Fontan-associated liver disease in patients after Fontan palliation


Department of Congenital Heart Disease-Pediatric Cardiology (1)
Department of Pediatric Radiology, Charité- Universitätsmedizin Berlin (2)
Department of Hepatology and Gastroenterology, Charité- Universitätsmedizin Berlin (3)

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 γ-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.