ORIGINAL ARTICLE

Congenital Eye Diseases at Olabisi Onabanjo University Teaching Hospital, Sagamu, Nigeria

O.T. Bodunde FWACS, H.A. Ajibode FWACS, FMCOpth
Department of Ophthalmology, Olabisi Onabanjo University Teaching Hospital, Sagamu, Nigeria

ABSTRACT

Background: *Congenital eye disorders, though rare are important causes of childhood blindness. It can occur in isolation or in combination, or as part of a syndrome. This retrospective study was aimed at documenting the causes of congenital eye diseases at Olabisi Onabanjo University Teaching Hospital (OOUTH), Sagamu, Nigeria.*

Method: *A review of the case notes of patients presenting at the eye clinic with congenital eye diseases between January 1994 and December 2004 was carried out.*

Result: *The most common congenital disorders are cataract 50(47.6%), congenital glaucoma 15(14.3%), Dacryostenosis 11(10.5%), and corneal opacity 6(5.7%) which are causes of preventable blindness. Less common congenital disorders are microcornea (1%), aniridia (1%), retinal atrophy (1%), and congenital anomaly of the optic disc (1%), which are congenital causes of irreversible childhood blindness.*

Conclusion: *We conclude that screening programmes should be instituted at the maternity centers before babies are discharged for early detection of congenital eye diseases and treatment of those that can cause preventable blindness. Also we recommend that Government should strengthen our welfare system by providing adequate measures for rehabilitation and care of those with irreversible blindness.*

KEYWORDS: Congenital; Eye diseases; Sagamu.

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INTRODUCTION

Congenital eye disorders, though rare are important causes of childhood blindness. It is estimated that the major structural ocular malformations such as anophthalmos, microphthalmos and coloboma account for 16.7% of childhood blindness globally. Kainbo[1] *et al* in Zaire reported a prevalence of 2.2% for congenital eye malformations while Bermajo[2] *et al* in Spain found a prevalence of 3.68/10,000 newborns. Also El-Gilary[3] *et al* in their own study in Egypt found that congenital causes accounted for almost half of the cases of the 113 blind people that were studied. Similarly Dawodu[4] in Nigeria found that 42.86% of blind children in Benin City had their problems from birth. Gilbert[5] has also reported that hereditary diseases are important causes of blindness in children in the industrialized countries.

Several factors are associated with the development of congenital abnormalities. These include: intrauterine infections, drugs, consanguinity, maternal metabolic disturbances e.g. folic acid deficiency, diabetes, cretinism and alcoholism[6].

Though much attention has not been given to congenital diseases as a cause of visual morbidity because they are rare and not of much impact, however, if considered singly, collectively the impact on vision is significant[7]. Some Nigerian authors have reported cases of congenital anomalies in Nigeria[8,9,10]. This study was conducted to determine the prevalence of congenital eye diseases at Olabisi Onabanjo University Teaching Hospital (OOUTH), Sagamu and its pattern of presentation.

MATERIALS AND METHODS

This 10-year retrospective, non-comparative study was carried out by retrieving the records of all congenital eye disease cases seen at the eye clinic of OOUTH, Sagamu between January 1994 December 2004.The diagnosis were made by Consultant ophthalmologists who see all new cases at their first visit/presentation mainly by clinical examinations and sometimes after an examination under anaesthesia and supportive laboratory investigations have been performed. The sex, age, and type of congenital eye disorders were recorded. Frequency tables of results were generated. Visual acuities were checked with snellen's chart in school children, pictorial chart in children aged 2-4 years and subjectively by finding out if the children could follow light in those below two years. Surgeries were offered and performed in those who required it and who gave informed consent to surgery. Follow- up was done at the eye clinic after treatment.

The functional definition of children is any one from birth and below 16years while congenital disease consist of

Correspondence to: **Dr. O.T Bodunde**
E-mail: bbodunde@yahoo.com

all forms of developmental defects present at birth.

RESULTS

A total of 14,389 new cases were seen during the study period Children constituted 27.31% (3,930) of all new cases. The cases of congenital eye disorders were 105, making 0.73% of all new patients and 2.67% of all new paediatric cases seen during the study period. There were sixty-two males (59.05%) and forty-three (40.95%) females. The male: female ratio was therefore 1.4:1. The age range at presentation was 4 hours 14 years with a mean of 4.18 years (+5.05 years). Seventy two (68.6%) presented on or before their 5th birthday Table I.

The most common congenital eye disease was cataract, seen in 50(47.6%) patients followed by congenital glaucoma 15(14.3%) and Dacryostenosis 11(10.5%) (Table II). Sixteen of the cases of cataract were bilateral, and the condition was more common in males (31) than females (19). Congenital corneal opacity was the fourth most common congenital disease and was seen in 6(5.7%) patients. Two of them were due to corneal dystrophies while the remaining four were due to unknown aetiology. The 0ther less common congenital diseases were anterior segment cleavage syndrome 2.9%, toxoplasmosis 3.8%, congenital ptosis 2.9%, congenital ectropion 1.9%, retinal atrophy 1.9%, squint 1.9%, microcornea 1%, congenital ankyloblepharon 1%, aniridia 1% and hereterochronic iridis 1%. The only case of unilateral aniridia also had bilateral cataract.

Only 4 cases of congenital cataract presented before one month (4 weeks) of age, with many more presenting above 1 year Table III.

All cases of Congenital ptosis were found in male patients. Two were unilateral affecting the left eyes while the remaining one was bilateral.

Table I. Age at Presentation of Patients

AGE	FREQUENCY(percentage)
<1yr - 1	53 (50.5)
>1yr - 5 yrs	19 (18.1)
>5 yrs	33 (31.4)

Table II. Congenital Eye Diseases seen in OOUTH Sagamu between January 1994 December 2003

DIAGNOSIS	FREQUENCY	PERCENTAGE (%)
CONG. CATARACT	50	47.6
CONG. GLAUCOMA	15	14.3
DACRYOSTENOSIS	11	10.5
CORNEA OPACITY	6	5.7
TOXOPLASMOSIS	4	3.8
ANTERIOR CLEAVAGE SYNDROME	3	2.9
CONG. PTOSIS	3	2.9
CONG. ECTROPION	2	1.9
RETINAL ATROPHY	2	1.9
SQUINT	2	1.9
MICROCORNEA	1	1.0
CONG. RUBELLA SYNDROME	1	1.0
CONG. ANOMALY OF OPTIC DISC	1	1.0
ANKYLOBLEPHARON	1	1.0
ANIRIDIA	1	1.0
HETEROCHROMIC IRIDIS	1	1.0
DERMOID CYST	1	1.0
TOTAL	**105**	**100.0**

Table III. Age of Presentation of Congenital Cataract

AGE	FREQUENCY	PERCENTAGE (%)
0- 1 MONTH	4	8
1 MONTH - 1 YR	21	42
>1YR - 6 YRS	11	22
> 6 YRS	14	28
TOTAL	**50**	**100**

DISCUSSION

Congenital eye diseases accounted for 0.73% of all new patients and 2.67% of all new paediatric out patient cases seen at the eye clinic in Olabisi Onabanjo University Teaching Hospital during the study period.

In this study congenital cataract, accounting for 47.6% of cases was the most common congenital disorder followed by glaucoma14.3%, dacryostenosis 10.5%, and congenital corneal opacity 5.7%. This is

similar to Kainbo's finding in Zaire [1] and Magulike's finding at Enugu, Nigeria[7]. Congenital cataract though much less common than age related cataracts, accounts for 1/10th of all causes of childhood blindness globally[11].

Any part of the eye and its adnexia may be affected by congenital malformation. Anomalies may occur in isolation, in combination, or as part of systemic malformation syndrome. Some of the identified causes include heredity, genetic factors, intrauterine diseases, maternal diabetic mellitus, galactoseamia, and toxic substances such as irradiation[12]. Shephard [13] in his report said that in 40-65% of congenital malformations the cause is unknown, 8-10% are due to specific teratogenic agent, while 15-25% are due to chromosomal abnormalities.

Treatment of congenital cataract is surgical and it is recommended that this should be done at about 4 weeks of life for optimal visual outcome and to reduce the risk of secondary glaucoma [14,15], but most of our cases presented later than this age .We however do extracapsular cataract surgery as soon as the children are fit for anaesthesia. Congenital glaucoma is the second most common congenital disorder but most of these cases did not return for follow-up when they were told about the need for surgery. These children will eventually grow up to become blind. The high cost of surgery and the fact that children are dependent are some of the major reasons for this default; we therefore suggest that surgical treatment for children be subsidized by Government and non-governmental organizations.

Congenital dacryostenosis (artresia of lacrimal duct) is due to delayed canalization near the valve of Hasner[16] and is characterized by epiphora and eye discharge. We usually recommend lacrimal sac massage and prophylactic topical antibiotics till two years of age followed by probing and syringing if massage fails. Up till now none of our patients have required surgical intervention (dacryocystorhinostomy). Our mode of treatment seems to conform to Clark's [17] who found that sequential nasolacrimal duct probing in congenital dacryostenosis has a high success rate and that age does not appear to have an impact on the success of probing.

Two of the cornea opacities were due to corneal dystrophy while majority of the cases were undetermined aetiology. According to Rezende R.A *et al* [18], the most common primary cause of congenital cornea opacities is Peter's anomaly, followed by sclerocornea, dermoid and metabolic diseases, 9.7% of their cornea opacities were also idiopathic in origin. However Babalola [19] in his own study in Nigeria, only 0.7% of the congenital cornea opacity was of undetermined aetiology. 66.7% of the cases of congenital ptosis were unilateral. This is similar to Baiyeroju's study in Ibadan[20] which also reported that most of their cases were congenital. However, ophthalmologist should be alert to the possibility of co-existing structural defects [21], since it may be associated with more than one ocular and/or systemic malformation.

The case of congenital ectropion responded well to temporary tarsorrhaphy like in Dawodus studies[9].
Most of the other less common congenital disorders have no definitive management e.g. microcornea, retinal atrophy; anomaly of the optic disc, aniridia and heterochromic irides. These children may eventually grow up being blind and need mainly rehabilitation from specialized welfare units.

In conclusion, congenital eye disorders are important causes of visual morbidity and mortality in children. We recommend screening programmes at the maternity centers before babies are discharged by pediatricians and ophthalmologists for early detection so that treatment can be started early to prevent blindness in cases such as congenital cataract, glaucoma and ptosis. Government should also subsidize the cost of surgery for children to reduce the rate of default from medical treatment. Steps should be taken to strengthen our welfare system through the provision of adequate equipment and manpower for the rehabilitation of those whose blindness is inevitable and irreversible at no cost to them. In addition the provision of necessary teaching materials in our schools for the blind will also help to improve their social and academic condition.

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