

**Supplementary Table 1: Human genes captured**

Gene name	Chromosomal locus	Group <sup>A</sup>	Recessive NSHL	Dominant NSHL	X-linked NSHL	Human syndrome	Number of captured exons	Number of captured bp
<i>ABR</i>	17p13.3	Mouse					25	10527
<i>ACAN</i>	15q26.1	Mouse					18	12140
<i>ACTB</i>	7p22	Human SHL, mouse				Juvenile-onset dystonia	7	2837
<i>ACTG1</i>	17q25.3	Human NSHL		DFNA20/26			5	2793
<i>ALDH1A2</i>	15q21.3	Mouse					15	5716
<i>ALMS1</i>	2p13	Mouse					23	16184
<i>AP3D1</i>	19p13.3	Mouse					31	9139
<i>APAF1</i>	12q23	Mouse					27	9495
<i>AQP4</i>	18q11.2	Mouse					5	5966
<i>ATF2</i>	2q32	Mouse					14	4663
<i>ATOH1</i>	4q22	Mouse					1	1201
<i>ATP2B2</i>	3p25.3	Mouse					26	12188
<i>ATP6V1B1</i>	2p13.1	Human SHL				Distal renal tubular acidosis (RTA) with sensorineural deafness	16	5190
<i>ATP8B1</i>	18q21.31	Mouse					28	9080
<i>AXIN1</i>	16p13.3	Mouse					11	4770
<i>BARHL1</i>	9q34	Mouse					3	2403
<i>BBS1</i>	11q13.1	Mouse					18	5895
<i>BBS4</i>	15q22.3	Mouse					16	5735
<i>BCR</i>	22q11.23	Mouse					23	11706
<i>BCS1L</i>	2q33	Human SHL				Björnstad syndrome, HL and pili torti; Leigh syndrome	28	3428
<i>BDNF</i>	11p13	Mouse					21	11541
<i>BMP4</i>	14q22	Mouse					5	2566
<i>BSN</i>	3p21.31	Mouse					12	16738
<i>BSND</i>	1p32.1	Mouse					4	1845
<i>CACNA1D</i>	3p14.3	Mouse					50	15080
<i>CACNB2</i>	10p12	Mouse					19	8340
<i>CACNG2</i>	22q13.1	Mouse					4	4368
<i>CASP3</i>	4q34	Mouse					8	4090

<i>CCDC50</i>	3q28	Human NSHL		DFNA44		12	10148
<i>CDH23</i>	10q22.1	Human NSHL, SHL	DFNB12		Usher, USH1D	67	21442
<i>CDKN1B</i>	12p13.1	Mouse				3	2724
<i>CDKN2D</i>	19p13	Mouse				2	1643
<i>CELSR1</i>	22q13.3	Mouse				35	15836
<i>CHD7</i>	8q12.1	Mouse				38	15355
<i>CHRNA9</i>	4p14	Mouse				5	2525
<i>CKB</i>	14q32	Mouse				6	2286
<i>CLDN11</i>	3q26.2	Mouse				6	3206
<i>CLDN14</i>	21q22.13	Human NSHL	DFNB29			7	3006
<i>CLDN9</i>	16p13.3	Mouse				1	2161
<i>CLIC5</i>	6p21.1	Mouse				7	8891
<i>CLRN1</i>	3q25	Human SHL, mouse			Usher, USH3A	4	3166
<i>COCH</i>	14q12	Human NSHL		DFNA9		12	4661
<i>COL11A1</i>	1p21	Human SHL, mouse			Stickler, STL2; Marshall; Fibrochondrogenesis	69	17185
<i>COL11A2</i>	6p21.32	Human NSHL, SHL	DFNB53	DFNA13	Stickler, STL3; Fibrochondrogenesis 2; Otospondylomegaepiphyseal dysplasia; Weissenbacher-Zweymuller syndrome	66	16278
<i>COL2A1</i>	12q13.11	Human SHL, mouse			Stickler, STL1; Czech dysplasia; Epiphyseal dysplasia, multiple, with myopia and deafness; Otospondylomegaepiphyseal dysplasia; Spondyloepiphyseal dysplasia congenita	54	13553
<i>COL4A3</i>	2q36	Human SHL, mouse			Alport	53	14732
<i>COL4A4</i>	2q35	Human SHL			Alport	49	17021
<i>COL4A5</i>	Xq22.3	Human SHL			Alport	51	14759
<i>COL9A1</i>	6q12	Human SHL, mouse			Stickler, STL4	38	9866
<i>COL9A3</i>	20q13.3	Human NSHL				32	7081

<i>CPLX1</i>	4p16.3	Mouse			4	2485	
<i>CRYM</i>	16p12.2	Human NSHL		DFNA40	12	3292	
<i>DDB2</i>	11p12-p11	Human SHL			Xeroderma pigmentosum	10	4337
<i>DDR1</i>	6p21.3	Mouse				21	6930
<i>DFNA5</i>	7p15.3	Human NSHL		DFNA50		11	4812
<i>DFNB31</i>	9q32	Human NSHL, SHL	DFNB31		Usher, USH2D	13	6334
<i>DFNB59</i>	2q31.2	Human NSHL	DFNB59			7	2407
<i>DIAPH1</i>	5q31.3	Human NSHL		DFNA1		28	9442
<i>DIO2</i>	14q24.2	Mouse				6	7328
<i>DIO3</i>	14q32	Mouse				1	2161
<i>DLX2</i>	2q32	Mouse				3	2844
<i>DLX5</i>	7q22	Mouse				3	2124
<i>DMD</i>	Xp21.2	Mouse				85	28443
<i>DSPP</i>	4q22.1	Human NSHL		DFNA36		5	2806
<i>DVL1</i>	1p36	Mouse				14	5990
<i>DVL2</i>	17p13.2	Mouse				15	4971
<i>DVL3</i>	3q27	Mouse				15	6038
<i>EDN3</i>	20q13.2	Human SHL, mouse			Waardenburg, WS4	5	3445
<i>EDNRB</i>	13q22	Human SHL, mouse			Waardenburg, WS4	8	5883
<i>EPHB1</i>	3q21	Mouse				16	6939
<i>EPHB2</i>	1p36.1	Mouse				16	6776
<i>EPHB3</i>	3q21	Mouse				17	5854
<i>ERBB4</i>	2q33.3	Mouse				28	15733
<i>ERCC1</i>	19q13.32	Human SHL			Xeroderma pigmentosum	10	4653
<i>ERCC2</i>	19q13.3	Human SHL			Cockayne syndrome, Xeroderma pigmentosum	23	5803
<i>ERCC3</i>	2q21	Human SHL			Cockayne syndrome, Xeroderma pigmentosum	15	5692
<i>ERCC4</i>	16p13.12	Human SHL			Xeroderma pigmentosum; XFE progeroid syndrome	11	8176
<i>ERCC5</i>	13q33	Human SHL			Cockayne syndrome, Xeroderma pigmentosum	15	9095
<i>ESPN</i>	1p36.31	Human NSHL	DFNB36			13	5047

<i>ESR2</i>	14q23.2	Mouse				11	9056
<i>ESRRB</i>	14q24.3	Human NSHL	DFNB35			11	4570
<i>EYA1</i>	8q13.3	Human SHL, mouse			Branchio-oto-renal, BOR1	18	7283
<i>EYA4</i>	6q23.2	Human NSHL		DFNA10		21	8888
<i>FABP4</i>	8q21	Mouse				4	1444
<i>FAS</i>	10q24.1	Mouse				9	6550
<i>FBXO10</i>	9p13.2	Suspected				11	6013
<i>FBXO2</i>	1p36.22	Mouse				5	2407
<i>FGF3</i>	11q13	Mouse				3	1864
<i>FGFR1</i>	8p11.2	Mouse				21	9979
<i>FGFR2</i>	10q26	Mouse				21	7861
<i>FGFR3</i>	4p16.3	Mouse				20	6313
<i>FIGN</i>	2q24.3	Mouse				3	4925
<i>FOXG1</i>	14q13	Mouse				1	2082
<i>FOXI1</i>	5q34	Human SHL, mouse			Enlarged vestibular aqueduct	2	2443
<i>FZD3</i>	8p21	Mouse				8	12977
<i>FZD6</i>	8q22.3	Mouse				8	5128
<i>GATA3</i>	10p15	Mouse				6	3648
<i>GBX2</i>	2q37	Mouse				2	1684
<i>GFI1</i>	1p22	Mouse				9	3923
<i>GJA1</i>	6q21-q23.2	Human SHL, suspected NSHL			Oculodentodigital dysplasia	3	3362
<i>GJA1P1</i>	5q21.3	Suspected				1	3121
<i>GJB1</i>	Xq13.1	Human SHL			Charcot-Marie-Tooth Neuropathy X	10	2163
<i>GJB2</i>	13q12.11	Human NSHL, SHL	DFNB1A	DFNA3A	Bart-Pumphrey syndrome; Hystrix-like ichthyosis with deafness; Keratitis- ichthyosis-deafness syndrome; Keratoderma, palmoplantar, with deafness; Vohwinkel syndrome	2	2442
<i>GJB3</i>	1p34.3	Human NSHL		DFNA2B		3	3726

<i>GJB4</i>	1p34.3	Human SHL, suspected NSHL			Palmoplantar Keratoderma, Erythrokeratoderma variabilis et progressiva	2	2123
<i>GJB5</i>	1p35.1	Suspected				2	1242
<i>GJB6</i>	13q12.11	Human NSHL	DFNB1B; digenic <i>GJB2/GJB6</i> deafness	DFNA3B		5	3162
<i>GLI3</i>	7p13	Mouse				15	10379
<i>GPR98</i>	5q13	Human SHL, mouse			Usher, USH2C	90	31270
<i>GPSM2</i>	1p13.3	Human NSHL	DFNB82			16	7173
<i>GPX1</i>	3p21.3	Mouse				1	1322
<i>GRHL2</i>	8q22.3	Human NSHL		DFNA28		16	7816
<i>GRID1</i>	10q22	Mouse				16	8339
<i>GRXCR1</i>	4p13	Human NSHL	DFNB25			4	1404
<i>GSTM1</i>	1p13.3	Sensitivity to hearing loss				8	2304
<i>GSTP1</i>	11q13	Sensitivity to hearing loss				7	1897
<i>GSTT1</i>	22q11.23	Sensitivity to hearing loss				5	2447
<i>GUSB</i>	7q21.11	Mouse				12	3958
<i>HAL</i>	12q22	Mouse				20	5668
<i>HES1</i>	3q28	Mouse				4	1952
<i>HES5</i>	1p36.32	Mouse				3	1321
<i>HGF</i>	7q21.11	Human NSHL	DFNB39			19	6699
<i>HMX2</i>	10q25.2	Mouse				2	1803
<i>HMX3</i>	10q26.13	Mouse				2	1284
<i>HOXA1</i>	7p15.3	Mouse				2	2763
<i>HOXA2</i>	7p15	Mouse				2	1762
<i>HOXB1</i>	17q21.3	Mouse				2	1322
<i>IFT88</i>	13q12.1	Mouse				28	8430
<i>IGF1</i>	12q22	Mouse				6	8091
<i>ILDR1</i>	3q13.33	Human NSHL	DFNB42			8	3570
<i>ITGA8</i>	10p13	Mouse				30	7350

<i>JAG1</i>	20p12.1	Mouse			26	10162
<i>JAG2</i>	14q32	Mouse			23	7484
<i>KCNE1</i>	21q22.12	Mouse		Jervell & Lange-Nielsen, JLNS2	6	3409
<i>KCNJ10</i>	1q22	Mouse			2	5325
<i>KCNMA1</i>	10q22.3	Mouse			31	21289
<i>KCNQ1</i>	11p15.5	Human SHL, mouse		Jervell & Lange-Nielsen, JLNS1	17	5859
<i>KCNQ4</i>	1p34.2	Human NSHL	DFNA2A		14	5694
<i>KIT</i>	4q11	Mouse			21	7796
<i>KITLG</i>	12q22	Mouse			10	6613
<i>LAMA2</i>	6q22	Mouse			65	18502
<i>LARGE</i>	22q12.3	Mouse			17	6608
<i>LFNG</i>	7p22.2	Mouse			13	4432
<i>LHFPL5</i>	6p21.31	Human NSHL	DFNB67		4	2205
<i>LHX3</i>	99q34.3	Human SHL		Hypopituitarism and hearing loss	6	3593
<i>LMO4</i>	1p22.3	Mouse			5	6050
<i>LMX1A</i>	1q22	mouse			10	5251
<i>LOXHD1</i>	18q21.1	Human NSHL, mouse	DFNB77		44	14385
<i>LRIG3</i>	12q14.1	Mouse			20	6860
<i>LRP2</i>	2q24	Mouse			79	26282
<i>LRTOMT</i>	11q13.4	Human NSHL	DFNB63		11	5853
<i>MAFB</i>	20q11.2	Mouse			1	3164
<i>MAP1A</i>	15q13	Mouse			6	10940
<i>MARVELD2</i>	5q13.2	Human NSHL	DFNB49		8	3409
<i>MCOLN3</i>	1p22.3	Mouse			12	5233
<i>MIR182</i>	7q32.2	Suspected			1	241
<i>MIR183</i>	7q32.2	Suspected			1	
<i>MIR96</i>	7q32.2	Human NSHL	DFNA50		1	469
<i>MITF</i>	3p14.2	Human SHL, mouse		Waardenburg, WS2A; Tietz albinism-deafness syndrome	13	7776
<i>MKKS</i>	20p12	Mouse			7	3567
<i>MOS</i>	8q11	Mouse			1	1201
<i>MPV17</i>	2p23.3	Mouse			9	2715

<i>MSRB3</i>	12q14.3	Human NSHL	DFNB74			8	5171
<i>MSX2</i>	5q34	Mouse				2	2403
<i>MTAP</i>	9p21.3	Human NSHL		Translocation		9	4974
<i>MUTED</i>	6p25.1	Mouse				5	3287
<i>MYH14</i>	19q13.33	Human NSHL, SHL		DFNA4	Peripheral neuropathy, myopathy, hoarseness, and hearing loss	43	12540
<i>MYH9</i>	22q12.3	Human NSHL, SHL		DFNA17	Epstein syndrome; Fechtner syndrome; Macrothrombocytopenia and progressive sensorineural deafness; May-Hegglin anomaly; Sebastian syndrome	41	13763
<i>MYO15A</i>	17p11.2	Human NSHL	DFNB3			64	20241
<i>MYO1A</i>	12q13.3	Human NSHL		DFNA48		29	7428
<i>MYO1C</i>	17p13	Human NSHL				32	8677
<i>MYO1F</i>	19p13.3-p13.2	Human NSHL suggested				28	7104
<i>MYO3A</i>	10p12.1	Human NSHL	DFNB30			35	12345
<i>MYO6</i>	6q14.1	Human NSHL	DFNB37	DFNA22		35	12353
<i>MYO7A</i>	11q13.5	Human NSHL, SHL	DFNB2	DFNA11	Usher, USH1B	50	15050
<i>NAV2</i>	11p15.1	Mouse				40	17935
<i>NDP</i>	Xp11.4	Human SHL, mouse			Norrie, NDP	3	2324
<i>NEU1</i>	6p21.3	Mouse				6	2804
<i>NEUROD1</i>	2q32	Mouse				2	2797
<i>NEUROG1</i>	5q23	Mouse				1	1762
<i>NF1</i>	17q11.2	Mouse				60	24863
<i>NOTCH1</i>	9q34.3	Mouse				33	14296
<i>NOX3</i>	6q25.1	Mouse				14	3508
<i>NOXO1</i>	16p13.3	Mouse				7	2455
<i>NR2F1</i>	5q14	Human SHL			Profound deafness, history of feeding difficulties, dysmorphism, strabismus, developmental delay	4	2647
<i>NR4A3</i>	9q22	Mouse				9	7815
<i>NTF3</i>	12p13	Mouse				2	1603

<i>NTN1</i>	17p13	Mouse			7	6449
<i>NTRK2</i>	9q22.1	Mouse			23	20733
<i>NTRK3</i>	15q25	Mouse			21	7941
<i>OC90</i>	8q24.22	Mouse			14	4456
<i>OTOA</i>	16p12.2	Human NSHL	DFNB22		30	6989
<i>OTOF</i>	2p23.3	Human NSHL	DFNB9		47	13890
<i>OTOG</i>	11p15.1	Mouse			22	7662
<i>OTOP1</i>	4p16.3	Mouse			6	2647
<i>OTOR</i>	20p12.1-p11.23	Suspected			4	2125
<i>OTX1</i>	2p13	Mouse			5	3849
<i>OTX2</i>	14q21	Mouse			5	3004
<i>P2RX2</i>	12q24.33	Human NSHL			12	2803
<i>PAX2</i>	10q24	Mouse			12	6337
<i>PAX3</i>	2q35	Human SHL, mouse		Waardenburg, WS1 and WS3; Craniofacial- deafness-hand syndrome	9	5370
<i>PCDH15</i>	10q21.1	Human NSHL, SHL	DFNB23	Usher, USH1F	43	18815
<i>PDZD7</i>	10q24.31	Human SHL digenic gene		Usher, USH2C, digenic; modifier of USH2A	13	6499
<i>PHEX</i>	Xp22.2	Mouse			22	6182
<i>PLDN</i>	15q21.1	Mouse			5	4488
<i>PMP22</i>	17p12	Human SHL		Dejerine-Sottas disease; Charcot-Marie-Tooth disease; Hereditary motor and sensory neuropathy Type 1A; Hereditary neuropathy with liability to pressure palsies.	5	2687
<i>PNOC</i>	8p21	Mouse			4	2725
<i>POLH</i>	6p21.1	Human SHL		Xeroderma pigmentosum	11	6097
<i>POU1F1</i>	3p11	Mouse			6	2166
	Xq21.1	Human NSHL, mouse		DFNX2 (DFN3)	1	1402
<i>POU3F4</i>						
<i>POU4F3</i>	5q31	Human NSHL	DFNA15		2	1442
<i>PROP1</i>	5q35.3	Mouse			3	1883



<i>PRPS1</i>	Xq22	Human NSHL		DFNX1 (DFN2)	Arts syndrome; Charcot-Marie-Tooth disease X; Gout; Phosphoribosylpyrophosphate synthetase superactivity	7	3167
<i>PRRX1</i>	1q24	Mouse				5	4528
<i>PRRX2</i>	9q34.1	Mouse				4	1444
<i>PTK7</i>	6p21.1	Mouse				20	6970
<i>PTPRQ</i>	12q21.31	Human NSHL	DFNB84			42	12779
<i>RARA</i>	17q21	Mouse				11	5734
<i>RARB</i>	3p24	Mouse				9	4370
<i>RARG</i>	12q13	Mouse				11	4772
<i>RASA1</i>	5q13.3	Mouse				25	8015
<i>RDX</i>	11q22.3	Human NSHL	DFNB24			18	7460
<i>S1PR2</i>	19p13.2	Mouse				2	2725
<i>SCARB2</i>	4q21.1	Mouse				12	5934
<i>SCRIB</i>	8q24.3	Mouse				33	9525
<i>SERPINB6</i>	6p25.2	Human NSHL	DFNB91			8	4851
<i>SIX1</i>	14q23.1	Human NSHL, SHL, mouse		DFNA23	Branchio-oto-renal, BOS3	2	2883
<i>SIX5</i>	19q13.32	Human SHL			Branchio-oto-renal, BOR2	3	3444
<i>SLC12A2</i>	5q23.3	Mouse				27	12237
<i>SLC12A6</i>	15q13	Mouse				28	11471
<i>SLC12A7</i>	5p15	Mouse				24	8399
<i>SLC17A8</i>	12q23.1	Human NSHL		DFNA25		12	4895
<i>SLC19A2</i>	1q23.3	Mouse				6	4327
<i>SLC1A3</i>	5p13	Mouse				10	5931
<i>SLC26A4</i>	7q22.3	Human SHL			Pendred, PDS; Enlarged vestibular aqueduct	21	8267
<i>SLC26A5</i>	7q22.1	Human NSHL	DFNB61			21	5500
<i>SLC30A4</i>	15q21.1	Mouse				8	4690
<i>SLC4A11</i>	20p12	Mouse				21	6037
<i>SLC4A7</i>	3p22	Mouse				26	11751
<i>SLC9A1</i>	1p36.1	Mouse				12	6469
<i>SMPX</i>	Xp22.1	Human NSHL		DFNX4		6	1645

<i>SMS</i>	Xp22.1	Mouse				11	3214
<i>SNAI2</i>	8q11	Human SHL, mouse			Waardenburg, WS2D	3	2405
<i>SOBP</i>	6q21	Mouse				7	8254
<i>SOD1</i>	21q22.11	Mouse				5	2207
<i>SOX10</i>	22q13.1	Human SHL, mouse			Waardenburg, WS4, WS2E; PCWH syndrome	5	3805
<i>SOX2</i>	3q26.3	Mouse				1	2603
<i>SOX9</i>	17q24.3	Mouse				3	3807
<i>SPRY2</i>	13q31.1	Mouse				2	3444
<i>ST3GAL5</i>	2p11.2	Mouse				8	4050
<i>STRC</i>	15q15.3	Human NSHL	DFNB16			29	9526
<i>SYNE4</i>	19q13.12	Human NSHL	DFNB76			9	2488
<i>SYNJ2</i>	6q25.3	Mouse				27	12621
<i>TBL1X</i>	Xp22.3	Human NSHL (late onset)				18	8650
<i>TBX1</i>	22q11.21	Mouse				11	3171
<i>TBX10</i>	11q13.2	Mouse				7	2488
<i>TCOF1</i>	5q32	Human SHL			Treacher Collins, TCOF1	28	9199
<i>TECTA</i>	11q23.3	Human NSHL	DFNB21	DFNA8/12		23	9823
<i>TFB1M</i>	6q25.1-q25.3	Suspected				7	3648
<i>TGFA</i>	2p13	Mouse				6	5726
<i>TGFB2</i>	1q41	Mouse				8	10780
<i>THRA</i>	17q11.2	Mouse				11	4333
<i>THRB</i>	3p24.2	Mouse				12	8175
<i>TIMM8A</i>	Xq22.1	Human SHL			Deafness, dystonia and additional features; Deafness and Dystonia- Optic Neuronopathy Syndrome; Jensen syndrome; MOHR- TRANEBJAERG syndrome	3	2274
<i>TJP2</i>	9q21.11	Human NSHL		DFNA51		25	11127
<i>TMC1</i>	9q21.13	Human NSHL	DFNB7	DFNA36		24	6079
<i>TMIE</i>	3p21.31	Human NSHL	DFNB6			4	2204
<i>TMPRSS3</i>	21q22.3	Human NSHL	DFNB8/10			13	5173

<i>TMPRSS5</i>	11q23.2	Human NSHL, suggested			13	3933
<i>TNFRSF11B</i>	8q24	Mouse			5	3126
<i>TPRN</i>	9q34.3	Human NSHL	DFNB79		3	2313
<i>TRIOBP</i>	22q13.1	Human NSHL	DFNB28		32	14731
<i>TRPV4</i>	12q24.1	Mouse			16	5417
<i>TSHR</i>	14q31	Mouse			11	6331
<i>TUB</i>	11p15.5	Mouse			15	8258
<i>TYRP1</i>	9p23	Mouse			8	3968
<i>UCN</i>	2p23	Mouse			2	843
<i>USH1C</i>	11p15.1	Human NSHL, SHL	DFNB18	Usher, USH1C	28	7396
<i>USH1G</i>	17q25.1	Human SHL, mouse		Usher, USH1G	3	3524
<i>USH2A</i>	1q41	Human SHL, mouse		Usher, USH2A	72	29525
<i>VANGL2</i>	1q22	Mouse			9	5970
<i>WFS1</i>	4p16.1	Human NSHL, SHL	DFNA6/14/38	Wolfram syndrome; Wolfram-like syndrome	9	5168
<i>XPA</i>	99q22.3	Human SHL		Xeroderma pigmentosum	7	2767
<i>XPC</i>	3p25	Human SHL		Xeroderma pigmentosum	16	5870
					SUM:	4421 1,981,479

<sup>A</sup>NSHL, non-syndromic hearing loss; SHL, syndromic hearing loss