

**PATTI MCGOWAN:** Hello. My name is Patti McGowan and I am one of the two family liaisons with the Pennsylvania Deaf-Blind Initiative of PaTTAN. I am also a parent to a son with deaf-blindness due to Usher syndrome type II. Usher syndrome is one of many etiologies of deaf-blindness. Today, I'd like to share some information with you on Usher syndrome.

The definition of Usher syndrome means a child is born with deafness or hearing impairment, and then will lose vision due to the eye disease retinitis pigmentosa, or known as RP, which is a slow, progressive disease of the retina. Both conditions must be present and together, and are caused by a single recessive mutated gene carried by both parents. There are three types of the syndrome. Type I is characterized by a profound congenital hearing loss, poor balance, and retinitis pigmentosa before the age of ten. Type II presents moderate to severe hearing loss, normal balance, and retinitis pigmentosa develops in the late teens or early 20s. Type III is characterized by progressive hearing loss and retinitis pigmentosa progressing at a verifiable rate, generally with onset around the second or third decade of life.

Usher syndrome was named after a British ophthalmologist, C.H. Usher, in 1914. He'd described how there was a link between congenital deafness and retinitis pigmentosa. Typically, the first signs of Usher syndrome are night blindness. Some symptoms of night blindness are: children cannot see when coming in from bright sunlight, tripping over things in dim light, and staying near a light in a dark room or at night. Yet another symptom of Usher's is being glare-sensitive, or squinting and shading eyes in bright or fluorescent lighting, or appearing awkward when exiting from the inside of the building to the outside and to bright lighting. The general symptoms of Usher syndrome can be summarized as night blindness, peripheral vision loss, difficulties adapting to lighting changes, blind spots, glare discomfort, balance problems, and acuity problems.

Usher syndromes is diagnosed by a series of tests including visual function test, the visual fields, and the electroretinogram, which detects retinitis pigmentosa. They also base results on the retinal examination, hearing test, and balance test for all patients around the age of ten and up -- older. Deaf-blindness is a combined vision and hearing loss that impacts an individual's learning, communication, and interaction with the world. The primary causes of Usher syndrome are hereditary disorders.

Children with Usher syndrome may be withdrawn, preoccupied with their bodies, or inwardly focused. Having Usher's syndrome compromises communication with others, mobility is greatly limited, it is complicated to obtain information, and their independence is threatened in school for children

suffering from Usher syndrome -- can sometimes be a challenge for more reasons than their learning abilities.

As individuals with this condition experience a loss of vision, they may encounter mobility difficulties walking through indoor and outdoor areas, including tripping, falling, bumping, and loss of balance. To help adjust with these vision changes, teachers will often show children how to be -- how to use scanning techniques to help adjust to the classroom. By looking to the right and to the left in the classroom, it will help adjust to the class as well as keep the child involved with communication.

As a general rule, tactile sign language should be introduced to students with Usher's syndrome at the earliest age opportunity. Along with tactile sign language, relevant instruction is taught at an early age to the best learning opportunity to children with Usher's. Orientation and mobility should also be taught to the child. This will not only be important for safety reasons, but for their independence.

Children with Usher syndrome need to learn to describe visual and auditory and specific support needs to others: their self-advocacy. This will help the child become independent. Using scanning techniques, visual tracking, sign language, tactile sign language, oral communication, using an assertive listening device, using interpreters, using appropriate lighting, assistive technology, and a quiet learning environment are just a few ways to help a child cope with the school setting while also dealing with the problems that come along with Usher's syndrome.

There are many modifications that can help a student with Usher's in the classroom environment. Perhaps when lecturing, teachers should stand in the students' line of vision or stand near the students to best fit their needs of seeing and hearing a teacher. A visually cluttered classroom and unseen movement should be avoided. Because bright lighting bothers children with Usher's syndrome, the student should always be seated in front of windows, never facing them. Some students should be allowed to even wear a ball cap inside the school building to help with glare or nuisance lights.

Students in the classroom should be seated comfortably, and the setting should provide easy movement through the classroom so the student cannot get hurt due to mobility issues. Contrasts will be needed on the chalkboard or whiteboard. Test, homework, and worksheets may need to be adapted, and font may need to be made larger. Many students use a CCTV or a closed-circuit television. This CCTV helps enlarge prints on a worksheet and reading materials. Educational teams and families should consider requesting an assistive technology evaluation for the student. Teachers should be made aware of any vision or hearing changes because this can decrease academic performance, reduce participating

in class, and cause isolation from classmates and group activities. Students should learn in the classroom to communicate their needs and to self-advocate as this will continue their independence.

As children get older, Usher's syndrome can get progressively worse. Unfortunately, the condition of retinitis pigmentosa can change rapidly, unpredictably, and sustainably. Both families and schools and the school teams help these children cope with Usher syndrome. Families and teachers can do many things at home and in the classroom to help make the child feel comfortable in their environment. Families will always be there to help children in the real world. Having Usher syndrome does not stop children from having a good education and fulfilling their goals in life.