Identifying signs of retinoblastoma

Retinoblastoma (Rb) is the most common malignant tumour of the eye in children (Wallach et al, 2006; Butros et al, 2002) and accounts for 3% of all childhood cancers. Between 40 and 50 cases are diagnosed each year in the UK.

The disease can occur unilaterally or bilaterally, and often in the first five years of life. It is heritable in 45% of cases, and children who inherit the altered Rb gene have a 90% chance of developing retinoblastoma, so screening from birth at an Rb centre is vital for children with affected relatives (tinyurl.com/CHECT-retinoblastoma).

The Childhood Eye Cancer Trust (CHECT) has identified that 72% of GPs do not make urgent referrals, as recommended by the National Institute for Health and Clinical Excellence (2005).

Signs and symptoms
Several signs could indicate Rb including:
» Leukocoria – (intermittent) white pupillary reflex seen in dim lighting or in photographs;
» Strabismus – squint (retinoblastoma must be ruled out for all cases of squint in babies and children, using a red reflex test);
» A change in colour of the iris;
» An absence of “red eye” in one eye only in flash photographs;
» Red, sore or swollen eye without infection;
» A deterioration in vision.

If a child presents with any of the above, a red reflex test must be performed. Details of this test are available at tinyurl.com/red-reflex.

All suspected cases of RB must be referred urgently to an ophthalmology department.

Although this cancer has a very high survival rate, many children live with the consequences of a delayed diagnosis. This includes loss of one or both eyes, a visual impairment or complete blindness. In unilateral cases, 70% of children need the eye removed to save their life.

Symptoms of Rb are listed in the Personal Child Health Record (the Red Book).

Screening
Retinoblastoma is heritable, so adults who have had Rb and wish to have children should be offered genetic counselling and testing. Testing includes pre-implantation genetic diagnosis, chorionic villus sampling at 11 weeks of pregnancy or amniocentesis at 16 weeks of pregnancy.

Second cancers
There is an increased risk of second primary cancers in people with the heritable form of Rb (Halford et al, 2008). Patients surviving this form of the disease have an increased risk of developing sarcomas within 5-25 years of treatment. They are also at an increased risk of developing other forms of cancer in later life.

It is important for Rb survivors to be alert to unexplained or persistent pain, lumps or new moles or changes to an existing mole. Those with a visual impairment may need frequent appointments with their GP or practice nurse for a complete body skin check, since they will not be able to see any potentially malignant skin lesions.

Further information
As part of the CHECT awareness campaign, a toolkit for health professionals is being distributed to surgeries and clinics around the country. If your surgery or clinic has not received this, contact CHECT at info@chect.org.uk or 020 7377 5578.

For information about the Childhood Eye Cancer Trust, visit www.chect.org.uk

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References

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