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Congenital lung lesions: Prenatal diagnosis and intervention

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ABSTRACT

Congenital lung lesions are common sonographic findings in pregnancy, usually detected at the routine 20 weeks scan. The most common is cystic adenomatous malformation of the lung (CCAM). This usually causes few prenatal problems; however, fetal hydrops occurs in about 5%. Prenatal intervention for these is possible in many to allow survival to birth. Bronchoplumonary sequestration (BPS), with an aberrant "feeder" vessel arising from the aorta may co-exist but is detectable as a separate entity by visualization of this vessel. Symptomatic or curative prenatal intervention is again possible in the few severe cases where hydrops or pleural effusions develop. Pleural effusions may be due to a primary leak usually of chylous fluid: prenatal thoracoamniotic shunting may prevent pulmonary hyoplasia or cure the consequent fetal hydrops. More often, however, effusions are a consequence of an underlying abnormality, including many structural or chromosomal abnormalities that may also cause co-existing fetal hydrops. Congenital high airway obstruction (CHAOS) is commonly fatal but cases potentially amenable to prenatal intervention or to immediate perinatal management may be identified using ultrasound or MRI.

Introduction

The incidence of congenital lung lesions has increased due to the detection of these lesions on prenatal screening scans in mid trimester. With increase in experience and better scanning machines, the lesions may be further defined as solid, cystic, or with the presence of an aberrant vessel. Complex lesions may be amenable to intervention to salvage the pregnancy. At any stage the maternal risk will always override the fetal risk. This article covers the diagnostic and therapeutic modalities available for prenatal lung lesions.

Pleural effusion

There is a collection of fluid in the pleural space(s) in the fetal thorax, more commonly on the right. Pleural effusions can be primary, or as a part of generalized fetal edema or hydrops.

The etiology of primary and secondary effusions overlaps: fluid accumulation in the chest, such as congenital chylothorax, may lead to fetal hydrops in severe cases, presumably due pressure on the heart and blood vessels in the fetal chest. Equally the fluid may simply be isolated, as well as part of fetal hydrops.

Effusions may occur with many fetal abnormalities, in particular chromosomal abnormalities or genetic syndromes, such as Trisomy 21, Turner's syndrome and Noonan's syndrome, or structural abnormalities such as bronchopulmonary sequestration, or viral infections such as CMV or parvovirus. Where hydrops is present, the most common additional underlying causes are structural, particularly cardiac, disease and fetal anemia due to blood group incompatibility, parvovirus infection, or fetomaternal hemorrhage.

Fetal hydrops predisposes to maternal pre-eclampsia, or "mirror syndrome." Under these circumstances, delivery is indicated for maternal safety, but reversal of the syndrome with successful treatment of the hydrops has been described.[1]

Sonographic appearances

Sonographic appearances differ according to severity, from only a thin hypoechogenic rim of fluid around the lungs to severe accumulation of fluid with collapsed lung unilaterally or bilaterally (Figure 1). Fetal hydrops and other structural abnormalities may be present.

Pregnancy management

Primary assessment should include a detailed anomaly scan, with fetal echo, and fetal Doppler studies in particular middle cerebral artery (MCA) Doppler, karyotyping usually by amniocentesis, a check of maternal CMV and toxoplasmosis

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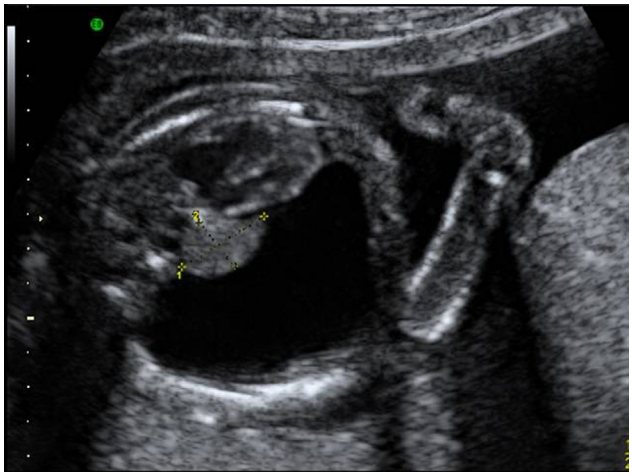

Fig. 1. Isolated pleural effusion.

testing and parvovirus, and maternal antibodies such as red cell alloimmunisation.

For small isolated effusions, observation is appropriate and these may appear to come and go. The management is for the underlying cause. If none is found, the possibility of congenital chylothorax should be considered. A large lesion may lead to pulmonary hypoplasia or progress to hydrops. Both may be prevented by fetal intervention. Options available in pregnancy include thoracocentesis, thoracoamniotic shunt, pleurodesis, and intrapleural injection of autologous blood. Thoracocentesis can be used as diagnostic and therapeutic measure. Pleural fluid is aspirated and checked for cell count, viral screen, and cultures. In most of the cases, however, fluid will accumulate again and a thoracoamniotic shunt will be appropriate.[2]

Possibly more commonly, the fetus is hydropic by the time shunting is considered. Shunt placement is a highly invasive procedure with a risk of complications, ie, premature rupture of membranes and preterm labor (approximately 15%), bleeding, infection, shunt migration, blockage (need for reshunting), and injury to the fetus, including intercostal artery laceration. Shunting may reverse fetal hydrops if the pleural effusion(s) has caused the hydrops, eg, for congenital chylothorax, although the sequence of events may not be clear before the procedure. A survival rate of approximately 50% is to be expected where a hydropic fetus with no other discernible cause undergoes shunting.[3]

Where generalized hydrops due to fetal anemia is present, the middle cerebral artery (MCA) peak systolic velocity will almost invariably be raised. Under these circumstances, in utero blood transfusion will usually lead to reversal of the hydrops. This is most appropriate for parvovirus infection or for red cell alloimmunisation. Where the fetus is not anemic or shunting has failed, the prognosis is usually very poor.

It is essential that observation for the development of pre-eclampsia is performed. Polyhydramnios, increased volume of amniotic fluid, may develop and, if extreme, can be treated with amnioreduction. The aim is to reduce the risk of extreme preterm labor.

Where a fetus with large pleural effusions with or without hydrops is to be delivered, thoracocentesis just before delivery may allow easier immediate postnatal ventilation.

Congenital cystic adenomatoid malformation (CCAM) of the lung

Congenital cystic adenomatoid malformation (CCAM) of the lung develops during first 6 weeks of gestation. The exact pathogenesis is still unknown. There is failure of maturation of bronchiolar structures during pseudoglandular stage of lung development. It results in overgrowth of the terminal bronchioles without corresponding alveoli. CCAM communicates with the tracheobronchial tree and has normal blood supply from pulmonary arteries. The incidence of prenatally diagnosed lesions has increased with better ultrasound usage and resolution. Approximately 95% of antenatally diagnosed CCAMs will be born alive.

Sonographic appearances

A CCAM is usually detected at the “anomaly” scan at 18–21 weeks’ gestation, can affect any lobe of the lung as a mass within the lung tissue, and is usually unilateral. It has been categorized into 3 types: Type I is macrocystic (25%) and is characterized by 1 or more cysts of variable size up to 10 cm (Figure 2), Type II (25%) is mixed with cysts with adjacent area of increased echogenicity on ultrasound, and Type III (50%) is microcystic (< 5 mm) and appears as an echogenic area within the fetal lungs (Figure 3). Where there is no hybrid (co-existing pulmonary sequestration) element, no separate blood supply from the aorta will be seen as perfusion is from the pulmonary vasculature; with hybrid lesions, a separate blood supply from the aorta, including from below the diaphragm, can often be seen using color or power Doppler.

With larger lesions there is often mediastinal shift and the lesion appears to cross the midline and may cause flattening of the diaphragm. Polyhydramnios may occur because of esophageal compression and overproduction of fluid by the lung lesion but is seldom severe unless there is hydrops. Non-immune hydrops, the accumulation of fluid in 2 or more parts the fetus, is reported to occur in approximately 5% of antenatally diagnosed lesions referred to a tertiary center.[4] It is probably due to inferior vena cava and cardiac compression. The cystic adenomatoid malformation volume ratio (CVR) is calculated from the size of the lesion and head circumference (HC), (length × height × width × 0.52)/HC. A CVR of more than 1.6 is predictive of increased risk of hydrops, with an 80% of fetuses developing hydrops.[5]

Pregnancy changes and management

A detailed anomaly scan and fetal echo are required. Associated abnormalities are unusual but occur more often with Type II. Lung echogenicity, particularly in CCAM type III, alters over gestation.

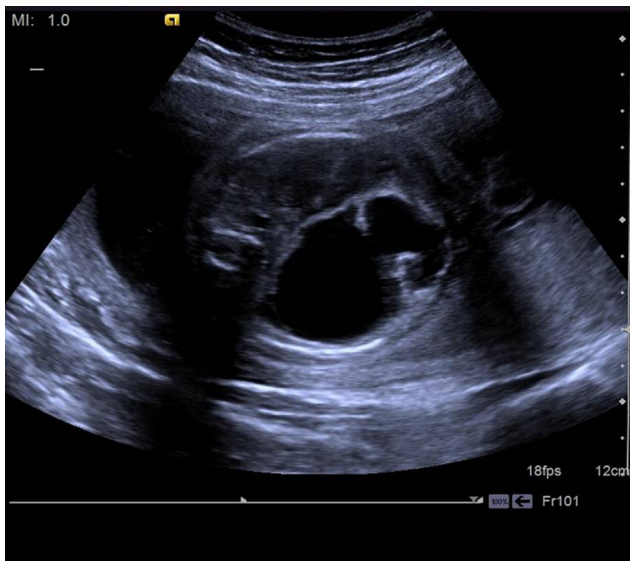

Fig. 2. Large Type I CCAM.