Nursing at Retinoblastoma Treatment
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What is retinoblastoma?

- The most common malignant intraocular, retinatumor (eye cancer) in children
- Occurs in heritable and nonheritable forms
- Mean age 18 months
- 24 months old in unilateral forms (one eye disease) and 12 months old in bilateral forms
- Bilateral forms (tumor in both eyes) 25-35%
- Incidence is 1:15 000 – 1: 18 000 live births, there is no increase over time
- Occurs around the world, no racial or gender differences
- Retinoblastoma has a good prognosis in most countries
Retinoblastoma in the Nordic Countries

Dr Rustam Usmanov

Finland: 4
Norway: 3
Sweden: 6
Denmark: 4
Iceland: 1 in 3 y
Estonia: 1
Lithuania: 2
Latvia:
About the eye

Retinoblastoma

Healthy Eye

Retinoblastoma

- Sclera
- Retina
- Ciliary muscle
- Iris
- Cornea
- Aqueous humor
- Vitreous humour
- Lens

Cancerous cells
Retina

The retina is the nerve tissue that lines the inside of the back of the eye. The retina senses light and sends images to the brain by the optic nerve.
Retinoblastoma
The international Retinoblastoma Staging System is used for staging in Huch Helsinki

- **Group A**: small tumors
- **Group B**: all remaining tumors confined to the retina, subretinal fluid
- **Group C**: local subretinal fluid or vitreous seeding
- **Group D**: diffuse subretinal fluid or seeding
- **Group E**: more than 2/3 of the eyeball is filled with tumor
Retinoblastoma occurs in heritable and nonheritable forms

- 60% non-hereditary, one eye disease is diagnosed at the age of 1-2 years old
- 15% hereditary, one eye disease is diagnosed under 1 year old
- 25% hereditary, both eyes disease is diagnosed under 1 year old
- All bilateral forms and 15% of unilateral forms are related to mutation of the RB-1 gene
- RB-1 gene is localized on chromosome 13q14
Congenital (hereditary) retinoblastoma vs sporadic (non-hereditary) retinoblastoma

**Heritable form** when one of the following is true:

- Heritability risk 50%
  - There is a family history of retinoblastoma
  - There is a certain mutation (change) in the RB1 gene
  - There is more than one tumor in the eye or there is a tumor in both eyes
  - There is a tumor in one eye and the child is younger than 1 year

**Nonhertitable form**:

- Heritability risk 14-95%
  - There is no family history
  - There can be tumors both eyes or only one eye, but two or more tumors and only one eye, one tumor
Genetic research and genetic counseling

- Examination of the inheritance of the retinoblastoma is important for parents, family members and the patient (child).
- Eye oncologist makes a referral to genetic counseling.
- Geneticist meeting will be arranged within 2-3 months.
- Ophthalmologist must examine the parents and siblings eyes as well as new members of the family.
- A child who has heritable Rb has an increased risk of trilateral Rb and other cancers.
### Signs and symptoms of retinoblastoma

<table>
<thead>
<tr>
<th>Condition</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Leukocoria</td>
<td>56%</td>
</tr>
<tr>
<td>Strabismus</td>
<td>20%</td>
</tr>
<tr>
<td>Red painful eye</td>
<td>7%</td>
</tr>
<tr>
<td>Poor vision</td>
<td>5%</td>
</tr>
<tr>
<td>Asymptomatic</td>
<td>3%</td>
</tr>
<tr>
<td>Orbital cellulitis</td>
<td>3%</td>
</tr>
<tr>
<td>Unilateral Mydriasis</td>
<td>2%</td>
</tr>
<tr>
<td>Heterochromia iridis</td>
<td>1%</td>
</tr>
<tr>
<td>Hyphema</td>
<td>1%</td>
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</tbody>
</table>
Symptoms

Leukocoria:

White pupil reflex coming from the tumour’s surface which can be seen through the pupil.
Retinoblastoma patient pathway in Finland

The process takes 24 hours to few days

1. Referral from health center or local hospital
2. HUCH Ophthalmology
3. Call to the patient
4. Eye examination by eye oncologist in Helsinki
5. Diagnose and treatment
Multiprofessional team involved

Team is formed by:
- eye oncologist
- anesthesiologist
- nurses and nursemaid
- social worker

As required there will be:
- eye prosthesis maker
- psychologist
- eye rehabilitation instructor
- optician / optometrist
Preoperative Care

The child is called as soon as possible to the Eye Clinic

1) Nurse will organize timetables with the eye oncologist and operating theater
   - usually assistant nurse manager will do that

2) Phone call to the parent and make the hospital income interview
   - child’s health history, allergies, other notices, listen to parent and encourage to ask!

3) Provide information about coming to the hospital
   - time of arrival
   - fasting times before anesthesia: food, milk 6 hours and water 2 hours
   - clothes, toy
Eye examination and diagnose

- Eye examination with maximally dilated pupils under general anesthesia
- Ophthalmoscopy
- Slit-lamp biomicroscopy
- Retcam ocular imaging
- Ultrasound examination
- MRI (magnetic resonance imaging)
Nursing tasks during diagnostic examinations

- Receive the child and family, introduce each other
  - parents are encouraged to participate in the care of the child all the way
  - the presence of the parents increases the child’s sense of security!
- Inform about the dayplan and program
- Introduce the ward, books, toys, Dvds for child
- Check the current health, weight, fasting times, the child’s clothing
- Put the identification wristband for a child
- Eye drops and pre-medication (if needed f.ex. Dormicum / ordered by anaesthesiologist)
Apply eye drops to both eyes x 2-3

Oftan Tropicamid and Minims Phenylephrine 2,5% are given

Guide the parent to keep the child in place

Tell the child and the parent that you will apply eye drops before you do it!

There is usually some stinging and burning immediately after applying drops
Operating theater
Slit-lamp biomicroscopy, ophthalmoscopy and intra-ocular pressure measurement
Retcam ocular imaging - photo documentation
Ultrasound examination
B-scan ultrasonography shows intraocular mass
Mri scan of the brain and orbits

Magnetic resonance imaging

Series of detailed pictures of areas inside the skull and the eye

Ensures intraocular retinoblastoma and extraocular retinoblastoma (metastatic)
The goals of treatment

- The aim is to destroy the tumor
- To prevent the spread of the tumor
- Retain useful vision

When the diagnosis of the retinoblastoma is confirmed:
- the Eye Oncologist will meet the family, discuss and tells about the disease, prognosis and treatment options
Treatment Options
Depend on the following:

- Whether the cancer is in one or both eyes
- The size and number of tumors
- Whether the tumor has spread to the area around the eye, to the brain or other parts
- The age of the patient
- How likely it is that vision can be saved in one or both eyes
Treatment of the unilateral retinoblastoma

1) Enucleation or removal of the eye
   - Effective and safe treatment when the tumor is large and useful vision is lost and the other eye is healthy
   - The advantage of the enucleation is that the tumor will be fully removed and the child receives prosthetic eye
   - Patient visits to the Eye Clinic will be reduced

2) Chemotherapy
   - If the tumor is not on the optical axis and there is vision left
   - Systemic chemotherapy drugs will be given in Huch Children’s Hospital, Chemotherapy Ward
Eye prostheses
Treatment of the bilateral retinoblastoma

1) Chemotherapy
   - In most cases effective method that can shrink large tumors

2) Thermotherapy
   - A good method to reduced tumors
   - The heat of the thermotherapy destroys cancer cells

3) Cryotherapy
   - Is a treatment that uses an instrument to freeze and destroy cancer cells
Group B retinoblastoma, before (A) and after (B) primary chemotherapy
Type IV regression after chemotherapy and focal laser consolidation
Treatment of the bilateral retinoblastoma

4) **Radiation therapy**

- In most cases used internal radiation therapy called Plaque / disc radiotherapy
- Destroys medium-sized and smaller tumors
- Radioactive seeds are placed on one side of a thin piece of metal (disc) and the disc is inserted onto the outside wall of the eye
- Radiation disc stays usually 1-2 days on the eye and will removed when the total radiation dose is reached
- The advantage of this treatment: accurate targeting of the tumor and helps to protect other areas of the eye
Plaque Radiotherapy of the Eye
Postoperative Care

- Nursing tasks:
  - Observation of the child’s general wellbeing
  - Pain management, specially after enucleation, cryotherapy and plaque radiotherapy
  - Drop eye drops if needed
  - Emotional and informational support to the family by giving them time
  - Individual patient guidance will support the family
Retinoblastoma Controls

- Ophthalmologist will evaluate by child’s age, treatment and the risk of new tumors
- Eye examinations monthly (in the first quarter) and during chemotherapy treatment, sometimes earlier f.ex. two weeks from a radiotherapy
- Depending on the response to treatment, every 2-3 months in the first year
- Every 2-4 months in the two years when the child is over 1 year and there has not been new tumors
- Every 6 months to 6-8 years old
- Once a year adulthood
Retinoblastoma is the most common intraocular cancer overall globally

Needs our attention

Ystäväni Ronja

A national TV "star" in Finland

Children’s program; My friend Ronja