



Cardiac involvement in children with respiratory chain disorders

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Letter to the Editor

In their article, Yaplito-Lee et al. reported about the cardiac findings in 89 children with mitochondrial disorders (MIDs) [1]. This retrospective study raises some concerns. It would be useful to know if all cardiac investigations applied to assess cardiac involvement (CI) were carried out in each of the 89 patients. We should be informed if focal myositis mentioned in table 2 was attributed to the underlying mitochondrial disorder as MIDs may induce inflammatory reactions [2]. It should be also explained why the “structural heart defect” was not attributed to the underlying disorder.

The authors found CI in 29 of 89 patients (33%). However, chest X-ray was carried out only in 58 patients, ECG only in 36 and echocardiography in 36 patients? How can they refer to 89 patients if cardiac investigations were carried out only in a limited number of

patients? Were patients included which did not undergo any of these three investigations? It should be indicated how many had X-ray, ECG, or echocardiography alone, and how many underwent combined examinations? Obviously, none of the patients underwent 24h-ambulatory ECG, cardiac MRI, or coronary angiography.

Noncompaction is not restricted to patients with Barth syndrome but is increasingly recognized among patients with primary MIDs [3]. How many of the other patients had noncompaction? In how many was it associated with cardiac disease other than hypertrabeculation. According to which criteria was noncompaction diagnosed [4]? Heart transplantation (HTX) is increasingly carried out in patients with MIDs and therapy-resistant heart failure [5]. How many of the 89 underwent HTX and which were the long-term results? How long did the transplanted patients survive? According to table 1 one patient with

Kearns-Sayre syndrome (KSS) did not present with CI. According to our experience and previous reports, however, CI is a dominant feature of KSS, most frequently manifesting as arrhythmias, why these patients often require a pacemaker or an ICD [6]. CI in KSS is age-dependent. Was the patient without CI very young or was he not properly investigated for cardiac disease? How was subclinical CI defined? How many ECG abnormalities needed to be present to classify subclinical CI? CI needs to be thoroughly defined. The authors mention that an X-ray was carried out in 7 of 11 patients with subclinical CI. Subclinical was thus diagnosed only upon the ECG and echocardiography? Usually, it is also difficult to confirm that the described abnormalities are indeed a manifestation of the MID. Which differentials were considered and how were they excluded? Contrary to the statement in the discussion, cardiomyopathy has not been described together with mutations in the *SURF1* gene [7].

The conclusion that MID patients with CI have a less favorable outcome than patients without CI appears reasonable but a prospective design of the study would have strengthened these conclusions. Overall, MIDs appear to be associated with cardiac disease. Due to the retrospective design of the study, however, the conclusions, which can be drawn from this study are limited. Regular follow-up investigations are required to optimally manage CI in MIDs.

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