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论文

GJB2基因条件敲除小鼠耳蜗扫描电镜观察

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摘要：

目的 进一步探讨Cx26突变导致耳聋的机制。方法 采用扫描电镜观察技术观察GJB2基因条件敲除小鼠模型（cCx26ko mice）小鼠出生后耳蜗表面形态的超微结构变化。结果 观察发现P8(出生第9天)和P12（出生第13天）小鼠耳蜗Corti器表面形态、毛细胞以及Deiters细胞等支持细胞均呈正常发育的一个演变过程，P21（出生第22天）小鼠耳蜗Corti器表面形态由底圈到中圈可见外毛细胞广泛散在性静纤毛的融合、脱落及外毛细胞缺失，Corti器网状板皱缩，并可见局部Corti器网状板破损，内毛细胞未见明显的病理变化。结论 GJB2基因缺失可以引起小鼠耳蜗Corti器外毛细胞静纤毛的缺失，Corti器网状板皱缩，局部Corti器网状板破损。对耳蜗结构的影响发生在小鼠出生后21d以前。

关键词： 缝隙连接26；突变；毛细胞，听觉；扫描电子显微镜

The morphologic presentation of cochlear in GJB2 conditional knock out mice under scanning electron microscopy

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Abstract:

Objective To explore the mechanism of hereditary hearing loss. Methods Cochlea from cCx26 mice on P8（the 8th postnatal day）, P12 （the 12th postnatal day）and P21（the 21st postnatal day） were examined with scanning microscope (SEM). Results On P8 and P12, the external morphology of inner hair cells (IHC), outer hair cells (OHC) and supporting cells developed normally. But in P21 stage, extensive coalesce or absence of stereo cilia of OHC and loss of OHC in the basal and middle turn of cochlea were seen. And the cuticular plate was shriveled and damaged, however, the IHCs in the cochlea were intact without any obvious change. Conclusion Mutation of GJB2 may cause the absence of stereo cilia of OHC, the shrinkage and the damage of Corti′s cuticular plate in mice. All these morphologic changes of cochlea may occur before P21.

Keywords: Connexin26; Mutation; Hair Cell, Auditory; Scanning Electron Microscopy

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