# [Increased extracellular volume in the liver of pediatric Fontan patients.](https://www.ncbi.nlm.nih.gov/pubmed/31303178)

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**Take Home Points:**

* In Fontan survivors, elevated systemic venous congestion, reduced cardiac output and a multitude of other hepatic insults including often multiple prior cardiac surgeries results in Fontan associated liver disease (FALD) characterized by hepatic congestion, and fibrosis.
* In Fontan patients, varying degrees of FALD is universal and progressive. Cirrhosis and hepatocellular carcinoma are known morbidities of advanced FALD.
* The natural history of FALD and factors influencing its progress are unclear. Traditional serological markers of hepatic function fail to provide adequate insight into severity and progression of FALD until late in the disease process.
* Cardiac magnetic resonance imaging T1 mapping of liver and young Fontan patient is feasible.
* Fontan survivors showed elevated native T1 and ECV values compared to both bidirectional cavopulmonary anastomosis subjects and controls suggesting diffuse hepatic congestion and/or fibrosis.
* Subjects with Fontan as well as bidirectional cavopulmonary anastomosis showed elevated native T1 compared to control indicating hepatic congestion even in subjects with bidirectional cavopulmonary anastomosis.
* In Fontan subjects, T1 values correlated with exposure to cardiopulmonary bypass time but not to central venous pressure or age.
* Native T1 and ECV values may be useful imaging biomarkers to evaluate hepatic congestion and fibrosis in FALD.
* ECV does not provide clinically significant additional information compared to native T1 analysis. Relying on native T1 analysis obviates the need for gadolinium and reduces the postprocessing and scan time.



***Commentary from Dr. Shaji Menon (Salt Lake City), section editor of Pediatric Cardiology Journal Watch:*** This is a retrospective cross-sectional cohort study of pediatric patients with bidirectional cavopulmonary anastomosis and Fontan procedures. Hepatic native T1 times and ECV was measured using cardiac short axis modified Look-Locker inversion recovery sequence retrospectively from images displaying the liver. A multivariate regression model was performed for evaluating factors associated with native T1 times and ECV. Hepatic native T1 time were increased in Fontan patients (n = 62, 11.4 ± 4.4 years, T1 762 ± 64ms) versus BCPC patients (n = 20, 2.8 ± 0.9 years, T1 645 ± 43ms, p = 0.04). Both cohorts had higher T1 than controls (n = 44, 13.7 ± 2.9 years, T1 604 ± 54ms, p < 0.001 for both). ECV was 41.4 ± 4.8% in Fontan and 36.4 ± 4.8% in BCPC patients, respectively (p = 0.02). In Fontan patients, T1 values correlated with exposure to cardiopulmonary bypass time (R = 0.3, p = 0.02), systolic and end diastolic volumes (R = 0.3, p = 0.04 for both) and inversely with oxygen saturations and body surface area (R = -0.3, p = 0.04 for both). There were no demonstrable associations of T1 or ECV with central venous pressure or age after Fontan.

**Figure 1**



**Figure 2**

