What is Usher Syndrome?
Usher syndrome is the most common condition that affects both hearing and vision.

Who is affected?
In the US, 4 in every 100,000 births have Usher Syndrome.
• Approximately 3-6% of all children who are deaf and another 3-6% who are hard of hearing have Usher syndrome.

Three clinical types of Usher Syndrome
Type 1: profoundly deaf and have severe balance problems, and do not benefit from hearing aids. They develop vision problems in early childhood (by the time they reach ten) and begin with difficulty seeing at night, but tend to progress rapidly until they are completely blind.
Type 2: Born with moderate to severe hearing loss & normal balance, but can benefit from hearing aids. The visual problems progress more slowly than in Type 1 with retinitis pigmentosa not apparent until the teens.
Type 3: Normal hearing at birth, but worsen over time. Type 3 develop hearing loss by the teens blind spots appear in the late teens to early adulthood and by mid adulthood the individual is legally blind.

Symptoms
- Hearing loss
- Eye disorder called retinitis pigmentosa, RP

Treatment
There is no cure for Usher syndrome. The best treatment is early identification so that educational programs can begin as soon as possible.

What causes Usher syndrome?
Usher syndrome is inherited as an autosomal recessive trait.

References