

## Non-invasive assessment of liver abnormalities in pediatric Fontan patients

Vandekerckhove K.(1), De Bruyne R.(2), Hendrickx F.(3), Van Overschelde H.(3), Vande Walle C.(4), De Groot K.(1), Panzer J.(1), De Wolf D.(1), Van Biervliet S. (2), Bové T.(5), François K. (5)  
(1) Department of Pediatric Cardiology, Ghent University Hospital, Ghent, Belgium; (2) Department of Pediatric Gastroenterology, Hepatology and Nutrition, Ghent University Hospital, Ghent, Belgium; (3) Faculty of Medicine and Health Sciences, Ghent, Belgium; (4) Department of Pediatric Radiology, Ghent University Hospital, Ghent, Belgium; (5) Department of Cardiac Surgery, Ghent University Hospital, Ghent, Belgium.

### Introduction

Pediatric data about liver abnormalities after Fontan palliation are scarce. We assessed the prevalence and degree of liver problems through non-invasive investigations suitable for longitudinal follow-up.

### Methods

35 Fontan patients (median age 11,8yrs; range 5,2-16,6yrs; 27 boys; median time since Fontan 3,29yrs) were evaluated. A liver ultrasound (US) was performed evaluating nodularity, coarsened echotexture, ascites, liver and spleen size. The diameters of inferior vena cava (IVC), portal vein (PV) in in- and expiration and the IVC collapsibility index (IVCCI) were measured. The pulsatility ratio (PR) of the PV and hepatic vein (HV) and the damping index (DI) were calculated. The resistance index (RI) of the PV, hepatic artery (HA) and superior mesenteric artery (SMA) was examined. Fibroscan (Echosens) was used to perform transient elastography (TE). Blood values of AST, ALT,  $\gamma$ GT, Alk Phos, bilirubin, total protein, albumin, alpha-foetoprotein, platelet count, cholesterol and Apo-lipoprotein A1 were measured.

### Results

Nodularity was found in 2/35 patients and irregularity of the liver surface in 2/35 other patients; hepatomegaly was present in 32% of patients, splenomegaly in 15%. PV mean flow velocity was < 15 cm/sec in 19 (54%) patients, correlating with portal hypertension. 22 patients (63%) showed IVCCI values below 17%, indicative of venous congestion (2). HA RI and SMA RI were inversely correlated with time post Fontan ( $p < 0,05$ ;  $r^2 = -0,369$  and  $r^2 = -0,365$  resp.). Liver stiffness was significantly increased compared to controls, with a median(range) of 12,6 kPa (6,6-25,7) versus 4,6 kPa (2-9,5) ( $p < 0,001$ ) from early after Fontan. AST, ALT,  $\gamma$ GT and direct bilirubin were abnormally increased in respectively 12 (34%), 5 (14%), 24 (69%) and 7 (20%), platelet count was decreased in 7 (20%).

### Conclusion

Non-invasive investigations were not able to confirm or differentiate fibrosis from hepatic congestion. We propose follow-up with serial measurements of lab values (ALT,  $\gamma$ GT, direct bilirubin, alpha-foetoprotein, platelet count and clotting), US and Doppler parameters (morphology, IVCCI, PV flow velocities, HA RI, SMA RI and PV pulsatility index) and TE. The use of reliable and accurate non-invasive techniques to assess liver fibrosis in children after Fontan remains a major topic for future research.