

Assessment of transient elastography (FibroScan®) and liver function in patients with Fontan circulation

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Objective:

Hemodynamic characteristics of the Fontan circulation are increased systemic venous pressure and reduced cardiac output. This altered hemodynamic status may result in chronic liver dysfunction thus leading to liver cirrhosis with the risk of hepatocellular carcinoma. Early detection of liver fibrosis is mandatory for identifying patients at risk. Transient elastography is a noninvasive, rapid method to assess liver fibrosis by measuring liver stiffness. However, there are limited data concerning the feasibility of this method in patients with a Fontan circulation.

Methods:

A total of 24 patients after Fontan procedure with a median age of 15 yrs were studied by transient elastography using a FibroScan® (Echosens, Paris) from August 2008 to January 2011. Values were correlated with clinical (invasive hemodynamic data, echocardiography, VO₂max, fenestration in conduit) and biochemical parameters (liver enzymes, BNP, coagulation tests).

Results:

Measurements of liver stiffness could be performed in 23 of 24 patients who underwent a Fontan procedure. Liver stiffness increased directly after Fontan completion and was significantly higher compared with age-matched healthy controls (16.9 ± 5.4 kPa vs 4.8 ± 1.2 kPa, $n=17$) or compared with patients with a bidirectional cavopulmonary anastomosis (BDCPA) (5.4 ± 1.7 , $n=8$, $p < 0.0001$). No significant difference was seen between patients with a BDCPA and healthy controls. Liver stiffness did not depend on the time interval since Fontan completion. There was no significant difference in liver stiffness in patients with ($n=10$) or without ($n=13$) a fenestration in the conduit. Hemodynamic parameters obtained by cardiac catheterization (mean pressure in IVC and PA as well as EDP in the ventricle) did not correlate with the data from transient elastography ($n=12$). However, liver stiffness correlated positively with BNP and GPT and negatively with ATIII ($p < 0.05$).

Conclusion:

Liver stiffness as assessed by transient elastography increases rapidly after completion of the Fontan procedure, while patients after BDCPA do not show any difference from normal controls. Biochemical parameters of impending liver dysfunction and cardiac failure in Fontan patients correspond to a further increase in liver stiffness. Therefore, transient elastography may be a sensitive diagnostic tool for early detection of patients at high risk for deteriorating liver and hemodynamic function.