Waltzer 11 Jackson, a new spontaneous mutation in the *Cdh23* gene

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Mutation (allele) symbol:*Cdh23* $^{v\text{-}11J}$

Mutation (allele) name: waltzer 11 Jackson

Gene symbol: *Cdh23*

Strain of origin: B6(Cg)-Tyr$^{c\text{-}2J}$/J

Current strain name: B6(Cg)-*Cdh23*$^{v\text{-}11J}$/J

Stock #:008288 (As of March 6, 2009 this allele will be available only as DNA from The Jackson Laboratory DNA Resource.)

Phenotype categories: neurological/behavioral: motor capabilities/coordination/movement anomalies/deafness/circling

Origin and Description

The recessively inherited spontaneous mouse mutation waltzer 11 Jackson (*v-11J*) was identified in a B6(Cg)-Tyr$^{c\text{-}2J}$/J research colony. Mutant mice display head bobbing as well as a mild circling pattern of movement, both of which are usually indicative of vestibular dysfunction and probable hearing loss. Mutant mice were crossed to C57BL/6J and intercrossed and the Tyr$^{c\text{-}2J}$ mutation was bred out by selective mating; creating the new inbred strain B6(Cg)-*Cdh23*$^{v\text{-}11J}$/J (stock number 008288). The colony is currently maintained by brother/sister mating of a heterozygous female and a homozygous mutant male.

Genetic Analysis

An intercross was performed with CAST/EiJ mice and 54 mutant F2 animals were analyzed. Using our standard mapping protocol, the mutation was mapped to a region of Chromosome 10 where the *Cdh23* gene is located. Due to the mapped proximity to *Cdh23* a complementation test was performed between mice heterozygous for the new mutation and mice heterozygous for waltzer 2 Jackson (*v-2J*), a mouse mutation previously identified in the *Cdh23* gene. The test-cross produced 4 litters with a total of 16 mice, 5 of which were mutant, thus confirming allelism.

Pathology

A routine pathological screen of two 11 week old male mice, one mutant and one heterozygous control, revealed no gross abnormalities between the mutant and control except in the inner ear. A degenerated organ of corti was observed in the mutant inner

ear, with no abnormalities seen in the heterozygous littermate. The other organs of the inner ear appeared normal. The auditory brainstem response was used to assess the hearing of one mutant mouse and two littermate controls at 6 weeks of age. The mutant mouse was deaf showing no response at the highest stimulus presented (100 dB SPL) while the littermate controls exhibited good hearing. A clinical eye exam revealed no abnormalities.

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