



Supplementary Figure 6. Strategy for targeted replacement of the wild-type *Dfnb59* allele with *Dfnb59*^{tmlUgds}. The mouse *Dfnb59* gene consists of only six exons, since there exists no exon equivalent to human untranslated exon 1. An asterisk indicates the position of the engineered R183W mutation in mouse exon 3. Scale bar: 1 kb.

Figure 6. Expression of pejvakin in the afferent auditory system of P30 mice from pure 129/Sv genetic background. The block diagram shows the main ascending pathways that project both ipsilaterally and contralaterally (for the sake of clarity, only projections from one cochlea are presented). Panels show expression of pejvakin in the somas of neurons located in the spiral ganglion (**a**), cochlear nuclei (**b**), superior olivary complex (**d**), and inferior colliculus (**e**). Pejvakin expression was not detected in fiber bundles, such as the trapezoid body (**c**). Note that, while pejvakin expression is detected in all spiral ganglion neurons, only a subset of neurons in brainstem auditory nuclei show pejvakin labeling, as indicated by co-immunolabeling with neuronal marker NeuN on adjacent sections (data not shown). Scale bars: 10 μm (**a, c**) and 50 μm (**b, d, e**).