

# Community Screening and Community Participation Along with Hospital Based Multidisciplinary Treatment Can be the Key Factors for Both Early Detection, Life and Globe Salvage of Retinoblastoma Patients in Developing Countries – Our Experience

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**Abstract.** Each year nearly 8000 new cases of retinoblastoma are detected worldwide. Revolutionary management strategy increased the survival rate of retinoblastoma above 95% but still it is a deadly cancer worldwide. Survival from retinoblastoma depends on income >90% vs 40% (in high to low income countries) and metastases is higher in low-income countries. Forty three percent of global burden lives in 6 countries of Asia. An estimated worldwide death rate is more than 40% and most from Asia and Africa. Bangladesh is one of the developing countries of the South- East Asia region and retinoblastoma constitutes 83% of all pediatric cancer bellow 4 years age group. Early detection of cases and proper referral may decrease the advance disease which may achieved by strong awareness program. To save life and vision multidisciplinary team approach is a time demand now. Chittagong Eye Infirmary is a tertiary eye care center is serving retinoblastoma patients with a multidisciplinary team approach from 2017 and set up chemotherapy facilities in ophthalmic institute. This institute also started screening and local community training program to detect the disease easily and upgraded treatment facility from 2019. From January 2017 to June 2021 total 284 retinoblastoma patients were diagnosed, 104 received chemotherapy and 53 underwent enucleation with long optic nerve. Two hundred fifty six children were screened, 169 primary school teacher and 408 health worker were trained about key sign of retinoblastoma from July 2019 to February 2021. This effort was able to create awareness which showed early presentation of retinoblastoma and seeking treatment of white pupillary reflex than previous period. The hospitals also take care for visual and psycho-social rehabilitation of the retinoblastoma survivals.

**Keywords:** Retinoblastoma, awareness program, early detection, team approach, proper referral.

## INTRODUCTION

Retinoblastoma is the most common primary intraocular malignancy worldwide. It is the commonest intraocular tumor of childhood representing 11% cancers developing in the first year of life [1] with a of incidence 1: 16000 to 1: 18000 live birth [2]. According to Retinoblastoma Collaborative Study 90% cases are detected and diagnosed before 6

years and approximately 99% before 10 years. The median age at diagnosis is 1.5 year (18 months) [3].

But delayed presentation are common in developing and underdeveloped countries. Lack of awareness, lack of organized treatment facilities and economic factors are the prime factors. Patients who presented late, more than 50% of them die from this disease [4]. Bangladesh is a developing country and facing the same problems like



**Figure 1.** Field activity. (A) Training program. (B) Screening of under 6 children.



**Figure 2.** Hospital activity. (A) Chemotherapy. (B) Counselling. (C) TTT application.

other developing countries in managing the retinoblastoma cases.

This tumor was uniformly fatal once, but now a days due to improved treatment modalities, it has become one of the highest survival cancer among all pediatrics malignancies [1, 5, 6]. In developed countries the survival rate is about 95% with a high ocular salvage [7]. But still retinoblastoma is a deadly cancer worldwide, with an estimated death rate of more than 40% and most of them from Asia and Africa. So, for developing countries we should focus on early detection of cases to prevent delayed presentation along with organization of existing treatment facilities and develop a good referral system. Chittagong Eye Infirmary is one of tertiary eye care center of Bangladesh is provide services for retinoblastoma science its inception

and from 2017 we are providing multidisciplinary treatment facilities. To detect early cases of retinoblastoma and other childhood blinding diseases, awareness creation is a must and realization this situation, this center has started field level awareness program and upgraded the treatment facilities from 2019. Here we sharing our hospital and field level activities.

## RETINOBLASTOMA THE PRESENT WORLD STATUS

Every year worldwide about 8000 and in India more than 1400 new cases are detected [8]. Though it can affect various socio-economic group equally, but lower socio

economic groups present with more advanced stages. The mortality rates in different region of the world are different. It is estimated that, the highest mortality rate is in Africa (70%). The rate in other regions are Asia without Japan (39%), Japan (3%), North America (3%), Latin America (20%) and Oceania (10%) [9]. Survival rate in developed countries is highly impressive. One survey shows the percentage of survival rate in high and low income countries are 90% vs 40% [10]. Literature also shows that occurrence of metastases is higher in low-income countries (32% vs 12% in middle-income) [11]. One study showed, approximately 43% of global burden lives in 6 countries of Asia like India, China, Indonesia, Pakistan, Bangladesh & Philippines [10].

## BANGLADESH AND RETINOBLASTOMA

According to Sarwar et al., the most common childhood cancer types in Bangladesh were leukaemia, retinoblastoma and malignant bone tumors in 0–14 year's age group and retinoblastoma constitute 25% of all pediatric and adolescent cancers. In case of 0–4 years age group retinoblastoma was the most predominant malignancy and was 83% [12].

In our country, there are few tertiary centers where treatment of retinoblastoma is available but not the total care. For this reason patients have to move different centers for further treatment. Most of patients are lost in this way and ultimately endangers the life of children. Also parents are not aware about the white pupil so most of patients are late presenter and this sometimes create difficult situations for health facilitators. These problems are almost same in other developing and underdeveloped countries of Asia and Africa. The probable difficulties that are facing these countries –

- Delay in seeking medical attention – due to lack of awareness about RB.
- Lack of National Screening Program on eye diseases.
- Lack of trained personal specialized in treating RB
- Very few well equipped treatment centers.
- Lack of information
- Socio-economic factors, financial issues, religious belief, gender bias
- Poor compliance to treatment
- Lack of one-stop multi-disciplinary team in one roof
- Lack of proper counselling and support group
- Poor referral system

## CURRENT MANAGEMENT PROTOCOL OF RETINOBLASTOMA

Retinoblastoma needs multidisciplinary management in a team approach. The team consists, ocular Oncologist, oculoplastic surgeon, retina specialist, pediatric ophthalmologist, pediatrician, pediatric oncologist, radiation

oncologist, anesthesiologist, histopathologist, ophthalmologist and counselor.

The management includes –

- Proper diagnosis of the disease
- Treatment
- Timely follow up
- Genetic & general counseling
- Sibling screening
- Rehabilitation of RB survivor

Treatment plan of retinoblastoma depends on presentation of the disease, either bilateral or unilateral; grading and staging of tumor and extent of metastasis.

The management of retinoblastoma has dramatically changed over the past two decades from previous radiotherapy methods to current chemotherapy strategies. Chemotherapy has become the first choice of treatment. Intravenous chemotherapy 6 cycles at an interval of 3–4 weekly is accepted as an international treatment standard by most centers of the world [13, 14]. Combination of systemic chemotherapy associated with local treatments called sequential aggressive local therapy (SALT) [15] is the most popular conservative treatment for intraocular tumor now a days. But enucleation still remains the gold standard for some case of unilateral retinoblastoma.

The treatment options are presented below as tabulated form in Table 1.

## RETINOBLASTOMA AND ROLE OF CHITTAGONG EYE INFIRMARY

Chittagong Eye Infirmary and Training Complex is a tertiary eye care and referral center of Bangladesh. This institute was treating retinoblastoma from its inception and nowadays serving in a team approach consisting of ocular oncologist, oculoplastic surgeon, retina specialist, pediatric ophthalmologist, pediatrician, oncologist, anesthesiologist, histopathologist and ophthalmologist which is a time demand. Along with chemotherapy and surgical treatment the hospital is providing other services like sibling screening, general and genetic counseling, timely follow up of the patients, visual and cosmetic rehabilitation of RB survivors, awareness programs, field level screening and training programs, reliable histopathological services and proper data preservation.

The hospital started chemotherapy services for retinoblastoma children with the support of a German NGO named Children Eye Cancer Foundation from January 2017 and up to June 2021, 284 cases were diagnosed, of which 88 children has received vincristine, etoposide, carboplatin (VEC) chemotherapy and 14 are receiving. Fifty three children underwent enucleation with long optic nerve. As treatment cost of any cancer is costly and sometimes it is a burden to a family, we are providing services at free of cost or at minimal cost. Children Eye



**Table 1.** Current treatment options for retinoblastoma.

Treatment options	
Local therapy	Laser photocoagulation (Green laser) Transpupillary thermo therapy (Diod laser) Cryo therapy
Chemotherapy	Local chemotherapy <ul style="list-style-type: none"> <li>• Intravitreal</li> <li>• Periocular</li> <li>• Intra cameral</li> </ul> Intravenous chemotherapy Intra- arterial chemotherapy Intrathecal chemotherapy
Radiation therapy	Plaque radiation therapy (Brachytherapy) External beam radiation therapy Proton beam therapy
Surgery	Enucleation (Intraocular) Exenteration (Extraocular)

**Table 2.** Hospital and field level activity of CEITC.

Hospital activity			Field activity		
Time period 2017	Diagnosis	284	Time period 2019	Screened	256
Jan–2021	Chemotherapy received	102	Aug–2021	Training of school teacher	169
June	Enucleation with long optic nerve	53	Feb	Training of health worker	408
	TTT & Cryo	76			

Cancer Foundation is giving support for chemotherapeutic medicine and hospital is providing support for surgery and local therapy. Schedule of chemotherapy, physical condition of patients and their reports are monitor regularly by telephone and using different apps. Many children needs blood transfusion as adverse effect of chemotherapy. For managing these situation we have blood donor team also. We also trained our junior doctors and midlevel ophthalmic assistant about the chemotherapy preparation and management of complication.

From 2019 with support of International Rotary club, Chittagong Eye Infirmary has started screening and field level awareness program by training primary school teacher and health worker by incorporating retinoblastoma in 'Fighting Small Children Blindness and Death under 6" project. Under this program, from July 2019 to Feb 2020 total 169 primary school teacher and 408 health worker were trained about features and key sign to identify retinoblastoma. Two hundred fifty six children were screened at different schools and EPI centers of Chittagong district. Due COVID pandemic the field activity are stopped but hospital activity was going on. The hospital also started Transpupillary Thermo Therapy (TTT) under this project. Currently Transpupillary thermo therapy (TTT) is the most commonly used adjuvant therapy. Treatment can also be undertaken alone as a primary treatment [16] This awareness programs has increased the referral to our center and new treatment modalities

are giving the surgeon new strength to fight against this deadly tumor.

## FUTURE PLAN

- Spreading the awareness program for early detection of cases countrywide.
- Extend the RB screening program.
- Strengthening the existing oncology services.
- Start genetic screening test for RB.

## CONCLUSION

Management of retinoblastoma is a long process and cumulative treatment cost of this disease is high. Most of the patients of developing countries are from low and medium income families and most of time it is a burden for families to bear the whole treatment and follow up cost. Strong awareness program and proper referral will detect cases earlier which will save both life, sight and unnecessary treatment cost. Set up of multidisciplinary team approach will increase the patient compliance. All these effort will able to decrease the economic burden of countries and countries. To make this effort successful, donation and support from individuals, institutions, government and different NGOs are needed.

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