Myocardial fibrosis detected by cardiac magnetic resonance imaging in children with hypertrophic cardiomyopathy

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Background: Contrast-enhanced cardiovascular magnetic resonance with late gadolinium enhancement (LGE) has emerged as an in vivo marker of myocardial fibrosis, although its role in stratifying sudden death risk in pediatric hypertrophic cardiomyopathy (HCM) remains incompletely understood.

Here, we sought to determine the prevalence of LGE in children with HCM with different causes and its relation with left ventricular mass and geometry.

Methods
CMR protocol included T2 weighted sequence in short axis view, TRIPLE IR FSE sequence, cine SSFP in short axis, two-chamber, three and four chamber view without contrast and perfusion analysis and late enhancement after injection of contrast agent. If left ventricular wall thickness was asymmetric, the size and location of relatively thickened segments were noted.

Results
A total of 74 children were included. Age at diagnosis was 3 years (range 1 day to 16 years). Mean age at CMR was 11 years (range 1-18 years). CMR was successfully performed in all patients, revealing a better performance in comparison to echocardiography to define precisely the anatomy of LV hypertrophy. Mean LV mass was estimated at 94 ± 41 gr/m². LV hypertrophy was concentric in 35 patients, asymmetric in 39 patients, with evidence of LV non-compaction aspect in 10 patients. Right ventricular hypertrophy was observed in 7 cases. LGE was detected in 6 patients within the septum and in 1 patient in apical wall. Perfusion defects were present in 5 patients in papillary muscles. LV ejection fraction was < 55% in 7 patients. We found no correlation between LV mass, LV outflow tract obstruction or geometry of the hypertrophy and the presence of LGE. Children with LGE did not have more cardiac events than those without.

Conclusion
In our series, the prevalence and extent of LGE in children with HCM was rare compared to adults with HCM. Furthermore, LGE was not associated to adverse events, such as sudden death that also occurred in patients without LGE.