

on a routine basis to detect cardiac involvement in DMD.<sup>5</sup> These tests should be performed at the time of the diagnosis, repeated every 2 years until the age of 10, and then yearly; ECG and echocardiography are to be recommended as the first tests.<sup>5</sup> These recommendations take into account the well-known natural history of the cardiomyopathy in DMD. Preliminary reports suggest that natriuretic peptides have a low negative predictive value in patients with DMD in the prediction of cardiac involvement, possibly due to the low physical activity of these patients.<sup>6,7</sup>

Secondly, therapeutic options should be considered in these high-risk patients. Previously, treatment was mainly supportive of heart failure, integrating angiotensin converting enzyme (ACE) inhibitors and beta-blocking agents.<sup>5</sup> The former have proven to be effective in the particular setting of DMD.<sup>8</sup> One may also speculate that these active drugs have a preventive effect if administered earlier in the course of the disease. Two recently published studies confirmed this hypothesis.<sup>9,10</sup> These studies have methodological limitations, i.e. the first is uncontrolled and evaluates short-term improvement in left ventricular ejection fraction (LVEF),<sup>9</sup> and the second is a two-phase study (a placebo, double-blind randomized period of 3y followed by open-label treatment with ACE inhibitor in all children).<sup>10</sup> However, both are highly clinically relevant and demonstrate that early treatment with ACE inhibitors at a pre-clinical stage might reduce deterioration of LVEF, an important prognostic factor in DMD. These results are in accordance with pharmacological hypotheses including afterload reduction and antifibrotic effects.<sup>11,12</sup> Finally, while steroids have demonstrated promising results on skeletal muscle function their effect on cardiac function needs to be established.

Should we apply the results of these studies in clinical practice? This effect may be transient, while DMD is a progressive disease. Definite conclusions will come from large trials using mortality as an end-point. However, as DMD is a rare disease, we should not expect several mortality trials to be published; studies using substitution criteria are to be considered. In the meantime, one should recommend simple use of validated attitudes including international expert consensus management recommendations<sup>5</sup> and more recent studies with concordant results.<sup>9,10</sup>

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## Erratum

'Social integration of adults with cerebral palsy'

Michelsen et al.

DMCN Vol **48**: 643–649

We would like to correct an error that was printed in the above mentioned article:

p 649: In Appendix 1 the values in the *n* columns and the percentage columns have been transposed: the values on the left are the percentages and those on the right are the *n* values.

We sincerely apologize for this error.

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