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Can Electrocardiogram Help to Differentiate Emery-Dreifuss Muscular Dystrophy from Other Muscular Dystrophies?

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Emery-Dreifuss muscular dystrophy (EDMD) is a rare X-linked muscular disease with a potentially lethal cardiac arrhythmia.

The purpose of this study was to determine if electrocardiogram (ECG) was useful to differentiate EDMD from other muscular dystrophies, mainly facioscapulohumeral dystrophy and limb-girdle muscular dystrophy that usually have a benign course and a good prognosis.

Our own experience consists of 3 patients of the same family with EDMD, 2 of them having a persistent atrial standstill during 22 years follow-up. Therefore we have reviewed the ECG patterns of 125 cases of EDMD reported in the literature and found persistent atrial standstill (PAS) in 37 patients (30%) and complete (31 patients, 25%) or incomplete (22 patients, 18%) A-V block.

On the contrary no case of atrial standstill was recorded in the patients with the diagnoses of facioscapulohumeral dystrophy or other types of muscular dystrophies based on rigorous clinical criteria.

On the other hand, in a survey of the etiologies of 109 cases of PAS, we have found 36 patients with EDMD (33%). Moreover in 27 patients with EDMD who had a 12 lead surface ECG, PAS was associated with a left anterior fascicular block (LAFB) in 14 cases (52%). Conversely in 51 patients without EDMD, PAS was associated with a LAFB in 5 cases only (10%, $p < 0.01$).

Conclusion. 1 The presence of persistent atrial standstill can probably help to differentiate Emery-Dreifuss muscular dystrophy from other muscular dystrophies more especially as a left anterior fascicular block is associated.

2 Pacemaker insertion is recommended in that case to avoid the risk of sudden death.

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Importance of Holter Monitoring, as Compared to Exercise Testing, in Revealing ST-segment Changes in Patients with Syndrome X

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Despite growing interest and a large body of studies on syndrome X (anginal chest pain and angiographically normal coronary arteries), there are no data concerning the respective diagnostic value of exercise testing (ExT) and ambulatory Holter monitoring (HM) in revealing ischemic ST-segment changes in these patients. To address this problem, we performed ExT and 24-hour HM, off therapy, in 38 syndrome X patients (27 women, age 54 ± 8 years). ExT showed ST-segment depression (STd) in 28 patients (74% Group 1), whereas it was normal in 10 patients (26%, Group 2). Anginal chest pain during ExT was reported by 10 patients (36%) of Group 1 and 9 patients (90%) of Group 2 ($p < 0.01$). On the other hand, STd episodes were detected on HM in 22 patients (59%), 17 (61%) of whom belonging to Group 1 and 5 (50%) to Group 2 ($p = NS$). On the whole, 129 STd episodes, 14 (11%) of which associated to anginal pain, were found in the 22 patients (mean 5.8 ± 4.6 episodes/patient). A total of 21 STd episodes (5 associated to anginal pain) were detected on HM in the 5 patients with negative ExT (mean 4.3 ± 3.5 episodes/patient, range 1–10). In these 5 patients, heart rate at 1 mm STd during HM was significantly lower than that achieved at peak exercise during ExT (111 ± 19 vs. 136 ± 22 bpm, $p = 0.01$). The prevalence of positive HM in patients with negative ExT in our syndrome X patients was significantly higher than that observed in an unselected group of 76 patients with stable angina, significant coronary artery disease and negative ExT (50% vs 8%, $p < 0.01$). Thus, our data show that HM, with respect to ExT, has a significant additional diagnostic value in revealing the occurrence of ischemic ST-segment changes in patients with syndrome X. These findings also support the hypothesis that a microvascular vasoconstriction, rather than an impaired vasodilation, plays a major role in the pathophysiologic mechanisms leading to transient ischemia in these patients.

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Neural Sprouting and Ventricular Tachycardia

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Damaging the axons of a nerve may result in initial degeneration of the distal axon, followed by a regenerative process which forms aberrant synapses (neural sprouting). Because myocardial infarction (MI) can result in cardiac nerve damage, neural sprouting will follow, and could contribute to the generation of ventricular tachycardia (VT). To test this hypothesis, we retrospectively studied 7 pts with drug-resistant sustained monomorphic VT who underwent surgical subendocardial ablation. The age ranged from 54 to 77 years. All pts had a history of MI. Five pts had discrete ventricular aneurysm

and 2 pts had a history of sudden cardiac death. Computerized mapping was used to determine the VT origin. Surgical treatment was performed by endocardial excision techniques. Paraffin-embedded tissue samples from the sites where VT were originated were sectioned and stained with Hematoxylin and Eosin (H & E), and for S-100 protein using standard immunohistochemical techniques to identify Schwann cells. The results of S-100 protein stain were classified on a scale of 0 to 4+ based on light microscope examination, with 0 being negative and 4+ being strongly positive. The H & E stain showed various degrees of fibrosis in all specimens, compatible with old MI. The S-100 protein stain was positive in 6 of 7 pts, showing numerous small regenerated nerve twigs within the region of MI not seen in normal myocardium. The average strength of S-100 protein stain was $1.7 \pm 1.0+$ (range 0+ to 3+). Tyrosine hydroxylase stain was performed in 2 pts. Both showed positive staining. We conclude that sympathetic neural sprouting is present at the origin of VT and may play a role in the generation of VT and sudden death after MI.

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Assessment of Myocardial Perfusion and Function by MR

Monday, March 20, 1995, 3:00 p.m.–5:00 p.m.

Ernest N. Morial Convention Center, Hall E

Presentation Hour: 3:00 p.m.–4:00 p.m.

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Contrast Wash-in During MRI Myocardial Perfusion Imaging in Patients with Q-Wave Myocardial Infarction

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Eleven patients (7 anterior and 4 inferior) with Q-wave myocardial infarction (QMI) undergoing radionuclide imaging were studied using MR perfusion imaging techniques and quantitative computer analysis of the signal intensity (SI)-time curves. The purpose of this study was to determine the relative accuracy of slope (wash-in) and peak SI when compared to radionuclide images in myocardial regions with corresponding Q-waves on the electrocardiogram.

Patients underwent MR perfusion imaging using an ultrafast interpolated klyhole acquisition during bolus administration of gadoteridol (0.1 mmol/kg). Perfusion images were obtained under resting conditions and after dipyridamole (0.56 mg/kg). Radionuclide images were obtained the same day by either thallium exercise testing or hybrid thallium-^{99m}Tc sestamibi rest-dipyridamole stress testing and available for comparison. The slope of the SI-time curve (regression analysis) and peak SI were compared between normal and infarcted myocardium. The slope and SI were normalized to left ventricular cavity values, averaged, and compared using t-test statistical analysis.

Eight QMI patients with scarred myocardium determined by the radionuclide study had decreased slope (0.14 vs 0.47, $p < 0.001$) and peak SI (0.38 vs 0.58, $p = 0.01$) compared to normal regions. Slope (0.14 vs 0.23) and SI (0.38 vs 0.46) increased in the scarred regions following dipyridamole but not significantly. Normal regions had increased slope (0.47 vs 0.66, $p = 0.02$) but not SI following dipyridamole. Three QMI patients had reversible ischemia in Q-wave regions. Slopes were higher when compared to patients with scar (0.39 vs 0.14, $p = 0.02$) and more closely approximated normal regions (0.39 vs 0.55, $p = NS$). Ischemic regions had no significant change in slope (0.39 vs 0.40) or SI (0.54 vs 0.50) following dipyridamole.

These data suggest that slope may be a stronger indicator of artery patency and closely parallels the results of radionuclide imaging.

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Altered Myocardial High-Energy Phosphate Metabolism in Aortic Valve Disease: Failing Myocardium or Inadequate Blood Supply?

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³¹P MR spectroscopy has become a valuable tool for assessment of alterations in myocardial high-energy phosphate (HEP) metabolism in vivo. In this respect, depression of the phosphocreatine (Pcr) to ATP ratio has been suggested as an early marker of heart failure. This may be of clinical relevance during natural history of aortic regurgitation. To address this issue, ³¹P MR spectroscopy (ECG triggered 1D ISIS technique with correction for blood contamination) was performed preoperatively in 9 male patients (Group I, mean age 53.7 ± 10.3 years) with aortic stenosis (mean aortic valve gradient 58.1 mmHg), 9 male patients (Group II, mean age 45.1 ± 9.5 years) with aortic regurgitation, and in 12 age and sex matched healthy volunteers. All patients underwent right and left heart catheterization, left ventricular and coronary angiography, and routine echocardiography.