IMPORTANCE OF THE RED REFLEX TEST IN THE DIAGNOSIS OF CANCER

The photographs below depict the asymmetry or absence of red reflex that can occur in several serious eye conditions, including retinoblastoma. Red reflex examination is essential at post natal, 6-week and routine development examinations, and at any consultation where a parent is anxious about their child's eyes, vision and/or eye appearance.

It is vital to note a child with retinoblastoma may appear well, apart from one or more of the signs noted below. Signs of retinoblastoma can be very subtle and difficult to describe.

Technique In a darkened room sit in front of the child and parent at arm's length; use a good quality halogen light ophthalmoscope set close to zero focussed on the parent's eyes. This shows the test is non-invasive and offers recognition of the normal red reflexes in that particular ethnic group. Then examine the child. The corneal reflex can be checked at the same time. If it is unclear whether the reflex is normal then dilate pupils with drops.

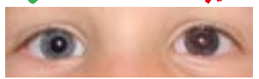
Normal red reflex and corneal reflex The colour and brightness of the red reflex, and the location of the small white corneal reflex, is identical in each eye.

Red reflex absent An absent red reflex in one or both eyes demonstrates serious eye disease (such as cataract or retinoblastoma) requiring urgent referral.

Red reflex is abnormal A red reflex that is the wrong colour or brightness in one or both eyes suggests serious eye disease (such as cataract or retinoblastoma) requiring urgent referral.

Red reflex with a squint A squint is a common indicator of retinoblastoma. Always do a red reflex test on a child presenting with a squint in order to rule out serious eye disease. An abnormal red reflex with squint requires urgent referral.

Other signs of retinoblastoma A change in colour to the iris can occur. This may be in one iris or one part of the iris. One or both eyes can become, red, sore or swollen without any infection. A child may have deterioration in their vision or they may have had poor vision from birth.



A RED REFLEX TEST SHOULD BE DONE FOR ANY OF THE SYMPTOMS ABOVE.

What to do next: If you are unable to confidently rule out retinoblastoma with a red reflex test, NICE guidelines state an urgent referral must be made for children with:

- a white pupillary reflex (leukocoria). Pay attention to parents reporting an odd appearance in their child's eye.
- a new squint or change in visual acuity if cancer is suspected.
- a family history of retinoblastoma and visual problems. (Screening is necessary from birth.)

Urgent referrals should be made to the local ophthalmology department. We recommend you call through to alert them to this case and to find out the speed with which their urgent referrals are seen (in some cases it can be longer than two weeks).

Tel: 020 7377 5578 Email: info@chect.org.uk.
See our website for more information: www.chect.org.uk
This poster was produced by the Childhood Eye Cancer Trust (CHECT) Registered Charity No. 327493.

The effects of Rb on a patient are lifelong.

Anyone who has had Rb should be monitored to detect and manage any long-term problems caused by the disease or treatment in order to ensure the best possible quality of life.

Effects of treatment:

- Late effects of chemotherapy for retinoblastoma can include hearing problems, kidney problems and second malignancies.
- Late effects of intra-arterial chemotherapy for retinoblastoma can include swelling around the eye, flushing of the skin around the eye and forehead and droopiness of the eyelid. Children may also experience a minor drop in their blood count.
- Late effects of radiotherapy can include cataracts, dry eye, facial asymmetry, retinal detachment.

Artificial eyes and vision:

- Many patients have one or both eyes enucleated. **People in the UK can visit the NHS National Artificial Eye Service (NAES)** or a private ocularist about their artificial eye. GPs can refer to these services.
- **Fewer socket problems occur if the patient is under the care of a prosthetist.** If problems persist a referral to a local oculoplastic/orbital surgeon via the GP is important as reconstruction of the socket may be possible or necessary.
- **Some patients will suffer severe vision loss as a result of their retinoblastoma. This may be in one or both eyes.**

Second cancers:

There is an increased risk of second primary cancers to those with the heritable form of Rb. This group includes cases of Rb where the tumour is bilateral or multifocal or where there is a family history. Patients surviving this form of Rb have an increased risk of developing sarcomas within about 5-25 years of treatment. They are also at an increased risk of developing other forms of cancer into later life. The level of risk depends also on the treatment given for the Rb.

Please be alert to any reports of unexplained or persistent pain, lumps or new moles or changes to an existing mole. Adults who had Rb should be seen regularly in an adult oncology clinic.

For referrals and information contact the Rb teams in London and Birmingham at:

The Retinoblastoma Service, The Royal London Hospital, 020 3594 1419
www.bartsandthelondon.nhs.uk/

The Retinoblastoma Service, Birmingham Children's Hospital, 0121 333 9 475
www.bch.nhs.uk/

Retinoblastoma
Would you recognise it?

Fighting Retinoblastoma
Childhood Eye Cancer Trust

What is Retinoblastoma?

Retinoblastoma (Rb) is the most common malignant tumour of the eye in children and accounts for 3% of all childhood cancers. It can occur either unilaterally or bilaterally and generally develops before the first five years of life. Tumours develop in the retinal cells which are developing rapidly in early life. The process of cell development continues throughout infancy and the retina is fully developed at approximately five years of age. It is a life-threatening disease but 98% of children survive retinoblastoma in the UK. Over the past four decades, the management of this disease has evolved tremendously, changing from a deadly childhood cancer to a largely curable disease.

A swift referral pathway for a suspected case is vital to reduce loss of vision and the risk of mortality.

Presentation

There are several signs (below) which could indicate retinoblastoma (Rb) but it is important to remember that a child with retinoblastoma may appear systemically well. The initial signs are confined to the eye. If any child presents with one of the following, a red reflex test (see reverse of leaflet) must be performed with a direct ophthalmoscope. All suspected cases or cases where retinoblastoma cannot be ruled out by the test must be referred urgently to a local ophthalmology department.

- Leukocoria – (intermittent) white pupillary reflex noticed in dim lighting or a photo.
- Strabismus – squint (retinoblastoma must be ruled out for all cases of squint in babies and children using a red reflex test).
- A change in the colour of the iris or part of the iris.
- Inflammation, redness or increased pressure in or around the eye without an infection.
- An absence of red reflex when doing a red reflex test.
- Deterioration of vision in one or both eyes or poor vision from birth.
- Wandering eyes (nystagmus).
- Parental history of retinoblastoma – the condition can be heritable so children of an affected parent with retinoblastoma must be screened from birth.
- Parental concern over vision or eye appearance.

Epidemiology

- Occurs in about 1:20,000 live births.
- Between 40-50 cases are diagnosed each year in the UK.
- It can be unilateral or bilateral.
- There is no gender or race predisposition.
- Retinoblastoma can be heritable so adults who had retinoblastoma, who wish to have children, should be offered genetic counselling and testing.
- Babies who may have inherited retinoblastoma must be screened from birth.

A simple red reflex test (see reverse of this leaflet) can determine whether an urgent referral is necessary. Early diagnosis is vital in order to save life and reduce the life-long impact of the disease on the child.

Why is the Childhood Eye Cancer Trust (CHECT) raising awareness of Rb with health professionals?

Although this cancer has very high survival rates, many children live with the consequences of a delayed diagnosis. Late diagnosis for a child with Rb can mean loss of one or both eyes, life with an artificial eye, a visual impairment or, in some cases, complete blindness. In unilateral cases 70% of children will need their eye removed to save their life.

It is vital that every health professional knows the signs of Rb, carries out the red reflex test when Rb is suspected and makes an urgent referral if concerned. Full details of the test and referral protocol are detailed in this leaflet.

Anecdotal evidence from CHECT members indicates that more than half the new cases of Rb identified in 2012-13 had a significantly delayed diagnosis because an urgent referral was not made and followed up.

REMEMBER:

- **Carry out a red reflex test in every case of parental concern**
- **White pupillary reflex (Leukocoria) or a squint (strabismus) are the most common signs**
- **Children with Rb usually show no other signs of illness to alert you to the condition**
- **Just because it's rare, doesn't mean it isn't there.**

Screening

Rb is heritable in 45% of cases and a child who inherits the altered Rb gene has a 90% chance of developing retinoblastoma, so screening from birth at an Rb treatment centre is vital.

In cases where there is family history of Rb, patients of child-bearing age should be seen by a geneticist who will explain the implications and considerations. **Information on the genetics of Rb was scarce 20 years ago therefore many patients who carry the altered gene may not be aware of the risks associated with this.**

Patients planning a family should be referred to a geneticist who can offer genetic counselling. The patient may wish to consider testing, which can include pre-implantation genetic diagnosis; chorionic villus sampling at 11 weeks of pregnancy or amniocentesis at 16 weeks of pregnancy.

Anyone considering having testing in a pregnancy should be seen by a geneticist before they plan a pregnancy so that the necessary background work can be done.

Find out more about Rb and the work of the Childhood Eye Cancer Trust at:

www.chect.org.uk