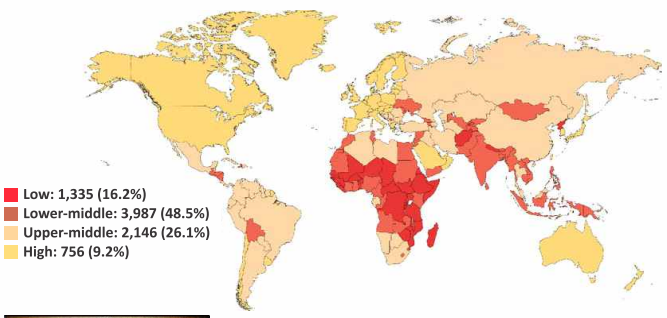
CHILDHOOD CANCER AWARENESS CAMPAIGN

"12 MAHINE: 12 POSTERS"

Pediatric Hematology and Oncology Chapter
Indian Academy of Pediatrics

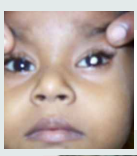
Shining Light on Retinoblastoma: Awareness Saves Sight and Life



- Over 80% of newly diagnosed cases of RB in LMIC
- Delayed presentation in developing compared with developed countries(18 months vs 36 months)
- India has highest contribution with 2000 new cases/year

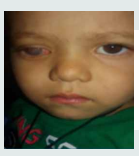
1. Retinoblastoma is highly curable tumour
2. Most common primary intraocular tumor in pediatric age group
3. About 90% are diagnosed by 3-4 years of age (98% by 5 years)
4. 70% RB present with intraocular disease
5. Hereditary retinoblastoma patients often have multifocal/bilateral disease, and they are at increased risk of developing other cancer
6. RB Contributes to 5% of causes of childhood blindness

Clinical Presentation


White reflex (cat's eye reflex) Leukocoria (81.9%)


Squint (54.8%), diminished vision


Phthisis bulbi


Staphyloma

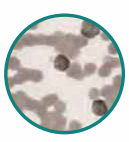

Orbital cellulitis- pain, redness and swelling

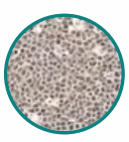

Overt orbital disease with proptosis, fungating mass


Signs/symptoms of metastasis (LN enlargement, anemia, bleeds, bony swellings, cranial nerve palsies etc)

WHO Global initiative for childhood cancers(GICC)

Retinoblastoma one of the six index cancers


Acute Lymphoblastic Leukemia
Most common worldwide


Burkitt Lymphoma common in many low-income countries


Hodgkin Lymphoma
Common in adolescents

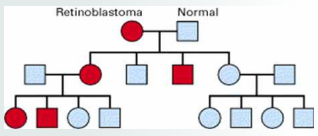

Retinoblastoma
Connecting communities for early diagnosis


Wilms Tumor
Connecting multidisciplinary services

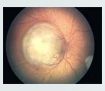

Low-Grade Glioma
Connecting health systems

Diagnosis

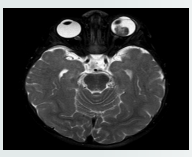
History
Age of onset, laterality
Family history
Make a family pedigree

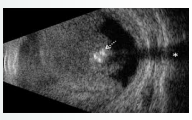


Ophthalmological evaluation
Visual assessment: Red reflex test
Tonometry
Examination Under Anaesthesia
Eye examination of parents important

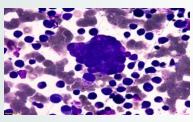


Disappearance of red glow important
Biopsy not required for diagnosis
Genetic studies important





Imaging
USG B-Scan- Presence of calcification
MRI Brain & orbit (contrast)- Involvement of extra orbital, optic nerve and CNS disease

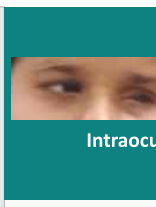


Metastatic work up

CSF cytology (Neuraxial dissemination)

Bilateral bone marrow biopsy (BM metastasis)

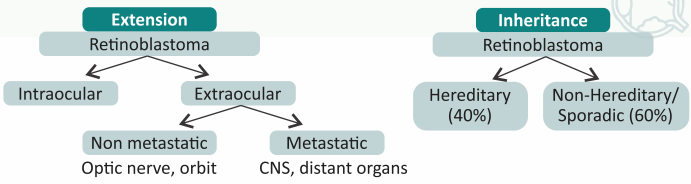
PET-CT (bone & visceral metastasis)/Bone scan


Intraocular RB (IORB)

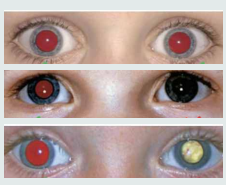
IORB vs EORB


Extraocular RB (EORB)

RB Classification/types



Seeing red: The red & white reflex



- Normal Red Reflex: Light entering the eye is reflected off the retina's blood vessels, giving the pupil a reddish-orange appearance
- Consult if the pupil is white/has no red reflex
- A white reflex in the eye (leukocoria), is an abnormal reflection of light from the retina, appearing white, gray, or silvery instead of the normal red "red reflex".

Screening should be done for children <5 years of age (Immunization centers & preschool); By simple eye examination using a torch/direct ophthalmoscope in dark room

Importance of family history

Approximately 40- 45% of children with retinoblastoma have the **heritable** form Family history maybe absent

When there is no previous family history, the disease is called **sporadic**

If retinoblastoma is **newly diagnosed in a family, parents are encouraged to have an eye examination**
(to diagnose undetected/ asymptomatic RB)

Prospective parents with a family history of retinoblastoma should be referred for genetic counselling

Not all white reflex/leukocoria is a retinoblastoma

Leukocoria can be seen in

- Congenital cataract
- Coats disease
- Retinopathy of prematurity
- Persistent hyperplastic primary vitreous (PHPV)
- Coloboma, Toxocariasis

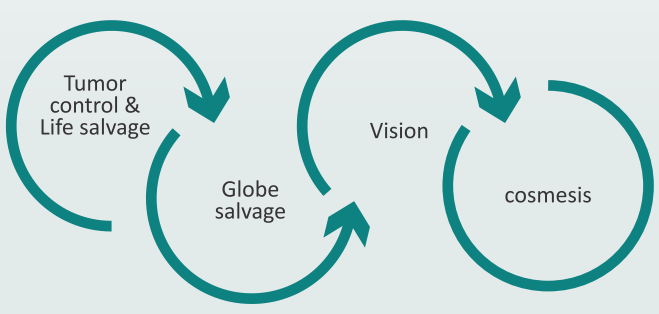
Treatment of Retinoblastoma

Multimodal

(Ophthalmologist, pediatric oncologist, radiation oncologist, ocular pathologist, radiologist)

- Focal therapy
  - Cryotherapy
  - Transpupillary Thermotherapy
  - Laser photocoagulation
  - Plaque brachytherapy
  - Peri-ocular Chemotherapy
- Chemotherapy (focal/systemic/ intra arterial) → Localized /advanced disease
- Enucleation → Primary/secondary
- Radiation → Refractory IORB & EORB
- HSCT → Metastatic RB

Goals of treatment



KEY MESSAGE:

- Survival depends on early diagnosis and appropriate treatment
- Spotting white eye reflex is important in early diagnosis
- Treatment needs multimodal team approach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