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Mutation of desmoplakin gene and assessment of right ventricle function in patients with arrhythmogenic right ventricular cardiomyopathy
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Purpose: Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an inheritable disease characterized by life-threatening ventricular arrhythmias. Approximately 50% of symptomatic individuals harbor a mutation in one of the five major components of the cardiac desmosome, among which desmoplakin (DSP) is the common mutation genes. Little is known about desmosomal protein gene mutation in Chinese ARVC patients. Pathologically, ARVC is characterized by a progressive replacement of myocytes of the right ventricle (RV) with fibrous and fatty tissue. Assessment of the RV by two-dimensional (2D) echocardiography is the most important screening method, but the identification of RV abnormalities using echocardiography is still challenging. We here sought to delineate the clinical characteristics and genetics of ARVC patients and characterize whether Doppler tissue imaging and 2D speckle tracking could usefully assess right ventricle function in Chinese ARVC patients.

Methods: Forty subjects fulfilling modified Task Force criteria were included. Genetic analysis was screened for mutations in familial ARVC. Information on medical history was obtained using echocardiography. Morphologic and functional study were performed for LV and RV (diameters, volumes, peak systolic velocity from tissue Doppler and longitudinal strain measurement) based on speckle tracking. The global and segment longitudinal strain were measured from the basal and mid segments in both ventricles. LV and RV longitudinal strain measurement was feasible in first degree relatives and controls on 85 and 78% but less in patients (60%). On first degree relatives, Segmental longitudinal peak systolic strain on RV (and basal lateral segments) was significantly lower than that of controls (−28.9±3.2% vs. −32.3±3.2%; P = 0.002 and −26.6±4.3% vs. −31.2±3.6%; P = 0.03 respectively) but significantly greater than that evaluated in ARVC patients (−20.6±4.7%; P = 0.001 and −19.5±4.2%; P = 0.01). For the left ventricular global strain, no differences were found between the control group and the first degree relatives (22.6±5.5% vs. −21.3±6.8%), but we found a difference between control group and patients (17.3±4.2%; P = 0.01).

Conclusion: Longitudinal strain of LV and RV segments was significantly lower in patients than in relatives and controls. The global and segment longitudinal function assessed by echocardiographic might help to detect early ARVC disease in firs degree relatives.

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Myocardial iron overload in sickle/thalassemia patients of Italian origin
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Purpose: Sickle-thalassemia is an inherited hemoglobin disorder resulting from the combined heterozygosity for sickle-cell and δ-thalassemia genes. Myocardial iron overload in patients with sickle-thalassemia has been poorly studied; however, a report characterized by lethal of cardiac iron in a small group (n=10) of multitransfused Arab patients. The current study aims to further evaluate cardiac iron overload in a larger group of Italian patients using a T2* multislice approach in firs time.

Methods: Fifty-nine sickle-thalassemia patients (29 males, mean age 35.6±14.1 years), enrolled in the MIOT network were considered. Three parallel short-axis views of the left ventricle were acquired and analyzed with a dedicated software (HIIPPO MIOT) providing the T2* value on each of 16 segments as well as the global T2* value averaged over all segmental T2* values and the T2* value in the mid-ventricular segment averaged over the mid-anterior and the mid-inferior septum. Results: We found 55 (93%) patients had all 16 segmental T2* values normal (<20 ms). Of the 4 patients with abnormal segmental T2* values, all showed an homogeneous MIO (some segments were weakly inversely in Doppler tissue imaging and LV mass and A/E ratio. EF weakly correlated with basal rotation, interesting, there was a strong inverse relationship (P=0.001, r = −0.75) between LV mass index and T2* values within the HCM subset.

Comparison of deformation data - HCM and

Longitudinal strain Longitudinal strain – systolic Longitudinal strain – diastolic Rotation – mitral value Rotation – apical value Rotation – ventricular value
HCM −16.5±3.3 −19.0±2.6 −20.3±2.9 0.08±0.02 0.08±0.02 0.11±0.03 0.19±0.03 controls −17.1±3.5 −12.5±2.8 1.13±0.7 −7.6±3.0 8.3±3.6 16.0±5.0

*p<0.05 HCM, hypertrophic cardiomyopathy.

Conclusions: Hypertrophic cardiomyopathy pts present abnormal deformation patterns despite preserved ejection fraction. Depressed longitudinal strain, strain rate and diastolic strain rate are paralleled by increased eft ventricular torsion resulting from enhanced apical rotation. Interestingly, within HCM subset, LV mass is negatively correlated with rotation indices with possible "inverse J-curve" relationship.

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Altered two-dimensional strain measures of the right and left ventricle in patients with arrhythmogenic right ventricular cardiomyopathy and their first degree relatives
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Introduction and aim: The identification of right ventricular abnormalities in patients with arrhythmogenic right ventricular cardiomyopathy (ARVC) in early stages is still difficult. The aim of this study was to investigate if longitudinal strain based on speckle tracking can detect subtle right (RV) or left ventricle (LV) dysfunction as an early sign of ARVC.

Methods and results: We have investigate 26 patients, fulfilling Task force criteria for ARVC, 16 were male, 34 (17-77) years old, 35 first degree relatives (21 were male, 29 years mean old and 20 healthy subjects (12 were male, 29 years mean old). Patients and relatives were explored by un electrocardiogramme and un echocardiography. Morphologic and functional study were performed for LV and RV (diameters, volumes, peak systolic velocity from tissue Doppler and longitudinal strain measurement) based on speckle tracking. The global and segment longitudinal strain were measured from the basal and mid segments in both ventricles. LV longitudinal strain measurement was feasible in first degree relatives and controls on 85% and 67% but less in patients (78%). On first degree relatives, Segmental longitudinal peak systolic strain on LV (and basal lateral segments) was significantly lower than that of controls (−28.9±3.2% vs. −32.3±3.2%; P = 0.002 and −26.6±4.3% vs. −31.2±3.6%; P = 0.03 respectively) but significantly greater than that evaluated in ARVC patients (−20.6±4.7%; P = 0.001 and −19.5±4.2%; P = 0.01). For the left ventricular global strain, no differences were found between the control group and the first degree relatives (22.6±5.5% vs. −21.3±6.8%), but we found a difference between control group and patients (17.3±4.2%; P = 0.01).

Conclusion: Longitudinal strain of LV and RV segments was significantly lower in patients than in relatives and controls. The global and segment longitudinal function assessed by echocardiographic might help to detect early ARVC disease in firs degree relatives.