

Craniofacial Clefts in a Black African Population

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Seven black children with craniofacial anomalies are reported. Holoprosencephaly and hemifacial microsomia featured prominently among the cases presented. These cases represented a large percentage of all cleft cases reported in the region served by the Port Harcourt Teaching Hospital (UPTH). The social, genetic, and surgical implications are discussed.

KEY WORDS: *cleft lip, cleft palate, holoprosencephaly, hemifacial microsomia, craniofacial anomalies*

Congenital cleft lip or cleft lip and palate is uncommon in black Africans (Iregbulem, 1982; Leck, 1983; Harrison et al, 1985). Other craniofacial anomalies are much more unusual (Oluwasanmi and Kogbe, 1975). Craniofacial anomalies in the African population are reported infrequently. The implications of such anomalies in the African population are different than in developed countries such as the United States and should therefore be reported. In this paper, seven black children with facial clefting seen at the University of Port Harcourt Teaching Hospital (UPTH) over a period of 3 years, 1985 to 1987, are presented and discussed.

CASE REPORTS

Case 1

The boy shown in Figure 1 was born following a full-term gestation to a young Nigerian couple. The mother had a low grade fever in early pregnancy but otherwise uneventful antenatal care at the Okirika General Hospital. The family resided near a petroleum refinery.

Physical examination showed a child with microcephaly, an occipitofrontal circumference of 26.5 cm at the age of 5 days, a finger tip patent anterior fontanelle, and a nonpatent posterior fontanelle. Orbital hypotelorism, proptosis, and an upward obliquity of the eyes were present. There was a true median cleft of the upper lip and a very wide cleft of the palate. There was no premaxilla, no nasal septum, and no columella. The nose was severely dysmorphic and nearly absent. The upper pole of each pinna was sharply angulated. The facial findings were consistent with the diagnosis of premaxillary agenesis type holoprosencephaly.

Case 2

This boy (Fig. 2) was the first child of young Nigerian parents. The mother had no antenatal care and was deliv-

ered by a traditional birth attendant in Port Harcourt. She had a mild fever in pregnancy for which she was treated. She remembered neither how early in pregnancy this occurred nor what medication she was given.

Seen at the age of 2 days, the baby had microcephaly with an occipitofrontal circumference of 26 cm. The anterior fontanelle was very small, and the posterior one was



FIGURE 1 Case 1, a 5-day-old child with premaxillary agenesis type holoprosencephaly. Note the true median cleft lip.

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FIGURE 2 Case 2, another neonate with premaxillary agenesis type holoprosencephaly. Note the flexed hypertonic posture of the limbs, which is often seen in this type of case.

closed. There was orbital hypotelorism. The eyes were proptotic, and the palpebral fissures had an upward obliquity. The nose was flat and underdeveloped, thereby appearing as a skin-covered cartilaginous mass traversed by a pin-hole nostril. There was a true median cleft of the upper lip. Except for a slight notching of the maxillary alveolus in the midline, there was no palatal cleft. The findings were consistent with premaxillary agenesis type holoprosencephaly.

Case 3

First seen at the age of 1 month, the girl shown in Figure 3 was born in Ahoada after a full-term pregnancy. The child had a right-sided incomplete cleft of the lip, a duplication of the nasal septum to the right, and a correspondingly wide and grooved alar dome. No other malformations were noted.

Case 4

First seen at the age of 4 months, this boy (Fig. 4) was delivered following a full-term gestation at the Okirika General Hospital after an uneventful pregnancy. The mother was 35 years old, but the father was between 65 and 70 years of age.

Malformations include a right-sided cleft of the oral commissure which extends toward a right-sided grade III microtia. The left ear was structurally normal, but there was an ear tag anterior to the tragus.

Case 5

This male child presented with an incomplete cleft lip, a left-sided incomplete cleft of the alveolus, a right-sided grade II microtia, a right-sided facial paresis, and an um-

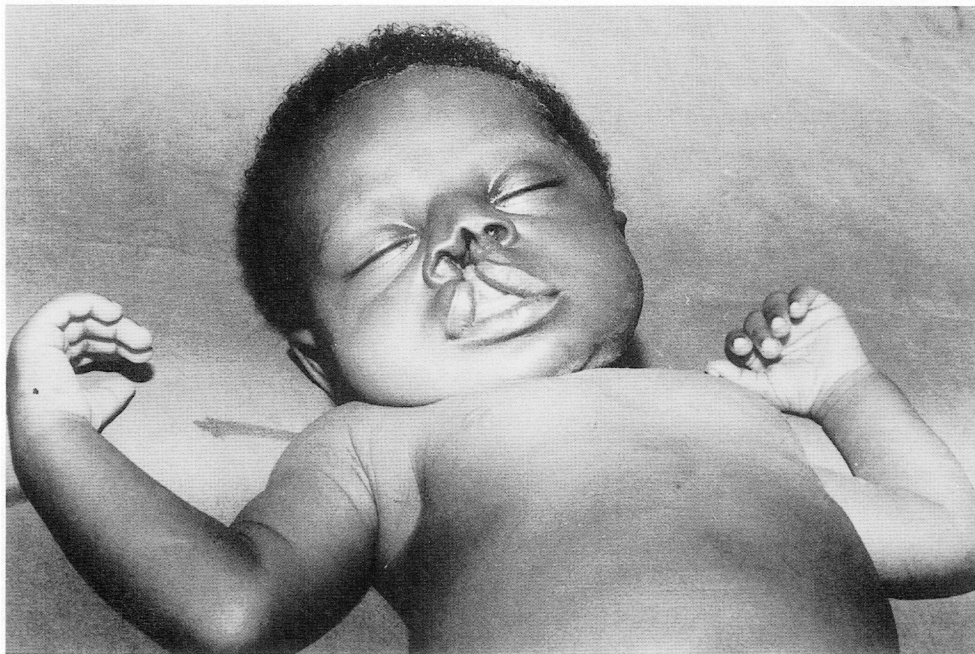


FIGURE 3 Case 3, an infant with duplication of the nasal septum associated with a right-sided cleft of the lip.

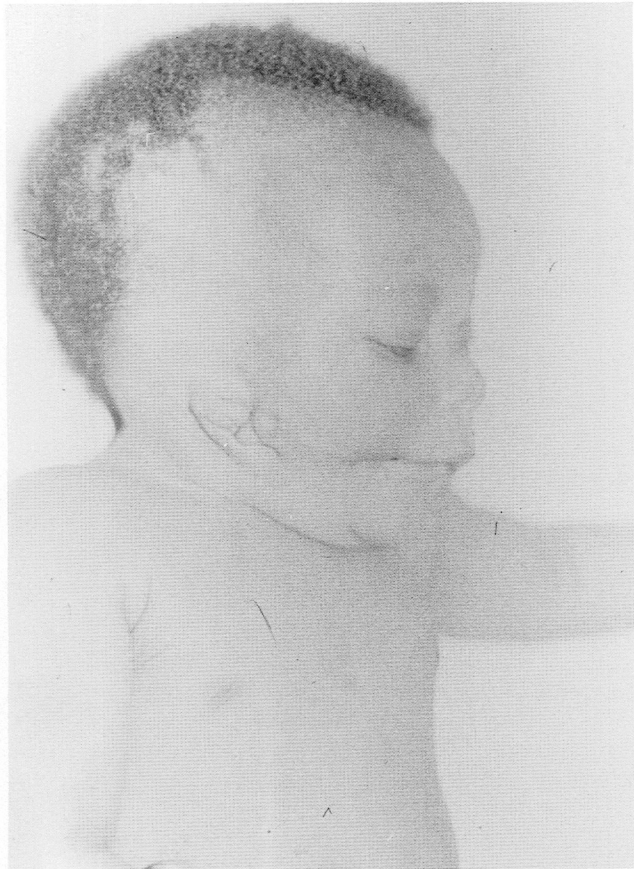
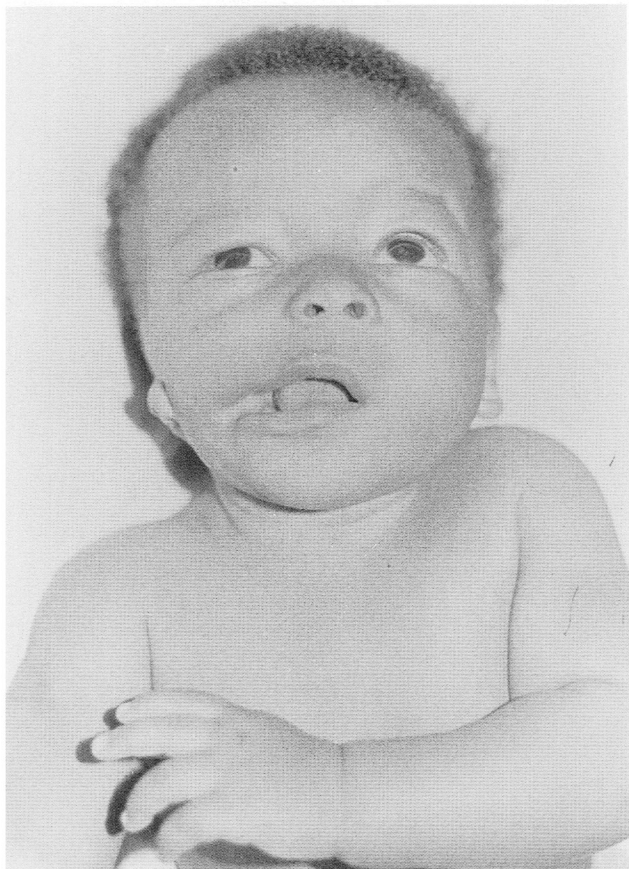


FIGURE 4 Case 4, a neonate with right-sided hemifacial microsomia, showing a right commissural cleft (left) and a right-sided grade III microtia (right).

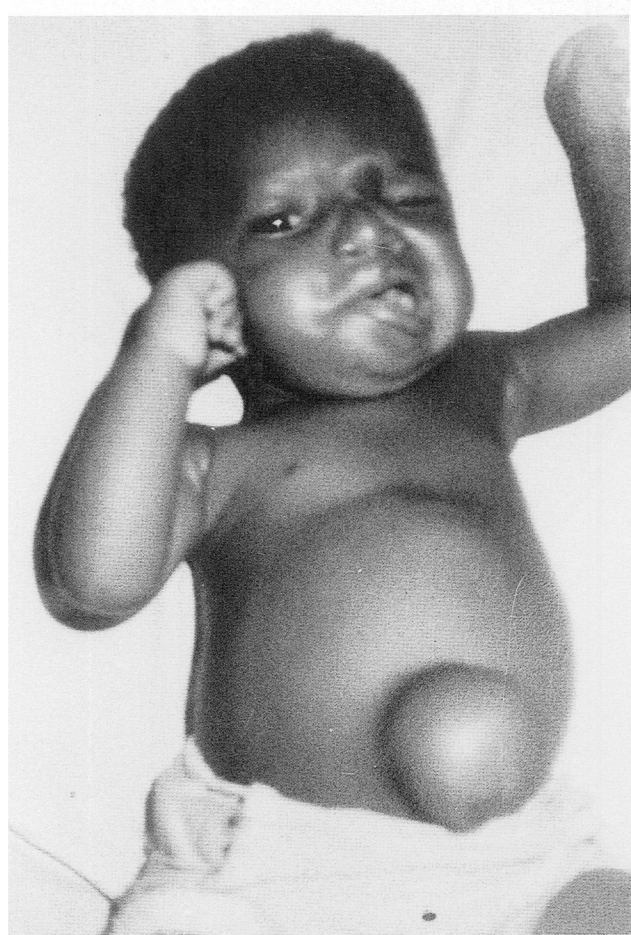
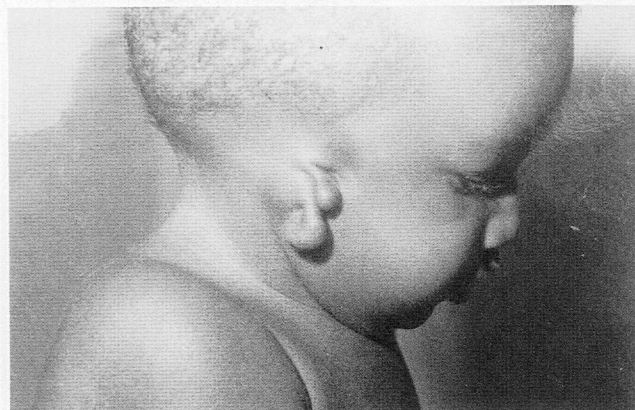
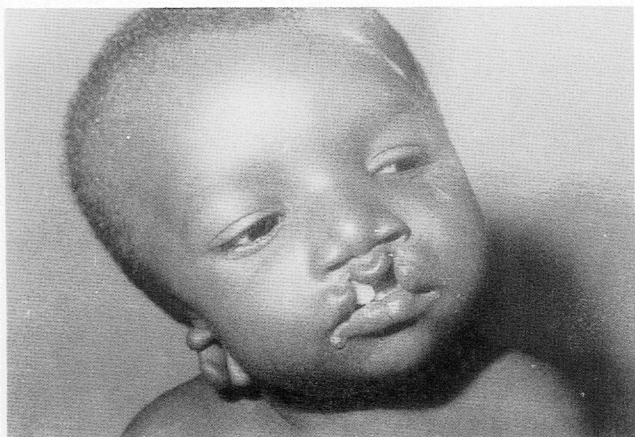


FIGURE 5 Case 5, an infant with incomplete bilateral cleft lip (top left), right-sided grade II microtia (bottom left), and right-sided facial paresis (right). Note the large umbilical hernia (right).

bilical hernia (Fig. 5). The palate was intact. He was born to very young parents in Okirika following a normal full-term gestation.

Case 6

This male child was seen at the age of 2 years with mild clefting of the right alar rim and a congenital inguinal hernia. No other malformations were found. No photographs are available.

Case 7

Clinical data for the child shown in Figure 6 were not available, but the dysmorphic facial features are reported. They include a right-sided cleft of the oral commissure, a cleft of the right alar dome, and orbital hypertelorism. There was a grade I microtia of the right ear.

DISCUSSION

The cases reported here represent seven patients out of a total of 31 with facial clefts who had been seen at the University of Port Harcourt Teaching Hospital during the 3-year period from 1985 to 1987. Unlike the remaining 24 who had more typical clefts of the lip, lip and palate, or palate alone, these children had unusual craniofacial anomalies. The relatively large number of supposedly rarer facial clefts among the population of this district is of interest. Is

this unusual proportion attributable to the lower prevalence of "typical" clefting among the black population with no similar reduction in the prevalence of rarer clefts? Or is there a higher prevalence of atypical clefts?

The parents of these children were under severe social pressure following their birth. Very often they refused to have their babies discharged until surgical correction had been undertaken. This is because there is a social taboo attached to the birth of such grossly malformed babies in the typical African setting. At home, the birth of any baby is difficult to hide. Neighbors are expected to visit for celebration without invitation. Parents therefore put a great deal of pressure on the surgical team to modify normal treatment plans. This may result in long hospital stays for some patients and psychotherapy for the parents. Surgical intervention is not given when the child has no developmental potential, as in the first two cases with holoprosencephaly.

The first two patients presented had a characteristic pattern of median craniofacial anomaly. DeMyer et al (1964) reported that the premaxillary agenesis facial type is pathognomonic for holoprosencephaly. They postulated that, in this condition, the prechordal mesoderm, which gives rise to the median facial bones, may be defective. Consequently, there is failure of induction of the rostral ectoderm by the defective prechordal mesoderm, thus resulting in arrested cleavage and subsequently in a monoventricular brain. This condition is etiologically heterogeneous and may be familial (Khan et al, 1970). Chromosomal abnormalities (Salmon, 1978), such as trisomy 13 and 18 (McDermott et al, 1968; Salmon, 1978) may cause holoprosencephaly. The need for chromosomal analysis in these children was recognized, but the facilities were not available for such studies.

The findings of microtia, macrostomia (commissural clefting), and facial asymmetry confirm the diagnosis of hemifacial microsomia in cases 4 and 5. The clefting of the lip and the facial palsy are recognized components of this etiologically heterogeneous congenital defect.

The notching of the alar dome in cases 3 and 6 tends to put them in type 1 clefting of Tessier's anatomic classification of craniofacial clefts (Tessier, 1976). However, the duplication of the nasal septum is seen in type O clefting, thus complicating the classification in case 3.

The cases presented were from Okirika, Port Harcourt, and Ahoada, which are local government areas in the River State of Nigeria. These are only three of the 10 local government areas served by this hospital (Fig. 7). All of the families involved belong to the lower socioeconomic class and cannot be described as being westernized in their way of living. There was no family history of similar anomalies or of any form of clefting. The role of febrile illness and medication in early pregnancy in these cases is not certain. The patients came from a malaria endemic zone and chloroquine is commonly ingested for most febrile illnesses, although malaria is not always the culprit.

Because the cases presented are varied, it is probable that there are multiple etiologies. Accurate documentation of these and other cases, detailed epidemiologic studies, and examination of the roles of febrile illnesses, medication in early pregnancy, and environmental pollution (Longo, 1980) may shed more light on the etiology of these conditions.

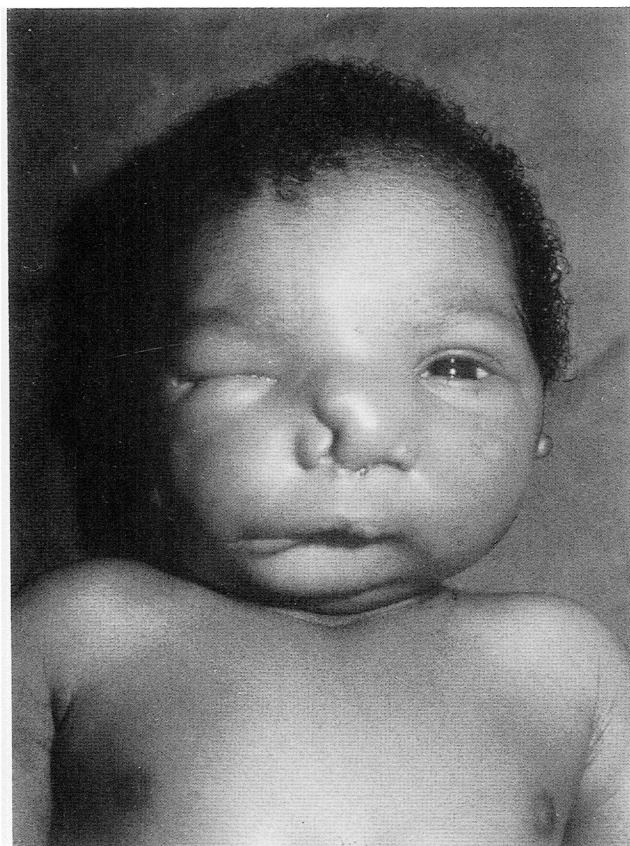


FIGURE 6 Case 7, showing a right-sided nasal cleft, right-sided commissural cleft, and hypertelorism. A right-sided grade I microtia is not visible in this photograph.

RIVERS STATE: LOCATION OF REFINERY, PETROCHEMICAL AND FERTILIZER INDUSTRIES.

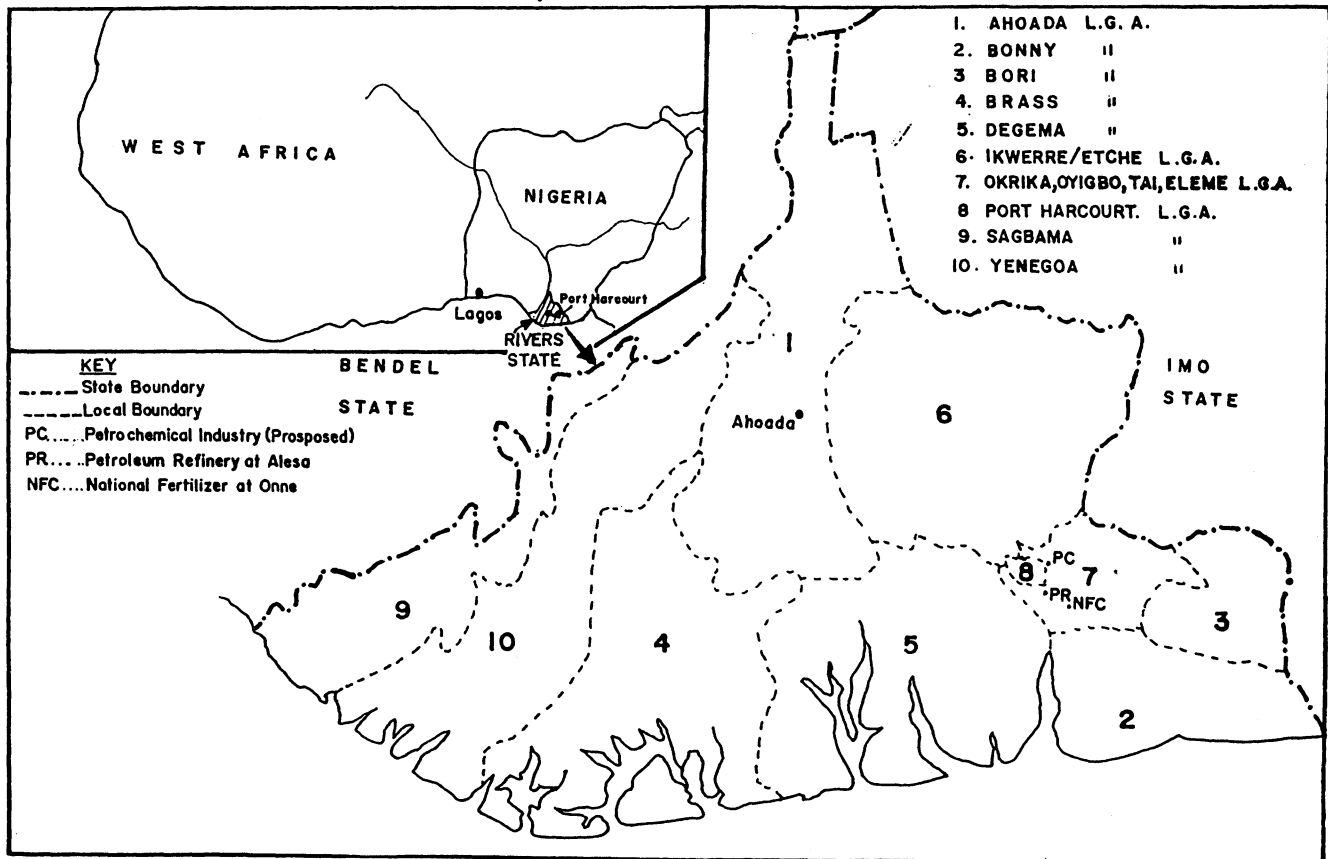


FIGURE 7 A map of River State (shaded area) in Nigeria that shows local government boundaries.

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Commentary

The seven interesting cases presented by Datubo-Brown confirms previous observations that facial clefts are heterogeneous in etiology regardless of the population sampled. The speculation by the author that atypical clefts may make up a higher proportion of all facial clefts in a black African population is certainly deserving of further study. It would be interesting to know whether similar observations have been made by investigators in adjacent countries or whether similar (or diverse) observations have been made in other racial subgroups.

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EDITOR'S NOTE: *The Editor would like to reinforce Dr. Jones' comments and call for the readership to submit any materials similar to those of Datubo-Brown to the Journal.*