

# Gfi1<sup>1b/1b</sup>-mouse model

## A model for studying hearing loss

### Background

Gfi1 is a transcriptional repressor being expressed in the hematopoietic and nervous system. Loss of function leads to severe defects in hematopoiesis and inner ear development.

Gfi1 is highly similar to its paralogue Gfi1b with almost identical sequences in their respective protein domains.

### Technology

A knock-in mouse model was created in which the Gfi1 coding region was replaced by Gfi1b.

The Gfi1<sup>1b/1b</sup>-mice did not exhibit significant defects in the hematopoietic system as previously described for Gfi1<sup>-/-</sup>-mice, suggesting that Gfi1b is able to rescue the loss of Gfi1-function.

Interestingly, however, the Gfi1<sup>1b/1b</sup>-mice showed significant defects in inner ear development, i.e. hair cell formation, leading to hearing loss.

These results point to some interesting mechanisms on the molecular level for the differentiation and maintenance of hearing.

This makes the Gfi1<sup>1b/1b</sup>-model a valuable tool for the development of novel approaches for treating hearing loss.

### Further Reading

Fiolka et al. (2006): Gfi1 and Gfi1b act equivalently in haematopoiesis, but have distinct, non-overlapping functions in inner ear development. *EMBO Reports*, 7(3): 326-333

Möröy (2005): The zinc finger transcription factor Growth factor independence 1 (Gfi1). *Int J Biochem Cell Biol.* 37: 541-546

Wallis et al. (2003): The zinc finger transcription factor Gfi1, implicated in lymphomagenesis, is required for inner ear hair cell differentiation and survival development. *130: 221-232*

Saleque et al. (2002): The zinc-finger proto-oncogene Gfi1b is essential for development of the erythroid and megakaryocytic lineages. *Genes Dev.* 16: 301-306

### Current Status

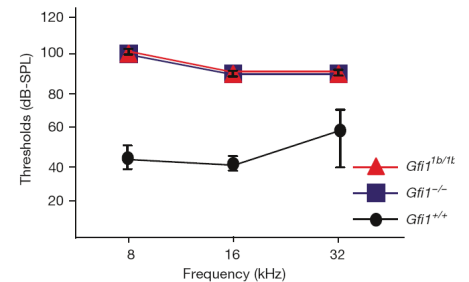
On behalf of the University of Duisburg-Essen, PROVendis offers access to the mouse model under a Material License Agreement.

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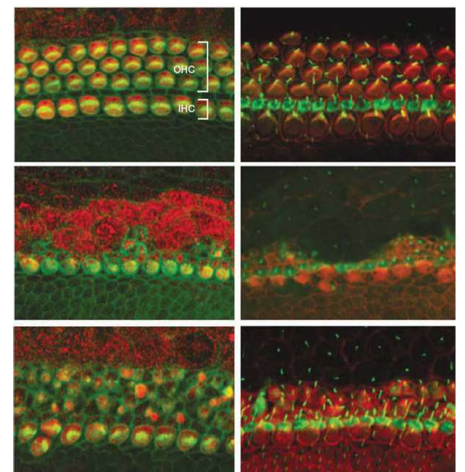
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### Evaluation of hearing capability



Auditory brainstem response thresholds of both Gfi1<sup>-/-</sup> (square) and Gfi1<sup>1b/1b</sup> (triangle) were determined to be over 100 dB at 8 kHz, whereas wild-type control mice (circle) were in normal range

### Expression of hair cell markers in cochlear epithelia



Left column: Cochlear epithelia of P0-mice was stained with antibody for myosin (red) labelling hair cells cuticular plate/cytoplasm and with phalloidin (green) labelling filamentous actin. Right column: Respective tissue was stained with antibody for acetylated tubulin (green) labelling hair cell kinocilia and various other tubulin-based structures and with phalloidin (red)  
Top: wild-type; mid: Gfi1<sup>-/-</sup>; bottom: Gfi1<sup>1b/1b</sup>

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