

LIVING WITH USHER SYNDROME

Usher syndrome affects between 3 and 6 out of every 100,000 people.

Usher syndrome is a rare genetic disorder that causes vision loss, hearing loss, and sometimes, balance issues. Symptoms are often present during birth or develop during childhood. Although there is no cure, there are many ways to manage the symptoms of Usher syndrome.

01 | Causes

Usher syndrome is an inherited disorder passed down from unaffected parents to their children. Genetic mutations can affect people in different ways. For example:

- Hearing loss happens when genetic mutations affect nerve cells in the cochlea of the inner ear, so nerve cells don't transmit sounds as they should.
- Progressive vision loss is caused by retinitis pigmentosa (RP). RP causes cells in the retina to break down, leading to a loss of night vision and, later, a narrowing of the visual field.

02 | Types

There are three clinical types of Usher syndrome. They are distinguished by the severity and age at which the signs and symptoms appear:

- **Type 1:** This type is characterized by profound hearing loss or deafness at birth and severe balance problems that cause trouble sitting or walking. Vision problems often begin before the age of 10, starting with a loss of night vision.
- **Type 2:** This type is characterized by moderate to severe hearing loss at birth, but no balance problems. Vision issues typically develop in the later teen years.

- **Type 3:** This type is characterized by normal hearing and vision at birth. People with type 3 begin to develop hearing loss in late childhood and vision loss in early to mid-adulthood. This is the rarest type of Usher syndrome.

03 | Treatment

Treatment for Usher syndrome is specific to each person. Some treatment options may include:

- Cochlear implants
- Hearing aids
- Vision aids like eyeglasses

For more information on Usher syndrome, please visit [usher-syndrome.org](https://www.usher-syndrome.org).

Did You Know?

Usher syndrome is the most common genetic cause of combined deafness and blindness.

References

<https://www.usher-syndrome.org/resources/>

<https://www.nei.nih.gov/eye-health-information/eye-conditions-and-diseases/usher-syndrome>

<https://my.clevelandclinic.org/health/diseases/15046-usher-syndrome>

<https://www.hopkinsmedicine.org/health/conditions-and-diseases/usher-syndrome>