Incidence, clinical presentation and outcome of gastroschisis and omphalocele in Sudan

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Original Article

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Encyclopedia of Incidence, clinical presentation and outcome of gastroschisis and omphalocele in Sudan

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Abstract

Gastroschisis and omphalocele have different etiologies and clinical presentations which dictated different approaches of surgical management. The aim of this study was to assess the clinical presentation and outcome of surgical management of these anomalies in Sudan, their prenatal diagnosis, associated anomalies and risk factors.

Methods

Fifty-one cases from all pediatric surgery units in Sudan were studied retrospectively and prospectively over the two years from June 2011 to June 2013. A data collection proforma including history, clinical examination, operative management and follow-up was filled.

Results

The incidence of omphalocele was 3:100000 live births, male to female ratio was 1:1.5 while the incidence of gastrochisis was 3:1000.000 live births with equal gender ratio. Thirty-three (72%) children with omphalocele were discharged well, 14 (28%) died, 7 of them died before surgery. Of these, four had congenital heart disease (CHD), two had other anomalies and one died suddenly. Post operatively, six patients developed sepsis and one died on the operating table. Only one infant (25%) with gastrochisis recovered and was discharged well. Fetal ultrasound diagnosed four out of 28 cases of omphalocele, and two out of four cases of
gastroschisis. Seventy-two percent of patients with omphalocele and 75% of those with gastroschisis had no other demonstrable associated anomalies.

Conclusions
There is low incidence of gastroschisis and omphalocele in Sudan with suboptimal prenatal diagnosis. The majority of cases had no associated anomalies and there were no apparent risk factors. Outcome of omphalocele was satisfactory but there was poor outcome of gastroschisis.

Keywords: Gastroschisis, omphalocele, abdominal wall defect, Sudan

Introduction
Gastroschisis and omphalocele have different etiology and clinical presentation which dictated different approaches of surgical management. Survival of newborns with omphalocele and gastroschisis has improved, but controversy remains regarding etiology, anatomy, embryology, the role of prenatal diagnosis, mode of delivery and initial management. In gastroschisis, young maternal age and interaction between smoking and gene polymorphisms have been suggested as predisposing factors\(^1\), while occurrence of omphalocele in twins, in consecutive siblings, in different generations of the same family as well as the incidence with trisomy 13, 18, and 21 (accounting for 25-50% of cases), and with Beckwith-Wiedemann syndrome suggest a genetic etiology\(^2\). The incidence and outcome of the management of omphalocele and gastroschisis in Sudan has not been documented previously.

Material and Methods
This is a descriptive observational combined prospective and retrospective cross sectional hospital-based study done at all Paediatric Surgery units in Sudan (Soba university hospital, Khartoum teaching hospital, Omdurman teaching hospital, Military hospital, Police hospital, Khartoum north hospital, Wadmadani Hospital Sennar Hospital and Eldammer Hospital) and included all cases of gastroschisis and omphalocele that presented during a period of 2 years from June 2011 to June 2013. A data collection sheet including history, clinical examination, operative management and follow up was filled. SPSS was used in data analysis.

Results
According to the statistics department of the federal ministry of health (FMoH) of the Sudan, the total number of live births from June 2011 to June 2013 was approximately 1.5 million. The total number of studied cases was 51 of whom 4 (7.8%) had gastroschisis and 47 (92.2%) had omphalocele. There were 29.8% exomphalos major and 70.2% exomphalos minor. The incidence of omphalocele was 3 in 100000 live births and the incidence of gastroschisis was 3 in 1000000 live births.
The male to female ratio was 1:1.5. There was only one neonate with a developmental sex disorder (DSD intersex). Three babies were preterm (6.4%) while the majority was born at term (93.4%) and the mean birth weight was 2.8kg (+/- 0.76 SD). The mean maternal age was 29.7 years with age less than 20 years in 4% and more than 30 years in 38% of the cases. Parity had no significant impact on the incidence of omphalocele, 54% of mothers had between 2-5 children. Pregnancy was complicated by febrile illness in 14% of the cases, anaemia in 3%, polyhydramnios in 2%, threatened abortion in 5% and while the majority (76%) had an uneventful pregnancy. Associated congenital abnormalities were encountered as follows: gastrointestinal tract in 4% (imperforate anus), cardiac in 4% (cyanotic congenital heart disease), genitourinary in 4% (undescended testes), neural tube defect in 7% (hydrocephalus-spina bifida) and musculoskeletal system in 7% polydactyly–talipes equinovarus (TEV). There were no associated anomalies in the remaining majority of 72%. Fetal ultrasound was done to 28 cases (59.6%). Only four cases of omphalocele were diagnosed prenatally. Out of 47 cases, 33 were discharged well while 14
died (Table 1).

Table 1: Shows the outcome of omphalocele in neonates who presented to paediatric surgery units in Sudan from June 2011 to June 2013.

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Omphalocele minor</th>
<th>Omphalocele major</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Discharged well</td>
<td>21</td>
<td>12</td>
<td>33</td>
</tr>
<tr>
<td>Death</td>
<td>12</td>
<td>2</td>
<td>14</td>
</tr>
<tr>
<td>Total</td>
<td>33</td>
<td>14</td>
<td>47</td>
</tr>
</tbody>
</table>

P value = 4.2

In 7 cases, death occurred before surgery, four of them had possible congenital heart disease and were awaiting echocardiography, in addition, one of the four had cleft lip and palate with polydactyly and another had Beckwith-Wiedemann syndrome. Two of the neonates had associated anomalies in a form of hydrocephalous with myelomeningocele, talipes equinovarus (TEV) and femur fracture and he died immediately while the other had imperforate anus with genitourinary fistula. The last one had omphalocele with no other anomalies, scheduled to be operated on later but he died suddenly at home. The other 7 were operated on; six of them died with postoperative sepsis, while one had ruptured omphalocele and was an intraoperative death.

Gastroschisis

Only four cases of gastroschisis were reported over two years period, male to female ratio was 1:1. Two of the cases (50%) were diagnosed during antenatal fetal ultrasound. One of the male neonates had undescended testis (UDT). Three babies were operated on while the 4th died preoperatively. Out of the three neonates who were operated on only one survived after successful staged repair using blood bag (Fig 1). The second one underwent primary reduction and died intraoperatively. The last one underwent staged repair using polyproline mesh network covered with Vaseline gauze and died postoperatively.

Discussion

According to the Statistics department of the FMoH of the Sudan, the total number of live births was approximating 1.5 (actual figure 1,439,768) in the period from June 2011 to June 2013. The incidence of omphalocele was
3:100,000 and the incidence of gastroschisis was 3:1,000,000. A hidden mortality (terminations - miscarriages - stillbirths and patients who died before coming to the surgical units) was not included. This incidence is low compared to that in other countries such as Japan and England. In Japan, the incidence of gastroschisis increased from 0.131 cases per 10,000 births from 1975-1980 to 0.467 cases per 10,000 births from 1996-1997; the incidence of omphalocele increased from 0.322 cases per 10,000 births from 1975-1980 (which was similar to the reported incidence of omphalocele in Sudan in 2011-2013) to 0.626 per 10,000 births in 1996-1997. In England and Wales, the incidence of gastroschisis doubled from 1.13 cases per 10,000 births in 1987 to 1.35 per 10,000 births in 1991(68 fold compared to Sudan) while the incidence of omphalocele decreased from 1.13 cases per 10,000 births in 1987 to 0.77 per 10,000 births in 1991(3).

In our study, although most of the babies were term, they were of low or very low birth weight which is a suspected risk factor for the disease(4,5,6), furthermore this could be affected by poverty and poor nutrition which may lead to intrauterine growth retardation. On the other hand, other known etiological risk factors influencing the incidence of omphalocele such as smoking, alcohol consumption(7,8) are rare among women in child bearing age in Sudan. Moreover, there were no reported significant maternal diseases or use of medications during pregnancy. The international survival rates of omphalocele increased significantly from 60% in 1960 to 90% recently(9) due to the improvements in respiratory care, pharmacology, anesthesia and surgery especially in developed countries. In Sudan the survival rate was 72%. In the literature, omphaloceles were known to be associated with other anomalies in more than 70% of the cases and the severity of the associated anomalies determines the prognosis (10). The mortality rate was 80% when associated anomalies were present, and it increased to 100% when chromosomal and cardiovascular abnormalities were present. Most associated anomalies were chromosomal(6).

In our data, the low incidence of associated anomalies with omphalocele (28%) may have helped reduce the mortality rate.

Regarding prenatal diagnosis, only four cases out of the 28 cases that underwent fetal ultrasound had been positively diagnosed with omphalocele. This may be due to difficulty in differentiation between a midline omphalocele and physiologic midgut herniation in early pregnancy(6,11). The correlation of fetal ultrasound to the gestational age was not assessed in this study. In the International data, routine fetal ultrasonography between 10 to 36 weeks had 100% sensitivity in detecting omphalocele(12) the low sensitivity in our data 14% could be explained by the dependency of ultrasound on the operator's experience, the reliability and quality of machine resolution and the fetal gestational age.

Gastroschisis

Low socioeconomic status, young maternal age, intra uterine growth retardation and subsequently low birth weights increase the risk of gastroschisis(4,5,6). Although these risk factors are prevalent in Sudan, they did not seem to increase the incidence of gastroschisis, perhaps this can be explained by lack of other incriminated risk factors such as the use of recreational drugs,(13-25) which are known predisposing factors for gastroschisis. The low incidence of gastroschisis may also draw attention to the genetic susceptibility that has been reported in the international data, in which African Americans have lowest risk for gastroschisis(26) while Hispanics have the highest one (2,13,25,27,28,29). This limited number of reported cases in our study may not represent the true scale of the incidence. In Sudan, many of the deliveries take place at home and, accordingly, some of these births may have included gastroschisis.
that either had been neglected and left to die or failed to reach surgical services before the death of the newborns.

In our study there were no reported dysmorphic features, and no other associated anomalies, except one who had undescended testis. This is consistent with available literature which documented the observation of absence of associated anomalies with gastroschisis apart from bowel atresia (10%) and undescended testis (31%)\textsuperscript{30,31}.

The type of repair affects the outcome of gastroschisis. As shown in this study, out of the three neonates who were operated on, only one survived after staged repair, using blood bag as a substitution to silo bag that is widely used in the staged repair of gastroschisis in the developed countries. Blood bag is a cheap, easy to use, smooth, soft flexible, impermeable, anticoagulated and sterile bag which can be readily available as a safe alternative to silastic silo and the spring loaded silo bags.

In conclusion, low incidence of abdominal wall defects in Sudan may be due to lack of environmental factors such as drugs, smoking and alcohol abuse. Prenatal diagnosis of abdominal wall defects showed low sensitivity when compared to other data. Infrequent coexistence of associated anomalies may have contributed to good outcome of the management of omphaloceles. Use of blood bag in the management of gastroschisis is a feasible, practical and safe alternative to silo bag.

References