

Cardia Magnetic Resonance in young patients with Hypertrophic Cardiomyopathy.

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Background: although the role of cardiac magnetic resonance (CMR) in adult with hypertrophic cardiomyopathy (HCM) has been widely validated, limited data existed on the role of CMR young patients with HCM. We sought to assess hypertrophy extension of HCM as well as late gadolinium enhancement (LGE) in a young population by CMR and evaluate their impact on event or procedure at follow-up.

Methods: we retrospectively evaluated 75 HCM or HCM-like (associated to a genetic syndrome) young patients (aged 10.4 ± 5.2 years, 49 males) who underwent CMR in 2 tertiary pediatric cardiac centers. Left ventricle volume, mass and function, wall thickness and LGE were assessed. According to the extension of the hypertrophy we divided our population in 3 groups: Focal: less than 3 ventricular hypertrophic segments; Intermediate: from 3 to 8 hypertrophic segments and Diffuse: more than 8 hypertrophic segments. Clinical history was abstracted from medical record. During a median follow-up of 2 years, one patient died, 2 underwent surgical procedure associated with ICD implant in one; ICD was implanted in 2 others patients, one of them was subsequently transplanted.

Results: thirty-two patients (43%) have a family history of HCM. In 22 Patients HCM was associated to a genetic syndrome. A gene mutation was present in 14 patients, in 4 patients any mutation has been found and genetic testing was unknown in the remaining patients. 55 % of the whole population presented intermediate hypertrophy extension, 27 % a diffuse one. The maximum wall thickness was 14.7 ± 6 mm. LGE was present in 22%, mainly in the interventricular septum. Patients with LGE had a greater wall thickness and left ventricular mass index than those without LGE (respectively: 20 ± 6.8 g/m² vs. 13 ± 4.9 g/m², $p=0.001$ and 97 ± 29 g/m² vs. 76 ± 26 g/m², $P=.02$). Moreover, the presence of LGE was associated with subsequent surgical/ICD procedure at follow-up (Chi²:6.1, $P=.01$).

Conclusions: In our population the intermediate pattern of HCM is the most frequent. LGE is less prevalent in young population than in adult HCM patients but it is associated with subsequent surgical/ICD procedure. Further longitudinal studies are needed to evaluate the progressive development of LGE and its prognostic significance.