

| Supplementary material | | | | | | | | | |
|---------------------------------------|--------------|------------|-------------|--------------|-------------|------------|--------------|------------------|----------------|
| | | | | | | | | | |
| Table S1: 215 deafness genes screened | | | | | | | | | |
| ABHD1 2 | CDH23 | DIABL O | G1PC3 | ILDR1 | MITF | NLRP 3 | PRKCB | SLC 52A 3 | TN C |
| ACTG1 | CEACA MI6 | DIAP H1 | G4TA3 | JAG1 | MPZL2 | NPTN | PRORP | SLC 9A1 | TPR N |
| ADCY1 | CEP78 | DIAP H3 | GATA2 | KARS | MRPS7 | OPA1 | PRPS1 | SLC 9A3 R1 | TR1 OBP |
| AIFM1 | CHD7 | DMXL 2 | GGPS1 | KCNE1 | MSRB3 | OSBP L2 | PTPN11 | SM PX | TRP P3 |
| ALMS1 | CLDNI4 | DNFB 59 | GJB1 | KCNJ10 | MT -TH | OTO A | PTPRQ | SN AI2 | TSH Z1 |
| ANKRD 11 | CLIC5 | DSPP | GJB2 | KCNQ4 | MT- RNR1 | OTOF | RAF1 | SOX 10 | TSP EA R |
| ATP1A3 | CLPP | EDN3 | GJB3 | KCNQI | MT-TE | OTOG | RDX | SPT BN5 | TUB B4B |
| ATP2B2 | CLRN1 | EDNR B | GJB4 | KDM6A | MT-TF | OTOG L | ROR1 | STR C | TW NK |
| ATP6V1 B1 | CLRN2 | ELMO D3 | GJB6 | KIAA119 9 | MT-TI | PAX3 | RPGR | SY NE4 | TW NL4 A |
| ATP6V1 B2 | COCH | EPS8 | GPR98 | KITLG | MT-TK | PCDH I5 | S1PR2 | TBC 1D2 4 | TYR |
| BCS1L | COL11A 1 | EPS8L 2 | GPRAS P2 | KMT2D | MT- TL1 | PDXD CI | SERPIN B6 | TBL IXR 1 | US H1 C |
| BDP1 | COL11A 2 | ERAL1 | GPSM2 | LHFPL5 | MT-TS1 | PDZD 7 | SIX1 | TEC TA | US H1 G |
| BSND | COL2A1 | ESPN | GRHL2 | LMX1A | MYH14 | PEX1 | SL1TRK 6 | TIM M8 A | US H2 A |
| C0L9A2 | COL4A5 | ESRP1 | GRHLI | LONPI | MYH7 B | PEX26 | SLC17A 8 | TJP 2 | VEZ T |
| C1B2 | COL4A6 | ESRRB | GRXC R1 | LOXHD1 | MYH9 | PEX6 | SLC19A 2 | TM C1 | VPS 13B |

| | | | | | | | | | |
|-------------|--------|------------|-------------|--------------|------------|------------|-------------|----------------------|----------|
| C1SD2 | COL9A1 | EYA1 | GRXC R2 | LRTOMT | MYO15 A | PEX7 | SLC22A 4 | TM C2 | WB P2 |
| CABP2 | COL9A3 | EYA4 | HARS | MAF | MYO1 A | PHYH | SLC26A 4 | TM EM 126 A | WF S1 |
| CACNA 1D | CRYM | FAM6 5B | HARS2 | MARVEL D2 | MYO3 A | PLCB 3 | SLC26A 5 | TM EM 132 E | |
| CCDC5 0 | DAMT1 | FDXR | HGF | MCM2 | MYO6 | PMP2 2 | SLC29A 3 | TMI E | |
| CD151 | DCDC2 | FGF3 | HOME R2 | MCOLN3 | MYO7 A | PNPT I | SLC44A 4 | TM PRS S1 | |
| CD164 | DFNA5 | FGFR3 | HSD17 B4 | MET | NARS2 | POU3 F4 | SLC4A1 1 | TM PRS S3 | |
| CDCI4A | DFNB31 | FOXII | IARS2 | MIR96 | NLRP1 2 | POU4 F3 | SLC52A 2 | TM TC2 | |

Table S2: PCR primer sequences used for cloning into the luciferase reporter construct (5' - 3')

| | | | genomic position chr13 (hg38) | | distance from the <i>GJB2</i> transcripti onal start site (kb) | | |
|-----------|---|--|----------------------------------|----------------|---|----------------|--|
| PGJ B2 | F | CGATCTAAGTAAGCTTCGGTGAATTTAAAC GTTTGGTGGC | 20,192, 394 | 20,193, 934 | | | |
| | R | CCGGAATGCCAAGCTTCCGCAGAATCCTATC AGTTTCC | | | | | |
| C3 | F | ATAAGGATCCGTCGACCTGCTTCGGCCTCCA AATAG | 20,419, 403 | 20,420, 445 | - 221. 8 | - 222. 9 | |
| | R | AAGGGCATCGGTCGACCTTCTTCTGGAGTCTT TTCTGG | | | | | |

Table S3: PCR primer sequences used for mutagenesis (5' - 3')

| | | | genomic position chr13 (hg38) | |
|-----------------------|---|----------------------|----------------------------------|------------|
| PromGJB2_rs1425012952 | F | ccgcggcgcggcccccctcc | 20,193,013 | 20,193,031 |

| | | | | |
|-----------------------|---|---------------------|--|--|
| PromGJB2_rs1425012952 | R | ggagggggccgcgccgcgg | | |
|-----------------------|---|---------------------|--|--|