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CLEFT LIP AND PALATE: THE JOS EXPERIENCE
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ABSTRACT

Objective: **To determine the pattern of occurrence of cleft lip/palate and the factors that may have influenced treatment outcome.**
Design: **Descriptive Study.**
Setting: **Jos University Teaching Hospital, Jos, Nigeria.**
Subject: **This study included 107 consecutive patients with cleft lip/palate managed between January 1991 and June 1997.**
Main outcome measures: **The pattern of occurrence of cleft lip/palate, the peculiarities of the malformation in this environment as well as factors that influenced treatment outcome.**
Results: **The 107 patients were aged between one day and twenty-six years at presentation. There were three adults aged between 18 and 26 years with a mean of 22.3 years and 104 children with a mean age of 9.5 months. The male/female ratio was 1. 1:1, the anomaly was 2.4 times commoner on the left and the cleft lip alone was the most frequent mode of presentation (52%). The incidence was higher in the 3rd and 4th siblings. In 13% of these patients, there were other associated congenital anomalies such as Van der Woude's Syndrome, Down's Syndrome and congenital heart disease. Complications were noted in 16(14.9%) patients and these included dehiscence after repair in 8(7.5%) patients, oronasal fistulae in 2(1.9%), nasal speech in 4(3.7%) and hypertrophic scar in 2(1.9%). Eighty nine percent of these repairs were satisfactory to the parents of the affected children. All the adult patients were satisfied with their repair.**
Conclusion: **The low incidence of this anomaly in our environment may be due to under-reporting in the rural areas. Public enlightenment programs should help improve early presentation.**

INTRODUCTION

The reported incidence of cleft lip and palate varies between races. It is commonest among the Orientals (2-3/1000 live births), followed by the Caucasians (1-2/1000 lives births) and lastly by the Negroid and American Indians with about 1/2000 lives births(1-3). These anomalies, which can, therefore, be said not to be uncommon, occur in isolation or in association with other non-syndromic and syndromic congenital anomalies. Some of these syndromes include Down's syndrome (and other trisomies), Pierre Robins sequence (micrognathia, glossoptosis and cleft palate), Treacher Collins syndrome (mandibulofacial dystosis) and Apert's syndrome (acrocephalosyndactyly)(4).

Others like Van der Woude's syndrome(5) (hypodontia and lower lip pits) and the Kabuki make-up syndrome, a combination of Van der Woude syndrome, dermatoglyphic skin patterns, a typical facial appearance and mental retardation are less common(6). Most of these syndromes have been shown to be inherited as autosomal dominant genetic disorders located at chromosome 1q32.

There have been reports from the West-African sub region particularly Western Nigeria describing these anomalies and peculiar problems from the sub-region(3,7-13). Late presentation including adult patients, social issues, associated anomalies and poor follow-up visits were some of the problems discussed in these studies. However, there has been no report from the Middle Belt of Nigeria on the subject to describe the anomaly or state local problems and peculiarities. This study was therefore, aimed at determining the pattern, the factors that influence outcome and peculiarities of this anomaly in Jos, Nigeria.

MATERIALS AND METHODS

The hospital record of 107 consecutive patients managed at the Jos University Teaching Hospital (JUTH) between January 1991 and June 30, 1997 with cleft lip/palate deformities were reviewed. Their demographic data, clinical presentation, management options and outcome were entered on proforma and analysed using the Epi-Info version 6.

RESULTS

A total number of 107 patients aged one day to twenty six years were included in the study. There were three adults aged 18, 23, 26 years (means 22.3 years). The age range of the 104 children was one day to eight years (Table 1). Table 2 shows the distribution of the cleft anomaly by sex. The overall male to female ratio was 1.10:1 and the isolated cleft lip was the most frequent anomaly which occurred in 52% of the cases. The anomaly was 2.4 times more common on the left than on the right (Table 3). The incidence was significantly higher in the 3rd and 4th siblings and this constituted 45.6% (Table 4). This deformity appeared to have been familial in 15 (14%) patients. Another sibling was involved in six cases, a parent in five, a first-degree cousin in five and an aunt in one.

Table 1

Average age at presentation and at surgery (mean ± sd in months)

		Mean ± SD (months)
At presentation		9.53 ± 19.06
At surgery		14.50±21.16
Average duration before surgery		3.39±4.60
Average age of presentation of the patients with cleft palate	(N-47)	2.48±5.05
Average age at surgery	(N-16)	23.27±8.29
Mean duration before surgery	(N-16)	16.60±2.80

Table 2

Distribution by sex and type of cleft

	Male	Female	Total	Ratio (m: f)
Lip only	25	31	56(52%)	1:1.27
Lip + palate	28	19	47(44%)	1.47:1
Palate only	2	1	3	2:1
Median cleft	1	0	1	1:0
Total	56	51	107	1.10:1

Table 3

Distribution according to sibling positions

		No.	%
First	–	13	12.1
Second	–	15	13.9
Third	–	26	24.3
Fourth	–	23	21.3
Fifth	–	15	13.9
Sixth	–	9	8.4
Seventh	–	4	3.7
11th	–	2	1.9
Total	–	107	100

Table 4

Distribution of associated congenital anomalies

Anomaly	Type of cleft	No. of patients
Van der Woude's syndrome	Lip/Palate	1
Down's syndrome	Lip/Palate	2
Tongue tie	Lip/Palate + Palate	2
Ventricular septal defect(VSD)	Lip/Palate	3
Right congenital hydrocele	Lip/Palate	1
Undescended Testis	Lip/Palate	2
Sinus arrhythmias + right ventricular hyertrophy (RVH)	Lip	1
Patent ductus arteriosus (PDA) + Arrhythmias	Lip/Palate	2
Total		14

There were 14(13%) associated congenital anomalies recorded in this study and 90% of these occurred in patients with a combination of cleft lip and palate. Six anomalies constituting 42.8% of all the anomalies and 5.6% of the cases were related to the heart (Table 4). Complications were noted in 16 (14.9%) patients including dehiscence following repair in eight (7.5%) patients, oranasal fistulate in two (1.9%), nasal speech in four (3.7%) and hypertrophic scar in two (1.9%). In 12 of these (11.2%), the results were assessed to have been unsatisfactory by the surgeon and the parents patients. Three patients each had dehiscence following the lip repair, palatal fistulae and nasal speech. Two patients had oronasal and one had a hypertrophic scar.

DISCUSSION

The Jos University Teaching Hospital (JUTH) is located in Jos, the capital city of Plateau State in the central part of Nigeria. It caters for the "Middle Belt" states of Plateau, Nasarawa, Benue, Bauchi, Gombe, Southern Kaduna State, The Federal Capital Territory, Abuja and Taraba State. Over a six and half-year period an average of about 16 cleft deformities were seen per year. This is considered to be a low incidence for a major referral centre like ours as this area has a population of over fifteen million people. This concurs with the low incidence quoted for our environment(3).

It is unfortunate that we still have adult patients presenting three decades after the initial study from Nigeria(3). This delay we suspect is as a result of ignorance of parents and traditional birth attendants who conduct many deliveries in rural areas (60% in this study) on treatment options. Patients with a combined cleft lip/palate presented earlier (meal age 2.48 ± 5.05 months) as compared to those with cleft lip alone (mean age 9.48 ± 19.06 months). This may

be due to associated feeding problems with the palate defect, which caused the parents to bring these children to the hospital earlier. This finding is similar to that of Osuji et *al.* (4) who reported that 60% of similar patients presented within two months of birth. They had observed that the children in their study were presented early for rehabilitation with feeding plates and pre-surgical orthodontics rather than for the repair of their clefts. In their study, 76% of the patients had combined cleft lip/palate as opposed to 43% in this study. The late presentation affected the average age at surgery, for the cleft lip, this was 14.50 ± 21.16 months. The mean duration before surgery of 3.39 ± 4.6 months confirmed, however, that patients were not unduly delayed before their surgeries.

A notable major finding not previously alluded to from the region was the large number of patients who did not return for palatal surgery following the lip repair. Indeed only about one third came for palatoplasty. A possible reason for this is a feeling on the part of the parents that having repaired the cleft lip, the cosmetic defect was no longer manifest and other problems were considered less critical. This is because the older children feed on solid food thereby reducing the feeding difficulties. Careful education/counselling of the patients at the clinic of presentation should help reduce this serious problem.

The male to female ratio of 1. 1: 1 and the higher frequency of the defect on the left side of the lip agrees with previous reports from Nigeria (7-10). There were only three patients with isolated cleft palate deformities in the study and one median cleft. This contrasts with the report from Sweden, which shows isolated cleft palate deformities to be as high as 39% of all clefts(14).

In this study, however, a positive family history and the incidence of associated anomalies was lower than previous reports from Nigeria and other parts of the world. There was a positive family history in 15 patients (14%). Eleven of these were first degree relatives. There was no particular disposition to a specific abnormality. Hagberg, *et. al.*(12) found a positive family history in 26.2% of the cases in their study. The inability of the more severe forms of cleft anomalies to survive in our poor rural environment may account for this low incidence of familial cleft deformities in addition to the paucity of documentation(9). Although 14 mothers gave a history of a febrile illness during pregnancy among other complaints, any cause-effect relationship is difficult to ascertain at this stage. The incidence of anomalies associated with cleft lip/palate varies with different authors and from different regions and countries. The incidence ranges from 4.3% to as high as 63.4% (14-19). Ademiluyi *et. al.*(11), Milerad *et al.*(14) and Lilius(17) found associated anomalies in 28.3%, 28% and 21.8% of cleft lip/palate respectively. In the study of Ademiluyi *et. al.*(11), cleft lip alone had associated anormalies in 19% of the patients; while cleft lip/palate in 35% and cleft palate alone 28%. Milerad *et. al.*(14) documented associated anomalies in only 8% of the patients with isolated cleft lips. We found associated anomalies in 13% of clefts and 90% of these were in patients with combined cleft lip/palate. The low incidence of anomalies in association with cleft lip alone agrees with previous studies especially that of Milerad *et. al.*(14). The generally low incidence of associated anomalies as compared to most studies may be due to under-reporting as our study shows that only 40% of deliveries were supervised by trained personnel. As such some of the more severe anomalies that may not survive the early neonatal period will not reach a medical facility as ours for documentation. This may be the reason why we did not record any cranial and spinal abnormalities though these anomalies were common in other studies(11,19,20). We also observed that the frequency of the defect peaks among and fourth siblings, the import of which remains to be determined.

The commonest complications were dehiscence of the lip after repair and palatal fistulae. Otherwise 90% of patients were satisfied after operation. In conclusion, we recommend the introduction of a national register for cleft lip/palate to document and co-ordinate the work of surgeons treating these patients in developing countries as recommended by Hammond and Stassen(21). Improved coverage of antenatal care and supervised deliveries would be key to this and should reduce problems of late presentation.

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