Fontan associated liver disease - prevalence of ultrasound and laboratory abnormalities in different age groups

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Introduction: Liver fibrosis is increasingly recognized as a potentially serious morbidity associated with Fontan circulation (Fontan associated liver disease, FALD). The purpose of this study was to assess the prevalence and progression of liver abnormalities with standard investigations. Methods: Fontan patients were screened for liver abnormalities by abdominal ultrasound and routine laboratory tests. Patients were divided into three groups based on follow-up since Fontan surgery (<5 years, 5 to 10 years, >10 years). Laboratory test were interpreted based on age and gender adjusted reference values. Results: 197 Fontan patients seen for routine follow-up between January 2013 and November 2018 who had abdominal ultrasound together with routine laboratory tests were included. Of them, 139 (70.6%) were re-investigated after a median follow-up of 2.4 (IQR 1.4-3.3) years. Median age at latest follow-up was 12.1 (IQR 8.7-16.7) years, the follow-up since Fontan surgery 9.4 (IQR 6.1-14) years. Sonographic signs of fibrotic changes included heterogeneous parenchyma, surface nodularity or hyper-echoic lesions in 103 (52%) patients. The prevalence of fibrotic changes increased with longer follow-up since Fontan completion (<5 years: 9/30 (23%); 5-10 years: 31/65 (48%); >10 years: 63/93 (68%); p<0.001). New or progressive fibrosis was seen in 24 of 63 (38%) patients with repeat ultrasound after 2.8 (IQR 1.5-3.2) years. Thrombocytopenia, a feature of portal hypertension, was observed in 3.2% and 2.9% of cases with less than 5 or 5 to 10 years of follow-up, but in 27.4% with more than 10 years of follow-up (p<0.001). Elevated Gamma-glutamyl-transpeptidase (gGT) levels were common in all groups, but less frequent with more than 10 years of follow-up (<5 years: 90%; 5-10 years: 97%; >10 years: 72%; p<0.001). Mean platelet count was lower (222 ±84/nl vs. 242 ±91/nl, p=0.041) and gGT levels were higher in patients with sonographic signs of fibrosis (67 ±42 U/l vs. 53 ±37 U/l, p=0.002), while absolute values of other laboratory data showed no differences.

Conclusions: The prevalence of ultrasound and laboratory abnormalities suggestive of FALD increases with time since Fontan surgery. Among laboratory abnormalities the platelet count might serve as a surrogate for progression of liver changes.