Deafness and Hereditary Hearing Loss Encyclopedia Article

Deafness and Hereditary Hearing Loss

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Deafness and Hereditary Hearing Loss

Genetic forms of hearing loss can be due to malformations of the inner ear or from conductive nerve damage (sensorineural hearing loss). Hearing loss may be congenital (shown at birth) or delayed in onset, progressive or nonprogressive, unilateral or bilateral. It may be part of a syndrome (involving identifiable physical characteristics in other systems) or nonsyndromic (hearing loss only).

Approximately 75% to 80% of genetic hearing loss is attributed to autosomal recessive genes, 18% to 20% to autosomal dominant genes and the remainder are X- linked or chromosomal disorders.

At least 100 genetic syndromes that involve hearing loss have been identified. They may be classified on the basis of other organ systems involved: craniofacial/cervical (head and neck), skeletal, integumentary (involving skin), ocular (eyes), neurologic (nerve), renal (kidney), metabolic, and other syndromes.

The following three disorders have been studied extensively and have had their gene locations mapped. Usher's syndrome is an autosomal recessive disorder of sensorineural hearing loss (SNHL) and retinitis pigmentosa (vision loss due to a gradual deterioration of the light sensitive cells of the retina). The syndrome has two variants. Patients with the first variant have normal vestibular function with a progressive SNHL, while those with the second are born with no labyrinth canal and deafness.,

Waardenburg's syndrome (also known as Klein-Waardenburg syndrome) is an autosomal dominant disorder with typical phenotypic characteristics of a white forelock, patchy depigmentation of the skin, dystopia canthorum (malformation of the inner fold of the eyelid) and irises with more than one colour or a pale colour (heterochomia of irides). It also has two recognized variants, both with SNHL.

Hereditary deafness has also been associated with a subtype of diabetes (both Insulin Dependent and Non-Insulin Dependent Diabetes). This rare syndrome is caused by a single **mutation** in **mitochondrial DNA**. Brcause mitochondrial **DNA** is inherited maternally, this syndrome has been named Maternally Inherited Diabetes and Deafness (MIDD) and affects approximately 1.3% of all diabetic individuals.

Classifications of nonsyndromic hearing impairments (hearing loss only) are generally based on audiologic characteristics, presence or absence of progression, age of onset and mode of **inheritance**. At least 40 chromosomal locations have been mapped for nonsyndromic deafness. Within these locations, five genes have been identified to date. The identification of the specific genes responsible for hearing loss may assist with genetic counselling for suspected carriers and in the future may direct **gene therapy** for the prevention or treatment of hereditary deafness.