RB Study : A prospective collaborative protocol for newly diagnosed Retinoblastoma patients ~~with Retinoblastoma~~ in India.

I. Study team

Please include

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Hypotheses

Considering abandonment, progression, relapse and death as events, we hypothesize that our standardized treatment strategy will result in 5-year event-free survival rates:

- >90% for intraocular retinoblastoma.
- >80% for retinoblastoma with regional extension.
- >80% for bilateral retinoblastoma.
- ~~>50% for metastatic disease~~. <20% only

Patient enrolment and eligibility criteria's-

1. All new patients must be staged and should be enrolled within 4 weeks of diagnosis and staging. If staged previously, they must be formally reassessed.
2. Patients with all types and Stages of classified Retinoblastoma (unilateral, bilateral, trilateral retinoblastoma) would be included in this protocol.
3. An informed consent should be sorted from all parents for enrolling the subjects.

Leukocoria, Strabismus, Poor vision, Red painful eye, Phthisis bulbi, Sterile orbital cellulitis, Proptosis

RetCam (Mandatory) or similar wide field funds photography

While intravenous chemotherapy remains the most extensively used modality of treatment (chemotherapy is used only beyond group c and most extensively used treatment is actually focal therapy TTT trans pupillary thermo therapy), other tools available for the therapeutic intervention in retinoblastoma include chemotherapy using different delivery routes - intra-arterial chemotherapy (IAC), periocular chemotherapy (POC), intravitreal chemotherapy (IVitC), focal treatment with cryotherapy, laser TTT, radiotherapy by teletherapy (external beam, EBRT) or brachytherapy (plaque radiotherapy), enucleation or orbital exenteration. The treatment must be tailored to every child, depending on the laterality and the group and stage of the disease at presentation.

Table 8: Management of unilateral retinoblastoma	
ICRb group	Preferred primary treatment option
Group A	• Chemoreduction IAC>IVC, when tumor located in visually critical area (fovea, juxtapapillary) (Move this below cryo) • Laser TTT • Cryotherapy for an anterior tumor
Group B	• Chemoreduction IAC>IVC, when tumor is located in visually critical area (fovea, juxtapapillary), or tumor size >4 mm (move this too below Laser) • Laser TTT/Cryotherapy for tumors ≤4 mm in size

Although enucleation and external beam radiotherapy are standard of care therapies for Group B tumors, (not the standard of therapy any more) in local institution studies, they have been successfully avoided, using vincristine, carboplatin and etoposide. However, this therapy puts patients at some risk for acute and long-term sequelae, including secondary leukemia related to etoposide exposure. While the cumulative dose of relatively small. In addition, some parents and investigators have chosen standard therapy (i.e. enucleation or external beam radiotherapy) to avoid this even small potential risk for an often fatal secondary leukemia related to etoposide exposure in this group of patients with such an exceptional long-term survival. Moreover, etoposide is myelosuppressive and increases the risk for Grade 4 cytopenias and infection.

*maximum dose 2 mg (HD chemo: VCN is same as standard dose 0.05mg/kg and Etop 10-12 mg / kg 200mg /m2 for above 3 years and carboplatin remains the same 560mg/m2 for more than 3 yrs)

Radiotherapy
(CTV volume for intraocular RB, with or without ant chamber involvement, post operative orbital RB cases has to be defined) will send you by email

I. Indications and guidelines for plaque brachytherapy

Episcleral plaque radiotherapy is a form of brachytherapy wherein the source of radiation is placed on the episclera adjacent to the tumor, and the tumor receives radiation, sparing other healthy ocular tissues from the ill-effects of external radiation (Table 16).

Indications
(1) Recurrent tumor that is > 3 mm in thickness (and less than 15 mm diameter) which is not suitable for treatment by other forms of focal therapy (TTT, cryotherapy or laser photocoagulation)