Conclusions: Despite normal ECGs, repolarisation abnormalities predisposing to arrhythmias might exist in asymptomatic relatives of ARVC patients.

1207-84 Are Right Ventricular Wall Motion Abnormalities Reliable for the Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy? A Cardiac Magnetic Resonance Imaging Study in Healthy Subjects Using a New Segmented Model

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Aims: To evaluate right ventricular wall motion abnormalities in healthy individuals using a new segmented model for the right ventricle.

Methods and results: 29 healthy individuals (9 female, 20 male, mean age 48.9±15 years) underwent magnetic resonance imaging (1.5 Tesla MR: Siemens, Erkelenz, Germany) to evaluate cardiac function and identify possible right ventricular wall motion abnormalities. True-FISP gradient-echo sequences with steady-state free precession were used for image acquisition. Right ventricular wall motion abnormalities were assessed and classified according to a segmented model for the right ventricle. In 27 (93.1%) of the 29 individuals right ventricular wall motion abnormalities were found. Dyskinesis was found in 22 (75.9%), hypokinesia in 11 (37.9%) and bulging in eight individuals (27.6%). The number of diagnosed wall motion abnormalities in the transverse plane (86.2%) was significantly higher compared to those found in the short axis plane (13.8%) or in the horizontal longitudinal plane (41.4%) (p<0.001).

Conclusions: Right ventricular wall abnormalities can be found in healthy individuals. Since these wall motion abnormalities are a criterion for the diagnosis of arrhythmogenic right ventricular cardiomyopathy, wall motion abnormalities around the insertion of the membranous Partials of the right ventricle should be excluded to prevent an incorrectly positive diagnosis.

1207-85 Diastolic Dysfunction After Neurocardiogenic Injury: Prevalence and Association With Pulmonary Edema


Introduction: ECG changes, troponin release, and reduced left ventricular ejection fraction (LVEF) have been described after subarachnoid hemorrhage (SAH). Little is known if this was associated with the following endpoints (by logistic regression): pulmonary edema (present vs. absent), LVEF ≤50% or <50%, and cTn (≥0.4 or >1.0 ug/L).

Methods: Over a period of 2 years, echocardiographic, clinical, chest X-ray, and cardiac troponin I (cTn) data was obtained on day 1, 3, and 6 following enrollment in 173 consecutive SAH patients admitted to UCSF medical center. Each echocardiogram included Doppler recordings of mitral inflow (early E and atrial contraction A) velocities, E wave deceleration time (DT), and pulmonary venous flow (systolic S and diastolic D) velocities. For adverse cardiac outcomes and may explain the development of pulmonary edema in some SAH patients.

Results: The study included 173 subjects, 154 had technically adequate Doppler data. Diastolic dysfunction was observed on at least one echocardiogram in 68% of subjects. The prevalence of diastolic vs. systolic dysfunction in 25 patients with pulmonary edema (1207-85) was significantly higher compared to those found in the short axis plane (13.8%) or in the horizontal longitudinal plane (41.4%) (p<0.001).

Conclusions: Despite normal ECGs, repolarisation abnormalities predisposing to arrhythmias might exist in asymptomatic relatives of ARVC patients. This study describes our experience of LVNC in paediatric patients seen at the Royal Children's Hospital, Melbourne, the sole paediatric cardiology service for a population of 4 million.

Aims: Anderson-Fabry disease (AFD) is an X-linked recessive disorder. Recent data have shown that cardiac involvement in males is common, however, the prevalence of cardiovascular abnormalities in females is uncertain. The aim of this study was to characterise the cardiac structure and function in female patients referred for family screening or for cardiac assessment.

Methods: Fifteen female heterozygotes (mean age 52.3±14.9 years, range 24-81) were identified either by mutation analysis or plasma a-galactosidase A (a-Gal) activity (mean 3.8 nmol/ml/h ± 1.19, range 1.6-6.6nmol/ml/h). All patients had an ECG and 2-D images and 4-chamber echocardiography. Twelve patients were able to perform maximum cardio-lunar exercise testing.

Results: Eleven of the 15 patients presented ≥20 years (mean 55 ± 11.4, range 40-78) with cardiac symptoms. Four patients had hypertension. Thirteen patients had abnormal ECGs: short PR interval (n=2), Romhilt-Estes criteria for left ventricular hypertrophy (n=5) and T wave changes (n=13). Left ventricular cavity dimensions and aortic root diameters were normal in all patients. Eleven patients had valvular abnormalities; 11 patients had an increased left ventricular mass index (2110 mg/m2) mean 133.9g/m2 ± 37.88, range 96-245g/m2, 4 patients had septal hypertrophy (18.8 mm ± 5.62, range 13-26mm). On transmitral Doppler, 5 patients had impaired relaxation with a reduction of their E/A ratio, and 1 patient had a restrictive left ventricular Doppler filling pattern. One patient had a low peak oxygen consumption ≤ 80% predicted on metabolic exercise testing.

Conclusions: This study suggests that cardiac involvement in females with Anderson-Fabry disease may be more common than previously reported.