Hepatopulmonary Syndrome: A Rare Complication of Chronic Liver Disease in Children

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Abstract

An 11-year-old boy with congenital hepatic fibrosis presented with cyanosis at the National University Hospital. Echocardiogram revealed a structurally normal heart with good ventricular function. A pulmonary cause of his cyanosis was suggested on macroaggregated albumin scan and selective pulmonary artery angiogram. Arterial hypoxaemia secondary to intrapulmonary arteriovenous shunting in chronic liver cirrhosis can lead to permanent cyanosis. The potential for a complete reversal of this condition after liver transplantation indicates that arterial hypoxaemia, rather than being a contraindication, should be a reason for early liver transplantation.


Key words: Arteriovenous, Cyanosis, Hepatic fibrosis, Pulmonary, Transplant

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