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Cardiac magnetic resonance imaging and disease progression in young patients with hypertrophic cardiomyopathy

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Introduction: Cardiac MRI including stress perfusion provides additional information for assessing sudden cardiac death risk in hypertrophic cardiomyopathy patients. However, the pathophysiological meaning of those findings in young patients is not fully inspected.

We investigated the relationship between CMR findings and clinical feature to speculate the mechanism of disease progression.

Method: From September 2011 to September 2016, 17 patients, with a mean age of 14.7 years, with hypertrophic cardiomyopathy underwent comprehensive cardiac MRI (1.5-T Philips Achieva). Cine images, stress and rest perfusion images, and late gadolinium enhancement (LGE) images were acquired successively. Adenosine triphosphate was intravenously injected at a dose of 0.14 mg/kg/min during stress perfusion. Both perfusion images were acquired by infusion of 0.1 mmol/kg meglumine gadopentetate at an infusion rate of 3.5 ml/s respectively.

Result: Stress induced perfusion defect (SPD) was detected in 10/15 patients (67%). LGE was recognized in 14/16 patients (88%). Of those with LGE, the extent was 10±13% of the myocardial mass. Left ventricular mass index was 162±61% of normal value and ejection fraction was 66±10%. In five patients with a history of syncope or ventricular fibrillation, LV mass index and plasma brain natriuretic peptide (BNP) level were significantly higher than in those with no history (206 vs 144 % of Normal, $p = 0.048$, 635 vs 130 pg/ml, $p=0.002$, respectively). LV mass index was also higher in those with SPD than in those without SPD (184 vs 129 % of Normal, $p = 0.041$), while LGE extent was not significantly different. Out of 3 patients who underwent the second cardiac MRI 2 years after the initial examination, 2 patients with SPD presented the expansion of LGE area along with plasma BNP elevation □ LGE extent; 6 to 13%, 4 to 8% and BNP; 107 to 274, 100 to 280 pg/ml, respectively □.

Conclusion: Microvascular dysfunction, which is proportional to magnitude of hypertrophy, can be one of the key role players in worsening disease severity. Comprehensive and periodical cardiac MRI including stress perfusion can be feasible and clinically valuable in young patients with hypertrophic cardiomyopathy.