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Prenatal prediction of pulmonary hypoplasia

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A B S T R A C T

Pulmonary hypoplasia, although rare, is associated with significant neonatal morbidity and mortality. Conditions associated with pulmonary hypoplasia include those which limit normal thoracic capacity or movement, including skeletal dysplasias and abdominal wall defects; those with mass effect, including congenital diaphragmatic hernia and pleural effusions; and those with decreased amniotic fluid, including preterm, premature rupture of membranes, and genitourinary anomalies. The ability to predict severe pulmonary hypoplasia prenatally aids in family counseling, as well as obstetric and neonatal management. The objective of this review is to outline the imaging techniques that are widely used prenatally to assess pulmonary hypoplasia and to discuss the limitations of these methods.

1. Introduction

Pulmonary hypoplasia is a rare condition, affecting nine to 11 per 10,000 live births. The diagnosis includes a spectrum of respiratory complications ranging from neonatal death to less severe manifestations, including chronic respiratory failure, pulmonary hemorrhage, bronchopulmonary dysplasia, or even transient respiratory distress [1–4]. The condition is caused by defective development leading to reduction in the number of lung cells, airways, and alveoli, resulting in lungs sufficiently small to impair gas exchange [2,5]. The severity of pulmonary hypoplasia relates to the timing of injury in relation to embryologic lung development [6,7]. Despite advances in neonatal support, mortality approaches 70% in most series [4]. Decreased lung weight:body weight ratio and low radial alveolar counts are used to make a postmortem diagnosis [8,9]. In survivors, the postnatal diagnosis of pulmonary hypoplasia is made in the setting of severe respiratory insufficiency, requiring high ventilatory support in the absence of obstruction or with abnormal radiologic findings (elevated diaphragm, bell-shaped chest) [4,10].

Prenatal counseling of patients at risk for a fetus with pulmonary hypoplasia is challenging. The more severe cases may be diagnosed prenatally but that is a small percentage of at-risk individuals. The ability to accurately predict pulmonary hypoplasia prior to delivery would improve counseling of parents. More accurate counseling could aid in obstetric management, guide neonatal resuscitation, and determine surgical management in cases with structural abnormalities, such as congenital diaphragmatic hernia or giant omphalocele. Current methods for the prediction of pulmonary hypoplasia and the limitations of these methods for accurate prenatal diagnosis are important to understand for appropriate counseling.

2. Fetal lung development and etiology of pulmonary hypoplasia

2.1. Stages of fetal lung development

The fetal lung develops and changes throughout gestation and goes through the following stages: embryonic, pseudoglandular, canalicular, saccular, and alveolar [11]. During the embryonic stage, which is completed by the eighth week of gestation, the lungs arise from the ventral aspect of the foregut and begin branching [12]. By the end of this stage, the major airways are formed and mesenchyme adjacent to the airways begins to develop into blood vessels, smooth muscle, and connective tissue [7,12].

The pseudoglandular stage, which is completed by the 16th week, is a period of continued branching of the bronchial tree [2]. During this period, interactions between the epithelial airways and surrounding mesenchyme result in budding and development of the acini, so that all of the major elements of the conduction system are formed [2,13]. The pulmonary arteries and veins also develop in a pattern corresponding to mature lung during this time [13].

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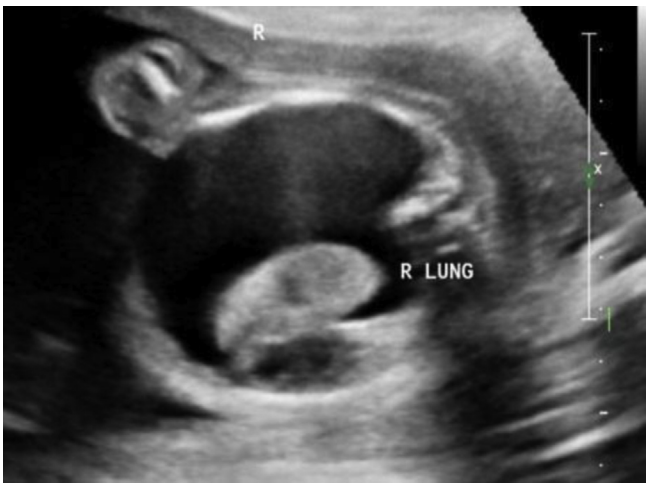

Fig. 1. Transverse section through the fetal chest showing bilateral lung compression with unilateral right pleural effusion.

The canalicular stage, which overlaps with the end of the pseudoglandular stage and extends to the 28th week, is the period during which gas exchange first becomes possible [2]. The airway epithelium thins and differentiates into type I and type II pneumocytes that are responsible for gas exchange and surfactant production, respectively [2,7].

The saccular stage extends from the 28th week until birth, during which time the acini further develop with a rapid increase in the gas exchange surface of the lung and thinning of the interstitium [2,12]. The type II pneumocytes also mature during this time-period [12].

The alveolar stage begins in the late third trimester and continues after birth with maturation of the alveoli and surrounding capillary network [2].

2.2. Etiology of pulmonary hypoplasia

Normal structural and functional lung development as outlined above requires adequate thoracic space for the growing lungs, as well as sufficient fluid exchange to distend the developing airways. Many fetal and obstetric complications may lead to pulmonary hypoplasia, and the timing of these insults influences the severity of pulmonary disease [4].

Conditions resulting in inadequate thoracic space may be related to restriction of the thorax, as is seen with lethal skeletal dysplasias. In these cases, the heart occupies a large portion of the fetal chest and there is a typical bell-shaped chest with small thoracic size. In pulmonary hypoplasia from mass effect, the result will vary depending upon the gestational age when the mass effect was initiated. It will also depend upon the tissue characteristics of the mass itself and the corresponding pressure applied to the fetal lungs. Pulmonary hypoplasia secondary to mass effect can be seen with large pleural effusions (Fig. 1), congenital diaphragmatic hernia (CDH) (especially if liver is present in the fetal chest), or congenital pulmonary airway malformation (CPAM). Restriction of normal diaphragmatic movement with subsequent failure to fully expand the chest can be seen with neuromuscular disorders and abdominal wall defects. Long-standing oligohydramnios may be associated with preterm premature rupture of membranes (PPROM), renal anomalies, or lower genitourinary tract obstruction. This leads to inadequate fluid exchange, which is necessary for adequate pulmonary development.

3. Prenatal diagnosis of pulmonary hypoplasia

3.1. Two-dimensional ultrasonography

Many biometric parameters using two-dimensional ultrasound have been used to assess risk for pulmonary hypoplasia. The most frequent are thoracic circumference (TC), thoracic circumference:abdominal circumference ratio (TC:AC), and lung area (LA). These measurements are taken in an axial plane at the level of the four-chamber view of the heart (Fig. 2) [14–16]. TC and LA are

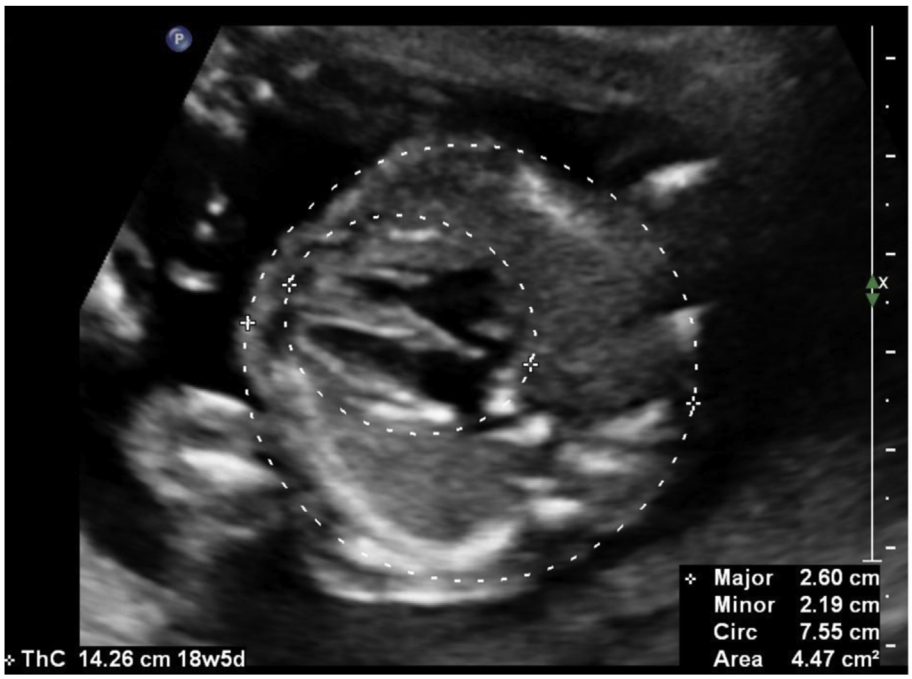

Fig. 2. Four-chamber view of heart demonstrating plane for thoracic circumference and lung area.

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