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Predicting perinatal outcome in isolated congenital diaphragmatic hernia using fetal pulmonary artery diameters

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Key words:

Pulmonary arteries; Congenital diaphragmatic hernia; Fetal lung; Pulmonary hypoplasia; Fetal malformation;

Predictors of mortality

Abstract

Objective: The aim of the study was to evaluate the potential of fetal pulmonary artery (PA) diameters to predict perinatal death and pulmonary arterial hypertension (PAH) in congenital diaphragmatic hernia (CDH).

Study Design: In this prospective observational study, observed PA (main, right, and left) diameters were measured at the level of the 3 vessels in 21 fetuses with isolated CDH and in 85 controls at 22 to 36 weeks. The observed/expected (o/e) diameters of the main, contralateral, and ipsilateral PAs were calculated by comparing these measurements with reference values obtained in our previous study and correlated with perinatal death and postnatal PAH.

Results: The o/e PA diameters were significantly reduced in fetuses with CDH compared to controls (P < .001) and in fetuses with CDH who died (P < .050). However, there was no significant association between PA diameters and PAH $(P \ge .050)$.

Conclusions: The PA diameters might be useful to predict perinatal death in isolated CDH but not postnatal PAH, suggesting that PA diameters are probably related to the severity of pulmonary hypoplasia.

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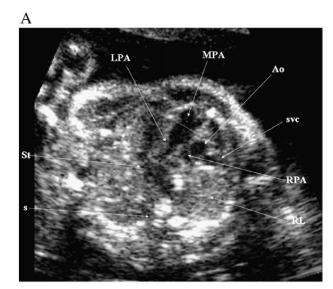
Congenital diaphragmatic hernia (CDH), whose incidence is approximately 1 in 2200 livebirths [1], presents an overall neonatal mortality rate of 50% in cases with a

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prenatal diagnosis [2]. Prenatal prediction of neonatal prognosis remains a challenge and is crucial for immediate neonatal care and for the selection of candidates for prenatal therapy such as intermittent tracheal occlusion by fetoscopy [2,3].

Because neonatal mortality is directly related to severe pulmonary hypoplasia, many prognostic factors have been suggested based on either direct (fetal lung volumes [4-7], lung-

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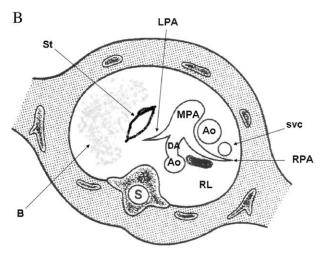


Fig. 1 A, Cross-sectional ultrasound image through the fetal chest showing the site of measurement of the main and branch PA diameters in a fetus with CDH at 24 weeks of gestation. B, Diagram of a cross-sectional ultrasound image through the fetal chest showing the site of measurement of the main and branch PA diameters in a fetus with CDH. MPA indicates main pulmonary artery; RPA, right pulmonary artery; LPA, left pulmonary artery; Ao, aorta; svc, superior vena cava; RL, right lung; s, spine; St, stomach herniated in fetal thorax.

to-head ratio (LHR) [8,9], and lung diameter—to—thoracic circumference ratio [10]) or indirect (liver and stomach positions [8,11-13], left-right ventricular ratio, amniotic fluid volume, and mediastinal shift) [14] prenatal assessment of lung size. However, experience shows that neonatal death occurs even in a few cases despite markedly high pulmonary weights and volumes, mainly when the onset of pulmonary arterial hypertension (PAH) has been linked to neonatal death. Recently, we have demonstrated that it is possible to evaluate quantitatively pulmonary vascularization by a 3-dimensional power Doppler histogram in 21 cases with isolated CDH, which may improve the prediction of neonatal outcome (death or PAH) [15]. However, this technique is still operator dependent.

In 2000, Suda et al [16] demonstrated that postnatal main pulmonary artery (MPA) diameter is correlated with postnatal death in infants with CDH, suggesting that PA

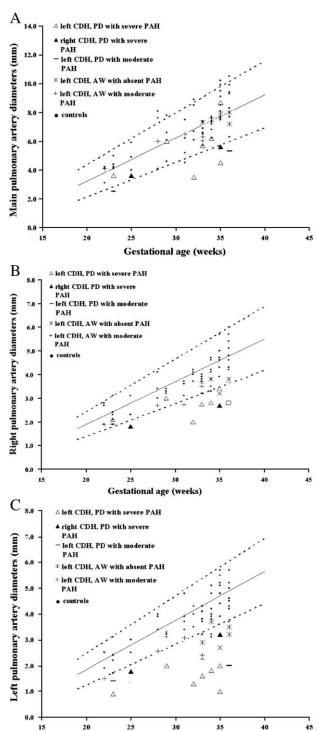


Fig. 2 A, Nomograms of MPA diameters in 21 fetuses with CDH and in 85 controls plotted against gestational age. B, Nomograms of RPA diameters in 21 fetuses with CDH and in 85 controls plotted against gestational age. C, Nomograms of LPA diameters in 21 fetuses with CDH and in 85 controls plotted against gestational age. PD indicates perinatal death; AW, alive and well.

Gestational age (weeks)

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