

Texas Children's Cancer Center and Hematology Service

Retinoblastoma

What is retinoblastoma?

Retinoblastoma is a disease in which cancer cells are found in the retina, which is a thin membrane on the back of the inside of the eye. It is curable in most cases.

Retinoblastoma is not very common. It occurs mostly in children less than five years old. Retinoblastoma may be found in one eye or in both eyes. Some children have retinoblastoma that runs in families (hereditary retinoblastoma). Brothers and sisters of children with retinoblastoma should be checked by an eye doctor, or should have laboratory tests to find out whether they are at risk for developing the disease. If retinoblastoma occurs in one eye, it may not be hereditary. If it is in both eyes, it is always hereditary.

Treatment option overview

Treatment for retinoblastoma depends on the stage of the disease and the age and general health of the child being treated.

Surgery is a common treatment for retinoblastoma.

- A surgical procedure called enucleation removes the entire eye. A child may have normal vision in the eye that is **not** removed.

- Cryosurgery kills the cancer cells by freezing them. This procedure is used for tumors that are very small.
- Photocoagulation uses a narrow beam of very strong light to kill blood vessels that feed the tumor caused by the cancer.

Radiation therapy can also be used to treat retinoblastoma. Radiation therapy uses high-energy x-rays to kill cancer cells and shrink tumors.

Chemotherapy uses drugs to kill cancer cells. With **systemic chemotherapy**, chemotherapy drugs enter the bloodstream and travel through the body. These drugs can kill cancer cells outside the eye. If cancer cells are found in the fluid that surrounds the brain and spinal cord, chemotherapy drugs may be put into the fluid surrounding the spinal cord using a needle. This is called **intrathecal chemotherapy**. Radiation and/or chemotherapy may be used depending on the spread of the tumor beyond the eye.

Stages and treatment

Once retinoblastoma is found, more tests will be done to find out how much cancer is in the eye and to find out if it has spread to other parts of the body. This is called staging. Your child's doctor needs to know the stage of the disease to plan treatment.

Intraocular retinoblastoma

Tumors are found in one eye or in both eyes, but have not spread to tissues around the eye.

Treatment

Your child's treatment depends on whether the cancer is in one eye or both eyes.

If the cancer is one eye, your child's treatment may include one or more of the following:

1. surgery to remove the eye (enucleation)
2. external beam radiation therapy
3. photocoagulation, with or without external beam radiation therapy
4. cryosurgery

If the cancer is in both eyes, your child's treatment may include one or more of the following:

1. surgery to remove the eye with the most cancer, followed by radiation therapy to the other eye
2. radiation therapy to both eyes
3. chemotherapy initially, followed by radiation therapy

Extraocular retinoblastoma

Retinoblastoma cells have spread to tissues around the eye, or to other parts of the body.

Treatment

Your child's treatment may include one or all of the following:

1. radiation therapy to the eye
2. intrathecal chemotherapy
3. systemic chemotherapy

Recurrent retinoblastoma

Recurrent disease means the cancer cells have come back after treatment.

Treatment

Your child's treatment depends on where the cancer came back. If it came back only in the eye, your child may have surgery, photocoagulation, cryosurgery or radiation therapy. If the cancer came back in another part of the body, your child may receive chemotherapy or radiation therapy.

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