

Retinoblastoma - Childhood - Treatment Options

This section has been reviewed and approved by the [Cancer.Net Editorial Board \[1\]](#), April / 2013

Treatment Options

ON THIS PAGE: You will learn about the different ways doctors use to treat people with this type of cancer. To see other pages in this guide, use the colored boxes on the right side of your screen, or click ?Next? at the bottom.

In general, cancer in children is uncommon, so it can be hard for doctors to plan treatments unless they know what has been most effective in other children. That's why more than 60% of children with cancer are treated as part of a clinical trial. [Clinical trials \[2\]](#) are research studies that compare standard treatments (the best proven treatments available) with newer treatments that may be more effective. Studying new treatments involves careful monitoring using scientific methods, and all participants are followed closely to track their health and progress.

To take advantage of these newer treatments, all children with cancer should be treated at a specialized cancer center. Doctors at these centers have extensive experience in treating children with cancer and have access to the latest research. A doctor who specializes in treating children with cancer is called a pediatric oncologist. In many cases, a team of doctors works with a child and the family to provide care; this is called a [multidisciplinary team \[3\]](#). Pediatric cancer centers often have extra support services for children and their families, such as child life specialists, nutritionists, physical and occupational therapists, social workers, and counselors. Special activities and programs to help your child and family cope may also be available.

Several types of therapies are used for retinoblastoma, and more than 90% of children can be cured. In addition to curing the retinoblastoma, an important goal of treatment is the preservation of vision. Many of these treatment approaches described below have become available as a result of clinical trials. The [Children's Oncology Group \[4\]](#) has recently developed treatment clinical trials that include retinoblastoma.

Descriptions of the most common treatment options for retinoblastoma are listed below. Treatment options and recommendations depend on several factors, including the type and stage of cancer, possible side effects, and the patient's preferences and overall health. Take time to learn about your child's treatment options and be sure to ask questions about things that are unclear. Also, talk about the goals of each treatment with the doctor and what your child can expect while receiving the treatment. Learn more about [making treatment decisions \[5\]](#).

Surgery

Surgery is the removal of the tumor and surrounding tissue during an operation. Surgery to remove the eye is called enucleation. Children with a tumor in one eye only can often be cured with this treatment. For children with a tumor in both eyes, enucleation is used only if the ocular oncologist (a doctor who specializes in eye cancer) determines that preserving vision using other treatment is not possible. Surgical treatment for retinoblastoma should be performed by a specially trained pediatric ophthalmologist.

If enucleation is the best treatment to cure the cancer, there will be some visual loss. This may result in trouble with depth perception, but most children adapt well to these differences over time. Talk with your doctor about what to expect if enucleation is recommended and what support services are available to help adapt to this change.

Many people worry about what they will look like when they have an eye removed. The cosmetic surgery available today usually has good cosmetic results. To fill the area left by the missing eye, the person is fitted for an artificial eye, called a prosthesis. The prosthesis will look and behave almost the same as a natural eye. For example, the artificial eye will move along with the person's remaining eye, just not as much as a natural eye moves. Family members may be able to tell that the eye is not real, but it is unlikely that strangers will know. If enucleation is required, talk with your doctor about a prosthesis; it may take many weeks for patients to receive the prosthesis. Learn more about [cancer surgery \[6\]](#).

Radiation therapy

Radiation therapy is the use of high-energy x-rays or other particles to kill cancer cells. A doctor who specializes in giving radiation therapy to treat cancer is called a radiation oncologist. The most common type of radiation treatment is called external-beam radiation therapy, which is radiation therapy given from a machine outside the body. Radioactive plaque therapy, also called internal radiation therapy or brachytherapy, is the delivery of radiation therapy directly to the eye with a disc containing radiation. A radiation therapy regimen (schedule) usually consists of a specific number of treatments given over a set period of time.

Fatigue, drowsiness, nausea, vomiting, and headache are common side effects of radiation therapy, and these usually go away after treatment. Radiation therapy in young children can interfere with normal growth, including growth of the bones around the eye, depending on the dose. The

increased risk of additional tumors later in life for children with the hereditary form of retinoblastoma is further increased after external-beam radiation therapy. These effects are not seen after radioactive plaque therapy. Learn more about [radiation therapy](#) [7].

Cryotherapy

Cryosurgery, also called cryotherapy or cryoablation, uses liquid nitrogen to freeze and kill cells. More than one freezing may be needed.

Laser therapy

Laser therapy uses heat in the form of a laser to shrink a smaller tumor. It may be called thermotherapy (or TTT for transpupillary thermotherapy), and it may be used alone or in addition to cryotherapy or radiation therapy. Photocoagulation is a different type of laser therapy that uses light to shrink tumors.

Chemotherapy

Chemotherapy is the use of drugs to kill cancer cells, usually by stopping the cancer cells' ability to grow and divide and may be used to shrink an eye tumor. Systemic chemotherapy is delivered through the bloodstream to reach cancer cells throughout the body. Chemotherapy is given by a pediatric oncologist or a medical oncologist, a doctor who specializes in treating cancer with medication. A chemotherapy regimen usually consists of a specific number of cycles given over a set period of time.

Chemotherapy is also delivered through the blood vessel feeding the eye and the tumor by inserting a catheter in the child's groin area. The doctor maneuvers the catheter with the help of imaging technology to reach the blood vessel in the head. This procedure is used in a few medical centers for children with advanced retinoblastoma with some success in an attempt to prevent removal of the affected eye.

Chemotherapy often makes it possible to completely get rid of any remaining smaller tumors with the following focal (localized) measures, outlined above:

- Thermotherapy or photocoagulation (laser therapy)
- Cryotherapy
- Radioactive plaque therapy

Chemoreduction (using chemotherapy to reduce the size of the tumor) is a treatment approach that is often used for children with retinoblastoma in both eyes in the hope of avoiding enucleation and preserving vision in at least one eye. The ophthalmologist, working closely with the pediatric oncologist, will determine if this treatment is appropriate.

Children with low-risk disease and some children with intermediate-risk disease may be able to avoid postoperative chemotherapy or reduce the intensity of chemotherapy. Talk with the doctor about whether this approach is appropriate for your child. Both doctors will regularly monitor how the treatment is affecting the cancer and may recommend additional treatment to prevent the cancer from returning.

The drugs used most often for retinoblastoma are vincristine (Oncovin, Vincasar PFS), carboplatin (Paraplatin), and etoposide (Toposar, VePesid). Depending on the extent of the tumor, a combination of two or more drugs will be recommended. All chemotherapy has side effects that occur during treatment. Some drugs have the potential for specific long-term side effects. Talk with your doctor about the specific drugs used and the possible long-term side effects before treatment begins.

Learn more about [chemotherapy](#) [8] and [preparing for treatment](#) [9]. The medications used to treat cancer are continually being evaluated. Talking with your child's doctor is often the best way to learn about the medications prescribed for you, their purpose, and their potential side effects or interactions with other medications. Learn more about your child's prescriptions by using [searchable drug databases](#) [10].

Stem cell rescue/transplantation

For some children with Stage IV extraocular retinoblastoma, a stem cell transplant may be recommended. The child is first treated with high doses of chemotherapy (and/or radiation therapy) to destroy as many cancer cells as possible, then the transplantation takes place. A stem cell transplant is a medical procedure in which healthy, highly specialized cells, called hematopoietic stem cells, are given to a patient, either to replace bone marrow that contains cancer or to help the patient's bone marrow recover after other cancer treatment. Hematopoietic stem cells are found both in the bloodstream and in the bone marrow. Today, this procedure is more commonly called a stem cell transplant, rather than bone marrow transplant, because it is the stem cells in the blood that are typically being transplanted, not the actual bone marrow tissue.

Before recommending transplantation, doctors will talk with the patient about the risks of this treatment and consider several other factors, such as results of any previous treatment and the patient's age and general health. Learn more about [bone marrow and stem cell transplantation](#). [11]

Getting care for symptoms and side effects

Cancer and its treatment often cause side effects. In addition to treatment to slow, stop, or eliminate the cancer, an important part of cancer care is relieving a person's symptoms and side effects. This approach is called palliative or supportive care, and it includes supporting the patient with his or her physical, emotional, and social needs.

Palliative care can help a person at any stage of illness. People often receive treatment for the cancer and treatment to ease side effects at the same time. In fact, patients who receive both often have less severe symptoms, better quality of life, and report they are more satisfied with treatment.

Palliative treatments vary widely and often include medication, nutritional changes, relaxation techniques, and other therapies. Your child may also receive palliative treatments similar to those meant to eliminate the cancer, such as chemotherapy, surgery, and radiation therapy. Talk with your

child's doctor about the goals of each treatment in the treatment plan.

Before treatment begins, talk with your child's health care team about the possible side effects of your child's specific treatment plan and supportive care options. And during and after treatment, be sure to tell your child's doctor or another health care team member if your child is experiencing a problem so it is addressed as quickly as possible. Learn more about [palliative care](#) [12].

Recurrent retinoblastoma

A remission is when cancer cannot be detected in the body and there are no symptoms. This may also be called "no evidence of disease" or NED.

A remission can be temporary or permanent. This uncertainty leads to many survivors feeling worried or anxious that the cancer will come back. Although many remissions are permanent, it's important to talk with your child's doctor about the possibility of the cancer returning. Understanding the risk of recurrence and the treatment options may help you and your child feel more prepared if the cancer does return. Learn more about [coping with the fear of recurrence](#) [13].

If the cancer returns after the original treatment, it is called recurrent cancer. It may come back in the same place (called a local recurrence), nearby (regional recurrence), or in another place (distant recurrence).

When this occurs, a cycle of testing will begin again to learn as much as possible about the recurrence, including whether the cancer's stage has changed. After testing is done, you and your child's doctor will talk about your treatment options. If retinoblastoma returns after treatment, the treatment plan depends on where the cancer recurred and how aggressive the new tumor is. The doctor may recommend surgery, radiation therapy, chemotherapy, and/or focal measures, such as photocoagulation, thermotherapy, or cryotherapy (see above). The doctor may also suggest clinical trials that are studying new ways to treat this type of recurrent cancer.

People with recurrent cancer and their families often experience emotions such as disbelief or fear. Parents are encouraged to talk with their health care team about these feelings and ask about support services to help them cope. Learn more about [dealing with cancer recurrence](#) [14].

If treatment fails

Although treatment is successful for the majority of children with cancer, sometimes it is not. If a child's cancer cannot be cured or controlled, this is called advanced or terminal cancer. This diagnosis is stressful, and it may be difficult to discuss. However, it is important to have open and honest conversations with your child's doctor and health care team to express your family's feelings, preferences, and concerns. The health care team is there to help, and many team members have special skills, experience, and knowledge to support patients and their families.

Parents or guardians are encouraged to think about where the child would be most comfortable: at home, in a home-like setting elsewhere, in the hospital, or in a hospice environment. Hospice care is a type of palliative care for people who are expected to live less than six months. It is designed to provide the best possible quality of life for people who are near the end of life. Nursing care and special equipment can make staying at home a workable alternative for many families. Some children may be happier if they can arrange to attend school part-time or keep up other activities and social connections. The child's health care team can help parents or guardians decide on an appropriate level of activity. Making sure a child is physically comfortable and free from pain is extremely important as part of end-of-life care. Learn more about [caring for a terminally ill child](#) [15] and [advanced cancer care planning](#) [16].

The death of a child is an enormous tragedy, and families may need support to help them cope with the loss. Pediatric cancer centers often have professional staff and support groups to help with the process of grieving. Learn more on [grieving the loss of a child](#). [17]

Choose "Next" (below, right) to continue reading to learn more about clinical trials, which are research studies. Or, use the colored boxes located on the right side of your screen to visit any section.

Links:

- [1] <http://www.cancer.net/about-us>
- [2] <http://www.cancer.net/node/19584>
- [3] <http://www.cancer.net/node/25356>
- [4] <http://www.curesearch.org/>
- [5] <http://www.cancer.net/node/24582>
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