

**Evaluation of diffuse myocardial fibrosis by T1 mapping in children and adolescents with primary cardiomyopathies – results from the RIKADA study**

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**Introduction:**

Diffuse myocardial fibrosis is a common finding and associated with adverse outcome in adult cardiomyopathy (CM), yet data on its role in pediatric patients are sparse. Cardiovascular magnetic resonance (CMR) T1 mapping is a non-invasive tool to quantify diffuse myocardial fibrosis as reflected by extracellular volume (ECV). This study aimed to assess markers of diffuse myocardial fibrosis and their correlation with ventricular function in children and adolescents with primary CM.

**Methods:**

In total, 13 prospectively enrolled patients (mean age  $11.6 \pm 5.0$  years) with primary CM (dilated CM,  $n=4$ ; hypertrophic CM,  $n=4$ ; left ventricular non-compaction CM,  $n=5$ ) were compared to 16 healthy controls (mean age  $25.1 \pm 2.7$  years). T1 maps were generated with Modified Look-Locker Inversion recovery (MOLLI) T1 mapping in a midventricular short axis plane before and 15 minutes after bolus application of Gd-DOTA. ECV was calculated from T1 values of the left ventricular (LV) circumference, T1 from LV blood pool, and hematocrit. In the patient group, T1 and ECV were compared to volumetric CMR data.

**Results:**

Median (interquartile range) native T1 in patients and controls was 1028 (44) ms and 995 (40) ms, respectively ( $p=0.009$ ). No significant difference in ECV between patient and control group was found (27 [7] % and 26 [5] %, respectively;  $p=0.5$ ). In patients, native T1 correlated significantly with LV-end-diastolic volume (EDV) indexed to body surface area ( $r=0.8$ ,  $p=0.003$ ). Correlations between ECV and parameters of ventricular function were not observed.

**Conclusions:**

Our results demonstrate a tendency towards increased diffuse myocardial fibrosis associated with LV-EDV in young patients with CM. However, the overall non-significant elevation of ECV may indicate an only minor role in the course of pediatric CM. Further longitudinal studies are necessary to investigate the diagnostic and prognostic value of non-invasive ECV measurements in children and adolescents with primary CM.