 sudden cardiac death is a dramatic, undesirable event that can often result from cardiomyopathies. To investigate the validity of the QT dispersion (QTd) in revealing regional heterogeneity of repolarisation, with consequent possibility of sudden death, we evaluated ECGs of patients affected by Becker muscular dystrophy (BMD). This is an X linked recessive muscular dystrophy caused by dystrophin anomalies in striated muscles with myocardial involvement and consequent dilated cardiomyopathy, ventricular arrhythmias and, in 30% of cases, sudden cardiac death.

**METHODS**

The study was retrospective using the clinically validated database of ECGs and echocardiograms (echo recorded together with the ECG) from 30 BMD patients (mean (SD) age 25 (10) years) with variable stages of myocardial involvement, and 26, age matched controls. All subjects underwent a physical examination, blood analyses, and M mode and two dimensional echocardiography. The diagnosis of the type of muscular dystrophy in Becker patients was confirmed by DNA analysis (polymerase chain reaction) and by reduced dystrophin labelling from the immunohistochemical examination of biopsy samples.

We excluded the ECGs from subjects with S-T T anomalies on the 12 lead ECG, and with sustained ventricular arrhythmias at 24 hour Holter monitoring, electrolyte imbalance, or QRS duration longer than 120 ms and from subjects undergoing cardiovascular treatment.

The standard 12 lead ECG, recorded at a paper speed of 25 mm/s (gain 10 mm = 1 mV) on a three channel recorder, was analysed for each subject by a blinded observer using a digitiser connected to a computerised system. QRS, RR, QT, JT (the latter obtained from the formula QT-QRS) were measured over three consecutive cycles. When U waves were present, the end of the T wave was considered the nadir of the curve between the T and the U waves. The QTd was defined as the difference between the maximum and minimum QT values. Extra and post-extra systolic cycles were excluded from the analysis.

For each parameter, a mean value was calculated for each lead dividing their sums by the total number of leads—at least 10 per subject. A study was also performed on the variability of QT measurements: 20 ECGs were coded, duplicated, and measured twice by the same blind observer. Percentage difference in QT measurements for the same ECG ranged from 1.5–2.8% for intra-observer variability. In addition, ejection fraction and fibre shortening were calculated from echocardiograms performed at the same time with ECG recordings.

Among the Becker patients, eight died from sudden cardiac death within 12 weeks after the registration of the examined ECG. There were no differences in ventricular arrhythmias, or echocardiographic findings between survivors and patients who died suddenly.

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Mean (SD) values and significance of the electrocardiographic and echocardiographic parameters in 30 Becker patients and in 26 age and sex matched controls</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>Becker patients</td>
</tr>
<tr>
<td>Sex</td>
<td>Men</td>
</tr>
<tr>
<td>Ejection fraction</td>
<td>53.00 (9.98)</td>
</tr>
<tr>
<td>Fibre shortening</td>
<td>779.10 (121.86)</td>
</tr>
<tr>
<td>RR interval</td>
<td>108.81 (15.98)</td>
</tr>
<tr>
<td>JT interval</td>
<td>277.3 (31.77)</td>
</tr>
<tr>
<td>QTc value</td>
<td>299.29 (49.68)</td>
</tr>
<tr>
<td>QT interval</td>
<td>386.14 (35.42)</td>
</tr>
<tr>
<td>QTc value</td>
<td>436.32 (30.30)</td>
</tr>
<tr>
<td>QTd interval</td>
<td>110.62 (28.76)</td>
</tr>
</tbody>
</table>

The results obtained comparing the electrocardiographic data from the eight Becker patients who died suddenly and from those who survived lend support to the use of the QT dispersion to foresee the risk of sudden death.

**RESULTS**

Findings from the electrocardiographic and echocardiographic parameters in BMD patients and in controls are presented in table 1. No significant difference between the two groups was found in RR, QT, JT and JTc. A significant increase in QRS duration and a highly significant difference in ventricular recovery time dispersion indexes (QTc, QTd) were observed in Becker patients compared to normal subjects.

The results obtained comparing the electrocardiographic and echocardiographic parameters from the eight Becker patients who suddenly died and from those who survived lend support to the use of the QT dispersion to foresee the risk of sudden death.

**Abbreviations:** BMD, Becker muscular dystrophy; QTd, QT dispersion
Our findings demonstrate that the analysis of dispersion of ventricular activation in the 12 lead surface ECG (QTd) was significantly increased in BMD patients; this parameter is principally related to non-homogeneous electrical activity. The case with which the QTd can be calculated, and the possibility of using the QTd value as a valid parameter to predict the risk of sudden death, may provide opportunities through anti-arrhythmic drug administration, or an implanted cardioverter-defibrillator, to prevent this dramatic event.

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IMAGES IN CARDIOLOGY

Magnetic resonance angiography in the evaluation of aortic pseudoaneurysm

Magnetic resonance angiography (MRA) and transesophageal echocardiography together are very useful imaging techniques in the evaluation of thoracic aortic pseudoaneurysms. A 63 year old hypertensive man was admitted to a community hospital by his primary care physician with three weeks history of night sweats, chills, fever, fatigue, and generalised myalgia. Blood cultures done in the outpatient clinic revealed group D salmonella bacteraemia. Transesophageal echocardiogram, abdominal computed tomography (CT), and ultrasound were normal. CT of the chest showed increased density within the anterior mediastinum, suspicious of an aortic pseudoaneurysm.

MRA of the thoracic aorta (left) showed two infected pseudoaneurysms, one in the descending aorta starting below the aortic arch (A), extending 7 cm caudally, and another smaller one in the lesser curve of the aortic arch (B). The latter (B) was missed on transesophageal echocardiography, although the former (A) was seen clearly. Preoperative coronary angiography showed critical three vessel disease. The patient successfully underwent resection of aneurysms with dacron patch repair of the lesser curve and Hemashield tube graft repair of the descending aorta. He was given ceftriaxone for a week and later switched to levofloxacin, which he has been taking long term.

Once invariably a fatal condition, infected aortic pseudoaneurysms can now be cured in most cases if diagnosed early and treated with appropriate surgical technique and antibiotic. Both MRA and transesophageal echocardiography complement each other in the evaluation of this condition.

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Is the value of QT dispersion a valid method to foresee the risk of sudden death? A study in Becker patients

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