

Details of the Excellence in Research work

Retinoblastoma

Retinoblastoma is the most common malignant intraocular tumor of childhood. The incidence of retinoblastoma is 1 in 16,000 to 1 in 18,000 live births.^{1,2} It represents 11% of cancers developing in the first year of life, and 3% of the cancers developing among children younger than 15 years.³ It is estimated that, worldwide, 7202 to 8102 new cases are detected each year, of which 5819 to 6545 cases (81% cases) are from Africa and Asia.⁴ The most common presenting complaint in children with retinoblastoma is leukocoria or a white pupillary reflex (60% to 70% cases). The second most common complaint is strabismus (20% to 25% cases).^{5,6} In our large study from India, we showed that the most common presenting complaints in India are leukocoria and proptosis.⁷ Strabismus is the third most common symptom⁷ unlike in the developed countries.



Figure 1 showing white reflex (leukoria) in the right eye of the three children with strabismus in child 1 and 2



Figure 2 showing proptosis in the right eye of both children

The diagnosis of retinoblastoma is delayed in the developing countries when compared to that of developed countries. This hypothesis has been proven by our International Collaborative study.⁸ High-income countries have a shorter lag time between symptoms and treatment compared to middle-income and low-income countries. While availability and accessibility of healthcare facilities may differ based on national income level, programs focusing on increasing awareness about retinoblastoma among the care givers and general practitioners would play a crucial role in decreasing the lag time and improving patient outcomes.

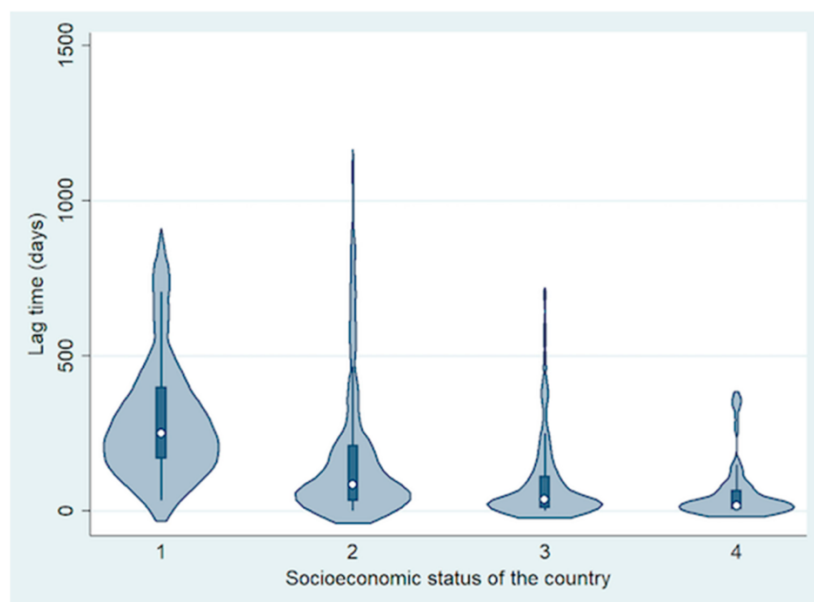


Figure 3. Violin plot showing the difference in lag time from first symptom to treatment based on socioeconomic status of the country.

The management of retinoblastoma depends on the severity of retinoblastoma. The treatment of choice in less advanced cases is chemotherapy and in more advanced cases is enucleation. With our studies on high-risk retinoblastoma, we have shown that high-risk intraocular retinoblastoma is more common in Asian Indians compared with Americans and differs with age.^{9,10} The survival of the eye and life also depends on age at presentation. Younger children are more likely to have higher chances of globe and life salvage compared

to older children.¹¹ The survival can be further improved with early diagnosis by effective screening strategies.¹²

In summary, with our research over the years, we are able to establish several new facts.⁷⁻¹⁶

1. Retinoblastoma in Indian children is associated with 92% life salvage and 50% globe salvage
2. Lag time between first symptom and initiation of treatment is longer in low-income countries, long in middle-income countries, and longer in low-income countries
3. The screening strategies for retinoblastoma are different in different parts of the world
4. High-risk retinoblastoma varies based on age at enucleation
5. The chances of globe salvage and life salvage in retinoblastoma is better in younger children compared to older children
6. Multimodality treatment achieves globe salvage of at least one eye in 58% cases and useful vision salvage in atleast 48% salvaged eyes even when the patients have advanced bilateral retinoblastoma
7. Fundus screening of parents can serve as a surrogate marker for germline mutation and may save the patients form the costs of genetic testing
8. In children presenting with advanced retinoblastoma in India, retinal detachment and seeds at presentation result in high chances of tumor recurrence and thus necessitates aggressive follow-up in these children
9. Second primary cancers occurs in <1% of Asian Indian retinoblastoma survivors and needs life-long monitoring
10. Asian Indians have a 5-fold greater risk of having optic nerve invasion and 3-fold greater risk of massive choroidal invasion compared with Americans

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