Pulmonary Sequestration

What is Pulmonary Sequestration?

Pulmonary sequestration is a piece of abnormal lung tissue that develops in the lung or beside the lung. This lung tissue does not work like normal lung tissue.

What makes pulmonary sequestration different from other lung masses is that it receives blood supply from a blood vessel that comes directly from the aorta, the main artery of the body, instead of the pulmonary artery that normally supplies blood to the lungs.

Pulmonary sequestration can often be identified during a prenatal ultrasound at 18-20 weeks of pregnancy, but some cases are not found until later in pregnancy or after birth.

What are the causes of Sequestration?

Pulmonary sequestration is a rare lung condition. Doctors do not yet fully understand the exact cause of pulmonary sequestration. It appears to be a random event that can happen to anyone.

What does this mean?

Pulmonary sequestration can vary in size and can change in size as pregnancy progresses. Some may grow with the baby, some may remain the same size as the baby continues to grow and the pulmonary sequestration becomes relatively small. Many pulmonary sequestrations may even shrink or disappear before birth. In all these cases, the outlook for a normal life is excellent.

Pulmonary sequestrations are seldom associated with genetic problems.

Rarely, babies with pulmonary sequestrations develop fluid in the chest or even fetal hydrops. Fetal hydrops is an abnormal collection of fluid in the baby’s skin, chest or belly. Fetal hydrops may indicate heart failure. Babies with hydrops are very sick and may need treatment before birth. A large sequestration that is fluid filled may be able to be drained or emptied with a needle or special catheter called a shunt. This is done at a specialized hospital.

It is important to remember that most babies with pulmonary sequestration do not require any procedures during pregnancy.

What other tests should we consider?
Other tests may include a detailed ultrasound and a fetal heart ultrasound (echocardiogram). The detailed ultrasound is used to assess the baby’s other organs. A referral to a paediatric surgeon (specialist in surgery of babies and children) will be made to discuss with you the type of surgery the baby may need after birth.

Ongoing ultrasounds are used to monitor the growth of the pulmonary sequestration and to look for early signs of hydrops.

**What will happen around the time of the baby’s birth?**

Babies with pulmonary sequestration may need to be born at a hospital with a neonatologist (specialist in newborn babies) and a pediatric surgeon. Babies with pulmonary sequestration may require care in a neonatal intensive care unit.

Some babies may not require this increased level of care and will be able to deliver at your local hospital. Your doctor will be able to tell you where your baby needs to be delivered.

**What does this mean for my baby’s future?**

Many pulmonary sequestrations shrink or disappear before birth and these babies do not require treatment after birth. Some babies will require surgery to remove the sequestration but do very well after and continue to develop normally. The long-term outcome for babies with pulmonary sequestration is excellent. These children appear to grow as well as other healthy children.

**What do we do now?**

You will meet with a doctor that specializes in high-risk obstetrics. You may also meet the neonatologist and the pediatric surgeon. These doctors will discuss with you in detail your options for further testing, discuss with you test results and provide you with treatment options. They will be able to answer any questions you may have.

**Where can I get more information?**

The Children’s Hospital of Philadelphia
Breathing Easier: Fetal Lung Anomalies Video
http://www.chop.edu/video/breathing-easier-fetal-lung-lesions/home.html?item=1

Boston Children’s Hospital
Cystic Lung Disease
http://www.childrenshospital.org/az/Site2087/mainpageS2087P0.html