The Prevalence and Perinatal Outcome of Obvious Congenital Malformations among Inborn Babies at Aminu Kano Teaching Hospital, Kano

M Mukhtar-Yola*, M Ibrahim*, R Belonwu*, Z Farouk*, A Mohammed*

Summary


Background: The prevalence of congenital malformations varies from place to place. With a declining economy, increased utilization of traditional medications, and the traditional culture of consanguinity in Kano area, we decided to examine the incidence and pattern of presentation of congenital malformations in the city.

Objectives: To document the prevalence, pattern, contribution to perinatal mortality and outcome in babies with obvious congenital malformations.

Patients and Methods: A retrospective review of obvious major congenital malformations in neonates delivered at the Aminu Kano Teaching Hospital, Kano over a six-year period, was carried out. Data were retrieved from the case files of the neonates.

Results: The overall prevalence of congenital malformations was 5.5/1000 total births with a perinatal mortality rate of 60.7/1000 total births. Congenital malformations contributed 1.3 percent to the overall perinatal mortality. Gastrointestinal, central nervous system and unclassified malformations were the commonest. Malformations of the cardiopulmonary system had the highest case fatality rate.

Conclusion: The contribution of congenital malformations to perinatal mortality in Kano may be higher than reported in this study as the diagnosis of congenital malformations is often missed because of low autopsy rates and non-availability of advanced diagnostic facilities in our institution.

Key words: Congenital malformations, inborn babies, Kano

Introduction

CONGENITAL malformations are defined as structural abnormalities found at birth, or during the first week of life. These include congenital defects which cause irreversible functional disturbance of organs, cells or cell constituents resulting from disorders in either genetic constitution or adverse antenatal environment. The

Aminu Kano Teaching Hospital, Kano

Department of Paediatrics
* Consultant Paediatrician / Lecturer 1
* Professor
**Consultant Paediatrician/ Senior Lecturer
* Resident

Correspondence: Dr Mariya Mukhtar Yola.
E-mail: mariyamukhtar@hotmail.com

number of recognizable patterns of congenital malformations has more than tripled in the last 25 years. The potential prenatal effects of various drugs, chemicals and environmental agents are better appreciated and the number of genetic and non-genetic defects in which prenatal detection is possible has increased. The focus of discussion on congenital malformations in the developed world has shifted from a consideration of clinical presentation and management, to more sophisticated research on prenatal detection, treatment and prevention. However the pattern, prevalence, clinical peculiarities and the contribution of these malformations to perinatal morbidity and mortality in the developing world is yet to be fully ascertained. The incidence of congenital malformations varies from one part of the world to the other and indeed, within the same country. With worsening levels of socio-economic status, utilization of traditional medications, increased
patronage of patent medicine stores and purchase of “off the counter” potentially teratogenic drugs, and the traditional culture of consanguinity in Kano, it would be instructive to document the prevalence, pattern and outcome of congenital malformations and its contribution to perinatal mortality. It is against this background that this study was undertaken in order to document our experience of cases of congenital malformations seen at the Aminu Kano Teaching Hospital (AKTH), Kano, over a period of six years.

Patients and Methods

The records of all deliveries including stillbirths, with birthweights of 500 grammes and above that took place at AKTH between October 31, 1998 and November 1, 2004 were analysed retrospectively. The routine in the hospital was that all newborns delivered were examined initially by the duty obstetrician and nurse and later by the paediatrician. Babies with major malformations and others requiring further treatment and observation were then transferred to the special care baby unit. Diagnosis of malformation was made clinically and in some cases, with the support of investigations.

Details of deliveries during the study period were obtained from the labour ward and neonatal records, and from these, neonates with congenital malformations were identified. Relevant information about the babies and their mothers were extracted from the case notes. Autopsies were not performed on any of the stillbirths or the neonates that died because of cultural and religious barriers.

The data obtained were analysed using SPSS 10.1 version. The data were then tabulated according to the classification codes established by WHO. The types of congenital malformations were grouped as organs and systems.

Results

During the study period, a total of 13,619 babies were born at the hospital. Of these, 75 babies had major congenital malformations, thus giving a prevalence rate of 5.5/1000 total births. There were 30 males and 45 females with a M: F ratio of 2:3. Of the 75 babies with malformations, 68 (90.7 percent) were term babies and only seven (9.3 percent) were preterm. Table I summarizes the maternal characteristics and obstetric profile. Table II shows the perinatal outcome of the babies. Eight of the babies (10.7 percent) had multiple system malformations, while the remaining 67 (89.3 percent) had single system malformations. Five of the babies had recognizable syndromes; of these, two had Down syndrome, and one each had Prune Belly syndrome, Beckwith-Wiedemann syndrome and Pierre Robin syndrome. Among the single system malformation group, gastrointestinal (GIT) malformation was the commonest accounting for 38.7 percent, followed by central nervous system (CNS; 29.2 percent), unclassified (8 percent), musculoskeletal system (MSS; 6.7 percent), cardiopulmonary system (CPS; 6.7 percent), chromosomal malformations (5.3 percent), cutaneous (4 percent) and genitourinary (GU; 1.3 percent).

During the same period, 828 perinatal deaths were recorded in the labour ward and neonatal unit of the hospital giving an overall perinatal mortality rate of 60.7/1000 total births; 703 (84.9 percent) were stillbirths while

### Table I

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>No of Cases</th>
<th>Percent of Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primigravidae</td>
<td>20</td>
<td>26.7</td>
</tr>
<tr>
<td>Multigravidae</td>
<td>53</td>
<td>70.6</td>
</tr>
<tr>
<td>Parity not stated</td>
<td>2</td>
<td>2.7</td>
</tr>
<tr>
<td>Mean maternal age (yrs) ± SD</td>
<td>24.4 ± 4.31</td>
<td></td>
</tr>
<tr>
<td>Age range (yrs)</td>
<td>15-40</td>
<td></td>
</tr>
<tr>
<td>Age not stated</td>
<td>12</td>
<td></td>
</tr>
<tr>
<td>Residence</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Kano</td>
<td>58</td>
<td>77.3</td>
</tr>
<tr>
<td>Jigawa</td>
<td>12</td>
<td>16.0</td>
</tr>
<tr>
<td>Katsina</td>
<td>5</td>
<td>6.7</td>
</tr>
<tr>
<td>Ethnic Group</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hausa/Fulani</td>
<td>68</td>
<td>90.7</td>
</tr>
<tr>
<td>Igbo</td>
<td>6</td>
<td>8.0</td>
</tr>
<tr>
<td>Yoruba</td>
<td>1</td>
<td>1.3</td>
</tr>
</tbody>
</table>
the remaining 125 (15.1 percent) were early neonatal deaths. Nineteen (25.3 percent) of the seventy-five neonates died; thus constituting 1.3 percent of the total perinatal deaths.

Further analysis as shown in Table III, revealed that the cardiopulmonary system had the highest case fatality rate followed by GIT, unclassified multisystem anomalies, chromosomal and the CNS. There was no death recorded in the MSS, GU or cutaneous malformation groups.

Discussion

Although much reliance has been placed on hospital data to provide an insight into problems, the limitations of a hospital based study in correctly assessing the magnitude of such problems in the society is well recognized. Unfortunately in many developing countries, community based studies are rendered virtually impossible because of the paucity of records of births, diseases and deaths.

The prevalence of congenital malformations in the present study which was also the first study of its type at our centre was 5.5/1000. This prevalence is similar to those reported from other centres in Nigeria, but lower than the 15.8/1000 reported from Lagos. This difference may be due to the cosmopolitan, multiethnic and industrial nature of Lagos. Malformations of the

Table II

Perinatal Outcome in Babies with Congenital Malformations

<table>
<thead>
<tr>
<th>Outcome</th>
<th>No of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total births</td>
<td>13,619</td>
</tr>
<tr>
<td>Total perinatal deaths</td>
<td>828</td>
</tr>
<tr>
<td>Total early neonatal deaths</td>
<td>125</td>
</tr>
<tr>
<td>Overall perinatal mortality rate (per 1000 total births)</td>
<td>60.7</td>
</tr>
<tr>
<td>Number of babies with congenital malformations</td>
<td>75</td>
</tr>
<tr>
<td>Early neonatal deaths among malformed babies</td>
<td>19</td>
</tr>
<tr>
<td>Overall incidence of congenital malformations (per 1000 total births)</td>
<td>5.5</td>
</tr>
<tr>
<td>Contribution of congenital malformations to overall perinatal mortality rate</td>
<td>1.3</td>
</tr>
</tbody>
</table>

Table III

Outcome in Babies with Congenital Malformations by Specific System

<table>
<thead>
<tr>
<th>System</th>
<th>No. of Cases</th>
<th>No. Alive</th>
<th>No. Referred</th>
<th>Discharged Against Medical Advice</th>
<th>No. of Deaths</th>
<th>Case Fatality %</th>
</tr>
</thead>
<tbody>
<tr>
<td>GIT</td>
<td>29</td>
<td>14</td>
<td>3</td>
<td>2</td>
<td>10</td>
<td>34.5</td>
</tr>
<tr>
<td>CNS</td>
<td>22</td>
<td>8</td>
<td>10</td>
<td>1</td>
<td>3</td>
<td>13.6</td>
</tr>
<tr>
<td>MSS</td>
<td>5</td>
<td>2</td>
<td>0</td>
<td>1</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>CPS</td>
<td>5</td>
<td>3</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>40.0</td>
</tr>
<tr>
<td>Chromosomal</td>
<td>4</td>
<td>3</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>25.0</td>
</tr>
<tr>
<td>Cutaneous</td>
<td>3</td>
<td>3</td>
<td>0</td>
<td>0</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Genitourinary</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Unclassified</td>
<td>6</td>
<td>4</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>33.0</td>
</tr>
</tbody>
</table>
Table IV
Spectrum of Congenital Malformations in 75 Newborns

<table>
<thead>
<tr>
<th>Malformation</th>
<th>No of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Meningocele/Myelomeningocele</td>
<td>12</td>
</tr>
<tr>
<td>Omphalocele</td>
<td>12</td>
</tr>
<tr>
<td>Encephalocele</td>
<td>8</td>
</tr>
<tr>
<td>Imperforate anus</td>
<td>8</td>
</tr>
<tr>
<td>Hirschsprung’s disease</td>
<td>4</td>
</tr>
<tr>
<td>Congenital heart disease</td>
<td></td>
</tr>
<tr>
<td>(3 VSD, 1 TGA)</td>
<td>4</td>
</tr>
<tr>
<td>Hydrocephalus</td>
<td>3</td>
</tr>
<tr>
<td>Skeletal malformations</td>
<td>3</td>
</tr>
<tr>
<td>Multiple congenital malformations</td>
<td>3</td>
</tr>
<tr>
<td>Cleft lip and palate</td>
<td>2</td>
</tr>
<tr>
<td>Down syndrome (Trisomy 21)</td>
<td>2</td>
</tr>
<tr>
<td>Choanal atresia</td>
<td>2</td>
</tr>
<tr>
<td>Others</td>
<td>11*</td>
</tr>
<tr>
<td>Total</td>
<td>75</td>
</tr>
</tbody>
</table>

VSD-ventricular septal defect
TGA- Transposition of the great arteries
* One case each of anencephaly, conjoined twins, sacrococcygeal teratoma, inguinal hernia, Patau syndrome (Trisomy 13), Pierre Robin syndrome, Beckwith-Wiedemann syndrome, congenital cataract, tracheoesophageal fistula, cystic hygroma, ectopia vesicae

GIT and CNS were the commonest in our series; the CNS, GIT and unclassified malformations however, had the highest case fatality rates. This is similar to findings of other workers\textsuperscript{7} in Nigeria. Congenital malformations accounted for 1.3 percent of all the perinatal deaths in this study. It is much less than the 26 - 34 percent reported from the United Kingdom.\textsuperscript{9-10} This difference is most likely related to the fact that the more technically developed countries have experienced a decline in most of the major contributors to perinatal mortality with a result that congenital anomalies are now taking leading roles as major causes of perinatal mortality. Factors which had led to such decline include improved obstetric and neonatal management with reductions in the incidence of low birth weight and premature deliveries, improved facilities for prenatal diagnosis, early management of congenital anomalies and timely effective treatment of diseases that result from factors in gestation and during labour and delivery.\textsuperscript{11}

It has been reported that the prevalence of congenital malformations among stillbirths is high.\textsuperscript{12} In the present series, 85 percent of the perinatal deaths were stillbirths and so if autopsies had been carried out, the contribution of congenital abnormalities to perinatal mortality might have been higher. Furthermore, the absence of special investigation facilities such as karyotyping, cytogenetic analysis, special staining techniques, molecular testing, special biochemistry or electron microscopy impede the quality of research undertaken in developing countries.\textsuperscript{13}

The prevalence of congenital malformations may be reduced by improving obstetric practices such as routine biochemical and ultrasonic evaluation of 'at risk' pregnancies, provision of adequate nutrition in pregnancy, preconception folic acid supplementation,\textsuperscript{14} avoidance of harmful traditional medications and potentially teratogenic orthodox medications.\textsuperscript{4} The role of counselling for parents at risk of having another malformed child cannot be overemphasized.\textsuperscript{14}

References
Unusual Presentation of Infantile Hypertrophic


