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Congenital Glaucoma: Epidemiological, Clinical and Therapeutic Aspects in the Ophthalmology Department of Abass Ndao Hospital

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Summary

Introduction: The objective of this study was to evaluate epidemiological, clinical, therapeutic and prognostic characteristics of congenital glaucoma in our context.

Material and methods: This was a retrospective descriptive and analytical study over 11 years, including the medical records of all children followed for congenital glaucoma. We collected the following data: age of the child, age of discovery of symptoms, sex, history, clinical and therapeutic data. The monitoring parameters were the appearance of the filtration bubble, the corneal diameter, the transparency of the cornea, the ocular tone and the appearance of the papilla. After 6-month follow-up, the surgical intervention was considered successful if the final IOP was less than 15 mmHg without or with hypotonizing medical treatment.

Results: Congenital glaucoma represented 4.5% of ophthalmic-pediatric consultations. The average age was 9 months and the sex ratio 1.54. Megalocornea found in 85% of cases. The average corneal diameter was 14 mm and corneal edema was found in 80% of cases. The average ocular tone was 22.63 mmHg. The fundus was accessible in 36.4% of cases. The average response time was 20.4 days. Trabeculectomy was done in 69.5%. After a 6-month post-operative follow-up, an average ocular tone of 14 mmHg was noted in 88% of cases. There was a lightening of the cornea in 92% of cases, a stabilization of the corneal diameter in 100% of cases and a normal optic disc in 24% of cases with a regression rate of the excavation of 26.66%. A failure rate of 12% was observed.

Conclusion: The frequency of congenital glaucoma is quite high in our context, with a high rate of consanguinity. Despite a delay in diagnosis and treatment and difficult post-operative follow-up, a success rate at 6 months post-therapy of 88% was noted.

Keywords: Megalocornea; Ocular Hypertonus; Trabeculectomy; Functional Prognosis

Introduction

Primary congenital glaucoma is defined by ocular hypertonia following an abnormality of the iridocorneal angle existing at birth or appearing in the first three years of life [1,2]. It is a serious pathology responsible for 5% of childhood blindness and 0.01 to 0.04% of all blindness cases according to the WHO [3,4]. In sub-Saharan Africa, very few epidemiological studies have been devoted to it. Hospital frequencies of 0.10% and 0.08% were found respectively in Senegal [5] and Togo [6]. It constitutes a diagnostic and therapeutic emergency. Its management is difficult in Africa. Indeed, there are few ophthalmologists, the diagnosis is often late, the follow-up is long with many therapeutic failures. The prognosis depends on the type of glaucoma, in fact, it is very different depending on whether it is unilateral glaucoma, of recent onset, around the age of three months or bilateral buphthalmia diagnosed at the birth with total edematous corneal opacity [7]. The aim of this study was to evaluate the epidemiological, clinical, therapeutic and prognostic characteristics of congenital glaucoma in our context.

Materials and Methods

This was a retrospective study over 11 years, from January 1, 2010 to December 31, 2020. It included the medical records of all children with congenital glaucoma, whether operated or not, who

had undergone at least two control examinations under general anesthesia (GA), with a 6-month follow-up. Children operated on in another hospital structure, juvenile and secondary glaucoma and those with no more than two GA control examinations were not included. We analyzed the following data: the child's age and the age of discovery of symptoms, sex, history, clinical and therapeutic data. The clinical examination was carried out under general anesthesia and made it possible to measure the corneal diameter, assess its transparency, look for other ocular anomalies, measure the IOP using the Perkins applanation tonometer and perform the fundus if possible. Surgery was performed at the same time either by trabeculectomy associated with peripheral iridectomy or by deep sclerectomy. Hypotonizing medical treatment or diode cyclo-weakening were carried out if necessary.Postoperative care included topical, locoregional and systemic corticosteroids and antibiotics. Postoperative monitoring was carried out on day 1 (D1), D3, D7 and D15 and examinations under general anesthesia control were carried out on 1 month (M1), M3, M6 and then twice a year. The monitoring parameters were the appearance of the filtration bubble, the diameter and transparency of the cornea, ocular tone and the appearance of the papilla. The intervention was considered successful if the final IOP was less than 15 mmHg without or with hypotonizing medical treatment. The success was complete, if the IOP, without any medical treatment of glaucoma was less than 15mmHg. Success was partial if the IOP was maintained below 15mmHg with the addition of hypotonizing treatment. An IOP greater than 15mmHg, with combination of hypotonizing treatment, was considered a failure. In cases where the intraocular pressure was greater than 15 mmHg, treatment with hypotonizing eye drops was instituted. In cases where intraocular pressure was too high during a check-up, revision was performed by trabeculectomy or by transscleral diode laser cyclophotocoagulation. The other criteria for success were: corneal transparency improvement, stabilization of the diameter of the cornea, the presence of a filtration bubble, regression or stabilization of the excavation and recovery of good visual acuity. The analysis was carried out with the following software: Excel 2016 and Epi info

Results

Epidemiological Data

During our study period, 42,944 patients were consulted in the department, including 2,940 children aged under 15 years old. Out of 132 files of glaucomatous children consulted, 102 (177 eyes) were retained. Congenital glaucoma represented 4.5% of ophthalmic-pediatric consultations and 0.3% of general consultations. Congenital glaucoma was bilateral in 73.1% of cases and unilateral in 26.9% of cases. The average age was 9 months with extremes of 1 month and 12 years (Figure 1). The average age of discovery was 6 months ±11 with extremes of 1 and 48 months. The median was 2 months. The sex ratio was 1.54.

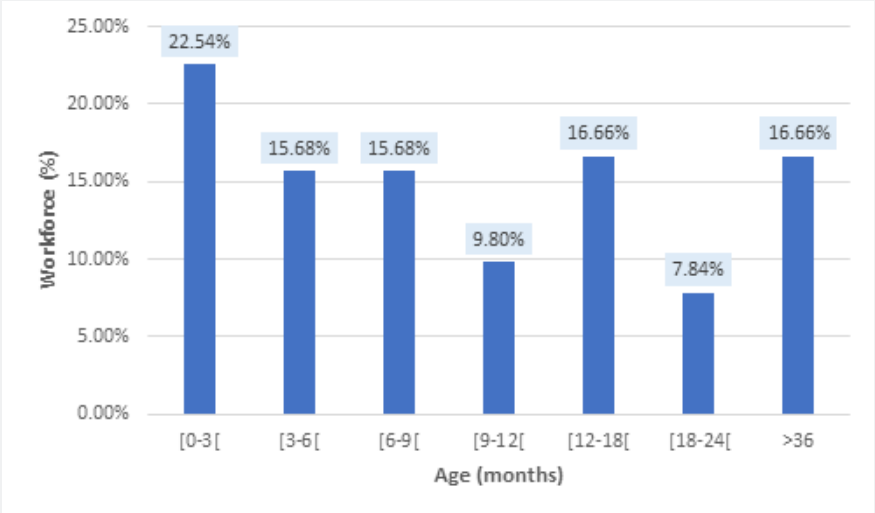

Figure 1: Distribution according to different age groups.

Background

Neonatal suffering was noticed in 9.8% of cases (10 children), psychomotor and weight delay in 27.45% of cases (28 children), trisomy 21 in 2.9% of cases (3 children), Von Recklinghausen's neurofibromatosis in 0.98% of cases (1 child), sickle cell disease in 1.96% of cases (2 children) and a notion of congenital glaucoma in siblings in 1.96% of cases (2 children). The notion of parental consanguinity was found in 49.6% of cases.

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Clinical Data

Megalocornea was found in 85% of cases (150 eyes), tearing in 55% of cases and photophobia in 34% of cases (Figure 2). The horizontal corneal diameter at the time of surgery varied between 10.5 and 17 mm with an average of 14 mm. Corneal edema was noted in 80% of cases and a clear cornea in 15% of cases (Figure 3). Before treatment, average intraocular pressure was 22.63 mmHg with extremes of 9 and 41 mmHg (Table 1). The fundus was accessible in 36.4% of eyes and showed among them, an excavated papilla in 66.66% of cases. An ocular abnormality was associated with congenital glaucoma in 23.7% of cases (Table 2).

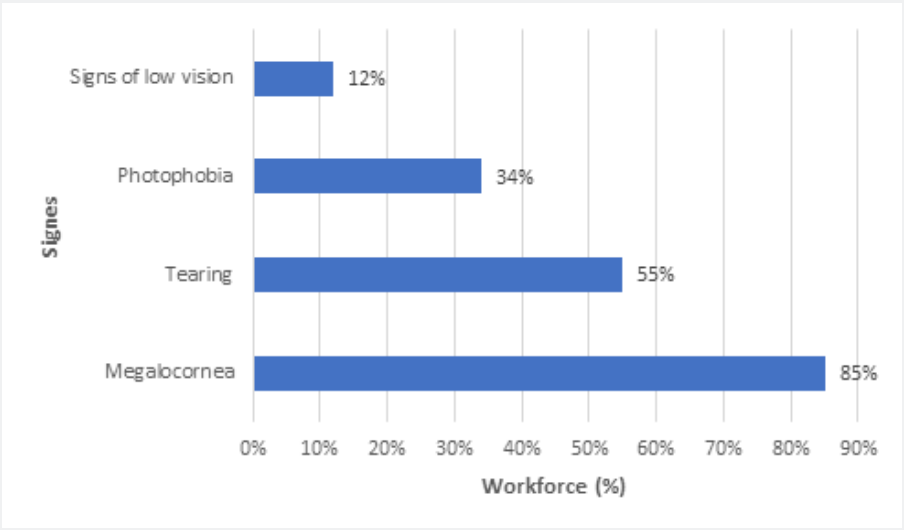

Figure 2: Distribution of patients according to the circumstances of discovery.

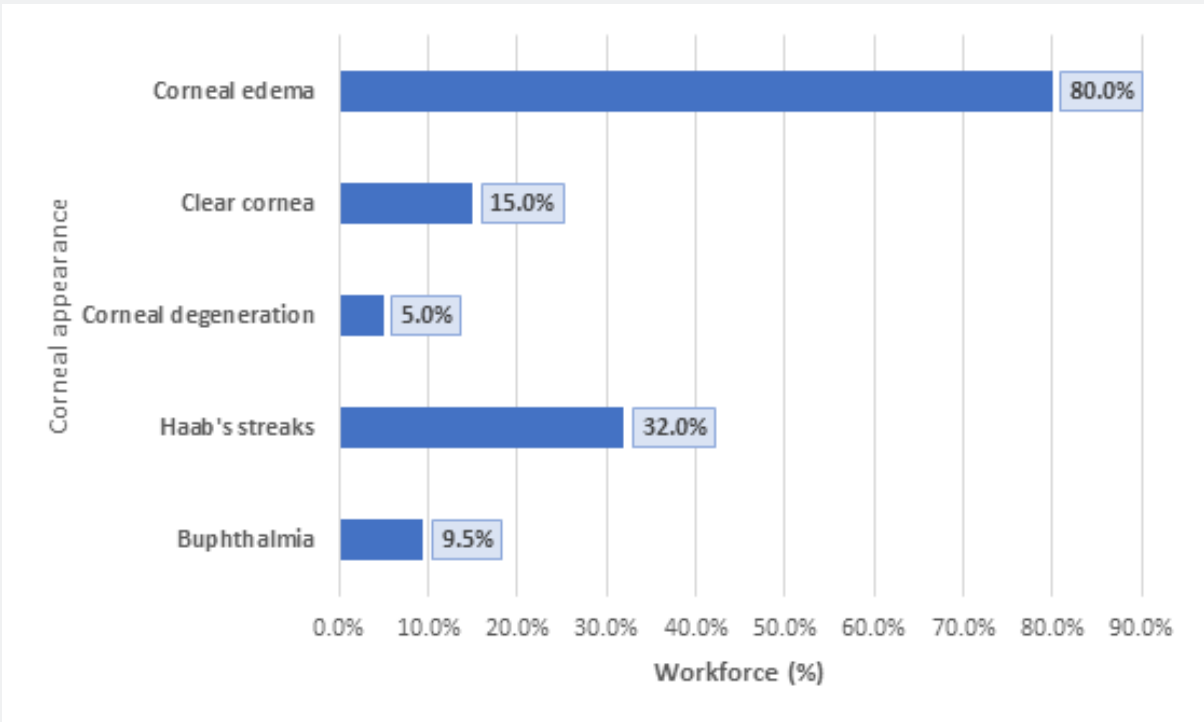

Figure 3: Distribution according to corneal appearance..

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Table 1: Distribution of the eyes according to preoperative intraocular pressure.

Intraocular Pressure (mmHg)	Number of Cases	Percentage (%)
[9 - 15]	30	17
[15 - 20]	32	18
[20 - 25]	63	35,6
[25- 30]	45	25,4
>30	7	4

Table 2: Distribution according to associated ocular lesions.

Associated Ocular Abnormalities	Number of Cases	Percentage (%)
Posterior embryotoxon	2	1,12%
Cataract	8	4,5%
Peters' anomaly	4	2,26%
Lens subluxation	9	5,08%
Incomplete aniridia	2	1,12%
Strabismus	12	6,78%
Nystagmus	7	3,95%

Therapeutic Data

The average intervention time was 20.4 days with extremes of 7 and 45 days and a standard deviation of 8 days. The median was 20 days. Trabeculectomy was performed in 69.5% of cases (123 eyes), deep sclerectomy in 5.08% (9 eyes), hypotonizing medical treatment alone in 20.3% (36 eyes) in forms with normotonia and a transscleral diode laser cyclophotocoagulation in 1.70% of cases. Intraoperative complications were observed in 5 cases (3.76%), such as vitreous leak in 2 cases (1.5%), abundant hyphema in 2 cases (1.5%) and athalamia in 1 case (0.75%). Postoperatively, complications were noted in 12 cases (9.02%). There were 6 cases of hyphema (4.5%), 4 cases of hypothalamia (3%) with conjunctival Seidel and 2 cases of retinal detachment (3.25%). After one month of post-operative follow-up, there was clearing of the cornea in 65% of cases and a still cloudy cornea in 35% of cases. The average ocular tone was 13 mmHg with extremes of 8 and 22 mmHg. The filtration bleb was present in 89.5% of cases (120 eyes), flat in 7.45% (10 eyes) and encysted in 2.98% (4 eyes). Excavation was also noted in 65% of cases with a regression rate of 1.6%.Total success was noted in 52% of cases and partial in 33% of cases (Table 3). Revision surgery was necessary for 20 eyes (11.3% of cases). At 3 months postoperatively, the filtration bubble was good in 65.5% of cases, flat in 24.7% of cases and encysted in 9.8% of cases. There was clearing of the cornea in 75% of cases (90 eyes) and a still cloudy cornea in 25% of cases (30 eyes). The average ocular tone was 15 mmHg. The total success rate was 42.5% (Table 4). After 6 months of postoperative follow-up, the bubble was present in 50.5% of eyes, flat in 31.50% of cases and encysted in 18.00% of cases. Of the 95 eyes checked at M6, there was clearing of the cornea in 92% of cases (87 eyes) and a still cloudy cornea in 8% of cases. There was stabilization of the diameter in 100% of the eyes with an average diameter of 14.5 mm. The average ocular tone was 14 mmHg. The partial success rate was 58% (Table 5). A re-intervention at M6 was carried out for 10 eyes; there were 8 trabeculectomy revisions and 2 transscleral diode laser cyclophotocoagulation procedures.

Table 3: Distribution of eyes according to tonometric success at M1.

Result	Number of cases	Percentage (%)
Total success	92	52
Partial success	58	33
Failure	27	15
Total	177	100

Table 4: Distribution of eyes according to tonometric success at M3.

Result	Number of cases	Percentage (%)
Total success	51	42,5
Partial success	56	46,5
Failure	13	11
Total	120	100

Table 5: Distribution of eyes according to tonometric success at M6.

Result	Number of cases	Percentage (%)
Total success	29	30
Partial success	55	58
Failure	11	12
Total	95	100

Discussion

Congenital glaucoma represents 46% of all childhood glaucomas and 5.7% of all glaucomas [8]. Despite its rarity, congenital glaucoma represents 18% of causes of blindness in children worldwide [8,9]. Its prevalence at birth varies across the world. It is 1 case in 5000 to 22000 births in the West [10]. In Senegal, previous studies carried out by Wade [11] in 1976 and by Ndiaye [5] in 1990 had found frequencies of 0.16% and 0.10% respectively. In our series, a frequency of 0.3% was observed. This could be explained by demographic growth and better knowledge of the disease. The average age at diagnosis varies depending on the region, it was 9 months in our study, 14 months in Morocco, 2.3 months in Spain and 3.8 months in France [2]. Megalocornea was the main warning sign of congenital glaucoma in our study while photophobia and clear tearing were classically described as the first warning signs. This indicates an often-late diagnosis in our series.The normal corneal diameter is 9.5 mm at birth, 10 mm at 6 months, 11.5 mm at 1 year and 12 mm at the age of two years [12]. Any corneal diameter exceeding these values by 1 to 2mm is increased and defines the megalocornea [13]. In our

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study, the corneal diameter was between 10.5 and 17 mm with an average of 14 mm; our results were similar to those of Hilal [14] who observed diameters between 11 and 18 mm with an average of 14.05. ±1.8mm. Essuman noted in a study involving 19 eyes of 12 patients, a corneal diameter of between 12 and 16 mm [15]. This disparity in results compared with data in the literature can be explained by the variation in the age of patients from one series to another. In our series, the average IOP measured by the Perkins tonometer under GA with fluothane was 22.63 mmHg with extremes of 9 and 41 mmHg. In the literature, it varies between 24 and 36 mmHg, thus AL-Hazmi found an average IOP of 30 mmHg in a series of 254 eyes [16]. We were able to visualize the fundus in only 36.4% of cases, and among these we noted an excavation in 66.66% of cases, optic atrophy in 25% of cases and a normal fundus in 8.33% of cases. Our results were comparable to the data in the literature, in particular to the study by Al Anazi carried out in 2013 on 325 eyes in which visualization of the papilla was only possible in 31% of cases and found an average C/D ratio of 0.69 [17]. In our series, these figures were underestimated as papillary excavation could not be assessed in 63.6% of cases due to frank corneal oedema making the FO inaccessible. These cases correspond to the most serious forms. The papillae masked by the oedema are a priori more excavated because of the major hypertonia [12]. Trabeculectomy was the main surgical technique used in our series. It was performed in 69.5% of eyes. It is the technique most frequently used in several series described in the literature, as it has several advantages in the treatment of congenital glaucoma. It is a well-known technique with a short learning curve. However, in our series, 43 eyes (24.7%) did not undergo surgery and received supplementary hypotonising medical treatment. This was the involutional form of congenital glaucoma described by Lockie [18].Therapeutic success was judged on the normalization of IOP, the clearing of the cornea, the stabilization of corneal diameter and the regression or stabilization of papillary excavation in our study. A success rate of 88% was obtained after 6 months post-surgery. Detry-Morel [19] reported a success rate of 71.05% 12 months after surgery and Agarwal [20] 86.7% after 18 months. Our success rate appears to be better than that reported in the literature, but these results should be put into perspective given the short follow-up time. Repeat surgery was performed in 30% of cases in our series. This could be explained by fibroblastic proliferation, which is very important in newborns and occurs in the first few days after surgery. Post-operative complications were noted in our study. These accounted for 6.2% of cases and included hyphaema, hypothalamia with conjunctival Seidel and retinal detachment. Our complications are similar to those described by Mandal [4].

Conclusion

In our series, congenital glaucoma represented 4.5% of childhood pathologies and 0.3% of general consultations. Megalocornea was the most frequent sign. Trabeculectomy was our main treatment method. Our therapeutic results were satisfactory after a 6-month follow-up.

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