Challenges in the Management of Omphalocele at University Teaching Hospital, Zambia.

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Introduction

Omphalocele is the herniation of abdominal viscera at the base of the umbilical cord. The visceral is covered externally by the amnion, being at times separated by Whartons Jelly. The Defect is covered by membranous sac with the sizes ranging from 2 to 12cm. It has an incidence of 1:6000 live births. Associated malformation ranges from 30 to 88% Nicolaides 1992.

The most important point in our environment is decision making on a patient with Omphalocele; the decision pertaining to whether to operate or not to operate. If the decision is made, the surgeon has to know that parental feeds are not available in our Institution and a patient has to start early oral feeds if it has to survive.

Incidences of mortality among these patients seem to depend on the decision taken by the treating Paediatric Surgeons in view of non-available of parental feeds and prosthesis. Availability of prosthesis gives a Surgeon confidence to carryout stage operation, while parental feed allow a patient to be kept alive without increasing intra-abdominal space in a patient with already increased intra-abdominal pressure from eviscerated abdomen contents.

Neonatal and Paediatric surgical wing of Surgical Department was opened in 1985 as a referral centre. In the Past 20 years (1985 to 2005), a total number of 360,000 patients attended this specialized hospital and consisted of neonates and children from few hours after birth to about 15 years of age. Of these, 210 (0.06%) were treated for omphalocele. The patients came from Provincial, District and Missionary Hospitals of Zambia. Flying Doctors service was used in collecting these patients from various health centres.

This paper describes our experience in the management of this congenital malformation.

Patients and Methods

Management of Neonates with omphalocele was divided into two groups:

1. Patients with intact omphalocele and
2. Patient with ruptured omphalocele.

Investigations done included plain abdominal x-rays and ultrasound.

Intact omphalocele were treated in two ways.

i) Conservative treatment by exposure methods, using antiseptic or alcohol, or merthiolate,
ii) Small omphaloceles usually with small sac which is either intact or ruptured and usually containing intestines were treated by Primary repair.

Omphaloceles with Ruptured Sac: In cases where there was enough skin available, primary closure was done under general anaesthesia. When successful the second repair was deferred until the infant was 6 months to one year of age at which ventral herniorrhaphy could be performed.

Results

A total of 210 patients with omphalocele were treated in Neonatal and Paediatric Surgical Units of the University of Zambia. Their ages ranged from a few hours after birth to 72 hours. Patients from labour ward of the University Teaching Hospital were seen within as soon as possible following birth, while those from referred from Provincial, District and Missionary Hospitals were brought in by Flying Doctor's Services and were seen between 24 and 72 hours depending on the distance from the Neonatal and Paediatric Surgical Centre.

Of the patients, 106 patients (50.5%) were males while 104 (49.5%) were females giving a ratio of almost 1:1 between male and female in this study. On admission there were 59 patients (28.1%) who had intact membrane while 20 patients (9.5%) had ruptured membrane. Table 1 shows the clinical classification of all the patients seen with omphalocele.

Large Omphaloceles with a diameter above 8cm posed difficulties in their management in our institution which is associated with lack of

parental feeds and prosthesis. Huge but intact omphalocele were treated conservatively with skin grafts. The result ended with a big abdominal hernia. The Pendulum hernia was kept in cosett. Mortality was Nil. Small

Table 1. Clinical Varieties of Omphalocele

Type I							
Omphalocele with a sac intact	**No.**	**%**	**M**	**%**	**F**	**%**	**Treatment**
Containing only intestines	95	45.3	49	23.3	46	21.9	Conservatively
Containing intestines and other intra abdominal strictures	20	9.5	12	5.7	8	3.8	Conservatively
TOTAL	**115**	**54.8**	**61**	**29.0**	**54**	**25.7**	
Type II							
Ruptured Omphalocele Sac	**No.**	**%**	**M**	**%**	**F**	**%**	**Treatment**
Ruptured of the sac during delivery	15	7.1	9	4.3	6	2.8	Primary repair
Prenatal rupture of the sac	10	4.8	6	2.8	4	1.9	Flaps
TOTAL	**25**	**11.9**	**15**	**7.1**	**10**	**4.7**	**25**
Type III							
Omphalocele Associated with major defects.	**No.**	**%**	**M**	**%**	**F**	**%**	**Treatment**
Ectopic Cordis	2	0.95	2	0.95	0	-	One repair No operation
Diaphragmatic hernia	11	5.2	5	2.4	6	2.9	All repaired
Exstrophy of bladder	45	21.4	23	10.95	22	10.5	Repaired
or Cloaca	12	5.7			12	5.7	Repaired
TOTAL	70	33.3	30 +	14.3	40	19.1	70
Type IV							
Gastroschisis	**No.**			**Dead**			**Mortality**
Conservative treatment	95			3			1.4%
Primary suturing	26			5			2.3%
Skin flaps	9			4			1.9%
Inoperable	70			70			33.3%

Huge ruptured omphalocele with diameter above 8cm posed a major problem. Operation was attempted in patients with minor congenital abnormalities such as cleft lip. The treatment consisted of full thickness abdominal flaps which allowed primary suturing.

The raw areas in the flanks were later covered with skin graft. Mortality in this group was 60%. Patients with big and ruptured omphalocele

associated with severe malformation such as cardiac, spinal bifida, and urinary tract, were left to the nature to take its course**.**

Discussion

Omphalocele (Exomphalos) is a defect of the abdominal wall at the umbilicus with herniation of abdominal contents. There may be a sac or not. The incidence here in Zambia is not known but the world incidence is about 1 in 6000 live births[25,38]. Credit has been given to Ambrose Pare' for the first description and to Hey and Hamitton for the first successful closure of omphalocele[19,20].

The problems encountered in closing a large omphalocele in this study led to staged procedures with wide skin mobilization to cover the defect[16,33,55]. Modern Institutions use temporary prosthetic materials to accomplish closure at one sitting[39]. Due to these technical advances and associated with better understanding of preoperative and postoperative care including transport, Anaesthesia and improved current Nursing Management and Nutritional support. Omphalocele has passed from being pathological curiosities to entities with rapidly improving survival rates[2,5,24,28-30,35,43,44,51]. More importantly even in our hands, these infants develop into normal citizens.

There is some controversy about the origin of omphalocele. Several authors think that the basic defect is a failure of anterior extension of the lateral abdominal folds which eventually become the lateral abdominal walls. Because of the arrest in this extension, there is a continuous communication between the true abdomen and yolk sac through the umbilicus[14,19].

Intestines and other abdomen organs would occupy both spaces, depending on the size of the defect[9,13,22]. Others[14] feel that the fault lies in the arrest of the normal migration of the elongating intestines from the yolk sac into the true abdominal cavity. In either case, the arrest or teratogenic insult occurs at about the tenth week of gastation. Omphalocele are sometimes associated with Ectopia Cordis, Pentalogy of Cantrell, and bladder extrophy[10,46]. Arrested development of the Cephalic fold as well as the lateral abdominal folds could explain ectopia cordis and other thoracic defects. Similarly, arrest of the Caudal and lateral folds could explain the association with bladder exstrophy[22,24].

Diagnosis of omphalocele is usually very obvious. In omphalocele, there is a defect involving the base of the umbilical cord which can be quite small. Small ones usually contain small portion of small intestines and large intestines where as the large ones may contain intestines and liver[26,27].

Rickham[38] noted 205 malformations in 83 patients with omphalocele of which the most common ones involving gastrointestine tract. Malformation of the intestine was present ion most cases. In decreasing frequency were Meckel's diverticulum, patent omphalo-mesenteric, intestinal atresia or stenosis, malformation with volvulus, intestinal duplication, meconium ileus and billiary atresia[40,45].

In this study, fifty-nine patients had genitourinary malformation. 28 patients had intra-thoracic malformations of which 10 were congenital heart defects. There were anomalies of jaw, tongue, tumours and haemangiomata, limb abnormalities, inguinal hernias, absent abdominal musculature and several cranial, ophthalmic and vertebral anomalies[47].

All Neonates with omphalocele went through detailed clinical examination in order to exclude associated congenital abnormalities. Inspection was of great importance. 20 Neonates (9.5%) were found with big tongues an indication of Beckwith's Syndrome (Macroglosia, omphalocele, and hypoglycaemia), early treatment was important to avoid any episode of hypoglycaemia with seizure since one attack can be sufficient to result in permanent brain damage. The prophylactic treatment consisted of intravenous administration of glucose solution which was started immediately. All infants with omphalocele and in the event of hypoglycaemia the blood sugar was regulated by titration of concentrated intravenous glucose[39,41,49].

Absence of Xiphoid suggested association of Pentalogy of Cantrell and cyanosis implied association of intracardiac malformation or diaphragmatic hernia[46].

The incidence of congenital malformation involving various parts of the body has increased. Hence the high number of omphalocele. This may be due to HIV/AIDS Pandemic affecting the Southern African Region. The second thought may be the introduction of Health Education, the third point may be the introduction of Health Education when prenatal assessment are non-existence. Lack of antenatal assessment allows high incidence of live births with malformations,

which had been detected, would have necessitated early legal abortion.

Treatment

In our hands omphaloceles were treated either by conservative method or by surgical procedure. In conservative method it was important to note that methiolate can result in absorption of sufficient mercury from omphalocele membrane and can cause mercury poisoning. It was therefore emphasized that it should only be applied not more than three times.

Conservative treatment was reserved for big omphalocele (4cm and above) which had intact sac. In case of surgical treatment some surgeons use local anaesthesia in order to observe the degree of respiratory compromise reduced by reduction of omphalocele, although this method sounds superior, our experience was with general anesthesia.

In this study, general anaesthesia was used followed by mechanical ventilation for several days until the abdominal muscles have released enough to permit weaning from the ventilation.

If the sack contained lever, this could compress on inferior vena cava and create obstruction. Release of the liver from the skin was often enough to prevent the acute vena caval angulation.

Prosthetic Reduction

Large omphalocele which precluded primary repair for which no sufficient skin was not available to permit primary skin closure. Silastic $Silo_3$ could be used to cover the defect. This is gradually reduced in the theatre under local anaesthesia, by the 14^{th} day the prosthetic can be removed and the muscles be closed. As the abdominal muscles gradually stretch to accommodate the abdominal volume in the same way in which the abdomen accommodates the enlarging uterus during pregnancy. In this study, prosthetic materials were difficult to come by. We used skin flaps which ended up in a large abdominal hernia.

Parental Alimentation

This is the most significant way of treating large omphalocele, mortality stands high in our environment since parental feedings were not available. All our patients got early feeds consisting of patient's mothers' milk, using nasal gastric tubes.

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