ARE CONGENITAL ANOMALIES COMMON IN JOS- NIGERIA?

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ABSTRACT

Objective

The study was prospectively conducted in the Special Care Baby Unit of the Jos University Taeching Hospital, Jos over a 28-month period to assess the pattern of congenital anomalies (CA).

Method:A register of all newborn infants seen with congenital anomalies was kept in the special care baby unit.

Result:A total of 104 babies with CA were seen accounting for 6.1% of all admissions into the unit, with an inborn incidence of CA being 5.3 per 1000 live-births during the period. Gastrointestinal lesions accounted for majority, 33.7% (35/104), of the lesions but myelomeningocoele was the single most common lesion seen accounting for 19.2% (20/104) of all CA. The different lesions seen were found to have affected most of the systems and a few lesions 5.8% (6/104) could not be readily classified. Mortality rate was 33.7 % (35/104) among the patients with CA, being worse in outborns. Mortality was 100 % (9/9) among patients with chromosomal anomalies followed by patients with gastrointestinal lesions with 45.7% (16/35).

Conclusion:CA are common in this center. The lesions involve most of the organ systems. Prompt action is required to decrease mortality especially in case of correctible lesions and it is recommended that carcful examination of all newborn prior to discharge would help in early detection of congenital anomalies.

Key words: Congenital, anomalies, newborns, mortality

Introduction

Congenital anomalies (CA) are an important cause of morbidity and mortality in the newborn in developing countries. They accounted for 3.3% of all admissions and up to 8% of all mortalities in a newborn unit in Tanzania (1). In India, it has been observed that they contribute 22% of all early neonatal deaths (2).

One report suggests that congenital central nervous system (CNS) anomalies, particularly myelomeningocoeles are the most common congenital anomalies (1). The incidence of congenital CNS anomalies in Nigeria varies from 1.37/1000 to 5.2/1000 live births (3,4). An earlier report from our unit gave an incidence of 7/1000 live births (5) for CNS anomalies and showed that myelomeningocoeles are the commonest congenital CNS anomalies.

Besides a few reports of congenital CNS anomalies (5,6), there has been no comprehensive report of the pattern of congenital anomalies in Jos, a city in the middle belt of Nigeria. A simple register of congenital anomalies was, therefore, opened in our Special Care Baby Unit (SCBU) on the 1st of March 1998. JUTH is a referral centre for Plateau and other neighbouring states in the Central and North Eastern part of Nigeria.

Patients and Methods

This report presents the pattern of CA seen in the SCBU of the Jos University Teaching Hospital (JUTH) in the 28 month period between 1st March 1998 and 30th June 2000. All babies with congenital anomalies seen in the SCBU of JUTH had the following data recorded in the CA register:

name, age at presentation, gestational age, and description of the lesions seen, treatment given and outcome. Patients were categorised based on the predominant lesion. Associated lesions (such as talipes equinovarus in a patient with myelomeningocoele) were not recorded separately. Attempts were made to classify cases with multiple CA into clinically recognizable syndromes (including chromosomal anomalies), using Smith's Recognizable Patterns of Human Malformation (7) as a guide. No postmortem examinations or chromosomal studies were done on any of the patients.

Results

A total of 104 babies with CA were seen during the 28 month period. They accounted for 6.1% of all the 1692 admissions during the same period and consisted of 64 males, 35 females and 3 intersex. Of the 104 patients, only 31 (29.8%) were born in our hospital; 73 (70.2%) were referred. During the period of study, there were 5850 live births in the hospital. The incidence of CA among babies born in JUTH was therefore 5.3 per 1000 births.

Birth weight: Birth weight was available for only 56 babies and ranged from 0.90-5.00 kg (mean 2.66±0.95). Twenty-four (42.9%) of them, weighed below 2.50 kg at birth. The gestational age was determined by dates in 99 babies of which 19 (19.2%) were preterm.

Age: The age at presentation ranged from 10 minutes to 28 days (mean 3.28± 4.97 days). The median duration of hospital stay was 9 days (range 1-81).

Lesions: The different lesions are as outlined in the table, which shows the numbers of patients with different conditions and the corresponding percentage of the total number of CA that each constitutes. The number of mortalities for individual lesions is also shown. However, the percentage mortality was based on the number of cases within an organ system.

The single most common CA was the myelomeningocoele accounting for 20(19.2%) of all anomalies. CNS anomalies altogether accounted for 27 cases (26.0%). Figures 1 & 2 show patients with frontonasal encephalocoeles. The patient in figure 2 was admitted at the age of 5 hrs, and had the mass excised but developed meningitis thereafter. He improved with antibiotics, staying a total of 81 days in the Hospital.

As a group, gastrointestinal lesions accounted for the majority 35(33.7%) of the lesions. Out of these imperforate anus and omphalocoeles topped the list, each accounting for 11(10.6%) of all the CA.

Six patients with cardiovascular (CVS) lesions were seen. Five of them were preterms with patent ductus arteriosus(PDA) which responded to the use of indomethacin. The sixth was an infant with cyanotic heart disease and associated complete heartblock. Two other patients had ventricular septal defects but were classified elsewhere based on their predominant lesions.

Musculo-skeletal lesions accounted for 14(13.5%) of all the CA. One of them is the conjoint twin shown in figure 3. Ultrasonographic examination on this patient revealed two hearts, one in the normal position (left precordial area) with a ventricular septal defect and the second with a single ventricle, in the left hypochondrial area.

The patient with cystic hygroma shown in figure 4, died after the infected multilocular mass was drained of 300 mls of pus. The mass had shown evidence of inflammation including erythema, tenderness and fever soon after admission. There had been no previous attempt at draining it. The four patients with polydactyly and one with an accessory nipple were admitted primarily because of neonatal sepsis. These lesions should really be classified as minor CA.

There were 9 patients with recognizable chromosomal lesions as shown on the table. One of these, the only one with trisomy 21 had a Tracheo-oesophageal fistula. The patient died of

pneumonia prior to surgical intervention. One patient each had the following clinically recognised synromes:Prune-Belly, Pierre-Robin, Treacher-Collins and Potter. Based on clinical patterns, three anomalies were recognized: Trisomies 13, 18 & 21. Six patients could not be classified (see the table).

Mortality: Thirty-five of the 104 babies died, giving a mortality rate of 33.7%. This was significantly higher than the 19.5% overall mortality in the unit during the same period (p=0.0001). Similarly, outborn babies had a significantly higher mortality of 29/73(39.7%) when compared with the inborns 6/31(19.4%; P=0.04). Deaths from CA accounted for 10.6% (35/330) of all mortalities in the SCBU during the period of study. The immediate causes of death were not analyzed. Intra-group mortality rates are as depicted in the table. All the patients with chromosomal anomalies died (100% mortality). This was followed by those with gastrointestinal lesions with 16/35 deaths (45.7%). None of the patients with isolated CVS lesions died.

DISCUSSION

This report has demonstrated the very wide range of CA seen in the SCBU of the Jos University Teaching Hospital, Jos. The emergence of myelomeningocoele as the single commonest CA (19.2% of all cases) is consistent with what was recently reported from East Africa(1). In the 1960s, however, other Nigerian workers had reported anencephaly (3) and hydrocephalus (4) as the commonest CA of the central nervous system. The incidence of CA among the inborns in the current report (5.3 per 1000), is low compared to 16.5 per 1000 from a Beirut hospital (8) and 1.31% from a population-based study in Leeds (9). This difference may be attributed to better referral systems in Leeds and Beirut.

The classification of CA here has been very simple. It can be argued, and rightly so, that polydactyly and accesssory nipples included in this study are really minor CA. Their inclusion has been based on the fact that the objective of the study was to describe the pattern of CA seen in the SCBU. Most minor CA are not referred to the SCBU. A clearer picture of such minor CA, and indeed all CA, would be obtained if all newborns were screened at birth in the labour room.

Our inability to conduct post-mortems and chromosomal studies may have prevented proper classification of the six unclassifiable CA. It is becoming increasingly evident that multiple CA are associated with chromosomal anomalies (10-12). Chromosomal studies should, therefore, be encouraged in such cases. The lack of post-mortems may also have prevented the identification of some other lesions, such as transposition of the great vessels, which could be incompatible with life.

That the majority of the patients (70.2%) were referred from elsewhere is not unusual for a referral centre such as ours. The significantly higher mortality among babies with CA compared to the overall mortality in the SCBU during the period of review is noteworthy. Recent work has shown that CA are associated with a 17-fold increase in the mortality of Caucasian infants (13). The significantly higher mortality rate among our out born babies may be a result of delayed presentation to hospital. The high mortality rate associated with gastrointestinal lesions (45.7% overall, 80% for upper gastrointestinal obstruction and 66.7% for omphalocoeles) may be attributable to late presentation and supervening infections.

It is pertinent to point out that outcome, whether survived or died, was simply assessed for the duration of patients' stay in hospital. It is possible that more patients died at home after discharge. This may be true especially for babies with unclassifiable CA for whom no definitive treatment could be offered.

This preliminary report supports the casual observation that CA are common in Jos. Prompt action is required to decrease the high mortality

especially in the case of correctable lesions. We would agree with Richards et al(9) that a local CA register like ours, is a very valuable instrument, and could prove very useful in the monitoring and evaluation of public health interventions when such are introduced. Early antenatal ultrasonography and the routine examination of all newborns prior to discharge should be encouraged as this will improve the detection of CA. Better liaison with surgical teams should decrease the mortality rate among babies with CA.

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***Table: Distribution of 104 Congenital Anomalies seen in the Special Care Baby Unit of the Jos University Teaching Hospital.**

System/Lesion	Number	%	Mortality (%)
CNS			
Myelomeningocoele	20	19.2	3
Craniosynostosis	1	1.0	0
Encephalocoele	5	4.0	3
Hydrocephalus	1	1.0	0
Total	**27**	**26.0**	**6 (22.2)**
Cardiac			
Congenital Heart Block	1	1.0	0
Patent Ductus Arteriosus	5	4.8	0
Total	**6**	**5.8**	**0**
Musculo-skeletal			
Polydactyly	4	3.8	0
Conjoined twins	1	1.0	1
Growing tooth	1	1.0	0
Saccrococcygeal tumour	1	1.0	0
Osteogenesis imperfecta	1	1.0	0
Achondroplasia	1	1.0	0
Syndactyly	1	1.0	0
Phocomelia	1	1.0	0
Accessory nipple	1	1.0	0
Cystic Hygroma	1	1.0	1
Anterior abdominal wall cyst	1	1.0	0
Total	**14**	**13.5**	**2 (14.3)**
Gastrointestinal			
Cleft lip/palate	1	1.0	0
Upper GI obstruction	5	4.0	4
Gastroschisis	1	1.0	1
Imperforate anus	11	10.6	3
Omphalocoele	11	10.6	7
Hirschsprung's	6	5.8	1
Total	**35**	**33.7**	**16 (45.7)**
Urogenital lesions			
Polycystic kidneys	1	1.0	0
Ambiguous genitalia	1	1.0	0
Ectopia vesicae	1	1.0	0

Total	**3**	**2.9**	**0**
Clinical syndromes			
Pierre-Robin	1	1.0	0
Prune-Belly	1	1.0	0
Treacher-Collins	1	1.0	0
Potter	1	1.0	1
Total	**4**	**4.0**	**1 (25.0)**
Chromosomal anomalies			
Trisomy 13	4	3.8	4
Trisomy 18	4	3.8	4
Trisomy 21	1	1.0	1
Total	**9**	**8.6**	**9 (100.0)**
Unclassifiable	**6**	**5.8**	**1 (16.7)**

*All numbers in bold represent subtotals for individual classes.
The percentages given in the mortality column represent percent mortality in the individual classes only.

Figure 1

A patient with Frontonasal Encephalocoele.

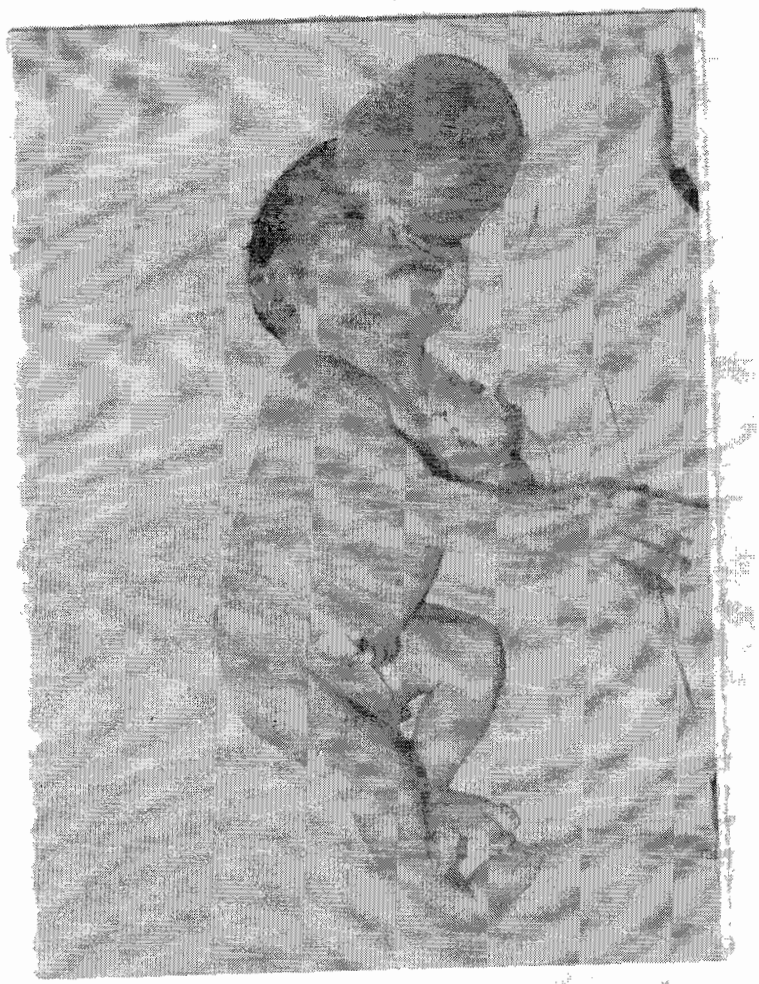

Figure 2

Ruptured Frontonasal Encephalocoele in a newborn infant.

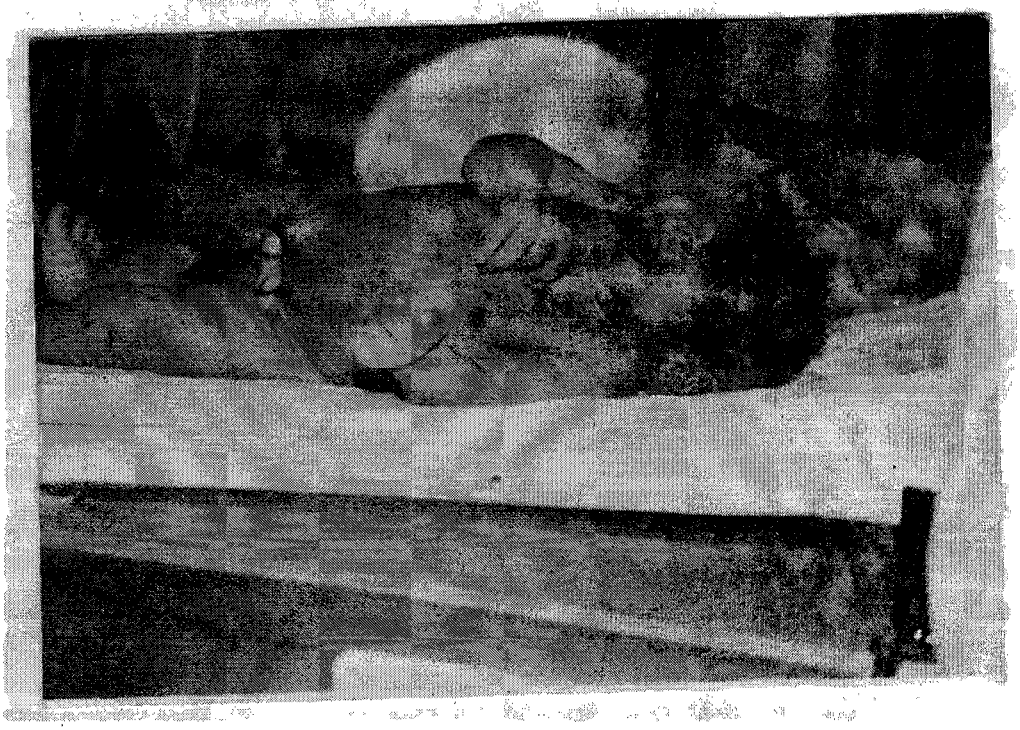

Figure 3

Conjoined twins seen at age 7 days.

Figure 4

A newborn infant with Cystic Hygroma