Impaired Hearing in Mitochondrial Disorders

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To the Editor: With interest, we read the article reported by Liu *et al*.[1] about the frequency of central and peripheral auditory system affection in 73 patients with a mitochondrial disorder (MID) due to mitochondrial DNA (mtDNA) point mutations or a single mtDNA deletion. We had the following comments and concerns.

How can the authors be sure that impaired hearing (IH) was due to mtDNA-mutations and not due to one of the frequent other genetic defects associated with IH?[2] Among those with a positive family history for IH, did the transmission follow a maternal trait in each case? How were genetic causes of IH other than the mtDNA-defect excluded?

It would be interesting to know how quickly IH progressed over time? Were patients repeatedly investigated and did IH progress with the same degree in all patients? Did those with initially normal findings develop abnormalities over time? How many patients profited from hearing devices and or cochlear implants?

Involvement of the peripheral nerves or the cranial nerves is a frequent finding in MIDs but often neglected by physicians.[3] In how many of the patients was IH attributable to neuropathy of the 8th cranial nerve and in how many of them was there affection of cranial nerves other than the acoustic nerve or of peripheral nerves? In how many patients was IH unilateral at onset and over which time did it progress to a bilateral problem?

The statement in the discussion that temporal asynchrony in central pathways or the auditory cortex should be diagnosed as auditory neuropathy spectrum disorder is contradictory. Why do the authors attribute a central nervous system problem to a peripheral nervous system disease?

Among those with IH due to affection of the central auditory pathways, was cerebral imaging indicative of an affection of the central auditory pathways as well?

There are indications that all MIDs sooner or later become multisystem diseases but that many patients do not survive that long. It is quite likely that also the auditory system will be sooner or later affected even in mono-organ MIDs, like mitochondrial myopathy, hepatopathy, hypothyroidism, nephropathy, chronic progressive external ophthalmoplegia, or dilative cardiomyopathy. In addition, it should be stated that nonsyndromic MIDs with IH are much more frequent than syndromic MIDs with IH.

Since subgroups were not evenly distributed, it would be interesting to know if the results differed with regard to gender or with regard to young and old age.

There are several reasons why IH is frequently missed by the treating physicians. First, doctors do not know about the frequent association between MID and IH; second, doctors do not ask for IH when taking the history; third, doctors recognize IH but do not attribute enough attention to it; and fourth, patients do not mention IH because they do not want to admit that they are handicapped.

Overall, this interesting study could profit from more extensive evaluation of its data and from a long-term follow-up to eventually show that the frequency of IH increases with age, and that the auditory system may be affected even in MID-patients with an initially mono-organ problem.

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