Let's Talk About...

Prenatal Counseling: Congenital pulmonary airway malformation (CPAM)

What is a congenital pulmonary airway malformation?

A **congenital pulmonary airway malformation** [kun-JEN-uh-tull PULL-moh-nehr-ee mal-for-MAY-shun] **(CPAM)** is the term for a portion of the lung that developed abnormally in the fetus. A healthcare provider usually notices a CPAM during an early pregnancy ultrasound.

There are 4 types of CPAMs, including:

- **Congenital cystic adenomatoid** [ad-en-OH-muh-toyd] **malformation (CCAM).** A CCAM is a piece of abnormal lung where the tissue grows faster than normal and forms abnormal air spaces, or cysts [SISS-ts]. It is the most common CPAM. Although it is usually found in the lower lung, a CCAM can grow in any part of the lung. Types of CCAMs include:
  - **Type I:** Large, cystic, fluid-filled mass
  - **Type II:** Fluid-filled microcyst (small cyst)
  - **Type III:** Solid mass (can be confused with a BPS)
- **Bronchopulmonary** [BRON-ko-PULL-moh-nehr-ee] **sequestration** [see-kwes-TRAY-shun] **(BPS).** A BPS is a piece of lung tissue that develops without being connected to the airway in the lung. This can develop either inside (intralobar) or outside (extralobar) the lung. The BPS can connect to surrounding structures, including parts of the lung or even the esophagus. In addition, the blood vessels that go to the BPS do not form normally. The BPS gets blood from the aorta, which is different than the normal lung.

- **Congenital lung emphysema** [em-fih-ZEE-muh], **(CLE).** CLE is a condition in which air enters the lungs but can't escape, causing the lungs to overinflate.
- **Hybrid lesion.** Sometimes a healthcare provider finds a combination of both a CCAM and a BPS. A hybrid lesion usually has an airway connection and an abnormal artery connecting it to the aorta.

A healthcare provider may not be able to tell what type of CPAM your child has until they're born and their lungs fill with air.

What causes a CPAM?

Doctors don't know what causes a CPAM, but it probably occurs during very early lung development. Nothing the mother does or has done during pregnancy causes this problem.

How is the CPAM diagnosed?

A healthcare provider diagnoses a CPAM when an ultrasound shows a baby has a cystic or solid lung mass. Rarely, the CPAM may grow large enough to push the heart to the side or press down on the diaphragm.

The healthcare provider may also perform a Doppler ultrasound to evaluate the lung mass. A Doppler ultrasound shows the blood flow in the lung and mass, which can help determine whether it is a CCAM or BPS. However, this isn't always possible.

How does my healthcare provider monitor the CPAM?

To watch the CPAM's size and location (it usually grows until 26 to 28 weeks into the pregnancy), a healthcare provider does several ultrasounds to get a measurement called **CVR**. A CVR:

- **Less than 1** means the CPAM is small and rarely leads to problems during or after pregnancy.
- **Greater than 1** is more concerning and typically requires close monitoring.
- **Greater than 1.6** can cause problems during pregnancy and after delivery. It must be monitored and may need early intervention.

Your healthcare provider may also recommend a fetal echocardiogram [ek-oh-CAR-dee-oh-gram] [ECG], a heart ultrasound to see the heart's function and structure.

These tests help the healthcare team tell whether your baby is likely to have problems so you can make the best possible decision about treatment.

How does a CPAM affect my baby?

Most babies with a CPAM develop normally with normal lung function. After 28 weeks gestation, the baby continues to grow faster than the developing lungs. This makes the CPAM appear to shrink and the CVR to go down.

The CPAM can cause heart and breathing problems, because it takes up room in the chest and can push on the heart and lungs. The CPAM's size and location determine how it will affect your baby.

- **A small CPAM (CVR less than 1) may not cause problems** and can be removed before your baby is 1 year old.
- **A larger CPAM (CVR greater than 1.6) can cause breathing trouble** and may need to be removed immediately after delivery.

A CPAM can grow too large and press on your baby's heart, which can cause heart failure. Fluid build-up in multiple areas of your developing baby (called hydrops) is a sign of heart failure. The CPAM can also press on the lungs and make them too small for your baby to survive.

How is a CPAM managed during pregnancy?

The fetal center will help determine the CPAM's size, location, and severity. Healthcare providers will tell you more about the CPAM, recommend ways to manage your pregnancy, and have you meet the specialists who'll care for your baby after delivery.

Many CPAMs, even large ones, can be managed with close follow-up and steroids. Most babies with CPAMs can be delivered close to their due date.

Questions for my doctor

Is there fetal intervention for CPAMs?

It is rare for a CPAM to grow large enough during pregnancy that there is a need to intervene while your baby is still in the womb. This may be an option if your baby has hydrops.

Under most circumstances, you will be given prenatal steroids first. Steroid injections can be effective, especially in large masses with small cysts (type II CCAM).

If the steroids don't stop the CPAM's growth, open fetal surgery to remove the mass can be a lifesaving option for your baby.

How is a CPAM managed during and after delivery?

Your baby will be evaluated and treated after delivery.

- **If your baby has a small CPAM (CVR less than 1),** you can usually deliver normally with your primary obstetrics provider. Healthcare providers will watch your baby closely in the hospital for breathing trouble. If your baby is doing well and is breathing normally, they can go home with you and see the pediatric surgeon for follow-up care.
- **If your baby has a moderate to large CPAM (CVR greater than 1),** you should deliver at the University of Utah Hospital. A pediatric surgeon will evaluate your baby and manage the CPAM in a neonatal intensive care unit (NICU) that specializes in CPAMs. Your baby may need oxygen or may even need a breathing machine (ventilator). They will be admitted to the NICU for close observation. The pediatric surgeon may recommend removing the CPAM before your baby goes home.

A CPAM does not go away on its own, so your baby should see the pediatric surgeon when they're about 3 months old. They should have a chest x-ray called a CTA before seeing the pediatric surgeon. This helps the surgeon see the CPAM and its blood supply and decide how to treat the CPAM (and whether it should be removed).

How is the CPAM removed?

A CPAM is usually removed thoracoscopically [thor-AS-oh-SCAH-pic-lee], or using a small camera, through 3 small openings on the chest. Most children who have this surgery will go home in 1 to 2 days.

The pediatric surgeon will tell you when and how the CPAM should be removed, what happens after surgery, and your child's expected recovery.

What outcome can I expect after surgery?

If your baby doesn't have any other problems, their outcome with a CPAM is generally very good. Almost all babies will develop normally without breathing or heart problems.

After surgery, your baby should not need long-term follow-up with the pediatric surgeon. They can see a pediatrician for care instead.

For additional information

Utah Fetal Center
intermountainhealthcare.org/locations/primary-childrens-hospital/medical-services/utah-fetal-center

Kids Health
kidshealth.org/en/parents/fetal-lung-mass.html?ref=search#catdigestive

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