

OCULAR ONCOLOGY

The Ocular Oncology service at Bascom Palmer Eye Institute is dedicated to helping you and your family cope effectively from the time of the initial diagnosis of eye cancer through treatment and the follow-up period.

Because eye tumors are so rare, patients may have to travel long distances to seek expert opinion and treatment from an ophthalmologist who specializes in the diagnosis and management of eye cancer. Patients travel from all regions of the United States and from around the world because of Dr. Timothy Murray's years of experience in treating eye tumors, his international reputation, and his excellence in professional and compassionate patient care.

Bascom Palmer Eye Institute of the University of Miami School of Medicine is recognized throughout the world as a leading center for ophthalmic care, research and education. Its full-time faculty of internationally respected physicians and scientists encompasses every ophthalmic subspecialty and, through the years, has contributed fresh insight and direction to many of today's advanced ophthalmic techniques, diagnoses and treatments. Consequently, the facility has been rated one of the best eye hospitals in the nation for 12 consecutive years based on a survey of board-certified ophthalmologists conducted by U.S. News & World Report.

This information is intended to help patients and their families better understand a diagnosis of eye cancer and prepare for treatment. The information presented is for educational purposes only and should not be relied upon as medical advice. This information is also available on our website at www.eyecancermd.org.

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RETINOBLASTOMA



Cancer is always a frightening diagnosis, and when it affects children, the situation is overwhelming. Eye cancer in children, though rare, is the third leading cancer of childhood. Retinoblastoma, a cancer of the retinal eye tissue, is the most common eye cancer in children. It affects approximately one in eighteen thousand live births without regard to race or gender. This number, in fact, probably underestimates the number of cases because children remain at risk for retinoblastoma during the first five years of life.

Retinoblastoma can be a particularly distressing form of cancer as it is potentially fatal and can result in vision loss of one or both eyes.

Fortunately, almost all children in North America can be successfully treated for retinoblastoma. Not only are children no longer dying of this disease, in most cases, with early diagnosis and treatment, children with retinoblastoma do not have to have an eye removed and will keep normal vision.

When they first learn of their child's diagnosis, many families want to gather as much information as possible. Because it is such a rare disease, there is very little information about retinoblastoma available to families. This website is designed to give you more information about retinoblastoma. It will discuss diagnosis, treatment options, follow-up, and the genetic aspects of retinoblastoma. It will also include common issues faced by parents that have been faced with a diagnosis of retinoblastoma in their child. Our aim is to help families better understand and effectively cope with retinoblastoma.

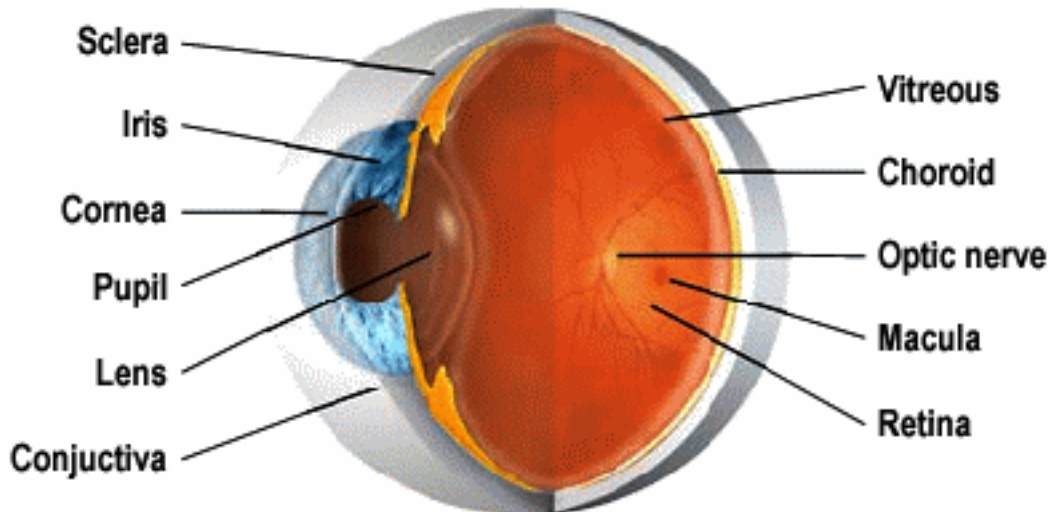
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ANATOMY OF THE EYE

In many ways, our eyes resemble an extraordinarily sensitive camera. Much like a camera lens the lens of the eye can change shape to focus on near or distant objects. The lens projects images on the retina, a layer of light sensitive cells on the back of the eye, functions much like the film in a camera.



The eye is connected to the brain by the optic nerve, a bundle of over one million nerve fibers. The cornea (the clear portion of the front of the eye) bends light rays through the pupil to the lens. The pupil is the black opening in the iris. The iris, the colored ring of tissue, regulates the amount of light entering the eye by adjusting the size of the pupil. The eye also has fluids that bathe the various parts of the eye and help to maintain the correct pressure within the eye. The anterior chamber, in the front of the eye, is filled with aqueous humor, a watery fluid. The back portion of the eye is filled with a clear jelly like substance, called the vitreous humor.

The eye can be divided into three layers (or tissues). The white outer coat of the eye, the sclera, is similar to the shell of an egg. Along with the cornea, the sclera protects the eye from trauma and maintains the delicate structure of the internal eye tissues. The middle tissue layer, the choroid, is the blood supply to the eye wall and outer retinal structures. This tissue is responsible for transporting nutrients and oxygen to the outer sclera and inner retinal tissue.

The innermost layer of the eye is the retina, a thin translucent tissue composed of ten microscopic layers. The retina is responsible for translating light images into electrical impulses that can be recognized and processed by the brain. The macula is the area of the retina that is responsible for central vision. The center of the macula is called the fovea, and it is responsible for very sharp vision. The retina receives images of light and transmits them to the brain via the optic nerve. The brain interprets these messages as sight.

DIAGNOSIS

We all know that life is always not fair and that there are no guarantees. This becomes painfully evident when a child is diagnosed with cancer.

There are two types of retinoblastoma. **Familial retinoblastoma** is hereditary, is passed from parent to child, and is bilateral (affects both eyes). Familial retinoblastoma represents 10% of cases. It is associated with a long-term predisposition to other types of cancer. The second type of retinoblastoma, responsible for 70% of all new cases, is **unilateral** (only one eye is affected). It represents the non-heritable form of the disease, and carries no increased risk of a second tumor.

Ninety percent of all retinoblastoma cases are diagnosed within the first three years of the child's life. On average, children with familial retinoblastoma typically are diagnosed at four months of age. When there is no family connection, the cancer is usually diagnosed when the child is approximately one to two years of age.

Well Baby Screening

Pediatricians can screen for vision and/or life threatening eye diseases. The minimal "well baby" screening for newborns is done during the first three months of life should include the following exams:

- **The Red Reflex:** checks for a normal red reflection in the eye that occurs when light travels inside the eye, hits the retina and the blood tissue, and is reflected back.
- **The Corneal Light Reflex:** when a light is shined into each cornea a symmetrical beam of light is reflected back in the same spot on each eye. This helps to determine whether the eyes are crossed.
- **An Eye Examination:** to check for any structural abnormalities. From six to twelve months of age, the eyes' ability to fix and follow objects both individually and together is evaluated. Between the ages of three and five, the examination of the eyes is coupled with testing for visual acuity, color vision and depth perception.

Parents also may want to look for eye abnormalities by directly examining each of the child's eyes under good lighting conditions. Both eyes should appear equal in size, be aligned, and should be able to move together and focus forward. Parents can observe the red reflex can be seen by dimming the room lights and using a flashlight to shine light directly into the child's eyes.

Unfortunately, because retinoblastoma is a rare cancer, some pediatricians may fail to detect it early enough or sometimes mistakenly diagnose it as:

- **Persistent hyperplastic primary vitreous (PHPV):** an unusual congenital anomaly whereby the eye is shorter than normal, develops a cataract, and may present with whitening of the pupil.
- **Coat's disease:** a typically unilateral disease, affecting young boys, leading to blood vessel abnormalities in the retina and retinal detachment.
- **Toxocara canis:** an infectious disease of the eye associated with exposure to infected puppies. This infection causes a retinal lesion leading to retinal detachment, which can mimic retinoblastoma.
- **Retinopathy of prematurity (ROP):** involves damage to the retinal tissue. It is associated with low birth weight infants who receive supplemental oxygen in the period immediately after birth.

Signs and Symptoms

Often the first sign of retinoblastoma that is noticed is **leukocoria**, a whitening of the pupil that looks like a "cat's eye". This whiteness can be seen in certain lighting conditions. It is often noticed in photographs of the child taken with a flash, which usually causes the eyes to appear red in the picture. Instead of the normal red reflex, you may notice a white pupil in the photo, which comes from the white surface of the tumor itself.

Examples of leukocoria:



Leukocoria is the most common sign of retinoblastoma and can be seen in 60% of patients. Other signs may include **strabismus** or crossing of the eyes, which is noticed in 20% of children. Furthermore, in approximately 10% of children, eye swelling with pain and redness occurs.

It is extremely important that a child suspected of having retinoblastoma be evaluated by a team of specialists, including an ocular oncologist, a pediatric ophthalmologist, a radiation oncologist, and a pediatric oncologist within an ocular oncology center. Children with this rare cancer require the most advanced testing and management to ensure the cure of the cancer with preservation of the greatest amount of vision.

Specialized testing is very important to confirm diagnosis, as there are no

blood tests available to confirm a diagnosis of retinoblastoma. Unlike tumors in other parts of the body, a biopsy cannot be performed due to the risk of spreading cancer cells outside the eye.

The Initial Eye Exam

Before the initial eye examination, your child may need a MRI or CT of the brain and orbits (eye sockets), with and without contrast (dye). These tests help to confirm the diagnosis of retinoblastoma and look for involvement of retinoblastoma with the optic nerve and any cancer outside of the eye.

Before the ophthalmologist examines your child, an optometrist or ophthalmic technician may take a medical and family history. Your child will receive eye drops that will dilate the pupils of the eye, which allows the ophthalmologist to examine the interior of the eye. The ophthalmologist will discuss his initial findings and possible treatment options.

The doctor may recommend an Examination Under Anesthesia (EUA) and any possible procedures that may be necessary during the EUA. Before the EUA a routine blood sample will be obtained to evaluate the components of the blood. Blood is obtained by a small prick made in a fingertip and a few drops are drawn off. If any of the tests are abnormal, more tests may be necessary to find out the reason. Tests may be performed to measure:

- **Hemoglobin:** the substance in the red blood cells that carries oxygen and is responsible for the blood's red color. Lower hemoglobin amounts than indicate anemia.
- **Hematocrit:** a measure of the amount of red blood cells, expressed as a percentage of the whole blood that is made up of red cells. A low count may indicate anemia.
- **Platelets:** the component of the blood that helps stop bleeding in case of injury.
- **White Blood Cell Count (WBC):** the components of blood that fight infection. Children receiving chemotherapy generally have a lower white cell count than normal.

Specialized Testing

Evaluation and treatment of children with retinoblastoma involves a variety of diagnostic testing. Many of these tests are performed when your child is first diagnosed and are repeated at intervals over the course of treatment to monitor progress and response to therapy.

Imaging Studies

Both a CT scan and a MRI require sedation to help a child lie quietly. There usually are no side effects from either of these tests, but children may be drowsy from the sedation. The difference between a normal eye and an eye containing retinoblastoma is very obvious, as you will see when the doctor shows you the x-rays. The CT scan or MRI usually shows the tumor(s) within the eye, since calcium accumulates in retinoblastoma tumors and is visible on the scan.

- **Computerized Tomography (CT):** A CT scan of the head is a computerized X-ray that provides a very clear picture of the eyes, the surrounding tissue and the brain. During the scan, the child's head is placed on a special headrest to allow proper positioning and to minimize movement during the scanning procedure. Unlike an ordinary X-ray machine, which takes one picture at a time, the CT scanner takes a number of small pictures as it rotates around the patient. This procedure takes about one hour.
- **Magnetic Resonance Imaging (MRI):** Magnetic Resonance Imaging uses magnetic fields and radio waves linked to a computer to create pictures of areas inside the body. The child lies on a table that slides into a tunnel-shaped piece of equipment. Antennas within the MRI machine pick up the radio waves within and feed them into a computer that assembles a picture. Because MRI can "see" through bone, it can provide clearer pictures of tumor located near bone and in the orbit. This procedure also takes about one hour.

Ultrasound (Echography)

During an ultrasound examination sound waves are directed towards the tumor by a small probe placed on the eye. Because tumors generate different "echoes" than normal tissue, sound waves above the range of human hearing can be bounced off tissue and then changed electronically into images. The pattern made by reflection of the sound waves helps to confirm that tumors are present and assists the doctor in establishing what type of tumor is present. Ultrasound also helps to determine the thickness or height of the tumor.



Bone Marrow and Lumbar Puncture

A bone marrow test or lumbar puncture may be performed as part of a child's initial EUA to make sure the cancer has not spread to the child's bones or brain. Both the bone marrow test and the lumbar puncture are completed in approximately 15 minutes. Usually there is only temporary tenderness at the site.

Bone marrow is the substance inside the larger bones of the body where healthy new blood cells are made. A small amount of the bone marrow is removed from the hip through a small needle and examined in the laboratory for cancerous cells. Since retinoblastoma can also spread into the brain, tumor cells may be floating in the cerebral spinal fluid (CSF) that bathes the brain and the spinal column (inside the backbone). To obtain the CSF, a small amount of fluid is drawn through a narrow needle at the base of the spine. The CSF is then tested for cancerous cells.

EXAMINATION UNDER ANESTHESIA (EUA)

Because a complete examination for retinoblastoma can be difficult for babies and young children, your doctor may recommend an Examination Under Anesthesia (EUA) as the best way to thoroughly examine your child's eyes. The EUA, may involve the following procedures:



- External examination of both eyes
- Internal examination of both eyes
- Drawings of the involved eye
- Photographs of both eyes
- Ultrasound
- Treatment procedures on the involved eye(s) such as cryotherapy or laser treatment
- Any necessary surgical procedures will be performed only after discussion with the family

Before your child undergoes an EUA your doctor may require a clearance for anesthesia from your children's pediatrician. Your doctor and hospital will give you complete instructions prior to the EUA, which probably will include no solid food for at least 8 hours before the procedure. Typically clear liquids (such as Gatorade, apple juice, grape juice, cranberry juice and water, but NOT milk or orange juice) may be given 6 hours before surgery, but after midnight, nothing to eat or drink.

Prior to the EUA, eye drops are administered to dilate the pupil. Because the eye drops sting (for about 30 seconds), your child may cry for a short time. Parents may want to help the nurses by holding the child still while the eye drops are given. This lets your child know that you are there. Usually after the eye drops are administered, the anesthesiologist will see your child and clear them for surgery.

After the anesthesiologist examines your child, your child will receive an injection to mildly sedate him or her. Typically a nurse will give this shot in your child's thigh about 10 minutes before your child goes to the Operating Room. Again, it is both helpful and comforting for you to hold your child while the shot is being administered. Once in the Operating Room, the anesthesiologist administers general anesthesia by placing a mask over the mouth or nose. Once the child is asleep, a tube may be placed in the throat to help regulate breathing.

When the procedure is finished, your child will be taken to the Recovery Room and remains there until fully awake. When your child returns from Recovery, he or she may be confused or upset. It is normal for a child to cry or be restless following an EUA. Many parents worry that the child is in pain. This is not usually the case. Generally, the child is reacting to the anesthesia wearing off, and your child may be a little disoriented during this time. Crying may actually assist in removing the anesthesia from your child's system and help make them more alert. The best thing you can do is to try to comfort your child and wait.



Following the EUA most children are either sleepy and/or hungry. If your child falls asleep, he or she may sleep for about an hour. If your child is hungry, you may offer clear liquids. Breast fed infants may be given breast milk. Bascom Palmer has some liquids (as well as Popsicles and crackers), but may not have your child's favorite drink so you might want to bring it with you.

Depending on what procedures are performed, your child usually will be ready to go home the same day. However, if any eye surgery is required, a night's stay in the hospital may be necessary. When your child is discharged, you will receive complete instructions and follow-up appointments.

TREATMENT OPTIONS

Your doctor will recommend treatment based on the size, location, and the extent of the tumor. Your child may have more than one type of treatment. For example, chemotherapy may be recommended as a primary treatment with laser therapy as a secondary treatment. It is critical that you discuss treatment options with your doctor, and that you understand the benefits and possible side effects of treatment.

Photocoagulation (Laser Therapy)

Laser therapy is performed on an outpatient basis during the EUA. Laser therapy focuses a powerful beam of light through the dilated pupil or through the wall of the eye onto the cancer and the surrounding tissue. This focused and high-powered light destroys cancerous tumors inside of the eye. The small burns produced by the laser destroy the blood supply to the tumor and cause the tumor to shrink. Multiple photocoagulation sessions may be necessary. Since laser therapy is done under anesthesia, there is generally no pain associated with the procedure. For this reason, post-operative medications typically are not necessary.

Depending on the location, small tumors can be treated with laser therapy alone. Tumors that are larger in size or involve the optic disc or macula may need the help of chemotherapy to reduce the size of the tumor. In this case, chemotherapy and laser therapy work together for tumor control.

Cryotherapy (Freezing Treatment)

Cryotherapy, which freezes small tumors, is performed in conjunction with an EUA. A probe (which looks like a pen) is placed on the sclera nearest to the tumor. Using a very cold gas, the tumor is frozen and thawed several times. This destroys the tumor cells and leaves a flat, pigmented scar with no signs of tumor. If tumor is evident, the treatment will need to be repeated to successfully destroy all of the tumor cells.

Cryotherapy can cause the eye and the lid of the eye to swell for several days. This is normal, and your doctor may advise the use of eye drops or ointment to reduce the swelling.

Chemotherapy

Chemotherapy is the treatment of cancer with drugs that can destroy cancer cells. These drugs are often called "anti-cancer" drugs. Normal cells grow and die in a controlled way. When cancer occurs, cells keep dividing and forming more cells without control. Anti-cancer drugs destroy cancer cells by stopping them from growing or multiplying. Because some chemotherapy drugs work better together than alone, often two or more drugs are given at the same time. This is called combination chemotherapy.

Typically, chemotherapy is used in addition to laser therapy, cryotherapy or radiation therapies in order to:

- Shrink a tumor before other therapies. This is called **neo-adjuvant therapy**.
- Help destroy cancer cells that remain after therapy. This is called **adjuvant chemotherapy**.
- Help destroy cancer if it spreads or recurs.

How often and how long your child gets chemotherapy depends on the size, location and the number of tumors in your child's eye. In addition, the specific drugs that are used, and how your child's body responds to the drugs are considered. Your child may receive treatment every few weeks or every month. Chemotherapy is often given in cycles that alternate treatment periods with rest periods. This gives your child's body a chance to build healthy new cells and regain strength. Sticking to the treatment schedule is VERY important for the drugs to work correctly. If you must miss a treatment session, you should contact your doctor immediately.

Chemotherapy can be given in several different ways, but is most often given intravenously (IV), through a vein. Because children who receive chemotherapy will likely be getting several cycles of drugs, a device may be surgically implanted which permits easy access to a vein. There are several devices, which may be considered.

Most commonly, a **port** is used. Ports are subcutaneous, meaning that they are placed under the skin. At one end of the port is a small metal chamber with a rubber top. This metal chamber lies under the skin on the right side of the chest, close to the collarbone. A flexible tube (called a catheter) is attached to the metal chamber, and is fed through the large blood vessel of the neck, and into the heart. Whenever chemotherapy drugs need to be given, a needle is inserted through the skin and into the rubber top of the port. The rubber portion on a port is self-sealing after needles are removed, and are designed to handle years of needle pricks. In addition, should blood need to be taken, or fluids, or medication delivered, the port may be used.

Types of Chemotherapy

There are three types of chemotherapy that usually are used with retinoblastoma:

- **Vincristine** (pronounced Vin-CRIS-teen): sometimes referred to as Oncovin or VCR. Vincristine is made from the periwinkle plant, a perennial, evergreen herb. Vincristine has anti-cancer properties because it causes cells to stop dividing.
- **Carboplatin** (pronounced car-BO-plat-in): also known as Paraplatin, is made from the metal, platinum. Carboplatin interferes with cell growth.
- **Etoposide** (pronounced E-TOE-poe-side): sometimes referred to as VP-16 or VePesid. Etoposide, works by stopping DNA (the part of the cell that directs reproduction). It also kills dividing cells.

Chemotherapy Side Effects

While chemotherapy kills cancer cells, healthy cells that divide at a rapid rate (like those that line the mouth, stomach, intestines, and hair follicles) may be harmed. Injury to these healthy cells is what causes chemotherapy side effects. Fortunately, these injured cells usually repair themselves after chemotherapy is completed. Chemotherapy side effects often can be controlled by medication.



An important thing to remember about chemotherapy is that not every child reacts in the same way or experiences the same side effects. Some children have virtually no side effects to chemotherapy. In part, this is due to revolutionary new drugs, which have changed the way children react to chemotherapy. These drugs work to prevent problematic symptoms, making side effects less prominent.

If your child does experience side effects, the most common ones are loss of appetite, nausea and vomiting. Be sure to tell the doctor or nurse if your child nausea or vomiting last for more than a day, or if fluids will not stay down.

HELPFUL HINT: When the port is accessed by a needle, children commonly feel a slight prick. A cream or spray is available which can be applied to the port access area that numbs the skin prior to a needle poke.

Localized Plaque Radiotherapy

Another method of radiation therapy used in the treatment of retinoblastoma, localized plaque radiotherapy, was developed in the 1930's. In this type of treatment, a small radioactive plaque (disc) is custom made and attached to the eye. Plaque radiotherapy has advantages over external beam radiotherapy in that it delivers radiation in a more localized fashion, thereby minimizing exposure to other eye structures. It should be stressed that only a small percentage of patients with retinoblastoma are candidates for plaque radiotherapy.

Children must be hospitalized for this procedure. The procedure requires two operations: the first to insert the plaque and the second to remove it. Plaques stay attached to the eye for three to seven days while the child is in the hospital. After the plaque is removed in a second surgery, eye drops are prescribed to prevent swelling and infection.

Enucleation

Enucleation is the surgical removal of the eyeball, leaving eye muscles and the contents of the eye socket intact. This procedure is done when there is no other way to remove the cancer completely from the eye. Unfortunately, loss of vision for the eye removed is permanent because an eye cannot be transplanted.



Due to earlier tumor detection and improved and increased use of conservative eye-sparing treatments, there has been a significant decrease in the enucleation of patients with retinoblastoma over the past 40 years. However, enucleation is indicated for unilateral tumors that fill over half of the eye, or when there is extensive seeding of tumor into the vitreous, total detachment of the retina, or involvement of other eye structures by the tumor.

The removal of the eye is done under general anesthesia, and surgery takes approximately one hour. The eye is removed, and a spherical implant made of coral or hydroxyapatite is placed into the orbit. The implant is wrapped in donor sclera. This allows the blood vessels to grow into the porous coral material. The muscles that help give movement to the eye are sutured to the implant, which allows for some movement of the prosthesis. The eye is then patched and children go to the Recovery Room. Most children go home the same day with post-operative instructions.

Approximately six weeks after enucleation, your child can see the ocularist to begin fittings for a custom made prosthesis (artificial eye).

External Beam Radiotherapy (XRT)

Cancer cells are growing and dividing more rapidly than many of the normal cells around them. External beam radiotherapy, also called radiation therapy or irradiation, uses an invisible form of high-energy (like x-rays) to kill cancer cells or keep them from growing and dividing. Radiation therapy is used to treat cancer because cancer cells are growing and dividing more rapidly than many of the normal cells around them.

The radiation treatment is painless. It is similar to having an x-ray taken, except that the child needs to hold still for a few minutes longer. A large machine called a linear accelerator directs radiation to the precise area of the eye needing treatment. The treatment is given in doses measured in units called centigrays (cGy). A custom fit, immobilization device is used to ensure that the radiation beam is delivered to the precise location of the tumor.

Depending on the age of your child, sedation or anesthesia may be required to calm him/her during the procedure to make certain that they are perfectly still during radiation therapy. Most facilities use an anesthetic that allows your child to recover quickly. The treatment takes only a few minutes and can be stopped at any time if the child experiences any difficulty. The entire procedure generally takes from 30 to 60 minutes. Nausea and vomiting are occasional side effects of anesthesia, but are well controlled by anti-nausea drugs such as Zofran.

You will not be allowed in the room during treatment, as this would expose you to needless radiation. Younger children may find it frightening to be left alone in the room during therapy. It may be reassuring to explain that you are just outside the room. In some hospitals, viewing windows or closed circuit television allow you to watch your child during treatment. Your child may feel easier knowing that you can see him or her throughout the treatment. A trip to the radiation therapy room ahead of time may also help relieve fears about the treatment.

Radiation is usually given every day for a specific number of days. A radiation oncologist, a medical doctor specialized in using radiation as a treatment, will work along with your ophthalmologist to determine the amount of radiation to be given. The currently accepted protocol is 3,500 to 4,000 centigrays (cGy) of external beam irradiation delivered in divided doses over 4 to 5 weeks given once or twice daily. The radiation oncologist will explain how the radiation will be given, what radiation is and how it works, and discuss any potential side effects.

Your child will not be radioactive during or after radiation therapy. Neither you nor anyone else need fear contact with your child. The skin around the area receiving radiation may appear to be "sunburned" or red and sore. This will go away soon after the treatment has finished. Some hair may be lost from the back of the head. This loss is usually temporary. Hair growth will begin about 3 months after the radiation treatment is completed.

Retinoblastoma is extremely sensitive to radiation; however, radiation therapy can cause mild, short-term side effects, and sometimes there is permanent damage that may not be evident until months or years after treatment. External beam radiotherapy is most often used to treat patients with bilateral retinoblastoma who are not able to receive local treatment (such as laser or cryotherapy). Radiation is generally preferred when a tumor recurs after initial treatment, a tumor extends into the bones around the eye (orbit), when the second eye contains a tumor larger than 16 mm in diameter, when the tumor is near the optic disc or center of vision (fovea), when multiple tumors are present, or when there is extensive vitreous seeding. It may also be used to treat the eye socket after an eye has been removed if studies show extension beyond the area that had been removed to prevent further spread of retinoblastoma.

Complications from radiation treatment include cataract (radiation can damage the lens of the eye and cause a cataract), dry eye, and abnormal orbital bone development (mid-facial hypoplasia). An important long-term complication of external beam radiation is the possibility of developing radiation-induced tumors. Several studies suggest that patients with hereditary retinoblastoma have an increased incidence of secondary tumors, and that the incidence rate is further increased in those who receive radiation therapy. Your child's ophthalmologist or radiation oncologist can tell you more about long-term effects in relation to your child.

PROSTHESIS

The Ocularist: What to Expect During Your First Visit

On the first visit, the ocularist will measure your child's socket, look at the shape of the socket and match eye color. The conformer that was inserted during surgery is removed and is replaced by a temporary prosthesis. This temporary prosthesis is as close a fit and match as possible to your child's existing eye. The temporary prosthesis is used to help your child get used to wearing a prosthesis. In addition, it helps to shape the socket, and to aid in the healing and development of the socket.



Can you tell which is the artificial eye?

Although this first visit may be emotional, it is not painful for your child. It may be helpful to speak with other parents who have been through this experience.

Caring for the Prosthesis

Adjustments to the prosthesis will be necessary as your child grows. How your child's prosthesis is fit and maintained plays a major role in the development of the socket. Without constant maintenance, polishing and necessary adjustments, their orbit will not develop and look like the natural eye. If the prosthesis is not properly maintained, your child may not have normal anatomical function or a good cosmetic result as an adult. The importance of having a proper fitting prosthesis for anatomical function and appearance cannot be overstated.

Parents are taught how to handle and clean the prosthesis. As your child grows up, he or she will accept the care of the prosthesis as part of the daily routine. Many children learn how to take the prosthesis out and put it back in on their own. For children under the age of 6, enlargements usually are necessary every 6-12 months. Polishing should be done at least twice a year. This can vary for each child. Cleaning varies from patient to patient. If the child has allergies or a cold, there may be an increased amount of mucus and the prosthesis will need to be cleaned more often.

The Conformer Came Out...Now What?

Occasionally a frantic parent will call saying, "that little plastic lens just fell out of my child's eye." That clear plastic piece is called a conformer, and although it is very important and needs to be put back in, it is not something to be afraid of. The conformer was placed in the socket after the eye was removed. It allows the socket to heal and form a space for the artificial eye, or prosthesis, to be inserted. It is not unusual for a conformer to come out or for your child to take it out. However, it is very important that it is placed back into the socket as soon as possible.

To insert the conformer, wash it completely with soap and water, and rinse it very well before reinserting. Raise the upper eyelid with the thumb of one hand and gently slide the conformer under the upper lid, just enough to make room to push the conformer into the socket and slide the conformer into place. Eventually you will insert the artificial eye the same way you inserted the conformer. As you become comfortable inserting the conformer, your child will be more comfortable and fitting your child with the prosthesis will be much easier.

The Importance of Protective Eyewear

Any child who has retinoblastoma, especially those with a prosthetic eye, should wear eyeglasses with polycarbonate lenses regardless of visual status. Polycarbonate lenses are made of the strongest available optical material used in protecting the vision of your child's eyes or remaining functional eye.

Due to certain orbital changes, vision defects can be made less noticeable using prisms that are added to the lenses, which may not be corrected by fit alone. Your ophthalmologist can guide you in identifying the appropriate protective eyewear and prescribe the correct lenses that are needed. Additional eye protection may be needed if your child participates in certain sports like soccer, basketball, football or tennis.



COPING WITH CHILDHOOD CANCER

We all know that life is always not fair and that there are no guarantees. This becomes painfully evident when a child is diagnosed with cancer.

Our assumptions, security, and worldview are dramatically altered when the health of someone we love is threatened. As parents, the safety and security of our children is our responsibility. We teach them to brush their teeth to protect them from tooth decay. We teach them to stay away from strangers to protect them from being harmed or mistreated by other people. However, in reality, we have no control over so many things in our children's lives, and we cannot protect them against cancer. Subsequently, when a child is diagnosed with cancer, families often feel the same sense of helplessness, anger, depression and fear of people who suffer through any traumatic event.

Being told a child has cancer is certainly a traumatic event. The shock of diagnosis often results in an overwhelming number of intense emotions. Both patients and their families experience these emotions. Many people have heard about the emotional stages we go through in dealing with trauma: denial, anger, bargaining, depression, and acceptance. It is important to remember that we don't go through these stages in a neat and tidy progression. When you've gone through anger to bargaining, it does not mean that you will not be angry again at a later time. Many of these emotions reappear at different times during your child's treatment.

Denial can take many different forms. Some parents may keep going from doctor to doctor, looking for someone to tell them that the diagnosis of retinoblastoma is wrong. While denial may prevent some parents from getting their child treatment, it can also serve to protect you from the overwhelming impact of your child's diagnosis and keep you from being immobilized by your emotions.

Like denial, anger is a normal part of the healing process. You may be angry at yourself, your spouse, God, your doctor, and even with your ill child. It is important to remember that while feeling angry is normal, the ways you choose to express this anger may not be helpful.

It can be helpful to take a moment to recognize the source of your anger. When something so awful and beyond our control happens in life, it is understandable to want to find someone or something to blame. We want someone or something to be responsible for the pain we suffer.

Anger at yourself can take the form of guilt, another common and normal response to childhood cancer. This reaction may be a way of helping to cope with the necessity of accepting a situation that you want to change but cannot. Parents tend to blame themselves and physicians for delays in diagnosis. They may look for things that they may have done to cause their child's illness. Parents may ask: "What could I have done to prevent this?", "Was there something in the environment that caused this?", or "Why didn't I recognize the disease sooner?" Mothers may want to know if they did something wrong during pregnancy to cause the cancer.

Parents need to remember that, as far as scientist can determine, **nothing they did or didn't do caused their child's illness to occur.**

Making deals, such as "If my child gets better, I promise I will always be a perfect parent", is part of the bargaining stage. Depression, feelings of helplessness, hopelessness and sadness can occur throughout the cycle of grieving and beyond. Eventually, most parents come to accept the diagnosis and that nothing they did or didn't do caused their child's illness and that with their support and good medical treatment, their child will go on to have a happy, healthy life.

Taking Care of Yourself

It can be difficult to juggle the many roles parents have. In addition to being a parent, you may have job responsibilities and important relationships with your spouse, your parents, your siblings, and your friends. A child's illness can put additional demands on your finances, your time, your energy, and your emotions.

Think of times you may have been on an airplane and the flight attendant says that if there is a problem, you should place the oxygen mask over yourself before helping you child. The reason is that if you pass out from a lack of oxygen, obviously you will not be able to help anyone else. The same is true in any crisis. If you don't take care of yourself, emotionally and physically, you won't be able to take care of your child.

Eating healthy, exercise and rest are extremely important. Talking with family or friends about their feelings is helpful to some parents. However, sometimes family members are so overwhelmed with their own feelings that they cannot be helpful to you. Even well meaning friends may not know how to be of help to you. In addition, this is a time when your child may be particularly sensitive to your moods and feelings. While it is important for parents to talk about their feelings, expressing these feelings too strongly in front of your child may create problems. This does not mean that you should not let children know that sometimes you may be angry, worried, or sad. However, also let them know that you are an adult and is not their fault that you have these feelings and that it is not their job to comfort or take care of you. Children who feel it is their job to emotionally protect their parents may stop talking to their parents about their own feelings, thereby cutting them off from their most important source of support.

While expressing your feelings directly to your child may be problematic, parents do need to find healthy ways to deal with the emotional turmoil they may be experiencing. Many parents find counseling or a support group of parents dealing with similar problems to be very helpful.

Taking Care of Family

The repercussions of retinoblastoma, its treatment, and its outcome are felt throughout the family. It affects the daily functioning of each individual family member as well as the family system. Families, like other systems, seek an equilibrium, or balance. Think of the family system as being like a mobile. When one part of the mobile moves, all the other parts also move until balance is restored. When an external stressor, like cancer, impacts the system, the family must somehow adapt to the new stress, and reestablish a sense of balance.



Families are often unaware that their "system" is out of balance. Signs of family system imbalance include:

- An increase in family arguments
- Problems concentrating at work or school
- Mood swings
- Increased or decreased appetite
- Insomnia (problems falling or staying asleep) or hypersomnia (sleeping too much)
- A loss of interest in activities
- Fatigue or loss of energy

Illness also can be a drain on your family's financial resources. Even with insurance, there are likely to be some costs you will incur and many parents lose time from work during a child's treatment. Dealing with insurance companies, billing errors, and payments is a major stressor for many parents. Read your insurance policy carefully to better understand the benefits and claims procedures. Keep all hospital bills, doctors bills, insurance explanation of benefits, prescription records, other tax deductible receipts (tolls, parking, meals, hotels) and all medical correspondence in a separate file may help prevent a financial catastrophe. State and federal programs may be sources of financial assistance for some families.

When so much of a family's energy is focused on an ill child, it may be difficult to find the time to spend with other family members. If a child with retinoblastoma has older brothers or sisters, in addition to adjusting to a new baby in the family, they must adjust to having a baby who may require an extraordinary amount of parental time and attention. Most siblings worry about their sick brother or sister and may be afraid they or their parents will get cancer too. Despite their concern, it is natural to feel jealous of all the attention the child with retinoblastoma receives, to be angry and blame the sick child for the family turmoil, and then feel guilty about having these feelings. Even if you have carefully explained what is happening to your other children, you may need to tell them over and over again what is going on.

You may not have large amounts of time, but it is important to be available to listen, to let your children know that their feelings matter, and that they are loved and cherished every bit as much as their sibling with cancer.

Over time, most families are able to adapt to a child's illness by incorporating it and its effects into the way the family makes sense of the world. How quickly a family is able to do this is dependent upon many factors including:

- The prognosis, treatment and course of the illness
- The prior level of family functioning
- The amount of parental insight into their own, as well as their children's reactions
- The availability and use of social support systems

A variety of therapies and activities can provide families with the tools to move forward in the coping and recovery process. Even families that have adequate resources to cope "on their own" may find that periodic counseling can actually accelerate the recovery process.

For adults, adolescents, and older children, traditional family or individual counseling often is effective. Younger children may respond to art, play, and music therapy. These therapies can benefit the entire family as well as at the individual. At the family level, therapy can:

- Improve family communication
- Decrease the likelihood of one family member becoming identified as the source of the family's problems

For an individual, therapy can:

- Decrease feelings of isolation
- Provide an opportunity to ventilate feelings
- Improve individual coping abilities
- Improve socialization skills
- Improve interpersonal relationships with medical personnel and family members
- Provide a sense of control and mastery that may otherwise be lost while involved in ongoing treatment

When combined with social support and existing coping strategies, counseling and other therapies can have a significant impact in helping families recover from the initial trauma of having a child diagnosed with retinoblastoma. For further information about these therapies, feel free to contact **The Courtelis Center**, University of Miami at (305) 243-4129 or <http://www.miami.edu/courtelis-center>.

Taking Care of Your Child

Being a parent is tough enough without having the additional stress of cancer. While you probably have a whole medical team advising you on how to best care for your child's physical health, you may have fewer resources helping you with your child's emotional well being.



It is unlikely that very young children will have a good understanding of their medical condition and treatment needs. What they are more likely to understand is that a lot of people in white coats keep poking at them, that sometimes they feel bad, and that the important adults in their lives are in turmoil.

When so much of your focus is on a child's illness, it may be difficult to remember that when this crisis is over, your child will need the same life skills any child needs to be a well functioning adult. While it is natural for parents to overprotect and overindulge a child with retinoblastoma to "make up" for all they are going through, spoiling a sick child has the same result as spoiling a healthy child. A child will feel normal only if he or she is treated normally. You should expect the same age-appropriate skills and enact the same discipline as you would if your child didn't have cancer.

Good parents want to provide their children with the best possible experiences for learning and developing. Children with visual impairments need to enjoy the same day-to-day and special experiences that other children enjoy. All children need to learn to dress themselves, do chores, play, and get along with others. While a child with a visual impairment at first may need some extra help, you should expect your child to learn the skills he or she will need to live independently. Here are some suggestions that other parents have found to be helpful.

- Help your child learn to use sight, smells, sounds, surface changes, and textures can be used as cues to the environment. For example, the change from tile to rug may alert a child that he or she is moving from the kitchen to the family room, or cooking smells mean that it's time to eat.
- Create opportunities and challenges for your child to learn. Children cannot learn what they don't experience. Use real objects and experiences as much as possible. Help your child understand that things don't just happen. For example, milk doesn't just magically appear on the table. Let your child experience opening the refrigerator door, picking up the milk carton, pouring milk into a cup, and placing the cup on the table.

- Point out visual features of objects, for example a toy's color or texture. "Look" and "see" are perfectly good words to use even if your child is totally blind.
- Direct indoor lighting or sunlight coming from behind, over your child's shoulder, may help your child use his or her vision more effectively. Try to minimize glare from light reflection off an object as glare can make it more difficult to see things.
- It usually is easier to see bright colored objects placed against an obviously different background. You can change the color of a surface to provide greater contrast by placing a light or dark cloth over it.
- Try not to bombard your child with sensory overload. Too many toys or too much stimulation can make it difficult to focus on a task.
- Children with a visual impairment may not be able to see facial expressions or body language that tell them that they are being spoken to. Use your voice to convey meaning. Especially if there are others in the room, speak to your child by name. Teach your child to lean his or head toward the person who is speaking.
- Use hand-over-hand or co-active movements to introduce your child to a new skill.
- You might want to get an idea of what you are asking your child to do before you have your child do it. Try to experience the world for you child's point of view.
- Help your child learn on his or her own instead of always doing things for your child. Trust your child to succeed and encourage your child to work through frustration.

Going Back to School

Since 98% of children with retinoblastoma are diagnosed before the age of 5, returning to school may not be an issue for your child. However, retinoblastoma can leave your child with unique educational needs. Late side effects from treatment may include hearing or visual loss or social difficulties that can impact your child's learning.



Some children with retinoblastoma may need extra help in the school environment. The Individual with Disabilities Act (IDEA) requires that every public school provide free and appropriate education in the least restrictive environment to all handicapped individuals between the ages of 3 and 21 years. Public schools are required to provide an individually designed instructional program for every eligible child. At-risk infants and toddlers are eligible for early intervention programs. If services are denied, parents can challenge the decisions of the school system, with disputes being resolved by an impartial third party.

If you believe that your child would benefit from special education services, you may ask the school to develop an "Individualized Education Program" or IEP. Unfortunately, some school districts don't offer special services unless parents ask for them, and many parents are not aware what services are available. Once the school district has received your request for an IEP, they will need your written permission to evaluate your child. This evaluation will determine whether your child needs special education services. When the IEP evaluation is complete, the school district will review the evaluation with you and write an education plan that fits your child's needs.

You may want to schedule a conference before your child's first day of school. In some cases it may be helpful to share some basic information about your child's diagnosis and treatment. For example, if your child has had an enucleation, it can be helpful to explain that your child has a prosthetic eye and needs to wear protective eyewear. If your child has a visual impairment, seating in the front of the classroom may be beneficial. Explaining your child's situation may help open the lines of communication and decrease curiosity, discomfort or even teasing by other students.

Some parents may conceal their children's medical problems for fear that their child will be treated differently. This might unintentionally signal others that retinoblastoma is something to be ashamed of or feared. It's important not to focus solely on your child's limitations. Some teachers may need to be reminded that first and foremost your child is a student, not a patient, in the classroom.

CANCER RESOURCES

New England Retinoblastoma Support Group

Provides emotional support and resources for families with retinoblastoma. Information and referral, phone support, conferences, literature, advocacy.

Phone: 1-800-562-6265

Address: New England Retinoblastoma Parents Support Group, P.O. Box 317, Watertown, MA 02272

American Cancer Society

Health information, coping with cancer, new treatment and research, discussion groups, locate resources in your community.

Phone: 1-800-227-2345

www.cancer.org

Children's Cancer Association

Visit this site and request a copy of The Kids Cancer Pages. It is a comprehensive national resource directory on childhood cancer. It was created to help connect and network families to organizations, information, resources, and to each other on a national basis.

Phone: 1-503-244-3141

www.ChildrensCancerAssociation.org

National Cancer Institute

The U.S. Government's primary agency for cancer research and information.

Phone: 1-800-4-CANCER

www.nci.nih.gov

Young People With Cancer

This book provides parents with information on the most common types of cancer, treatments and side effects and common issues that arise when a child is diagnosed with cancer.

Phone: 1-800-4-CANCER (1-800-422-6237) and select the option to order publications Monday through Friday, 9:00 a.m. to 4:30 p.m. local time (within the U.S. and its territories).

The National Children's Cancer Society

Provides information, emotional and financial support to children with cancer and their families for expenses related to cancer treatment. The website has special links just for kids and information on healthy caregiving for families.

Phone: 1-800-532-6459

www.children-cancer.com

Eye Cancer Network

Educational, diagnostic and treatment information about ocular cancers.

www.eyecancer.com

Band-Aids and Blackboards

A website about growing up with a medical problem from the perspective of kids and teens.

www.faculty.fairfield.edu/fleitas/sitemap.html

National Eye Institute

The U.S. Government agency conducts and supports research to help prevent and treat eye diseases. The website provides health information in English and Spanish.

Phone: 1-301-496-5248

www.nei.nih.gov

Patient-Centered Guides

Childhood Cancer Survivors; A Practical Guide to Your Future

Childhood Cancer: A Parent's Guide to Solid Tumor Cancers

Books written to give survivors and their families the information they need.

www.patientcenters.com

Phone: 1-800-998-9938

Candlelighter's Childhood Cancer Foundation

A national non-profit membership organization whose mission is to educate, support, serve, and advocate for families of children with cancer, survivors of childhood cancer, and the professionals who care for them.

Phone: 800-366-2223

www.candlelighters.org

Visual Impairment Resources

Visually impairment can be described as partially sighted, low vision, hard-of-seeing, or blind based on the degree of visual impairment. A person is considered LEGALLY blind when the best-corrected visual acuity is 20/200, or the person's visual field is 20 degrees or less. Few legally blind persons have no sight; in fact, most have some remaining vision. Partially sighted or low vision generally refers to a severe visual impairment, not necessarily limited to distance vision. Low vision applies to all individuals with sight who are unable to read the newspaper at a normal viewing distance, even with the aid of eyeglasses or contact lenses. They use a combination of vision and other senses to learn, although they may require adaptations in lighting or the size of print, and, sometimes, Braille. Generally, the partially sighted individual has a central visual acuity range of 20/70 to 20/200 in the better eye with best correction by glasses.

Florida Division of Blind Services (DBS)

This state agency provides vocational rehabilitation and employment services for blind and visually impaired Florida residents. Services include school to work transition, job placement, counseling, vocational and academic training, orientation and mobility training, personal and social adjustments services and post-employment services. A part of DBS, the Florida Department of Education specialists from Children and Family Services serve eligible children who are blind or visually impaired so that they may reach educational and independent living goals.

Phone: 1-800-342-1330

www.myflorida.com/dbs

National Association for Parents of Children with Visual Impairments (NAPVI)

A national non-profit organization that enables parents to find information and resources for their children who are blind or visually impaired. NAPVI offers information and referral services, initiates outreach by providing seed money to promotes the development of state and local organizations, has a quarterly publication, advocates for educational needs, and offers emotional support and networking opportunities.

Phone: 1-900-562-6265

www.spedex.com/napvi

V.I. Guide

An Internet site that contains information on topics is pertaining to parenting and teaching children with a visual impairment. Information and links to other sites about vision-related services, special education, assistive technology and products, laws and legislation, entertainment, research, and other organizations and groups that might be of interest.

www.viguide.com

The Hadley School for the Blind

The Hadley School offers Parent/Child and Parent/Family correspondence courses on a variety of topics. Parent/Child courses include: Reach and Teach, which provides step-by-step teaching techniques and guideline to help parents assess and build on a child's strengths; Child Development; Partners in Education; Learning, Play, and Toys; and Early Independence. Parent/Family course topics include Abacus I and II; Accessing Local Sports and Recreation; Braille Reading for Family members, College Bound, the Human Eye; Independent Living; Personal Computers; self-esteem.

Phone: 1-800-323-4238

www.hadley-school.org

Blind Children's Center

This Los Angeles organization has a number of videos and publications (some in Spanish) that can provide useful information for families of blind or visually impaired children. Topics include: beginning nurturing, social, play and language interactions; teaching young children who are visually impaired; common concerns for the preschool child who is visually impaired or blind; feeding a child with a visual impairment; movement development for babies who are visually impaired; orientation and mobility for preschool children designing and constructing simple adaptive mobility devices for preschool children; multiple disabilities; and parents talk about their feelings.

Phone: 1-800-222-3566

www.blindcntr.org

Prosthesis

SNG Prosthetic Eye Institute

www.prostheticeye.com

Other Organizations

American Council of the Blind

Phone: 1-800-424-8666

www.acb.org

American Federation for the Blind

Phone: 1-800-232-5463

www.afb.org

American Printing House for the Blind

www.aph.org

Lighthouse International

www.lighthouse.org

National Federation of the Blind

www.nfb.org

Links to Other Websites

Eye Resources on the Internet

webeye.ophth.uiowa.edu/dept/websites/eyeres.htm

International Organization for the Blind

www.io4b.org

Blindness Resource Center

www.nyise.org/blind

A Blind Net

www.blind.net

Snuggl' Up, Inc

Phone: 305-931-6333

www.snugglup.com

Travel

Angel Flight

Angel Flight is a non-profit, volunteer pilot organization that coordinates free air transportation on corporate aircraft for those with medical needs.

Phone: 1-800-352-4256

www.angelflightse.org

Midwest Express Miracle Miles

One free flight, thereafter flights are discounted. No financial requirements.

Phone: Call Natalie Fuerst at 1-414-570-3644

Corporate Angels

Private jets used by corporations that have empty seats, free flights. No financial requirements.

Phone: 1-914-328-1313

www.corpangelnetwork.org

Northwest Airlines, Kid Cares Program

Those with financial need have priority.

Phone: 1-612-726-4206

www.nwa.com/corpinfo/aircares/about/kidcares.shtml

American Airlines, AAdvantage Miles for Kids in Need

Helps those in financial need.

Phone: 1-817-963- 8118

The National Patient Travel HELPLINE

Helps those in financial need.

Phone: 1-800-296-1217

www.patienttravel.org

Housing

American Cancer Society's Winn-Dixie Hope Lodge

The American Cancer Society understands the challenges that families face when a loved one needs cancer treatment far from home. It can be uncomfortable and extremely costly to stay in hotels for weeks at a time. That's where Hope Lodge can help. It offers a friendly environment with no limit on the length of stay. Hope Lodge provides several kitchen areas where families can cook their own meals. While Hope Lodge does not provide medical, counseling, or religious services, it is far more than a hotel. It's a home-away-from-home when it's needed most.

Phone: 1-800-ACS-2345; In Miami: 305-547-2210; Other areas: 1-317-415-5000

Ronald McDonald House

Since 1974, families around the world have been calling Ronald McDonald House a "home-away-from-home."

Address: Ronald McDonald House Charities, One Kroc Drive, Oak Brook, Illinois, 60523

Phone: 1-630-623-7048

www.rmhc.com

CONTACT INFORMATION

Mailing Address:

Bascom Palmer Eye Institute
University of Miami School of Medicine
Ocular Oncology Service
Timothy G. Murray, MD
900 Northwest 17th Street
Miami, Florida 33136

Phone Number:

305-326-6000

Toll Free:

800-329-7000

Fax:

305-326-6551

Email:

For questions about retinoblastoma, contact us at retinoblastoma@eyecancermd.org. For all other questions or to request an appointment at Bascom Palmer Eye Institute please visit: www.bpei.med.miami.edu/contact/index.asp.

Please note: It would be inappropriate for us to try to diagnose or treat any illness via e-mail. Please visit a doctor to receive the attention and care necessary for proper treatment.