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Mitochondrial mutation associated with hearing loss

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Jonathan B Weitzman
Email: jonathanweitzman@hotmail.com

The identification of alleles associated with complex hearing defects, such as presbyacusis (age-related hearing loss, AHL), presents a formidable challenge to geneticists. In the February Nature Genetics Johnson *et al*. describe their use of elegant mouse breeding experiments to identify the first example of a mitochondrial DNA (mtDNA) mutation that acts as a modifier of a nuclear AHL locus (*Nat Genet* 2001, **27:**191-194). They performed a series of reciprocal backcrosses between three hearing-impaired inbred strains and a wild-type strain. Johnson *et al*. identified an mtDNA locus in one of these strains, the A/J strain, which affects hearing loss when mice are homozygous for the nuclear Ahllocus on mouse chromosome 10. Analysis of the A/J mtDNA genome revealed a single adenine nucleotide insertion in the *tRNA-Arg* gene. This unique example of nuclear-mitochondrial interaction will shed light on our understanding of human hearing impairmentconditions.

References

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