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50 Years Ago in *THE JOURNAL OF PEDIATRICS*

An Early Description of Persistent Pulmonary Hypertension of the Newborn

Siassi B, Goldberg SJ, Emmanouilides GC, Higashino SM, Lewis E. Persistent Pulmonary Vascular Obstruction in Newborn Infants. *J Pediatr* 1971;78:610-5.

In 1971, Siassi et al described 5 full-term infants with delayed transition from fetal to neonatal circulation. All developed cyanosis and tachypnea soon after birth without grunting or retractions. On radiographs, findings of parenchymal lung disease such as granularity and air bronchograms were notably absent. An electrocardiogram and diagnostic cardiac catheterization were performed to exclude congenital heart disease. These tests showed a structurally normal cardiac anatomy with right ventricular hypertrophy, right-to-left shunting at the ductus arteriosus, and a pulmonary artery blood pressure equal to or greater than the aortic blood pressure. All infants received supplemental oxygen, and 4 of the 5 survived to discharge. One died on the third day, and autopsy of this infant showed smooth muscle hypertrophy of pulmonary arterioles.

In the intervening 50 years, the diagnosis and treatment of persistent pulmonary hypertension of the newborn (PPHN) has seen a dramatic evolution. Although most cases of PPHN are mild, severe PPHN at the time of this publication was commonly lethal, as it was in one of the infants described in the series. In the 1980s, extracorporeal membranous oxygenation was shown to be effective at decreasing mortality in severe PPHN.¹ In 1999, the US Food and Drug Administration approved the use of inhaled nitric oxide for PPHN, and it has since become the mainstay of therapy, sparing most patients from extracorporeal membranous oxygenation.² Pulse oximetry and echocardiography, noninvasive alternatives to cardiac catheterization, are now the standard of care for the evaluation of this disease. The rapid advancement in the treatment of PPHN has paralleled the progress of research in the field of pulmonary vascular biology. Hopefully, continued work in this field will lead to similar improvements in the outcomes of patients affected by other forms of neonatal pulmonary vascular disease.

Maxwell Mathias, MD

Division of Neonatology

Ann and Robert H. Lurie Children's Hospital

Chicago, Illinois

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