

## MP2-17

### Contrast echocardiography for screening of pulmonary arterio-venous fistula in children with portal hypertension

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#### Introduction:

The hepato-pulmonary syndrome consists of intrapulmonary shunting (IPS) secondary to pulmonary arterio-venous fistulas (PAVF) in the setting of chronic liver disease and portal hypertension. The objective of this study was to establish the diagnostic value of contrast echocardiography (CE) as a screening technique for IPS in children with portal hypertension using lung scintigraphy as the gold standard.

#### Methods:

CE was performed in children with portal hypertension using an intravenous line inserted preferably in the antecubital region. CE was done using microbubbles created by hand-agitated saline solution. IPS positivity was defined as the appearance of microbubbles in the left atrium within < 5 heart cycles after complete opacification of the right atrium. A lung scintigraphy was performed within 4 weeks with IPS presence graded from 1-4 according to the amount of shunt.

#### Results:

30 children with portal hypertension underwent CE and lung scintigraphy. The mean age was  $9.9 \pm 3.8$  years (2.1-16.6), with no difference in gender distribution (18 girls, 60%). The origin of portal hypertension was pre-hepatic in 6 (20%), intra-hepatic in 22 (73%) and secondary to a congenital or surgical shunt in 2 (7%). Scintigraphy was negative in 24 (80%) and positive for IPS in 6 (20%). All patients with a negative lung scintigraphy had a negative CE. In the 6 patients with a positive lung scintigraphy, 5 had a positive CE while 1 had a negative CE (false negative). The sensitivity of CE was 83% (CI 36.5-99.1) with a specificity of 100% (CI 82.8-100). The negative likelihood ratio was 17% (CI 0.03-0.99) and the negative predictive value 96%.

#### Conclusion:

CE is a simple and reliable screening tool for PAVF and IPS in children with portal hypertension, with a sensitivity of 83% and a negative predictive value of 96%. Technical limitations may be an issue particularly in small children when rapid saline injection is limited by the small size of the intravenous catheter. Detection of IPS is of utmost importance because hepato-pulmonary syndrome is now considered an indication for liver transplantation since the shunting disappears following liver transplantation.

**Table:** Sensitivity, Specificity, Positive (LHR +) and Negative (LHR -) Likelihood Ratio Values, Positive (PPV) and Negative (NPV) Predictive Values of Contrast Echocardiography for Shunt Prediction

Sensitivity (% [95% CI])	Specificity (% [95% CI])	LHR + (% [95% CI])	LHR - (% [95% CI])	PPV (%)	NPV (%)
83.3 [36.5 - 99.1]	100 [82.8 - 100]	$\infty$	0.17 [0.03 - 0.99]	1	0.96