제목: Incidence and clinical significance of intrapulmonary shunt in biliary atresia

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초록:

**Purpose:** Intrapulmonary arteriovenous shunt (IPS) is one of the long-term complications in chronic liver disease. If arterial oxygen defect in patients with IPS with chronic liver disease is found, hepatopulmonary syndrome (HPS) can be diagnosed and liver transplantation is needed for the treatment. However, it is not well described which characteristics of patients with IPS can be developed to HPS. Therefore, we investigated the incidence and the clinical significance of IPS in biliary atresia.

**Methods:** We prospectively evaluated the 72 patients with biliary atresia during March 2010 to May 2013. For the diagnosis of IPS, contrast-enhanced echocardiography was performed. For the confirmation of HPS, arterial blood-gas analysis (ABGA) was conducted additionally. Clinical data were reviewed retrospectively by grouping of non-IPS group, IPS without HPS group, and HPS group.

**Results:** IPS was identified in 41 patients (56.9%). Except six patients who did not examined ABGA after identification of IPS, IPS without HPS group was confirmed in 20 patients and HPS was diagnosed in 15 patients (20.8%). Bilirubin level at IPS evaluation was significantly increased by groups (Total: 0.6mg/dL, 1.5mg/dL, and 1.9mg/dL, p=0.005, Direct: 0.2mg/dL, 0.8mg/dL, and 1.2mg/dL, p=0.001 respectively). Liver stiffness score was significantly increased by groups (8.7kPa, 15.5kPa, and 29.3kPa, p<0.0001). The liver transplantation after evaluation of IPS significantly more performed according to groups (0%, 5%, 33.3%, p=0.001, respectively).

**Conclusions:** The worse clinical outcomes, such as bilirubin level and liver stiffness score were appeared in HPS group than IPS only group. The presence of IPS in biliary atresia might be considered as the transient step forward to HPS. Therefore, the identification of IPS and the close-monitoring of patients with IPS in biliary atresia is the substantial in that favorable outcome by the early liver transplantation.