

Generating Awareness and a Planned Multidisciplinary Treatment Approach Can Save Both the Sight and Life in Retinoblastoma in Developing Countries

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While rare, retinoblastoma is the most common (1:16000 – 18000 live births) intraocular and life threatening tumor of childhood [1,2]. According to the World Health Organization (WHO), 66% of children present with symptoms before 2 years of age and 95% before 5 years of age. About 8000 new cases are detected annually with the highest incidence in Africa and India. In fact, more than 1400 cases each year are from India [3]. According to Mukesh et al., 43% of the global burden lives in 6 countries of Asia (India, China, Indonesia, Pakistan, Bangladesh & Philippines) [4].

Though white pupillary reflex is the most common presenting sign of retinoblastoma, delayed presentation is still a prevalent issue in low and lower middle income countries [5,6]. Late presentation of the disease in patients correlates to a greater than 50% chance of mortality [7]. Occurrence of metastases is also higher in low-income countries (32%) than in middle-income countries (12%) [8]. The mortality rate from retinoblastoma is also highest in Africa (70%), followed by Asia, excluding Japan, (39%), and Latin America (20%) [9]. The opposite situation is observed in developed countries where the survival rate is about 95-98% and most efforts are now concentrated at increasing the rate of eye preservation [10,11], whereas, in developing countries, the survival rate is unsurprisingly only about 40%. There is a long list of difficulties and almost all developing countries are facing the same problems in managing retinoblastoma including lack of awareness, lack of a screening program for the disease, delayed presentation, few treatment centers and trained personnel in retinoblastoma, lack of accessibility, socio-economic and financial factors, religious beliefs, gender bias, poor treatment compliance, a lack of one stop multidisciplinary treatment facilities, a lack of proper counseling, and many

others [4]. These difficulties, even with governmental intervention, cannot be surmounted simultaneously. Proper planning in combination with governmental organizations, non-governmental organizations, community members and donor agencies is needed. Generating awareness and establishing multidisciplinary treatment facilities are now what the situation demands.

Generating awareness will facilitate the early detection of cases of retinoblastoma, increasing the survival rate and salvaging the eye. Achieving awareness could be accomplished by running a pilot project on the community level involving the healthcare workers, preschool teachers, and parents who have children under five-years-old. They could be taught about nature of the disease, ease of detection by torch light, its fatality rate, availability of treatments, treatment outcomes and the importance of follow up and sibling screening. This may be done by arranging group meetings, posters, flyers, and video demonstrations. The awareness program and screening can also be incorporated into an expanded initiative involving immunization and school eye health programs. When community members and health workers are aware of the disease, they should be taught about proper referral. Proper referral will help parents adequately manage their child's condition and prevent delay in seeking treatment.

Due to current management strategies, the survival and globe salvage rates have dramatically improved and survival rate is better than 95% in developed countries [12]. The management strategy includes proper diagnosis and treatment of the disease, maintaining follow up appointments, counseling and screening of the siblings. For proper management of retinoblastoma, the establishment of multidisciplinary treatment facilities are

very much essential [13,14]. The multidisciplinary team should include one or more ophthalmologists (ocular oncologist, retina specialist, pediatric ophthalmologist and/ or oculoplastic surgeon), a pediatric oncologist, a radiation oncologist, an anesthesiologist, one or more histopathologists, an ocularist, a counselor and social support group. It has been observed that when a patient is referred from one center to another, the parents become confused, a loss of treatment compliance occurs, follow ups are lost and ultimately, treatment is discontinued.

As the total combination of different disciplines seems initially difficult for low and middle income countries, efforts have been taken to train ophthalmologists as retinoblastoma specialists through fellowships available at different institutes. The one stop services will increase the compliance of the patients dramatically.

Retinoblastomas are mainly diagnosed clinically, but sometimes imaging (CT or MRI) is needed in confusing cases or to see the extent of the disease [15]. As this disease has some unusual presentations [16], the ophthalmologist should have a high index of suspicion in cases where symptoms and history do not correlate. After diagnosis of retinoblastoma, grouping, staging and a management plan should be established to help the ophthalmologist provide accurate treatment and can also prevent child mortality. Sometimes, only one cycle of chemotherapy can change the whole situation and may lead the ophthalmologist to reach an erroneous decision.

Different institutes follow different chemotherapy schedules; however, recently Vincristine, Etoposide, Carboplatin (VEC) therapy of 6 cycles, 3 to 4 times weekly is the standard protocol followed in most centers of the world [17,18]. Recent treatment strategies showed the excellent results of combined chemotherapy and local ocular therapy termed sequential aggressive local therapy (SALT) [19]. A diode laser is the best laser for the treatment, but if not available, argon laser or frequency doubling Nd-YAG laser can also work. In cases of enucleation, the optic nerve should be excised more than 10 mm behind the globe from its attachment to globe, with minimal manipulation of 'no-touch' surgical techniques [20]. Every time the enucleated eye should be sent for histopathology. Collaborations with oncologists and histopathologists are also essential. In most developing countries the branch of ocular histopathology has not yet been developed so trained retinoblastoma specialist have to provide information about the patient and should ask about the histopathological high-risk factors present. In most developing countries, enucleation is the primary modality of treatment. As such, both the ophthalmologists and histopathologists should be aware about the presence or absence of histopathological risk factors on which the life of the child depends sometimes. The same situation

applies in cases of oncologists who are less aware about recent treatment protocols for retinoblastoma. Thus, they need to be informed and sensitized.

Treatment of retinoblastoma is completed when the parents are properly counseled, the child is properly followed up and screening of siblings performed. When parents are counselled properly, they will rely on the treatment procedure, receive the treatment, and the patients' loss will be reduced. Timely follow up and sibling screening will detect the recurrence and new lesions earlier which will save the life and sight of the child. The final step of management is the rehabilitation of retinoblastoma survivor both visually and psychologically. The growing survivors sometimes suffers from depression regarding their loss of vision and/or the eye. They should provide information regarding the nature of the disease and should be counseled to lessen mental trauma, if present. Their awareness will make them more vigilant for future offspring and themselves as hereditary survivors have a chance to develop other cancers in future life.

Lastly, we need devotion for this difficult work and financial support if it is to be accomplished. The whole treatment cost of retinoblastoma cannot be carried by patients themselves, so financial support for treatment, hospital stay, and transport should be provided, if needed. Financial support will also be needed to facilitate awareness program and to establish one stop center with multidisciplinary treatment facilities. In developing countries, only the government support is not enough for this. To save the life and sight, national and international non- governmental organizations and different donor agencies should come forward.

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