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Buphthalmos: a Twelve-year Review at Ibadan

CO Bekibele*, BA Olusanya**

Summary

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Objective: To review the presentation, management, follow-up and outcome of management of children presenting with buphthalmos at the University College Hospital (UCH), Ibadan.

Methods: This was a retrospective review of cases of buphthalmos seen at the UCH Ibadan, between 1991 and 2002. The case notes of patients seen in the Eye Clinic during the period were reviewed. All children diagnosed with buphthalmos or congenital glaucoma were identified, and their case notes studied in greater details.

Results: Twenty seven eyes in 15 children with buphthalmos were reviewed. This represented 0.1 percent of all new patients, and 0.6 percent of new paediatric cases seen within the study period. Their ages ranged between 22 days and nine years (mean, 25.5 months). Sixty percent were aged 18 months or less, 80 percent were boys and the disease was bilateral in 80 percent of cases. Only one (6.7 percent) patient presented with the classical triad of epiphora, photophobia and blepharospasm, while four (26.7 percent) presented with both epiphora and photophobia. Two (13.3 percent) patients had associated cardiac abnormalities. Of the 27 buphthalmic eyes, 21 (77.8 percent) had trabeculectomy, with good intraocular pressure control (i.e. < 21mmHg) after surgery in 14 eyes (66.7 percent). Of these, three (21.4 percent) required medication to achieve such control. The duration of follow-up from the time of initial presentation ranged between 2.5 months and 6.75 years (mean, 25.9 months). Six (40 percent) of the patients were followed up for more than a year, while 60 percent were lost to follow-up. Out of 10 eyes in which the visual acuity was objectively measured, two (20 percent) had good visual outcome (i.e. visual acuity 6/18 or better), while six had poor outcome due to complications associated with late presentation. Due to lack of appropriate facilities, visual acuity was not assessed objectively in 17 eyes.

Conclusion: Buphthalmos is a rare disease in Ibadan and occurs predominantly in males. Majority of cases are bilateral. In this study, trabeculectomy achieved adequate pressure control in the majority of cases and as such, it appeared to be a good option in the management of buphthalmos in black patients. The main problems were late presentation and poor follow up. The public needs to be educated on making early use of available health services. There is also a need to introduce, equip and provide support for paediatric ophthalmology units.

Introduction

BUPHTHALMOS (ox eye), defined as enlargement of the eyeball, is a characteristic feature of congenital glaucoma.[1] It occurs as a result of stretching and thinning of the sclera and cornea from increased intraocular pressure occurring before three years of age. Buphthalmos does not usually occur after the age of three years because beyond that age, the sclera and cornea become resistant to stretching. It has an estimated incidence of 1 in 12,500 live births in Western Europe.[1] It is an important cause of preventable blindness in children, accounting for 2.5 to 10 percent of all registered blind children.[2] Buphthalmos is responsible for 10.9 percent of cases of childhood blindness in south eastern Nigeria,[3] and four percent of childhood blindness in Northern Ireland.[4] Approximately 65 percent of affected patients are male, and the disease is bilateral in 75 percent of cases.[5] Its inheritance is autosomal recessive with incomplete penetrance but it seems more likely to be multi-factorial.[1] In primary congenital glaucoma, the commonest variety of

University College Hospital, Ibadan

Department of Ophthalmology

* Lecturer

** Registrar

Correspondence: Dr CO Bekibele.
E-mail: cob150@yahoo.com

congenital glaucoma, a developmental abnormality of the anterior chamber angle leads to obstruction of aqueous outflow without a consistent association with other ocular or systemic developmental abnormalities. Most patients present within the first year of birth,[5] with the usual presenting symptoms being epiphora, blepharospasm, and photophobia. Cloudiness of the cornea and poor vision are also common. The raised intraocular pressure also causes the associated cupping of the optic discs, and the gradual development of visual field defects.

The management of buphthalmos is primarily surgical with goniotomy and trabeculotomy being the procedures of first choice.[6] Trabeculectomy used to be a secondary procedure when geniotomy had failed but recently, it has been advocated that trabeculectomy should be performed as the primary procedure, with success rates ranging from 75-87 percent.[6,7] When detected early, and when surgery is successful, optic disc cupping is reversible and the visual prognosis is good.[1] Causes of visual loss in congenital glaucoma include optic nerve damage, media opacities, and amblyopia.

The literature on congenital glaucoma in Nigeria is scanty. Baiyeroju *et al*,[7] in 2002, reviewed 89 trabeculectomy operations carried out on 56 patients under the age of 30 years; in 36 (40.4 percent) of the eyes, the operation was for congenital glaucoma. They observed that intraocular pressure control was better in the older than younger patient, possibly due to the effect of young age on healing; there was however, no report on the visual outcome and the factors affecting it. The aim of the present series is therefore, to highlight the features of congenital glaucoma as well as review the outcome of care with the aim of making recommendations for improvement.

Materials and Methods

A retrospective review of case notes of patients seen over a period of twelve years [1991 to 2002] was carried out at the Eye Clinic, University College Hospital, Ibadan. All children diagnosed with buphthalmos or congenital glaucoma were identified from out-patient and in-patient registers. Their case notes were retrieved and studied. Twenty-one patients were identified but six patients were excluded because of inadequate records. The case notes were reviewed according to a standard protocol for the following variables: age at presentation, sex, laterality, family history, pregnancy history, systemic anomalies, ocular features, management, duration of follow-up, and outcome of treatment.

Results

Fifteen children with features of buphthalmos were reviewed. These constituted 0.1 percent of 13,960 new patients, and 0.6 percent of new paediatric cases seen within the study period. There were 12 (80 percent) boys and three (20 percent) girls with ages ranging from 22 days to nine years (mean, 25.5 months); nine (60 percent) were aged 18 months or less at presentation (Fig. 1); only seven (46.7 percent) patients presented within the first year of life.

Both eyes were involved in 12 (80 percent) patients while it was unilateral in three (20 percent). Two patients who had a positive family history were siblings, while another two (13.3 percent) had cardiac anomalies detected on systemic examination and investigations; one had a ventricular septal defect (VSD) and the other had cardiac dysrrhythmia in form of premature atrial beats. There was no history of maternal illness during pregnancy in any of the patients.

Clinical features included epiphora, photophobia, blepharospasm, enlarged eyes, poor vision, corneal haziness, pale and cupped optic discs, and elevated intraocular pressures (Table 1). Only one patient presented with the classical triad of epiphora, photophobia, and blepharospasm, while four had both epiphora and photophobia. Fourteen patients (93.3 percent) required Examination under anaesthesia was carried out to assess the corneal diameter, intraocular pressures, and optic discs in 14 (93.3 percent) of the patients; one patient was old enough to be examined without anaesthesia. The corneal diameter was measured in 24 of the 27 eyes. It ranged from 12.5 mm to 17mm

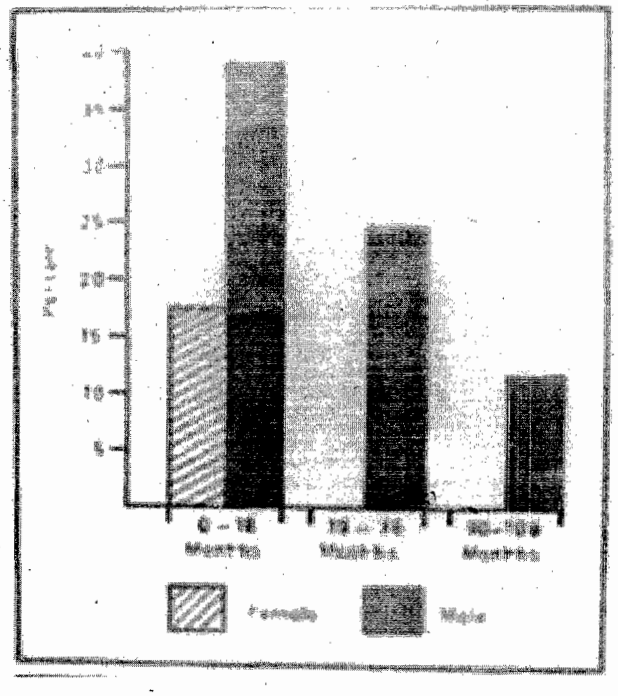

Fig. 1 Age and sex distribution of patients with buphthalmos

Table I

Frequency of Clinical Features in Patients with Buphthalmos

Feature	*Number of Eyes*	*Percent*
Buphthalmos	25	92.6
Corneal haziness	22	81.5
Raised IOP	20	74.1
Poor vision	16	59.3
Pale cupped disc	16	59.3
Photophobia	12	44.4
Epiphora	10	37.0
Blepharospasm	2	0.1

IOP = intraocular pressure

(mean ± SD, 14.6 mm ± 1.54 mm). Intraocular pressures were measured in 25 eyes using either the Schiötz or the Perkins tonometers. The values ranged from 6mmHg to 45.5mmHg. Six eyes (24 percent) had intraocular pressures of less than 21 mmHg; three of these eyes were on medication prior to examination. Eleven eyes (44 percent) had intraocular pressures of between 21 and 30mmHg, while eight eyes (32 percent) had intraocular pressures of greater than 30mmHg. The cup:disc ratio was evaluated in 16 eyes. One eye had a ratio of 0.3 or less, four had ratios between 0.4 and 0.6, another four had ratios between 0.7 and 0.9 while seven had ratios equal to 1.0. The cup:disc ratio could not be estimated in 11 eyes due to severe media opacities.

Trabeculectomy was performed on 21 (77.8 percent) eyes. Eight patients had bilateral trabeculectomies, either at one or two sittings, while five patients had unilateral surgery. Three eyes (in two patients) were not operated upon because of low intraocular pressures, one eye had been operated upon elsewhere and one patient (two eyes) was lost to follow-up before surgery could be done. The mean (±SD) pre-operative intraocular pressure in the eyes that were operated upon was 28mmHg (± 8.4mmHg). Of the eyes that had surgery, 14 (66.7 percent) had satisfactory intraocular pressure control (intraocular pressure less than 21mmHg as at last visit) although three (21.4 percent) of the eyes required medication (Timolol, Pilocarpine) to achieve the control. The mean postoperative intraocular pressure in these 14 eyes was 12.6mmHg (SD 4.5 mmHg). In five of the eyes, the intraocular pressures were poorly controlled with a mean postoperative pressure of 34.1 mmHg ± 7.8mmHg. Although repeat trabeculectomy was scheduled in respect of four (80 percent) of these, only one case underwent repeat surgery with unsatisfactory results, while the remaining three were lost to follow-up. Complications of surgery included vitreous loss (three eyes), endophthalmitis (two eyes) and bullous keratopathy (one eye).

The duration of follow-up from the time of initial presentation ranged from 2.5 months to 6.75 years (mean ± SD, 25.9 months ± 28.6 months). Six (40 percent) patients were followed up for more than one year, while nine (60 percent) were lost to follow-up. Out of 10 eyes in which the visual acuity was measured objectively post-operatively, two (20 percent) had a good visual outcome (visual acuity 6/18 or better), while the remaining eight eyes had visual acuities ranging from 6/36 to no perception of light. Causes of poor visual outcome in the eight eyes ranged from corneal opacity (two), cataract (one), advanced glaucoma (one), and endophthalmitis (one), to possible amblyopia (three).

Table II

Outcome of Management in Patients with Buphthalmos

Characteristic	*Number of Eyes*	*Percent*
IOP control		
d" 21mmHg	14	73.7
> 21mmHg	5	26.3
Cornea clarity		
Better	15	71.4
Unchanged	6	28.6
Visual acuity		
≥ 6/18	2	20.0
< 6/18	8	80.0

Visual acuity was not tested objectively in 17 eyes
IOP=intraocular pressure

The visual acuity was not assessed objectively in 17 eyes because the patients were too young to respond verbally, and in addition, there was a lack of appropriate facilities to assess vision in infants and young children. None of the patients had undergone amblyopia therapy. Reversal of corneal haziness was recorded in 15 (71.4 percent) of the eyes that had trabeculectomy. Six (28.6 percent) eyes had residual corneal opacities.

Discussion

The prevalence of buphthalmos in this study, was similar to that reported by Franks and Taylor for Western Europe.[1] It occurred predominantly in males with 80 percent of the patients in the present series being males, a percentage that is slightly higher than that quoted by Song *et al.*[5] The laterality observed is in keeping with findings in other parts of the world.[5]

Although about 80 percent of cases are reportedly diagnosed by one year of age,[5] in this study, only 46.7 percent had presented by their first birthday. This suggests that late presentation may be a problem in this environment. This could be due to a lack of awareness among the general population and the fact that the enlarged eyes especially when bilateral, may not be worrisome to the parents as they may be considered attractive. The delay in diagnosis that results from late presentation could have been responsible in part, for the poor visual outcome in these patients.

The successful control of intraocular pressure with trabeculectomy in this study compares relatively well with what obtained in other series.[4,6-9] This success rate strengthens the suggestion that trabeculectomy should be performed as the initial procedure for congenital glaucoma in black patients.

Follow-up in the present series was poor with only 40 percent of the patients attending for more than one year. This problem is not peculiar to buphthalmos but the effects of inadequate follow-up are particularly severe in these patients because intraocular pressure elevation can recur at any age and amblyopia therapy is often required. Poor follow-up may be due to lack of patient education and the illiteracy of the general population. Satisfactory visual outcome (visual acuity 6/18 or better) was observed in only 20 percent of those who had objective visual acuity assessment. This is significantly lower than that recorded in other series. Morin & Bryars,[4] as well as Morgan *et al*,[9] reported that 47 and 58 percent of their patients respectively, had visual acuities of 6/18 or better. The poor visual outcome in the present series may be due to the late presentation of patients (with irreversible cornea opacity and advanced glaucoma in three eyes), poor follow-up, and inadequate amblyopia therapy.

The main problems identified in the management of buphthalmos in Ibadan are late presentation of patients, poor follow-up, lack of appropriate facilities to determine visual acuity objectively in very young patients, and lack of adequate amblyopia therapy. Other problems include technical difficulties at surgery as a result of the scleral thinning that occurs in buphthalmos. Screening and public enlightenment may aid earlier diagnosis and thus result in better visual prognosis. Good patient education and counselling may also be beneficial in achieving better follow-up of these patients. Establishing well equipped paediatric ophthalmology centres to which patients can be referred, will aid in prompt management, while the pooling effect thus created, would aid better surgical skill acquisition and improved quality of care.

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