

MP1-11

PULMONARY HYPOPLASIA ASSOCIATED TO CONGENITAL HEART DISEASES: A FOETAL STUDY

*Bonnet D. (1,5), Ruchonnet-Metrailler I (2), Bessieres B.(3), Vibhushan S.(4), Delacourt C.(2,5)
AP-HP, Hôpital Necker-Enfants Malades, Cardiologie Pédiatrique, M3C-Centre de référence pour les Malformations Cardiaques Congénitales Complexes Paris, France (1)
AP-HP, Hôpital Necker-Enfants Malades, service de Pneumologie Pédiatrique, Centre de Référence pour les Maladies Respiratoires Rares de l'Enfant, Paris, France (2)
AP-HP, Hôpital Necker-Enfants Malades, Cytogénétique, Paris, France (3)
INSERM, U955, IMRB, Equipe 04, Créteil, France (4)
Université Paris-Descartes, Paris, France (5)*

Background. Abnormalities of the fetal pulmonary vasculature may alter lung morphogenesis. Postnatal studies have suggested that pulmonary hypoplasia (PH) may be associated to congenital heart diseases (CHDs). However, no fetal study have been carried out to determine whether these abnormalities have a common developmental origin, early in morphogenesis, or whether they gradually become established after birth.

Objective. To determine the prevalence of PH associated with CHDs, and to identify the types of CHDs associated with the highest risk for lung growth impairment.

Methods. Between January 2006 and December 2010, fetuses with CHD obtained following the termination of pregnancies due to fetal abnormalities were examined in a prospective manner for the detection of heart and lung defects. CHDs were classified into five pathophysiological groups. PH was defined as a fetal lung weight to body weight ratio <0.015 before 28 weeks, and < 0.012 after 28 weeks. The expression of CD31 and VEGF in lungs was evaluated by immunohistochemistry.

Results. PH was detected in 15 of the 119 fetuses analyzed (13%). It was significantly associated with CHD with right outflow obstruction, independently of chromosomal abnormality and associated extra-cardiac anomaly ($p < 0.03$). Right outflow obstruction was present in 60% of fetuses with CHD and PH, but in only 32% of those with CHD but no PH. In fetuses with right outflow obstruction, no difference was observed between those with PH and those without PH, in terms of the ratio of the pulmonary artery diameter to the aortic diameter, lung CD31 expression, or lung VEGF expression.

Conclusion. CHDs with right outflow obstruction are a significant risk factor for prenatally acquired PH. The occurrence of fetal PH is not correlated to abnormalities of the pulmonary vasculature, suggesting the involvement of perfusion-independent mechanisms.