Liver stiffness: a useful tool in the longitudinal follow-up of patients with Fontan circulation

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Introduction: Congestive hepatopathy usually appears and develops after a Fontan operation (FO), often without obvious clinical features, but it may lead to life-threatening complications.

Objective: We aim to assess the potential usefulness of the liver stiffness (LS), assessed by elastography, in the longitudinal follow up of Fontan patients.

Methods: In our center, patients with a Fontan circulation were prospectively evaluated since 2012 through an annual work up including physical examination, laboratory tests, trans-thoracic echocardiography and LS using transient elastography (TE). This work up was also performed in case of clinical complications, which were classified as follows: cardiac complications (including arrhythmias or catheter interventions) and subdiaphragmatic complications (including clinical sign of portal hypertension or protein losing enteropathy (PLE)).

Results: Forty eight patients (21.7± 8.2 years of age and 9.7±6.5 years post-Fontan) were included, 28 of them (58%) had least two LS measurement. Mean time between first and last LS measurements was 3.27 ±1.9 years. Mean LS at baseline was 15.3±6.9kPa (4.3-47.2 kPa) No significant correlation was found between LS and age (r=0.8, p=0.73), time since Fontan surgery (r=0.3, p=0.64). LS did not vary regarding the presence of a fenestration (15±6.8 vs 15.1±6.7 kPa, p=0.82).

During the follow-up, a clinical complication occurred in 19 patients (39.6%) including 8 cardiac complications and 11 subdiaphragmatic. Among the cardiac complications group, 6 had atrial flutter and 2 had catheter-based intervention (occlusion of vein-venous collateral). Among the subdiaphragmatic complications group, 3 had PLE and were referred for heart transplant and 8 developed portal hypertension. LS was significantly higher in patient with liver complication (17.2±7.7 vs 13.8±5.9, p=0.01). Patient with an increasing LS value during the follow up had a higher complications rate than patients with a decreasing or unchanged LS (4/11 (36%) vs n=5/17 (29%); p=0.04).

Conclusion: LS measurement using TE is a good tool for the non-invasive follow-up of patient palliated with FO. Indeed, a significate elevation of the LS is associated with the occurrence of liver and/or cardiac complications.