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

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Table.S5

Supplementary table S5. Effect of renal function on the extent of hearing deterioration

	Patient 7	Patient 8-1	Patient 9	Patient 11
COQ6 Genotype	Q229P/ P261L	K64del/ P261L	Q229P/ P261L	P261L/ P261L
Change of renal function after CoQ10 administration	Aggravated	Aggravated	Improved	Not changed
Classification of hearing after CoQ10 administration	Non-responder	Responder	Non-responder	Responder
Correlation of renal function with extent of hearing deterioration	Possible	No	No	No

Note: Four patients were possible to analyze how CoQ10 supplement therapy affects not only hearing but also kidney function simultaneously. As for Patient 1-6, the renal function was in progress to ESKD before CoQ10 administration. As for Patient 8-2 and Patient 10, audiological follow-up was not available for more than 1 year after CoQ10 administration, thereby excluding from this analysis.