

Psychosocial Factors Influencing Parental Decision to Allow or Refuse Potentially Lifesaving Enucleation in Children with Retinoblastoma

Rolando Enrique D. Domingo, Maria Socorro W. Toledo, Beverlee Verona L. Mante

Department of Clinical Epidemiology, College of Medicine, University of the Philippines, Manila, Philippines



Corresponding author: Rolando Enrique D. Domingo, MD

Department of Ophthalmology and Visual Sciences, College of Medicine,
University of the Philippines, Manila, Philippines

Tel: +63 920 9540523

E-mail: rddomingo2@up.edu.ph

Received: June 05, 2016, Accepted: August 01, 2016

ABSTRACT

Objective: Retinoblastoma is the most common malignancy of the eye and ocular adnexa in the Philippines. It is curable when treated early, but delay in enucleation is common due to the parental refusal of surgery for varied reasons. The aim of this study is to identify the psychosocial barriers and facilitating factors for accepting versus refusing enucleation as treatment for retinoblastoma. **Methods:** This is a cross-sectional descriptive study utilizing structured interviews and a questionnaire. It was conducted at the Retinoblastoma Clinic of the Philippine General Hospital. A questionnaire using the Likert scale was constructed after performing key informant interviews and focus group discussions. It was pretested and revised before parents of patients with retinoblastoma were invited to participate in the study. Descriptive statistics, quantitative

item analyses using inter-item correlations and item-total correlations was performed. **Results:** Factors that correlate with refusal to enucleate are the beliefs that cancer is a fatal illness, the fear of unacceptable esthetic outcome of the surgery, and the cost of treatment. Favorable factors include value of life, high regard for the opinion of medical practitioners, and appreciation of the efficacy of treatment. **Conclusions:** There are several favorable factors and barriers that health practitioners must consider in facilitating parental decision-making toward enucleation for retinoblastoma.

Key words: Decision-making, enucleation, psychosocial factors, retinoblastoma

Access this article online

Quick Response Code:



Website: www.apjon.org

DOI:
10.4103/2347-5625.207736

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

Cite this article as: Domingo RE, Toledo MS, Mante BV. Psychosocial factors influencing parental decision to allow or refuse potentially lifesaving enucleation in children with retinoblastoma. *Asia Pac J Oncol Nurs* 2017;4:191-6.

Introduction

Retinoblastoma is the most common tumor of the eye and ocular adnexa in the Philippines.^[1] Local tumor registries have shown an incidence of 7.7–8.9 per million children aged 0–14 years.^[2] In the Philippine General Hospital (PGH), records show that the majority of eyes with retinoblastoma were enucleated at an advanced stage of the disease.

The majority of children with retinoblastoma present with leukocoria or a cat's eye reflex. Other less common signs are strabismus, eye redness, glaucoma, heterochromia and, in advanced cases, proptosis or bulging eyes.^[3] Clinical examination combined with imaging procedures such as computed tomography scan and ultrasound leads to diagnostic accuracy of up to 95%.^[4] The incidence of enucleation with a histologic diagnosis of a mimicking lesion (pseudoretinoblastoma) is low, with specialized clinics such as the PGH Retinoblastoma clinic having less than 5%.^[5]

Retinoblastoma is a very aggressive and rapidly growing tumor. The initiation of treatment immediately after the onset of symptoms is important. The condition is uniformly fatal if untreated, and prognosis worsens with advancing stage of the disease. However, if the condition is diagnosed and treated early, survival rates in the most favorable subset of patients are almost 100%.

Enucleation, or removal of the eye, is still the most common treatment for retinoblastoma. Done in early stages of the disease, enucleation can offer complete cure. It is recommended only if there is no hope for vision, the tumor involves more than half of the retina, glaucoma is present, and/or there is seeding into the vitreous. These findings are usually present by the time parents notice anything unusual in the child's eye.

In developed countries, children with retinoblastoma are diagnosed and treated early with the full armamentarium of therapeutic options. Eye-sparing treatment combining chemotherapy, radiation, cryotherapy, and laser surgery is done, but due to the natural progression of the disease, enucleation is still performed in more than 80% of patients until the previous decade.^[6-8] With newer treatment modalities such as intra-arterial chemotherapy, enucleation rates are going down. Most conservative treatment options are available in the Philippine setting and eyes with very small tumors are spared removal. However, since most patients are brought in with advanced disease, enucleation is indicated for almost all unilateral cases. In cases of bilateral retinoblastoma, the worse-affected eye is enucleated and the better eye is treated conservatively.

In patients with intraocular tumor but with high-risk factors on histopathology such as optic nerve involvement,

scleral, and choroidal invasion, enucleation combined with chemotherapy still carries very good prognosis.^[9] The overall survival rate in developed countries is higher than 90%.^[10]

In developing countries of Asia and Latin America, the late diagnosis and treatment of retinoblastoma is still a major concern. At the PGH retinoblastoma service, a large percentage of patients submit to surgery at late extraocular stages of the disease. Advanced cases where the tumor extends beyond the eye have dismal prognosis, particularly if there is direct extension to the brain.

In many Asian countries, a major concern is the delay in treatment after retinoblastoma is diagnosed. One big obstacle is the refusal to enucleate on initial consult. Researchers in these and other countries have shown that treatment delay directly correlates with advanced stage of disease at the time of surgery and decreased survival rate. A study in Indonesia^[11] showed that only 47.3% of patients undergo treatment immediately after diagnosis with 31.5% delaying treatment and 18.2% refusing treatment completely.

In India,^[12] treatment abandonment rate is almost 50% with financial difficulties (30%) and refusal to undergo enucleation (20%) as the main reasons for such. Although Taiwan is a developed and affluent country, a study done in its northern regions showed that their survival rates are lower than western countries, and this is partially attributed to the fact that parents do not allow enucleation early in the diseases opting to try traditional treatments and agreeing to surgery only when the tumors are at an advanced stage.^[13]

The observation in PGH is similar to our neighboring countries. The patients are usually diagnosed by a pediatrician or an ophthalmologist and are advised enucleation. Many parents initially refuse but eventually return for the treatment once the tumor progresses and symptoms such as pain and proptosis become increasingly difficult to bear. By the time they submit to treatment, the surgical procedure is more difficult and extensive, expensive chemotherapy is imperative, and prognosis is poor.

The disclosure process is always difficult and most doctors are neither confident nor comfortable in telling parents that their child has eye cancer and that the eye has to be removed. The session always ends up with parents and grandparents crying. The situation is never easy to manage, even in the hands of experienced clinicians.

A study in Israel involving the experience of twelve couples with a child with retinoblastoma showed that distress levels increased upon detection of the first symptoms, decreased upon consulting a center, increased again at the time of therapeutic decision, decreased when consent to enucleate was given, and increased again after discharge.^[14]

Although the significance of treatment delay in retinoblastoma is widely acknowledged, no published studies investigating the reasons for the refusal of potentially lifesaving enucleation in children diagnosed with retinoblastoma could be found. This study will hopefully lead to a deeper understanding of these factors and aid in the development of strategies to improve health-seeking behavior in this subset of patients.

Methods

This is a cross-sectional descriptive study utilizing structured interviews and a questionnaire. It was conducted at the Sentro Oftalmologico Jose Rizal of the Department of Ophthalmology and Visual Sciences of the PGH. Informed consent was obtained from all participants, and the procedures followed were in accordance with the Helsinki Declaration.

A questionnaire using the Likert scale was constructed by the investigators after developing the conceptual framework, performing four key informant interviews and two focus group discussions involving physicians, nurses, and patients' relatives. It was then pretested on four subjects and revised in terms of language and order.

The working framework for this study is based on the Theory of Reasoned Action which focuses on the interplay between societal norms, personal attitude, and behavioral intention.^[15] The investigators consider the most important factors that play a role in the decision-making for enucleation are as follows [Figure 1].

Once the questionnaire was ready, parents of patients with retinoblastoma admitted at the Sentro Oftalmologico Jose Rizal or following up at the Retinoblastoma clinic of the Department of Ophthalmology and Visual Sciences of the PGH within 12 months after enucleation were invited to participate in the study. In the absence of parents, legal guardians actively involved in treatment decision-making were invited. When filling up the questionnaire, all participants were encouraged to ask clarificatory questions at any time.

The parents of patients who underwent surgery more than 12 months earlier were excluded to prevent recall bias. Companions who were not present or involved during the decision-making for the previous surgery were also excluded. Difficulty in understanding and replying in the Filipino language was also a cause for exclusion.

A total of 10 consecutive participants were invited to participate and they all gave their consent to answer the questionnaire. They were parents/guardians of 10 different patients. The demographic profile of the respondent such as age, educational attainment, religion, province of origin, number of children, and monthly family income were collected. Patient data such as sex, age, birth order, and

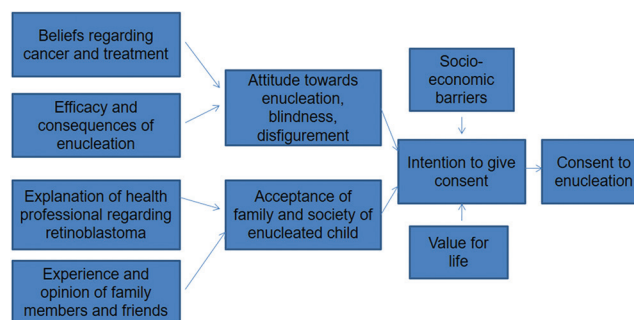


Figure 1: Conceptual framework for decision making towards enucleation

laterality of retinoblastoma at the time of consult in PGH were also recorded.

Data gathered from the questionnaires were encoded using Excel. They were subjected to quantitative item analyses using inter-item correlations, item-total correlations, and Cronbach's alpha using STATA 12 (StataCorp LP, College Station, Texas, USA). After item analysis, it was found that three questions needed to be recoded giving better correlation. Overall analysis showed that the sample size was too small, and a descriptive analysis was found to be more appropriate.

Results and Discussion

The demographic data obtained were not surprising, and most respondents were young mothers, Catholic, with one to three children, and had high school level education. The mean monthly family income is about the same cost as an enucleation at a public hospital [Table 1].

When the Likert scores for each respondent were totaled and averaged, a correlation between the score and the likelihood of giving consent to enucleation can be appreciated. The respondent with the highest Likert score of 2.67 is the only case who refused enucleation. The five patients who said they had to give a lot of thought before giving their consent had an intermediate average score of 2.13 (range: 1.83–2.13). The lowest average score of 1.87 (range: 1.67–2.08) was obtained by respondents who claimed that they immediately agreed to surgery after consulting the doctor [Table 2].

Closer examination of the individual questions and the responses [Table 3] reveals that the highest Likert scores (average = 3.3) were on Question 1 which deals with the issue of cancer as a fatal illness. Nine out of ten respondents agree or strongly agree that cancer causes death. It is understandable that such a belief would make parents question whether going through surgery is worthwhile when one feels that the patient will die of the disease anyway. It is the responsibility of the health worker during the time of disclosure to explain that retinoblastoma is a cancer that can be cured 90% of the time, particularly

with early intervention. Explanation on the course and prognosis of retinoblastoma may make the attitude of the parents more favorable toward seeking surgery.

The second highest scores (average = 2.8) were in response to Question 11 on the appearance of an enucleated child. Again, this affects the attitude of the decision-maker toward intervention. Three out of ten respondents feel that an enucleated eye is esthetically unacceptable while seven out of ten disagreed. It should be noted that most of these respondents already agreed to have their child enucleated and it might be difficult for them to accept that their child will be disfigured by an operation that they consented to. Clinicians must bear in mind that a good esthetic result after surgery is an important consideration and that parents usually imagine it to be a lot worse than it actually will be.

The third item that may be considered as a barrier is the cost of the surgery covered by Question 4 (average = 2.4). The respondents are equally divided with five saying that it is a hurdle and the other five saying that it is not. Considering that based on the declared incomes most of the families will have to make sacrifices to pay for the operation, it is heart-warming to know that they would do so for the sake of their child.

Questions 7 and 13 have more favorable average Likert scores of 2.2. Seven out of ten respondents consider the opinion of friends important (Question 7) which identifies potential allies for the clinician when dealing with reluctant parents. Only one out of ten respondents admitted to considering going to a faith-based healer (Question 13) which is encouraging because it means that scientific and rational treatment modalities are preferred.

The rest of the questions gave very favorable Likert scores. Nine out of ten respondents agree that the opinion of relatives is important and this corresponds to social norms that affect many decisions. Eight out of ten respondents believe in the efficacy of enucleation as treatment for retinoblastoma and this positively affects the attitude toward surgery. On Question 3, all respondents agree that surgery and chemotherapy will be helpful to the patients.

Value of life is reflected in Question 2 and nine out of ten respondents do not agree that it is better to die than to be blind. This is particularly important since four out of the ten patients were cases with bilateral advanced retinoblastoma who would be completely blind after surgery and their parents would accept this as long as they survive.

A very favorable average Likert score of 1.6 was obtained for Questions 5 and 9. All respondents agree or strongly agree that they value a doctor's recommendation highly (Question 5) and this puts a lot of responsibility on the physician to choose his/her words carefully and use this power he/she has to the patient's greatest advantage. Likewise, all respondents feel that a thorough understanding

Table 1: Demographic profile of respondents (n = 10)

Characteristics	n
Age (years)	
21-25	4
26-30	3
31-35	0
36-40	2
41-45	1
Relationship to patient	
Mother	7
Father	2
Other relative	1
Educational level	
College level	3
High school	6
Elementary	1
Income (US\$/month)	
< 120	4
121-240	2
241-360	2
361-480	2
> 480	0
Religion	
Roman Catholic	9
Protestant	1
Number of children	
1	3
2	3
3	3
4	1

Table 2: Respondents' decision to consent to surgery and their average Likert scores

Decision to consent to surgery during the first consult	n	Range of average Likert scores
Yes	4	1.67-2.08 (1.87)
Will think about it before saying yes	5	1.83-2.50 (2.13)
No	1	2.67

of the surgical procedure is important (Question 9). These findings correlate well with the finding that the trait that they look for most in a physician is the ability to explain well.

Surprisingly, the question with the highest Likert score (average = 1.5) deals with personal conviction. All the respondents proclaim that they will decide on what they think is best for their child even if this is contrary to what their friends and relatives say. This tells us that the person who needs convincing the most is the mother as her decision is the most valuable of all.

Conclusion

Parents with more favorable Likert scores are more likely to consider and give their consent to enucleation. The

Table 3: Average Likert scores per question

Question number	Question	Strongly agree (n)	Agree (n)	Disagree (n)	Strongly disagree (n)	No answer (n)	Average Likert score*
Q1	Fatality of cancer	0	1	5	4	0	3.3
Q2	Choice for life despite blindness	3	6	1	0	0	1.8
Q3	Value of surgery and chemotherapy	3	7	0	0	0	1.7
Q4	Issue of cost	2	3	4	1	0	2.4
Q5	High regard for MD's recommendation	4	6	0	0	0	1.6
Q6	Opinion of relatives	1	9	0	0	0	1.9
Q7	Opinion of friends	1	6	3	0	0	2.2
Q8	Personal conviction	5	5	0	0	0	1.5
Q9	Appreciation of outcome of enucleation	5	4	1	0	0	1.6
Q10	Efficacy of enucleation	3	5	2	0	0	1.9
Q11	Acceptability of esthetic outcome of enucleation	0	3	6	1	0	2.8
Q13	Attitude toward faith-based healers	0	7	2	0	1	2.2

*Likert scores - 1: Strongly agree, 2: Agree, 3: Disagree, 4: Strongly disagree. Q12 of the questionnaire was a clarificatory question regarding medical advice not answered on a Likert scale. N: Number of respondents

questionnaire identifies the concept of retinoblastoma as a fatal illness as the most important barrier to the acceptance of enucleation as a treatment for retinoblastoma. The unacceptable esthetic outcome of enucleation and the cost of surgery are other barriers that must be addressed thoroughly.

The high regard for a doctor's recommendation is something that must be carefully and responsibly exploited. The appreciation of the value of interventions such as surgery and chemotherapy, belief in the efficacy of enucleation, and an appreciation of the outcome of enucleation ranks high as important facilitating factors.

The choice for life despite blindness is a strong facilitating factor for influencing the decision toward enucleation. Relatives and friends are important potential allies as their opinions matter, but the most important person to talk to is the mother as she will make the decision in the end and her strong personal conviction is the strongest facilitating factor of all.

Physicians and health workers can facilitate parental decision toward acceptance of enucleation by reinforcing the identified facilitating factors and breaking down the barriers by addressing the fears and misconceptions.

Limitations

The study has a small sample size and is limited to a single institution and population. However, the center treats more than half of all retinoblastoma cases in the country, and the sample is representative of the Philippine setting. Cultural and socioeconomic similarities with neighboring countries make the study useful, at the very least as a basis for developing their own investigations on the subject.

Recommendations

The findings are useful for all health workers, particularly nurses who spend a lot of time caring for

patients and sharing information with their families helping them make difficult clinical decisions. Further study would greatly expand understanding of the problem. The development of a treatment decision aide based on the findings may be helpful to health workers and parents of children with retinoblastoma faced with the decision to enucleate or not.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

- Domingo RE, Manganip LE, Castro RM. Tumors of the eye and ocular adnexa at the Philippine Eye Research Institute: A 10-year review. *Clin Ophthalmol* 2015;9:1239-47.
- Ngelangel CA, Wang EH. Cancer and the Philippine Cancer Control Program. *Jpn J Clin Oncol* 2002;32 Suppl 1:S52-61.
- McLean IW, Burnier MN, Zimmerman LE, Jakobiec FA. Tumors of the Eye and Ocular Adnexa. Washington, DC: Armed Forces Institute of Pathology; 1993. p. 103-5.
- Ghosh S, Mukhopadhyay S, Dutta SK, Chattopadhyay D, Biswas K. Diagnostic accuracy in retinoblastoma. *J Indian Med Assoc* 2010;108:509, 512-3.
- Valenzuela RM, Domingo RE, Rancho JM, Manganip LE. A review of pseudoretinoblastoma cases at a tertiary hospital. *Philipp J Ophthalmol* 2010;35:25-31.
- Mallipatna AC, Sutherland JE, Gallie BL, Chan H, Héon E. Management and outcome of unilateral retinoblastoma. *J AAPOS* 2009;13:546-50.
- Berman EL, Donaldson CE, Giblin M, Martin FJ. Outcomes in retinoblastoma, 1974-2005: The Children's Hospital, Westmead. *Clin Exp Ophthalmol* 2007;35:5-12.
- Aung L, Chan YH, Yeoh EJ, Tan PL, Quah TC. Retinoblastoma: A recent experience at the National University Hospital, Singapore. *Ann Acad Med Singapore* 2009;38:693-8.
- Chantada GL, Dunkel IJ, de Dávila MT, Abramson DH. Retinoblastoma patients with high risk ocular pathological

- features: Who needs adjuvant therapy? *Br J Ophthalmol* 2004;88:1069-73.
10. Honavar SG, Singh AD, Shields CL, Meadows AT, Demirci H, Cater J, *et al.* Postenucleation adjuvant therapy in high-risk retinoblastoma. *Arch Ophthalmol* 2002;120:923-31.
 11. Sitorus RS, Moll AC, Suhardjono S, Simangunsong LS, Riono P, Imhof S, *et al.* The effect of therapy refusal against medical advice in retinoblastoma patients in a setting where treatment delays are common. *Ophthalmic Genet* 2009;30:31-6.
 12. Kumar A, Moulik NR, Mishra RK, Kumar D. Causes, outcome and prevention of abandonment in retinoblastoma in India. *Pediatr Blood Cancer* 2013;60:771-5.
 13. Kao LY, Su WW, Lin YW. Retinoblastoma in Taiwan: Survival and clinical characteristics 1978-2000. *Jpn J Ophthalmol* 2002;46:577-80.
 14. Hamama-Raz Y, Rot I, Buchbinder E. The coping experience of parents of a child with retinoblastoma-malignant eye cancer. *J Psychosoc Oncol* 2012;30:21-40.
 15. Ajzen I, Fishbein M. *Understanding Attitudes and Predicting Social Behavior*. Englewood Cliffs, NJ: Prentice-Hall; 1980.