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

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Introduction: Liver stiffness (LS) assessed by Transient elastography (TE) is known to be elevated in patients with a Fontan circulation and is considered as a promising tool to evaluate hepatopathy in these patients. However the longitudinal evolution of this parameter has never been studied.

Methods: We conducted a prospective ongoing study since 2007 including all patients with a Fontan circulation who underwent an annual TE assessment in addition to their systematic work-up and/or in case of a clinical complication.

Results: In this ongoing study, 20 patients were screened with TE and 17 had at least two LS evaluations. Median age at the first TE was 21 years [range 4–32], median duration between the Fontan surgery and the first TE was 4 years [range 1–12], and 6.6 years [range 3–15] at the last TE screening. The median LS at the first evaluation was 12.8 kPA [range 6-47] and 12.4 kPA [range 6-27] at the last evaluation. No significant modification of LS values was observed in our cohort, except for 3 who developed a failing Fontan and are currently waiting for cardiac transplant.

Conclusions: Liver stiffness assessed by TE is stable over time in Fontan patients but significantly increases in patients with a failing Fontan. This tool might be useful in the longitudinal follow-up of these patients for anticipating the decrease of cardiac status and to evaluate non-invasively the effects of targeted therapies before heart transplant.