Treating Retinoblastoma

This information represents the views of the doctors and nurses serving on the American Cancer Society’s Cancer Information Database Editorial Board. These views are based on their interpretation of studies published in medical journals, as well as their own professional experience.

The treatment information in this document is not official policy of the Society and is not intended as medical advice to replace the expertise and judgment of your cancer care team. It is intended to help you and your family make informed decisions, together with your doctor.

Your doctor may have reasons for suggesting a treatment plan different from these general treatment options. Don’t hesitate to ask him or her questions about your treatment options.

General treatment information

Retinoblastoma is rare, so not many doctors other than those in specialty eye hospitals and major children’s cancer centers have much experience treating it. Children with retinoblastoma and their families have special needs that can best be met by these children’s cancer centers. These centers have teams of specialists who know about retinoblastoma and the unique needs of children with cancer. This gives the child the best chance for recovery and, if possible, keeping their sight.

If your child has retinoblastoma, be sure he or she is treated at a children’s cancer center that has expertise in treating children with this rare form of cancer. Ask about the services offered at your treatment center. Your child’s doctor or nurse can tell you what is available to help with any problems you or your child might have.

Children with retinoblastoma are treated by a team of doctors that often includes:
- A **pediatric ophthalmologist**: a doctor who treats eye diseases in children
- An **ocular oncologist**: a doctor (usually an ophthalmologist) who treats cancers of the eye
- A **pediatric oncologist**: a doctor who treats children with cancer
- A **radiation oncologist**: a doctor who treats cancer with radiation therapy

The team might also include other doctors, physician assistants (PAs), nurse practitioners (NPs), nurses, therapists, child psychologists, social workers, genetic counselors, and other professionals. Having a child go through cancer treatment often means meeting lots of specialists and learning about parts of the medical system you probably haven’t been exposed to before. For more information, see [Children Diagnosed With Cancer: Understanding the Health Care System](#).

Once the cancer is found and the needed tests have been done, the cancer care team will discuss treatment options with you. It’s important to discuss all of the options as well as their possible side effects with your child’s doctors to help you make an informed decision. For a list of some questions to ask, see the section [What Should You Ask Your Child’s Doctor About Retinoblastoma?](#) Then add your own.

If time permits, it can often be helpful to get a second opinion if you have questions about the recommended plan (or if you just want to confirm this is the best option). This can give you more information and help you feel more confident about the treatment plan you choose. Check with your insurance provider about their policy on second opinions.

**Treatment principles**

The goals of treatment for retinoblastoma are:

- To get rid of the cancer and save the child’s life
- To save the eye if possible
- To preserve as much vision as possible
- To limit the risk of second cancers later in life, which can be caused by treatment, particularly in children with hereditary retinoblastoma

The most important factors that help determine treatment are:

- Whether the tumor is just in one eye or both
- How good the vision in the eye is
- Whether the tumor has extended outside the eye

Overall, more than 9 in 10 children with retinoblastoma are cured. The chances of long-
term survival are much better if the tumor has not spread outside the eye.

The main types of treatment for retinoblastoma are:

- **Surgery**
- **Radiation therapy**
- **Photocoagulation** (using lasers to kill small tumors or the blood vessels that feed them)
- **Cryotherapy** (using cold to freeze and kill small tumors)
- **Thermotherapy** (using a type of laser to apply heat to kill small tumors)
- **Chemotherapy**
- **High-dose chemotherapy and stem cell transplant**

Sometimes more than one type of treatment may be used. The treatment options are based on the extent of the cancer and other factors.

**Surgery (Enucleation) for Retinoblastoma**

Surgery is not needed for all retinoblastomas, especially for smaller tumors. But if a tumor gets quite large before it is found, vision in the eye has often already been destroyed, with no hope of getting it back. The usual treatment in this case is *enucleation*, an operation to remove the whole eye, plus part of the optic nerve attached to it. This is done while the child is under general anesthesia (in a deep sleep).

During the same operation, an orbital implant is usually put in to take the place of the eyeball. The implant is made out of silicone or hydroxyapatite (a substance similar to bone). It is attached to the muscles that moved the eye, so it should move the same way as the eye would have.

Your child will probably be able to leave the hospital the same day or the next day.

After several weeks, you can visit an *ocularist*, who will create an artificial eye for your child. This is a thin shell that fits over the orbital implant and under the eyelids, like a big contact lens. It will match the size and color of the remaining eye. Once it is in place, it will be very hard to tell it apart from the real eye.

When retinoblastoma occurs in both eyes, enucleation of both eyes would result in complete blindness. If neither eye has useful vision because of damage already caused
by the cancer, this is the best way to make sure all of the cancer is gone. But if there is
any chance of saving useful vision in one or both eyes, doctors may advise trying other
types of treatment first.

**Possible side effects:** The most obvious side effect of enucleation is the loss of vision
in that eye, although most often the vision has already been lost because of the cancer.

Removing the eye also can affect the future growth of bone and other tissues around
the eye socket, which can make the area look somewhat sunken. Using an orbital
implant can sometimes lessen this effect. ([Radiation therapy](#), the other major treatment
option in such cases, may cause the same side effect.)

- **References**
  See all references for Retinoblastoma

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## Radiation Therapy for Retinoblastoma

This treatment uses high energy x-rays or particles to kill cancer cells. Radiation therapy
is an effective treatment for some children with retinoblastoma. Compared with surgery,
it has the advantage of possibly saving vision in the eye. But radiation therapy also has
some disadvantages (see “Possible side effects” below).

Two types of radiation therapy can be used to treat children with retinoblastoma.

### External beam radiation therapy

External beam radiation therapy (EBRT) 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
are not well-controlled with other treatments.

Radiation is usually given 5 days a week for several weeks. Before treatments start, the
radiation team takes careful measurements with [imaging tests](#) such as MRI scans to
determine the correct angles for aiming the radiation beams and the proper dose of radiation.

Each treatment is much like getting an x-ray, but the dose of radiation is much higher. 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 – usually 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 is not painful, but young children may be given medicine to make them sleep so they will stay still during treatment.

**Newer forms of radiation therapy**

Many centers now use newer types of external radiation therapy, which can target the tumor more precisely. This lowers the doses that surrounding normal tissues get, which may help reduce side effects.

**Intensity modulated radiation therapy (IMRT):** IMRT lets doctors shape the radiation beams and aim them at the tumor from several angles, as well as adjust the intensity (strength) of the beams to limit the dose reaching the nearby normal tissues. This may let the doctor deliver a higher dose to the tumor, while reducing side effects. Many major hospitals and cancer centers now use IMRT.

**Proton beam therapy:** Protons are positive parts of atoms. Unlike the x-rays used in standard radiation, which release energy both before and after they hit their target, protons cause little damage to tissues they pass through and then release their energy after traveling a certain distance. Proton beam radiation may be able to deliver the same level of radiation to the tumor while causing much less damage to nearby normal tissues. Early results with proton beam therapy are promising, but it’s still fairly new, and there is very little long-term data on its use for retinoblastoma. There are only about 15 centers that do proton beam therapy in the United States at this time.

**Possible side effects of EBRT**

Some of the side effects of EBRT will go away after a short while and are usually not serious. Short-term problems might include effects on skin areas that receive radiation, which can range from mild sunburn-like changes and hair loss to more severe skin reactions.

More importantly, EBRT can damage nearby normal body tissues. This might eventually
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 tissues near the eye, which can affect the way the area around the eye looks over time.

External radiation therapy can also increase the risk of developing a second cancer in the area. This is especially important in children with the hereditary form of retinoblastoma, who are already at increased risk for developing other types of cancer.

Newer forms of radiation therapy, such as IMRT and proton beam therapy, target the tumor more precisely and spare more normal tissue. This may make some of these side effects less likely than in the past.

**Brachytherapy (plaque radiotherapy)**

The use of brachytherapy, also known as *internal radiation therapy* or *episceral plaque radiotherapy*, is limited to small tumors. During brachytherapy, a small amount of radioactive material is placed on the outside of the part of the eyeball where the tumor is for several days. The radioactive material is put in a small carrier (known as a *plaque*), which is shaped like a very small bottle cap. The plaque is made of gold or lead to shield nearby tissues from the radiation. The radiation travels a very short distance, so most of it will be focused only on the tumor.

The plaque is sewn in place on the eyeball with tiny stitches during a short operation. It is then removed during a second operation several days later. Both operations are done while the child is under general anesthesia (in a deep sleep). The child typically stays in the hospital while the plaque is in place.

**Possible side effects:** Brachytherapy is less likely to cause side effects than external radiation. The main concern is damage to the retina or optic nerve, which can affect vision many months later. Recent advances in treatment may make this problem less likely. Brachytherapy has not been linked to an increased risk of developing a second cancer.

For more information on radiation therapy, see [A Guide to Radiation Therapy](#).

- [References](#)
  
  See all references for Retinoblastoma

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Laser Therapy (Photocoagulation) for Retinoblastoma

Photocoagulation is a type of treatment that uses a laser beam aimed through the pupil. The laser is focused on the blood vessels that surround and supply the tumor, destroying them with the heat caused by the beam. Photocoagulation is effective only for smaller tumors toward the back of the eye.

Your child will be under general anesthesia (in a deep sleep) during the treatment. The treatment is usually given 2 or 3 times, with about a month between treatments.

Possible side effects: In some cases, laser therapy can damage the retina, which can lead to blind spots or temporarily cause the retina to detach from the back of the eyeball.

- References

See all references for Retinoblastoma

Cryotherapy for Retinoblastoma

In cryotherapy, the doctor uses a small metal probe that is cooled to very low temperatures, killing the retinoblastoma cells by freezing them. It is only effective for small tumors toward the front of the eye. It is not used routinely for children with several tumors.

The child will be under general anesthesia (in a deep sleep) during the treatment. After the child is asleep, the probe is placed on the outer surface of the eyeball next to the
tumor, which is then frozen and thawed several times. Cryotherapy is usually given 2 or 3 times, with about a month between treatments.

**Possible side effects:** Cryotherapy can cause the eye and eyelid to swell for a few days. As with laser therapy, cryotherapy can damage the retina, which can lead to blind spots or temporarily cause the retina to detach from the back of the eyeball.

- References
  See all references for Retinoblastoma

Thermotherapy for Retinoblastoma

For thermotherapy (also called *transpupillary thermal therapy*, or TTT), the doctor uses a different type of laser than that used in *photocoagulation therapy*. The laser applies infrared light to heat and kill the tumor cells. The temperatures are not quite as high as those used in photocoagulation therapy, so some of the blood vessels on the retina may be spared.

Thermotherapy can be used alone for very small tumors. For larger tumors, it can be used along with *chemotherapy* (called *thermochemotherapy*) or with *radiation therapy* (called *thermoradiotherapy*). Heat seems to help these other treatments work better.

The treatment is given while the child is asleep, usually for about 10 minutes at a time. Typically, 3 treatments about a month apart are needed to control each tumor. When used as part of thermochemotherapy, the heat is usually applied at a lower temperature over a slightly longer period of time, starting within a few hours after chemotherapy.

**Possible side effects:** Thermotherapy can sometimes cause part of the iris (the colored part of the eye) to shrink. Other possible effects include clouding of part of the eye lens or damage to the retina, which might affect vision.

- References
  See all references for Retinoblastoma
Chemotherapy for Retinoblastoma

Chemotherapy (chemo) is the use of anti-cancer drugs to treat retinoblastoma.

Ways of giving chemotherapy

Chemo can be given in different ways.

**Systemic chemotherapy:** In most cases, chemo drugs are injected into a vein (IV) or given by mouth. These drugs enter the bloodstream and reach throughout the body. This is known as *systemic chemotherapy*.

**Periocular (subtenon) chemotherapy:** For some advanced intraocular cancers, higher doses of chemo are needed inside the eye. Along with systemic chemotherapy, one of the drugs (carboplatin) may be injected in the tissues around the eye, where it slowly diffuses into the eyeball. This is called *periocular or subtenon chemotherapy*. These injections are done while the child is under anesthesia (asleep). This can cause redness and swelling around the eye.

**Intra-arterial chemotherapy:** A newer approach sometimes used instead of systemic chemotherapy is to inject chemo directly into the ophthalmic artery, the main artery that supplies blood to the eye. In this technique, a very thin catheter (a long, hollow, flexible tube) is inserted into a large artery on the inner thigh and slowly threaded through the blood vessels all the way up into the ophthalmic artery. (This is done with the child asleep under general anesthesia.) The chemo is then infused into the artery. The drug used most often is melphalan, but other drugs such as carboplatin and topotecan can also be used. This process may then be repeated every few weeks, depending on how much the tumor shrinks.

Because the chemo is put directly into the artery feeding the eye, doctors can use much smaller doses of chemo drugs (less than 10% of the doses used for systemic chemo). Therefore, there are fewer side effects from the chemo.
Early results with this technique in eyes with advanced tumors have been promising, generally with good tumor control and few side effects. In most cases it has allowed doctors to save an eye that otherwise would have needed to be removed.

**Intravitreal chemotherapy:** In this newer approach, chemotherapy is given directly into the vitreous humor, the jelly-like substance inside the eye. This approach is discussed in the section [What’s New in Retinoblastoma Research and Treatment?](#)

**Uses of chemotherapy**

Chemotherapy may be used in different situations:

- Chemo can be used as the first treatment to shrink some tumors that have not spread outside the eye. This is called *chemoreduction*. These tumors can then be treated more effectively with focal therapies such as *laser therapy*, *cryotherapy*, *thermotherapy*, or *brachytherapy*.
- Systemic (IV) chemo may be given to children whose tumors don’t seem to have spread beyond the eye, but might be likely to spread because of the tumor’s size and/or location.
- Chemo is sometimes used when the eye has already been removed, but the tumor was found to extend into some areas in the eye that make it more likely to have spread. This type of treatment is called *adjuvant chemotherapy*.
- Systemic chemo is also used to treat children whose retinoblastoma has spread beyond the eye, a much more critical situation. If the cancer has spread to the brain, chemo may also be given directly into the cerebrospinal fluid that surrounds it (known as *intrathecal chemotherapy*). Tumors outside the eye may shrink for a time with standard doses of chemo, but they will usually start growing again. For this reason, doctors often prefer to give more intense chemo, usually along with a stem cell transplant. (See the section [High-dose Chemotherapy and Stem Cell Transplant](#).)

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

Some of the drugs used to treat children with retinoblastoma include:

- Carboplatin
- Cisplatin
- Vincristine
- Etoposide
- Cyclophosphamide
- Topotecan
- Doxorubicin

Most often, 2 or 3 drugs are given at the same time. A standard combination used to shrink intraocular retinoblastomas is carboplatin, vincristine, and etoposide, although for very small tumors, only carboplatin and vincristine may be enough. Other drugs might be used if these are not effective.

**Possible side effects**

Chemo drugs attack cells that are dividing quickly, which is why they work against cancer cells. But other cells in the body, such as those in the bone marrow (where new blood cells are made), the lining of the mouth and intestines, and the hair follicles, also divide quickly. These cells are also likely to be affected by chemo, which can lead to side effects.

Children tend to have less severe side effects from chemo and to recover from side effects more quickly than adults do. One benefit of this is that doctors can give them the high doses of chemo needed to kill the tumor.

The **side effects** of chemo depend on the type of drugs, the doses used, and how long they are given. Possible short-term side effects include:

- Hair loss
- Mouth sores
- Loss of appetite
- Nausea and vomiting
- Diarrhea or constipation
- Increased chance of infections (from having too few white blood cells)
- Easy bruising or bleeding (from having too few blood platelets)
- Fatigue (from having too few red blood cells)

Most of these side effects go away after treatment is finished. There are often ways to lessen these side effects. For example, drugs can be given to help prevent or reduce nausea and vomiting. Be sure to discuss any questions about side effects with your child's cancer care team, and let them know if your child has side effects so they can be managed.

Along with those listed above, certain chemo drugs can cause specific side effects. For example:
• Cisplatin and carboplatin can affect the kidneys. Giving the child plenty of fluids during treatment can help reduce this risk. These drugs can also cause hearing loss in young children, especially in babies younger than 6 months. Your child’s doctor may check your child’s hearing with tests during or after treatment. When carboplatin is injected directly into the tissues near the eye (periocular chemotherapy), it can cause redness and swelling in the area.
• Vincristine can damage nerves. Some children may feel tingling and numbness, particularly in their hands and feet.
• Some drugs, such as etoposide, doxorubicin, and cyclophosphamide, can increase the risk of developing a cancer of white blood cells known as acute myeloid leukemia (AML) later in life. Fortunately, this is not common.
• Doxorubicin can damage the heart. The risk of this happening goes up with the total amount of the drug given. Doctors try to limit this risk as much as possible by not giving more than the recommended doses and by checking the heart with an echocardiogram (an ultrasound of the heart) during treatment.
• Cyclophosphamide can damage the bladder, which can cause blood in the urine. This risk can be lowered by giving this drug along with plenty of fluids and with a drug called mesna, which helps protect the bladder.

For more information about chemotherapy, see the Chemotherapy section of our website.

• References
See all references for Retinoblastoma

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High-Dose Chemotherapy and Stem Cell Transplant for Retinoblastoma

Doctors are studying the use of this type of treatment in children with retinoblastoma that has spread outside the eye and who are unlikely to be cured with other treatments.
A stem cell transplant (SCT) lets doctors give higher doses of chemotherapy (chemo) than could safely be given otherwise). In the past, this type of treatment was commonly referred to as a bone marrow transplant.

The bone marrow is the soft, inner part of some bones where new blood cells are made. Chemo drugs can affect quickly dividing cells like those in the bone marrow. Even though higher doses of these drugs might be more effective in treating tumors, they can’t be given because they would cause severe damage to bone marrow cells, leading to life-threatening shortages of blood cells.

To try to get around this problem, the doctor may treat the child with high-dose chemo (sometimes along with radiation therapy) and then use a stem cell transplant to “rescue” the bone marrow.

How is it done?

The first step in a SCT is to collect, or harvest, the child’s own blood-making stem cells to use later. (These are the cells that make the different types of blood cells.)

In the past, the stem cells were often taken from the child’s bone marrow, which required a minor operation. But doctors have found that these cells can be taken from the bloodstream during a procedure similar to donating blood. Instead of going into a collecting bag, the blood goes into a special machine that filters out the stem cells and returns the rest of the blood to the child’s body. The stem cells are then frozen until the transplant. This process may need to be done more than once.

Once the stem cells have been stored, the child gets high-dose chemotherapy in the hospital, sometimes along with radiation therapy. When the treatment is finished (a few days later), the stem cells are thawed and returned to the body in a process similar to a normal blood transfusion. The stem cells travel through the blood and settle in the bone marrow.

Over the next few weeks, the stem cells start to make new blood cells. Until this happens, the child is at high risk of infection because of a low white blood cell count, as well as bleeding because of a low platelet count. To avoid infection, protective measures are taken, such as using special air filters in the hospital room and having visitors wear protective clothing. Blood and platelet transfusions and treatment with IV antibiotics may also be used to prevent or treat infections or bleeding problems.

The child can usually leave the hospital once their blood cell counts return to a safe level. They may then need to make regular visits to the outpatient clinic for about 6
months, after which time their care may be continued by their regular doctors.

**Practical points**

A SCT is a complex treatment that can cause life-threatening side effects. If the doctors think your child can benefit from a transplant, the best place to have it done is at a nationally recognized cancer center where the staff has experience with the procedure and with managing the recovery period.

A stem cell transplant often requires a long hospital stay and can be very expensive, often costing well over $100,000. Be sure to get a written approval from your insurer before treatment if it is recommended for your child. Even if the transplant is covered by your insurance, your co-pays or other costs could easily amount to many thousands of dollars. It’s important to find out what your insurer will cover before the transplant to get an idea of what you might have to pay.

**Possible side effects**

The possible side effects from SCT are generally divided into early and long-term effects.

**Short-term, early side effects:** The early complications and side effects are basically the same as those listed in the Chemotherapy for Retinoblastoma section, but they can be more severe because the drug doses are higher. Side effects can include:

- Low blood cell counts (with fatigue and an increased risk of infection and bleeding)
- Nausea and vomiting
- Loss of appetite
- Mouth sores
- Diarrhea
- Hair loss

One of the most common and serious short-term effects is an increased risk of serious infections. Antibiotics are often given to try to keep this from happening. Other side effects, like low red blood cell and platelet counts, may require blood product transfusions or other treatments.

**Long-term and late side effects:** Some complications and side effects can last for a long time or might not occur until months or years after the transplant. These can include:
• Radiation damage to the lungs
• Problems with the thyroid or other hormone-making glands
• Problems with fertility
• Damage to bones or problems with bone growth
• Development of another cancer (including leukemia) years later

Be sure to talk to your child’s doctor before the transplant to learn about possible long-term effects your child might have. For more on the long-term effects of this and other treatments, see the section Late and Long-term Effects of Treatment for Retinoblastoma.

To learn more about stem cell transplants, see Stem Cell Transplant for Cancer.

• References
See all references for Retinoblastoma

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Treatment of Retinoblastoma, Based on Extent of the Disease

If your child has retinoblastoma, a number of factors can affect the treatment options your child’s doctor recommends. Some of these include:

• Whether tumors are in one or both eyes
• The size and location of the tumor in the eye(s)
• The chance for saving vision in the eye(s)
• Whether the tumor is still confined within the eye(s) or has spread elsewhere

If the retinoblastoma is only in one eye, treatment depends on whether vision in the eye can be saved. If the chance to save vision is poor, the treatment is often surgery to remove the eye. Surgery was used more often in the past even for smaller tumors, as it offered the best chance to ensure all of the cancer was removed. But in recent years, doctors have become more comfortable with using other treatments (often intra-arterial chemotherapy plus some form of local treatment) if there is a good chance of saving
vision in the eye.

If the retinoblastoma is in both eyes, doctors will try to save at least one eye if at all possible so that the child maintains some vision. Most children with retinoblastoma in both eyes will be treated with chemotherapy (intra-arterial or systemic) first to shrink the tumors (called chemoreduction), followed by some form of local treatment and possibly radiation therapy. Surgery (removal of the eye) is reserved for the most advanced tumors and for those that do not respond to chemoreduction and local treatments.

Many children will get several types of treatment. Treatment might be needed for months or even years, especially in eyes treated with cryotherapy and/or photocoagulation after chemotherapy.

No matter which types of treatment are used, it’s very important that they are given by experts at centers experienced in treating these tumors.

**If the eye can see and probably can be saved**

For some smaller tumors, local treatments such as laser therapy (photocoagulation) or cryotherapy may be the only treatment needed.

More often, tumors are larger or in hard-to-treat areas. Treatment usually includes a combination of chemotherapy and focal treatments. If systemic chemotherapy (chemotherapy given by vein) is used, it is given for about 6 months to shrink the tumor as much as possible. More recently, many centers have begun to give chemotherapy directly into the artery that feeds the eye (known as intra-arterial chemotherapy) instead of systemic chemotherapy. Usually a few treatments are needed, each given a few weeks apart.

Depending on how much the tumor shrinks and where it is in the eye, different focal treatments can then be applied, usually starting after the first or second cycle of chemotherapy. Treatment options may include brachytherapy (plaque radiotherapy), cryotherapy, laser therapy (photocoagulation), or thermotherapy. External radiation therapy may also be given, but it is usually delayed until the end of chemotherapy.

If the combination of these treatments doesn’t control the disease, surgery to remove the eye may be needed.

**If the eye cannot see or cannot be saved**

If there is no vision in the eye, if the tumor is so advanced within the eye that there is no
hope of cure by other means, or if there is painful glaucoma, then surgery is done to remove the eye and place an orbital implant in the socket.

If the cancer affects only one eye, no other treatments may be required. But sometimes, after looking at the removed eye under the microscope, the doctors find that some retinoblastoma cells might have escaped the eye and thus may come back later in other parts of the body. These children may be given chemotherapy, possibly along with radiation therapy to the area around the eye, to try to lower this risk.

In some instances where there are large tumors in both eyes, chemotherapy may be used first to try to shrink the tumors and avoid the need for surgery in both eyes. If chemotherapy shrinks the tumors enough, local therapies such as brachytherapy (plaque radiotherapy), cryotherapy, laser therapy (photocoagulation), or thermotherapy might allow at least one eye (and some vision) to be saved.

Some children with retinoblastoma in only one eye may actually have the hereditary form of retinoblastoma, which means they will probably develop disease in the other eye as well. Therefore, it is very important that children with retinoblastoma in one eye continue to have the other eye examined regularly after treatment.

**If the cancer has spread outside the eye**

In this uncommon situation, treatment is usually a combination of chemotherapy, radiation, and in some cases surgery.

If the cancer has spread only to the orbit (the area around the eye), treatment with chemotherapy, surgical removal (enucleation) of the eye, and radiation therapy to the orbit is often successful.

If the cancer has spread outside the orbit to distant parts of the body such as the liver or the bones and bone marrow, the chances of a cure using standard chemotherapy and other treatments are very low. In these cases, using higher doses of chemotherapy followed by a stem cell transplant can often be successful.

Cancers that have spread to the brain (including trilateral retinoblastomas, which are retinoblastomas that usually start in the pineal gland) are very hard to treat, and the chances of a cure using standard treatments are low. High-dose chemotherapy and stem cell transplant has shown some promise for these cancers in a recent small study. Clinical trials using other newer treatments may be an option in this situation as well.

**If the cancer comes back in the eye after initial treatment**
Treatment of cancer that recurs in the eye depends on the size and location of the tumor and on what treatments were used the first time. If the tumor is small, the child's sight can often be saved while the cancer is destroyed with local treatments such as cryotherapy, laser therapy, radiation therapy (if not already used), or other treatments. Chemotherapy may be given first. If the child's sight cannot be saved, the eye may need to be removed with surgery. Either way, the chance of a cure is very good as long as the cancer is still confined to the eye.

If the cancer comes back outside the eye after initial treatment

Cancers that recur outside the eye are harder to treat. Options may include chemotherapy and radiation, or high-dose chemotherapy with a stem cell transplant in some cases. In this situation, the treatment and the chances of cure are similar to what is described above (when the cancer has spread outside the eye before initial treatment).

Summary

If the cancer is in only one eye and the potential for saving sight is good, chemotherapy (either by vein or directly into the artery of the eye) and local treatments such as cryotherapy laser therapy (photocoagulation), thermotherapy, or plaque radiation are used (although very small tumors, which are very rare, can be treated with local treatments alone). Otherwise the eye will likely need to be removed.

If the cancer is in both eyes, then the doctors will try to save as much vision as possible. The treatment usually starts with chemotherapy, followed by local treatments.

In all cases, children who have had retinoblastoma need to be followed closely for some time after treatment.

- References
See all references for Retinoblastoma

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