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Letter to the Editor

Cardiac Abnormalities in Duchenne and Becker Muscular Dystrophy

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In recent article, Kirchmann *et al.*, (2005) reported about the cardiac findings in 42 juvenile patients with Duchenne-muscular-dystrophy (DMD and 17 juvenile patients with Becker-muscular-dystrophy (BMD). The presented data evoked several objections.

It is not clear how often and at which intervals an individual patient was investigated. The authors speak of a "study period" but its duration is not mentioned. For determining the "onset" of a FS<25% repeated echocardiographic investigations are necessary.

According to table 1 12 DMD and 5 BMD patients have an FS<25%, whereas in the text 8 DMD and 4 BMD patients with an FS<25% are mentioned. The confusion is increased by figure 1 showing 1/17 BMD patient with a FS>24% at age 40, which makes 6%, but not 35%.

The analgetic medication is not mentioned although it is well known that non-steroidal anti-rheumatic drugs promote heart-failure.

Why did any of the DMD patients receive b-blockers, although 17% of the patients had sinus-tachycardia and 19% a FS<25%? B-blockers were previously recommended as an adjunctive therapy of tachycardious rhythm abnormalities and heart-failure (Bushby, K. *et al.*, 2003). Were b-blockers generally avoided because of their side-effects?

The authors seem to be unaware of left ventricular hypertrabeculation/noncompaction as a feature of CI in BMD (Stollberger, C., & Finsterer, J. 2004) but also DMD (Finsterer, J. *et al.*, 2005). Left ventricular hypertrabeculation is a congenital or acquired abnormality of the left ventricular myocardium, defined echocardiographically or autoptically according to meanwhile 4 different proposals (Burke, A. *et al.*, 2005; Chin, T.K. *et al.*, 1990; Oechslin, E.N.*et al.*, 2000; Stollberger, C., & Finsterer, J. 2004). Did the authors systematically look for left ventricular hypertrabeculation?

How many of the 59 mothers were systematically investigated for CI, since mothers of patients with dystrophinopathies frequently present with CI, requiring even heart-transplantation in single cases (Finsterer, J., & Stöllberger, C. 2003).

Was digoxin in the DMD-group given for atrial-fibrillation? Did the DMD-patient with atrial-flutter receive oral anticoagulation? What was the indication for digoxin in the BMD-patient?

How to explain autonomic involvement in >50% of DMD-patients but in none of the BMD-patients? Was this due to the young age of the BMD-patients, who had not yet developed autonomic impairment? Was there any indication for autonomic neuropathy in the included DMD-patients?

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It has been repeatedly reported that CI in dystrophinopathies is associated with certain preferred locations of deletions and point mutations in the Dystrophin-gene. It would be interesting to perform a genotype-phenotype correlation study in the 59 included patients addressing this issue.

Overall, we agree that there is a need to carefully monitor DMD and BMD-patients for CI as soon as they have been diagnosed or become symptomatic and to initiate adequate therapy.

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