What is a Bronchopulmonary Sequestration?
A Bronchopulmonary Sequestration (also known as pulmonary sequestration or BPS) is a piece of lung tissue that develops without being connected to the airways, sometimes inside the lung and sometimes outside of it. The blood vessels going to the BPS also form abnormally. The BPS does not get blood flow like the rest of the lung. Rather it steals blood flow away from the body via a separate artery.

These lung masses can cause breathing problems, infection, or other complications if untreated. Most of the time a BPS does not cause problems for the fetus. In these cases, the baby can be monitored throughout the pregnancy and the BPS can be removed after the baby is delivered.

If the BPS is more severe, however, prenatal surgery may be needed. BPSs are a very rare birth defect. Nothing the mother does or has done during pregnancy causes this problem.

How is a Bronchopulmonary Sequestration diagnosed during pregnancy?
Typically around 20 weeks of gestation, a routine ultrasound will show if the baby has a lung mass. A series of follow-up ultrasounds can give the physicians more information about whether it is a BPS and its level of severity. Another test that may be recommended is a fetal echocardiogram (echo). This is an ultrasound of the heart performed by a pediatric cardiologist. This test is recommended to rule out structural heart defects and assess heart function. We also perform a fetal MRI to determine the nature and size of the mass, as well as how well the rest of the lung has developed. With all of this information, we can help patients and their families make the best possible decision about treatment.

How will a Bronchopulmonary Sequestration affect my baby?
BPSs usually cause problems by compressing the adjacent lung and heart. They usually continue to grow throughout the pregnancy, so we watch the growth very carefully by ultrasound examination. Very small sequestrations may not cause any complications, and can be removed within the first year of the baby’s life. However, larger sequestrations can be life-threatening both before birth and immediately after birth.

Before birth, the BPS can grow rapidly in the confined space of the chest. As shown in the picture above, this can push the heart and lungs to the opposite side of the chest and cause heart failure. A sign of heart failure is the accumulation of fluid in multiple areas of the developing fetus. If this process continues, the baby can die before birth. If no heart failure develops, the BPS can still be life-threatening at birth, by making the lungs too small to survive or preventing the lungs from expanding within the chest.

How are Bronchopulmonary Sequestrations managed and treated during pregnancy?
The primary goal of an evaluation at the SSM Health St. Louis Fetal Care Institute is to determine the severity of the BPS. This may take several evaluations in which we determine if the BPS is growing rapidly and how it is impacting the fetus.

Depending on the size and severity of the BPS, different types of surgery may be needed to treat it—in some cases before birth, and in some cases after birth.
What are the fetal surgery options for a Bronchopulmonary Sequestration?

**Laser Ablation:**
The Fetal Care Institute is one of the only places in North America to offer this groundbreaking treatment for severe BPS.

» During this minimally invasive fetal intervention, a small needle is inserted into the BPS and a laser fiber is targeted at the abnormal blood vessel going to the BPS. The laser blocks the blood flow, causing it to stop growing. The goal of the operation is to stop or decrease the blood flow to the mass. After the surgery, the BPS steals less blood flow from the fetus, and the heart and lungs start growing more normally as the BPS shrinks in size.

**Open Fetal Surgery:**
If the laser ablation operation is not possible, or is not successful, then open fetal surgery may be needed.

» In this operation, the fetal surgeon opens the mother’s uterus, exposing the baby’s chest. The chest is opened and the BPS is removed. The uterus is closed and the pregnancy continues. After open fetal surgery, the baby must be delivered by Cesarean birth since the uterine wound from fetal surgery cannot tolerate labor.

How is a Bronchopulmonary Sequestration treated during and after delivery?

If fetal intervention is not necessary the infant will be evaluated and treated at delivery. Most babies with a small BPS can be delivered vaginally without any apparent complications. These babies typically go home and are followed as an outpatient in two to four weeks after birth. The baby will be referred to a pediatric surgeon, who will help determine if, when, and how the BPS should be removed.

Babies with a moderately large BPS may have some difficulty breathing after birth. Usually, these babies breathe very quickly and sometimes they require oxygen. This quick breathing can make eating difficult for the baby. These babies need to be in a neonatal intensive care unit (NICU) for stabilization until surgery is performed to remove the BPS, and to allow for the remaining normal lung to function optimally. The baby will remain in the NICU until breathing and eating improve.

Sometimes, a very large BPS can compress the lung so severely that it prevents the lungs from expanding and functioning immediately at birth. We try to predict this problem based on the degree of shift of the lungs and heart to the opposite side of the chest. To avoid a crisis in the baby's breathing at birth, an EXIT (Ex Utero Intrapartum Treatment) procedure may need to be performed for delivery.

During an EXIT procedure, the mother has a Cesarean birth while asleep under general anesthesia. The placenta and umbilical cord remain attached to the mother and support the baby while we evaluate the baby's lung function. This gives the fetal surgeon time to evaluate the BPS and make sure that breathing will be adequate after birth. A breathing tube is inserted and breathing is assisted using a ventilator machine. If the baby breathes well, then the baby can be completely delivered and we can plan for removal of the BPS later. If the baby’s breathing is compromised, then the BPS needs to be removed immediately. The open chest operation of the baby is performed while the mother’s placenta supports the baby.

When the operation is finished, the umbilical cord is cut and the baby can breathe better without the compression of the BPS. Extracorporeal membrane oxygenation (ECMO) can also be used as a backup in case the lungs need more time to function optimally. The mother’s Cesarean incision is repaired and she is allowed to recover just like any Cesarean delivery.

What happens after surgery?
The outcomes for Bronchopulmonary Sequestration after surgery are excellent. In nearly all cases, these babies will develop normally without breathing or heart problems.