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that children with parents or siblings with a history of retinoblastoma have this genetic test done during the first few weeks after birth. The results of the test then help define how often eye exams should be done. For example, children at risk because they have an *RB1* gene change might need an eye exam every month or so during the first year, and then every few months until at least age 4 or 5.

Children with the heritable form of retinoblastoma usually develop tumors in both eyes (bilateral retinoblastoma). Most heritable retinoblastomas develop and are diagnosed in infants only a few months old. Usually, if tumors develop in both eyes, it happens at the same time. But in some children, tumors develop in one eye first, then a few months (or even years) later in the other eye. So even if retinoblastoma is diagnosed in only one eye, these children will still need regular exams of the other eye for several years after treatment.

If a child has retinoblastoma that is thought to be hereditary, many doctors also recommend magnetic resonance imaging (MRI) scans of the brain at regular intervals for up to 5 years to check for a trilateral retinoblastoma (which includes a brain tumor such as a pineoblastoma). For more information, see [Tests for Retinoblastoma](#).

Hyperlinks

1. www.cancer.org/cancer/types/retinoblastoma/causes-risks-prevention/what-causes.html
2. www.cancer.org/cancer/risk-prevention/genetics.html

References

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Last Revised: December 3, 2018

Signs and Symptoms of Retinoblastoma

- [White pupillary reflex](#)
- [Lazy eye](#)
- [Other possible signs and symptoms](#)

Retinoblastomas nearly always occur in young children. They are often found when a parent or doctor notices that a child's eye looks unusual.

White pupillary reflex

This is the most common early sign of retinoblastoma. Normally when you shine a light in the eye, the pupil (the dark spot in the center of the eye) looks red because of the blood vessels in the back of the eye. In an eye with retinoblastoma, the pupil often appears white or pink instead, which is known as a white pupillary reflex (or leukocoria).

A parent might notice this white glare after a flash photograph is taken, especially if the pupils are different colors. It also might be noted by the child's doctor during a routine eye exam.

Lazy eye

Sometimes the eyes don't appear to look in the same direction, a condition often called lazy eye. (Doctors call this **strabismus**.) There are many possible causes of this in children. Most of the time lazy eye is caused by a mild weakness of the muscles that control the eyes, but it can also be caused by retinoblastoma.

Other possible signs and symptoms

Less common signs and symptoms of retinoblastoma include:

- Vision problems
- Eye pain
- Redness of 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 when exposed to bright light
- A different color in each iris (the colored part of the eye)



Tests for Retinoblastoma

doctor who specializes in eye diseases), who will examine the eye closely to be more certain about the diagnosis. The ophthalmologist will use special lights and magnifying lenses to look inside the eye. Usually, the child needs to be asleep (under general anesthesia) during the exam so that the doctor can take a careful and detailed look.

If a diagnosis of retinoblastoma seems likely based on the eye exam, imaging tests will be done to help confirm it and to find out how far it might have spread within the eye and possibly to other parts of the body. Usually an ophthalmologist who specializes in treating cancers of the eye (called an **ocular oncologist**) will make the final determination. This doctor should also be part of the team of doctors treating the cancer.

Imaging tests

Imaging tests use x-rays, sound waves, magnetic fields, or radioactive substances to create pictures of the inside of the body. Imaging tests may be done for a number of reasons, including:

- To help tell if a tumor in the eye is likely to be a retinoblastoma
- To determine how large the tumor is and how far it has spread
- To help determine if treatment is working

Children thought to have retinoblastoma may have one or more of these tests.

Ultrasound of the eye

[Ultrasound](#)² uses sound waves to create images of tissues inside the body, such as the inner parts of the eye. For this test, a small ultrasound probe is placed up against the eyelid or eyeball. The probe gives off sound waves and detects the echoes that bounce off the tissues inside and around the eye. The echoes are converted by a computer into an image on a screen.

Ultrasound is one of the most common imaging tests to confirm a child has retinoblastoma. This test can also be very useful when tumors in the eye are so large they prevent doctors from seeing inside the whole eye.

This test doesn't hurt and doesn't expose the child to 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.

Optical coherence tomography (OCT) is a similar type of test that uses light waves instead of sound waves to create very detailed images of the back of the eye.

Magnetic resonance imaging (MRI) scan

An [MRI scan](#)³ creates detailed images using radio waves and strong magnets (instead of x-rays). MRI scans often are used for retinoblastomas because they provide very detailed images of the eye and surrounding structures, without using radiation. This test is also very good for looking at the brain and spinal cord.

Most children with retinoblastoma will have an MRI scan as part of their initial workup. For children with bilateral retinoblastomas (tumors in both eyes), many doctors continue to do MRI scans of the brain for several years after treatment to look for tumors of the pineal gland (sometimes called trilateral retinoblastoma).

Your child may have to lie inside a narrow tube for this test, which is confining and can be upsetting. This test also requires staying still for long periods of time. Young children may be given medicine to help keep them calm or even asleep during the test.

Computed tomography (CT) scan

[CT scans](#)⁴ can help determine the size of a retinoblastoma tumor and how much it has spread within the eye and to nearby areas.

Normally, either a CT or an MRI scan is needed, but usually not both. Most doctors prefer to use MRI, because CT scans use x-rays, which might raise a child's risk for other cancers in the future. However, a CT scan can show deposits of calcium in the tumor much better than an MRI, which can be very helpful when the diagnosis of

Biopsy

For most cancers, a biopsy (removing a tissue sample from the tumor and looking at it under a microscope) is needed to make a diagnosis. But trying to biopsy a tumor at the back of the eye can often damage the eye and may spread tumor cells, so this is almost never done to diagnose retinoblastoma. Instead, doctors make the diagnosis based on eye exams and on imaging tests, like those listed above. This is why it's very important that the diagnosis of retinoblastoma is made by experts.

Lumbar puncture (spinal tap)

Retinoblastomas can sometimes grow along the optic nerve, which connects the eye to the brain. If the cancer has spread to the surface of the brain, this test can often find cancer cells in samples of cerebrospinal fluid (CSF), which is the fluid that surrounds the brain and spinal cord. Most children with retinoblastoma don't need a lumbar puncture. It's used mainly when there is a reason to think retinoblastoma might have spread into the brain.

Typically for this test, the child is given anesthesia so they will be asleep and not move during the procedure. This can help ensure the spinal tap is done cleanly. The doctor first numbs an area in the lower part of the back over the spine. A small, hollow needle is then placed between the bones of the spine to withdraw a small amount of the fluid, which is then sent to the lab for testing.

Bone marrow aspiration and biopsy

These 2 tests might be done to see if the cancer has spread to the bone marrow, the soft, inner part of certain bones. These tests are usually not needed unless the retinoblastoma has grown outside the eye and doctors suspect it might have also spread to the bone marrow.

The tests typically are done at the same time. The samples are usually taken from the back of the pelvic (hip) bone, but sometimes they may be taken from other bones. Usually the child is given anesthesia so they will be asleep during the procedure.

For a bone marrow **aspiration**, a thin, hollow needle is then inserted into the bone, and a syringe is used to suck out (aspirate) a small amount of liquid bone marrow.

A bone marrow **biopsy** is usually done just after the aspiration. A small piece of bone and marrow is removed with a slightly larger needle that is pushed down into the bone. Once the biopsy is done, pressure is applied to the site to help stop any bleeding.

The samples are then sent to a lab to be tested for cancer cells.

Hyperlinks

1. www.cancer.org/cancer/risk-prevention/genetics/genetic-testing-for-cancer-risk/what-happens-during-genetic-testing-for-cancer.html
 2. www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/ultrasound-for-cancer.html
 3. www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/mri-for-cancer.html
 4. www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/ct-scan-for-cancer.html
 5. www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/nuclear-medicine-scans-for-cancer.html
 6. www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/imaging-radiology-tests-for-cancer.html
 7. www.cancer.org/cancer/types/retinoblastoma/causes-risks-prevention/what-causes.html
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most doctors now use. It divides intraocular retinoblastomas into 5 groups, labeled A through E, based on the extent of the cancer and on the chances that the eye can be saved using current treatment options.

Group A

Small tumors (no more than 3 millimeters [mm] across) that are only in the retina and are not near important structures such as the optic disc (where the optic nerve enters the retina) or the foveola (the center of vision).

Group B

All other tumors (either larger than 3 mm or close to the optic disc or foveola) that are still only in the retina.

Group C

Well-defined tumors with small amounts of spread under the retina (subretinal seeding) or into the jelly-like material that fills the eye (vitreous seeding).

Group D

Large or poorly defined tumors with widespread vitreous or subretinal seeding. The retina may have become detached from the back of the eye.

Group E

The tumor is very large, extends near the front of the eye, is bleeding or causing glaucoma (high pressure inside the eye), or has other features that mean there is almost no chance the eye can be saved.

The Reese-Ellsworth staging system

The Reese-Ellsworth system was developed in the 1960s, when most children were being treated with [external beam radiation therapy \(EBRT\)](#)¹. While this is no longer a common treatment, some doctors may still use this system to classify retinoblastomas that have not spread beyond the eye. This system can help determine the likelihood of preserving vision while still treating the tumor.

Terms such as favorable, doubtful, and unfavorable used in this staging system refer to

the likelihood that the cancer can be treated while saving the affected eye, **not** to the likelihood of the child's survival. Indeed, more than 9 in 10 children with intraocular retinoblastomas are cured. The major challenge is saving the vision in the affected eye.

To explain the groupings below, it helps to define a few terms.

- The **optic disc** is the end of the optic nerve where it is attached to the retina. Retinoblastomas are diagnosed by looking at the retina through the front of the eye, so doctors can't measure their size directly using a ruler. Instead they compare the size of the tumor with the size of the optic disc, which is usually about 1.5 millimeters (1/16 inch) across. For example, a tumor estimated to be 3 times the size of the disc (3 *disc diameters* or 3 DD) would be about 4.5 millimeters (3/16 inch) across.
- The **equator** is an imaginary line that divides the front and back halves of the eyeball.

The Reese-Ellsworth staging system divides intraocular retinoblastoma into 5 groups. The higher the group number, from 1 to 5, the lower the chance of controlling the retinoblastoma or of saving the eye or any useful vision.

Group 1 (very favorable for saving [or preserving] the eye)

- 1A: one tumor, smaller than 4 disc diameters (DD), at or behind the equator
- 1B: multiple tumors smaller than 4 DD, all at or behind the equator

Group 2 (favorable for saving [or preserving] the eye)

- 2A: one tumor, 4 to 10 DD, at or behind the equator
- 2B: multiple tumors, with at least one 4 to 10 DD, and all at or behind the equator

Group 3 (doubtful for saving [or preserving] the eye)

- 3A: any tumor in front of the equator
- 3B: one tumor, larger than 10 DD, behind the equator

Group 4 (unfavorable for saving [or preserving] the eye)

- 4A: multiple tumors, some larger than 10 DD
- 4B: any tumor extending toward the front of the eye to the ora serrata (front edge of the retina)

Group 5 (very unfavorable for saving [or preserving] the eye)

- 5A: tumors involving more than half of the retina
- 5B: vitreous seeding (spread of tumors into the jelly-like material that fills the eye)

Other staging systems

Other staging systems that include both intraocular retinoblastomas and those that have spread beyond the eye (extraocular retinoblastomas) may be used by some doctors. These can be especially useful in countries where these cancers are more likely to have spread by the time they are found. For example, the **American Joint Commission on Cancer (AJCC) staging system** takes into account 4 key pieces of information:

- **T:** The size of the main (primary) **tumor** and how far it has grown within and outside of the eye
- **N:** Whether or not the cancer has reached nearby lymph **nodes** (small, bean-shaped collections of immune cells, to which cancers sometimes spread) in the head or neck
- **M:** Whether or not the cancer has spread (**metastasized**) to distant parts of the body, such as the bone marrow, brain, skull, or long bones
- **H:** Whether or not the child has the [heritable form of retinoblastoma](#)²

This system can be used to describe the extent of retinoblastomas in detail, particularly for those that have spread outside the eye, which rarely happens in the United States.

The different staging systems used for 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.

Hyperlinks

1. www.cancer.org/cancer/types/retinoblastoma/treating/radiation-therapy.html
2. www.cancer.org/cancer/types/retinoblastoma/causes-risks-prevention/what-causes.html

References

American Joint Committee on Cancer. Retinoblastoma. In: *AJCC Cancer Staging Manual*. 8th ed. New York, NY: Springer; 2017: 819-831.

Burwitz R, Shields CL, Shields JA, et al. Chapter 27: Retinoblastoma. In: Pizzo PA, Poplack DG, eds.

Questions to Ask About Retinoblastoma

ask any question, no matter how small it might seem. Here are some examples.

If retinoblastoma has just been diagnosed

- How sure are you that my child has [retinoblastoma](#)¹?
- Is only one eye affected or are there tumors in both eyes?
- Do we know if this is the [heritable \(hereditary\) form of retinoblastoma](#)²? How can we find out? If it is, what would this mean?
- Has the cancer spread outside the eye?
- What is the [stage](#) of the cancer, and what does that mean?
- Has my child's vision been affected?
- Do we need any other [tests](#) before we can decide on treatment?
- How much experience do you have treating this type of cancer?
- Do we need to see any other types of doctors?
- Who else will be on the treatment team, and what do they do?

When deciding on a treatment plan

- What are our [treatment options](#)³?
- Can my child's sight be saved? If so, how much?
- What do you advise for treatment and why?
- Should we get a [second opinion](#)⁴? How do we do that? Can you recommend a doctor or cancer center?
- Are there any [clinical trials](#)⁵ we should consider?
- How soon do we need to start treatment?
- How long will treatment last? What will it be like? Where will it be done?
- What should we do to be ready for treatment?
- What are the risks and side effects of the suggested treatments?
- Which side effects start shortly after treatment and which ones might develop later on?
- Will treatment affect the growth of the area around my child's eye?
- Will treatment affect my child's ability to grow and develop?
- Could treatment affect my child's [ability to have children](#)⁶ later on?

During and after treatment

Once treatment begins, you'll need to know what to expect and what to look for. Not all of these questions may apply, but getting answers to the ones that do may be helpful.

- How will we know if the treatment is working?
- Is there anything we can do to help manage side effects?
- What symptoms or side effects should we tell you about right away?
- How can we reach you or someone on your team on nights, weekends, or holidays?
- Who can we talk to if we have questions about costs, insurance coverage, or social support?
- What are the chances of the cancer [coming back](#)⁷ after treatment? What are our options if this happens?
- Is there any risk of this type of tumor occurring in our other children or relatives?
- Should we consider [genetic counseling and testing](#)⁸?
- What type of [follow-up](#)⁹ will my child need after treatment?
- Does my child have a higher long-term risk of [other cancers](#)¹⁰?

Along with these sample questions, be sure to write down some of your own. For instance, you might want more information about recovery times so you can plan your schedules. You may also want to ask about nearby or online support groups, where you may be able to get in touch with other families who have been through similar situations.

Also keep in mind that doctors aren't the only ones who can give you information. Other health care professionals, such as nurses and social workers, may be able to answer some of your questions. You can find out more about speaking with your health care team in [The Doctor-Patient Relationship](#)¹¹.

Hyperlinks

1. www.cancer.org/cancer/types/retinoblastoma/about/what-is-retinoblastoma.html
2. www.cancer.org/cancer/types/retinoblastoma/causes-risks-prevention/what-causes.html
3. www.cancer.org/cancer/types/retinoblastoma/treating.html
4. www.cancer.org/cancer/managing-cancer/finding-care/seeking-a-second-opinion.html
5. www.cancer.org/cancer/managing-cancer/making-treatment-decisions/clinical-

[trials.html](#)

6. www.cancer.org/cancer/managing-cancer/side-effects/fertility-and-sexual-side-effects.html
7. www.cancer.org/cancer/survivorship/long-term-health-concerns/recurrence.html
8. www.cancer.org/cancer/risk-prevention/genetics/genetic-testing-for-cancer-risk/what-happens-during-genetic-testing-for-cancer.html
9. www.cancer.org/cancer/types/retinoblastoma/after-treatment/follow-up.html
10. www.cancer.org/cancer/types/retinoblastoma/after-treatment/follow-up.html
11. www.cancer.org/cancer/managing-cancer/finding-care/the-doctor-patient-relationship.html

Last Revised: December 3, 2018

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