

Retino Blastoma

The Parent's Guide



Retinoblastoma Information Booklet

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1. Introduction

If your child has just been diagnosed with retinoblastoma (Rb) you are probably a bit dazed by what it means and how it will affect your child, you and your family.

This booklet was compiled to attempt to make things a bit more digestible from the start. To help you understand and prepare you for what lies ahead. Accept that you have to deal with it, and time is often of the essence. Your child needs you to love him/her and be the strong one when they're around. Your frame of mind determines largely how your child will deal with this.

The gist of it is as follows:

The first year is the most difficult. The second year is still difficult, but you kind of get used to it. With Rb you might think it is over, and then another tumour develops and you start the process all over again. Know that if caught early the treatment is much less invasive.

You are going to hear lots of medical jargon and most of it will not have much meaning to you. When you are ready and able to surf the world wide web you will find that information is available from overseas, but not much on what it actually means and how it might impact you and your child.

You will battle to get the information and the answers you want, and you might have to make big decisions, like taking out one of your child's eyes, within a day or two. The medical world cannot give options and probabilities - each case is too individual and each child responds differently to different treatments. This can be frustrating and placing your trust in a stranger can be difficult.

Get used to hospitals - you will probably spend many hours there over the next few years (yes - years). Be prepared for the hospital visits - bring your own snacks, drinks, toys and blankies with as not all hospitals have these things readily available. It does get less and easier over time.

With Rb treatment the first priority is always to save the child's life, thereafter to save vision, and then only cosmetics (what he/she looks like to others).

It is a scary process, but also know that if diagnosed early your child has a 96% chance of getting through this just fine and lead a normal life, i.e. chances are GOOD!

Children are amazing beings and take things in their stride. They are much more capable of dealing with these things than us adults.

Know that you are not alone.

May God be with you.

2. What is Rb

Retino Blastoma (Rb) is a rare, fast growing childhood eye cancer (typically before 5 years of age). Tumours may affect one eye (unilateral) or both eyes (bilateral), and typically develop before 5 years of age. Some children are born with tumours.

Rarely, trilateral retinoblastoma can also occur, which is when children (almost always with the hereditary form of Rb) develop a tumour in the brain, usually the pineal gland.

Retinoblastoma arises from immature cells of the retina, the light-sensitive layer of cells at the back of the eye, which converts light into nerve signals.

Small fragments of tumour may break off the main tumour and invade the vitreous, the jelly-like centre of the eye. This is called vitreous seeding.

If retinoblastoma is not treated promptly, cancerous cells can spread outside the eye, travelling along the optic nerve to the brain, or through blood vessels to the bone marrow.

Incidence

Between 25-30 children are newly diagnosed each year in Canada, **Note from the Public Library** 300-350 in the USA, and 40-50 in the UK. Countries with high birth of **Science:** rates, like India, diagnose more than 1500 children with retinoblastoma each year.

Symptoms

The most easily recognizable sign is an abnormal appearance of the pupil, which reflects light as a white reflex (leukocoria), like a cat's eye. This is usually only visible in low artificial lighting or in photographs where a flash has been used. Occasionally a squint or crossed eyes can be the first sign of retinoblastoma.

Treatment

Current treatments include laser, cryotherapy (freezing the tumour), systemic and localized chemotherapy, brachytherapy (radioactive plaque sewn directly onto the tumour), radiotherapy and enucleation (surgical removal of the eye).

Prognosis

With early diagnosis and appropriate treatment, nine out of ten children with retinoblastoma can be cured today, creating one of the highest cure rates, and best success stories of all childhood cancers.

However, without early intervention, the cancer will quickly spread beyond the eye, traveling directly to the brain, or invading the bone marrow. Untreated retinoblastoma is fatal.

All of us begin our lives as a single cell (made when an egg and sperm fuse) which repeatedly divides into the few billion cells that constitute an adult human.

During these divisions cells become increasingly differentiated from each other, until in an adult almost all cells are highly specialized to perform a specific function - skin cells, liver cells, eye lens cells, nerve cells, etc.

Cancer is a collection of cells without a function, which grow when normal genetic controls of cell division are interrupted.

Cancer cells are less differentiated than normal cells - leading to the hypothesis that the final steps of differentiation prevent cells from becoming cancerous.

>From the Daisy Eye Cancer Fund website

3. Why did this happen

It is usually a genetic disease, i.e. if you have a family history of this disease your chances increase dramatically. Sometimes it just happens. When the egg is fertilized and the chromosomes connect, something just falls off.

Your feelings

As a parent, the fact that your child has cancer is one of the worst situations you can be faced with. You may have many different emotions, such as fear, guilt, sadness, anger and uncertainty. These are all normal reactions, and are part of the process that many parents go through at such a difficult time. Your child may also have a range of powerful emotions throughout their experience with cancer.

The body has a mechanism that stops certain cells developing once an organ is complete. With Rb this mechanism that stops eye cells from developing further is not in place, so the eye just keeps on making more cells. Compare it to a wound that makes a scab, but the scab doesn't stop growing. Imagine this inside the eyeball.

Genetic Counseling can be done to determine which genetic strain is responsible.

If there is a family history of Rb children must be checked as soon as possible after birth, and at regular intervals thereafter. Siblings of Rb babies should also be checked from time to time.

4. The process going forward

Regular hospital visits will be required. Typically it starts off with monthly check-ups and chemotherapy. As the tumours disappear the hospital visits will taper off to every second month, then every 3 months, every 6 months and then annually for the rest of the patient's life. These visits are very important as going forward you want to detect any new tumours as early as possible. This makes treatment more simpler and increases success dramatically.

As children and tumours develop and react differently to various treatments there is no one recipe that can be applied to everyone. Depending on the result of a check the applicable treatment will be scheduled. Usually (again depending on size, location and amount of tumours) chemotherapy is required to reduce the tumour(s) to a size treatable with laser. Laser is applied once the tumours are small enough, and several treatments might be required to completely get rid of the tumour(s). If the tumours are not responding as wished radiation or enucleation will be considered.

Many different specialists will be involved in the management, including oncologists, ophthalmologists, ocularists and geneticists.

5. The EUA

What is it?

Check-ups are done through an examination under anaesthetics (EUA). An EUA is exactly what it says: the child is put under anaesthetics (usually gas is used to sedate a child) and the doctor will examine the eyes with special tools to monitor existence, location, size and growth of tumour(s).

Preparation for theatre involves nil per mouth (no food or drink) at least 6 hours before anaesthetics to prevent complications. Dilating drops are inserted into the eyes to open up the pupil. This allows the doctor (ophthalmologist) to see inside the eye.

How will it impact you?

This will become a standard part of your life. You will have to remember to schedule your own appointments, and if you have medical aid, to get approval beforehand every time. You will need to fill in lots of paperwork every time you go. You will have to keep your child away from food and drink every time you go to hospital, and try and distract from the hunger. You will have to pack a goodies bag (preferably healthy foods) for when they wake from the anaesthetics. You will have to get up early to travel to the hospital as they usually need to be there early morning. You will have to be strong, patient and have faith to help your child go through this process so many times.

Notes from a mom:

The best way to prepare for an EUA is to play hospital-hospital with your child a day or (and) 2 before.

Practice putting eye drops in (water in a washed saline bottle or dropper will do), and get a mask (maybe your hospital has old ones, or buy something similar at a pharmacy). Practice putting it over the mouth & nose

Most hospitals require you to be there at 8am. Once booked in eye drops are inserted (usually 3 drops, 1 drop every 15 minutes), and then the kids are sent to theatre starting with the youngest. Sometimes the wait can take an hour or two. A parent can usually accompany the child into theatre and stay there until they sleep. Then it's to the waiting room to wait some more until the doctors are finished and the child starts waking. This can take anything from 15 minutes to an hour. The child will have to eat something and hold it down before the hospital will discharge them. It's good to avoid dairy after anaesthetics as this can cause nausea, and to include some sugar to increase blood sugar levels (clear fruit juice e.g. apple or grape works well).

whilst taking deep breaths, counting each one (5 should do). If they're old enough they can do it themselves.

This makes the process less scary for them as they know what to expect.

A pair of sunglasses comes in handy when they need to go outside with dilated pupils, and they look cool!

Some facts & info

Exams will be required for life - initially via EUA, and later when the child is cooperative via routine examination in the doctor's rooms (with no sedation/anaesthetics).

6. Treatment Modes •

Chemotherapy

What is it?

Chemo is medicine which is taken in via a drip. What you have seen on television is an overdramatisation. There are lots of different types of chemo medicine, but the typical combination used in RB is Vincristine, Etoposide and Carboplatin, or VEC for short.

A drip with fluids is also required to 'flush' the toxins out of the system i.e. to help the liver and kidneys deal with all the 'bad' things. An anti-nausea injection is also given. Chemo kills fast dividing cells, which are typically tumours, but also hair and the lining of the mouth and in the stomach. This can lead to hair loss and sores in the mouth and tummy. Your oncologist will watch out for signs of this and prescribe the necessary treatment. It affects the blood count levels in the body, of which there are three types: red blood cells (oxygen carrier), white blood cells (immune system) and platelets (blood clotter).

How will it impact you?

It usually involves staying at the hospital for a few days as the drip runs for most of the day. The schedule is typically chemo in week 1, for 3 - 4 days then back to hospital for bloods in week 2 which entails going in for the morning to have blood drawn and waiting for the analysis which will indicate the blood count levels. These results will determine whether a transfusion is needed and/or readiness for next chemo treatment.

Some facts & info

This is the most invasive treatment as it affects a person on a cellular level, and sometimes people can feel ill during the process. It also leads to the immune system dropping to very low levels, which means viruses and bacteria can easily cause other illnesses /infections. Prepare yourself for a transfusion at some stage as it is very rare that someone goes through chemotherapy and the blood levels remain acceptable throughout. Hair loss is par for course. Babies are not bothered by this, but some bigger children prefer wearing bandanas or wigs. Take your cues from your child.

Normal Blood Count Levels

HGB - 10.5 - 14.0 (They'll transfuse if less than 8.0)

ANC 1500 - 8000 (If your numbers are less than 1000 you're considered neutropenic (immunocompromised). If they get less than 500 there could be serious problems as you're severely neutropenic. This number indicates how well equipped the immune system is to combat disease)

Platelets 150 - 450 (They'll transfuse if less than 20)

Total White Count (TWC) - Normal is greater than 4.5

Notes from a mom:

Always wash hands before touching your child. Get siblings and visitors to do the same.

Stay away from areas with lots of people. Lots of people equal lots of germs. School is not a good idea during chemo.

If the child is small put toys in a bag in a freezer overnight. The temperature kills most germs & bacteria. Alternatively invest in a steamer to clean toys, floors etc. in your home.

It's ok if your child is not hungry during the chemo. Keep healthy snacks (fresh & dried fruit etc) available in case. Sometimes they make up for it a day or two afterwards.

• Laser - Trans PupillaryThermotherapy (TTT)/Cryotherapy.

What is it?

Laser therapy destroys tumours by means of creating an environment in which they cannot survive. Tumors can't tolerate extreme temperatures and die.

- o TTT (Trans Pupillary Thermotherapy) is a laser which heats the tumour to a temperature in which it cannot survive, i.e. "burns" the tumour. This laser is used if the tumours are close to the middle of the retina as well as smaller more peripheral lesions.
- o Cryotherapy is a laser which 'freezes' the tumor (like dry ice). This modality is used for larger tumours, usually more peripheral tumours.

How will it impact you?

The impact on the family is minimal as it does not require another hospital visit. TTT or Cryotherapy is usually done right after an EUA whilst the child is still under anesthetic. The treated eye will be red and sometimes a bit swollen, but there will be no major discomfort.

Some facts & info

TTT is the least invasive of all treatments. Although several treatments are usually required to completely get rid of a tumour(s) it does not require extra hospital visits or sleeping out. It usually forms part of the EUA.

There are only two places in South Africa where this type of laser can be done. The one being Groote Schuur Hospital in Cape Town, the other at the Johannesburg Eye Hospital.

• Radiation plaque/Brachytherapy

What is it?

Brachytherapy is a localized radiation therapy. This type of radiation is preferred to external beam radiation (as they used in the past) as longer term side effects are much less, i.e. it is not as invasive.

A medical physicist works with our own world renowned radiation therapist to make a customized radio-active plaque focusing on the specific patient's tumour(s). This plaque is inserted in the eye during a small operation (under anesthetics they cut the membrane surrounding the eye open and insert the plaque underneath). This must then stay in for a period of 4 - 7 days, depending on the size and location of tumour.

How will it impact you?

This procedure will require travelling to the Groote Schuur Hospital in Cape Town. It involves an operation on day one (similar to an EUA), staying in hospital for the duration of the plaque therapy, and another operation to remove the plaque on the last day. The doctors usually place the plaque so that the child can still see during the radiation process.

The challenge with this mode of treatment is in the fact that the patient is radio-active during this period. This means the patient must be kept in a room for the whole period, with limited contact with any people, including hospital staff.

A family member (usually the mother) is allowed to stay with the child at hospital, and visitors are allowed (if not pregnant and no children). A metal apron will be provided if the child is in the mother's arms most of the time.

Some facts & info

Our unit at Groote Schuur is one of the more experienced units, and is highly regarded.

Nutrition

A healthy diet is always paramount in managing any illness.

It is said that sugar and sometimes salt causes phlegm in our intestines, and this is what tumours feed on. Any doctor will agree that minimizing processed foods and food high in sugar should be replaced with freshly prepared balanced meals, at least 3 times per day. A balanced meal includes a healthy portion of vegetables with smaller portions of carbs (e.g. rice/pap) and protein (meat/fish/dairy).

Enucleation

What is it?

This is the process where they remove the eye. This is always a scary decision to make, but a necessary one once a tumour is of such a size that it is dangerous to leave it in due to the risk of spreading to the cavity or the brain. Once the retina has detached and the eye is blind it is often better to enucleate as the development of the scull can be impacted. The operation itself is much less invasive than one would think. The removal of the eye is one of the least painful procedures in this category. The patient will be in theatre for a few hours (say 1 to 5 hours), and will be up and about the following day. Only pain medication (for babies usually Panado) is required as aftercare.

How will it impact you?

The doctor will remove the eye, and then immediately insert a ball, which will be wrapped with membrane. You will have a choice between a silicone ball (it is easily removable if any problems arise) and a coral ball (this ball is porous and the membrane grows into the ball itself). Some say that the coral ball is better as more movement is possible, but removal/replacement is close to impossible.

Following the removal a conformer is inserted, which is a hollow, round cap, which keeps the socket open for healing and hygiene purposes as well as for proper development of the socket. Once the membrane has healed the process of making an artificial eye will commence. This process entails several visits to an Ocularist, which is a specialist in making prosthetic eyes. They are made from acrylic, and each one is hand painted to match the other eye.

Some facts & info

In children a new artificial eye is usually needed every 1 - 2 years due to growth and changes in eye colour. The human eye only stabilizes completely at the age of 76!

7. Alternative remedies

Many people that receive a cancer diagnosis gravitate towards alternative and/or complementary therapies such as herbs, homeopathy and nutraceutical (vitamin) treatments. This is natural, since it gives that individual a sense of empowerment. Conventional medical doctors usually discourage patients and family members from employing any such treatments. They will insist that these treatments have no evidence-base and only interfere with the drug treatments.

The truth is that there is an evidence-base for a number of anti-cancer and supportive treatments. However, it should only be employed under the supervision of practitioners trained and experienced in this field.

A number of different herbs, homeopathic remedies and nutraceuticals have been shown to work very well to support the body, particularly the immune system and the liver, during chemotherapy and radiation. At the same time they often have their own synergistic anti-cancer effects. Some herbs do interfere with drug treatments and are thus contra-indicated. Some, however, do not interact negatively.

Patients and care-takers of patients should not be tempted to self-medicate, simply due to their lack of knowledge and experience in this regard. In addition to this, herbs available in health shops are not of an adequate grade and generally do not provide the correct dosage. The herbs prescribed and dispensed by trained practitioners are produced in accordance with good manufacturing practices in order to guarantee quality and purity. These herbs are generally only available to registered practitioners.

Notes from a homeopath

What can you as a parent or patient do?

I would recommend that you nourish the body, especially during chemo and radiation. I find that smoothies work well, especially to assist with recovery. Pack the smoothies full of fruit and berries to get lots of antioxidants in. Add whey protein powder to help with protecting and recovering lean muscle mass, which can become compromised during chemotherapy. Whey protein also provides a lot of amino acids that are building blocks for glutathione, your body's own potent anti-cancer nutrient. Add organic, raw (unheated) cocoa powder to smoothies to give it a natural chocolate flavour. Raw, unheated cocoa has the highest anti-oxidant index (more than double that of goji-berries)! Make an appointment with a practitioner trained and experienced in using complimentary therapies such as herbs, nutraceuticals and IP6 to treat and support cancer patients.

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8. Statistics

There is a registry in South Africa which attempts to keep record of diagnoses made, treatment followed and outcome. Unfortunately this has been a challenge up to now as the infrastructure does not allow for this to happen with ease.

It would be appreciated if you could keep them informed of treatment and progress made so South Africa could compare their statistics and research with the rest of the world.

9. Contact list

Herewith some details on the type of specialists you will probably have to deal with, including names of some:

Ophthalmologists/Ophthalmi

c Surgeons

They are doctors specialising in eyes

Once diagnosed there are several ophthalmologists in SA that do examinations under anaesthetics (EUA's), but few with the necessary equipment. Herewith institutions with thermo- and cryotherapy laser equipment:

- o Dr Karen Lecuona (Groote Schuur, Cape Town)
- o Dr Louis Kruger (Johannesburg Eye Hospital)
- Oncologists (childhood cancer specialists)
Dr Janet Poole & associates (group of 8 oncologists) (JHB General Hospital)
Prof Mariaan Kruger (Pretoria)
- Pediatric Surgeons (port-o-catheter ("port"))
This type of doctor will typically insert the port for chemotherapy
- Radiation Specialists (radiation plaque/brachytherapy)
Prof Stannard (Groote Schuur Hospital, CT)
- Optometrists (B-Scan)
- Ocularists (builders of prosthetic eyes)
Jorgen Cieslik (The Artificial Eye Centre, Krugersdorp, Johannesburg)
- Genetic Coucelling (Genetic testing through bloods)
Lindsey Landley (NHLS, Braamfontein)



Shining like the morning sun and the vibrant joy of children, the gold ribbon is the official international symbol of childhood cancer. The ribbon is recognized and promoted by treatment centres, organisations, families, friends, and caregivers across the globe. Each gold ribbon speaks of children and their families who are currently battling childhood cancer, celebrates the victory of each survivor and honours the memory of every precious life lost.

10 Childhood Cancer Facts

- Cancer kills more people each year around the world than AIDS, malaria and tuberculosis combined.
- Globally, cancer is the second highest cause of death in children aged between one to fourteen years. It is the #1 killer of children in developed countries.
- More than 160,000 children are diagnosed with cancer each year around the world, and 90,000 of those children will eventually die. In reality these figures are significantly higher, as many countries in the developing world do not have established cancer registries, and many children die before being diagnosed.
- 84% of children with cancer live in developing countries, where medical care and expertise is often basic or non-existent. They receive less than 5% of global childhood cancer funds.
- Approximately 8,300 children develop retinoblastoma around the world each year.
- Due to global population distribution, 92% of children with retinoblastoma live in less economically developed countries.
- In developed countries, retinoblastoma accounts for about 3-4% of cancers in children aged under 14. In Africa, that figure is much higher due to larger numbers of children under the age of five years.
- Children do not die from primary retinoblastoma contained in the eye, but from cancer that has spread beyond the eye due to late diagnosis and inappropriate or incomplete medical care. With early detection and treatment, eye cancer in children is 100% curable.
- Overall, more than 96% of children treated for retinoblastoma in developed countries will be successfully treated.
- Global Rb survival is less than 20%. That equates to approximately 6,640 avoidable deaths from retinoblastoma each year - 6,640 mothers whose arms ache from being so empty.

10. Some reference sites

www.daisvsevecancerfund.org

www.orphancancer.org

www.candlelighters.org

Facebook group: Retinoblastoma SA