

**Midwall fibrosis in children and adolescents with primary inherited dilated cardiomyopathy**

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**Objectives:**

Midwall fibrosis as detected by late gadolinium enhancement (LGE) cardiovascular magnetic resonance (CMR) is a common finding predicting adverse outcome in adults with idiopathic dilated cardiomyopathy (DCM). So far, its relevance in pediatric patient is relatively unknown. The present study aimed to evaluate focal myocardial fibrosis by LGE in children and adolescents with primary inherited DCM.

**Methods:**

Patients  $\leq$  18 years with primary inherited DCM were prospectively enrolled into the study. Standardized CMR including volumetric analysis and LGE for assessment of focal myocardial fibrosis was conducted in all participants. LGE images were acquired 8 to 10 minutes after bolus administration of gadolinium-DOTA at a dose of 0.2 mmol/kg, covering the entire left ventricle (LV) in short axis orientation, and in 2-, 3- and 4 chamber view.

**Results:**

In total, 13 patients (median age (range) 11.6 (7.5-15.0) years, 7 female) were included. Median LV EF was 33 (14-53) % and LV EDV indexed to body surface area was 124.1 (107.1-225.6) ml/m<sup>2</sup>. LGE was present in 8 of the 13 patients (62%). In all 8, LGE was characterized as midwall fibrosis located in the basal and/or midventricular septum. Of the 8 patients with midwall fibrosis, 4 underwent cardiac transplantation (HTx). Prior mechanical circulatory support (MCS) was performed in 3, and additional implantation of an implantable cardioverter defibrillator for secondary prevention of sudden cardiac death in 1 of the 4 HTx patients. Of the 5 LGE negative patients, 1 received MCS and HTx.

**Conclusions:**

Our data suggest that midwall fibrosis appears frequently in children and adolescents with primary inherited DCM and may be associated with unfavorable clinical outcome in these patients. Further longitudinal studies are necessary to assess the incidence and prognostic role of midwall fibrosis in pediatric DCM.