CMR in pediatric HOCM: correlation with LV function, echocardiography and demographic parameters

Hypertrophic cardiomyopathy (HCM) is the most common inheritable cardiac disorder, with an estimated prevalence of 1:500. Although the disease is compatible with normal life expectancy, it is associated with premature death from ventricular arrhythmia, heart failure, and stroke in a minority of patients. Techniques which accurately measure left ventricular wall thickness, integrity and perfusion are necessary to evaluate the effectiveness of treatment strategies.

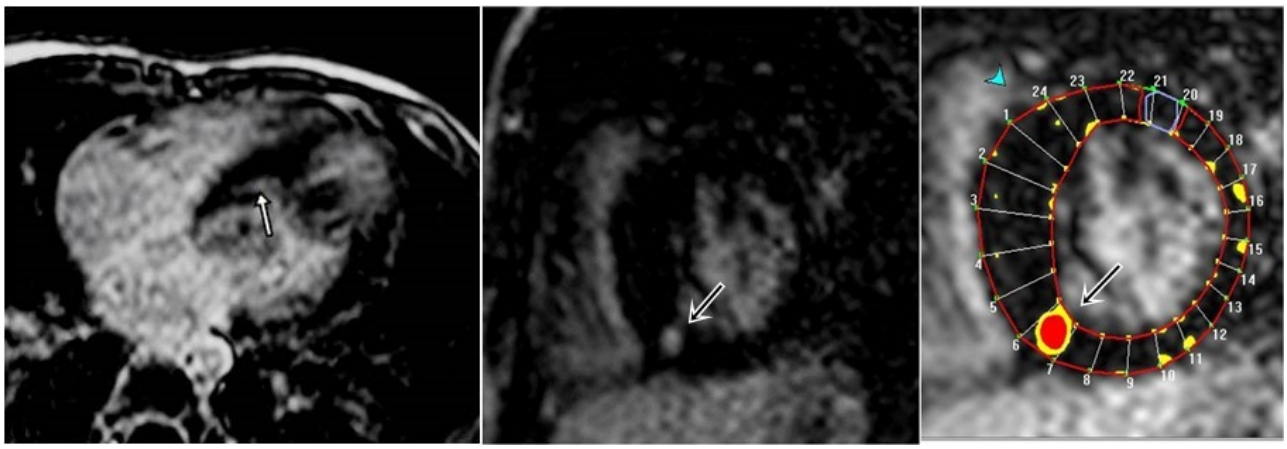

Fig. 1. Quantification of percentage of myocardial fibrosis. A focus of hyperenhancement at the inferior aspect of the septal wall (arrow) in a 7 year old patient.

Our aim was to detect the presence of fibrosis by Cardiac magnetic resonance imaging (CMR) in the pediatric age group and correlate CMR findings with demographic data, LV function and other echocardiographic parameters.

We studied 13 female and 27 male patients from 45 days up to 18yrs. 82.5% of the patients showed variable degrees of myocardial fibrosis mainly at the areas of most left ventricular thickening which was the interventricular septum in most cases. We found significant correlation between the severity of dyspnea of the patients and the grade of left ventricle obstruction caused by the hypertrophied ventricular wall as well as the percentage of myocardial fibrosis. The percentage myocardial fibrosis didn't correlate with any other demographic or LV functional cardiac parameters. As surprisingly the age of the patients didn't correlate with the thickness of the myocardium or the presence of fibrosis.

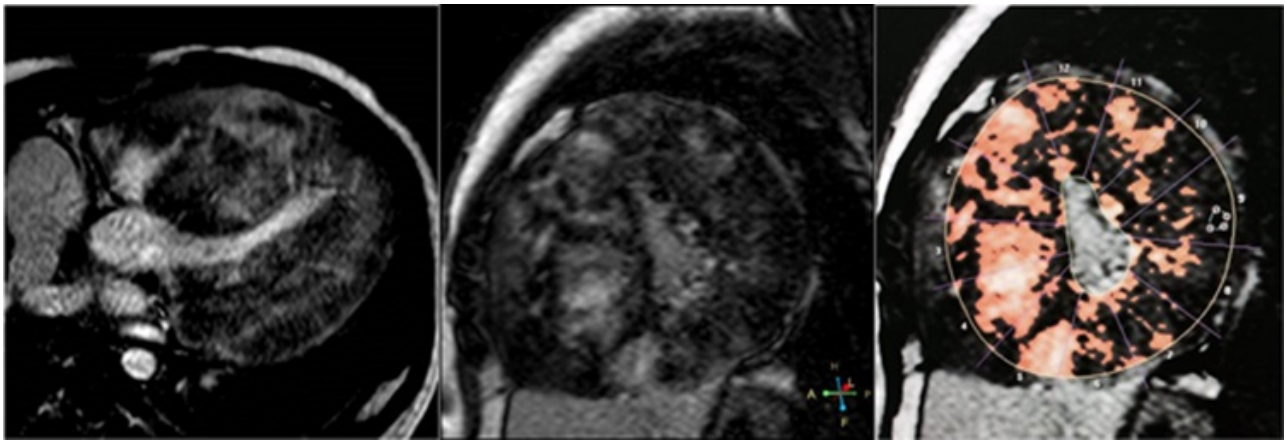

Fig. 2. Extensive patchy midwall areas of delayed enhancement indicating myocardial fibrosis with marked myocardial wall thickness in a 16year old patient.

Myocardial fibrosis is believed to be the main pathological hallmark for adverse outcomes hence, the presence of myocardial fibrosis is a risk factor for cardiac arrhythmia and sudden death. Our study results may help in better management of children with hypertrophic cardiomyopathy, planning of follow up and improving the prognosis of the disease. In addition, the ongoing study of those with myocardial fibrosis may lead to future changes in the guidelines especially regarding risk stratification.

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Publication

Delayed Myocardial Enhancement in Pediatric Hypertrophic Cardiomyopathy: Correlation with LV Function, Echocardiography, and Demographic Parameters.
El Saiedi S, Behairy NH, Kharabish A, Esmail R, Seliem ZS, Shafik M, El Mozy W
Pediatr Cardiol. 2017 Jun