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50 Years Ago in *The Journal of Pediatrics*

Hepatopulmonary Syndrome: Transplantation Is the Big Difference

Kravath RE, Scarpelli EM, Bernstein J. Hepatogenic cyanosis: arteriovenous shunts in chronic active hepatitis. J Pediatr 1971;78:238-45.

Fifty years ago, Kravath et al presented a girl with chronic hepatitis, portal hypertension, cyanosis, spider nevi, extertional dyspnea, and clubbing. From the onset of symptoms at the age of 10 years until she died 6 years later, she had several hospital admissions. With advancing liver disease, she had lower oxygen saturation in blood, lower arterial PaO_2 in, and a low single-breath carbon monoxide diffusing capacity. Postmortem examination revealed pulmonary arteriovenous anastomoses and dilated vascular channels.

Hepatopulmonary syndrome is characterized by impaired arterial oxygenation induced by intrapulmonary vascular dilatation in the setting of liver disease, portal hypertension, or congenital portosystemic shunts. In 1884, a woman with liver cirrhosis and cyanosis was described, and arteriovenous fistulas and dilatation of pulmonary vessels were found in 1956 thus in 1977 the term hepatopulmonary syndrome was suggested.[1-3]

Hepatopulmonary syndrome evolves insidiously in some children with chronic liver disease and is accompanied by complications and higher mortality. In severe cases, oxygen therapy may be required. Even though intrapulmonary vascular dilation could be subclinical without hypoxemia, pulse oximetry may be useful for screening. The mechanisms of hepatopulmonary syndrome are likely to involve endogenous vasodilators and pulmonary vascular remodeling.[4]

Fifty years ago, the severe liver disease progressed slowly despite a thorough workup and this patient eventually developed a coma and died. Hepatopulmonary syndrome often resolves after liver transplantation and the availability of transplantation is the major difference from the situation 50 years ago.

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