Retinoblastoma Causes, Risk Factors, and Prevention

Risk Factors

A risk factor is anything that affects your chance of getting a disease such as cancer. Learn more about the risk factors for retinoblastoma

- What Are the Risk Factors for Retinoblastoma?
- Do We Know What Causes Retinoblastoma?

Prevention

In adults, the risk for many cancers can be reduced by avoiding certain risk factors, such as smoking. But there are no known avoidable risk factors for retinoblastoma. If your child does develop retinoblastoma, it’s important to realize that you or your child did nothing to cause it.

Some gene changes that put a child at high risk of retinoblastoma can be passed on from a parent. Children born to a parent with a history of retinoblastoma should be screened for this cancer starting shortly after birth because early detection of this cancer greatly improves the chance for successful treatment.

What Are the Risk Factors for Retinoblastoma?

A risk factor is anything that affects a person’s chance of getting a disease such as cancer. Different cancers have different risk factors.
Lifestyle-related risk factors such as body weight, physical activity, diet, and tobacco use play a major role in many adult cancers. But these factors usually take many years to influence cancer risk, and they are not thought to play much of a role in childhood cancers, including retinoblastomas.

There are very few known risk factors for retinoblastoma.

**Age**

Most children diagnosed with retinoblastoma are younger than 3 years old. Most congenital or hereditary retinoblastomas are found during the first year of life, while non-inherited retinoblastomas tend to be diagnosed in 1- and 2-year-olds. Retinoblastomas are rare in older children and in adults.

**Heredity**

About 1 out of 3 retinoblastomas is caused by a mutation (change) in the \(RB1\) gene that is present in all the cells of the child’s body. But of these cases, only about 1 in 4 is inherited from one of the child’s parents. In the rest, the gene mutation is not inherited, but occurs during early development in the womb. Children born with a mutation in the \(RB1\) gene usually develop retinoblastoma in both eyes. Regardless of whether the mutated \(RB1\) gene was inherited from a parent or not, because these children have the mutated gene in all of their cells, they have a 1 in 2 chance of eventually passing it on to their children.

Most of the remaining 2 out of 3 retinoblastomas occur as a result of a random \(RB1\) gene mutation that occurs only in one cell of one eye. These mutations are not inherited from a parent, and children who have them do not pass on a greatly increased risk of retinoblastoma to their children. Non-hereditary retinoblastomas always affect one eye only.

The way in which inherited gene changes make certain children likely to develop retinoblastoma is explained in the section [Do We Know What Causes Retinoblastoma?](#)

- **References**

[See all references for Retinoblastoma](#)

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Do We Know What Causes Retinoblastoma?

Retinoblastoma is caused by mutations (changes) in certain genes. Over the past few decades, scientists have learned how certain changes in a person’s DNA can cause cells of the retina to become cancerous. The DNA in each of our cells makes up our genes, which are the instructions for how our cells function. We usually look like our parents because they are the source of our DNA. But DNA affects much more than how we look.

Some genes control when our cells grow, divide into new cells, and die at the right time. Certain genes that help cells grow, divide, or stay alive are called oncogenes. Others that slow down cell division or cause cells to die at the right time are called tumor suppressor genes. Cancers can be caused by DNA changes that turn on oncogenes or turn off tumor suppressor genes.

The most important gene in retinoblastoma is the $RB1$ tumor suppressor gene. This gene makes a protein (pRb) that helps stop cells from growing too quickly. Each cell normally has 2 $RB1$ genes. As long as a retinal cell has at least one $RB1$ gene that works as it should, it will not form a retinoblastoma. But when both of the $RB1$ genes are mutated or missing, a cell can grow unchecked. This can lead to further gene changes, which in turn may cause cells to become cancerous.

Hereditary or bilateral retinoblastoma

About 1 out of 3 children with retinoblastoma have a germline mutation in one $RB1$ gene; that is, all the cells in the body have a defective $RB1$ gene. In most of these children (75%), this mutation developed after conception while in the womb. The other 25% of children have inherited it from one of their parents.

About 9 of 10 children who are born with this $RB1$ germline mutation develop retinoblastoma. This happens when the second $RB1$ gene is lost or mutated. Most often the retinoblastoma is bilateral (in both eyes), but sometimes it is found early enough that it is still only in one eye.

These children have hereditary retinoblastoma. All bilateral retinoblastomas are
considered hereditary, although not all hereditary retinoblastomas are bilateral when they are found.

Every person has 2 RB1 genes but passes only 1 on to each of their children. (The child gets the other RB1 gene from the other parent.) Therefore there is a 1 in 2 chance that a parent who had hereditary retinoblastoma will pass the mutated gene on to his or her child.

Most children with hereditary retinoblastoma don't have an affected parent. But these children can still pass their RB1 gene mutation on to their children. This is why we call this form of retinoblastoma “hereditary” (even though neither of the child's parents may have been affected).

Non-hereditary (sporadic) retinoblastoma

Most of the remaining 2 out of 3 children with retinoblastoma do not have the RB1 gene mutation in all the cells of their body. Instead, the RB1 mutation happens early in life and first occurs only in one cell in one eye.

Whether the changes in the RB1 gene are hereditary or sporadic, it’s not clear what causes these changes. They may result from random gene errors that sometimes occur when cells divide to make new cells. There are no known lifestyle-related or environmental causes of retinoblastoma, so it’s important to remember that there is nothing these children or their parents could have done to prevent these cancers.

- References
  See all references for Retinoblastoma

Can Retinoblastoma Be Prevented?

In adults, the risk for many cancers can be reduced by avoiding certain risk factors, such as smoking or exposure to hazardous chemicals in the workplace. But there are
no known avoidable risk factors for retinoblastoma. If your child does develop retinoblastoma, it’s important to realize that you or your child did nothing to cause it.

In some cases, gene changes that put a person at high risk of retinoblastoma are hereditary (passed on from a parent to a child). People who have had retinoblastoma might want to consider genetic counseling before having children to learn more about the risks of passing on the gene change and perhaps to explore ways to avoid this. For example, an option some people might consider would be to use in vitro fertilization and implant only embryos that don’t have the gene change.

If a preventive option is not used, children born to a parent with a history of retinoblastoma should be screened for this cancer starting shortly after birth because early detection of this cancer greatly improves the chance for successful treatment. See the section Can Retinoblastoma Be Found Early? for more information.

- References
  See all references for Retinoblastoma

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