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Letter to the Editor

Renal Disease in Mitochondrial Disorders Is Divers

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With interest we read the review article by Govers *et al.,* about renal involvement in mitochondrial disorders (MIDs) (Govers, L. P. *et al.,* 2020). The authors delineated tubular and non-tubular manifestations and subdivided the tubular manifestations into proximal (renal Fanconi syndrome) and distal (hypomagnesemia, hypokalemia) dysfunction. Non-tubular manifestations included nephrotic syndrome due to focal segmental glomerulosclerosis (FSGS) and tubulo-interstitial nephritis (TIN) (Govers, L. P. *et al.,* 2020). We have the following comments and concerns.

The main shortcoming of the review is that the list of renal manifestations in MIDs is incomplete. In a recent review about renal involvement in MIDs we identified a reduced glomerular filtration rate as the most frequent renal abnormality in MIDs (Finsterer, J., & Scorza, F. 2017). Other renal manifestations in MIDs not discussed in the review by Govers *et al.,* are renal cysts, nephrolithiasis, nephrocalcinosis, and renal neoplasms. In a study of 42 pediatric patients with a MID, 1 presented even with hydronephrosis (Martín-Hernández, E. *et al.,* 2005).

The second shortcoming is that the list of mitochondrial syndromes which have been reported in association with renal dysfunction is incomplete. Renal involvement may not only occur in mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes (MELAS) syndrome, Kearns-Sayre syndrome (KSS), Leigh syndrome, Pearson syndrome (PS), primary coenzyme-Q deficiency, complex-IV deficiency but also in mitochondrial depletion syndrome (MDS), progressive external ophthalmoplegia (PEO), Leber's hereditary optic neuropathy (LHON), X-linked sideroblastic anemia (XLSA), growth retardation, Fanconi type aminoaciduria, cholestasis, iron overload, profound lactic acidosis, and early death (GRACILE) syndrome, 3-methyl-glutaconic aciduria, deafness, encephalopathy, Leigh-like (MEGDEL) syndrome, HUPRA, myopathy, lactic acidosis, sideroblastic anemia (MLASA), multiple systemic lipomatosis (MSL), pyruvate-dehydrogenase deficiency, and in non-syndromic MIDs (Finsterer, J., & Scorza, F. 2017).

Missing is a discussion about secondary, non-renal manifestations of renal involvement in MIDs. Non-renal complications include polyneuropathy, renal anemia due to decreased production of erythropoietin, and arterial hypertension. There are also reports that serum troponin levels can be increased. Arterial hypertension may result from increased production of angiotensin.

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When proposing renal transplantation as a therapeutic option for MID patients with renal end-stage disease, it should be considered that these patients require life-long immunosuppression and steroids. From steroids it is well-known that they cause mitochondrial myopathy and that they may worsen manifestations of a pre-existing MID. From tacrolimus and sirolimus it is known that they may interfere with mitochondrial energy production and biogenesis.

Overall, this interesting review has a number of shortcomings, which need to be addressed before final conclusion can be drawn. The spectrum of renal involvement in MIDs is more widespread than anticipated and the number of MID syndromes in which renal involvement has been reported is higher than anticipated. Since long-term immunosuppression for renal transplantation may worsen a MID, it has to be indicated with caution in this population.

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