Commentary: Retinoblastoma management—Where are we and where do we go from here?

We are presented with an excellent review of contemporary retinoblastoma management in this issue of the Indian Journal of Ophthalmology.[1] India along with other lower-middle-income countries contribute nearly 45% of retinoblastoma cases in the world.[2] We, in India, have made rapid strides in the last two decades in managing retinoblastoma with outcomes comparable with those of the developing countries. All the contemporary treatment modalities listed in the article are available to Indian retinoblastoma patients, albeit in select centers across our country.

Where do we differ in comparison with the developed world:

1. Delayed presentation of disease: Children with retinoblastoma present later in our country than those in advanced countries. This is with a more advanced stage of the intraocular disease and, more often, with the extraocular disease (in certain pockets of the country).[3-7] Improving awareness of retinoblastoma, increasing availability of ocular oncology care across the country, and possible adoption of universal eye screening of all infants can result in earlier diagnosis of retinoblastoma in our country and ultimately resulting in improved outcomes
2. Sparse use of intra-arterial chemotherapy (IAC): The high cost of IAC in India has restricted its widespread use resulting in the selection of intravenous chemotherapy (IVC) to treat even unilateral tumors.[8] The cost of consumables used and paucity of governmental and nongovernmental trust hospitals offering this treatment make it unviable for large-scale adoption of IAC in our country. Indigenous development of cost-effective consumables, rationalizing the costs, and increasing the number of centers offering the treatment can help the transition from IVC to IAC
3. Poor penetration of genetic testing: We do not perform genetic testing as often as we should, because of the paucity of testing centers and the high cost of the tests. While we have the technical finesse to offer prenatal sampling and diagnosis, the capabilities are restricted to a few urban centers in the country. Awareness among clinicians of the need for genetic testing, identifying/creating genetic testing centers, and rationalizing the cost of the tests can mitigate this lacuna in retinoblastoma care in India
4. There is a paucity of support groups to help counsel and educate the family and Retinoblastoma (RB) survivors in our country. The involvement of retinoblastoma specific nongovernmental organizations and a conscious effort by the existing caregivers to create such groups will aid in creating this much-missed support structure

5. The high note is the accessibility of retinoblastoma care in India to the less privileged. A combination of select governmental and nongovernmental trust hospitals offer free/subsidized world-class retinoblastoma care to a large segment of the patient population. A recent initiative to create a collective of all retinoblastoma centers in India promises enhanced funded care to the many more retinoblastoma patients in the near future.

Invading an eye with retinoblastoma for therapeutic purposes is less often being considered a taboo. We are slowly but cautiously adopting intraocular therapies in treating retinoblastoma. Intravitreal chemotherapy, pioneered by Kaneko *et al*., has been adopted to treat resistant vitreous disease, and following an established protocol in delivering the chemotherapeutic agents into the eye has not shown to increase the risk of extraocular disease.[9] Vitrectomy to treat vision-threatening complications, such as rhegmatogenous retinal detachment and vitreous hemorrhage, was seldom considered as a safe option in retinoblastoma and was withheld until complete, sustained regression of the tumor was ensured. On the other hand, there are reports of resistant retinoblastoma being treated with vitrectomy.[10] While such an aggressive approach may not be warranted, a judicious approach to adopting an early vitrectomy in retinoblastoma eyes with vision-threatening complications, such as rhegmatogenous retinal detachment, may allow improved visual recovery, particularly important in one-eyed patients. It is important to stress that invading an eye with retinoblastoma for therapeutic purposes is still fraught with the risk of converting a relatively safe intraocular retinoblastoma into a life-threatening extraocular disease. However, treading with abundant caution, select eyes with vision-threatening complications are being treated with earlier vitrectomy under cover of systemic chemotherapy and a continuous intraoperative infusion of chemotherapeutic agents. Special attention to sclerotomy sites by adopting techniques to minimize extraocular spillage of cells and prophylactic treatment of sclerotomy sites with cryotherapy tend to minimize the risks. Continued systemic chemotherapy and, if necessary, external beam irradiation of the eye and orbit also mitigate the risk of extraocular/metastatic disease after vitreous surgery in these eyes. A meticulous case selection and a pre-, intra-, and postsurgical protocol aimed at mitigating spillage and prompt treatment of possible spillage will allow the treatment of such extreme cases.

Managing retinoblastoma has changed considerably in the last three decades, but the basic tenets remain—saving a life takes priority followed by saving the eye and vision. Having improved the survival rates, we are moving toward salvage of eyes with advanced staging and those with vision-threatening complications. We need to improve on lowering the age at diagnosis of retinoblastoma, increasing penetrance of genetic testing and cost-effective IAC, and creating robust support groups as we cautiously explore salvaging more eyes with advanced disease and complications.

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