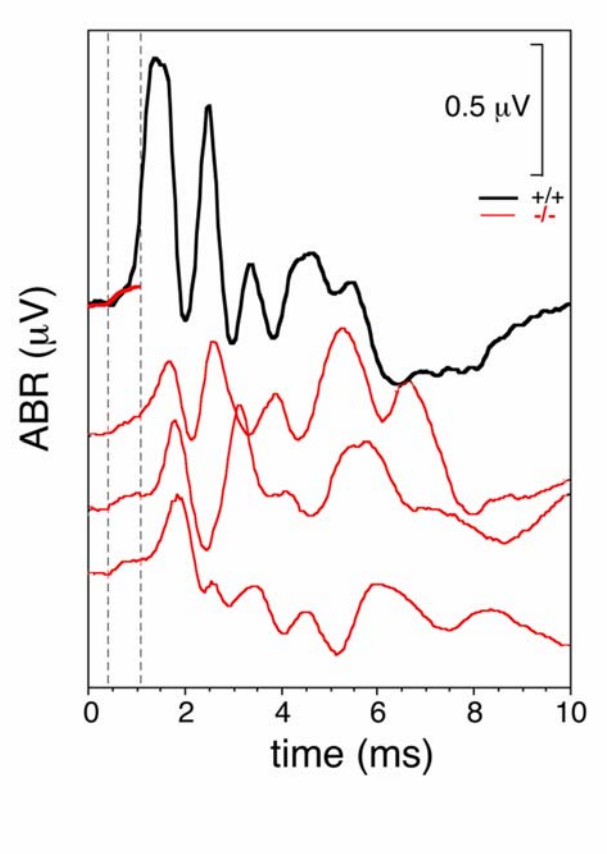

Supplementary Figure 5. Examples of ABR waveforms in response to 20 kHz tone bursts at 80 dB SPL in one ear of one wild-type (black) and three *Dfnb59* $^{tm1Ugds/tm1Ugds}$ mice (red) with increasingly delayed ABR waves (from top to bottom). Waveforms are offset for the sake of clarity. An early positive response is systematically present prior to ABR wave I, in the 0-to-1 ms post-onset time interval (vertical dashed lines). This early electrical response corresponds to the receptor potentials (summating potential, SP) of cochlear hair cells. Note that the amplitude of the response hardly changes between mutant mice, despite an increasing auditory dysfunction. The SP plateau is masked by the rise of ABR wave I in the wild-type ear, making it difficult to assess its amplitude. However, it is likely similar to the average SP of mutant ears (short red line superimposed on the wild-type ABR trace in the 0-to-1 ms time interval) and certainly does not exceed it.

Figure 5. Audiological characterization of postnatal day 30 *Dfnb59* knock-in mice. (**a**) Mean ABR thresholds of 8 wild-type (black squares) and 8 *Dfnb59* $^{tm1Ugds/tm1Ugds}$ (red diamonds) mice of mixed C57BL/6-129/Sv background are plotted against sound frequency. (**b**) Example of ABR waveforms at 20 kHz in one ear of a wild-type mouse (black lines, waves I to IV marked), superimposed on an example of ABR waveforms in one ear of a *Dfnb59* $^{tm1Ugds/tm1Ugds}$ (red lines, waves I to IV marked). Waveforms elicited at different hearing levels are offset for clarity. Labels on the right indicate stimulus levels (in dB SPL). An arrow indicates the peak of the wave corresponding to the electrical activity of hair cells, revealed here by the delay in wave I at stimulus levels over 90 dB. (**c**) Wave latency plotted as a function of the stimulus intensity at 20 kHz for the three more robust ABR waves (I, II and IV). Black lines mark the upper limit of the normative range in wild-type control mice of the same genetic background for waves I, II and IV (from bottom to top). Red lines show the results in individual ears from *Dfnb59* $^{tm1Ugds/tm1Ugds}$ mice exhibiting an ABR threshold elevation of more than 15 dB (shaded lines, wave I; dashed lines, wave II; continuous lines, wave IV). At all the frequencies tested, wave latencies in knock-in mice are longer than the maximum latency observed in their wild-type control littermates. (**d**) DPOAE growth functions at 15 kHz of wild-type (n=8; in black) and *Dfnb59* $^{tm1Ugds/tm1Ugds}$ ears exhibiting an ABR threshold elevation of more than 15 dB at 15 kHz (n=6; in red). Differences in the DPOAE growth functions between wild-type and *Dfnb59* $^{tm1Ugds/tm1Ugds}$ mice are not significant. Vertical bars, ± 1 s.e.m.