ORIGINAL ARTICLE

Congenital eye and adnexial anomalies in Kano, A five year review

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Abstract

Background: *Knowledge of prevalence and types of congenital eye and adnexial anomalies is important as some of these anomalies can lead to childhood blindness. The aim of the study is to determine the types of congenital eye and adnexial anomalies seen in the eye clinic of Aminu Kano Teaching Hospital Kano, Nigeria between the years 2001 to 2005. To determine the proportion of children below the age of 10 years who presented with congenital eye and adnexial anomalies, the proportion of eye surgeries due to such anomalies, and proportion of these anomalies amenable to (surgical) intervention.*

Methods: *The clinic and theater registers were used to obtain the total number of children less than 10 years of age seen and those that had surgery during the review period. A list of those with congenital eye and adnexial anomalies was compiled. The case folders of patients with such anomalies were retrieved and information on age, sex, type of anomaly, laterality of the condition, and types of surgical intervention offered, was obtained. The data was manually analyzed.*

Results: *There were 4163 children seen in the eye clinic and 268 had surgery during the review period. There were 109 eyes of 69 patients with congenital eye and adnexial anomalies. The prevalence of such anomalies amongst children (<10 years of age) is 1.7% and surgery for congenital eye and adnexial anomalies accounted for 25.7% of eye surgeries in the study popularion. The male to female ratio is 2.3:1. In 40 patients, the anomaly was bilateral and it was unilateral in 29 patients. The commonest congenital anomalies are buphthalmos in 38%, cataracts in 35%, and naso lachrymal duct obstruction in 14%. Other less frequent anomalies are anophthalmia/ micophthalmia, limbal dermoid cysts and aniridia. Eighty six percent of the patients had surgery to correct the anomaly.*

Conclusion: *Most of the congenital anomalies seen in our hospital can lead to childhood blindness and vigilance by eye care providers is advised to ensure prompt identification and intervention.*

Keywords*: Congenital, anomalies, buphthalmos, cataract, dacrostenosis*

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INTRODUCTION

Congenital malformations are morphological defects that are present at birth, although they may not become clinically apparent until later in life. Three percent of newborns have a major malformation having either cosmetic or functional significance.[1] The eye is a complex organ that develops through highly organized process during embryogenesis. Alterations in this process can lead to severe disorders that become clinically apparent at birth or, shortly after. Congenital eye defects occurs relatively frequently in the human population. Cataracts is one of the common such anomalies and it affects 23 million children under the age of 16 years and 1.4 million are blind.[2] Severe malformations are associated with infant mortality. Chromosomal aneiploides are often associated with ocular anomalies and the commonest type is trisomy 13 and trisomy 21. Trisomy 13 is associated with microphthalmia, coloboma, cataract and retinal rosette like structures.[3] Mutations that lead to clinically relevant phenotypes highlight important stages in eye development and such mutations may affect genes at the top of the regulatory hierarchy and hence initial stage of eye development. Genetic mutations such as PAX6 and SOX2 lead to microphthalmia, anophthalmia and aniridia.[4] Congenital malformations may affect any part of the eye and the ocular adnexia. Developmental malformations of the eye may occur in isolation or as part of larger systemic malformation syndrome.[5] The aim of the study is to evaluate the prevalence (amongst children less 10 years of age attending the eye clinic) and types of congenital eye and adnexial anomalies seen amongst children below the age of ten years in Aminu kano Teaching Hospital, Kano- Nigeria from 2001 to 2005 and determine the proportion of these anomalies that are amenable to some form of treatment intervention.

PATIENTS AND METHODS

The clinic outpatient register was used to obtain the total number of children below the age of ten years seen during the review period and the number of those found

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to have congenital eye and adnexial anomalies. The theater register was used to obtain the total number of children below the age of ten years who had eye surgery and the list of those who had surgery for congenital eye and adnexial anomalies between 2001 and 2005 was obtained. The case folders of all the patients with congenital eye and adnexial anomalies was retrieved and the following information extracted; age, sex, type of congenital eye or adnexiae anomaly, laterality of the condition and types of (surgical) intervention offered. The information obtained was manually analyzed.

RESULTS

During the review period 4163 children (<10 years of age) were seen in the eye clinic of our hospital and 268 children in this age group were operated. The prevalence of congenital eye and adnexial anomalies amongst children less than 10 years of age attending the eye clinic of our hospital is 1.7%. Surgery for congenital eye and adnexial anomalies accounted for 25.7% of all eye surgeries in children below the age of 10 years. There were 109 eyes of 69 patients seen with congenital eye and adnexial anomalies. Forty patients had bilateral manifestation and 29 were unilateral. The male to female ratio is 2.3:1. The common congenital anomalies seen are congenital glaucoma in 26 patients (38%), congenital cataracts in 24 patients (35%), and congenital naso-lachrymal duct obstruction in 9 patients (14%). Other less common anomalies are limbal dermoid cyst, microphthalmia/ anophthalmia and aniridia as shown in table I.

Sixty six percent of the patients presented before the age of 2 years (table II). Twenty six patients (35%) had trabeculectomy, 24 patients (35%) had extra capsular cataract extraction with primary posterior capsulotomy, and 9 patients (13%) had dacrocystorhinostomy (DCR). Overall, 86% of the patient's benefited from surgical intervention.

Table 1: Types of congenital eye and adnexial anomalies in 109 eyes of 69 patients

Congenital anomaly	Bilateral	Unilateral	Number (%)
Cataract	19	5	24 (35)
Buphthalmos	19	7	26 (38)
Naso lachrymal duct obstruction	-	9	9 (14)
Limbal Dermoid cyst	-	6	6 (8)
Microphthalmia	1	2	3 (4)
Aniridia	1	-	1 (1)
Total	**40**	**29**	**69 (100)**

Table II: Age at presentation and sex distribution of 69patients with congenital eye and adnexial anomalies

Age in months	Sex		Total (%)
	Male	Female	
<12	23	9	32 (46)
12-23	10	4	14 (20)
24-35	7	5	12 (18)
36-47	5	0	5 (7)
48-59	2	2	4 (6)
60+	1	1	2 (3)
Total	**48**	**21**	**69 (100)**

DISCUSSION

The incidence of congenital eye and adnexial anomalies in our environment is not known. This may not be unconnected with poor record keeping of medical illnesses in general, and the fact that many children afflicted by such conditions may not present in hospital. In the United Kingdom, the incidence of new diagnosis of congenital and infantile cataracts in the first year of life is 2.49 per 10000 children (95% confidence interval 2.10-2.87).[6] Congenital cataracts may present in isolation or in association with facial dysmorphism neuropathy syndrome. The later is of early onset and associated with microcornea, micro pupil and microphthalmos.[7] It is often difficult to trace the origin of isolated congenital cataracts in our environment. Congenital cataract can occur as an isolated autosomal dominant trait in 10% of cases.[8] Maternal rubella infection is a risk factor for congenital cataract that can be minimized by prompt maternal rubella immunization of mothers.[9] The possibility of rubella infection can not be ruled out as rubella specific IgM serology was not done on any of the patients with cataracts. Congenital rubella syndrome is rarely serologically diagnosed in our hospital though there is a reported case of serologically confirmed congenital rubella infection from Port Harcourt, Nigeria.[10] The incidence of primary congenital glaucoma is 1:10000 live births.[11] Congenital glaucoma is a distinct clinical entity following an autosomal dominant mode of inheritance.[12] In our study congenital glaucoma is marginally commoner than congenital cataracts. In Enugu, congenital cataract is most common in 42.6%, congenital glaucoma accounted for 22.2% and anophthalmia/ microphthalmia 9.3% each.[13] Similarly in Sagamu, the commonest eye anomaly is cataract in 50 patients (47.6%), congenital glaucoma in 15 patients (14.3%) and dacrostenosis in 11 patients (10.5%).[14] Congenital

anophthalmos/ microphthalmos is rare in our study, unlike in Edo State, Nigeria, where all the 2 cases seen with congenital eye anomaly had this condition.[15] Both sporadic and hereditary forms of lachrymal outflow dysgenesis may present in isolation or as a part of a systemic syndrome.[16] The study in Abuja showed that naso lachrymal duct obstruction was the commonest congenital anomaly accounting for 12.4%, cataract was seen in 5.1% and 2.9% had congenital glaucoma.[17] Few cases of congenital anophthalmos have been reported from various parts of Nigeria. Two cases of bilateral anophthalmos were reported in Lagos, one of the patients had associated polydactyly.[18] The 2 cases reported from Benin had unilateral anophthalmos.[19] A study involving the assessment of 2.5 million births in California (United States) indicated that the prevalence of anophthalmia was 0.18 per 10000 live birth and micophthalmos occurred in 0.22 per 10000 live birth.[20] Although a case of lid coloboma has been reported elsewhere,[21] none of the patients had congenital lid anomaly. Most of the patients in our study had congenital anomalies amenable to surgical treatment. There is need for vigilance to ensure early detection of potentially blinding congenital eye anomalies and prompt intervention.

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