**Bi-allelic inactivating variants in the *COCH* gene cause autosomal recessive prelingual hearing impairment**

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Pathogenic variants in *COCH* are a known cause of DFNA9 autosomal dominant progressive hearing loss and vestibular dysfunction with adult onset. Hitherto, only dominant nonsynonymous variants and in-frame deletions with a presumed dominant negative or gain-of-function effect have been described. Here, we describe two brothers with congenital prelingual deafness and a homozygous nonsense c.292C>T(p.Arg98\*) *COCH* variant, suggesting a loss-of-function effect. Vestibular dysfunction starting in the first decade was observed in the older patient. The heterozygous parents and sibling have normal hearing and vestibular function, except for the mother, who shows vestibular hyporeflexia and abnormal smooth pursuit tests, most likely due to concomitant disease.

This is the first report of autosomal recessive inheritance of cochlea-vestibular dysfunction caused by a pathogenic variant in the *COCH* gene. An earlier onset of hearing impairment and vestibular dysfunction compared to the dominant hearing loss causing *COCH* variants is observed.

Keywords: *COCH* mutation - autosomal recessive - prelingual deafness