# [Relation of Magnetic Resonance Elastography to Fontan Failure and Portal Hypertension.](https://www.ncbi.nlm.nih.gov/pubmed/31474329)

Alsaied T, Possner M, Lubert AM, Trout AT, Szugye C, Palermo JJ, Lorts A, Goldstein BH, Veldtman GR, Anwar N, Dillman JR.

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**Take Home Points:**

* Magnetic resonance elastography (MRE) is a non-invasive method of assessing liver stiffness and liver fibrosis.
* In this study of 70 adult Fontan patients, all had increased MRE liver stiffness (greater than 2.9 kPa is the cutoff used as this institution).
* Liver stiffness of 4.5 kPa or greater is associated with Fontan failure (sensitivity of 77% and specificity of 77%) and radiologic signs of portal hypertension (sensitivity of 69% and specificity of 65%).
* MRE-derived liver stiffness is associated with adverse hemodynamics, liver function, and clinical outcomes in Fontan circulation.



***Commentary from Dr. Maan Jokhadar (Atlanta GA), section editor of ACHD Journal Watch:***

Fontan associated liver disease (FALD) is associated with Fontan failure (FF) and appears to be multifactorial with contributing factors that include elevated central venous pressure and chronic liver congestion, low cardiac output, abnormal liver lymphatic drainage, and other factors. Assessing FALD and FF with liver biopsy is of limited reliability due to the non-uniform nature of fibrosis in this population. Noninvasive assessment of liver stiffness including magnetic resonance elastography (MRE) has shown mixed results.

Dr. Alsaied and colleagues from the University of Cincinnati performed a retrospective cohort analysis of 70 adult Fontan patients who were evaluated between 2012 and 2018 to determine the role of MRE as a measure of liver stiffness and its relationship to hemodynamics, liver enzymes, portal hypertension, and FF. In this study, FF was defined as death, heart transplant listing, or heart failure symptoms requiring escalation and diuretic therapy. The upper limit for normal liver stiffness at this institution is 2.9 kPa.

The median age was almost 25 years and 52% were men. The median follow-up from the time of MRE was 3.9 years. There is no significant correlation between MRE liver stiffness and age or time since Fontan. There was one case of hepatocellular carcinoma detected with MRE screening.

There was a moderate negative correlation of liver stiffness with ventricular ejection fraction and ventricular end diastolic volume. Patients with moderate to severe atrioventricular valve regurgitation had significantly higher liver stiffness compared to patients with no or mild regurgitation. Also, patients with a history of thromboembolism had higher liver stiffness compared to patients without prior thromboembolism. There was a very mild positive correlation between liver enzyme elevations and liver stiffness. There was no difference in laboratory values in patients with or without FF.

Patients with features of portal hypertension including splenomegaly, varices, or ascites had higher liver stiffness compared to patients without these features. Liver stiffness had a weak negative correlation with platelet count. Using ROC analysis liver stiffness had an area under the curve of 0.69 to differentiate portal hypertension and it should be noted that liver stiffness above 4.5 K PA differentiated portal hypertension with the sensitivity of 69% and specificity of 65%.

FF was present in 19 patients (4 deaths, 1 heart transplant, and 8 with worsening heart failure, 1 of whom received LVAD. FF patients had higher liver stiffness. A liver stiffness above 4.5 kPa differentiated FF with sensitivity of 77% and specificity of 77%.

All adult Fontan patients in this study had increased liver stiffness by MRE and this correlated with increased Fontan pressure, lower ventricular ejection fraction, atrioventricular valve regurgitation severity, FF, elevated liver enzymes, and radiologic signs of portal hypertension.

Liver stiffness in FALD is a function of both congestion and fibrosis and this may explain previous studies showing discrepant data regarding the correlation of MRE and biopsy measures of fibrosis. In contrast with previous studies, this study of adult Fontan patient showed that MRE liver stiffness was not associated with time since Fontan.

The study authors suggest that MRE could be used routinely in Fontan patients as part of routine assessment cardiac and liver “health”, which could be used as a global assessment of the Fontan circulation that correlates with the development of clinical symptoms and outcomes. Abnormal MRE liver stiffness could then trigger more detailed evaluation.

Additional prospective, multi centered studies with longer serial follow-up and changes with medication and/or intervention are needed before recommending routine MRE surveillance for Fontan patients. using this technique. Cost effectiveness should also be considered.