

# USHER SYNDROME

## What is Usher Syndrome?

- A rare autosomal recessive genetic disorder caused by a mutated (i.e. changed) gene
- Autosomal recessive means it affects both males and females equally and each parent carries the gene
- Leading cause of deaf-blindness in the world
- Characterized by progressive hearing loss, progressive vision loss until blindness, potential mobility and balance difficulties, possible speech and language differences.
- Retinitis pigmentosa (RP) is progressive vision loss that has variable onset and progression rates with this condition.

## Three Types of Usher Syndrome

- Type I: Child is born with sensorineural hearing loss (damaged ear nerves) and have severe balance problems. They typically do not respond to hearing aids. Visual loss usually occurs around age 10 with night blindness and progresses to total blindness.
- Type II: Child is born with sensorineural hearing loss and respond well to hearing aids. They have normal balance and a later onset of visual difficulties (adolescence).
- Type III: Born with normal hearing, then progressive hearing loss starts in early teens likely becoming profound hearing loss in their 40's. Vision loss starts as night blindness during puberty.

## Consequences of Communication Issues

- Lack of understanding in both hearing and deaf communities
- Limited ability to communicate with family, friends, and groups of people which may result in social isolation and depression.



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## Role of the Speech-Language Pathologist (SLP)

- Identify how the child communicates
- Introduce different communication methods that might better fit the child's future needs
- Work with other professionals on the child's team (e.g. physical therapist, occupational therapist, audiologist, educator of the deaf, teacher of the visually impaired, and special education teacher)
- Educate family and team members about ways to communicate with the child
- Audiologist may suggest hearing aids/cochlear implants
- Early intervention is KEY

## Classroom Modifications

- Use appropriate lighting and have contrasting colors
- Give the student time to visit and adjust to new buildings
- Release the student early to make exiting the building easier
- Assign a classroom buddy as a guide
- Tell the student about furniture changes in the classroom

## Modes of Communication

A Total Communication approach (i.e. encouragement of any communication technique) is essential

- Spoken language
- Sign language or tactile sign language
- Braille or enlarged print
- Assistive hearing devices
- Assistive vision devices

For more information about Usher Syndrome please visit:

National Institute on Deafness and Other Communication Disorders: <http://www.nidcd.nih.gov/>

American Speech-Language-Hearing Association: [www.asha.org](http://www.asha.org)



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