the potential need for specialized equipment or personnel at birth.

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How will my pregnancy be monitored?

Your doctor will suggest serial ultrasound monitoring of your baby's condition to assess whether the lung lesions have grown over time. Sometimes these lesions can impair the baby's ability to swallow amniotic fluid before birth and can result in an excess of amniotic fluid, called polyhydramnios. This can be apparent on ultrasound and if severe, can increase the risk of preterm birth. The fetus will also be monitored for a condition called hydrops fetalis in which excess fluid is found in the chest, abdomen or under the skin. Not all babies with these conditions will develop this complication. It is more common when there are large cysts in the baby's lungs. In these circumstances your doctor may suggest placing a hollow tube, called a "shunt" into the baby's chest cavity to get rid of excess fluid around the lung which can prevent the lung from developing. Additionally, in some cases your doctor may recommend administration of steroid medication to the mother in an attempt to reduce the growth of the lung lesions and improve hydrops. Recently, in utero laser surgery has been offered before birth in select cases to stop the blood supply to the lung lesion.

Sometimes a CPAM can get harder to see on ultrasound as the baby develops to term. Postnatal imaging is still required.

Due to the nature of these lung lesions and potential for complications affected pregnancies are managed in high risk perinatal centres with a specialized team of Fetal Medicine Specialists, Neonatologists and Pediatric Surgeons.

What does this mean for my baby after birth?

After birth a skilled pediatrician or neonatologist will want to examine the baby and assess the lungs with chest X-Ray and /or ultrasound. Sometimes additional imaging such as MRI may be suggested. Babies with CPAM are at risk for recurrent lung infections and at times surgery is recommended to remove the diseased portion of the lung. These babies do well after surgery.

Surgery to remove the disease portion of the lung may also be required if the lung lesion is particularly large or interferes with breathing.

What is the risk of this happening again in a future pregnancy?

These lung conditions are considered sporadic occurrences with no significant risk of recurring in future pregnancy. Nevertheless, a detailed ultrasound to rule out structural problems will likely be recommended in future pregnancy.

What other questions should I ask?

- How often do I need to have an ultrasound?
- Will my baby require surgery in the womb?
- Where should I deliver?
- Can I meet the doctors who will be caring for my baby after birth?

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