

## **BILATERAL RETINOBLASTOMA AT PEDIATRIC ONCOLOGY CENTRE GCRI, DATA BETWEEN 2001-2008.**

### **INTRODUCTION:**

**Most common intraocular cancer of childhood** with approximately 2000 cases diagnosed annually in India. The incidence of bilateral tumors is strongly age dependent with 42% of the occurring in children < 1 year of age .About one third of cases are bilateral. Bilateral and familial retinoblastomas are caused by a Germline mutation and are thus a heritable tumor. Approximately 40% of cases are Hereditary.

**The retinoblastoma gene is a tumor suppressor gene** responsible for the Hereditary/Bilateral cases. Patients with Retinoblastoma most frequently come to medical attention because of the presence of **Leucocoria ( White reflex )**. Strabismus, conjunctival erythema and decreased visual acuity are other presenting complaints. In our country most cases can present as an orbital mass due to late presentation.

The **goal of treatment** should be to save the child's life and salvage vision • Prognosis for vision in bilateral retinoblastoma depends on the Extent of tumor involvement and Effectiveness of treatment modalities.**Treatment options** include Enucleation, External beam radiotherapy (EBRT), Plaque radiotherapy, Laser photocoagulation, Intravenous chemoreduction, Subconjunctival chemoreduction ,Systemic chemotherapy for possible metastatic disease and lastly Orbital exenteration .

### **GCRI EXPERIENCE:**

**There were 275 cases of retinoblastoma that came to GCRI from 2001-08**, out of which

- Retinoblastoma accounts for approximately **8% of all pediatric malignancies at GCRI.**
- It is the **fifth most common pediatric malignancy at GCRI.**
- A total of **39 cases of Bilateral Retinoblastoma** were found.
- Making the **incidence of Bilateral Retinoblastoma as 14.1%.**
- **Age at presentation ranged from 2.5 months to 11 years.**
- **Median age at presentation was 2 years, >80% of cases were more than 1 year of age at presentation.**
- **Family history was present in 2 patients** (parent was affected in one and sibling in the other).
- The **most common presenting symptoms were orbital mass and leucocoria.** Other symptoms included Strabismus , Red eye, Vision loss , Periorbital cellulitis , Pruritus
- The **M:F ratio was 1.9:1**
- **56% of patients were stage 4** on presentation.
- **>90% of patients were treated with Enucleation and chemo radiation.**
- **Local therapy was used in 56% of cases** most common being Laser photocoagulation.
- **8 patients are alive and doing well.**

**We are currently having a family** with four of the five children having bilateral retinoblastoma. Jagruti has completed treatment and is in regular follow-up since 4 years Dhiru her younger brother died at the age of 3 years, he had CNS involvement. Shaila (3 years) and Naresh (7 months) are having early disease, **Laser photocoagulation have been done, and are currently undergoing chemoreduction.**

## **CONCLUSION:**

Retinoblastoma is a curable malignancy (5 year survival >90%) that requires early diagnosis and team work of medical oncologist, ophthalmologist, radiologist, radiotherapist, pediatric surgeon, Geneticist, genetic counselor and finally the medical staff.

For further information contact

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## **Jagruti**



- Diagnosed with bilateral retinoblastoma in 2003 at the age of 2 years
- Left eye enucleation with stage II disease
- Right eye treated with local RT
- Stage II protocol with VC till 57 weeks
- Asymptomatic at present

## **Shaila**



- Presented with Right eye leucocoria for 3 months.
- Diagnosed with Bilateral retinoblastoma at the age of 3 years
- Left Eye – Stage I ,treated with laser photocoagulation
- Right Eye- Stage II started on chemoreduction.

### Naresh



- Diagnosed to have Bilateral retinoblastoma on screening at the age of 7 months
- Right Eye – Stage I ,Photocoagulation Done
- Left Eye – Stage II started on chemoreduction.