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## If Your Child Has Retinoblastoma

- [What is retinoblastoma?](#)
  - [Different types of retinoblastoma](#)
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Retinoblastoma starts in the cells in the retina. This cancer almost always starts because of a change (mutation) inside the cells in a gene called *RB1*. The *RB1* gene change leads to the cells growing out of control.

and are also at higher risk for some other types of cancer.

In the **non-heritable (non-hereditary)** form, the changes in the *RB1* gene are only in the cancer cells. These children develop tumors in only one eye, and they are not at higher risk for other types of cancer.

### Questions to ask the doctor

- How sure are you that my child has retinoblastoma?
- Is there a chance it's not retinoblastoma?
- Would you please write down the kind of tumor you think my child has?
- What will happen next?

### How does the doctor know my child has retinoblastoma?

Retinoblastoma is often found when it causes certain [signs or symptoms](#)<sup>2</sup>.

**White pupil reflex:** This is the most common early sign of retinoblastoma. When you shine a light in the eye, the pupil (the dark spot in the center of the eye) should look red because of the blood vessels in the back of the eye. But in an eye with retinoblastoma, the pupil often looks white or pink instead. This might be noticed after a flash photo is taken, or it might be noted by the child's doctor during a routine eye exam.

**Lazy eye:** Sometimes the eyes don't seem to look in the same direction, a condition often called lazy eye. There are many possible causes of this in children. Lazy eye is more likely to be caused by a mild weakness of the muscles that control the eyes, but it can also be caused by retinoblastoma.

Other symptoms might include:

- Vision problems
- Eye pain
- Redness in the white part of the eye
- Bleeding in the front part of the eye
- Bulging of the eye
- A pupil that doesn't get smaller in response to bright light
- A different color in each iris

Many of these are more likely to be caused by something else. Still, if your child has signs or symptoms that could be from retinoblastoma, the doctor will want to get a medical history to find out more about the symptoms, and will do a physical exam.

Your child's doctor might want your child to be seen by an **ophthalmologist** (a doctor of eye diseases), who will look at the eye more closely. This doctor will use special lights and magnifying lenses to look inside the eye. Usually, the child needs to be under general anesthesia (asleep) during the exam so that the doctor can take a careful look.

Other [tests](#)<sup>3</sup> might be needed as well.

### Tests that may be done

[Ultrasound](#)<sup>4</sup> **of the eye:** This is often the first test done if the doctor thinks your child has an eye tumor. The test uses sound waves to make pictures of organs inside the body, such as the eye. This test doesn't hurt and does not use radiation, but the child may need to be given medicine to help keep them calm or even asleep so the doctor can get a good look at the eye and the nearby structures.

[MRI scan](#)<sup>5</sup>: This test uses radio waves and strong magnets to make detailed pictures of the inside of the body. Most children will have an MRI if retinoblastoma has been found. This test doesn't use radiation, but the child may need to be given medicine to help keep them calm or even asleep during the test.

[CT or CAT scan](#)<sup>6</sup>: This test uses x-rays to make detailed pictures of the inside of the body. MRI is often done instead of a CT scan in children with retinoblastoma, because it doesn't use radiation. But a CT scan might be done if it's not clear that the child has retinoblastoma. **Bone scan:** Most children with retinoblastoma don't need this test. But it can help show if the has spread to the skull or other bones.

**Genetic testing:** Sometimes a blood test might be done to look for the *RB1* gene change in cells outside the eye. This can help tell if a child has the heritable form of retinoblastoma, which can affect the type of follow-up a child needs after treatment. It might also affect if other children in the family need to be tested.

**Lumbar puncture (spinal tap):** Retinoblastoma can sometimes spread to the surface of the brain. This test might be done to look for cancer cells in the fluid that surrounds the brain (the cerebrospinal fluid or CSF). The child is given medicine before this test so they will be asleep. The doctor then puts a small, hollow needle between the bones of the spine to withdraw a small amount of CSF, which is sent to the lab for testing. Most children with retinoblastoma don't need this test.

**Bone marrow biopsy:** This test might be done to see if the cancer has spread to the bone marrow, which is the soft, inner part of certain bones. The test is usually not needed unless the cancer has grown outside the eye and doctors think it might have spread to the bone marrow. The child is first given medicine so they will be asleep during the test. The bone marrow samples are usually taken from the back of the hip bone using long, hollow needles. The samples are then sent to a lab to be tested for cancer cells.

### Questions to ask the doctor

- Who will do these tests?
- Where will they be done?
- Who can explain them to us?
- How and when will we get the results?
- Who will explain the results to us?
- What do we need to do next?

### How serious is my child's cancer?

If your child has retinoblastoma, the doctor will want to find out some key pieces of information to help decide how to treat it. The most important of these are:

- The [stage](#)<sup>7</sup> of the cancer, which is based on how much cancer is within the eye, and if the cancer has spread outside of the eye
- The chance of saving sight in the eye (and saving the eye itself)
- Whether the cancer is in just one eye or in both eyes

Staging systems describe how serious the cancer is and help doctors determine which treatment to use. They divide retinoblastomas into groups, based on the chance of saving the eye.

The staging of retinoblastoma can be confusing. Be sure to ask your child's doctor if you have any questions about the stage of your child's cancer.

### Questions to ask the doctor

- Where exactly is the cancer?
- How big is the cancer?

- Can all of the cancer be removed?
- Can the vision in the eye (and the eye itself) be saved?
- Has the cancer spread anywhere else?
- What is the stage of the cancer?
- How do these things affect our treatment options?
- What will happen next?

## What kind of treatment will my child need?

The main types of [treatment](#)<sup>8</sup> for retinoblastoma are:

- Surgery
- Radiation treatment
- Laser treatment
- Cryotherapy (freezing)
- Chemotherapy (chemo)

### Surgery

For most children with retinoblastoma, [surgery](#)<sup>9</sup> is not the first treatment. But if the tumor has already grown large enough so that the sight in the eye can't be saved, or if other treatments have not worked, surgery to remove the entire eye (known as **enucleation**) might be the best treatment option.

During the surgery, an implant is usually put in to take the place of the eyeball. The implant is attached to the muscles that moved the eye, so it should move the same way as the eye would have. A few weeks later, a specialist known as an **ocularist** will create an artificial eye for your child. This is a thin shell, like a very big contact lens, that will match the size and color of the remaining eye. It fits over the implant and under the eyelids. Once it's in place, it will be very hard to tell it apart from the real eye.

### Side effects of surgery

Any type of surgery can have risks and side effects, such as bleeding or infections. Ask your child's doctor or nurse what to expect. If your child has any problems, let them know. Doctors and nurses who treat children with retinoblastoma should be able to help you with any problems that come up.

The most obvious side effect is the loss of sight in that eye, although most often it has already been lost because of the cancer.

Removing the eye also can affect the future growth of bone and other structures around the eye socket, which can make the area look somewhat sunken. Using an implant can sometimes lessen this effect. (Some other types of treatment might also cause this side effect.)

## **Radiation treatment**

[Radiation](#)<sup>10</sup> uses high-energy rays (like x-rays) to kill cancer cells. Two types of radiation treatments can be used to treat children with retinoblastoma.

### ***Brachytherapy (plaque radiotherapy)***

This treatment can be used for some small tumors. A small amount of a radioactive substance is put into a small gold or lead carrier (known as a **plaque**), which is shaped like a very small bottle cap. During a short operation, the plaque is sewn in place on the outside of the eyeball where the tumor is, using tiny stitches. It is left there for several days, and then removed during a second operation. The radiation travels a very short distance, so most of it will be focused only on the tumor. The child typically stays in the hospital while the plaque is in place.

### **Side effects of brachytherapy**

The main concern with this treatment is damage to the retina or optic nerve, which can affect sight many months later. But advances in treatment may make this problem less likely.

### ***External radiation***

This treatment focuses radiation beams from a source outside the body on the cancer. This was once a common treatment for retinoblastoma. But because of the side effects it can cause, it is now most often used only for cancers that aren't well-controlled with other treatments.

Radiation is usually given 5 days a week for several weeks. Each treatment is much like getting an x-ray, but the dose of radiation is stronger. For each session, your child will lie on a special table while a machine delivers the radiation from precise angles.

The actual treatment each day lasts only a few minutes, but the setup time – getting

your child into place for treatment – takes longer. The child’s head is positioned in a custom-fitted mold that is similar to a cast used to treat broken bones. The treatment isn’t painful, but young children may be given medicine to make them sleep so they will stay still during treatment.

### **Side effects of external radiation**

Short-term problems might include effects on nearby skin areas, which can range from mild sunburn-like changes and hair loss to more severe skin reactions. These side effects tend to go away after a short while. Radiation can also damage other nearby structures. This might lead to cataracts (clouding of the lens of the eye) and damage to the retina or optic nerve, which could reduce vision. Radiation can also slow the growth of bones and other structures near the eye, which can affect the way the area around the eye looks.

Radiation can also raise the risk of getting a second cancer in the area. This is very important in children with the heritable form of retinoblastoma, who already have an increased risk of other types of cancer.

Newer forms of radiation treatment can target the tumor more precisely, which may make some of these side effects less likely than in the past.

### **Laser treatment**

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- If the cancer has spread to other parts of the body

Chemo can be given in different ways to treat retinoblastoma.

### ***Systemic chemo***

Chemo drugs can be injected into a vein (IV) or given by mouth. These drugs enter the bloodstream and reach throughout the body. Most often, 2 or 3 chemo drugs are given at the same time.

This type of chemo is given in cycles, with each treatment followed by a rest period to give the body time to recover. Each chemo cycle lasts for a few weeks, and the total length of treatment is often several months.

### ***Intra-arterial chemo***

Chemo can be injected right into the main artery that supplies blood to the eye (the **ophthalmic artery**). For this treatment, the child is given medicine to make them sleep. A very thin catheter (a long, flexible tube) is put into a large artery on the inner thigh and slowly threaded through the blood vessels all the way up into the ophthalmic artery. The chemo is then infused into the artery. This may be repeated every few weeks, depending on how much the tumor shrinks.

Because the chemo is put right into the artery feeding the eye, doctors can use much smaller doses of chemo drugs, so there are fewer side effects.

### ***Intravitreal chemo***

In this approach, a tiny needle is used to inject a chemo drug right into the **vitreous humor**, which is the jelly-like substance inside the eye. This is sometimes used (along with other types of chemo) to treat tumors that are widespread within the eye and haven't been helped by other treatments.

### **Side effects of chemo**

The side effects of chemo depend on how it's given and the types and doses of drugs used.

**Systemic chemo** can make your child feel very tired, sick to their stomach, or cause their hair to fall out. It can also make them more likely to get infections, and it might also



- Will my child need other types of treatment, too?
- What will these treatments be like?
- What's the goal of these treatments?
- What side effects could my child have from these treatments?
- What can we do about side effects that my child might have?
- Is there a clinical trial that might be right for my child?
- What about vitamins or diets that friends tell me about? How will we know if they are safe?
- How soon do we need to start treatment?
- What should we do to be ready for treatment?
- Is there anything we can do to help the treatment work better?
- What's the next step?

## What will happen after treatment?

You'll be glad [when treatment is over](#)<sup>16</sup>. But it's hard not to worry about the cancer coming back. Even if it never comes back, you might still worry about it. For years after treatment ends, your child will still need to see the doctor. At first, these visits may be every few months. Then, the longer your child is cancer-free, the less often the visits are needed.

Be sure your child goes to all of these follow-up visits. Your doctors will ask about symptoms, do physical exams, and may have tests done to see if the tumor has come back. They may also test to see if the cancer or its treatment has caused any long-term problems. If needed, they will help you and your child learn to deal with the changes.

### [For connecting and sharing during a cancer journey](#)

Anyone with cancer, their caregivers, families, and friends, can benefit from help and support. The American Cancer Society offers the Cancer Survivors Network (CSN), a safe place to connect with others who share similar interests and experiences. We also partner with CaringBridge, a free online tool that helps people dealing with illnesses like cancer stay in touch with their friends, family members, and support network by creating their own personal page where they share their journey and health updates.

## Hyperlinks

1. [www.cancer.org/cancer/types/retinoblastoma/about/what-is-retinoblastoma.html](http://www.cancer.org/cancer/types/retinoblastoma/about/what-is-retinoblastoma.html)

2. [www.cancer.org/cancer/types/retinoblastoma/detection-diagnosis-staging/signs-and-symptoms.html](http://www.cancer.org/cancer/types/retinoblastoma/detection-diagnosis-staging/signs-and-symptoms.html)
3. [www.cancer.org/cancer/types/retinoblastoma/detection-diagnosis-staging/how-diagnosed.html](http://www.cancer.org/cancer/types/retinoblastoma/detection-diagnosis-staging/how-diagnosed.html)
4. [www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/ultrasound-for-cancer.html](http://www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/ultrasound-for-cancer.html)
5. [www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/mri-for-cancer.html](http://www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/mri-for-cancer.html)  
[www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/ct-scan-for-cancer.html](http://www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/ct-scan-for-cancer.html)

**Radiation** (ray-dee-AY-shun) **therapy**: The use of high-energy rays (like x-rays) to kill cancer cells.

**Retina** (RET-in-uh): The inner layer of cells in the back of the eye. These are the cells in which retinoblastoma starts.

**Vitreous humor** (VIT-ree-us HYOO-mer): The jelly-like substance inside the eye

### How can I learn more?

We have a lot more information for you. You can find it online at [www.cancer.org](http://www.cancer.org)<sup>17</sup>. Or, you can call our toll-free number at 1-800-227-2345 to talk to one of our cancer information specialists.

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