

Retinoblastoma

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Key Points

- What is retinoblastoma
- Frequency of retinoblastoma
- Risk factors of retinoblastoma
- Clinical signs of retinoblastoma
- Treatment options for retinoblastoma

Retinoblastoma is a scarce type of eye cancer that is found in children and it arises in the retina. It is the most frequent type of ocular infant malignancy with a frequency of 1 in 15,000-20,000 live births.¹ Its frequency is different in various regions of the world like 4.1 per million in Europe² to 11.8 children per million in the USA.³ It appears that socio economic reasons factor in on the frequency of retinoblastoma cases like poor populations of the world tend to have a higher frequency.⁴ Conservative treatment for at least one eye is possible in most of the bilateral cases. It includes laser alone or combined with chemotherapy, cryotherapy and brachytherapy. The indication for external beam radiotherapy should be restricted to large ocular tumors and diffuse vitreous seeding because of the risk of late effects, including secondary sarcoma. Vital prognosis, related to retinoblastoma alone, is now excellent in patients with unilateral or bilateral forms of retinoblastoma. Long term follow-up and early counseling regarding the risk of second primary tumors and transmission should be offered to retinoblastoma patients.

The most common clinical signs of retinoblastoma are leukocoria (white reflection in the pupil) and strabismus.⁵

Affected population:

Despite being an uncommon disease, retinoblastoma is the most frequent cancer of the eye in children, accounting for around 3% of all paediatric cancers.

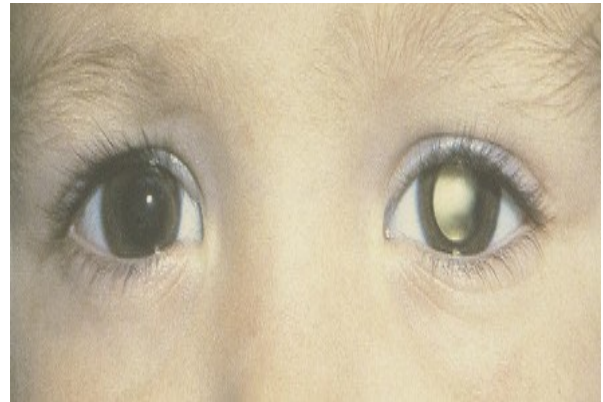


Figure: *Unilateral retinoblastoma*²

Males are slightly more likely than females to develop retinoblastoma. The incidence is predicted to be 2-5 children per 1,000,000 people in the general population in the United States and Europe. In the United States, the age-adjusted yearly incidence for children aged 0-4 is 10-14 children per 1,000,000. This is around 1 in 14,000-18,000 live births. The number of newly diagnosed persons with an illness in a given year is referred to as the incidence. More than 90% of retinoblastomas appear before the age of five years, and two-thirds of children are affected before the age of two.⁵

Risk Factors:

Age, Genetics (Hereditary), During pregnancy, mothers' diets are low in fruits and vegetables. Pregnancy and exposure to toxins in gasoline or diesel

exhaust, Radiation exposure of fathers and older age in fathers.⁶

Diagnostic Methods:

An examination of the ocular fundus under anesthesia leads to diagnosis. The lesion appears as a white tumor with angiomatous dilatation the vessels.⁷

Ocular Ultrasounds, MRI and Computed tomography⁷

Conclusion:

Due to diagnostic delay the reported cases of Retinoblastoma have had an increased incidence in places like India. Moreover the increase in unilateral retinoblastoma shows that environmental factors may contribute to an increase of the uninherited form in India.⁸

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