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Hearing Loss: Acquired or Genetic?

JAY WILEY

GENETIC TRAIT: Hearing loss can be an inherited condition.

While acquired hearing loss associated with age or noise exposure is more common than genetic hearing loss, congenital (inborn, inherent) deafness occurs in one of every 1,000 to 2,000 births. Autosomal recessive inheritance is the most common form, accounting for more than 75 percent of all congenital deafness.[1]

Of the 50 percent of the genetic forms of hearing loss, an estimated 70 percent are due to recessive causes, about 15 percent have a dominant cause, and the remaining 15 percent include all the other forms of inheritance.[2]

Genetic scientists classify genetic hearing loss into two general categories:

- non-syndromic (hearing loss only);
- syndromic (hearing loss with other clinical symptoms).

Two-thirds of all genetic hearing losses are non-syndromic. A single gene, known as Connexin 26 (abbreviated CX26), is reported to be responsible for half of all the recessive cases of hearing loss experienced by people (a recessive gene elicits a phenotype in a homozygous organism). While there are more than 400 known genetic causes involving hearing loss, aberrations in CX26 are responsible for an estimated one-third of all the cases of genetic hearing loss.[2]

The Importance of Genetics

Genetics is a branch of science that studies the process and product of biological inheritance. Genomics investigates the process and product of how genes interact with each other and the environment.

A trait, or phentotype, is a distinguishing characteristic, feature or quality exhibited by a person. People inherit genetic information and, therefore, traits from their parents. For example, a hearing loss from an ancestor may be passed to his or her descendent. However, genetic information alone does not fully describe how diseases impact people. A better explanation is offered by understanding the impact of both genetics and the environment.

Mutations in the CX26 gene are a relatively common genetic cause of hearing loss and follow an autosomal recessive pattern of inheritance. Autosomal means that the mutations occur on chromosomes other than the sex chromosomes X and Y.

Aminoglycoside-induced hearing loss is an example of how both genetics and the environment interact to produce hearing loss. It is most likely to occur with people who take aminoglycoside antibiotics and who carry a gene that is susceptible to interacting with this antibiotic.[1]

Inheriting genes that place a person at risk for hearing loss does not ensure that hearing loss will occur. For example, if a person never takes an aminoglycoside antibiotic, then being susceptible to the antibiotic's impact on the at-risk genes is of no consequence. Additionally, a person's genome may be impacted differently depending upon age and health.

more info

American Speech-Language Hearing Association
www.asha.org/default.htm

Boys Town National Research Hospital
www.babyhearing.org/HearingAmplification/Causes/genetics.asp

Genetic Counseling

People who have inherited genes that place them at risk for hearing loss have an option to seek genetic counseling. Genetic counselors have specialized graduate degrees and experience in the areas of medical genetics and counseling. They identify families at risk, investigate the problem present in the family, interpret information about the disorder, analyze inheritance patterns and risks of recurrence, and review available options with the family.[4]

While not as common as acquired hearing loss, genetic hearing loss remains an important reason why people do not hear normally. Genetic counselors are available to provide information to help these people make quality-of-life decisions.

REFERENCES

1. American Hearing Research Foundation: www.american-hearing.org/disorders/hearing/cong_hearing.html
2. Boys Town National Research Hospital: www.babyhearing.org/HearingAmplification/Causes/genetics.asp
3. American Speech-Language Hearing Association: www.asha.org/default.htm
4. National Society of Genetic Counselors: www.nsgc.org

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