EDITORIAL

Clinical Presentation and the Outcome of Management of Neonatal Abdominal Wall Defects at Gezira National Centre for Pediatric Surgery, March 2008 to February 2013

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Abstract:

Objective: Anterior abdominal wall defects represent unique challenges to pediatric surgeons and offer insight into normal fetal development. Our main goal is to study the ation and the outcome of management of abdominal wall defects in neonates managed at Gezira National Centre of Pediatric Surgery.

Patients andMethods: This is a retrospective, prospective, descriptive hospital based study of patients presenting with clinical aspects of abdominal wall defects who were admitted to hospital in Gezira National Center of Pediatric Surgery (GNCPS) in Gezira state, Sudan, over 5years between March 2008 and Feb. 2013.

Results: The common age group is neonates, and most cases presented earlier than 24 hours (64.1%). Regarding the presenting symptoms, (33.3%) presented with eviscerated the small intestine was the most commonly eviscerated in (17.5%), and the liver was eviscerated in (4.8%). The operative mortality was 30 %. T he analysis of the type of found that: 13 patients (20.6%) were delivered vaginally at hospital, 10 patients (15.9%) were delivered by C/S, and 40 patients (63.5%) were delivered vaginally According to the management, surgery was done to 30 cases (47.6%), non surgical treatment done for 33 cases (52.4%) with good outcome. Primary closure was done for 20 cases (66.7%), Blood bank bag operation underwent for 6 cases (20.0%), and delayed hernia defects repair was done for 4 cases (13.3%). The trend in the centre is to conserve patients with intact membrane. Those with ruptured omphalocele, gastroschisis bowl obstruction will undergo urgent surgery. The analysis of the which revealed: Mortality of 9 cases (30%) the majority was due to failure and sepsis. Six cases had late complications (20%) were ventral hernia, and 27 cases had early complications (50%) most of these were

The overall mortality (30 %) can be explained by the lack of neonatal intensive care unit and unavailability of long term total parenteral nutrition. In Lukman study the mortality rate was 30.4% which is similar to ours [(13)]. The overall survival in children with omphalocele and gastroschisis is now reduced to less than 10 % in developed countries especially in isolated cases without chromosomal anomalies or major organ malformation[(17)].

Conclusion: The management of the anterior abdominal wall defect is challenging and need urgent intervention. Early presentation improves the management out come and decreases the mortality and complications, this need proper antenatal care follow up.

EDITORIAL

Key words: Omphalocele, Gastroschisis, amniocentesis.

Introduction:

Abdominal wall defects represent unique challenges to pediatric surgeons and offer insight into normal fetal development.Omphalocele and gastroschisis are the major anomalies encountered in the neonate. The first description of an abdominal wall defect dates back to 1634, when Ambroise Paré first described an omphalocele is a defect in abdominal wall musculature and skin with protrusion of abdominal viscera contained within a membranous sac. Survival of an infant born with an abdominal wall defect, especially gastroschisis, was unusual before the advent of modern antibiotics, nutritional support, and neonatal intensive care capabilities [(1)]. In its very mild form, a small loop of intestine protrudes into the base of the umbilicus; this is a hernia into the umbilical cord. In the more severe form, the defect allows protrusion of small intestine and other viscera, pushing the umbilical cord forward and distending its base into a cystic mass containing the viscera. This constitutes an omphalocele. Omphalocele is more common, with a general incidence of 1:4,000 births. Gastroschisis occurs in 1:10,000 births; although this is less common than exomphalos, in the Western world an increased incidence of tenfold is noted in young mothers with substance abuse. Gastroschisis is not due to or associated with impaired organ formation, but there could be complications from mass protrusion of viscera through a small defect, including vascular compromise, which in early fetal life could result in bowel atresia. The prevalence in sub Saharan Africa is not known as there are no population based studies. While the birth prevalence of omphalocele has remained generally stable over the years, reports from industrialized countries (Europe, United States, and Japan) indicate that the rate for gastroschisis is on the increase. When omphalocele is associated with other abnormalities, the etiology is multifactorial and incidence varies with age of the mother. These abnormalities occur more in younger mothers; omphalocele alone is more prevalent in older mothers, however [(2)]. Infants with gastroschisis and omphalocele can be identified by prenatal ultrasonography [(3)].When gastroschisis is identified antenatally, serial ultrasonography is performed to identify impending threats to the intestine; amniocentesis is used to monitor lung maturity and determine when to induce labor [(4, 5, 6, 7,8)].

A baby with a ruptured omphalocele is treated the same way as a baby with gastroschisis [(9)]. Closure of giant omphaloceles containing the liver is always challenging [(10, 11, 12)].

General Objective:

To determine the clinical presentation and outcome of abdominal wall defects at neonates in National Centre 0f Pediatric Surgery from March 2008 to Feb. 2013 **.**

Specific Objectives:

1- To analyze the demographic features of the studied population.

2- To assess prenatal care and mode of delivery.

3 - To describe the surgical anatomy of the defects in the study population.

4- To determine other malformations associated with abdominal wall defects. 5 – To describe the operative management, complications and outcome.

Patients and Methods:

EDITORIAL

This is a retrospective, prospective, descriptive hospital based study of patients presenting with clinical aspects of abdominal wall defects who were admitted to hospital in Gezira National Center of Pediatric Surgery (GNCPS) in Gezira state, Sudan, over 5years between March 2008 and Feb. 2013. The data were collected by using a patient data sheet, and were analyzed by computer using statistical program for scientific science, SPSS. All patients seen in GNCPS center due to abdominal wall defects were evaluated clinically regarding the age of presentation, the symptoms at presentation, the size of the defects, the eviscerated organ and treatment modality. A total number of 63 cases who fulfilled the inclusion criteria were available for analysis at the end of the study period. All patients who were initially diagnosed and managed at GNCPS as omphalocele or gastroschisis were included in the study. Patients admitted in other hospitals and underwent surgical exploration, and came with complications were excluded. All neonates who presented to the Gezira National Centre of Pediatrics Surgery within the defined study period, with features of symptomatic Bladder Exstrophy and Cloacal Exstrophy were excluded. The data were analyzed by computer using SSPS. The study was conducted at GNCPS which is modrately equipped; three floors, 70 beds facility that located in Wad Medani town, the capital of Gezira state. This strategic location makes the center provide its services for very large area in the country from eastern, south east and middle states, and also from some western and southern states. The staff is headed by specialized pediatric surgeon and includes another two pediatric surgeons and general surgeons under training, registrars of general surgery, medical residences, house officers, and trained nurses. The follow up of patients is clinically at the refferal clinic two weeks after discharge and then monthly.

Results:

The data of 63 patients were collected using a patient data sheet, and analyzed by computer using statistical program for scientific science (SPSS).From this study the finding was that the age group presentation was distributed as follow: less than one day 39 patients (61.9%), followed by between 1-7 days, 17 patients (27%), more than 7 days age group, 7 patients (11.1%). Thirty five patients (55.6%) were males and 28 patients (44.4%) were females, 36 patients (57.1%) from rural areas, and 27 patients (42.9%) from urban areas. According to the presentation, the symptoms were analyzed and revealed, 21 patients (33.3%) presented with eviscerated organs, 42 patients with intact membrane (66.7%) figure 1. Other associated presenting symptoms were analyzed and resulted in: 6 patients presented with vomiting (9.5%), 4 patients presented with fever (6.3%), 3 patients presented with delayed passing of meconium (4.8%), and 2 patients presented with jaundice (3.2%). According to antenatal care follow up we found 27 patients mother (42.9%) were on regular follow up, and the remaining were not on regular follow up. In the analysis of the type of delivery the finding was that: 13 patients (21.3%) were delivered vaginally at hospital, 10 patients (15.9%) were delivered by C/S, and 38 patients (62.3%) were delivered vaginally at

EDITORIAL

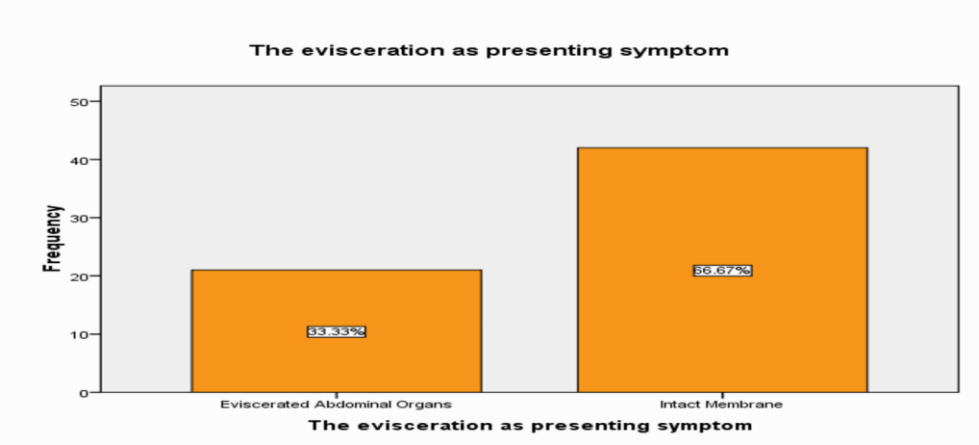

Figure 1: The distribution of evisceration as a presenting symptom during the neonatal period of 63 patients with anterior abdominal wall defects.

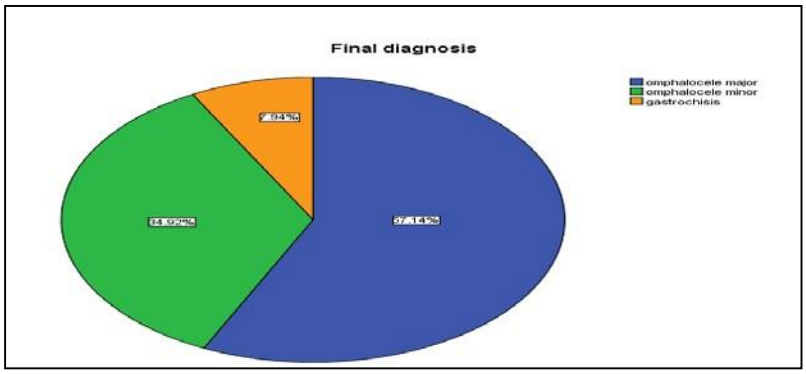

Figure 2: The distribution of the final diagnosis of 63 patients with anterior abdominal wall defects.

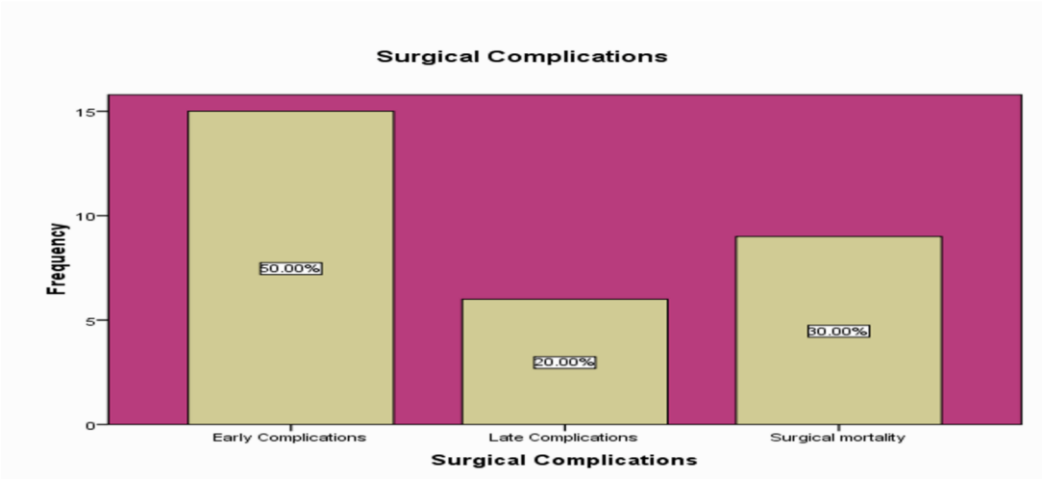

Figure 3: The distribution of the surgical complications of 30 patients with anterior abdominal wall defects.

EDITORIAL

home.The time lapse between the delivery and presentation was analyzed, and the finding was that 41 patients presented in the first day of onset of symptoms (65.1%), and 15 patients (23.8%) presented between 1 to 7 days, and 7 patients

(11.1%) presented after 7 days .The surgical anatomy of the defects were analyzed , measurements of the abdominal wall defects revealed: 35 patients had defect more than 5 cm in diameter (55.6%), and 24 patients had defect less than 5 cm (38.1%).The eviscerated organs were analyzed and the finding was that: the small intestine was eviscerated in 11 patients (17.5%), the liver was eviscerated in 3 patients (4.8%), the stomach and large intestine in one patient (1.6%), and there were more than one organ eviscerated in 6 patients (9.5%) included: small and large intestine in 2 cases (3.2%), liver, stomach and large intestine in 2 cases (3.2%), stomach, small and large intestine in 2 cases (3.2%). The analysis of the associated congenital malformation which revealed: cardiac anomaly in 4 patients (6.3%), skeletal anomaly in 4 patients (6.3%), genitourinary abnormality in 3 patients (4.8%), and neural tube defect in one patient (1.6%). Regarding the diagnosis the finding is that 36 patients were diagnosed as omphalocele major (57.2%), 22 patients were diagnosed as omphalocele mimor (34.9%), and 5 patients were diagnosed as gastroschisis (7.9%) figure 2. According to the management, surgery was done to 30 patients (47.6%) and conservative treatment done for 33 patients (52.4%). Primary closure was done for 16 cases (61.54%), Silo operation underwent for 6 cases (23.08%), and delayed hernia repair was done for 4 cases (15.38%). The analysis of the surgical complications which revealed : Mortality of 9 patients (30%) the majority was due to wound infections and sepsis , 6 patients had late complications (20%) were ventral abdominal hernia , and 27 patient had early complications (50%) most of this was infections figure 3. The high mortality can be explained by lack of neonatal intensive care unit and unavailability of long term total parenteral nutrition.

Discussion:

Increasing prevalence of omphalocele and gastroschisis has been reported in several regions of the world. Lukman et al quoted a figure of 2.4% of hospital admissions [13], in the study these defects accounted to 0.64 % of hospital admissions which is lower than Lukman figure. A large scale multi-centre study is needed to estimate the true prevalence in Sudan. From this descriptive study, the finding is that the main age group that presented with anterior abdominal wall defects is less than 1 day (61.9%). Most of our patients presented early and this is attributed to the alarming of bowl evisceration or a central defect seen by parents at birth. Thirty five patients (55.6%) were males and 28 patients (44.4%) were females, 36 cases (57.1%) from rural areas, and 27 cases (42.9%) from urban areas.The delayed presentation was observed among patients who were delivered at home and travelled from remote areas to attend the centre. In spite of the obvious anomaly there was a delay in presentation reported in a study done in Nigeria. In this study Lukman et al attributed late presentation to home delivery and remote residence [13]. In contrast all defects were diagnosed either prenatally or at birth in UAE [14]. There was no regular antenatal care follow up in (57.1%). Even among those with regular antenatal care only one case was diagnosed prenatally towards the end of the third trimester. If serial ultrasonography shows dilatation and thickening of the intestine in a baby with gastroschisis, delivery should occur as soon as appropriate. Once amniocentesis demonstrates lung maturity, delivery should be expeditiously induced. Postnatal evaluation to determine other congenital anomalies should occur prior to surgical intervention in those infants with omphalocele [15]. Prenatal diagnosis is of value in planning the mode of delivery which should be in a health facility capapable of treating such malformation. It is well known that delayed transfer leads to fluid loss and increases the incidence of infection. Babies with small omphaloceles have associated abnormalities more frequently, including intestinal problems (eg,

Meckel diverticulum, intestinal atresia), genetic syndromes (eg, BeckwithWiedmann, trisomy 18), and congenital heart disease. Babies with giant omphaloceles usually have small, bell-shaped thoracic cavities and minimal pulmonary reserve. Repair of the omphalocele may precipitate respiratory failure [16, 17]. The analys is of the associated congenital malformation which revealed: cardiac anomaly in 4 patients (6.3%), skeletal anomaly in 4 patients (6.3%), genitourinary abnormality in 3 patients (4.8%), and neural tube defect in one patient (1.6%). There are 4 cases associated with more than one anomaly. Lukman et al reported an incidence of 6.1 % of syndromic omphalocele. In the 1960s and 1970s, omphaloceles outnumbered gastroschisis 3:1, but over the last 20 years gastroschisis defects have predominated 2-3:1. This increase in incidence may represent increased selective termination of pregnancies of omphaloceles, more accurate classification of defects, or an actual increase in the gastroschisis birth rate [18]. Omphalocele maior is the most commonly diagnosed anterior abdominal wall defect (57.1%), gastroschisis is less common (7.9%), and omphalocele minor (34.9%). One third of cases presented with eviscerated organ (ruptured omphalocele and gastroschisis) (33.3%) and the most eviscerated organ is small intestine (17.5%), liver (4.8%), and more than one organ (9.5%).

Conclusion:

1- The management of the anterior abdominal wall defect is challenging and need urgent intervention.
2- Presentation is usually early because of the obvious defect
3- Prenatal diagnosis improves management and outcome. In the study area the percent of prenatal diagnosis is negligible.
4- Defects are observed to be large. This adds to the challenges and operative outcome.
5- Mortality is high due to inadequate transfer, large defects and lack of neonatal intensive care facility.

Recommendations:

1- Obstetrician and ultrasonographers need to increase their index of suspicion in the prenatal diagnosis of fetal malformations.
2- Though presentation is early, transfer of neonates occurs under less advantaged conditions like inadequate fluid replacement and leaving the eviscerated bowl exposed. This practice should be advised in patient transfer.
3- Tertiary canters should be prepared with well equipped neonatal intensive care units to reduce the overall mortality.

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EDITORIAL

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