Arrhythmia prevalence in patients with advanced cardiac dystrophinopathy

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Aims

To define the prevalence of tachy- and brady-arrhythmias in patients with advanced left ventricular dysfunction due to dystrophinopathy (LVEF ≤ 40% by echo-measurement) using continuous ECG-rhythm surveillance over six-month follow-up in a pilot observational study.

Methods and Results

Patients under regular follow up at a specialist cardiology-muscle clinic whose left ventricular ejection fraction was less than 40% at recent assessment were invited to participate. After providing informed consent, all had an ECG loop-recorder implanted, cardiac MR-imaging, signal averaging of their ECG and comprehensive rhythm surveillance for predefined arrhythmias - regardless of symptoms, for six months. All were already taking combination cardiac medications unless contraindicated (ie: ACE-inhibitor, beta-blocker, mineralocorticoid receptor antagonist) and none were on antiarrhythmic drugs.

Ten patients were studied [DMD 3, BMD 4, DMD-females 3; age 36.3 years (range 22.4-56.6); LVEF 32.9 ± 3.2%]. There were no deaths, brady- or sustained tachy-cardia episodes at any stage during follow-up. At least one self-limiting episode of arrhythmia was confirmed in seven (70%) – ventricular alone 3, atrial and ventricular 3 and atrial alone in one. None were symptomatic. Cardiac MRI showed chamber dilatation and varying extents of myocardial fibrosis in all and four had 'late potentials' on signal-averaged ECG. Further episodes of similar arrhythmias recurred during follow-up ranging 9-30 months. Ventricular arrhythmia features (eg: onset coupling intervals, irregularity, cycle length) suggested a variety of underlying mechanisms.

Conclusions

The results confirm that ventricular and atrial arrhythmias occur infrequently in patients with advanced cardiac dystrophinopathy. They overlap with multimodal indicators of poor cardiac prognosis and occur mostly secondary to advanced structural changes. Therefore, deploying specific anti-arrhythmic strategies prophylactically, such as cardioverter-defibrillators, would not be expected to improve group prognosis. Long-term survival in dystrophinopathy can best be achieved by deploying combination drug therapies optimally from an early age, to slow the decline in ventricular function and reduce myocardial fibrosis.

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