

Retinoblastoma (eye cancer in babies and children)

Retinoblastoma (Rb) is a rare form of eye cancer which affects babies and children mainly under the age of five. On average, around forty to fifty cases of the cancer are diagnosed in the UK per year. The signs and symptoms of Rb are often very subtle and confined to the eye, and most often detected by parents¹. The child usually appears well in themselves, so it is important to pay attention to parental concern about their child's eyes.

- A red reflex test must be carried out by a GP in every case of parental concern.
- White pupillary reflex or a squint are the most common signs.
- Children with Rb usually show no other signs of illness to alert you to the condition.
- Although Rb is a very rare condition, listen to parental concern over vision or eye appearance.

What is Retinoblastoma?

- Retinoblastoma (Rb) is a rare type of cancer of the eye which presents in babies and young children mainly up to the age of five.
- Early diagnosis is vital to ensure these babies are given every opportunity to save their sight and eyes, and the best chance of survival.
- Although 98% of children in the UK survive Rb², many children live with the consequences of a delayed diagnosis: the loss of one or both eyes; life with an artificial eye; a visual impairment or, in some cases, complete blindness.
- Health visitors play such an important role in the early stages of a child's life, it is crucial that they are aware of the signs and symptoms of Rb. Health visitors can make a difference through improved identification and early intervention, ensuring a swift referral pathway for suspected cases.

- Parents who have had Rb as a child, or have a family history of Rb, should be referred for genetic counselling and testing before a pregnancy occurs.

Incidence

- Rb occurs in about 1:20,000 live births, with around 40-50 cases diagnosed each year in the UK³.
- It can be unilateral (one eye) or bilateral (both eyes).
- There is no gender or race predisposition.
- Babies who may have inherited Rb must be screened from birth.

Health visitors should make parents aware of the useful section in the Personal Child Health Record Book titled 'Screening and routine reviews'. This includes a page with a checklist for "Can your baby see?" with the main symptoms of Rb.

Remember, Rb is rare and the symptoms could be caused by other things. But, parents should be advised to get their child's eyes checked by their GP or optometrist as soon as possible.

More information on page 2

For additional resources see www.ihv.org.uk

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Signs and symptoms of Retinoblastoma

White eye / leukocoria

- A white pupillary reflex, known as leukocoria, is the most common sign of Rb. This is sometimes described by parents as a 'cats eye glow' in appearance and is often seen on photographs taken using flash photography, instead of the common red eye. It can also be seen by the naked eye in certain lighting.



Squint / strabismus

- Squints are commonly picked up by health visitors. Although not all are due to Rb, data collected by the Childhood Eye Cancer Trust (2021)³ from parents of 334 children in the UK who were diagnosed between 2012 and 2021 showed this was a symptom for one third of them. So it is important to ensure that all squints are checked with a red reflex test (usually performed by a GP) to rule out eye cancer before any referrals to squint clinics are made, as this can unwittingly cause delays in diagnosis.



Change in colour

- Change in the colour of the iris or part of the iris.



Red, sore eye

- Inflammation, redness or increased pressure in or around the eye without an infection.

References

1. Parulekar MV. Detecting retinoblastoma. *Community Eye Health*. 2018;31(101):10. <https://bit.ly/3wnJKOs> [Accessed 12.5.22]
2. MacCarthy A, Birch JM, Draper GJ, et al. Retinoblastoma: treatment and survival in Great Britain 1963 to 2002 *British Journal of Ophthalmology* 2009;93:38-39. <https://bit.ly/3w3lYsk> [Accessed 12.5.22]
3. Jenkinson H. Retinoblastoma: diagnosis and management - the UK perspective. *Archives of Disease in Childhood*. 2015 Nov;100(11):1070-5. doi: 10.1136/archdischild-2014-306208. Epub 2015 May 4. PMID: 25940424. <https://bit.ly/3wldSj> [Accessed 12.5.22]

Poor vision

- In small babies, poor vision may present as the inability to 'fix and follow', a tendency to focus only on bright lights, or lack of interest in the face of carers. In young children, a reduction in visual acuity may be detected by reports of a toddler becoming more clumsy and bumping into things, sitting close to the TV, or bringing small objects very close to the eye to see them.
- Other signs and symptoms include:
 - An absent red reflex.
 - Nystagmus (oscillating eyes).

Referral

- Rb is a very rare condition but, if it is present, then it is important that the child is referred to see an ophthalmologist urgently. If you notice any of the signs or symptoms of Rb, you should advise the family to take the child to their GP or optometrist (eye tests are free for children under 16). Also advise parents to take along any photos showing the altered appearance of the eye so any changes can be noted (white eye in particular is often viewed intermittently). The GP or optometrist will carry out a dilation of the eyes and a direct ophthalmoscope examination by shining a light in the child's eye in a darkened room. This is known as a red reflex test. If the red reflex is absent, the child must be referred to a paediatric ophthalmologist urgently.
- NICE guidelines⁴ state that an urgent referral (an appointment within two weeks) for ophthalmological assessment for Rb must be considered for children with an absent red reflex.

Further information:

- **Childhood Eye Cancer Trust:** <https://bit.ly/36XbtNm>
- **NHS**
 - » Retinoblastoma: <https://bit.ly/3JcSdZ9>
 - » Squint: <https://bit.ly/3ue6qkz>
- **NICE:** <https://bit.ly/3yczo6z>
- **Children's Cancer and Leukaemia Group:** <https://bit.ly/3NOPCID>

4. Childhood Eye Cancer Trust (2021) <https://bit.ly/3v1rHgv> [Accessed 12.5.22]
5. National Institute for Health and Care Excellence (NICE), *Suspected cancer: recognition and referral*, last updated September 2020. <https://bit.ly/3yczo6z> [Accessed 12.5.22]

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