What is Usher Syndrome?

A **syndrome** is a condition which has at least two characteristics or symptoms. One such syndrome that causes deaf-blindness is **Usher Syndrome**, affecting half (50%) of the deaf-blind population. Usher Syndrome is a hereditary condition that results in hearing loss and progressive vision loss due to **retinitis pigmentosa (RP)**. Symptoms of RP include: night blindness, peripheral (side) vision loss, and eventual central vision loss. Hearing loss of varying degrees is usually present at birth, and the onset of vision loss varies with each individual. Usher Syndrome is inherited when each parent carries a recessive gene that is passed on to the child who eventually is faced with the condition. There are three general types of Usher Syndrome.

Symptom	Type I	Type II	Type III
Hearing Loss	Born deaf with profound hearing loss	Born hard of hearing with a sloping sensorineural loss from mild loss in low frequencies to severe-profound loss in high frequencies	Born with good hearing or mild hearing loss which gets worse over a decade or more
			Looks like Type II in teenagers and young adults; looks like Type I in older people
Balance	Absent inner ear balance	Normal inner ear balance	Progressive balance disturbance
Vision Loss from RP	Night blindness in infancy or early childhood	Night blindness begins in the teens	Night blindness in childhood or teens
	Tunnel vision usually by age 16	Tunnel vision in the late teens to early 20s	Tunnel vision becomes pronounced in the 20s

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For more information, go to www.ncdhhs.gov/dsdhh or call 800-851-6099 (V/TTY), 919-874-2212 (V/TTY) or 919-890-0859 (VP).

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