Table 1. Summary of syndromes associated with hearing loss described in this review.

	Inheritance pattern	Medical conditions in addition to hearing loss	Genes involved
Usher (Types IB-G, Type IIA-D, Type IIIA)	Autosomal recessive	Vision loss due to retinitis pigmentosa, vestibular function is affected in type I, vestibular function is not affected in type II type and vestibular function is variable in type III	Type I: *MYO7A*, *Harmonin (PDZ73), CDH23, PCDH15, SAN.* Type II*: Usherin, GPR98,* WHRN Type III: *Clarin-1*
Pendred	Autosomal recessive	goiter, enlarged vestibular aqueduct with or without the presence of cochlear hypoplasia	*SLC26A4,FOXI1*
Branchio-oto-renal	Autosomal dominant	Involvement of the second branchial arch with branchial cleft cysts, cervical fistulas and renal abnormalities	*EYA1, SIX1, and SIX5*
Waardenburg (Types I-IV)	Autosomal dominant	Pigmentary changes of hair and eyes, dystopia canthorum (wide spacing between eyes), limb abnormalities	*PAX3, MITF, SNAI2 (SLUG) END3, SOX10, EDNRB*
Jervell and Lange-Nielsen	Autosomal recessive	Arrythmia, cardiac conduction defects	*KCNQ1, KCNE1*
Townes-Brocks	Autosomal dominant	Imperforate anus, irregularly shaped ears, thumb malformations	*SALL1*