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Letter to the Editor

With interest we read the article by Louw et al. about measuring coenzyme-Q(CoQ)-levels in frozen muscle biopsies and work-up for the causes CoQ-deficiency.in 76 biopsies from patients with a suspected respiratory chain defect(RCD) [1]. We have the following comments and concerns.

Complex-II(CII) and complex III-(CIII)-deficiency not necessarily reflect primary CoQ-deficiency. CII/CIII-deficiency may also occur in patients with other RCD defects [2]. These RCD defects may go along with secondary CoQ-deficiency.

Patients with a MID or with CoQ-deficiency may require substitution with CoQ [3]. How many of the included patients were under a CoQ-therapy, necessitating the exclusion of such patients from the study.

Did the authors also apply their reference limits to patients with genetically confirmed primary CoQ-deficiency? Possibly, CoQ-levels are normal at least in some of these patients due to upregulation of CoQ-production in cellular compartments other than the mitochondria.

Since muscle CoQ-levels may also depend on lactate production [4], we should be informed how many of the included patients had in fact lactic acidosis.

Additionally, it would be useful to know which drugs the included patients were taking at the time of the muscle biopsy. Not only CoQ-substitution may influence muscle CoQ-levels, but also drugs, such as statins [5].

The physical status of a patient including muscle mass, physical fitness, and disabilities may further influence CoQ-levels. Thus, the severity of the muscular involvement, the patients' ability to ambulate, and the physical fitness should be reported. In which stage of the disease did the included patients undergo muscle biopsy? How many were disabled and dependent on a walking aid or a wheel chair and how many were bedridden?

Overall, this interesting study could be more meaningful by discussing the dependency of muscle CoQ on drugs, lactic acidosis, and the individual fitness. Upregulation of CoQ-production in compartments other than the mitochondria may further influence muscle CoQ.

Ethics approval and consent to participate

Not relevant

Consent for publication

Not relevant

Availability of data and material

Not relevant.

Competing interests

There are no conflicts of interest.

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Author contributions

All authors contributed equally, JF: design, literature search, discussion, first draft, SZ-M: literature search, discussion, critical comments.

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