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High ECG risk-scores correlate with late Gadolinium enhancement on magnetic resonance imaging in HCM in the young

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INTRODUCTION: Hypertrophic cardiomyopathy (HCM) is the most common inherited cardiac disease. HCM is characterized by gradual thickening of the myocardium and disturbed myocardial composition. ECG risk-score >5p has been shown to detect pediatric patients with subsequent cardiac arrest with high sensitivity. Myocardial fibrosis measured by cardiac magnetic resonance (CMR) has also been proposed as a risk-factor for cardiac events. Previous studies have shown disturbed myocardial perfusion surrounding the fibrotic areas. CMR is time consuming and expensive. We sought to assess whether ECG analysed by ECG risk-score could be used as an indicator of fibrosis or perfusion deficit in HCM as measured by CMR.
METHODS: A single center cohort of 50 individuals; 41 patients from 29 family-pedigrees, HCM-patients (n=24), age 7-31 years, 17 individuals at risk of HCM (phenotype-negative mutation-carriers,n=8; first-degree HCM-relative,n=9), and 9 healthy controls, underwent CMR and 12-lead ECG. CMR was performed using late Gadolinium enhancement (LGE) and Adenosine stress-test to identify fibrosis and myocardial perfusion defect. ECG was analyzed according to the ECG risk-score method (Eur Heart J 2010;31:439), and categorical data compared with Fisher´s exact test.
RESULTS: The majority of HCM-patients presented <19 years of age. ECG risk-score >5p was significantly more common in the HCM group, median 3p [IQR 0-9p], vs. controls and individuals at risk of HCM; median 0p [0-1p], (p=0.001); and significantly more common also compared to individuals at risk of HCM (p=0.005). In patients with positive LGE median risk-score was 8p [3-10p]. An ECG risk-score in the high-risk range (6-14p) correlated with positive LGE on CMR (p<0.001), with specificity 97%, sensitivity 57%, positive predictive value (PPV) 89% and negative predictive value (NPV) 85%. Including perfusion defect in analysis did not increase sensitivity, and specificity and PPV remained unchanged.
CONCLUSIONS: Myocardial fibrosis on CMR (LGE) correlate with high ECG risk-scores. ECG risk-score in this study is very specific and quite sensitive. Since ECG is easily performed and available in almost every healthcare facility, ECG risk-score allows you to prioritize your high risk patients and could be an in-expensive complementary tool in risk stratification of HCM in the young.