

MR elastography of the liver in patients status post Fontan procedure: A pilot investigation

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Introduction: The Fontan procedure is used to improve outcomes in patients with single ventricle physiology. With many Fontan patients now reaching adulthood, one of the factors that is increasingly affecting long-term prognosis is the presence of hepatic congestion and fibrosis, a known complication of Fontan physiology. When liver fibrosis progresses to cirrhosis, the risk of neoplasm and portal hypertension rises, and hence these patients may become candidates for liver transplantation. However, if detected early, the progression of hepatic fibrosis can be slowed or halted, and in some situations, reversed. Currently, liver biopsy is considered the gold-standard for assessing liver fibrosis. MR elastography (MRE) is a relatively new, non-invasive imaging technique that quantitatively measures liver stiffness and thus provides a measure of fibrosis. The purpose of this study was to evaluate the feasibility of performing MRE as a screening tool during surveillance cardiac MRI for elevated liver stiffness in patients status-post Fontan procedure.

Methods: A retrospective study was performed evaluating hepatic MREs in patients with a history of Fontan procedure. The MRE of the liver was performed in the same session as the cardiac MRI. Liver MRE would take only 1-2 minutes of additional scan time. Liver stiffness values was calculated by drawing regions of interest on the stiffness maps. The mean liver stiffness (measured in kilopascals) and its correlation with the length of time (years) since Fontan surgery was studied. Mean scan duration was also calculated.

Results: Four patients were included in this study ranging in age from 8 to 26 years. A representative MRE example is shown in Figure 1. All four patients had elevated liver stiffness values by MRE (range 3.4 to 4.2 kPa) as seen in Figure 2 (normal adult liver stiffness is 2.2 kPa), suggesting the presence of mild fibrosis (lower values) to cirrhosis (higher values). A trend of increasing liver stiffness value relative to number of years' post-Fontan completion was detected (see Figure 2).

Conclusions: Liver MRE is feasible in patients status-post Fontan procedure who are undergoing surveillance cardiac MRI, and in our small cohort, demonstrated elevated liver stiffness in all patients, with a trend towards stiffer livers with increased time from Fontan completion. Further investigation with histologic correlation is needed to determine the etiology and long-term sequela of elevated liver stiffness in this population. Our preliminary study shows that duration of hepatic congestion following Fontan procedure is related to liver stiffness and hepatic fibrosis.

References: [1] Marcelletti C, Mazzera E, et al. Fontan's operation: an expanded horizon. J Th. Cardiovasc Surg. 1980. [2] Yin M, Talwalkar JA, et al. Assessment of hepatic fibrosis with magnetic resonance elastography. Clin Gastro Hepatol 2007. [3] Serai S, Towbin AJ, Podberesky DJ. Pediatric MR Elastography. Dig. Dis. Sci. 2012.

