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RETINOBLASTOMA: ASSESSING THE LEVEL OF KNOWLEDGE OF TUMOUR BY MIDWIVES IN BRAZZAVILLE
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RETINOBLASTOMA: ASSESSING THE LEVEL OF KNOWLEDGE OF TUMOUR BY MIDWIVES IN BRAZZAVILLE

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ABSTRACT

***Objective*: To assess the level of knowledge of this tumour by midwives in two hospitals (Talangaï and Makélékélé) in Brazzaville.**
***Design* : An analytical cross-sectional study.**
***Setting* : Two maternity units (government hospitals) in the outskirts of Brazzaville**
***Subjects*: One hundred midwives who responded to a questionnaire in the form of multiple-choice question (MCQ). These midwives were randomly selected based on their availability to respond to questions. Each midwife was seen once and had 30 minutes to answer three questions, define the retinoblastoma and recognise its two main early clinical signs (leucocoria, strabismus).**
***Results*: Forty percent were able to define the retinoblastoma. For 60% leucocoria was the only sign suspect of retinoblastoma, for 80% strabismus was the only sign of retinoblastoma, only 10% had associated leucocoria and strabismus as two early signs of this cancer. Twelve percent had a score equal to 3/3, eight (8%) had a score equal to 2/3, and 80% had a score of less than or equal to 1/3.**
***Conclusion*: midwives in Brazzaville poorly understand the retinoblastoma. Training programmes should be tailored to effectively fight against mortality of this pathology.**

INTRODUCTION

Retinoblastoma is a genetic malignant tumour. The result of two mutations of the Rb gene located on the long arm of chromosome 13. This gene is inherited as an autosomal dominant mode, but works like a recessive gene because both alleles must be mutated for the tumour to develop (1). In rich countries, the average age of diagnosis is one year for bilateral forms and two years for unilateral forms (2). The average age is slightly higher in black Africa; Boubacar and *et al* (3) in Mali estimate it at 4.2 years.

Cure is the norm in developed countries (4). In black Africa, mortality from this cancer is high, because of the later consultations (3). Early diagnosis is a major asset in the strategies against mortality from this disease. In Congo, as in most countries in Africa midwives provide childhood immunisation and nutritional counselling to mothers. These paramedics are the first contact with the mother and child, they must be trained to be an effective link in the strategy against mortality from this cancer. They must be able to orientate the child to the ophthalmologist in case of suspicion of retinoblastoma.

Between 2010 and 2014, the ophthalmology department of the UHB has recorded 23 cases of retinoblastoma. They were referred for an exophthalmos. In 100% an anti-inflammatory treatment and/or anti-biotic was carried out for an average term of one month on suspicion of eyelid abscess.

MATERIALS AND METHODS

This was an analytical cross-sectional study. The study took place for a period of a month, January 2015. One hundred midwives working in two maternity units on the outskirts of Brazzaville, Talangaï and Makélékélé hospitals. These midwives had responded to a questionnaire in the form of MCQ. They were randomly selected based on their willingness to answer the questions. Each midwife 30 minutes to answer three questions. The first on the definition

of retinoblastoma. Retinoblastoma was defined as a serious ocular cancer in the childhood. The second and third were related to the recognition of two very early signs of this cancer, namely leukocoria and strabismus. Each question had three responses proposals, only one was correct. Total possible points was three (3), each correct answer was rated 1 point. Knowledge of retinoblastoma was deemed insufficient at rates of less than or equal to 1/3, average at 2/3 and sufficient at to 3/3.

RESULTS

Four percent of the midwives were able to define the retinoblastoma (Table 1).

Table 1
Assessment of the level of knowledge of midwives on the definition of retinoblastoma

Definition	Response (effective)	Frequency (%)
Correct	+ (n= 4)	4
false	- (n= 96)	96
Total	100	100

Effective: the number of midwife corresponding to the given answer
(+) Means that the answer is correct, and (-) in relation to an incorrect answer
For 60% leukocoria is the only early suspect sign of retinoblastoma. For thirty percent of them, strabismus was the only early sign of retinoblastoma. Only ten (10) % had associated leucocoria and strabismus as two highly suggestive signs of retinoblastoma.

Twelve percent had a score equal 3/3, eight (8%) had a score equal 2/3, and eighty (80) % had a score of less than or equal 1/3 (Table2).

Table 2
Assessment of the level of knowledge of midwives in retinoblastoma

Level of knowledge	Effective	Frequency (%)
Insufficient	80	80
Medium	8	8
Sufficient	12	12
Total	100	100

DISCUSSION

Good knowledge of clinical signs of retinoblastoma is essential for early diagnosis and determining prognosis. The white pupil or leukocoria and strabismus are the two major early signs of retinoblastoma (5- 8). Leukocoria indicates the presence of advanced tumours, whereas strabismus sign macular disease (5).

Leukocoria is the most common manifestation of retinoblastoma. It presents 60% of cases. This is usually the family, not the doctor who initially observed its appearance (8). The medical term Leukocoria, from Greek roots Leukos meaning white and pupil Korê have replaced today cat eye used in the first time in 1817 by Beer (9). This white pupil reflects the tumour in the pupillary area at the bottom of knowing lighting. This reflection is similar to that observed night in cat eye. In fact, parents use all kinds of expressions to translate this. They talk about weird reflection, shiny eye, strange glow, unusual look, intermittent light. Strabismus is the second early sign of the disease (6-8). It is generally binds a macular disease. It is found in nearly 20% of cases. The extension of the tumour to the macula or into the vitreous causes a loss of binocular vision. This is still the case in poor countries where exophthalmos and tumour externalising are often the first signs that motivate a medical. consultation (3, 10-13). In about two-thirds of cases (14) only one eye is affected (unilateral retinoblastoma); in the other third, tumours develop in both eyes (bilateral retinoblastoma). The number and size of tumours on each eye may vary. In certain cases, the pineal gland or the suprasellar or parasellar region (or in very rare cases other midline intracranial locations) is also affected (trilateral retinoblastoma). The position, size and quantity of tumours are considered when choosing the type of treatment for the disease. In children with the heritable genetic form of retinoblastoma there is a mutation on chromosome 13, called the RB1 gene. The genetic codes found in chromosomes control the way in which cells grow and develop within the body (15). If a portion of the code is missing or altered (mutation) a cancer may develop. Inherited forms of retinoblastomas are more likely to be bilateral. In addition inherited uni- or bilateral retinoblastomas may be associated with pineoblastoma and other malignant midline supratentorial primitive neuroectodermal tumours (PNET) with a dismal outcome; retinoblastoma that concurs with such a PNET is also known as trilateral retinoblastoma (16). A recent meta-analysis has shown that survival of trilateral retinoblastoma has increased substantially over the last decades (17). Some diseases have similar clinical symptoms of retinoblastoma, namely:

- *Persistent hyperplastic primary vitreous*: congenital developmental anomaly of the eye resulting from failure of the embryological, primary vitreous and hyaloid vasculature to regress, whereby the eye is shorter, develops a cataract, and may present with whitening of the pupil.

- *Coats disease*: a typically unilateral disease characterised by abnormal development of blood

vessels behind the retina, leading to blood vessel abnormalities in the retina and retinal detachment to mimic retinoblastoma.

- *Toxocara canis*: an infectious disease of the eye associated with exposure to infected puppies, which causes a retinal lesion leading to retinal detachment.

- *Retinopathy of prematurity*: associated with low birth weight infants who receive supplemental oxygen in the period immediately after birth, it involves damage to the retinal tissue and may lead to retinal detachment.

The priority of retinoblastoma treatment is to preserve the life of the child, then to preserve vision, and then to minimize complications or side effects of treatment. The exact course of treatment will depend on the individual case and will be decided by the ophthalmologist in discussion with the paediatric oncologist.(18). Children with involvement of both eyes at diagnosis usually require multimodality therapy (chemotherapy, local therapies). The various treatment modalities for retinoblastoma includes (18):

- *Enucleation of the eye*: most patients with unilateral disease present with advanced intraocular disease and therefore usually undergo enucleation, which results in a cure rate of 95%. In bilateral Retinoblastoma, enucleation is usually reserved for eyes that have failed all known effective therapies or without useful vision.

- *External beam radiotherapy (EBR)*: the most common indication for EBR is for the eye in a young child with bilateral retinoblastoma who has active or recurrent disease after completion of chemotherapy and local therapies. However, patients with hereditary disease who received EBR therapy are reported to have a 35% risk of second cancers (19).

- *Brachytherapy*: brachytherapy involves the placement of a radioactive implant (plaque), usually on the scleral adjacent to the base of a tumour. It used as the primary treatment or, more frequently, in patients with small tumours or in those who had failed initial therapy including previous EBR therapy.

- *Thermotherapy*: thermotherapy involves the application of heat directly to the tumour, usually in the form of infrared radiation. It is also used for small tumours

- *Laser photocoagulation*: laser photocoagulation is recommended only for small posterior tumours. An argon or diode laser or a xenon arc is used to coagulate all the blood supply to the tumour.

- *Cryotherapy*: cryotherapy induces damage to the vascular endothelium with secondary thrombosis and infarction of the tumour tissue by rapidly freezing it. Cryotherapy may be used as primary therapy for small peripheral tumours or for small recurrent tumours previously treated with other methods.

- *Systemic chemotherapy*: systemic chemotherapy has become forefront of treatment in the past decade, in the search of globe preserving measures and to avoid the adverse effects of EBR therapy. The common indications for chemotherapy for intraocular retinoblastoma include tumours that are large and that cannot be treated with local therapies alone in children with bilateral tumours. It is also used in patients with unilateral disease when the tumours are small but cannot be controlled with local therapies alone.

- *Intra-arterial chemotherapy*: Chemotherapeutic drugs are administered locally via a thin catheter threaded through the groin, through the aorta and the neck, directly into the optic vessels) (20).

- *Nano-particulate chemotherapy*: To reduce the adverse effects of systemic therapy, subconjuctival (local) injection of nanoparticle carriers containing chemotherapeutic agents (carboplatin) has been developed which has shown promising results in the treatment of retinoblastoma in animal models without adverse effects (21, 22).

Good prognosis depends upon early presentation of the child in health facility, in the developed world, retinoblastoma has one of the best cure rates of all childhood cancers (95-98%) (23, 24). Late presentation of the child in hospital is associated with poor prognosis (23, 25).

In conclusion, midwives in Brazzaville have limited knowledge on Retinoblastoma. Better training for midwives may contribute to reduction in the mortality of Retinoblastoma in Brazzaville.

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