Retinoblastoma

What is retinoblastoma?

Retinoblastoma is cancer of the eye. It begins in the retina, the layer of nerve cells lining the back of the eye. It happens when nerve cells in the retina change, growing in size and number. The cells eventually form a tumor. The cells usually spread in and around the eye. They can also spread to other parts of the body, including the brain and spine.

Retinoblastoma usually affects young children, though it can sometimes occur in adults. Children with retinoblastoma may have inherited a gene from their parents that causes this disease. These children tend to get retinoblastoma at an earlier age, and in both eyes.

What are the symptoms of retinoblastoma?

When light shines in the eye (as with flash photography), you may notice the usually dark pupil looks white.

Here are some other signs of retinoblastoma:

- eyes that appear to be looking in different directions
- redness of the eye
- swelling of the eye

If you notice anything unusual about your child's eyes, call your child's doctor right away. If you have a family history of retinoblastoma, be sure to let your child's doctor know. Ask the doctor about regular eye exams for your child to check for retinoblastoma.

Retinoblastoma is often first noticed through flash photography, when you may notice the dark pupil looks white. Notice the white reflection in the child's eye on the right.

How is retinoblastoma diagnosed?

An ophthalmologist will do a complete medical exam to diagnose retinoblastoma. The doctor will do imaging tests to see if retinoblastoma is affecting other areas around the eye.

Your child's ophthalmologist may refer you to other specialists, such as a cancer doctor or a genetic counselor.

How is retinoblastoma treated?

How retinoblastoma is treated depends on the tumor and whether the cancer has spread to other areas. The goal of treatment is to save vision whenever possible.

Chemotherapy. Chemotherapy is a drug treatment. It is when chemicals travel throughout the body to kill cancer cells. Chemotherapy may help shrink a tumor so that another treatment can be used to treat the remaining cancer cells. Chemotherapy may also be used to treat cancer that has spread outside the eyeball or to other areas of the body.

Radiation therapy. Radiation therapy uses beams of energy to kill cancer cells. In some cases, a small disk of radioactive material is placed in or near the tumor. This disk is left in place for some time to kill tumor cells. In other cases, radiation is applied to the tumor from outside the body. A large machine beams energy targeted at the tumor.

Laser therapy. A laser may be used to destroy blood vessels that feed the tumor, killing the cancer cells.

Cold treatments (cryotherapy). An extremely cold substance (such as liquid nitrogen) is used to freeze cancer cells. Once the cells freeze, the substance is removed and the cells thaw out. This process of freezing and thawing is repeated, killing cancer cells.

Heat treatments (thermotherapy). Extreme heat is targeted at cancer cells to kill them.

Surgery (enucleation). If a tumor has grown too much for other treatments, surgery may be needed to remove the eyeball. This may help keep the cancer from spreading to other areas of the body.

After removing the eyeball, the surgeon places an artificial eye implant in the eye socket. The eye's muscles are attached to this implant. Eventually the eye's muscles will move this eyeball just as they did with the natural eye. However, the implanted eyeball cannot see.

Several weeks after surgery, a custom-made artificial eye can be made to match the healthy eye. This new eye clips onto the eye implant and sits right behind the eyelids just as the natural eye does. Again, this eye will not see. However, it will look and move like the healthy eye.

Removing an eye will affect a child's vision. However, most children adapt very well over time. In many cases, it will not be obvious that the child has an artificial eye.

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Summary

Retinoblastoma is cancer of the eye that begins in the retina. It can eventually spread to other parts of the body. Retinoblastoma usually affects young children, though it can sometimes occur in adults. Children with retinoblastoma may have inherited a gene from their parents that causes this disease. These children tend to get retinoblastoma at an earlier age, and in both eyes.

Get more information about retinoblastoma from EyeSmart—provided by the American Academy of Ophthalmology—at **aao.org/retinoblastoma-link**.

COMPLIMENTS OF:

BRYAN VEKOVIUS, M.D.

OCULOPLASTIC SURGERY AND
NEURO-OPHTHALMOLOGY

450 ASHLEY RIDGE BLVD.
SHREVEPORT, LA 71106

(318) 675-3733

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