

Diffuse myocardial fibrosis is associated with type of adverse loading and location of the right ventricle in congenital heart disease – a cardiovascular magnetic resonance T1 mapping study

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Introduction:

While focal myocardial fibrosis is commonly detected by late gadolinium enhancement cardiovascular magnetic resonance (CMR) and has been related to adverse outcome in various congenital heart disease (CHD), the extent of diffuse myocardial fibrosis and its role in CHD are relatively unknown. This study sought to assess myocardial extracellular volume (ECV) reflecting diffuse myocardial fibrosis, and to investigate associations with clinical and functional parameters, type of adverse loading and location of the right ventricle (RV) in CHD.

Methods:

CHD patients (n=53, median age 26.1 years) with pressure and/or volume overload of the RV were prospectively enrolled and compared to healthy controls (n=19, 24.8 years). Participants received standardized CMR, laboratory and cardiopulmonary exercise testing. Modified Look-Locker Inversion recovery (MOLLI) T1 mapping was performed in midventricular short axis and axial orientation to quantify RV and left ventricular (LV) ECV.

Results:

RV and LV ECV were significantly higher in patients (median 31 and 28%) than controls (29 and 25%; $p=0.021$, respectively), with values above the upper limit of normal in 32 and 25% of the patients. Within the patient group, LV ECV correlated with N-terminal pro brain natriuretic peptide ($r=0.61$, $p<0.001$) and indexed LV stroke volume ($r=-0.37$, $p=0.017$). Increased RV ECV was related to volume overload ($p=0.024$), while greater LV ECV was associated with pressure overload of the RV in systemic vs. subpulmonary location ($p<0.001$).

Conclusions:

CHD patients had higher RV ECV as well as LV ECV in correlation with markers of heart failure, indicating an adverse ventricular interaction. Elevated RV and LV ECV were associated with the type of adverse loading and the location of the RV. Further studies are necessary to explore the clinical implications of diffuse myocardial fibrosis, and to assess the utility of non-invasive ECV measurements in supporting risk stratification and guiding treatment in CHD.