Increased diffuse myocardial fibrosis on cardiovascular magnetic resonance T1-mapping and intraventricular conduction delay on electrocardiogram on long-term follow-up after surgical septal myectomy in children with obstructive hypertrophic cardiomyopathy

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Introduction

The long-term effect on myocardial remodeling of surgical septal myectomy performed in children with severe obstructive hypertrophic cardiomyopathy (HCM) and its influence on cardiac conduction system disease or arrhythmias is unclear.

Materials and Methods

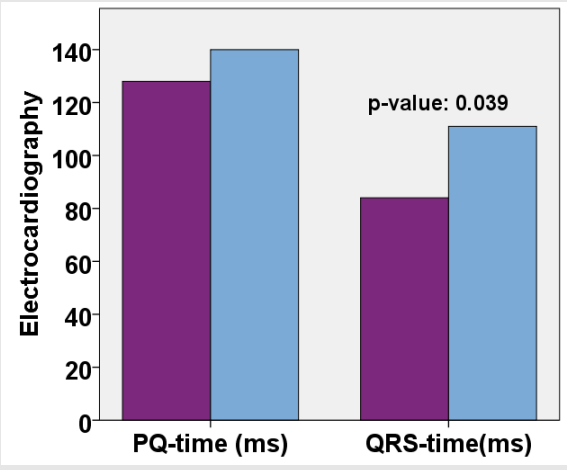
Patients with childhood HCM were evaluated by electrocardiogram (ECG), Holter ECG, cardiopulmonary exercise test, transthoracic echocardiography (TTE), genetic testing, and cardiovascular magnetic resonance (CMR).
Diffuse myocardial and focal fibrosis was assessed by myocardial extracellular volume fraction (ECV) with T1-mapping and late gadolinium enhancement, respectively, on CMR. Patients with were compared to patients without prior myectomy.

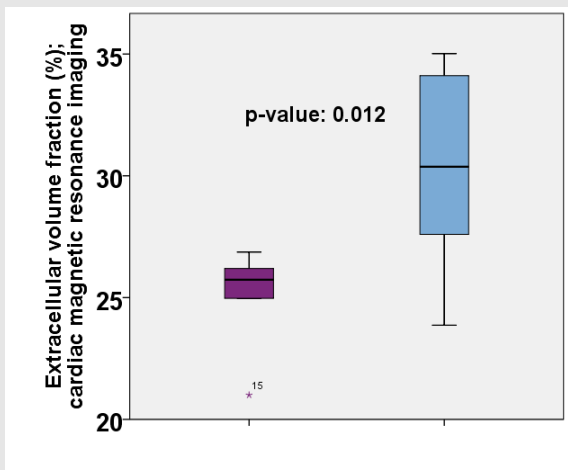
Results

	Septal myectomy (10)	No septal myectomy (5)
Male/Female (N)	8/2	4/1
Noonan Syndrome/ Non-syndromic HCM (N)	6/4	1/4
Age at surgery (years)	2.5 (0.2/16.1)	-
Age at study (years)	16.5±8.7	16.4±4.5

	Septal myectomy (10)	No septal myectomy (5)	P-value
CMR: late gadolinium enhancement (N)	4/10	3/5	NS
CMR: Left ventricular mass (g/m²)	123±90	79±29	NS
TTE: IVSd Z-score	2.9±2.6	3.1±2.2	NS
TTE: LVPWd Z-score	2.7±1.9	2.6±1.7	NS
TTE: Ejection fraction (%)	74±9	87±9	NS
Rhythm disturbances (N)	1/10	0/5	NS

Data expressed in mean±STD or median (min/max) according to sample distribution; N: number of patients; NS: no statistical significance; CMR: cardiac magnetic resonance imaging; TTE: transthoracic echocardiography; IVSd: enddiastolic interventricular septum thickness ; LVPWd: enddiastolic left ventricular posterior wall thickness





Conclusions

Long-term follow-up demonstrates increased diffuse myocardial fibrosis and intraventricular conduction delay after surgical septal myectomy performed during childhood for obstructive HCM. This might be due to intrinsic general myocardial remodeling in HCM after severe left ventricular outflow tract obstruction or secondary to inflammatory triggers during extracorporeal circulatory bypass in patients requiring cardiac surgery during childhood.