

*A Parent's
Guide to
Understanding*

Retinoblastoma



IRIS Medical *Education*
Understanding Retinoblastoma

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GLOSSARY OF TERMS

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Aqueous Humor - watery fluid which bathes and nourishes the front of the eye

Bilateral Retinoblastoma - cancerous tumor(s) in the retina of both eyes

CAT Scan - test which uses x-rays to view the eyes and brain

Choroid - the middle layer of the eye which contains blood vessels

Chromosome 13 - the chromosome which has a missing piece that is responsible for the development of retinoblastoma

Conjunctiva - thin membrane which lines the outside of the eye

Cornea - clear portion of the front of the eye which bends light rays

Cryotherapy - freezing treatment for small retinoblastoma tumor(s)

Disc Diameter - horizontal size of the optic nerve head approximately 1-1.5 mm used as a reference point to measure tumors

Enucleation - surgical removal of the eye

Equator - a circular reference zone approximately half way between the back of the eye and the front of the eye

External Beam Radiation - treatment which uses machines to give radiation to treat the tumors

Fundus Drawing - map of the eye with tumor sketches drawn by the ophthalmologist

Fundus Photographs - a photograph of the inside of the eye showing the retina

Indirect Ophthalmoscope - instrument used to view the retina

Iris - the colored portion of the eye

Lamina Cribrosa - the zone in the optic nerve which represents the anatomical end of the eye

Laser - light therapy used to treat small retinoblastoma tumor(s)

Leukocoria - white pupillary reflex; cat's eye reflex; the most common presenting sign of retinoblastoma

Macula - the area of the retina responsible for central vision

Metastasis - spread of a cancer to other parts of the body

Ora Serrata - portion of the retina near the front part of the eye

Optic Nerve - structure which carries impulses from the retina to the brain for interpretation

Pediatric Oncologist - physician who specializes in cancer of children

Prognosis - the overall outlook of treatment

Pupil - black hole in the center of the iris

Radioactive I-125 Plaque - a disc of radiation that is sewn to the eye to treat the retinoblastoma tumor(s)

Retina - inner light-sensitive layer of the eye

Retinoblastoma - a cancerous tumor of the retina of the eye

Sclera - the outer protective white coating of the eye

Strabismus - crossed eyes; second most common presenting sign of retinoblastoma

Ultrasound - test which uses sound waves to view the eye and tumor(s)

Unilateral Retinoblastoma - cancerous tumor(s) in the retina of one eye

Vitreous Humor - the transparent gel that nourishes the back of the eye

Vitreous Seeding - small balls of retinoblastoma that break off and extend into the vitreous



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WHAT IS RETINOBLASTOMA?

Retinoblastoma - (*Reh-tin-oh-blast-oma*) is a cancer of one or both eyes which occurs in young children. There are approximately 350 new diagnosed cases per year in the United States. Retinoblastoma affects one in every 15,000 to 30,000 live babies that are born in the United States. Retinoblastoma affects children of all races and both boys and girls.

The retinoblastoma tumor(s) originate in the retina, the light sensitive layer of the eye which enables the eye to see. When the tumors are present

in one eye, it is referred to as **unilateral** retinoblastoma, and when it occurs in both eyes it is referred to as **bilateral** retinoblastoma. Most cases (75%) involve only one eye (unilateral); the rest (25%) affect both eyes (bilateral). The majority (90%) of retinoblastoma patients have no family history of the disease; only a small percentage of newly diagnosed patients have other family members with retinoblastoma (10%).

This booklet will help you understand the eye and the diagnosis and treatment of retinoblastoma.



STRUCTURE & FUNCTION OF THE EYE

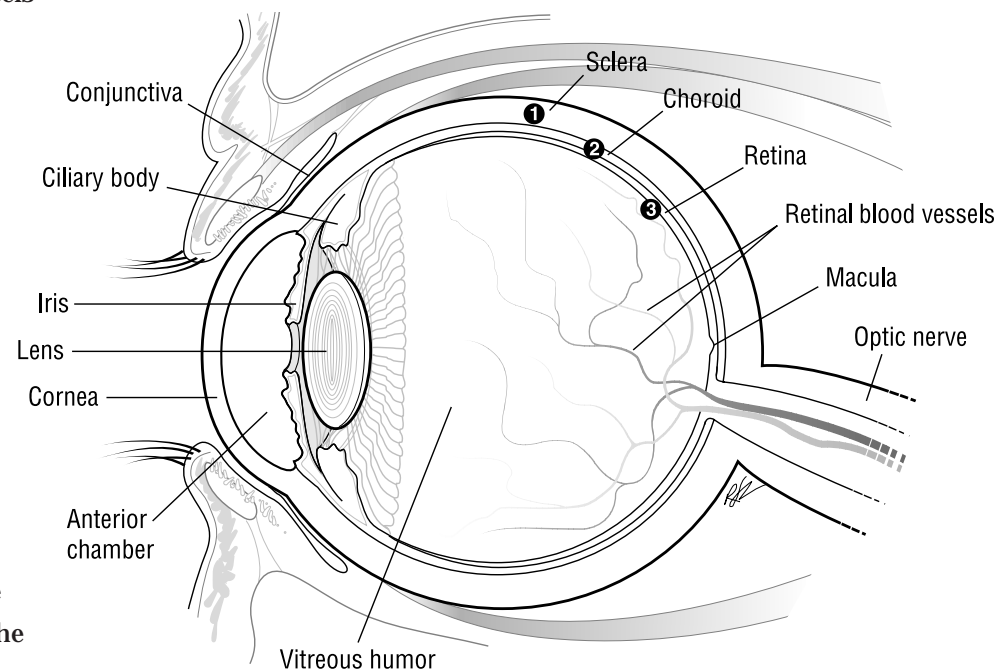
The eye of an adult measures about one inch from the front to the back of the eye; a child's eye measures about three-quarters of one inch.

The eye has three layers:

- 1) **Sclera** - the outer protective white coating of the eye
- 2) **Choroid** - the middle layer which contains blood vessels to nourish the eye
- 3) **Retina** - the inner layer which contains the nerves that bring information to the brain for seeing

The **cornea** is the clear portion of the front of the eye which bends light rays. The **conjunctiva** is a thin tissue which lines the eyelids and the eyeball up to the edge of the cornea. The **iris** is the

colored portion of the eye which is made up of a spongy tissue and is an extension of the choroid. The **pupil** is the opening in the iris (black) which allows light into the eye. The **lens** helps focus light rays onto the retina the way a camera lens focuses light onto film; the lens can change shape, or accommodate, to focus on near or distant objects.



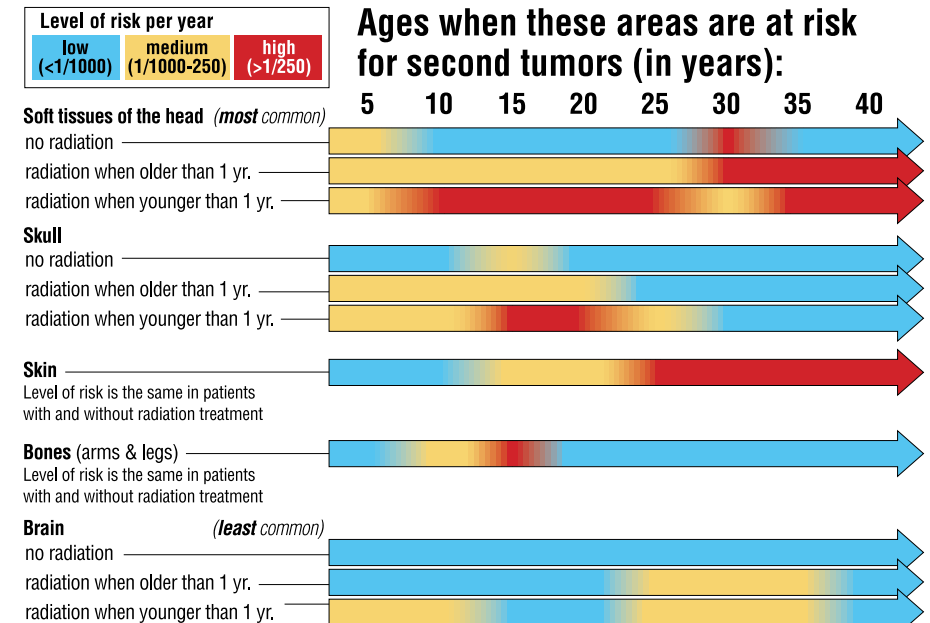
found it very helpful to talk to other parents who share similar concerns. Some institutions have newsletters or formal support groups for parents of children with retinoblastoma. Finally, some institutions have programs which can make the child's return to school, home and the community a bit easier.

Retinoblastoma is a life-threatening disease, but it is rarely a fatal one if treated appropriately. With

the correct treatment in the hands of an experienced ophthalmologist and appropriate follow-up both for eyes and for other cancers, the retinoblastoma patient has a very good chance of living a long, full, and happy life.

This chart shows the ages when parents and patients need to be aware of potential secondary cancers. Statistical analysis of over 40 years of cases has helped doctors recognize when and where these cancers develop.

A reference card explaining this and other important information may be available at your doctor's office.



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LONG TERM CONSEQUENCES OF RETINOBLASTOMA

The majority of children in the United States (over 95%) survive the cancer and have perfectly normal lives. All the children with unilateral retinoblastoma have one normal eye whose sight is not affected even though they may have had one eye removed. Children with one eye have normal vision, play sports, and later drive cars. It is, however, especially important for children with vision in only one eye to wear protective eyewear during sports and other hazardous activities. These children grow up and become famous actors, physicians, lawyers, nurses, accountants and parents themselves.

The majority of children with bilateral retinoblastoma retain at least one eye with good vision and many are able to retain the use of both eyes. They also live normal lives, go to school, enjoy life, have careers and families themselves. However, all children with bilateral disease and the 15% of unilateral patients who have the familial form of retinoblastoma will be at much higher risk for other cancers not involving the eyes throughout their lives. Five years after the initial diagnosis of retinoblastoma, more children with the genetic form

of retinoblastoma have died from these second tumors than from the original retinoblastoma from the eye. The most common second tumors are osteogenic sarcoma, a cancerous tumor which affects the bones, soft tissue sarcomas and cutaneous melanomas (tumors of the skin, muscle, and connective tissue). Although the reported incidences of these tumors vary widely, the risk appears to be about 1% a year.

Follow-up appointments are very important when a child is diagnosed with retinoblastoma. All children should be followed by an ophthalmologist and by a pediatric oncologist. Frequency of examinations depends upon the age of the child, the ophthalmologist's suspicion of new tumors, the involvement of one or both eyes, and the type of treatment that the child has received. Mothers and fathers are encouraged to talk to the nurse and to call with questions between visits. An example of the follow-up appointment schedule is listed on page 13.

Many parents meet other parents in the waiting room of the physician's office, clinic, or hospital who also have children with retinoblastoma. Some have

The eye is filled with fluids which help nourish and maintain the pressure within the eye. The **anterior chamber**, the front portion of the eye between the iris and the cornea, is filled with **aqueous humor**, a watery fluid, which nourishes the lens and maintains the pressure within the eye. The back portion of the eye is filled with **vitreous humor**, a transparent gel. The **retina** is made up of ten layers and contains over one million cells. The **optic nerve** has nerve fibers which transmit information to the brain for interpretation of objects seen.

The **macula** is the area of the retina that is responsible for central vision; its central portion is referred to as the **fovea** and is responsible for the sharpest vision. The macula houses the highest concentration of the cones which are responsible for color and sharp vision. The rods, which compose the rest of the retina, are more sensitive to light and are responsible for night vision and peripheral vision. Attached to the globe of the eye are six muscles which aid in the movement of the eye. Movement of the eye may be caused by one, a few or all of the eye muscles working together.



SIGNS & SYMPTOMS OF RETINOBLASTOMA

Retinoblastoma can present in a variety of ways. The majority of retinoblastoma patients present with a white pupil reflex or **leukocoria** instead of a normal healthy black pupil or a red reflex similar to the one seen when photographs are taken of a child looking directly into the camera. This abnormal white pupillary reflex is sometimes referred to as a cat's eye reflex (below).

Many times the parent is the first one to notice the cat's eye reflex. Other eye diseases can also present with this white pupillary reflex; leukocoria does not always indicate retinoblastoma. An ophthalmologist can determine the correct diagnosis.



A crossed eye or **strabismus** is the second most common manner in which retinoblastoma presents.

The child's eye may turn out (towards the ear), called **exotropia**, or turn in (towards the nose), called **esotropia**.

Retinoblastoma may also present with a red, painful eye, poor vision, inflammation of tissue surrounding the eye, (above), an enlarged or dilated pupil, different colored irides (heterochromia), failure to thrive (trouble eating or drinking), extra fingers or toes, malformed ears, or retardation. On rare occasions, retinoblastoma is discovered on a well-baby examination. Most often, the symptoms of retinoblastoma are first detected by a parent.



aiming the light through the wall of the eye and not through the pupil. Laser treatment is done under anesthesia and usually does not have any post-operative pain associated with it and does not require any post-operative medications. Laser can be used alone or in addition to radiation or cryotherapy.

Cryotherapy is another treatment which is performed under local or general anesthesia and freezes smaller retinoblastoma tumors. A pen-like probe is placed on the sclera adjacent to the tumor and the tumor is frozen. Cryotherapy usually has to be repeated many times to successfully destroy all of the cancer cells. Cryotherapy causes the lids and eye to swell for 1 to 5 days; sometimes the swelling is so much that the children are unable to open their lids for a few days; this can be frightening for the child and parents, but is usually harmless. Eye drops or ointment can be given to reduce the swelling.

Chemoreduction is the treatment of retinoblastoma with chemotherapy. Chemotherapy is given intravenously to your child. The drug passes through the blood stream and as it passes through the eye it causes the tumors to shrink within a few weeks. Chemotherapy, with one or more drugs, can be given once, twice, or more. Depending on the drug(s) and on the institution, the child may or may not be hospitalized during this process.

After chemotherapy, the child is re-examined and the remaining tumor is treated with cryotherapy, laser, or radioactive plaque. Children may require as many as twenty treatments with re-examinations of the eye under anesthesia every 3 weeks.

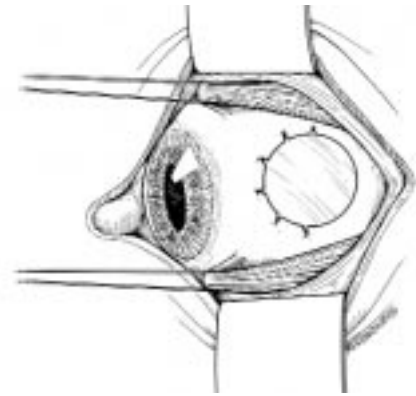
Although it is rare if the retinoblastoma is treated promptly, retinoblastoma can spread, or metastasize, outside of the eye to the brain, the central nervous system (brain and spinal cord), and the bones.

Chemotherapy is prescribed by a pediatric oncologist and is administered through the peripheral blood vessels or into the brain for months to years after initial diagnosis of metastatic disease.



Cryotherapy treatment for retinoblastoma

GENETICS OF RETINOBLASTOMA



Radioactive plaque attached to the eye

shrink, because the pinkish-grey tumor mass is replaced by white calcium. Immediately after treatment, the skin may be sunburned or a small patch of hair may be lost in the back of the head from the beam exit position. Following external beam radiation, long-term effects can include cataract, radiation retinopathy (bleeding and exudates of the retina), impaired vision, and temporal bone suppression (bones on the side of the head which do not grow normally).

Radioactive plaque are discs of radiation that were developed in the 1930's to radiate retinoblastoma. Today, the isotope iodine-125 is used and the plaques are custom built for each child (top). The child must be hospitalized for this procedure and undergoes two operations (one to insert the plaque and one to remove the plaque) over 3 to 7 days. No pregnant visitors or health care

professionals may visit or care for the child, and children under twelve years old are not permitted to visit according to state guidelines. Patients take eye drops following plaque surgery for 3 to 4 weeks to prevent infection and inflammation. Following the plaque, long-term effects including cataracts, radiation retinopathy, and impaired vision may occur.

Laser therapy is a non-invasive treatment for retinoblastoma. It can be performed on an out-patient basis under local or general anesthesia. Lasers very effectively destroy smaller retinoblastoma tumors. This type of treatment is usually done by focusing light through the pupil onto and surrounding the cancers in the eye. Recently a new delivery system of the laser, called a DioPexy™ Probe, has enabled treatment of the cancer by



Ninety percent (90%) of all children who develop retinoblastoma are the first one in their family to have eye cancer. In 10% of retinoblastoma cases, however, a parent, grandparent, sibling, uncle, aunt, or cousin also had retinoblastoma. When retinoblastoma is passed from parent to child, the disease is usually, but not always, bilateral. Much work has been done in the past 10 years to figure out how a genetic abnormality causes cancer. The genetic patterns and their implications for you and your family are listed on page 7.

Although it is not exactly understood why retinoblastoma occurs, it is known that in all of the cases this cancer is caused by an abnormality in chromosome 13, in which a piece of the chromosome is nonfunctional or missing. In 40% of the cases, the abnormality is present in every cell of the body including the eye and in 60% of cases, the abnormality is only found in the eye. Chromosome 13 is responsible for controlling retinal cell division. In children with

retinoblastoma retinal cell division continues unchecked, causing the retinal tumor(s).

If a parent had bilateral retinoblastoma:

If a parent has been treated for bilateral retinoblastoma and decided to have children, almost half (45%) of their children will develop retinoblastoma in their eyes. The child may have tumors in the eye at birth and may even have tumors that have spread through the body and into the brain at birth. On the other hand, many of these

Cancer Facts

- Cancerous growths have been discovered in Dinosaurs!
- Retinoblastoma was documented in children more than 2,000 years ago.



children do not have tumors in the eye at birth and develop them during the first few years of life. In our experience, all of these children begin to develop tumors in the eyes by 28 months and can continue to form them for 7 years.

The overwhelming majority of children born to a parent with bilateral retinoblastoma will also have bilateral retinoblastoma, but about 15% will develop tumors in only one eye (unilateral retinoblastoma).

Every time the bilateral retinoblastoma parent has another child, the chance of that child developing retinoblastoma is 45%.

If a parent had unilateral retinoblastoma:

If a parent had unilateral retinoblastoma, 7% to 15% of their offspring will have retinoblastoma. Interestingly, when a parent with unilateral retinoblastoma has a child who develops retinoblastoma, that child will usually (85% of the time) develop bilateral retinoblastoma. Many of these affected children do not have the tumor present at birth. But as with the situation above, if the child is going to develop retinoblastoma, they will begin to develop tumors by 28 months and can continue to form them for 7 years.

Every time the unilateral retinoblastoma parent has another child, the chance of that child developing retinoblastoma is 7% to 15%. On the other

hand, 85% to 93% of the offspring of a unilateral retinoblastoma parent will not develop the disease.

If neither parent had retinoblastoma:

The situation where neither parent has had retinoblastoma is the most common and the most difficult to explain. Approximately one out of every 100 patients who has retinoblastoma appears to somehow destroy the tumor without treatment or perhaps has a form of the disease which is not capable of spreading. This is why both parents are encouraged to have a dilated retinal exam. One percent of the time there is evidence of a cured or limited form of retinoblastoma in the eye of a parent who was never aware of having had retinoblastoma and was never treated for it. In that case, 45% of that parent's children will develop retinoblastoma.

More confusing, and rarer is the possibility that a parent has the gene for retinoblastoma and can pass it on to his or her children without having any evidence of retinoblastoma in the eye. This is called the carrier state and it means that the parent carries the gene but not the disease. In this case, 45% of their children will be affected. Each of the examples listed here explains how a parent who never had retinoblastoma can have many children affected with bilateral retinoblastoma.

The prosthesis is made of plastic by an ocularist (an artist/technician) to look exactly like the other eye. Since the extraocular muscles are not attached to the prosthesis, it does not move as well as a natural eye. The prosthetic eye tends to move better up and down than it does side to side. And of course, the prosthetic eye does not see. Unfortunately, there is currently no way to transplant or replace an entire eye.

When both eyes are involved, sometimes the more involved or "worse" eye is enucleated, while the other eye may be treated with one of the vision preserving treatments such as external beam radiation, plaque therapy, cryotherapy, laser treatment, and chemoreduction which are described below.

External beam radiation has been used since the early 1900's as a way to save the eye(s) and vision. Retinoblastoma is sensitive to radiation and frequently the treatment is successful. The radiation treatment is performed on an outpatient basis five times per week over a 3 to 4 week stretch. Custom-made plaster-of-paris molds are made to prevent the head from moving during treatment and sometimes sedatives (medicines which help relax the children) are prescribed prior to the treatment.



Laser treatment for retinoblastoma

Tumors usually get smaller (regress) and look scarred after external beam radiation treatment but they rarely disappear completely. In fact, they may even become more obvious to the parent as they

1959 Dr. G.M. Schwickerath (Essen, Germany) First Case of Retinoblastoma treated with Photocoagulation (Laser)

1967 Dr. H. Lincoff (New York, USA) First Case of Retinoblastoma treated with Cryotherapy

1996 Dr. D. Abramson (New York, USA) First Case of Retinoblastoma treated with DioPexy Laser

1996 Dr. D. Abramson (New York, USA) First Case of Retinoblastoma treated with Local (Peri-ocular) Chemotherapy

TREATMENT OF RETINOBLASTOMA

The treatment of retinoblastoma is individualized for each patient. The treatment depends upon the age of the child, the involvement of one or both eyes, and whether or not the cancer has spread to other parts of the body. Almost all parents choose some form of treatment for their child. No treatment may mean that the child would die. Goals of treatment from most to least important ranking are: saving life, maintaining the eye and vision, and preserving cosmetic appearance.

Enucleation (Eee-nuke-lee-a-shon) is the most common form of treatment for retinoblastoma. During an enucleation, the eye is surgically removed. This is necessary because it is the only way to remove the cancer completely. It is not possible to remove the cancer from within the eye without removing the entire eye. This can be done for some other eye cancers but is dangerous and may even contribute to the spread of the cancer for retinoblastoma patients.

The removal of the eye is done under general anesthesia (the child is asleep). A parent may

accompany the child to the operating room on the day of surgery and may be present until the child is anesthetized. The anesthesiologist will place a mask over the child's mouth and nose. Children usually fall asleep within a minute or two of breathing the anesthesia gases.

In the operation, the entire eye is removed along with a long piece of the optic nerve as one specimen, and is sent to a pathologist for examination under a microscope. The eyebrow, lids and muscles of the eye are all left in place. Blinking, tearing and movement of the brow are not affected from this surgery. The operation takes less than an hour and is not painful. Children go home the same day and are usually examined in the office on the following day when post-operative instructions and care are explained.

A ball of plastic, rubber or coral is placed where the eye had been so there is no cavity or hole. After the socket heals, it will look like the tissue on the inside of the lip. The child is fit for a prosthesis or false eye approximately 3 weeks after the operation.

1767
Dr. R. Hayes (London, England)
First Case of Retinoblastoma
treated with **Enucleation**

1903
Dr. H. Hilgartner (Texas, USA)
First Case of Retinoblastoma
treated with **Radiation**

1933
Dr. Foster-Moore (London, England)
First Case of Retinoblastoma
treated with **Radioactive plaque**

1953
Dr. C. Kupfer (Maryland, USA)
First Case of Retinoblastoma
treated with **Chemotherapy**

The most common situation is where neither parent has the gene for retinoblastoma but has a child born with retinoblastoma. If the parents are

genetically normal, the chance of another child having retinoblastoma is 1 in 15,000 to 1 in 20,000.

This chart shows the likelihood of a parent to give the retinoblastoma to a child. A reference card explaining this and other important information may be available at your doctor's office.

If Parent Was...

| | Bilateral | | | Unilateral | | | Unaffected | | | | | |
|--|-----------------|----------------|----------------|----------------|-----------------|-------------------|---------------|----------------|-----------------|----------------|--------------|----|
| Chance of offspring having retinoblastoma | 45% affected | | 55% unaffected | 7-15% affected | | 85-93% unaffected | <<1% affected | | 99% unaffected | | | |
| Laterality | 85% bilateral | 15% unilateral | 0% | 85% bilateral | 15% unilateral | 0% | 33% bilateral | 67% unilateral | 0% | | | |
| Focality | 100% multifocal | 96% multifocal | 4% unifocal | 0% | 100% multifocal | 96% multifocal | 4% unifocal | 0% | 100% multifocal | 15% multifocal | 85% unifocal | 0% |
| Chance of next sibling having retinoblastoma | 45% | 45% | 45% | 45% | 45% | 45% | 7-15% | 5%* | <1%* | <1%* | <1%* | |

*If parent is a carrier, then 45%



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Retinoblastoma Facts

- More than 95% of patients treated for retinoblastoma in the U.S. are cured
- More than 90% of patients retain at least one eye
- More than 80% of the children keep 20/20 vision
- Complete blindness is unusual in children who have been successfully treated for retinoblastoma

GENETIC TESTING

The gene responsible for retinoblastoma has been identified and studied in great detail. Approximately 40% of patients have the abnormal gene responsible for retinoblastoma.

There is now a test to detect the genetic defect. At the present time, the test is most accurate if the lab can study a specimen of tumor from the enucleated eye of the patient together with a blood sample. The test is technically more difficult when only blood specimens are available and is more straight forward in patients who have bilateral retinoblastoma than in patients with unilateral retinoblastoma. If the patient has the genetic abnormality, then other members of the family may



need to be screened or observed for development of the disease.

Did you know?

- Retinoblastoma is a cancer
- Retinoblastoma is the most common eye cancer in children
- Retinoblastoma is about as common as hemophilia
- Retinoblastoma affects boys as commonly as girls
- The average age for diagnosis of retinoblastoma when one eye is involved is 2.5 years
- There are 350 new cases of retinoblastoma per year in the U.S. and about 5,000 new cases worldwide
- Retinoblastoma affects all races equally

You should speak to your doctor or nurse about the usefulness and cost regarding the procedure involved with genetic testing.



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Nursing Issues

- It's okay to feel frightened and overwhelmed; this is a normal or common reaction to the diagnosis of cancer.
- You may find support by talking to other parents and children in the waiting room.
- Check with your local institution for support groups, one-on-one counseling, and newsletters. Many families have found these resources helpful.
- Children are remarkably adaptable. Your child may cope better than you!

EXAMINATION SCHEDULE FOR RETINOBLASTOMA SUSPECTS

FIRST EXAMINATION:

When: Within 24 - 48 hours of birth
Where: Newborn Nursery
By Whom: Ophthalmologist at your hospital; Contact pediatrician for referral
Anesthesia: NO
Dilation: YES

FOLLOW-UP EXAMINATIONS TWO THROUGH FOUR

To be done by your Ophthalmologist (Doctor's Name): _____

Phone/Contact information: _____

Anesthesia: NO
Dilation: YES

SECOND EXAMINATION: @ 3 weeks of age
THIRD EXAMINATION: @ 6 weeks of age
FOURTH EXAMINATION: @ 10 weeks of age

FOLLOW-UP EXAMINATIONS FIVE THROUGH FOURTEEN

Anesthesia: YES
Dilation: YES

FIFTH EXAMINATION: @ 16 weeks of age (4 months)
SIXTH EXAMINATION: @ 24 weeks of age (6 months)
SEVENTH EXAMINATION: @ 34 weeks of age (8 months)
EIGHTH EXAMINATION: @ 44 weeks of age (11 months)
NINTH EXAMINATION: @ 54 weeks of age (1 year, 1 month)
TENTH EXAMINATION: @ 66 weeks of age (1 year, 3 months)
ELEVENTH EXAMINATION: @ 78 weeks of age (1 year, 6 months)
TWELFTH EXAMINATION: @ 90 weeks of age (1 year, 9 months)
THIRTEENTH EXAM: @ 102 weeks of age (1 year, 11 months)
FOURTEENTH EXAM: @ 114 weeks of age (2 years, 2 months)

This chart shows the examination schedule for children when one parent has retinoblastoma.

A reference card explaining this and other important information may be available at your doctor's office.

CLASSIFICATION OF RETINOBLASTOMA

The Reese-Ellsworth classification system was developed by two physicians as a method to predict prognosis of eyes treated by methods which save or preserve the eye. The higher the Group number in the system, the poorer the prognosis is for saving the eye.

Reese-Ellsworth Classification for Retinoblastoma

Group I

- a. Solitary tumor, less than 4 disc diameters in size, at or behind the equator See Figure 1
- b. Multiple size tumors, none over 4 disc diameters in size, all at or behind the equator See Figure 2

Group II

- a. Solitary tumor, 4 to 10 disc diameters in size, at or behind the equator
- b. Multiple size tumors, 4 to 10 disc diameters in size, all at or behind the equator

Group III

- a. Any lesion anterior to the equator
- b. Solitary tumors larger than 10 disc diameters behind the equator

Group IV

- a. Multiple tumors, some larger than 10 disc diameters
- b. Any lesion extending anteriorly to the ora serrata

Group V

- a. Massive tumors involving over half the retina
- b. Vitreous seeding See Figure 3



Figure 1. Solitary tumor, less than 4 disc diameters in size, at or behind the equator



Figure 2. Multiple size tumors, none over 4 disc diameters in size, all at or behind the equator



Figure 3. An anterior segment photograph of vitreous seeding.

Recently, a staging system was developed to group retinoblastoma tumors confined to the eye and those tumors which spread outside of the eye and in other parts of the body. This system is referred to as the Abramson Staging System for Retinoblastoma.

The Abramson Staging System for Retinoblastoma

1. Intraocular disease

- a. Retinal tumors
- b. Extension into choroid
- c. Extension up to lamina cribrosa
- d. Extension into sclera

2. Orbital disease

- a. Orbital tumor
 1. Suspicious (pathology of scattered episcleral cells)
 2. Proven (biopsy proven orbital tumor)
- b. Local nodal involvement

3. Optic nerve disease

- a. Tumor beyond lamina but not up to cut section
- b. Tumor at cut section of optic nerve

4. Intracranial metastasis

- a. Positive CSF only
- b. Mass CNS section

5. Hematogenous metastasis

- a. Positive marrow/bone lesions
- b. Other organ involvement

DIAGNOSIS OF RETINOBLASTOMA

When there is a family history of retinoblastoma, newborn babies should be examined in the nursery at birth by an ophthalmologist, or an eye doctor. When there is no family history, it is frequently the parents who notice leukocoria or strabismus and bring their child in for an examination. Often the general ophthalmologist refers the child to an ophthalmologist who specializes in children with retinoblastoma and other cancers of the eye.

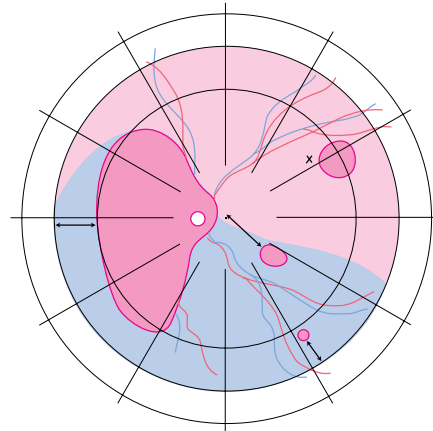
The ophthalmic examination by the specialist is best done under general anesthesia. Some very young and older patients can be examined without general anesthesia; this decision is made by the ophthalmologist. When the examination is performed without general anesthesia, the child is placed on his or her back and is wrapped in a sheet like a mummy to restrict the movement of the child's arms and legs. Dilating drops (which sting for approximately 30 seconds after they are placed in the eye) are placed into both eyes prior to the examination. These drops dilate the pupils of the eyes and allow the ophthalmologist to view the retina. Sometimes numbing drops are also placed in both eyes to numb the



Ophthalmic examination for retinoblastoma

surface of the eye.

If the child is to be examined under anesthesia, the anesthesiologist will put the child to sleep by placing a mask over his or her mouth or nose. A tube may also be placed in the child's throat to aid breathing and an intravenous line may be started. In order



Retinal drawing of retinoblastoma

to minimize the risks of anesthesia, the anesthesiologist will ask that the child not be given food or fluids for several hours before the examination. The child will usually fall asleep within a few minutes and the parent may stay until the child is anesthetized. If you have any doubts or questions about whether your child should have anesthesia, you should ask your ophthalmologist, anesthesiologist or nurse.

The ophthalmologist views the retina with an indirect ophthalmoscope, a special instrument

that permits a view of the retina, to determine the presence of tumors. A lid speculum or metal clip is placed between the eyelids to help keep the child's eye open. During the examination, the ophthalmologist uses a Q-tip or a metal scleral indenter (which looks like a pen with a flattened tip) to press on the scleral surface of the eye to push the most forward portions of the retina into view. Because the child is under anesthesia and his or her eye is numbed, they experience little or no discomfort from this process.

A sketch, or fundus drawing, is made of the tumor(s) of the eye. Sometimes photographs of the view through the indirect ophthalmoscope, fundus photos, are also taken of the tumor(s). Both the fundus drawing and the fundus photos serve as "maps"



Ultrasound of retinoblastoma

for the tumor(s) in the eye. Future examinations will be based on these original drawings and photographs; new drawings and photographs may be sketched and taken at each follow-up visit.

Ultrasound examination, which uses sound waves to penetrate and outline structures in the eye, confirms that retinoblastoma tumors are present and determines their thickness or height. Black and white polaroids of the ultrasound images may be taken initially and during the follow-up visits.

An X-ray test called a CAT scan, Computed Axial Tomography, is also performed to determine if there is any tumor outside of the eye or in the brain (right). An MRI, Magnetic Resonance Imaging, which uses magnetic waves to image the eyes, the orbits, and the brain, may also be performed.

Children who are diagnosed with retinoblastoma are also seen by a pediatric oncologist (children's cancer physician). The pediatric oncologist determines whether there is a cancer anywhere else in the child's body. A physical examination and sometimes blood tests, a spinal tap (to see if there are cancer cells in the central nervous system), and a bone marrow biopsy (a sample of the blood cells taken from the bone) are performed by a pediatric oncologist.



CT Scan of retinoblastoma

Diagnostic Facts:

- Retinoblastoma is diagnosed with the indirect ophthalmoscope, ultrasound, CT/MRI scans
- Biopsies are rarely necessary
- Anesthesia for examination and treatment may be necessary as often as every 2 weeks
- Examination of the parents may help in the diagnosis of the child