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Experience with the Management of Exomphalos and Gastroschisis in Ibadan

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Summary

Adeyokunnu AA and Akingbehin NA. Experience with the Management of Exomphalos and Gastroschisis in Ibadan. *Nigerian Journal of Paediatrics* **1981; 8: 45.** Ninety-eight cases of exomphalos and gastroschisis were managed in the Department of Paediatrics, University College Hospital, Ibadan, over a period of eight years (1970–1978). Fifty-two (53%) of the cases were given non-operative while forty-six (47%) had surgical management. Mortality was 13% in those treated non-operatively and 59% in those treated surgically. The advantages and disadvantages of both forms of management are discussed and factors contributing to prognosis analysed. Operative management seems ideal when exomphalos occurs in an infant who is full term and of good weight, when the defect is minor and the sac is healthy, and when the scout films of the chest and abdomen reveal no significant abnormality, provided a good team of anaesthetists and surgeons is available for expedient surgical correction. For the fragile premature infant or any infant with major defect and infected defects, the use of non-operative technique under antibiotic cover as in-patients seems to offer the best course of action.

Introduction

EXOMPHALOS is herniation of some intra-abdominal contents through an open umbilical ring. The resulting protrusion is covered by a translucent, avascular membrane which consists of a fusion of peritoneum, amniotic membrane, and Wharton's jelly. Exomphalos differs from umbilical hernia in that it has no skin covering except at its base. The sac of an exomphalos may rupture before or soon after birth and he condition may therefore be confused with gasttroschisis. In gastroschisis, evisceration of the abdominal contents occurs through a full thickness defect of the abdominal wall at a point other than the umbilicus, and the insertion of the umbilical cord into the abdomen remains in the normal position.[1]

Exomphalos is not a common anomaly. Various reports[2 6] put the incidence at between 1 in 5,000 and 1 in 10,000 births. In Nigeria, an incidence of about 1 in 1,400 births has been reported from one hospital.[7]

The embryologic basis of exomphalos has been the subject of intense research and reviews.[8 12] The most widely accepted view is that a small exomphalos is due to failure of the umbilical ring to contract thereby permitting a herniation into

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the umbilical cord while large exomphaloses are regarded as morphogenic failures of the four folds of the body of the embryo to converge properly.[12 13] The reasons for the failures that produce exomphaloses are unknown. There have been isolated reports of occurrence in siblings and twins, and chromosomal abnormalities have also been found in some cases[14 16] but on the whole, hereditary factors do not seem to play any key role in the causation of the condition. On the other hand, the frequent association of exomphalos with a wide range of other malformations including those of the jaws, tongue and gastrointestinal tract,[5] and genitourinary tract,[4] suggest a general interference with embryonic development at an early stage of intrauterine life.

The best method for the management of exomphalos remains controversial. In the past, the mode of treatment for all cases was surgical correction, but this was accompanied by a high mortality rate.[18 19] More recently, it has been shown that a significant reduction in the mortality rate could be achieved with non-operative method in cases of exomphalos that have not ruptured.[5 20 21] The present communication reports our experience with the management of 98 cases (96 exomphalos and 2 gastroschisis) seen over an 8-year period in the Children's Department, University College Hospital (UCH), Ibadan.

Patients and Methods

All the children in whom the diagnosis of exomphalos or gastroschisis were made on clinical grounds between 1970 and 1978 were studied. The age, sex, weight and maturity of each patient were recorded, and the interval between birth and the time of reporting in the hospital noted. The size of the defect, associated malformations as well as the results of radiological, bacteriological and biochemical investigations carried out on any of the patients, were noted. The mode and the outcome of management in each case were also studied.

The anomalies were divided on the basis of their sizes into three categories namely: minor, major and gastroschisis. Irrespective of whether the sac was intact, infected or ruptured, an exomphalos was classified as minor if the diameter of the fascial defect was less than 5cm (Fig 1).

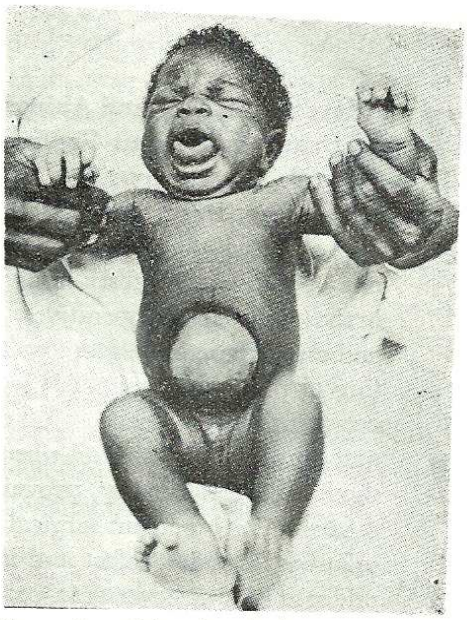

Fig. 1. Exomphalos minor, the diameter of the fascial defect is less than 5cm

Where the diameter of the fascial defect was greater than 5cm, the exomphalos was classified as major (Fig. 2), and cases in which a large portion of the gut had eviscerated were classified as gastroschisis.

Management was either surgical or conservative (non-surgical). The non-surgical management involved painting of the defect with aqueous solution of 2% mercurochrome. Our choice of the method of management was determined by the size and state of the defect, the state and maturity of the patient, and the presence and severity of associated malformations. Fifty-two of the 98 cases were treated conservatively while 46 were treated surgically.

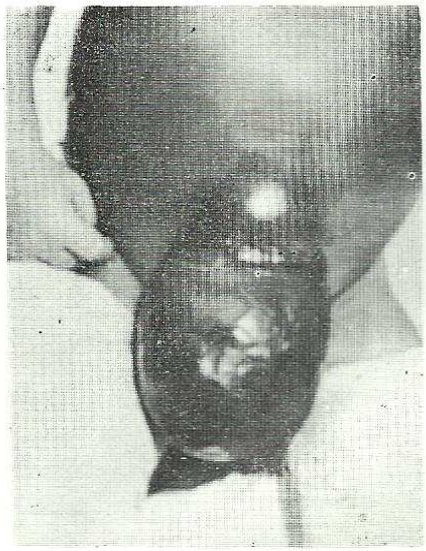

Fig. 2 Exomphalos major with diameter of fascial defect greater than 5cm. Both legs in plaster slabs to restrain the thighs rubbing on the defect.

Results

Sex, Birthweight and Age at Presentation

The 98 patients consisted of 63 males and 35 females (male to female ratio of 1.8:1). The birthweights of the infants are summarized in Table I. Thirty-six (37%) were premature by weight, their birthweights varying between 1.0 and 2.5 kg. Forty-eight (49%) weighed between 2.6 and 3.5 kg; and 14 others (14%) weighed 3.6kg and above.In comparison with the average birthweight of Nigerian babies[22 23] this last group of babies were significantly big.

TABLE I

Birthweights and Sex Distribution of Patients

Wt. (kg.)	*No. of Cases*		*Total*	*Percentage of Total*
	Male	*Female*		
1.0–2.5	23	13	36	37
2.6–3.5	31	17	48	49
3.6 and above	9	5	14	14
Total	63	35	98	100

The interval between birth and presentation was within 24 hours of delivery in 37 cases, between one and three days in 39 cases, between three and seven days in 18 cases and over one week in 4 cases.

Types of Defect and Associated Anomalies

A minor intact exomphalos was present in 24 (24%) and a major defect in 72 (73.5%) cases. The sac had ruptured in 15 of the major cases. Six infants were intially diagnosed clinically as gastroschises but only 2 were confirmed to be so at operation, the remaining 4 being examples of ruptured exomphalos major. Fifty-seven patients (58%) had associated anomalies (Table II); these were present in all organ systems but were commonest in the gastrointestinal tract.

Laboratory and Radiological Findings

Bacteriological cultures of exomphalos swabs and of blood were performed in 41 patients. These included all the 22 cases reporting after 72 hours of delivery, and 19 of the 39 cases who sought medical attention between 24 and 72 hours post-delivery. Positive cultures were obtained in 24 cases and the isolated organisms included *E. Coli* (9 cases; 7 from swabs and 2 from blood), *Pseudomonas* (7 cases; all from swabs), *Klebsiella* (5 cases; 3 from swabs and 2 from blood), *Proteus* (2 cases; both from swabs) and *Staph. aureus* (1 case; from blood).

Blood sugar was estimated in 27 cases including the 14 babies who weighed more than 3.6kg on admission. Blood sugar levels of less than 1.9 mmol/l (35 mg%) were obtained in 13 cases, all of whom also had somatic gigantism and macroglossia which are classic features of the Beckwith-Wiedemann syndrome, (Fig. 3). In 6 cases, the blood sugar level was less than 1.1mmol/l (20mg%) and this necessitated vigorous treatment for hypoglycaemia.

Fifty-six cases had radiographs of the abdomen and chest, and 32 of these showed evidence of either pneumonitis or consolidation of one or

both lungs. Fluid levels indicating intestinal obstruction occurred in 8 cases, while situs inversus (Fig. 4), absence of sternum, and hemivertebrae occurred in one case each.

TABLE II

Associated Malformations in 57 Cases of Exomphalos/Gastroschisis

Malformation	*No. of Cases*
FACE and TONGUE	
Macroglossia	13
Cleft lip and palate	5
GASTROINTESTINAL TRACT	
Malrotation of the gut	16
Meckel's diverticulum	6
Ileal atresia	3
Jejunal atresia	2
Persistent omphalo-mesenteric duct	1
Imperforate anus	7
Biliary atresia	2
GENITOURINARY SYSTEM	
Ectopic kidney	3
Duplex kidney	1
Bifid ureter	1
Ectopia vesicae	3
LIMBS	
Talipes equinovarus	8
CARDIOVASCULAR	8
CENTRAL NERVOUS SYSTEM	
Cataract	1
Microcephaly	1
Hydrocephalus	3
Meningomyelocele	1
VERTEBRAL	1
ABSENT STERNUM	1
CHROMOSOMAL DEFECTS	
Down's Syndrome	3
Trisomy 'E'	2
Total	92

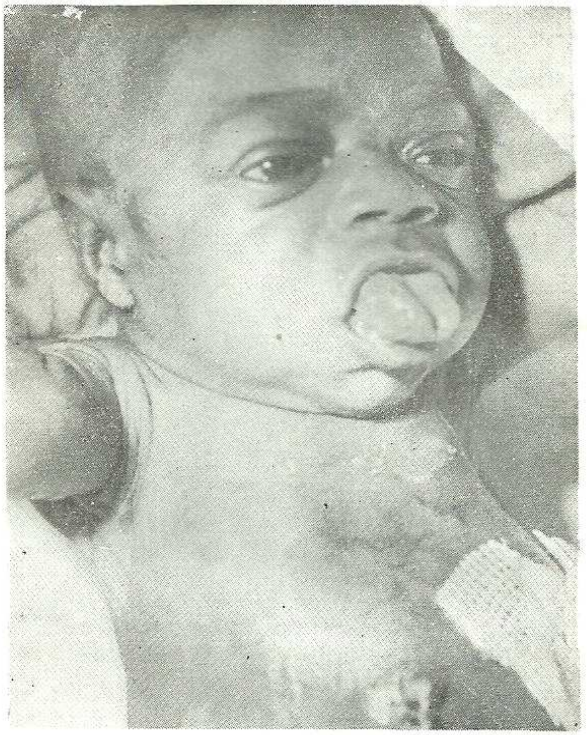

Fig. 3 Female infant with repaired exomphalos, macroglossia somatic gigantism and hypoglycaemia (example of Weidemnann-Beckwith Syndrome)

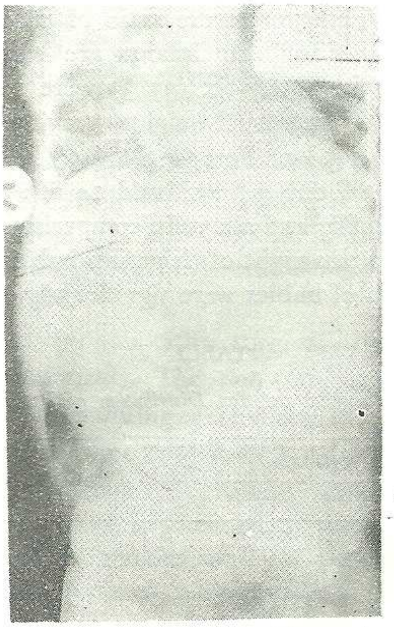

Fig 4 X-ray of chest and abdomen of a patient with exomphalos showing situs inversus and right-sided pneumonitis

Management and Outcome

Thirty-three out of the 57 cases with intact exomphalos major received non-operative treatment, and five of these died, (a mortality rate of 15%). The remaining 24 cases underwent surgery and 11 of these survived while 13 died (mortality rate of 54%). Among the 15 cases with ruptured sacs, two were treated conservatively because of gross infection and one of these survived and the other died. The remaining 13 cases underwent surgery and 11 of these died (a mortality of 84.6%). Seventeen of the 24 cases of exomphalos minor were operated upon and 2 died, a mortality of 28.5%. The two cases of gastroschisis were managed surgically, one of them died while the other survived.

The mortality in relation to the age and weight of the patients at presentation are summarized in Tables III and IV.

TABLE III

Mortality in Relation to Birthweights in 98 Cases of Exomphalos/ Gastroschisis

Wt. (kg.)	*No. of Cases*	*No. Alive*	*% of Cases*	*No. Dead*	*% of Cases*
1.0–2.5	36	19	52.8	17	47.2
2.6–3.5	48	33	68.8	15	31.2
3.6 and above	14	12	85.7	2	14.3
Total	98	64	65.3	34	34.7

TABLE IV

Mortality in Relation to Length of Delay before Hospitalization

	Length of Delay	*No. of Cases*	*No. Dead*	*% Mortality*
(*a*)	Less than 1 day	37	9	24.3
(*b*)	1 – 3 days	39	15	38.5
(*c*)	3 – 7 days	18	8	44.4
(*d*)	Over 1 week	4	2	50.0

Discussion

The present study showing a preponderance of males over females, a low birth weight in 37%, and exomphalos major in 73.5% of the cases, compares favourably with the findings of Jacho,[2] Aitken,[15] and Firor.[25] The presence of associated anomalies in 58% of our cases also compares with the incidence of 38% recorded by Hutchin,[1] and 44% reported by Smith and Leix.[26] This high incidence of associated anomalies is not surprising because all the organ systems involved, namely: the heart, the central nervous system, the renal tract and the gastrointestinal tract share the same embryogenesis.

The best method of management of exomphalos is controversial. Operative management enables the surgeon to explore the abdominal contents and if need be, correct any associated gastrointestinal tract anomalies. Compared with non-operative management, the period of hospitalization for surgical management is also short; thus one to four weeks was the average length of hospitalization for our surgically managed patients whereas the conservatively managed cases required eight to twelve weeks of hospitalization. Surgical correction was therefore practised in all cases until it was realized that non-operative management of exomphalos major was associated with reduced mortality.[4 19]

The conservative method of management of exomphalos was reportedly introduced in 1957 by Grob.[27] The method involves the painting of the exomphalos sac with 2% aqueous solution of mercurochrome. This dries the sac to a thick eschar under which granulation tissues form, followed by gradual epithelization and cicatrix formation. Good results have followed this regimen[4 19 20 28] but the disadvantages include, prolonged hospitalization and the attendant demand on the nursing services, the occasional delays in diagnosing intestinal anomalies, the risk of peritonitis during the early phase of resolution of the sac, and the uncommon but distinct possibility of mercury poisoning from absorption of mercurochrome.

The mortality rates from operative and non-operative methods of management in the present study were fifty-nine and thirteen % respectively. The disadvantages of operative management were most glaring in exomphalos major. Sixty-five % of patients with exomphalos major who underwent surgery died; whereas only 17 % of those managed conservatively, died. Surgical management of exomphalos major presents considerable technical problems. The small abdominal cavity may not be large enough to contain all the eviscerated structures, and therefore, reduction is often only accomplished under tension. This causes increased intra-abdominal pressure and respiratory embarrassment and predisposes to other complications such as intestinal kinking and obstruction, pressure necrosis of the intestine, and compression the inferior vena cava with the attendant reduction in the venous return to the heart. Surgeons of are aware of these complications and therefore do not always attempt a primary closure of big defects but sometimes resort to manipulative techniques such as multi-stage repair of the defect, and the use of Silastic Teflon Sheath. Massive adhesion formations however constitute a potential hazard of these alternative operative procedures.

Survival following surgery appears also to be partly related to the quality of available surgical services. For example, among the last 29 cases in the present series, only one death occurred out of the 13 cases surgically managed by a paediatric surgeon whereas 4 out of 5 babies in the same group who had their surgical correction performed by general surgeons, died.

The overall mortality rate of 34·7% in the present series compares with an average mortality rate of 50% in eleven centres reviewed by Hutchin.[1] Contributory factors to mortality in all series have included prematurity, associated congenital malformations, the size of the sac, and rupture of the exomphalos sac. Bacterial infections and delay in the institution of management constitute additional adverse factors in the present series. These two problems did not feature prominently in the series from developed countries, and are causally related to poor environmental sanitation and scarcity of the appropriate medical facilities prevailing in underdeveloped countries.

Encouraging as the non-operative management is, the length and cost of hospitalization make it unsuitable for poor countries. In a recent paper, Olowe[21] suggested that this major disadvantage of conservative management might be overcome if patients are treated on outpatient basis. In the light of this suggestion, we ventured outpatient management in some of our patients after 3 weeks of hospitalization, but were soon obliged to readmit them for further periods of 8 to 12 weeks each, because of infections of the exomphaloses at home. Outpatient management can therefore not be regarded as the best way for reducing cost and alleviating the disadvantages of prolonged hospital stay in these patients.

From the foregoing, it would seem that operative management of exomphalos is ideal in an infant who is full term and of good weight, whose defect is minor and the sac is healthy, and in whom the scout films of the chest and abdomen reveal no significant abnormality, provided a good team of anaesthetist and surgeon is available for expedient surgical correction. For the fragile premature infant or any infant with a major defect and those whose defects are infected, the use of non-operative technique under antibiotic cover as inpatients seem to offer the best course of action.

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