

<u>Surdités non syndromiques</u> Dominante (DFNA) (45 gènes)	ACTG1, CD164, CCDC50, CEACAM16, COCH, COL11A2*, CRYM, DIABLO (SMAC), DIAPH1, DMXL2, DSPP*, EYA4, GJB2*, GJB3, GJB6*, GRHL2, GSDME (DFNA5), HOMER2, <u>IFNLR1</u> , KCNQ4, KIAA1199 (CEMIP), KITLG, <u>LMX1A</u> , MCM2, MYH14, MYH9, MYO6*, MYO7A*, <u>NLRP3</u> , OSBPL2, P2RX2, <u>PDE1C</u> , <u>PLS1</u> , <u>POU4F3</u> , <u>REST</u> , SIX1*, <u>SLC9A3R1</u> , SLC17A8*, TBC1D24*, TECTA, TJP2, TMC1*, TNC, <u>TRRAP</u> , WFS1*
Récessive (DFNB) (77 gènes)	ADCY1, ATP2B2, BDP1, BSND, CABP2*, CDC14A, CDH23*, <u>CLDN9</u> , CIB2, CLIC5, CLDN14, COL11A2*, ELMOD3, EPS8, EPS8L2, <u>ESRP1</u> , DCDC2, ESPN, ESRRB, <u>GAB1</u> , GIPC3, GJA1, GJB2*, GJB6*, GPSM2, <u>GRAP</u> , GRXCR1, GRXCR2, HGF, ILDR1, KARS, LHFPL5, LOXHD1*, LRTOMT, MARVELD2, MET, <u>METTL13</u> , <u>MPZL2</u> , MSRB3, MTAP, MYO15A, MYO3A, MYO6*, MYO7A*, NARS2, OTOA, OTOF*, OTOG, OTOGL, PCDH15*, PDZD7*, PJKV (DFNB59), PNPT1*, <u>PPIP5K2</u> , PTPRQ, RDX, RIPOR2 (FAM65B), <u>ROR1*</u> , S1PR2, SERPINB6, SLC26A4*, SLC26A5, SLITRK6*, <u>SPNS2</u> , STRC, SYNE4, TBC1D24*, TMC1*, TMEM132E, TMIE, TMPRSS3, TPRN, TRIOBP, TSPEAR, USH1C*, <u>WBP2</u> , WHRN*,
Lié à l'X (6 +1)	AIFM1 (DFNX5)*, COL4A6, <u>GPRASP2</u> , POU3F4, PRPS1, SMPX, TBL1X
<u>Surdités syndromiques (N=88)</u> USHER Branchiooculofacial -BOF BOR cérebrooculofacial-COFS Alport Charge Pendred Perrault* Stickler Jervel et Lange Nielsen Norrie Waardenburg et Hirschprung Treacher Collins Kallmann Townes Brocks PHARC Harboyan Myopie surdité (ANSD) Déficit transporteur riboflavine (ANSD) Insuffisance hypophysaire et surdité Atrésie congénitale Alagille LAMM, Muenke Barakat (HDR) Microphthalmie Piebaldisme et surdité Mohr-Tranebjaerg (MTS) (ANSD) Wolfram Charcot-Marie-Tooth et surdité <u>Syndrome d'Emberger Duane</u> <u>microtie</u>	MYO7A*(USH1B), USH1C*, CDH23 (USH1D), PCDH15* (USH1F), SANS (USH1G), USH2A, ADGRV1(USH2C, GPR98), WHRN*(USH2D), CLRN1 (USH3), HARS, PDZD7 (USH3B) TFAP2A EYA1, SIX1*, (SIX5) ERCC2 COL4A3, COL4A4, COL4A5* CHD7, SEMA3E SLC26A4*, FOXI1, KCNJ10, CLPP, HARS2, HSD17B4, LARS2 COL2A1, COL9A1, COL9A2, COL11A1, COL11A2, KCNE1, KCNQ1 NDP ECE1, EDNRB, EDN3, MITF, PAX3, SNAI2, SOX10, POLR1C, POLR1D, TCOF1 ANOS1(KAL1), FGF8, FGFR1, PROK2, PROKR2 SALL1, SALL4 ABHD12 SLC4A11 SLITRK6* SLC52A2, <u>SLC52A3</u> LHX3 TSHZ1 JAG1 FGF3, FGFR3 GATA3 SOX2 KIT TIMM8A WFS1* PMP22 <u>GATA2</u> , <u>CHN1</u> , <u>HOXA2</u> , <u>MAN2B1</u>
<u>Neuropathies auditives (17)</u> Isolées (NA, RA, X) Syndromiques (ANSD)	AIFM1* AUNA, CABP2*, <u>CAV1</u> , LOXHD1, OTOF*, <u>ROR1*</u> , *SMAD4, SLC17A8*, TBC1D24*, <u>TMTC2</u> , C10ORF2 (TWNK), CMT1 (MPZ), ERAL1, ERCC4, ERCC6, ERCC8, FDXR, FXN, OPA1, PNPT1*, <u>PTRH2</u> , SLC19A2, TMEM126A

nouveaux gènes panel V4 , (*) gènes appartenant à plusieurs groupes