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The Pattern of Congenital Malformations at the Ladoke Akintola University of Technology, Osogbo

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Summary

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Introduction: This retrospective study was designed to determine the pattern and outcome of congenital malformations among paediatric admissions at the Ladoke Akintola University of Technology Teaching Hospital, (LTH), Osogbo, a relatively new tertiary medical centre in southwestern Nigeria.

Materials and Methods: The case notes of children with congenital malformations admitted to the paediatric unit of the hospital between July 1, 2001 and June 30, 2006 were studied. The malformations were classified into organ systems, while recognized syndromes were classified under miscellaneous.

Results: Out of a total of 3,826 paediatric admissions, 53 (1.39 percent) had congenital malformations; 42 (79.2 percent) of them were referred patients and the remaining 11 (20.8 percent) who were inborn, constituted 0.34 percent of 3,264 deliveries in the hospital maternity unit. The commonest malformations recorded were omphalocoele (13.2 percent), Hirschsprung's disease (11.3 percent), cleft lip and palate (11.3 percent) and cyanotic congenital heart diseases (9.4 percent). Nineteen (35.8 percent) of the 53 patients were discharged for follow-up in the paediatric outpatient department of the hospital, 10 (18.9 percent) were referred to other tertiary centres for surgery because of lack of requisite surgical facilities within the hospital, while nine (17.0 percent) were discharged against medical advice (DAMA) at the request of parents. The remaining 15 (28.3 percent) died during the neonatal period.

Conclusion: The present study emphasizes the need for promptness and improvement in the management of simple correctable malformations such as those causing intestinal obstruction, omphalocoele, as well as cleft lip and palate in the tertiary institutions where children with such malformations are often cared for. It is believed that increases in autopsy rates will help in defining the pattern as well as determining the prevalence of congenital malformations in the community.

Key words: congenital anomalies

Introduction

CONGENITAL malformations occur worldwide[1,2] and are responsible for a significant proportion of childhood morbidity and mortality in the developed world.[3,4] They may be defined as morphologic defects of an organ or region of a body as a result of intrinsic abnormal developmental processes. The International Classification of Disease System 10 (ICD 10) classifies malformations based on the systems affected.[5] The situation in Nigeria is similar to that in many developing countries where infections and nutritional disorders constitute major determinants of childhood morbidity and mortality, while congenital malformations are relatively less important causes.[6] It is believed that with the control of nutritional disorders and vaccine preventable diseases of childhood, the relative contributions of congenital malformations to morbidity and mortality in children will increase in these developing nations.[7,8]

Data on congenital malformations in Nigeria have been derived mainly from studies in newborns

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delivered at various tertiary medical centres[9,10] as well as paediatric admissions into these hospitals.[11] These data, like in India[1] and Iran,[12,13] where the prevalence figures of 2.72 and 1.01 percent respectively, have been reported, have demonstrated slight variations in the relative frequencies of the different malformations. Anomalies of the musculoskeletal system were the highest followed by the central nervous and genitourinary systems in the Iranian study, while musculoskeletal anomalies were highest followed by gastrointestinal, central nervous and genitourinary systems in the Indian study. Although tertiary hospital data may be unrepresentative of the overall incidence of congenital malformations in the country,[14] they highlight the patterns of malformations that exist in the populations studied and can be useful for planning in the various institutions where these studies carried out.[15]

Most of the available studies on congenital malformations in Nigeria were conducted over two decades ago.[6,9,10] In view of this and the fact that the pattern and outcome of patients seen with congenital malformations at our health facility is unknown, the present five-year retrospective study was carried out at the Ladoke Akintola University of Technology Teaching Hospital (LTH), Osogbo, southwestern Nigeria. This relatively new tertiary medical institution also provides primary and secondary health care to the inhabitants of Osun State and neighboring communities of Ekiti, Kwara, Oyo and Ondo states.

Patients and Methods

The patients consisted of children with congenital malformations who were admitted to the paediatric unit of the Ladoke Akintola University of Technology Teaching Hospital, Osogbo, from July 1, 2001 to June 30, 2006. Their case notes were reviewed and the information extracted in respect of each patient included the type of malformation, age on admission, sex, and the outcome of admission (whether discharged, referred to other health institution or dead). The records in the maternity unit were checked for the total number of both live and stillbirths. During the period studied, it was the practice to admit all children with congenital malformations for stabilization, treatment, including surgery where applicable, and counseling of their parents. Neonates with thoracic, abdominal or cardiac malformations were further investigated using plain or contrast radiographs, ultrasound and echocardiography when indicated. The subjects were grouped into those delivered in the hospital maternity unit and those referred to the hospital.

Malformations identified in the studywere classified into organ systems according to ICD-10.[1,5,7] Where more than one organ system was involved, the malformations were recorded as multiple organ malformations and classified under miscellaneous. Recognized syndromes were also classified under miscellaneous. None of the patients had karyotyping since the facility for the procedure was not available; the diagnosis of Down syndrome in the study was therefore mainly clinical. The results were analyzed as simple percentages and chi square test was used to compare data, where applicable.

Results

During the five-year study period, 53 (1.39 percent) of 3,826 paediatric admissions into the LTH had congenital malformations. Twenty-nine (54.7 percent) of these children were males, while the remaining 24 (45.3 percent) were females; a male to female ratio of 1.2: 1. Table I shows the age distribution of the patients on admission. Fifty-one (96.2 percent) of the children, which included 42 (79.2 percent) neonates, presented within the first year of life. Forty-two (79.2 percent) of the patients were referred to the hospital, while the remaining 11 (20.8 percent) were among the hospital deliveries and they constituted 0.34 percent of a total of 3,264 births in the hospital maternity unit during the study period. There were 304 (9.31 percent) stillbirths among these deliveries and none had autopsy.

Table I

Age Distribution in Patients with Congenital Malformations

Age	*Frequency*	*% of Total*
1–28 days	42	78.2
1 month – 1year	9	17.0
> 1 year - 5 years	1	1.9
> 5 years	1	1.9
Total	53	100.0

Anatomic sites affected by congenital malformations

The various types of congenital malformations and the categories to which they belong are shown in Table II. The most affected organ systems were the gastrointestinal (37.7 percent), cardiovascular, and central nervous systems (13.2 percent each). As for individual malformations, omphalocoele, (13.2 percent), Hirschsprung's disease (11.3 percent), cleft

Table II

Categories of Congenital Malformations

Categories of Malformations	*Types of Malformation*	*No of Cases*	*% of Total*
Gastrointestinal	Omphalocoele	7	13.2
	Hirschsprung's disease	6	11.3
	Intestinal obstruction	4	7.5
	Anorectal anomalies	2	3.8
	Tracheo-oesophageal fistula	1	1.9
Central Nervous System	Encephalocoele	3	5.7
	Microcephaly	2	3.8
	Hydrocephalus	2	3.8
Cardiovascular	Cyanotic congenital heart disease	5	9.4
	Acyanotic congenital heart disease	2	3.8
Facial/Palatal Cleft	Cleft lip and palate	6	11.3
Genitourinary	Hypospadias	2	3.8
	Posterior urethral valve	1	1.9
	Ambiguous genitalia	1	1.9
Chromosomal	Down syndrome	3	5.7
Ocular	Bilateral micro-ophthalmia	1	1.9
Miscellaneous	Multiple organ malformations	4	7.5
	Pierre Robin syndrome	1	1.9
Total		53	100.0

lip and palate (11.3 percent) and cyanotic congenital heart diseases (9.4 percent) were the commonest.

Outcome of admission

Table III shows the final outcome of admission in the 53 patients studied. Nineteen (35.8 percent) of the 53 were discharged for follow-up, 10 (18.9 percent) were referred to other tertiary hospitals for surgical procedures which could not be handled at the hospital during the period of the study, while nine (17 percent) were discharged against medical advice

Table III

Outcome of Admission in Patients with Congenital Malformations

Outcome	*Frequency*	*% of Total*
Discharged	19	35.8
Referred	10	18.9
DAMA*	9	17.0
Dead	15	28.3
Total	53	100.0

**DAMA = Discharge Against Medical Advice*

(DAMA) at the parents' request. There were 562 (14.7 percent) deaths among the 3,826 paediatric admissions. Fifteen (28.3 percent) of the 53 children with congenital malformations died compared with 547 (14.5 percent) of 3,773 children that did not have congenital malformations ($\div^2$ = 6.9; df =1, p = 0.009). All the 15 that died were neonates. Nine of them had gastrointestinal malformations, three had congenital heart disease, two had multiple congenital malformations and one had an encephalocoele.

Discussion

The total of 53 cases of congenital malformations during the five years covered by this study can be considered small when compared with reports of similar studies from the older centres of Ilorin[11] and Ibadan.[14] This may be partly due to the fact that this centre is relatively new compared with the cited older centres. We cannot say by how much the results obtained in this study have been affected by the cultural attitude of 'hide and destroy' with respect to children with congenital malformations in the Nigerian society as earlier described by Adeyokunnu.[16] This latter phenomenon may also partly account for the fact that 19 percent of the cases were discharged against medical advice (DAMA) at the request of their parents.

The prevalence of congenital malformations among the hospital deliveries was 3.4 per 1000 deliveries. This was much lower than the reported prevalences of 6.4-30/1,000 deliveries from various centres in Nigeria,[14] and that of 9.3/1,000 live births and 16.85/1000 live births reported from Libya[17] and Singapore,[18] respectively. Autopsies on stillbirths have been shown to increase the rate of congenital malformations in the perinatal period,[19] and performing autopsies on the 304 stillbirths could have increased the prevalence of congenital malformations recorded in the present study. A higher prevalence of congenital malformations among the hospital deliveries could also have been possible if there had been routine post-delivery follow-ups. This is because of the possibility that malformations that were asymptomatic in the neonatal period might have manifested with increasing age.[14]

The major systems affected by malformations in the present study were gastrointestinal, cardiovascular and central nervous systems. This distribution was different from those reported in studies on congenital malformations from other centres in Nigeria namely, Ilorin,[11] Ibadan,[14] Lagos[20] and Kano.[21] These might have been due to differences in study designs, socio-environmental factors, autopsy rates and the paucity of investigative procedures such as karyotyping and radio-imaging in our series.

The referral of 18.9 percent of our cases to other centres, as well as an overall mortality rate of 28.3 percent among our patients may be attributed to lack of surgical personnel and facilities to manage many of them on presentation at the time of the study. Observations from centres in Europe and North America show that many critically ill newborns often benefit from mechanical ventilation, the use of intravenous hyper-alimentation as well as facilities for prompt investigations. The non-existence of some of these procedures in the care of the newborn in many hospitals in Nigeria, including LTH, may also have contributed very significantly to mortality in this study, as the deaths occurred only in neonates.

This present study with its findings are consistent with those of previous Nigerian studies[11,14,20,21] in demonstrating the relative contributions of congenital malformations to childhood morbidity and mortality in the country. As the treatment and rehabilitation of children with congenital malformations are costly and complete recovery is often not realizable in some cases,[22] the economic reality in present day Nigeria where infections and malnutrition constitute major challenges to childhood survival, indicates that congenital malformations are not likely to receive priority consideration in national health planning in the near future. It is therefore suggested that tertiary institutions, which are presently the last hope for survival of these unfortunate children with congenital malformations, should endeavour to improve their skills and facilities in the diagnosis and treatment of simple correctable lesions especially those causing intestinal obstructions, cleft lip and palate, omphalocoele repairs and some others, for an improved reduction in childhood morbidity and mortality. Increase in autopsy rates in these hospitals will also help in accurately defining the patterns of congenital malformations in the respective communities.

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