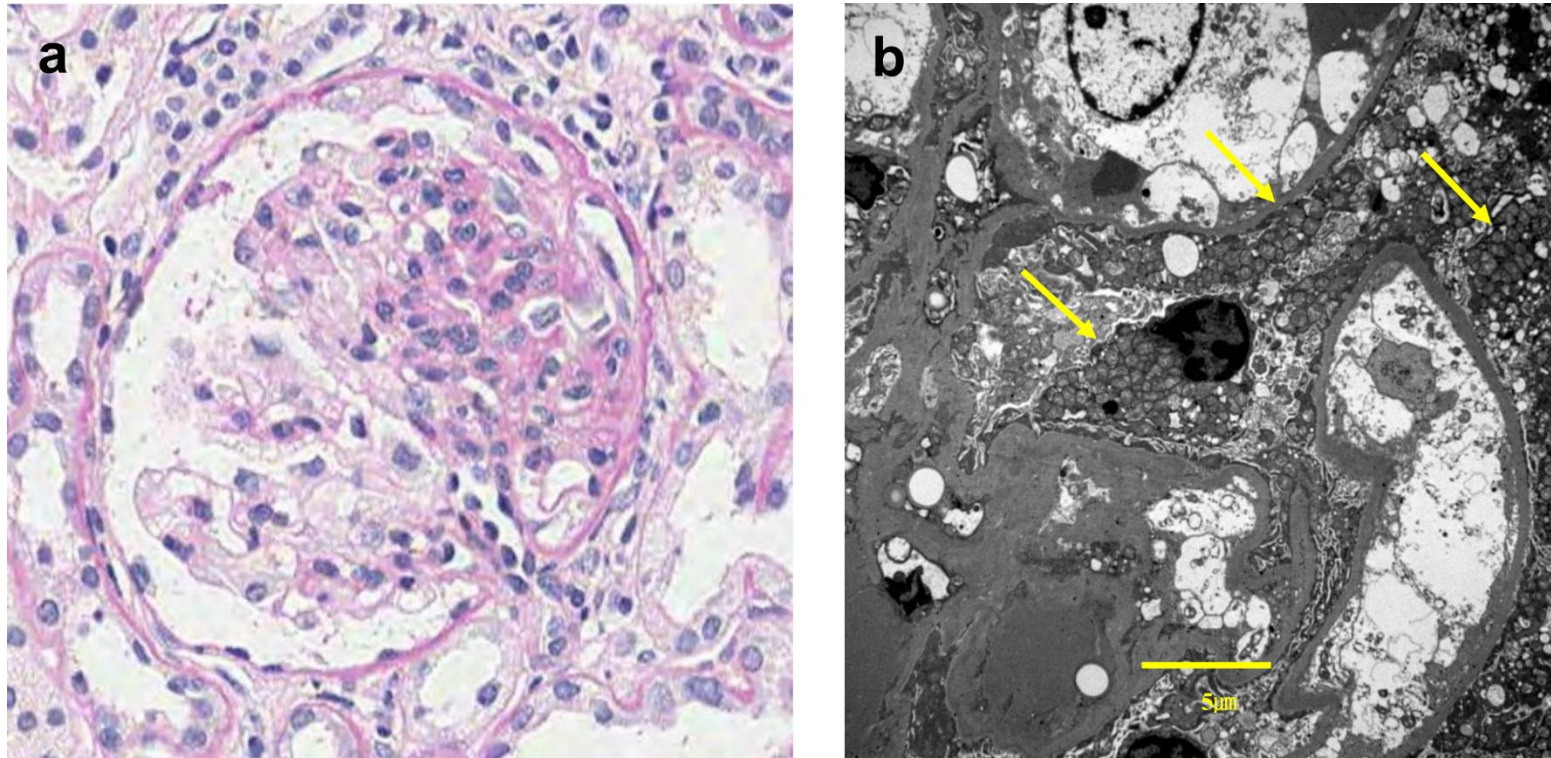


Effects of CoQ10 replacement therapy on the audiological characteristics of pediatric patients with *COQ6* variants

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Fig.S2



Supplementary figure S2. Light microscopic (a) and electron microscopic (b) findings of kidney biopsies of the patients with biallelic *COQ6* mutations manifesting as steroid-resistant focal segmental glomerulosclerosis and sensorineural hearing loss. A glomerulus featuring segmental sclerosis of the not-otherwise-specified variant (a: Patient 9, 400×, periodic acid-Schiff staining). A large collection of morphologically abnormal and distorted mitochondria (arrows) in podocyte cytoplasm (b: Patient 8-1).