

**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 ( $222 \pm 84/\text{nl}$  vs.  $242 \pm 91/\text{nl}$ ,  $p=0.041$ ) and gGT levels were higher in patients with sonographic signs of fibrosis ( $67 \pm 42 \text{ U/l}$  vs.  $53 \pm 37 \text{ U/l}$ ,  $p=0.002$ ), 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.