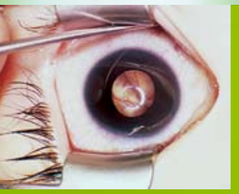


RETINOBLASTOMA
視網膜母細胞瘤

兒童癌病基金
Children's Cancer Foundation

1. What is RETINOBLASTOMA?

Retinoblastoma (RB) is a cancer of the eye that occurs in children. This cancer arises from the retina, the structure that allows vision, inside the eye. The cancer can affect one or both eyes. In the USA, the incidence rate of retinoblastoma is 1 in every 15,000 newborn per year. In Hong Kong (from 2009 to 2012), there are on average 6 newly-diagnosed cases each year. This cancer has no racial or gender predilection. In the developed world where this cancer is diagnosed early and treated appropriately, the survival rate is over 90%.

Unilateral RB refers to retinoblastoma affecting only one eye while bilateral RB refers to the cancer affecting both eyes. In general, 75% of patients suffering from retinoblastoma have unilateral disease while the remaining 25% have bilateral disease. Of those with bilateral retinoblastoma, 90% do not have any history of this cancer in their family.

2. What are the symptoms of RETINOBLASTOMA?

Common symptoms include:

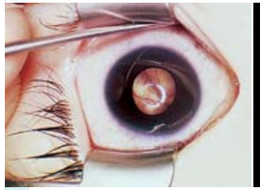
1) Leukocoria or loss of red reflex
2) Squint or strabismus

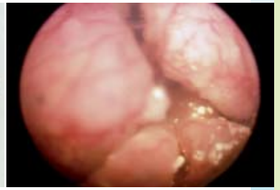
Other symptoms include: red eyes, pain, blurred vision, infection, variation of iris colours etc.

3. How is RETINOBLASTOMA diagnosed?

RB is diagnosed clinically through examination by an ophthalmologist. The ophthalmologist dilates the child's pupils and examines the retina with special equipment to make the diagnosis. Apart from this, other additional instruments or methods to make the diagnosis include:

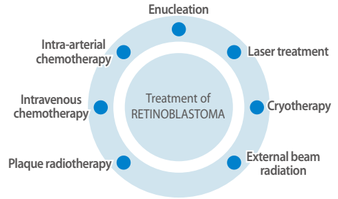
1) Ultrasound (USG) to assess for presence of calcification.
2) Computed tomography (CT scan) to assess for presence of calcification, extension of tumor outside the eye or into the brain.
3) Magnetic resonance imaging (MRI) to assess for extension of the tumor outside the eye or into the brain.
4) Assessment by pediatrician to rule out spread of cancer to other parts of the body. This may involve blood taking, lumbar puncture, bone marrow biopsy, etc.





4. Treatment of RETINOBLASTOMA

The aim of all cancer treatments is to save LIFE. The commonly used methods to treat retinoblastoma are:



5. How is RETINOBLASTOMA inherited?

Retinoblastoma results from defects arising from the Rb gene, which is located on chromosome 13. The normal Rb gene is responsible for regulating normal cell growth. A defect in this gene allows cells to grow excessively, giving rise to cancer. The exact cause of this defect on the Rb gene is still not fully understood. However, it is known that 40% of RB patients carry this gene defect in all cells of the entire body while 60% only carry this gene defect in retinal cells in the eye.

Retinoblastoma can be classified as sporadic or germline. 60% of the cases are sporadic mutation. The remaining 40% belongs to germline mutation, in which the mutation occurs in every cell of the body, and the mutation can be inherited by their offsprings. Those patients with germ line mutation may not necessarily have a positive family history. For germ line mutation cases, the patients usually have bilateral involvement with multiple tumors and earlier onset of disease at less than one year of age. Though unilateral involvement does not rule out a germ line mutation. The germline, or hereditary form of the illness usually gives rise to bilateral retinoblastoma, but can also be unilateral. The cancer typically occurs earlier in life at around the age of one. Sporadic retinoblastoma usually occurs later in life at around the age of two.

	Sporadic	Germline
Gene defect	Only in retinal cells	In all cells of entire body
Risk of other cancers	Nil	Bone tumour, melanoma, soft tissue tumour.
Inheritance	1 in 15,000	45 in 100

From the patients' parents

"My son was diagnosed with tumours in both his eyes when he turned 3 months old. The worst thing was that his paediatrician did not notice at the very beginning. I hope more attention can be given to this type of cancer so that tragedy happens less often. A child's eyes are priceless."

"I am the mother of a child with RB. I want to use this leaflet to make an appeal to doctors. You are the first point of contact with children who might have RB. Please do a careful checkup. With your timely intervention, parents can have their children examined at the earliest opportunity. Early detection and diagnosis means a greater chance of saving our children's eyesight. Thank you!"

An eye is just a small organ in the human body, but it is an important one too. Tumours in the eyes can ruin a child's life. A group of helpful doctors in the Hong Kong Eye Hospital, together with the Children's Cancer Foundation, patients and their families established 'Child's Vision' – a mutual support group for RB. The objectives of 'Child's Vision' are: 1) to assist families of newly-diagnosed children to learn more about RB, its treatment and follow up; 2) to exchange information and sharing among all those affected. We hope to help each other in the group and raise our children healthily together.

Children's Cancer Foundation website: www.ccf.org.hk