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Experience with Exomphalos in a Tertiary Health Center in Nigeria

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Abstract

Background: There is paucity of data on the pattern, presentation and outcome of management of exomphalos in developing countries.

Patients and Methods: A retrospective study of all cases of exomphalos managed within a 15-year period at the Obafemi Awolowo University Teaching hospital, Ile Ife, Southwestern Nigeria.

Results: Ninety-one cases of exomphalos were managed over the study period. Forty-six (50.5%) were males and 45 (49.5%) were females (M:F=1:1). The mean age at presentation was 2.48days (range 2hours - 15 days). The median weight was 3.0kg (range 1.6 - 4.95kg). Sixty four (7.3%) patients had major exomphalos (defect > 5cm) while 27 (29.7%) were minor (defect <5cm). The size of the defects ranged from 3cm to 16cm (median 8cm). Associated anomalies were present in 40(44%) patients. Fifty-five (60.4%) patients were managed non-operatively while 36 (39.6%) had emergency surgical closure.

Over two-thirds (66%) of the patients managed non-operatively survived compared to 47.2% in the surgical group, but this was not statistically significant (p=0.231). The mortality rate among those with ruptured exomphalos was 75%. The overall mortality rate was 45 %(41 of 91). Sepsis was the most common cause of death while respiratory distress was common among those who had surgical closure.

The average duration of hospitalization in the non-operative group was 34 days (range 18-82 days) and 16 days in those who survived surgical repair (range 4-28 days).

Conclusion: For the less developed countries a non-operative approach is advocated in the management of omphalocele because mortality from emergency operative closure is very high.

Keywords: Exomphalos, Management, Outcome, Developing country

Introduction

Exomphalos is one of the most common forms of anterior abdominal wall defects with an incidence of 1: 4000 to 7000 live births[1]. The incidence of exomphalos varies with geographical locations and ethnic groups[1]. In Nigeria and other parts of African continent the incidence is unknown. Exomphalo is associated with a high incidence of chromosomal and structural anomalies including cardiac, skeletal renal and neural tubes that negatively impact the outcome[2-5]. There is presently a paucity of data on the presentation and outcome of management of newborn with exomphalos in Nigeria. The aim of this study is to document the current practice and outcome of management of exomphalos in a teaching hospital in south western Nigeria.

Materials and Methods

This was a review of all cases of congenital anterior abdominal wall defects managed at the paediatric surgery unit of the Obafemi Awolowo University Teaching Hospital, Ile Ife, Nigeria, between January 1988 and December 2002. Information regarding the bio-data, antenatal history, and birth history, mode of presentation, associated malformations, intervention and outcome of management were extracted from the patients' case notes and operation register. Among the patients who died, the possible cause of death was determined and the follow up history among the survivors noted. The exomphalos was classified into minor (defects less than 5cm) or major (defect greater than 5cm).

All the patients were fully resuscitated with intravenous fluid, antibiotics and vitamin K. The sacs were routinely covered with warm sterile saline

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soaked gauze. Patients who had ruptured exomphalos were taken to theatre for immediate closure while the patients who presented with infected sacs, prematurity or large exomphalos were initially managed non-operatively. The decision to do primary closure was made if it was determined that reduction and closure could be achieved without tension. There were no facilities for ventilatory management or parenteral nutrition. Non-operative management consisted of daily application of 70%alcohol on sterile gauze covering the sac. This was continued until the eschar separated and the wound was allowed to heal by granulation tissue. Those who had non-operative management later had repair of their ventral hernia between the ages of 12 to 18 months. Mortality was compared between the non-operative and the surgically managed group using Chi-square. P-value < 0.05 was assumed to be significant.

Results

There were 91 cases of exomphalos over the 15 year study period. The patients' characteristics are summarized in table 1. There were 46 (50.5%) males and 45 (49.5%) females (M: F; 1:1). The mean age at presentation was 2.48days (range 2hours 15 days). About 60% of the patients presented within 24hours after birth. Their median weight was 3.0kg (range: 1.6 - 4.95kg). Sixty four (70.3%) patients had major exomphalos while 27 (29.7%) were minor. The size of the defects ranged from 3 cm to 16 cm (median 8cm). Eighty two (90.1%) of the exomphalos had intact sac at presentation while 9 were ruptured. Three other patients later had ruptured sacs while on admission. Ninety five percent of the patients were full term; 85% were delivered spontaneously by vaginal route. About 29% of the babies were delivered either at home or in the mission house.

A total of 44 associated anomalies were recorded in 40(44%) patients, the most common of which were macroglossia (7), Polydactyl (6) and Beckwith-Weidman syndrome (5) (Table 2).

Fifty-five (60.4%) patients were managed non-operatively, with survival rate of 66% (33/55) and mortality rate of 34%. The average duration of hospitalization in this group was 34 days (range 18-82 days).

Thirty six (39.6%) patients had emergency surgical closure. Primary fascial closure was employed in 26 patients while skin mobilization and improvised prosthetic staged closure was used in 5 patients each [Table 3]. In 12 (33.3%) of the 36 patients, the reason for the closure was due to ruptured sac. Postoperative complications observed in 11 (30.6%) of the survivors who had surgical closure are shown in Table 4. The mortality in the emergency surgical group was 47.2 %(

Table 1: Background Information

Number of Patients	91
Types	
Major (Greater than 5cm)	64 (70.3%)
Minor (Less than 5cm)	27 (29.7%)
Antenatal History	
Booked	78 (85.7%)
Unbooked	13 (14.35)
Presentation	
Within 24 hours	55 (60.4%)
2-7 days	31 (34.6%)
8-15 days	5 (5.5%)
Gestational Age	
>/=37 weeks	86 (94.5%)
>37 weeks	5 (5.5%)
State of Sac	
Intact	(86.7%)
Ruptured	12 (13.3%)
Treatment Modality	
Non-Operative	55 (60.4%)
Surgical	36 (39.6%)
*Outcome	
Overall Mortality	41/91 (45.1%)
Non-operative Group	22/55 (44%)
Surgical Group	19/36 (52.8%)
Hospital Stay in Survivors	
Non-Operative	34 days (range 18-82 days)
Surgical	16 days (range 4-28 days)

*No statistically significant difference in mortality between Non-operative and surgical group (p=0.231)

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Table 2: Showing Associated Malformations in 40 Babies with Exomphalos

TYPE OF ANOMALIES	
Syndromic	
Beckwith Weidman Syndrome (BWS)	5
OEIS Syndrome	3
Sternal defects (? Pentalogy)	2
Downs Syndrome	1
Non Syndromic	
Macroglossia	7
Polydactyl	6
Undescended Testis	4
Gut malrotation	3
Patent Urachus	2
Syndactyl	2
Situs Inversus	2
Sacrococcygeal tumour	1
Webbed Neck	1
External Auditory Meatus Stenosis	1
Microphthalmia	1
Imperforate anus	1
Micrognathia	1
Microcephaly	1

Table 3: Treatment Modalities in 91 Patients with Exomphalos

Mode of Treatment	Number of Patients
Non-Operative	55 (60.4%)
Surgical	36 (39.6%)
1. Primary Fascia Closure	26
2. Skin Mobilization	5
3. Improvised Silo	5
Total	91

Table 4: Postoperative Complications in 10 Survivors

Complication	Frequency
Respiratory Distress	4
Wound Infections	3
Wound Dehiscence	3
Incisional Hernia	3
Jaundice	1
Entero-cutaneous fistula	1
Upper GI Bleeding	1
Orchitis	

Table 5: Probable Cause of Death in 23 Patients with Exomphalos

Cause of Death	Conservative	Surgical Closure	Total
Overwhelming Sepsis	11	3	14
Respiratory Distress		7	7
Enterocutaneous Fistula		1	1
Anaesthetic Complications		1	1
Total	11	12	23

17 of 36). The overall mortality among those with ruptured exomphalos was 75% (9/12). The average hospital stay was 16 days in those who survived surgical repair (range: 4-28 days).

The overall mortality was 45 %(41 of 91). There was no statistically significant difference between the mortality in the two groups (p=0.231).

Data on causes of death was available in 23 patients (Table 5). Overwhelming sepsis was the most common cause of death while respiratory distress was common among those who had surgical closure.

Discussion

The management of anterior abdominal wall defects (AWD) continues to be a major challenge to paediatric surgeons worldwide. While the advent of prenatal diagnosis, well equipped neonatal intensive care units (NICU) availability of total parental nutrition and ventilatory management has improved the survival in newborns with AWD in developed countries, the same cannot be said of developing countries.

Diagnosis was made at birth in all our patients. Although the majority of our patients had antenatal care in different hospitals, they did not have the benefit of prenatal ultrasound diagnosis because of lack of this service. The benefit of counseling, possible termina-tion of pregnancy, planned delivery and surgery associated with prenatal diagnosis is therefore not available. This scenery is likely to continue until routine screening for congenital anomalies become a

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standard.

There was a high rate of associated anomalies in this series. About 30 to 70% of exomphalos have associated anomalies, and up to 30% of cases would have chromosomal abnormalities[2-5]. Exomphalos associated with chromosomal and other severe congenital anomalies carries a high mortality rate of up to 30%[4-7]. The prognosis in exomphalos associated with normal karyotype is excellent[8]. It is suggested that the teratogenic insult in exomphalos occurred early in embryonic life which explains why there is an increased incidence of associated malformations[9,10].

Routine chromosomal analysis and screening for cardiac anomalies are not done in our hospitals because we lack facilities for these services; therefore it is possible that many other anomalies may have been missed. Because of the retrospective nature of this study it is very difficult to relate the high mortality in this series to the presence of associated anomalies. Five patients presented with the typical features of Beckwith Weidman Syndrome (BWS). Some of the patients only had associated macroglossia, it is known that in a minority of patients, one of the cardinal features of BWS may be absent[11]. None of the patients with BWS however had neonatal hypoglycemia.

The optimal management of patients with AWD is prompt or early surgical closure, either by primary or delayed primary surgical closure using the silastic silo. This approach cannot be routinely followed in our setting because of lack of facilities. Therefore unless there is rupture of the sac the decision to operate primarily in exomphalos is based on the surgeon's conviction that primary closure will not cause undue rise in intra-abdominal pressure. There is, however, no facilities to monitor this pressure changes in our hospitals. It is therefore not surprising that respiratory distress was frequently observed among those who have emergency closure.

We used primary fascial closure, skin flap closure and improvised silo using collapsible intravenous fluid giving set in ruptured exomphalos. The outcome however is far less favourable when compared to those managed non-operatively, and was worse for ruptured exomphalos. In ruptured exomphalos obtaining cover for the exposed viscera can be very challenging as there is no silastic sheet available. This led to the improvising of the collapsible saline bag in these patients. It is possible that many of these patients could have been saved if silastic sheet, total parenteral nutrition and ventilatory support were availability.

Those patients who had ruptured exomphalos before admission did not do well, although they tended to present earlier than intact exomphalos. The poor outcome could be attributed to lack of organized mode of transportation, pre-transport resuscitation, poor temperature control, sepsis and metabolic derangement.

The majority of our patients were managed non-operatively primarily because of lack of facilities to support immediate surgical closure. Even in centres where facilities for ventilatory support and TPN are available, non-operative management are still preferable in patients with giant exomphalos, extreme prematurity and presence of severe associated congenital anomalies[9].

In this review, there was a high postoperative morbidity following primary repair of exomphalos. The mortality was less in patients managed non-operatively than those managed surgically. This trend was also observed in an earlier series from Jos and Zaria in the Northern part of Nigeria[12-14].

The mean hospital stay was longer among the survivors in the non-operative group compared with those in the surgical group. This is because it takes a long time for the eschar to form and separate, and for granulation and epithelialization to cover the defect. Because of the financial implication involved, a few of our patients requested for discharge before complete epithelialization of the wound and were subsequently followed up in the outpatient clinic.

As in other parts of Nigeria and West Africa[12,15], the mortality rate in exomphalos in our environment is still

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L'expérience De L'exomphalos Dans Un Centre Tertiaire À Ile Ife Au Nigeria

Résumé

Arrière-plan: Il y a peu de données sur le mode, la présentation ou le résultat de la gestion de l'exomphalos aux pays en voie de développement. ***Malades et méthode:*** Une étude prospective de 15 ans traitant de tous les cas d'exomphalos gérés au centre hospitalo-universitaire Obafemi Awolowo à Ile Ife au sud-ouest du Nigeria. ***Résultats:*** Quatre-vingt-onze cas d'exomphalos ont été gérés pendant la période de l'étude. Quarante-six (50,5%) étaient des garçons et 45 (49,5%) des filles (M:F=1:1). L'âge moyen à la présentation à l'hôpital était de 2,48 jours (gamme 2 heures à 15 jours). Le poids médian était de 3,0kg (gamme 1,6 4,95kg). Soixante - quatre (70,3%) malades avaient un exomphalos majeur (décalage> 5cm) alors que 27 (29,7%) étaient mineurs (décalage <5cm). La dimension des décalages variait de 3cm à 16cm (médian 8cm). Des anomalies connexes se sont manifestées chez 40 malades (44%). Cinquante-cinq (60,4%) des malades ont été soignés non opérativement alors que 36 (39,6%) ont subi une fermeture chirurgicale d'urgence. Plus des deux tiers (66%) des malades soignés non opérativement ont survécu par rapport à 47,2% dans le groupe chirurgical, mais ce n'était pas statistiquement important (p=0.231). La mortalité parmi ceux atteints de l'exomphalos éclaté était de 75%. La mortalité totale était de 45% (41 sur 91). La septicité était la cause la plus fréquente de mort, alors que la détresse respiratoire était fréquente parmi ceux qui avaient une fermeture chirurgicale. La durée moyenne d'hospitalisation dans le groupe non-opératoire était de 34 jours (gamme 18 à 82 jours), 16 jours pour ceux qui avaient survécu à la réparation chirurgicale (gamme 4 à 28 jours). ***Conclusion:*** Pour les pays moins développés, une approche non-opératoire est préconisée dans la gestion de l'omphalocelle parce que la fermeture chirurgicale d'urgence entraîne une mortalité très élevée. ***Mots-clés:*** Exomphalos, gestion, résultat, pays en voie de développement.

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