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**Medicine**

# Epidemio-Clinical Aspect of Intraocular Retinoblastoma at Sikasso Regional Hospital

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**Abstract** **Original Research Article**

***Introduction***: Intraocular retinoblastoma is a malignant neuroepithelial tumor occurring in infants and young children. It is the most common intraocular tumor. The early clinical sign is unilateral strabismus. Leukocoria already corresponds to a locally advanced stage. It is most often unilateral. In Mali, it represents 33.1% of all solid tumors. The aim of this work is to study the epidemiological and clinical aspects of intraocular retinoblastoma in the pediatric and ophthalmology department of Sikasso hospital. ***Patients and method*:** This is a retrospective study over 8 years which collected the intraocular forms of Rb. The intraocular forms without signs of clinical and radiological extraocular extension were included. The cases benefited from an ophthalmological and oncological assessment. The clinical stage of Rb was determined according to the IIRC classification. ***Results:*** Over 8 years, we have collected 29 out of 61 cases of ocular tumors, a frequency of 48%. The male sex predominated with 55% (sex ratio of 1.2). The average age was 1.23 years. 17.24% of patients had a family history of cancer. The first sign was leucocoria (68.96%) and the average consultation time was 3 months. The left eye was the most affected in 56%. The fundus shows 65.52% endophytic form. 68.97% were at stage D of the ABC classification. ***Conclusion*:** Retinoblastoma is the most common ocular tumor in children and the diagnostic delay was too long, compromising the patient's functional and vital prognosis. Early diagnosis is of utmost importance.

**Keywords:** Retinoblastoma, early diagnosis, enucleation.



## INTRODUCTION

Retinoblastoma is a malignant tumor of neuroepithelial origin occurring in infants and young children. Its prognosis is good since more than 90% of children are cured in industrialized countries. It is the most common intraocular tumor in children according to Arne [3]. Its incidence in France varies between 1/15,000 to 1/20,000 births.

It is unilateral in 60% of patients with a median age at diagnosis of about 2 years. It is bilateral in 40% of patients: the median age at the time of diagnosis is then around 1 year and tends to decrease in industrialized countries thanks to the screening of children at risk from the neonatal period. Infectious or environmental risk factors favoring the occurrence of retinoblastoma have been reported, but cannot be considered as established today.

The more frequent occurrence of retinoblastomas in children born from medically assisted procreation pregnancies has been reported, but as yet unconfirmed. Its frequency observed in Europe and America varies between 1/25,000 and 1/14,000 births. In Senegal, there are 15 cases in hospital consultation for 1000 consultants. In Morocco, its incidence is estimated at 50 new cases per year, most of which are sporadic forms.

In Mali in a study made on orbito-ocular tumors at IOTA about 130 cases, retinoblastoma was

the most common malignant tumor, 28 cases or 21.54%.

In a recent study carried out in Mali by Togo B *et al.*, from CHU Gabriel Touré, retinoblastoma represented 33.1% of all solid tumors and was second in frequency after lymphomas (39.7%).

Success in the management of retinoblastoma partly depends on early diagnosis, hence the purpose of our study.

## PATIENTS AND METHOD

We carried out a retrospective study over 8 years (from 2010 to 2018) which collected the intraocular forms of Rb. Recruitment took place respectively in the pediatric and ophthalmology departments of the regional hospital of Sikasso. We included intraocular forms without signs of clinical and radiological extraocular extension; who have benefited from a transfer to the UP of Bamako with the informed consent of the parents and/or the legal representative. All the cases included benefited from a complete ophthalmological assessment with fundus (FO) in indirect ophthalmoscopy (Schepens®), under general anesthesia (GA) whenever possible and an imaging assessment (orbitocerebral CT, ultrasound B). A systematic physical examination in pediatric oncology with evaluation of physiological constants was associated with the preoperative assessment and pre-chemotherapy if necessary. The clinical stage of Rb was determined according to the ABC classification.

**ABC RATING**

**A:** Small retinal tumors distant from the fovea and optic disc Tumor < 3mm in diameter Located more than 3mm from the fovea and more than 1.5mm from the optic disc without vitreous invasion, without associated retinal detachment.

**B:** All other tumors restricted to the retina; minimal serous retinal detachment. Retinal tumors limited to the retina but not tilting in group A and/or retinal serous detachment of less than 3 mm around the tumor base, with no visible subretinal fragment.

**C:** moderate localized serous retinal detachment; localized subretinal or intravitreal fragments. Isolated serous detachment (without subretinal fragment) between 3 and 6mm around the tumor base. Intravitreal or subretinal tumor fragments within 3mm of the base. of the tumour.

**D:** Extensive retinal serous detachment or diffuse subretinal or intravitreal spreading. Isolated serous detachment more than 3mm from the tumor base. Intravitreal or subretinal tumor fragments more than 3mm from the tumor base.

**E:** Presence of at least one of these pejorative prognostic factors for ocular preservation. Tumor masses taking up more than 2/3 of the globe Involvement of the anterior segment; Ciliary body damage; Irian neovascularization Neovascular glaucoma.

The data was collected on individual survey forms. Data entry was carried out on Microsoft office Word and Excel 2016. Data analysis was carried out on Epi info version 7.2.

## RESULTS

**Sociodemographic characteristics**

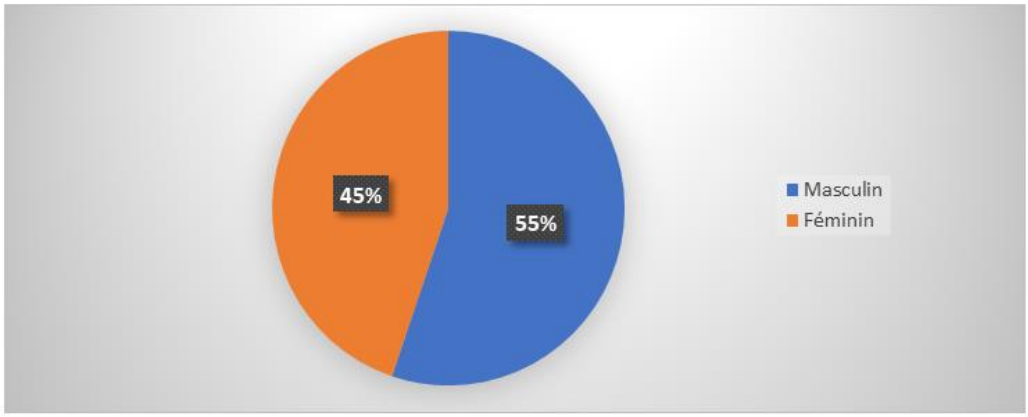


**Figure 1: Distribution of patients by gender**

The sex ratio is in favor of men 1.2.

**Table 1: Distribution of patients according to an ATCD of cancer in the family**

| Familial cancer ATCD | Workforce | % |
|---|---|---|
| YES | 5 | 17.24 |
| **NO** | **24** | **82.76** |
| Total | 29 | 100 |

The majority of patients did not have ATCD of cancer in the family, i.e. 82.76% of cases.

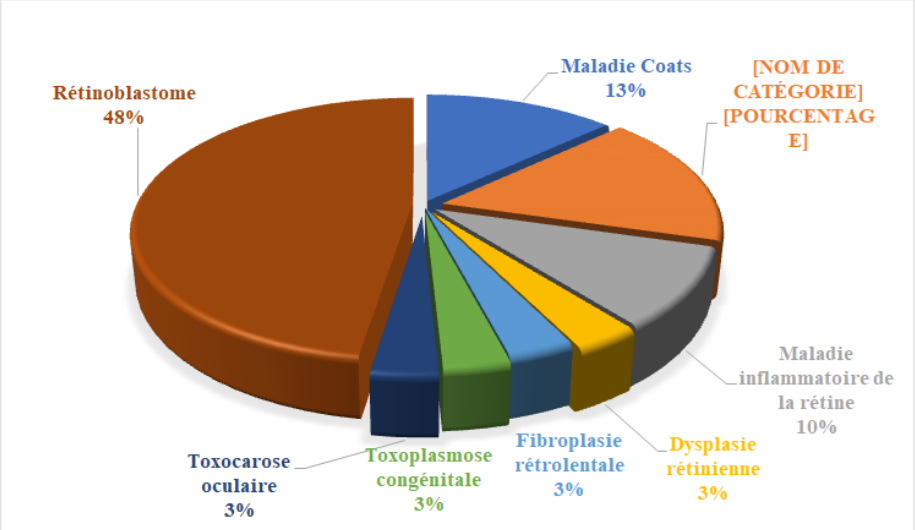


**Figure 2: Distribution of the different eye tumors compared to retinoblastoma compared to other pathologies of the eye**

Retinoblastoma accounts for 48% of cases.

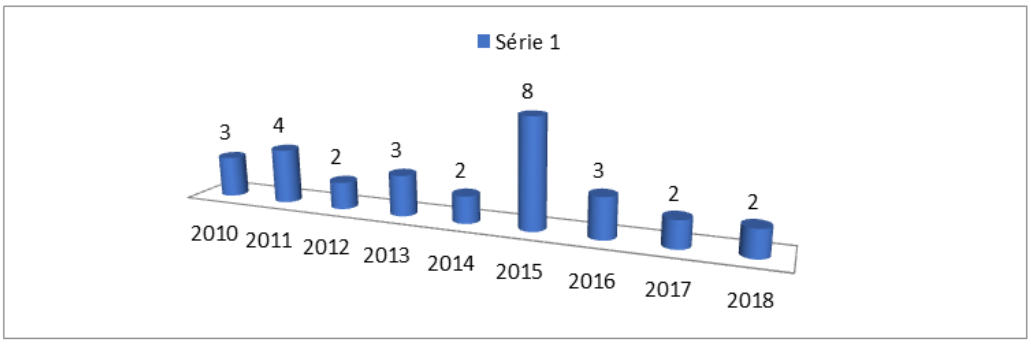


**Figure 3: Annual distribution of retinoblastoma cases (2010-2018)**

The frequency of retinoblastoma was higher in 2015, at 27.58 cases.

**Clinical data**

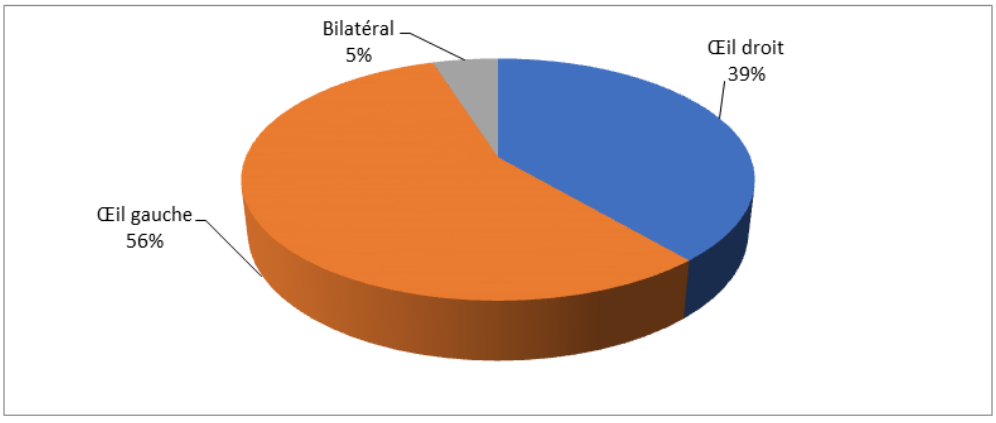


**Figure 4: Distribution of patients according to the affected eye**

The left side was the most affected with 55.17%.

**Table 2: Distribution of patients according to the first sign**

| Sign | Workforce | % |
|---|---|---|
| **Leukocoria** | **20** | **68.96** |
| Strabismus | 7 | 24.14 |
| Other associated sign** | 2 | 6.90 |
| **TOTAL** | **29** | **100** |

**Buphthalmia, glaucoma

Leukocoria was the most encountered sign with 68.96% of cases and strabismus was 24.14% of cases.

**Table 3: Distribution according to the number of intraocular tumors**

| Number of tumor | Workforce | % |
|---|---|---|
| Not mentioned | 10 | 34.48 |
| Multiple tumor | 6 | 20.69 |
| **A single tumor** | **13** | **44.83** |
| **TOTAL** | **29** | **100** |

The majority of children carried a single tumor with 44.83% of cases.

**Table 4: Distribution of patients according to the type of tumor**

| Type of tumor | Workforce | % |
|---|---|---|
| Exophytic | 6 | 20.69 |
| **endophytic** | **19** | **65.52** |
| **Not mentioned** | **4** | **13.79** |
| **TOTAL** | **29** | **100** |

The most represented tumor was endophytic with 65.52% of cases.

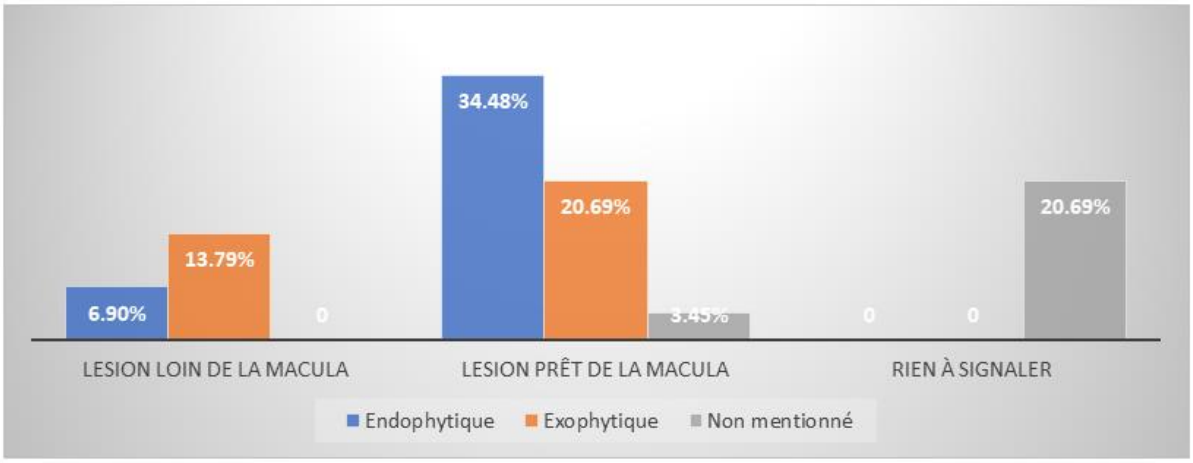


**Figure 5: Distribution by tumor site versus tumor type**

The endophytic tumor dominates at the level of lesions close to the macula.

**Table 5: Distribution of patients according to ABC classification**

| Classification of tumors | Workforce | % |
|---|---|---|
| **A** | **0** | **00** |
| B | 7 | 24.13 |
| C | 0 | 00 |
| **D** | **20** | **68.97** |
| E | O | OO |
| Unclassified | 2 | 6.90 |
| **TOTAL** | **29** | **100** |

**Stage D** was the most frequent with 68.97%

**Table 6: Distribution of patients according to the latest news.**

| Become Sick | Workforce | % |
|---|---|---|
| **FOLLOW-UP in Bamako** | **09** | **31.03** |
| NO NEWS | 20 | 68.97 |
| Total | 29 | 100 |

Patients referred to Bamako were in the minority with 31.03% of cases.

# DISCUSSION

**Frequency**

We collected 29 cases of retinoblastoma over 8 years, i.e. 3.6 cases/year. Of 61 cases of ocular tumors, 29 were retinoblastomas, i.e. 48%.

**Age and sex**

A male predominance was noted in our series, i.e. 55%; with a sex ratio of 1.2. The age group of 0-5 years was the most affected with 41.38% of cases. The mean age at diagnosis was 1.23 years. A male predominance was also recorded in Sidibé H with a sex ratio of 1.2 and an average age of 4.2 years [1]. Koné A, had found a clear male predominance (sex ratio at 2.2) and the average age was 2.5 years (3 months to 60 months) [2]. Our results agree with those of kangaroo which found age extremes ranging from 1 to 7 years to a predominance of male sex (sex ratio at 1.4).

**History of family cancers**

The interrogation and the ophthalmological clinical examination found 5 cases or 17.24% of retinoblastomas with a history of family cancers (osteosarcoma, leukemia).

The notion of family history has been reported in several studies. It was 17.1% in Jordan [3]; 7% in Brazil [4]; 9.5% in Italy [5] and 9.5 also in Algiers [6].

**Clinical aspect**

In our study, all patients were able to carry out normal activity at the time of diagnosis and the first sign was Leukocoria, i.e. 68.96%. In the majority of cases, the patients had consulted after 3 months of progression of the disease, ie 51.72%. This relatively long delay could be explained by the ignorance of the signs of retinoblastoma by local health workers.

**Tumor location**

In our study the left eye was the most affected with 56% of cases against 39% of the right eye and 5% of bilateral forms. In the series of Sidibé H, the left eye was also the most affected with 49.1% against 40% of the right eye and 10% of bilateral form [1]. C. Doutetien found 62.5% unilateral form in their study [5, 6]. Our results agree with those of H Sidibé concerning unilaterality and the predominance of left eye involvement [1].

**The fundus**

The fundus shows the presence of one or more tumors which are most often with endophytic, sometimes exophytic or mixed development [7, 8] rarely in 1 to 2% of cases, the tumor is in the form diffuse infiltrant [9, 10].

Our results agree with those of the literature: we had 65.52% of endophytic form.

**ABC RATING**

In the series provided by Palazzi MA *et al.*, from a reference center in Brazil, i.e. 91% of unilateral retinoblastomas were of stage D or E [4], whereas this percentage drops to 78.9% in the series from the institute curie reported by Lumbrosol [11] and 74% in the Batra R series from the reference center in England [12]. possibilities of preservation of visual function and even of the eyeball.

**Becoming referred patients in Bamako**

None of our patients had been seen after their evacuations to Bamako. This did not allow us to know the outcome of our patients.

# CONCLUSION

Retinoblastoma is the most common ocular tumor in children and the diagnostic delay was too long, compromising the patient's functional and vital prognosis. Early diagnosis is of utmost importance. If retinoblastoma is a therapeutic emergency, the intraocular form remains a diagnostic challenge for the doctor and requires education of all actors: the family for an alarming reaction to any leukocoria; health personnel for early referrals.

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