

## Original Research Article

# Frequency of Different Stage Presentation of Retinoblastoma Visiting Ophthalmology Department at a Tertiary Care Hospital

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### Abstract:

**Background:** Despite being a significant health burden in underdeveloped nations, little is known about the clinical appearance and stages of retinoblastoma. This study's primary goal is to offer a thorough examination of different presentation stages in a context with limited resources. It has been suggested that treatment outcomes are better when applicable knowledge about the predominant stage of presentation is available because this can result in more successful primary, secondary, and tertiary healthcare treatments.

**Materials and Methods:** Total 90 patients of both sexes who are aged six months to ten years, having retinoblastoma are eligible to participate. Individuals suffering from bilateral retinoblastoma, eye trauma and prior retinoblastoma ocular surgery or enucleation were excluded. The parents of each patient were asked for their informed consent. Following pupillary dilatation with Tropicamide 2% eye drops, the patient had a B-scan and indirect bio-microscopy examination while under general anesthesia. The disease was staged.

**Results:** The mean age in this study was  $4.19 \pm 1.78$  years, with a range of 6 months to 10 years. Seventy-one (78.89%) of the patients were between the ages of six months and ten. With a male to female ratio of 1:4.6, 30 (33.33%) of the 90 patients were men and 60 (66.67%) were women. In our study, the average length of illness was  $4.43 \pm 1.64$  months. In our study, 60.0% of the eyes with retinoblastoma had stage 2, 20.0% had stage 1 at initial presentation, 13.33% had stage 3, and 6.67% had stage 4.

**Conclusion:** In order to promote earlier discovery and enhance the chances for globe salvage and vision preservation, these findings highlight the critical need for increased public awareness, routine pediatric eye screening, and prompt referral systems.

**Keywords:** Retinoblastoma, pediatric, stage, presentation, advances.

## INTRODUCTION

Retinoblastoma accounts for about 4% of all pediatric cancers and is the most commonly reported intraocular malignant tumor in children. 1 in 14,000 to 1 in 20,000 live births is its incidence rate. 2 According to estimates, children under the age of five and those under ten have an annual crude incidence of Rb of 4.0/100,000 and 2.4/100,000, respectively. 3 If the condition is not identified in its early stages, it could lead to blindness or possibly

death. Recent technological developments have made it possible to diagnose Rb quickly and treat it effectively, boosting the survival rate to up to 95%. In order to reach this survival rate, developing nations continue to face numerous obstacles. Furthermore, there is no racial or gender preference for this cancer. 4

Both unilateral and bilateral tumors can develop from retinoblastoma. Leukocoria, strabismus,

decreased vision, anterior chamber inflammatory signs, spontaneous hyphema, and proptosis are the most common presenting symptoms of retinoblastoma; however, in developing nations, proptosis and metastasis are reported to be the most common presenting symptoms.<sup>5,6</sup> To define the clinical response to treatment and to stage intraocular illness, the International Intraocular Retinoblastoma Classification (IIRC) was created. Retinoblastoma is categorized in this method into groups A through E. Group A eyes have less disease and a higher chance of survival, while group E eyes have more disease and a worse prognosis when treated with chemotherapy alone.<sup>7</sup> Of the 90.2% of eyes that emerged at IRSS stages II to IV in one study, 55% of the eyes had retinoblastoma at IRSS stage II. At initial presentation, 15.6% of eyes showed overt orbital disease with metastatic retinoblastoma symptoms (IRSS stage IV), whereas 19.6% of eyes had regional extension of retinoblastoma at IRSS Stage III.<sup>8</sup>

Despite being a significant health burden in underdeveloped nations, little is known about the clinical appearance and stages of retinoblastoma. This study's primary goal is to offer a thorough examination of different presentation stages in a context with limited resources. It has been suggested that treatment outcomes are better when applicable knowledge about the predominant stage of presentation is available because this can result in

more successful primary, secondary, and tertiary healthcare treatments.

#### METHODOLOGY:

During the course of 6 months, this Descriptive, Cross-sectional study was carried out at the Department of Ophthalmology, Bahawal Victoria Hospital, Bahawalpur from 24 June 2025 to 23 September 2025. By using WHO calculator for single proportion, sample size of 90 cases has been calculated with 95% confidence level, 7.5% margin of error and taking percentage of stage IV as 15.6%.<sup>8</sup> Patients of both sexes who are aged six months to ten years, having retinoblastoma (presence of proptosis (abnormal forward displacement of globe >20 mm examined by Hurlter's exophthalmometer), Leukocoria (white pupillary reflex or cat's eye reflex on direct ophthalmoscopy) and presence of hyperattenuating intraocular areas consistent with calcifications (on CT scan)) are eligible to participate. Individuals suffering from bilateral retinoblastoma, eye trauma and prior retinoblastoma ocular surgery or enucleation were excluded.

The parents of each patient were asked for their informed consent. Following pupillary dilatation with Tropicamide 2% eye drops, the patient had a B-scan and indirect bio-microscopy examination while under general anesthesia. The disease was staged as under;

**Figure: 1**

Stage	Description
0	Patients treated conservatively
1	Eye enucleated, completely resected on histopathological examination
2	Eye enucleated, microscopic residual tumor
3	Regional extension a. Overt orbital disease b. Pre-auricular or cervical lymph node extension
4	Metastatic disease a. Hematological metastasis (without CNS involvement) 1. Single lesion 2. Multiple lesions b. CNS extension (with/without any other site of regional/metastatic disease) 1. Prechiasmatic lesion 2. CNS mass 3. Leptomeningeal and CSF disease

The computer program SPSS 25.0 was used to evaluate the collected data. The data's normality was examined using the Shapiro-Wilk test. Age and disease duration were used to compute the mean, standard deviation, or median (IQR). For gender, side affected (left or right), and illness stage (1/2/3/4), frequency and percentage were computed. The post-stratification chi square/fisher

exact test was used to control for effect modifiers such as age, gender, length of illness, and side effects. A P-value of less than 0.05 was regarded as significant.

#### RESULTS

The mean age in this study was 4.19 ± 1.78 years, with a range of 6 months to 10 years. Seventy-one

(78.89%) of the patients were between the ages of six months and ten. With a male to female ratio of 1:4.6, 30 (33.33%) of the 90 patients were men and 60 (66.67%) were women. In our study, the average length of illness was  $4.43 \pm 1.64$  months. Table I displays the distribution of patients with additional confounding variables.

According to Table II, 60.0% of the eyes with

retinoblastoma in our study had stage 2, 20.0% had stage 1 at initial presentation, 13.33% had stage 3, and 6.67% had stage 4. Table III displays the stratification of retinoblastoma's various stage presentations according to age, gender, duration, and side affected.

**Table I: Distribution of patients with other confounding variables (n=370)**

Confounding variables		Frequency	%age
Age (years)	6 mon-5 years	71	78.89
	6-10 years	19	21.11
Gender	Male	30	33.33
	Female	60	66.67
Duration of disease (months)	≤6	79	87.78
	>6	11	12.22
Side affected	Right	49	54.44
	Left	41	45.56

**Table-II: Frequency of different stage presentation of retinoblastoma (n=90).**

Stage	No. of Patients	%age
1	18	20.0
2	54	60.0
3	12	13.33
4	06	6.67

**Table III: Stratification of different stage presentation of retinoblastoma with respect to age, gender, duration and side affected.**

Variable	Category	Stage 1	Stage 2	Stage 3	Stage 4	P-value
Age (years)	6 months–5 years	18 (25.35%)	41 (57.75%)	06 (8.45%)	06 (8.45%)	0.005
	6–10 years	00 (0.0%)	13	06	00 (0.0%)	

			(68.42%)	(31.58%)		
<b>Gender</b>	Male	10 (33.33%)	14 (46.67%)	04 (13.33%)	02 (6.67%)	0.149
	Female	08 (13.33%)	40 (66.67%)	08 (13.33%)	04 (6.67%)	
<b>Duration of disease (months)</b>	≤6	15 (18.99%)	47 (59.49%)	11 (13.92%)	06 (7.59%)	0.714
	>6	03 (27.27%)	07 (63.64%)	01 (9.09%)	00 (0.0%)	
<b>Side affected</b>	Right	11 (22.45%)	32 (65.31%)	03 (6.12%)	03 (6.12%)	0.167
	Left	07 (17.07%)	22 (53.66%)	09 (21.95%)	03 (7.32%)	

## DISCUSSION

Using the International Intraocular Retinoblastoma Classification (IIRC) system, this study examined the clinical staging of retinoblastoma at the time of presentation, paying particular attention to age, gender, laterality, and symptom duration. Of the ninety patients, sixty percent had stage 2 retinoblastoma, twenty percent had stage 1 at initial presentation, thirteen percent had stage 3, and six and a half had stage 4. At presentation, the average age was  $4.19 \pm 1.78$  years. The results of our study are consistent with literature showing that retinoblastoma typically affects children under the age of five<sup>9</sup>, with the majority being unilateral.<sup>10</sup>

Lack of public knowledge, incorrect diagnoses by non-specialists, conventional healing methods, and financial and geographic obstacles to receiving professional care are some of the factors that contribute to delayed presentation.<sup>11</sup> Missed possibilities for early detection are a result of the limited integration of eye screening into regular pediatric or neonatal health examinations.<sup>12</sup> Knowing the distribution of clinical stages at presentation serves as a baseline for evaluating the effects of upcoming awareness campaigns and screening initiatives, as well as an indicator of how well the current healthcare system detects ocular cancers. Reducing the number of children who need high-dose chemotherapy or undergo enucleation can be achieved by moving toward earlier presentation trends.<sup>13</sup> There is still a dearth of published information on the stage distribution of retinoblastoma at diagnosis in Pakistan, especially when considering national trends.

International studies have shown similar trends, with bilateral cases being discovered sooner as a result of family screening procedures.<sup>14,15</sup> Bilateral eyes showed a significantly larger percentage of early stage (32.8%) than unilateral eyes (17.8%), despite the fact that both unilateral and bilateral cases mostly appeared in Groups D and E. This emphasizes how bilateral presentation might lead to an earlier clinical examination and the importance of genetic surveillance. These findings are in line with

recent research that identified no differences in retinoblastoma stage at diagnosis based on gender, suggesting that sociocultural gender biases had no discernible effect on the time to diagnosis in this sample.<sup>16,17</sup>

Compared to older children, the percentage of early-stage diagnoses was somewhat greater for children under three. This is also in line with earlier studies that indicate older children frequently appear at more advanced stages due to delayed diagnosis and a failure to recognize symptoms.<sup>18,19</sup> Advanced phases, however, continued to predominate even in younger age groups, suggesting structural obstacles to early detection. Children with Group D and E disease presented later, at 2.7 and 2.9 years, respectively, whereas children with Group B or C disease presented at a mean age of 2.3 to 2.4 years. The idea that diagnostic delays contribute to the course of the illness and that even brief time differences can cause intraocular retinoblastoma to significantly advance is further supported by this steady age-stage change. All things considered, these results are consistent with trends seen in a number of developing nations, where advanced-stage presentation and delayed diagnosis are caused by a lack of awareness, restricted access to specialized care, and a lack of frequent pediatric eye screening.<sup>20</sup>

Early identification of leucocoria in newborns, especially in children at risk, may improve clinical outcomes. The significance of genetic counseling in the treatment of genetic retinoblastoma and early cancer detection cannot be denied. The potential of social media to increase awareness of severe diseases like retinoblastoma can be used. Retinoblastoma was discovered early because to awareness campaigns about the condition that were started by a number of centers in underdeveloped countries, including Pakistan. It is essential to address these concerns at the national level. Better medical education and training for healthcare professionals on the diagnosis of retinoblastoma is essential in this regard.<sup>21,22</sup>

This study's strength is its use of standardized clinical staging to provide useful local

epidemiological data on the stage distribution of retinoblastoma upon presentation. However, generalizability and thorough interpretation are constrained by its single center design, limited sample size, and absence of socioeconomic or outcome data. To further understand the factors influencing stage at presentation and to inform strategies for early detection and better care of retinoblastoma, future multicenter research with bigger cohorts that incorporate socioeconomic determinants and long-term outcomes are advised.

#### CONCLUSION:

The majority of children with retinoblastoma arrive with the disease in an advanced stage. In order to promote earlier discovery and enhance the chances for globe salvage and vision preservation, these findings highlight the critical need for increased public awareness, routine pediatric eye screening, and prompt referral systems. They also reflect diagnostic delays.

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