

## MP3-9

### Contrast Echocardiography for Hepatopulmonary Syndrome Risk Stratification in Children With Normal Arterial Oxygenation

Chinali M., Pietrobattista A., Esposito C., del Pasqua A., Basso M.S., Franceschini A., Liccardo D., Nobili V., Rinelli G.  
Bambino Gesù Children Hospital, IRCSS

**BACKGROUND:** Hepatopulmonary syndrome (HPS) is defined as a triad including liver disease (and/or portal hypertension), intrapulmonary vascular dilatations (IPVD) and abnormal arterial oxygenation, and is associated with a pretransplantation mortality of 25%-46% in children. However, in the early phases of the disease abnormal oxygen saturation might not be found and thus echocardiographic identification of IPVD by bubble study contrast echocardiography (CE) is of paramount importance for risk stratification. Aim of this study was to determine the prevalence and associated signs and symptoms of IPVD (diagnosed by CE) in children with liver disease and portal hypertension without HPS.

**METHODS:** The study population included 44 patients with liver disease and portal hypertension with  $SpO_2 \geq 99\%$  (20 girls; age  $12 \pm 4$  years, range 2-18). IPVD were diagnosed by using CE and they were based on the presence of microbubbles in the left atrium with  $>3$  cycles and they were graded according to the Barzilai score (grade I to IV). Patients with evidence of intracardiac shunt were excluded ( $n=3$ ).

**RESULTS:** Specific diagnoses for the patients included: portal vein cavernoma (58%), biliary atresia (27%), congenital hepatic fibrosis (10%) and others (5%). Among the 41 included patients, only 10 patients showed no evidence of pulmonary shunting, while a positive CE was found in 31 patients (or 76%). In details: Grade I was found in 14 (34%) patients, Grade II in 8 (20%), Grade III in 4 (10%) and Grade IV in 5 (12%). A significant association was found between the severity of the disease, according to the semi-quantitative Barzilai score and the total serum proteins ( $r = -0.325$ ) and INR values ( $r = 0.261$ ; both  $p < 0.05$ ). No association was found between the length of the disease and the anthropometric parameters (weight, length, BMI, Z score BMI) with the presence of a shunt. A comparison of the groups, according to the Barzilai score at the CE, demonstrated a significant association with the presence of the esophageal varices ( $p$  for trend = 0.042; Figure).

**CONCLUSIONS:** Prevalence of positive CE in children with liver disease is a common finding. Accurate grading of positive CE is of paramount importance, as despite normal arterial oxygenation in the presence of significant shunting (Barzilai grade  $>II$ ), signs of disease severity can be found, including abnormal blood coagulation associated with the presence of significant esophageal varices, thus identifying pediatric patients at high risk of HPS development.

Prevalence of Esophageal Varices According to Pulmonary Shunt Grading by Contrast Echocardiography

