

## Abstract

**Background:** Diffuse fibrosis, as measured by T1 mapping and extracellular volume, may be associated with ventricular dysfunction in the Fontan circulation.

**Methods:** A retrospective cohort study at a moderate-altitude academic center of pediatric and young adult Fontan patients vs healthy controls exploring demographic differences, markers of fibrosis on cardiovascular magnetic resonance imaging, and associated clinical correlates.

**Results:** The Fontan cohort of 93 patients was stratified by field strength (n=65 at 1.5T, n=28 at 3T) and again by dominant left (n=47), dominant right (n=45) or combined (n=1) ventricles. Comparison was made to a control cohort of 49 patients (n=26 at 1.5T, n=23 at 3T). The Fontan cohort demonstrated increased native T1 values across varying field strengths of 1.5T (981.4 vs 1031.6 ms  $p<.001$ ) and 3T (1253.4 vs 1287.5 ms  $p=.031$ ). Extracellular volume was also increased (23.2 vs 26.0%  $p<.001$ ). There was no significant difference between dominant right and left ventricles among 1.5T native T1 (1040.9 vs 1023.1 ms  $p=.166$ ), 3T native T1 (1275.5 vs 1299.4 ms  $p=.289$ ) or extracellular volumes (26.3 vs 25.4%  $p=.321$ ). VO2 max was significantly reduced among the Fontan cohort (2.4 vs 1.3 LPM  $p<.001$ ), and negatively correlated with extracellular volume ( $R=-.26$   $p=.018$ ). Negative correlation with extracellular volume was also observed with age ( $R=-.35$ ,  $p<.001$ ), hematocrit ( $R=-.51$   $p<.001$ ), and diastolic blood pressure ( $R=-.38$   $p<.001$ ). Positive correlation was observed for Fick cardiac index ( $R=.36$   $p=.007$ ).

**Conclusions:** Markers of myocardial fibrosis were elevated among the Fontan cohort compared to healthy controls. There was a trend towards increased fibrosis among dominant right ventricles compared to dominant left. Fontan patients were shown to have a reduced exercise tolerance as measured by VO2 max. A negative correlation between ECV and VO2 max was observed across the Fontan group, suggesting a possible relationship between myocardial fibrosis and impaired exercise tolerance.