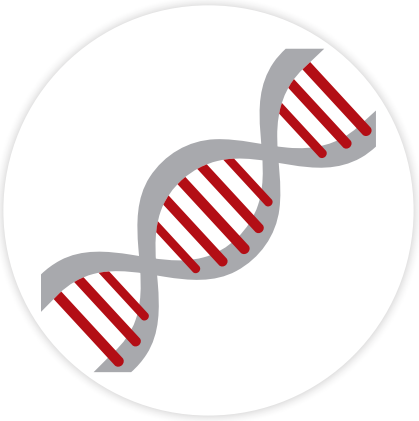
Understanding Genetic and Progressive Hearing Loss



In adults and children, loss of hearing can sometimes progressively worsen at a much faster rate than noise-induced or age-related hearing impairment.

These symptoms usually indicate a specific type of hearing loss, referred to as progressive hearing loss, which can stem from a variety of both detectable and unknown diseases. About one in 500 infants are born with or develop a hearing loss throughout early childhood.

If a patient of yours is concerned that their child is experiencing spinning, nausea, headaches, or hearing loss, they may be suffering from a genetic or progressive hearing loss. Our office provides comprehensive solutions that can help keep our community healthy and happy. Thank you for working with us to improve the quality of life in our local community!

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AUDIOLOGY, LLC

SOUND information brought to you
by your local audiology professionals

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Understanding Genetic and Progressive Hearing Loss

About one in 500 infants are born with or develop a hearing loss throughout early childhood. When children are born with a hearing impairment, this is considered "congenital," while the development of an impairment after birth is referred to as "acquired." There are many potential causes; for example, German measles, syphilis, and diabetes present a high risk to unborn children. But the most prominent causes are:

- A genetic disease passed down from parents
- An infection that affected the fetus during gestation or shortly after birth
- Exposure to an ototoxin as a fetus or infant

For some children, a cause may never be identified. A child can have normal hearing as an infant and may proceed to lose their hearing after one or two years of life; this type of "delayed onset" hearing loss usually happens when a child is born with a virus that no one is aware of until later. A child may also develop hearing loss from an ototoxic medicine used when the child is sick, and it may continue to worsen even after the child stops taking the identified medication.

Risk factors for a genetic or progressive hearing disorder include:

- A family history of hearing loss
- Premature birth, which is sometimes responsible for underdeveloped lungs that necessitate the use of a ventilator; excessive ventilator use is associated with lung infections that can spread to the ear canal
- Physical malformations of the head or ears
- Infections during pregnancy or birth

A newborn with known risk factors for progressive hearing loss should have their hearing tested prior to age 3, in case a previously undetectable hearing loss may have emerged since birth. Children born with the cytomegalovirus (CMV), for example, face a significantly higher risk of developing progressive hearing loss. Urine samples testing for CMV can serve as a warning sign to parents and caregivers. If the child doesn't seem to respond to the noises they used to, or if their speech patterns change, they may be experiencing a progressive hearing loss.

Over the course of several months, a bilateral progressive hearing loss can occur and can be diagnosed as autoimmune inner-ear disease, which happens when the body's immune system misdirects its defenses against the inner-ear structures to cause damage to this part of the body. This disease is typically managed with long-term corticosteroids and other drug therapy. Other illness, like meningitis, makes a present hearing loss more likely to get worse over time.

There are two developmental conditions, genetic in nature, that may cause a wide range of hearing impairment:

- Waardenburg syndrome is associated with hearing loss that ranges anywhere from mild to total deafness, and is characterized by a wide spacing between the eyes, a broad nose and bridge, and connecting eyebrows. This hearing loss is present from birth.
- Crouzon syndrome causes the plates of the skull to fuse together, preventing the brain from growing. Facial deformities, particularly of the inner ear, are common and may result in no ear canal whatsoever.

On occasion, progressive hearing loss is correctable through surgery. Cholesteatoma and otosclerosis are two causes of progressive hearing loss in which patients can see a total or near-total recovery of their hearing.

- Cholesteatoma is a disease of the ear in which a noncancerous skin cyst grows into the middle ear and the mastoid, eroding tissue and causing destruction of the ear. This could be caused by improper function of the Eustachian tube. Cholesteatomas can take years to form and are common in individuals with a history of middle-ear fluid and infections.
- Otosclerosis is a disorder of the stapes, a tiny bone in the middle ear that helps conduct sound to the inner ear. This is a genetic development problem, with about 50 percent of all patients showing a family history of otosclerosis. During surgery, the stapes is removed and replaced by a prosthesis to restore function.

For most other forms of progressive hearing loss, hearing aids are a viable option. If the patient's loss reaches a level where use of hearing aids no longer helps, a cochlear implant or a direct bone-conduction system are two advanced hearing solutions that should be considered to help patients hear their best.

Hearing loss is associated with twice as many visits to the emergency room, a tenfold increase in the risk of being held back at least one grade in school, and reduced earning potential as an adult. Many forms of progressive hearing loss cannot be reversed, but they can be treated through use of medication, surgery, and advanced better-hearing technology. By creating solutions, healthcare providers and hearing specialists can partner to create a healthier local community for hearing loss sufferers of all ages. ■

Centers for Disease Control and Prevention. A Parent's Guide to Genetics & Hearing Loss. U.S. Department of Health and Human Services.

Holt, J. Cholesteatoma and Otosclerosis: Two Slowly Progressive Causes of Hearing Loss Treatable Through Corrective Surgery. Clinical Medicine and Research. April 2003.

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