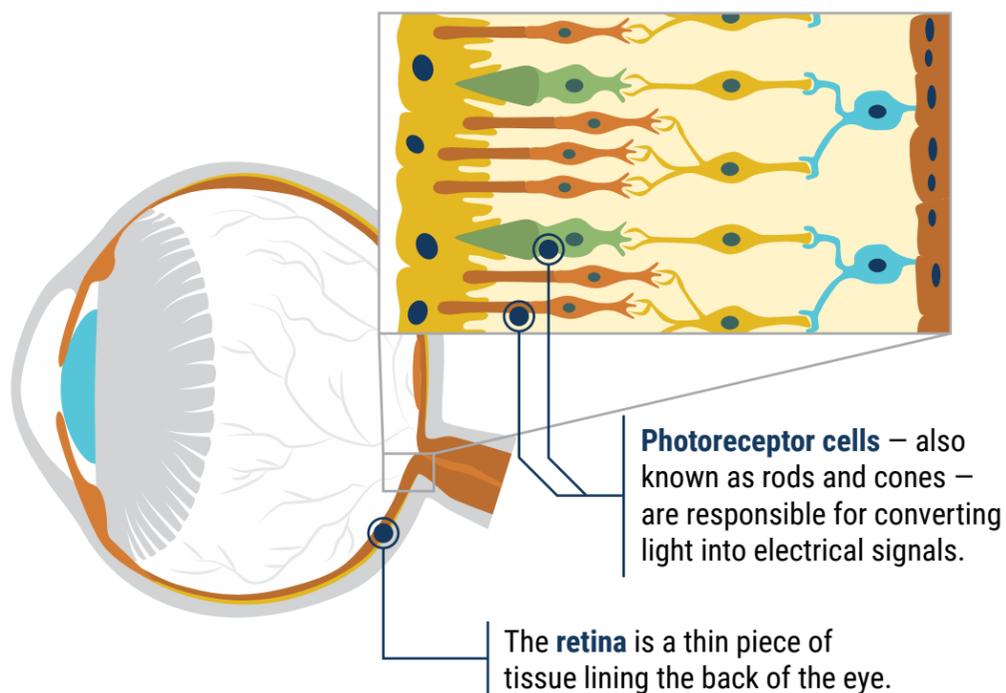


USHER SYNDROME

WHAT IS USHER SYNDROME?

Usher syndrome is an inherited disease causing combined hearing loss and vision loss. The vision loss is due to retinitis pigmentosa (RP), a degenerative condition of the retina.

Researchers estimate that as many as **25,000 people in the U.S.** have Usher syndrome. Worldwide, it is the leading cause of combined deafness and blindness. About **30 percent** of people with RP report some degree of hearing loss, and about half of them are diagnosed with Usher syndrome.



HOW IS USHER SYNDROME INHERITED?

Usher syndrome is autosomal recessive. That means it is **genetically passed** when both parents (unaffected carriers) have one gene for the syndrome paired with one normal gene.



There is a **25% chance** of their child inheriting the two Usher syndrome genes.



GENETIC TESTING

Genetic testing helps people definitively diagnose their condition and the risk of other family members or future offspring being affected. A genetic diagnosis can also help a person qualify for a clinical trial. Genetic counselors are excellent resources for discussing inheritability, family planning, genetic testing, and other related issues.



Hearing loss in Usher syndrome occurs, because the gene mutations affecting the retina also affect the cochlea, a sound-transmitting structure of the inner ear.

WHAT TO EXPECT WITH USHER SYNDROME:

THINGS TO KNOW:



Effects both vision and hearing.



Often diagnosed in childhood or adolescence.



Can also cause problems with balance.



Progression varies from person to person.

THERE ARE THREE GENERAL CATEGORIES OF USHER SYNDROME:

Type 1 (USH1):

- Usually born with severe hearing loss.
- Experience problems with balance.
- The first signs of RP usually appears in early adolescence.

Type 2 (USH2):

- Newborns have moderate to severe hearing impairment.
- Symptoms of RP typically start shortly after adolescence.
- Visual problems progress less rapidly than in Usher type 1.

Type 3 (USH3):

- Rarest form of Usher syndrome, first documented in 1995.
- Usually born with good or only mild impairment of hearing.
- Progressive hearing and vision loss, starting around puberty.