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Retinoblastoma

- Not Just Sight Threatening,
But Life Threatening As Well!

Good vision is something we take for granted and seldom pay attention to, especially when we are in a healthy state. Very often, we tend to overlook the subtle signs of potential eye diseases, which if not diagnosed and treated early, will lead to blindness and possibly premature death. Though this may happen in both children and adults, it is more often so in children. Therefore, it is necessary for parents to keep a lookout for such signs in their children and seek prompt medical attention.

What is Retinoblastoma?

Retinoblastoma is the most common primary malignancy affecting the eye in children. It is a childhood cancer arising from immature retinal cells in one or both eyes and typically seen between the ages of six months to three years in all races and both genders equally. In general, the incidence is one in 10,000 - 20,000 births, resulting in an average of two to three new cases every year in Singapore, though far more patients are seen from the neighboring countries.

Retinoblastoma is a life-threatening disease, but it is rarely a fatal one if treated appropriately. An early diagnosis with the correct treatment by an experienced ophthalmologist and appropriate follow-up is critical to the successful treatment of this disease.

Symptoms that parents should look out for

The common symptoms of retinoblastoma include:

- Abnormal light reflex (having a white 'glow' or 'glint' in the pupil) in one or both eyes
- Presence of a white pupil in a colour photo
- Deviation of the eye (crossed or misaligned eyes)
- Red painful eye
- Poor vision
- The iris may be a different colour in each eye

If you notice any of the above signs in your child, it should be brought to the attention of an ophthalmologist for a detailed eye examination.

Diagnosis of Retinoblastoma

Unfortunately, there are no effective screening tools for retinoblastoma. The most common symptoms are a white or altered red reflex in the pupil, crossed eye or deviated eye and sometimes even red eyes. Unfortunately, all the above are considered symptoms of advanced retinoblastoma. In children with family history (parent or sibling with retinoblastoma), routine screening of the younger sibling by dilated examinations by the ophthalmologist up to five to seven years of age may be able to diagnose lesions earlier.

Treatment

Treatment for each patient varies depending on the number, position and size of the tumours in the eye, whether one or both eyes are involved and also if the cancer has spread to other parts of the body. The aim of treatment is to get rid of the cancer and try to retain the eyesight. When confined to the eye alone, the cancer may be treated by the following techniques:

- External-beam radiation (IMRT or Stereotactic Radiotherapy)
- Plaque therapy
- Cryotherapy
- Laser treatment
- Thermotherapy
- Chemoreduction

Given the different modalities involved, this group of patients is often co-managed with the Paediatric Oncologists as well as the Geneticist, Radiotherapists and Social Worker.

If these measures fail, and both vision and globe are not salvageable, enucleation (removal of the eye) may be required with primary orbital implantation, followed by a 'customised artificial eye' fitting. This is necessary because it is the only way to remove the cancer completely, with negligible recurrence if properly performed. By reconstructing the socket with advanced oculoplastic techniques, this can be accomplished without cosmetic disfiguration and also restores the normal appearance of the patient.

Enucleation or removal of the eye is performed under general anaesthesia in children and usually coupled with a detailed dilated examination of the other eye. While the entire eyeball is removed intact, the other tissues of the eye socket (namely

the muscles, fat, eyelids) are left in place. If the distal end of the optic nerve (which connects the eye to the brain) is free of tumor, a spherical ball implant is placed at the same sitting and the wound is repaired with the placement of a conformer (space maintainer) for four to six weeks. Once healed, a customised artificial eye (prosthesis) is fabricated and placed in the eye socket. This is performed by a trained Ocularist, who is a specialist trained to fabricate artificial eyes for patients with blind, disfigured eyes and those who have had their eyes removed. The process may take one to three days so that comfort, symmetry, minimal discharge and thus a normal appearance is restored.

When it involves both eyes, the brain or the disease has spread beyond the eye, chemotherapy may be required as well. Once chemoreduction is achieved, the eye with the more advanced tumor may still require removal and the other eye with less advanced tumor may benefit from local treatment with lasers, radiation, heat therapy or other modalities discussed earlier.



Two-year old child with retinoblastoma – right eye presenting as altered light reflex (leukocoria)



Same child - post enucleation with artificial eye fitting right eye

CONTACT US

For more information or appointments, contact:

The NUH Eye Surgery Centre

Equipped with the latest state-of-the-art equipment and facilities, our Eye Surgery Centre provides a full and comprehensive array of diagnostic and therapeutic eye services for our patients.

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Services available at NUH Eye Surgery Centre

- Glaucoma
- Cataract
- LASIK Surgery
- Medical & Surgical Retina
- Eye Plastic & Aesthetic Surgery
- Cornea
- Inflammatory Eye Diseases
- Paediatric Ophthalmology

Ocular Prosthetic Service @ NUH Eye Surgery Centre

The ocular prosthetic service under the umbrella of services offered by NUH Eye Surgery Centre is aimed at rehabilitating those unfortunate who have lost their eyes from various causes. Using special techniques, a functional rehabilitation of patients with such eye disorders with an aesthetic angle is achieved.

Our Eye Plastic Surgeons

Senior Consultant Dr Shantha Amrith (Director, Oculoplastic Service)
 Consultant Dr Gangadhara Sundar (Director, Eye Aesthetic Service)

Both specialists are trained in the diagnosis and management of children and adults with diseases of the eyelid, tear ducts and eye socket.