Microscopic Lung Airway Abnormality and Pulmonary Vascular Disease Associated with Congenital Systemic-to-Pulmonary Shunt

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Links between impaired lung development/growth and pulmonary hypertension (PH) have been demonstrated in infants with bronchopulmonary dysplasia or congenital diaphragmatic hernia. Experimental studies also suggest that a complex interplay of epithelial-endothelial cells is required for normal pre/postnatal lung

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morphogenesis [1]. In this chapter, we focus on the role of lung development/growth abnormality in the development of pulmonary vascular disease (PVD) associated with congenital heart defect (CHD).

Children with large systemic-to-pulmonary shunt typically exhibit symptoms of heart failure in early infancy (tachypnea, failure to thrive), have PH with increased pulmonary blood flow and do not develop irreversible PVD if closure of defect is performed before 9 months to 2 years of age [2]. In general, the patient's age and type of lesion contribute to the risk of developing irreversible PVD. Conditions with pressure overload and high flow, such as large ventricular septal defect or patent ductus arteriosus, are likely to cause PVD earlier than low-pressure, high-flow lesions (pre-tricuspid shunt such as atrial septal defect). However, we sometimes encounter children with "atypical" PH which is associated with CHD but unexplained by the type or magnitude of coexisting CHD (e.g., pre-tricuspid shunt or small post-tricuspid shunt with severe PH and elevated pulmonary vascular resistance [PVR]; large post-tricuspid shunt lacking symptom of heart failure due to high PVR sustained from neonatal period).

Our recent observation suggests that microscopic airway abnormality caused by genetic or pre/postnatal conditions may be associated with PVD in infants with "atypical" PH.

References

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