



Usher Syndrome & Deaf-blindness

Usher Syndrome is an inherited condition that affects a person’s hearing and vision. It is the most common cause of Deaf-blindness. People with Usher Syndrome have vision loss caused by Retinitis pigmentosa (RP) that slowly causes tunnel vision. Tunnel vision can be like looking through a straw and in some cases causes total loss of vision. RP affects a person’s night vision first. Many people with Usher Syndrome also have a hard time with their balance.

Types of Usher Syndrome

Usher Type 1 – Children are born profoundly deaf and have problems with their balance. This means it takes longer for them to learn to sit up and walk. Children with Usher Type 1 have vision problems (limited vision and difficulty seeing at night) that start before they are 12 years old. Their vision loss begins with difficulty seeing at night and soon thereafter, then they start losing their peripheral vision and may become completely blind.

Usher Type 2- These children are born hard of hearing, with moderate or severe hearing losses. These children DO NOT usually have a hard time with balance. They experience slow vision loss in their teenage years, beginning first with loss of vision at night.

Usher Type 3 – Children are born with normal hearing and usually have normal balance. During puberty, they have progressive vision loss and hearing loss. By the time the person reaches adult they are most often legally blind.

Descriptions of Usher Syndrome Type 1, Type 2, & Type 3

Comparisons	Usher Type 1	Usher Type 2	Usher Type 3
Hearing	At birth, profoundly deaf	At birth, hard of hearing progressing to profoundly deaf in teenage years	At birth, normal hearing progressing to hearing loss in teenage years
Vision	By age 12, limited vision & difficulty seeing at night. Can progress to total blindness	Slow vision loss may begin in teenage years. Can stabilize or can progress to pin-hole vision in early adulthood	Vision loss begins in teenage years & progresses to legal blindness in adulthood
Balance	At birth, difficulties with balance that continue throughout life	Usually no balance problems	No balance problems

Important ideas for working with Deaf-blind students who have Usher Syndrome

1. Self-Advocacy/individual preferences: Always ask the person first what is preferred in terms of seating, lighting, colors and materials. Support student to initiate asking for what accommodations and adaptations he/she needs. For example, more time on tests, seating and materials preferences.
2. Lighting: Communication should be in full light without glare, teacher/communicator should not stand in front of windows; windows should be behind student.
3. Role Models: Find a local Role Model(s) with Usher Syndrome to connect with your student and his/her family.
4. Sign Language: Consider learning sign language (such as local sign language or American Sign Language-ASL). Make sure the size of ASL being presented is understandable (based on the student's vision loss). Consider the use of a sign language interpreter for students that have adequate vision and tactile signing when vision is more limited and the student cannot benefit from visual signs.
5. Orientation & Mobility: Assess potential needs of the student and begin O&M training when students still have usable vision.

Adapted from:

<http://www.nidcd.nih.gov/health/hearing/pages/usher.aspx>

<http://www.unr.edu/ndsip/tipsheets/usher.pdf>



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