

## **Editorial**

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# Continuing Challenges in the Management of Retinoblastoma

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The management of retinoblastoma has gradually changed over the past 10 years. Over 90% of children with localized retinoblastoma are cured with modern techniques. The challenge remains, however, in maintaining the eye and vision. There is a trend away from enucleation and external beam radiotherapy toward focal conservative treatments involving primary chemoreduction in conjunction with thermotherapy and cryotherapy. This is related to earlier detection of the disease, recognition of more effective chemotherapeutic agents, more focused local treatment modalities, and, most importantly, knowledge of the long-term risks of external beam radiotherapy. Enucleation is still preferable for retinoblastoma that fills most of the eye, especially when the disease is unilateral or when there is concern for tumour invasion into the optic nerve, choroid, or orbit. The orbital integrated implant is placed after enucleation and provides acceptable prosthesis motility and appearance. External beam radiotherapy is still vital for treating advanced retinoblastoma, especially when there is diffuse vitreous or subretinal seeding after failure of other methods and preservation of vision is a priority. One important recent advance in the management is, use of neoadjuvant chemotherapy termed “**chemoreduction.**”<sup>1</sup>

In a prospective, nonrandomized, single-centre clinical trial,<sup>2</sup> 158 eyes of 103 patients with retinoblastoma were managed with 6 cycles of chemoreduction (vincristine sulfate, etoposide, and carboplatin). The eyes were classified according to the Reese-Ellsworth classification and were also grouped on the

basis of clinical features as follows: group 1, tumour only; group 2, tumour plus subretinal fluid; group 3, tumour plus focal seeds (3a, focal subretinal seeds; 3b, focal vitreous seeds); group 4, tumour plus diffuse seeds (4a, diffuse subretinal seeds; 4b, diffuse vitreous seeds); and group 5, neovascular glaucoma or invasive retinoblastoma.

This was followed by tumour consolidation with focal measures such as thermotherapy, cryotherapy, and plaque radiotherapy. This strategy provided reduced tumour volume. External beam radiotherapy and enucleation can now be avoided in most cases of Reese-Ellsworth groups I (minimal disease) through IV (moderate disease) retinoblastoma. The most advanced stage of retinoblastoma, Reese-Ellsworth group V, continues to provide the greatest difficulty for management, and external beam radiotherapy and enucleation are often employed in addition to chemoreduction to save the child's life. A collaborative prospective study is currently under way to further study the benefits and risks of chemoreduction for minimal, moderate, and advanced retinoblastoma.

### Transpupillary Thermotherapy as Initial Treatment for Small Intraocular Retinoblastoma

Small retinoblastoma tumours can be treated with transpupillary thermotherapy (TTT) alone. Transpupillary thermotherapy is applied only when a tumour first appears or if growth subsequently occurs. Treatment is performed with patients under general anesthesia with the Iris diode laser (810 nm) on continuous

mode with a large 1.2 mm spot size. In a study<sup>3</sup>, the mean initial tumour base was 0.67 disc diameters (DD) (range, 0.101.5). Eighty-four tumours (92%) were cured with TTT alone in this report. Retinoblastoma tumour < 1.5 DD in base diameter can be successfully treated with TTT alone.

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