

## MP4-4

### Fontan-associated liver disease in patients after Fontan palliation

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**Objectives:** Despite improved long-term survival Fontan palliated patients are prone to the development of Fontan-associated liver disease (FALD) including liver fibrosis, liver cirrhosis and hepatocellular carcinoma. In this study we aimed to develop a diagnostic algorithm for early detection of FALD and identify patients at risk.

**Methods:** In the last 30 years 350 patients received Fontan palliation for single ventricle anatomy in our institution. To identify patients at risk for FALD we extended our routinely performed follow-up program with a detailed hepatic assessment: In the last 12 months 53 patients of the total cohort (median age 19.4 years [7; 49]) underwent a detailed age-adjusted liver examination including laboratory analysis, liver ultrasound (n=24), transient elastography (n=30), invasive measurement of Fontan pressure (n=29) and liver vein wedge pressure (n=10). In 12 patients a biomarker test (Fibrotest) was performed to estimate the level of liver fibrosis.

**Results:** Liver ultrasound revealed hepatic parenchymal changes in 18 of 24 patients. Most common findings were heterogeneous parenchymal echotexture, segmental atrophy/hypertrophy or destroyed intrahepatic vascular architecture. In 6 of 24 patients (median age 33.7 years [19; 47]) severe liver cirrhosis was detectable. The presence of hepatic parenchymal changes was associated with the duration of the Fontan circulation (median duration 13.3 years [1; 25];  $p=0.04$ ) and elevated liver vein wedge pressure ( $p=0.05$ ). Fontan duration also showed a strong correlation with hepatic stiffness measured by transient elastography ( $p=0.003$ ), the liver enzymes aspartat aminotransferase ( $p=0.034$ ) and  $\gamma$ -glutamyl transferase ( $p=0.011$ ) and the presence of hepatomegaly ( $p = 0.001$ ). Fibrotest correlated with transient elastography ( $p=0.019$ ), but not with the duration of the Fontan circulation ( $p=0.672$ ). Fontan pressure was significantly higher in patients with a failing Fontan circulation ( $p=0.001$ ) and correlated with liver stiffness measured by transient elastography ( $p=0.024$ ).

**Conclusions:** In 75 % of Fontan patients FALD was present. Liver damage detected by ultrasound, laboratory analysis and transient elastography strongly correlated with the duration of the Fontan circulation. A detailed hepatic assessment is indispensable for a long-term follow-up program for Fontan palliated patients.