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Primary Congenital Glaucoma in Kano, Northern Nigeria

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Abstract

Background: Primary congenital glaucoma is one of the causes of childhood blindness and it appears to be common in our environment. This study evaluated the pattern of presentation and management of primary congenital glaucoma in children.

Patients and Methods: This was a prospective questionnaire-based study of primary congenital glaucoma over a three year period. The information analyzed included patients' bio-data, vision, laterality of disease, presenting symptoms, family history, and history of traditional medicine application to the eye. Others included horizontal corneal diameter, intra ocular pressure and outcome of management.

Results: There were 26 patients, 20 were males and 6 were females (M:F=3.3:1). Their ages ranged between 3 months and 5 years. Over a half of the patients presented during the first year of life. Primary congenital glaucoma was bilateral in 19 patients (73%). The commonest presenting symptoms were large eyes (85%), cloudy eyes (77%), photophobia (46%) and lacrimation (31%). In 44 eyes (98%) the intra ocular pressure was 20-49 mmHg before surgery and below 20 mmHg after surgery. Ttrabeculectomy resolved almost all the presenting symptoms, reduced intra-ocular pressure (IOP) and improved vision in all the patients. No sight threatening complications were recorded.

Conclusion: Primary congenital glaucoma is common in our environment. Trabeculectomy remains a valuable treatment modality because it abolished most of the symptoms and IOP.

Key Words: Congenital Glaucoma, Childhood Blindness, Trabeculectomy.

Introduction

Primary congenital glaucoma (PCG) is rare with estimated incidence of 1:10000 live births[1]. The incidence is not known in our environment. PCG arises from maldevelopment of the trabeculum and irido-trabecular junction, and is not associated with other ocular anomalies. This form of trabecular dysgenesis is characterized by absence of angle recess and direct insertion of the iris to the trabeculum. PCG has autosomal recessive mode of inheritance[2], and it accounts for a large percentage of childhood blindness worldwide[3]. It is responsible for 2% of individuals in blind institutions[4]. PCG accounts for 10.8% of blind children in Sao Paulo, Brazil and is only second to cataract as a cause of childhood blindness[5]. In this study we wanted to evaluate the pattern of presentation and outcome of management of PCG in children.

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Patients and Methods

This was a questionnaire-based prospective study of all patients presenting with primary congenital glaucoma at the Aminu Kano University Teaching Hospital between June 2003 and June 2006. Information collected includes the patient's age, sex, tribe, laterality of disease, hospital number, vision, history of traditional eye medication (TEM) application, history of consanguinity, family history, number of health facilities visited before presentation, and socio economic status. The presenting symptoms and signs were also noted. Patients were carefully examined for the presence of other congenital malformations. Baseline investigations done includes hemoglobin, genotype, urea and electrolytes. Informed consents were obtained for trabeculectomy from the parents. Examination under anaesthesia (EUA) was performed under ketamine general anaesthesia. Topical amethocaine 1% was instilled

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and intraocular pressure (IOP) measured with Schiotz tonometer, 2 closest readings were considered. The post operative IOP was measured at 6 weeks. Edge to edge horizontal corneal diameter was measured in millimeters with calipers. The eye was examined and fundoscopy done with indirect ophthalmoscope (although the view was often poor due to corneal edema). All patients had trabeculectomy without applying anti fibrotic agents. A superior rectus 4/0 silk suture was carefully applied, then limbal based conjunctiva flap was raised, conjunctiva was carefully dissected to the limbus (often there was conjunctivisation of cornea hence the need for caution). Hemostasis was secured with cautery. 4x4 mm outer scleral flap was raised. An inner triangular flap was raised and excised, peripheral iridectomy was done. The outer flap was sutured at 4 edges with 9/0 silk. Anterior chamber was reformed with saline when necessary, continuous 6/0 vicryl suture was applied to the conjunctiva. Gentamycin (20mg) and Dexamethasone (4mg) was injected under the conjunctiva, and 1% Atropine eye drop applied before padding the eyes. Post operative eye drops given included Ofloxacin, 1% Prednisolone and 1% Atropine. Appropriate doses of oral Amoxicillin and Paracetamol were also given. Patients were discharged after a week with first follow up visit at 2 weeks. Post operative eye examination was done to assess corneal clarity, reduction of photophobia, cessation of corneal enlargement, and reduction in IOP. Refraction under anesthesia was done after 6 weeks and patching for amblyopia advised if there was indication to do so. All the patients were followed up for at least 6 months.

Results

There were 26 patients, 20 were males and 6 were females (M:F=3.3:1). Their ages ranged from 3 months to 5 years. Eighteen patients (69%) presented during the first year of life, 4 patients (15%) between 1 and 2 years, 2 (8%) at the age of 3 years, 1 (4%) at the age of 4 years, and 1 (4%) at the age of 5 years.

Table 1: Horizontal corneal diameter in 45 eyes of 26 patients with primary congenital glaucoma

Horizontal Corneal Diameter in Millimeters	Number of Eyes
12	12
13	11
14	9
15	8
16	4
17	1

Table 2: Distribution of intra ocular pressures before and after surgery in 45 eyes of 26 patients with primary congenital glaucoma

Before		After	
IOP in mmHg	No. of eyes (%)	IOP in mmHg	No. of eyes (%)
20-29	6 (13)	10-14	28 (62)
30-39	25 (56)	15-19	16 (36)
40-49	13 (29)	20-24	1 (2)
50+	1 (2)	25+	-

Surgery for congenital glaucoma accounted for 21% of surgeries done on children below the age of 5 years during the study period. Nineteen patients (73%) had bilateral disease while in 7 (17%) patients the disease was unilateral. Forty five eyes were involved, 24 right eyes and 21 left eyes. Four (15%) patients were products of consanguineous marriage; of this group, 3 had bilateral and 1 had unilateral buphthalmos. Twenty patients (72%) were of poor socio economic background.

Twenty four (92%) patients had been to other health facilities before coming to our hospital. Twenty three (88%) patients had not applied traditional eye medication, while 3 (12%) patients had applied topical traditional eye medicine. There was no family history of similar disease or other congenital abnormality in any of the children.

The commonest presenting complaints were large eyes in 22 (85%) patients, cloudy eyes in 20 (77%) patients, and tearing in 8 (31%) patients. All the

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patients had corneal edema, and 18 patients (69%) had healed breaks in the descemet membrane (Haab's striae). Forty eyes had corneal diameter of 12-15 mm, 5 eyes had corneal diameter of 16mm or, more (table 1). Forty four eyes had intra ocular pressure (IOP) of 20-49 mmHg; only 1 eye had IOP above 50 mmHg. Forty four eyes had post operative IOP of less than 20 mmHg (table 2).

All patients had reduced corneal edema from first post operative day and this improved remarkably before discharge. One child with bilateral PCG had failure of filtration surgery (due to fibrosis) after 11 months of treatment but improved with bleb revision. Four patients had shallow anterior chamber due to either excessive filtration or, wound leak which improved with pressure padding. No post operative endophthalmitis, or other sight threatening complications noted in all the patients.

Discussion

The high rate of surgery for PCG in this report indicates that the disease is common in our environment. PCG was commoner in boys than in girls in this study as in other reports[6]. Most of the patients presented during the first year of life. This probably accounted for the impressive outcome of management because early presentation and intervention is more likely to offer better prognosis than late presentation. In 10% of cases congenital glaucoma is transmitted as autosomal recessive trait[7]. Although consanguineous marriages can encourage transmission of autosomal recessive disorders[8], only 4 (15%) patients in this study were product of such marriages. Majority of congenital glaucoma are not hereditary and the risk of having another child after birth of a boy with the disease is less than 3%, while after birth of a girl the risk is mimnimal[9]. This low familial tendency reflects the absence of family history in the present study.

Most parents had visited other health facilities before presentation, resulting in delayed intervention with permanent opacification and blindness. Some patients applied TEM the nature of which could not be ascertained. Such medications can introduce infection or cause injury to the cornea. Application of TEM is reported as the cause of removal of the eye in an earlier study[10]. This practice is not unusual because traditional eye healers are easily accessible in rural areas; cultural beliefs may contribute to this practices.

All the patients had trabeculectomy. Goniotomy is said to be successful in intra ocular pressure control in 93% of patients even after 5 years[11], but, we lack surgical Gonio lens and Goniotome in our hospital. Besides, most of the patients presented with marked corneal edema, and the poor view negates against the procedure. Trabeculotomy on the other hand is possible even in the presence of corneal opacity[12,13]. Trabeculectomy was the only procedure possible in this study, although the procedure is more technically difficult because of scleral thinning and carries a higher risk of failure[14,15] than primary trabeculotomy and goniotomy[16]. We rather performed trabeculectomy despite report of poor long term glaucoma control with filtering surgery due to fibrosis[17], because it is the only procedure possible in our hospital. More recent studies indicates that

Le glaucome congénital primaire à Kano, au nord du Nigeria

Résumé

Arrière-plan: Le glaucome congénital primaire est une des causes d'aveuglement infantile et il semble être répandu dans notre environnement. Cette étude a évalué le mode de manifestation et la gestion du glaucome congénital primaire chez les enfants. ***Malades et méthodes:*** Il s'agit d'une étude à base de questionnaire du glaucome congénital primaire pendant une période de trois ans. Les données analysées comprennent les données biographiques des malades, l'état de la vue, la latéralité de la maladie, les symptômes manifestés, l'histoire familiale, le dossier du traitement de l'œil par la médecine traditionnelle. D'autres données sont le diamètre cornéen horizontal, la pression intraoculaire et le résultat de la gestion. ***Résultats:*** Il y avait 26 malades, dont 20 garçons et 6 filles (M:F=3,3:1). Ils étaient âgés de 3 mois à 5 ans. Plus de la moitié

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des malades se sont présentés pendant la première année de leur vie. Le glaucome congénital primaire était bilatéral chez 19 malades (73%). Les symptômes les plus fréquents étaient de grands yeux (85%), des yeux nuageux (77%), la photophobie (46%) et le larmoiement (31%). Dans 44 yeux (98%), la pression intraoculaire était de 20 à 49 mmHg avant la chirurgie et au-dessous de 20 mmHg après la chirurgie. La trabéculectomie a guéri presque tous les symptômes initiaux tout en réduisant la pression intraoculaire (PIO) et a amélioré la vue chez tous les malades. Nous n'avons enregistré aucune complication pouvant constituer un danger pour la vue. ***Conclusion:*** Le glaucome congénital primaire est répandu dans notre environnement. La trabéculectomie s'avère être un bon moyen de traitement parce qu'elle abolit la plupart des symptômes et la PIO. ***Mots-clés:*** Glaucome congénital, aveuglement infantile, trabéculectomie.

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