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Usher Syndrome

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Usher syndrome is a genetic disorder that affects hearing, sight, and balance. This condition is named after Charles Usher, the optometrist that first documented the disease. It is rare, occurring in only 4 to 17 out of 100,000 people.

Usher syndrome causes sensorineural hearing loss, which is a type of hearing loss generated in the inner ear. Later in life, retinitis pigmentosa (RP) occurs, which means the light sensitive cells in the back of the eye start to deteriorate. Typically this occurs 5-10 years after the onset of hearing loss. RP causes night blindness (the inability to see in the dark), then tunnel vision, and sometimes full blindness. The vestibular system, which is located in the inner ear and controls balance, is also negatively impacted.

There are three types of Usher syndrome with various subtypes. They all have different genetic origins and presentations:

Type I

Severe to profound hearing loss is present at birth. Night blindness starts within the first 10 years of life. Balance is also impacted, which may cause delays in sitting up and walking.

Type II

Hearing loss ranging from mild to severe is present at birth, especially impacting high frequency sounds. Many of those high frequency sounds, such as 's' and 'th,' occur in speech, and this impacts an individual's ability to hear conversations. However, the vestibular system remains intact, and balance develops typically. Night blindness begins in adulthood.

Type III

Hearing and vision are normal at birth. The onset of hearing loss begins around adolescence or early adulthood and progresses to a profound hearing loss in middle-age. Loss of vision also begins around adolescence and early adulthood, and individuals occasionally suffer from deficits in the vestibular system.

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Although there is no cure for Usher syndrome, there are many treatments and strategies offered by a team of professionals, including audiologists, low vision specialists, speech-language pathologists, counselors, and more. These strategies are implemented to improve an individual's quality of life and communication. Modifications can be made to the school and work environment to allow people with Usher syndrome to participate.

Depending on when hearing loss starts, a speech-language pathologist may recommend different modalities of communication. For someone who has hearing loss at birth, American Sign Language (ASL) may be recommended as a primary mode of communication. For someone with a later-onset hearing loss, a speech-language pathologist may develop strategies for that individual to use residual hearing and then transition to another mode of communication as hearing diminishes.

Since Usher syndrome causes sensorineural hearing loss, some individuals are candidates for cochlear implants, which are electronic devices surgically implanted to restore hearing. A speech-language pathologist can assist with aural rehabilitation, which helps a person with a hearing aid or cochlear implant perceive and interpret sound. Some individuals are also taught braille so that they preserve the ability to read after the loss of sight.

People with Usher syndrome and their families have many online resources offering information and support. Here are a few:

The Usher Syndrome Coalition

Hellen Keller Center for Deaf-Blind Youths & Adults

National Center on Deaf-Blindness

Resources:

"Aural Rehabilitation for Adults," accessed April 13, 2020, from https://hearinghealthfoundation.org/usher-syndrome-treatments

"Treatment for Usher Syndrome," accessed April 13, 2020, from https://hearinghealthfoundation.org/usher-syndrome-treatments

"Understanding Usher Syndrome," accessed April 13, 2020, from https://www.asha.org/content.aspx?id=8589967442

"Usher syndrome," accessed April 13, 2020, from https://ghr.nlm.nih.gov/condition/usher-syndrome#resources

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