PATTERN OF CLEFT LIP AND PALATE DEFORMITIES IN RIVERS STATE OF NIGERIA

Pages with reference to book, From 64 To 66

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Abstract

The pattern of congenital cleft lip and/or palate deformities in the Rivers State of Nigeria is being presented. Thirty nine patients seen over four years were retrospectively studied. Similar to the other published African series but unlike the Caucasian series, isolated cleft of the lip was the predominant lesion seen in this study (54%). In contrast, however, to the same African Series the isolated cleft of the palate was only seen in girls (15%). The unusual cleft deformities are highlighted while the significance of associated congenital malformations is examined (JPMA 40 : 64, 1990).

INTRODUCTION

Congenital cleft lip and/or palate deformities are uncommon among black Africans[1,2] . Iregbulem[3] examined 21,624 new born infants in Enugu, Nigeria and found only 8 cleft deformities, giving an incidence of 1:2,703 live births. This contrasts markedly with the incidence rate of 1:373 in the Japanese[4]or the Caucasian[5] of 1:500. This study is hospital based. Although, the University of Port Harcourt Teaching Hospital (UPTH) is the only reference centre for these malformations in the Rivers State of Nigeria, it is believed that certain children born with congenital cleft deformities may not have been seen in this study. The study, however, presents for the first time the pattern of cleft lip and palate deformities in this part of the world. This is compared with some other published African and Caucasian patterns.

PATIENTS AND METHOD

Thirty nine patients with congenital cleft lip and/or palate were seen and managed at the University of Port Harcourt Teaching Hospital (UPTH) between January 1984 and December 1987. The case files of these patients were analysed for sex, age at presentation, type of cleft deformity including side affected, presence of other congenital malformations and presence or absence of family history of cleft deformities.

RESULTS

A total of thirty nine patients were studied, at an average of ten patients each year. Nineteen of these were males and twenty females giving a dale: female ratio of 1:1 (Table 1).

TABLE I. Types of Cleft Deformities and their Male/Female Distribution.

Type of Cleft	M	F	Total No.	%	Comments
Cleft of left upper lip	10	2	12	30.8	4 had associated cleft palate
Cleft of right upper lip	2	7	9	23.1	
Bilateral cleft of the upper lip	3	4	7	17.9	3 had associated cleft palate
Cleft palate alone	0	6	6	15.4	All females
Median cleft of the upper lip	2	0	2	5.1	Holoprosencephaly, all with cleft palate
Cleft of the nose	1	1	2	5.1	
Lateral cleft of the lip	1	0	1	2.6	
Total	19	20	39	100.0	

M = Male
F = Female

There were more left sided cleft lips than right side. The cleft of the left lip occurred in ten boys out of twelve children, while the right lip deformity occurred in seven girls out of nine. The median and lateral clefts of the lip were found only in boys, the cleft of the palate alone was only seen in females. The cleft of the nose and bilateral cleft deformities were evenly distributed in both sexes.

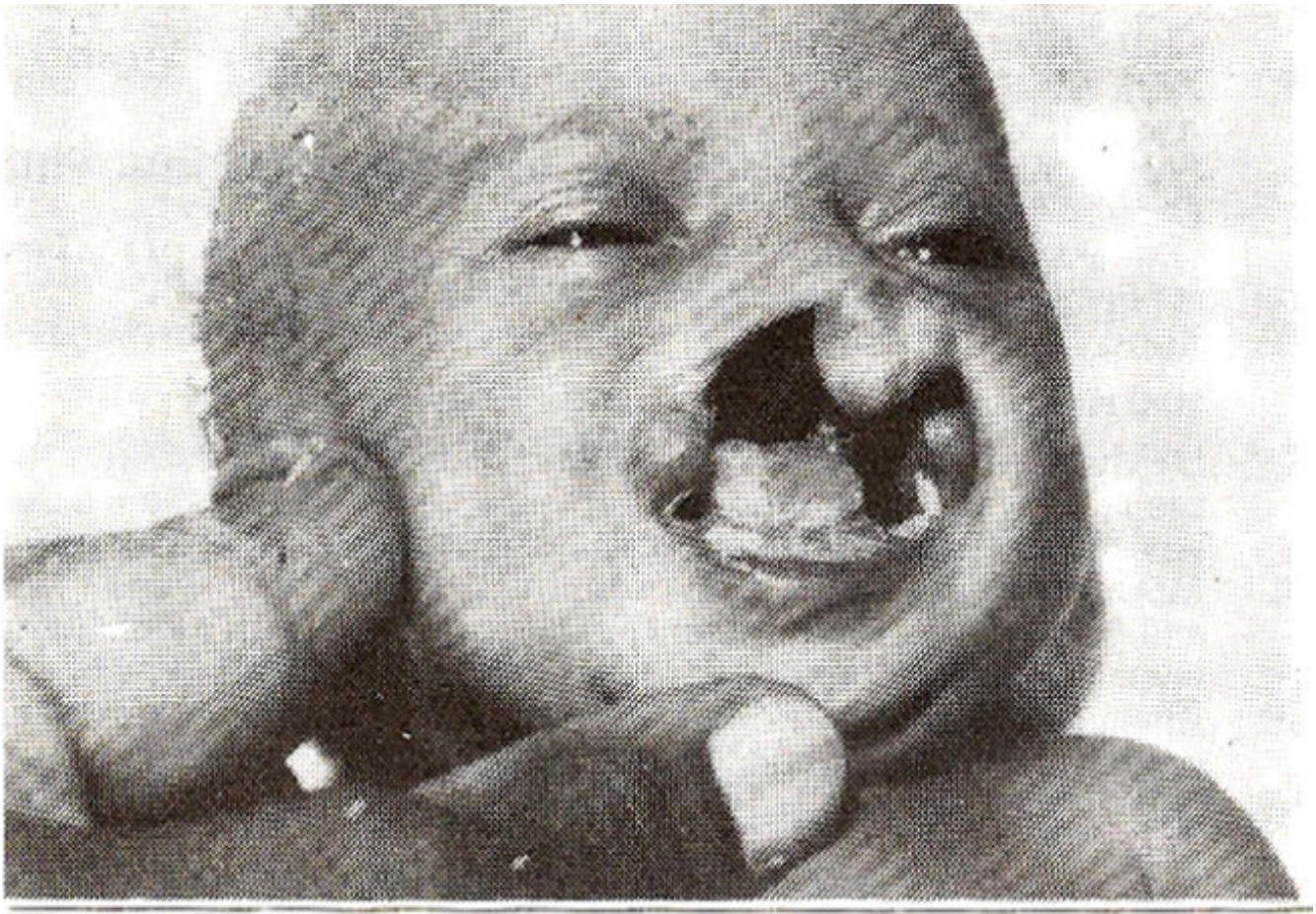

Figure 1. Bilateral complete cleft of the upper lip and cleft of palate in a one month old Nigerian girl.

Figure 1 illustrates bilateral complete cleft of the upper lip and clift of the palate.

TALE II. Associated Congenital Malformations.

Malformations	Frequency
Umbilical hernias	6
Accessory auricles	3
Hypotelorism	2
Microcephaly	2
Bulging eyes	2
Monoventricular brain	2
Facial palsy	1
Rudimentary pinna	1
Second nasal septum	1
Congenital inguinal hernia	1
Ventricular septal defect	1
Low set imperfectly fused pinna	1

Table II lists the frequency of the associated malformations. Two children with median clefts of the lip had mono-ventricular brains resulting in the holoprosencephaly syndrome which includes microcephaly, hypotelorism and mongoloid slant of the eyes (Figure 2).

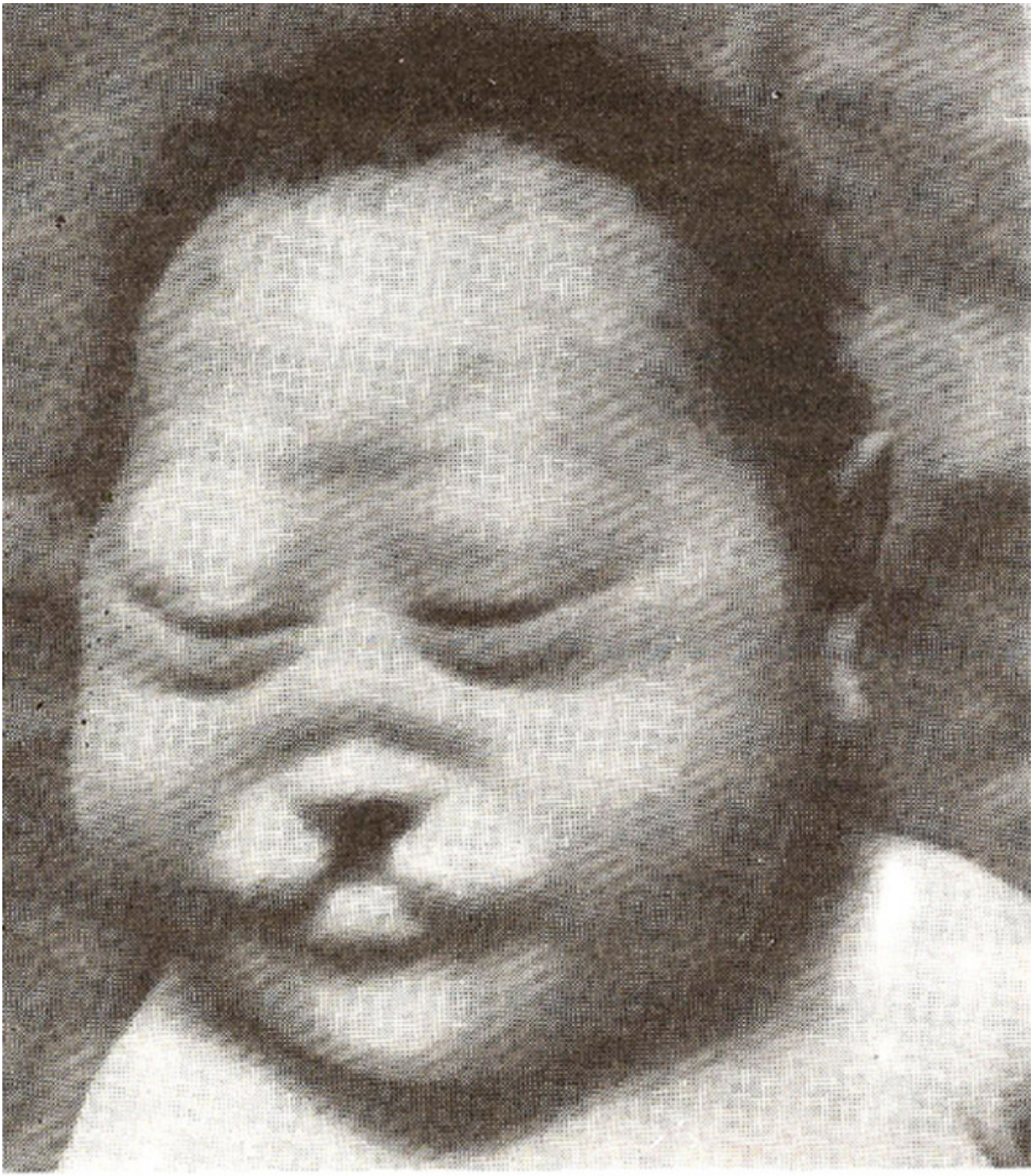

Figure 2. Median cleft associated with Holoprosencephaly in a one week old Nigerian boy.

Another child with bilateral incomplete cleft of the upper lip had imperfectly fused and low set right pinna with associated right facial palsy (Figure 3)

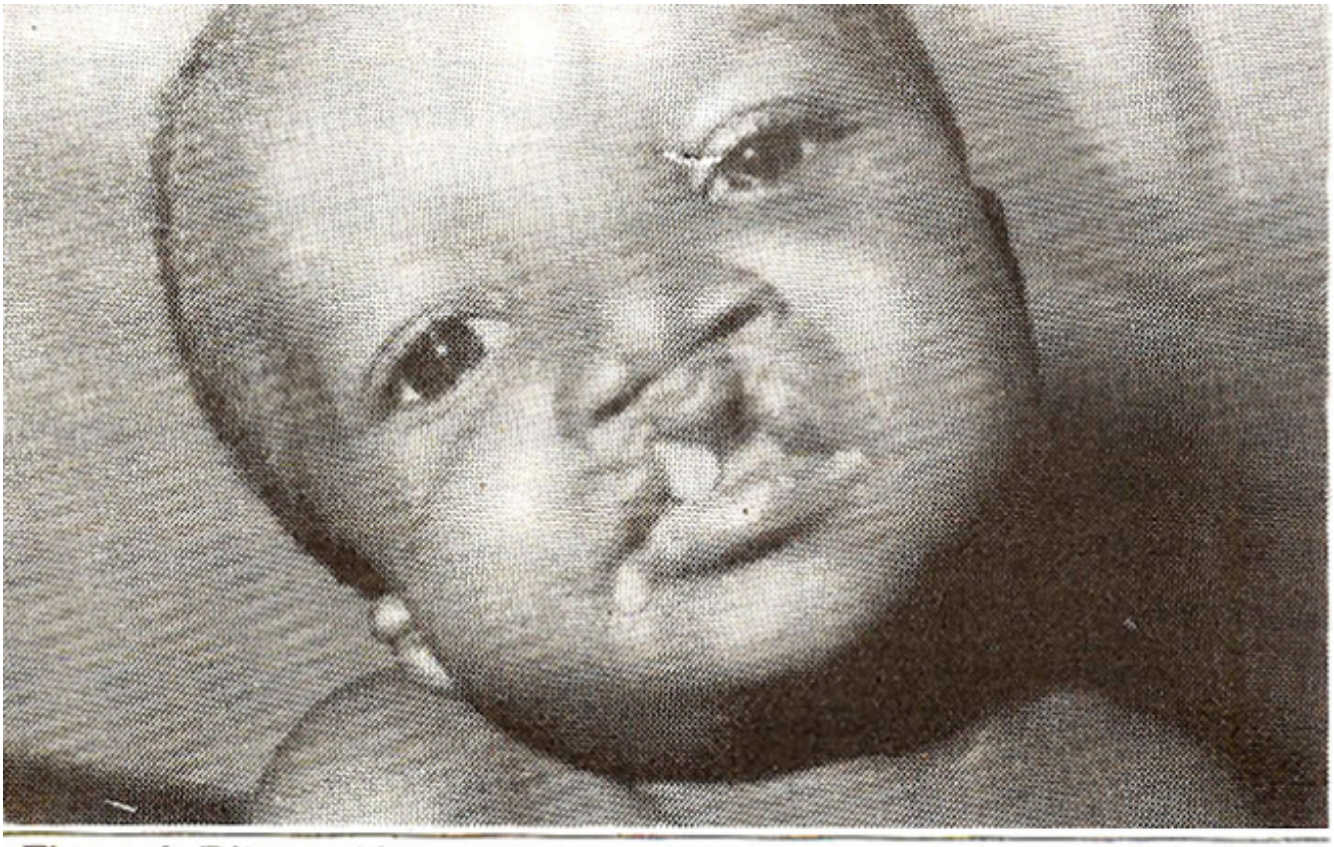

Figure 3. Bilateral incomplete cleft of the upper lip, imperfectly fused and low set right pinna, right facial palsy (apparent when child cries or smiles).

and umbilical hernia. Twenty six of these children presented before the age of 1 year, while only one presented at the age of 18 years. The remaining twelve were between 1-10 years of age at presentation. All the parents denied the presence of any cleft deformities in the family.

DISCUSSION

Congenital cleft deformity of the face is an in heritable disease[3]. The complete absence of a positive family history in this study must be interpreted with caution. The social taboos attached to such disfiguring congenital deformity as cleft lip is very strong in a typical African society. Affected families may, therefore, deny the existence of such deformities in any of its members, past or present. In the more advanced western societies, congenital cleft deformities conventionally present at birth reflect the level of health awareness and the availability of both human and material resources to correct the defects. In this study, the presence of cleft lip deformities in children of school age and even late teenage points to the low level of health awareness and inadequate material and human recources typical of the developing countries. The first author is the only Plastic Surgeon in the Rivers State with a population of over three million people. Many other neighbouring States of Nigeria have none at all. Some of the patients have been denied basic education because both school teachers and pupils were scared by their presence at school. The depression resulting from lack of social interaction in the older children is of course easy to understand. The pattern of congenital cleft deformities in this report is similar to some other published African series[2,6] in certain respects. The male: female ratio in isolated cleft palate was 1:1 in another series[3]. In this report, all isolated cleft palates were found in females. In

another study[7] a male: female ratio of 0.7:1 in isolated cleft palate was found in Northumberland and Durham pointing to a higher frequency of isolated cleft palate in girls. The literality of clefts in the two sexes is also interesting. While the cleft of the upper lip is predominantly seen in boys, right sided cleft lip occurs more in girls in this study. The significance of this is not clear. The more unusual forms of cleft deformities are a source of curiosity as they are not easily explained by the failure of fusion of the various processes that make up the face. The lateral cleft of the lip and clefts of the nose fall into this category. The patient with median cleft of the upper lip (Figure 2) have the characteristic facies that is pathognomonic of Holoprosencephaly[8]. In these children, the prechordal mesoderm in the embryo which gives rise to the median facial bones may be defective because of mechanical, genetic, or environmental teratogenic factors. Consequently, there is a failure of induction of the rostral neural ectoderm by the defective prechordal mesoderm resulting in arrested prosencephalic cleavage and therefore, monoventricular brain. This deformity which is often associated with chromosomal abnormalities[9] may be familial[10]. Easily identifiable external congenital malformations were routinely looked for in addition to listening to the heart sounds of the patients. Umbilical hernias were the most common associated exfrrnal malformation observed. We do not think that this has any specific sig nificance in relation to. cleft deformities as umbilical hernias are fairly common in the population under consideration. The childin figure 3 with bilateral incomplete cleft of the upper lip, imperfectly fused and low set pinna with associated facial palsy is a case of hemifacial microsomia, also known as the first and second branchial arch syndrome. We believe that the umbilical hernia is simply an unrelated incidental finding in the same patient.

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