The eL18-4 PureWave linear array transducer in the assessment of second trimester congenital pulmonary airway malformation (CPAM)

Overview
Diagnostic ultrasound is commonly used to diagnose fetal congenital pulmonary airway malformation (CPAM) and to follow lung volume ratios in order to assess prognosis and even possible in utero therapy options. CPAM was previously known as congenital cystic adenomatoid malformation.

A CPAM is a mass of abnormal fetal lung tissue that forms during pregnancy. This mass or lesion, is usually located in one lung and it does not function as normal tissue. A lesion can vary in size, appearance and location. It can change remarkably during pregnancy. Lesions can be filled with fluid or solid and can compress venous return and cause hydrops or death in the fetal state. Postnatally, they require removal secondary to the risk for malignant transformation. This condition is relatively rare, affecting about one in 25,000 pregnancies. Males are affected slightly more than females.

Patient history
A pregnant female patient was seen at 19 weeks and 4 days for an anatomy scan. The scan demonstrated a complex mass in the left chest measured at 2.86 x 2.79 x 3.42 cm, and the heart was deviated to the right side of the chest (Figures 1 and 2). The CPAM volume ratio (CRV) was 0.8, estimated using the formula for a prolate ellipse: CPAM volume = (length x height x width x 0.52).

A CRV ≤ 1.6 cm² at presentation suggested that the risk of hydrops developing in the absence of a dominant large cyst (cyst greater than 1/3 the size of the CPAM) was low. Lung lesions with a CRV < 1.2 cm² may be followed weekly; lesions 1.2 – 1.6 cm² twice per week depending on the gestational age and CRV ratio on the initial scan. A CRV > 1.6 cm² or a CPAM with a dominant cyst may be followed two to three times per week, as these states are high-risk for fetal sequelae.

It is common to see initial growth and then plateau or shrinking of the lesion, especially after 28 weeks.

The accuracy of the measurements thus is critically important for management, surveillance and prognosis. Traditionally the normal lung tissue can be quite difficult to tell apart from the CPAM tissue.
Due to the known CPAM seen in this fetus, the eL18-4 linear transducer was used to evaluate the mass at 33 weeks 4 days.

Diagnostic ultrasound is commonly used to assess fetal CPAM and volume ratios. Ultrasound has proven valuable in establishing sonographic images and indicators for the evaluation of fetuses at risk for hydrops and possible intervention. CPAM can change remarkably during pregnancy. The eL18-4 linear transducer's resolution is excellent for evaluating this CPAM throughout pregnancy, including later gestational age. These images clearly demonstrate the exceptional resolution of the eL18-4 linear transducer.

This 33-year-old G2P1001 female patient presented with no medical issues and prior pregnancy complicated by a right pelvic kidney which progressed to a multicystic dysplastic kidney on subsequent ultrasound studies (Figures 3 and 4). The family history of both parents was negative for congenital anomalies, chromosomal problems, genetic disorders, consanguinity or mental retardation.

The routine anatomy scan demonstrated a complex mass in the left chest. The mass measured 2.86 x 2.79 x 3.42 cm and the heart was deviated to the right side of the chest. There was a feeder vessel seen. The CRV was 0.8 cm² (Figure 5).

Protocol
With a routine anatomy scan, the maternal and fetal structures are evaluated. It is common to find some normal variations during the anatomy scan. In most anatomy scans, the C9-2 or C5-1 transducers are used for the evaluation (Figure 6). Here, the routine anatomy scan demonstrated a possible CPAM in the left chest; the heart was deviate to the right side of the chest, the CRV was 0.8 cm².

The following week, the CRV increased to 12 cm². The patient was referred to the Colorado Fetal Care Center (CFCC) for counselling and possible intervention. She was followed for several months at the CFCC and the ratio increased to the largest CRV of 1.6 cm². She was given steroids and the CRV dropped, with the most recent CRV of 0.8 cm² at 33 weeks 4 days. There were no signs of fetal hydrops, polyhydramnios or other issues.

The first image utilizing the C9-2 curved array transducer demonstrated the CPAM in the transverse plane (Figure 1) and the next image in the longitudinal plane (Figure 2). These images were taken prior to obtaining the eL18-4 linear transducer.

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Conclusion
Diagnostic ultrasound is commonly used to assess fetal CPAM and volume ratios. Ultrasound has proven valuable in establishing sonographic images and indicators for the evaluation of fetuses at risk for hydrops and possible intervention. CPAM can change remarkably during pregnancy. The eL18-4 linear transducer's resolution is excellent for evaluating this CPAM throughout pregnancy, including later gestational age. These images clearly demonstrate the exceptional resolution of the eL18-4 linear transducer.

Results from case studies are not predictive of results in other cases. Results in other cases may vary.

References
1 Congenital Pulmonary Airway Malformation. Cincinnati Fetal Center, Cincinnati Children’s Hospital Burnet Campus, 3333 Burnet Ave, Cincinnati, Ohio, 45229. https://www.cincinnatichildrens.org/service/f/fetal-care/conditions/congenital-pulmonary-airway-malformation