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Anaesthetic challenges in omphalocele repair at a tertiary hospital in Ibadan, Nigeria: A review of medical records from January 2008 through December 2017

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Abstract

Background: Omphalocele is the commonest congenital anterior abdominal wall defect. It is associated with herniation of abdominal contents and the challenges it pose to the surgeons, anaesthetists and neonatal intensivists are daunting especially in the developing country.

Methods: This was a retrospective study of the challenges encountered during the perioperative management of neonates who had repair of omphalocele over a 10-year period. Information retrieved were demographic data, perioperative complications, anaesthetic techniques and outcome. These were collected in a proforma and analyzed.
Results: There were 57 neonates comprising 34 (59.6%) males and 23 (40.4%) females with male: female of 1.5:1. They presented between 1st and 14th day of life with a mean age of 2.7±3.6 days and mean birth weight of 3.06 ± 0.54 kg. There were 33 (57.9%) major and 24 (42.1%) minor omphaloceles. About 77.2% of patients had American Society of Anaesthesiologists (ASA) Physical Status Classes 1 and 2 at the time of surgery, 15 (26.3%) children had associated comorbidities which included sepsis 10 (17.5%), anaemia 3 (5.3%) and jaundice 2 (3.5%). Associated congenital anomalies were noticed in 10.5% of cases. All neonates had general anaesthesia with relaxant technique. Intraoperative challenges encountered were difficult intubation 5 (8.8%), hypothermia 7 (12.3%), tachycardia 37(64.9%) and bradycardia 3 (5.3%). Postoperatively, 15 (26.3%) neonates needed mechanical ventilation and intensive care management, 43 (75.4%) neonates survived while 14 (24.6%) died.

Conclusions: General anaesthesia is the conventional method for omphalocele repair, it is safe but not without its challenges. These challenges should be anticipated, and facilities should be in place to manage them.

Keywords: anaesthesia, challenges, omphalocele, repair, Nigeria

Introduction

Omphalocele is the most commonly encountered congenital anterior abdominal wall defect. It is a defect in the midline with herniation of abdominal content through an extra embryonic part of the umbilical cord.

The aetiology is due to failure of the four embryonic folds to meet in the midline to form umbilical ring resulting in ventral abdominal wall defect of varying degree¹. Omphalocele can be minor or major. It is classified as omphalocele minor when the defect is less than 5 cm in diameter with the sac containing only bowel loops and major when the size of the defect is more than 5 cm with the covering sac containing liver, spleen, etc. or has associated major congenital anomalies²,³. However, omphalocele has been re-classified to include the ‘intermediate’ group when the defect is more than 5 cm in diameter with no associated major anomaly and bedside reduction can be achieved without compromise. Such omphaloceles can be closed primarily².

Its incidence in Nigeria is not known but the world incidence varies from 1:6000 live birth to 1:32000 when live birth and still born are considered⁴.
Omphalocele can occur as an isolated finding or in association with a number of congenital abnormalities like cardiac defect, genitourinary abnormalities and chromosomal abnormalities like Trisomy 13, 15, 18 & 21 and other syndromes like Beckwith Wiedemann syndrome. Surgical management of omphalocele includes primary closure of the defect or staged closure with silo. The anaesthetic management may be challenging and require anticipation of problems such as prematurity, low birth weight, hypothermia, sepsis, dehydration, presence of associated congenital anomalies and small abdominal cavity. Postoperatively, the abdominal cavity may be too small to accommodate the eviscerated organ when they are returned back into the abdomen with consequent respiratory failure.

Advances in anaesthesia and neonatology have greatly improved the survival of these neonates. Communication and cooperation between the surgeons, anaesthesiologists and neonatologists are of utmost importance to ensure the best possible outcome. The aim of this study was to outline the challenges encountered peri-operatively and outcome of surgical management of neonates during omphalocele repair in our institution.

Methods
This was a retrospective review of records of all patients with omphalocele repair at the main operating room of the University College Hospital, Ibadan over a ten - year period from January, 2008 to December 2017. Neonates with gastroschisis were excluded from the data. The operating room records were reviewed to identify all cases of omphalocele operated during the period. The case records of the patients identified were then reviewed. The data retrieved included age, gender, birth weight, size of the sac, associated congenital anomaly, American Society of Anaesthesiologists (ASA) physical status, induction technique and muscle relaxants used, type of surgery, intraoperative events and postoperative complications, length of hospital stay, admission to the Intensive Care Unit (ICU) and final outcome. Ethical approval for the study was obtained from our Institution Ethical Review Committee (UI/EC/18/0461). Data collection and analysis were conducted in accordance with the ethical standards of 1964 declaration of Helsinki and its later amendments.

Anaesthetic technique
The technique of anaesthesia for all neonates was general anaesthesia with endotracheal intubation, muscle relaxation and intermittent positive pressure ventilation. The cadre of anaesthesia providers were physicians,
consultants that were formally trained in paediatric anaesthesia and senior registrars that have spent at least four years in postgraduate training. General anaesthesia was induced with either intravenous ketamine or inhalational agent; halothane or sevoflurane and tracheal intubation were facilitated with suxamethonium. The sizes of endotracheal tubes for intubation were between 2.0 mm and 3.5 mm in internal diameter. All neonates were premedicated with atropine to reduce secretion and increase the heart rates. Controlled ventilation was used in all patients and anaesthesia was maintained with Isoflurane, 100% of Oxygen and Atracurium. Analgesia was with Fentanyl and Paracetamol. Drugs were given in dose per kilogram body weight. Monitoring included pulse oximetry, electrocardiograph, capnograph, temperature, urinary output and blood glucose. Postoperatively, after reversal of the neuromuscular blockade those with delayed recovery or respiratory distress were transferred to the general Intensive Care Unit (ICU) whereas those with full recovery with no distress were transferred to the recovery room.

Statistical analysis
Data were analyzed using Statistical Package for Social Sciences (SPSS) version 20.0, Chicago Illinois. Continuous data were presented as means ± standard deviation whereas categorical variables were presented as frequency and percentages. The level of significance was set at p value of < 0.05.

Results
A total number of 57 neonates with omphalocele had surgical repair during the study period comprising 34 (59.6%) males and 23 (40.4%) females with male: female of 1.5:1. The age at presentation ranged from 1 day to 14 days with a mean age of 2.70 ± 3.26 days. The mean birth weight was 3.06 ± 0.54 kg, 5 (8.8%) neonates had birth weight less than 3.0 kg. There were 33 (57.9%) major and 24 (42.1%) minor omphalocoeles at presentation. Four (7.0%) patients had American Society of Anesthesiologist’s (ASA) Physical Status Class 1, 40 (70.2%) had ASA Class 2, 11 (19.3%) had ASA Class 3 and 2 (3.5%) patients had Class 4. More than 90% of the mothers resided in the urban community and only two mothers had prenatal ultrasound diagnosis of omphalocele. (Table 1)

Ten (17.5%) neonates had ruptured omphalocele and sepsis was the commonest preoperative comorbidities in 10 (17.5%) neonates, anaemia in 3 (5.3%) and jaundice in 2 (3.5%) neonates. Other associated congenital anomalies were congenital heart disease, Beckwith Wiedemann syndrome and intestinal atresia in 2 (3.5%) neonates each. (Table 2)
Forty (70%) neonates had rapid sequence induction while 17 (30%) had inhalational induction. All but 5 (8.8%) patients had primary closure of omphalocele with or without closure of the facial. Of the 5 patients that did not have primary closure, a silo was constructed for them and these were the patients that had rupture of omphalocele at presentation.

Intraoperative complications recorded were difficult intubation in 5 (8.8%) neonates, hypothermia in 7 (12.3%), tachycardia in 37 (64.9%) neonates (heart rate >160 /mins) and bradycardia in 3 (5.3%) neonates (heart rate < 100/mins). (Table 3)

Post-operative complications included respiratory failure in 7 (12.3%) neonates, delayed recovery in 8 (14.0%), wound infection in 6 (10.5%), burst abdomen in 2 (3.5%) and persistent hypoglycemia in 1 (1.8%) neonate. (Table 4)

Ten neonates were admitted into the general ICU and mortality was recorded in 14 (16.9%) patients. The mortality included the 5 neonates that had ruptured omphalocele with silo construction, sepsis was observed to be the major cause of death in them. The length of hospital stay ranged from 7 to 30 days.

**Discussion**

Omphalocele is the commonest of the anterior abdominal wall defects encountered in surgical practice. There were 57 cases operated over a 10 - year period making approximately 6 cases per year, this is similar to an earlier report by Ihekwaba from the same center who reported 33 cases over a five - year period from 1973 to 1978. However, a higher figure was reported in Benin Nigeria with 96 cases over 10 years making the abnormality of public health concern. Surgical treatment is often not considered an emergency unless the covering sac is ruptured therefore; all neonates went through a detailed history, physical examination, laboratory and radiological investigations to rule out other congenital anomalies before anaesthesia.

Sepsis, the commonest (17.5%) preoperative complication observed on the patients is lower than 37.5% reported by Abdur-Rahaman et al., this may be due to unhealthy care of the intact sac which can easily be soiled by faeces.
and urine. Sepsis is believed to contribute significantly to increase morbidity and mortality in the management of omphalocele\textsuperscript{6}. The avascular sac is easily infected despite proper preoperative antibiotic therapy\textsuperscript{8}.

Children presenting for repair would have already been resuscitated preoperatively due to dehydration and electrolyte loss from exposed viscera as was done for our patients\textsuperscript{7}. Fluid and electrolytes deficit were corrected preoperatively and maintenance crystalloid fluid at 4ml/kg/hour was continued into the intra and postoperative periods.

The anaesthesized neonate is at risk of hypothermia. The finding of hypothermia in this series is similar to findings from previous reports\textsuperscript{7,10}. These patients have less insulator subcutaneous fat in addition to loss of heat from the exposed gut. Efforts were made to maintain normothermia in these babies by covering the sac with warm saline soaked gauze, transferring them with incubator to the operating room. The problem of hypothermia may be compounded by cold operating room environment and dry anaesthetic gases. Heat conservation strategies we adopted include placing the neonates on warm mattress and increasing the operating room temperature in addition to warming the intravenous fluid and wrapping up the neonates.

Hypoglycemia is a common problem in neonates due to so many factors which included sepsis and hypothermia but this could be worsened in a patient with associated Beckwith Wiedemann syndrome preoperatively\textsuperscript{7}, hence blood glucose was monitored in all the neonates with omphalocele studied in addition to standard monitoring in anaesthesia. The two neonates with Beckwith Wiedemann syndrome had hypoglycaemia which was corrected. Glucose containing fluids were commenced preoperatively and continued into the post-operative period.

General anaesthesia with endotracheal intubation and muscle relaxant is the usual anaesthetic technique of choice during surgical repair of omphalocele in our setting. However, general anaesthesia with regional techniques like caudal and spinal block\textsuperscript{11} and combined spinal epidural\textsuperscript{12} have been reported by some authors.

Intraoperative opioids are generally avoided in neonates due to the fear of opioid induced respiratory depression with increased need for postoperative ventilation\textsuperscript{13}. Remifentanil which is not always available can be used as it is preferred to more commonly available fentanyl because it is an ultra-short acting drug and is well tolerated by neonates.
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Non-depolarizing muscle relaxants are better avoided in neonates because they are more sensitive to its effects due to reduced release of acetylcholine from immature motor neurons. However, muscle relaxation is necessary for optimum closure of the abdominal defect and intubation is necessary to protect the airway against the risk of aspiration. Cis-atracurium is preferred to atracurium (which is more available in this environment) as cis-atracurium produces a more predictable duration of action.

Rapid sequence induction was employed in some neonates in this series when they were at risk of aspiration at induction of anaesthesia as a result of full stomach while some neonates with minor omphalocoeles had inhalational induction with halothane or sevoflurane especially when the patient was stable and fit. Intravenous induction versus gas induction remains controversial in neonates with omphalocele.

Difficult intubation as observed in some neonates is a common challenge, it occurs in 10% of patients with craniofacial anomalies and incidence increases in those with associated syndrome like Beckwith Wiedemann syndrome. Expert airway management is however essential for safe and effective administration of general anaesthesia. The anatomical airway peculiarities of the neonates including long and floppy epiglottis with relatively large tongue add to the incidence of difficult intubation.

Surgical treatment modalities of omphalocele include primary closure of the defect and staged repair using silo. The postoperative management is however a challenge as primary closure is often complicated with respiratory distress as a result of increased intra-abdominal pressure making extubation difficult. Measuring the respiratory airway pressure may be used as an indirect method of measuring intra-abdominal pressure after primary closure of the defect and surgeons were advised to reduce the abdominal pressure or staged the repair. Another indirect method of measuring the intra-abdominal pressure is the measurement of the intra-vesical pressure and if the pressure is less than 20 mm HgO the patient can be extubated.

Patients with delayed recovery and respiratory distress after surgery are usually managed in ICU for mechanical ventilation and postoperative monitoring to ensure good outcome. The lack of well-equipped standard Neonatal Intensive Care Unit (NICU) poses a great challenge to successful management of surgical neonate and is one of the factors that have contributed to the high morbidity and mortality in neonatal surgery experienced in Nigeria and other low and middle-income countries.
The practice of neonatal anaesthesia in our centre has improved over the last decade from postoperative manual ventilation in the first 1 to 2 years of this study to mechanical ventilation, provision of modern equipment, safe drugs administration and structured training of physician paediatric anaesthetists. The latter years witnessed a change from the use of manual ventilation to mechanical ventilation and this has impacted on the outcome of procedures including paediatric surgical cases. As with other major centres in the country\(^\text{20}\), the major challenge in our centre is the lack of a dedicated NICU and neonatal ventilator as these neonates are still nursed in the general ICU where adult equipment are used for them. Thus there is marginal improvement generally in morbidity and mortality in neonatal surgery including repair of omphalocele.

The survival of neonates with omphalocele in the developed countries has been attributed to the availability of NICU, parenteral nutrition and various requisite equipment needed to support the post-operative intensive care of these neonates.

The observed mortality rate of 16.9% is lower than the rate of 22.9% and 32.4% reported by Osifo DO et al\(^\text{9}\) and Abdur-Rahman et al\(^\text{10}\) in Nigeria respectively. It is also significantly lower than the reported rate of 64% by an earlier study carried out about four decades ago in our centre\(^\text{8}\).

Respiratory insufficiency was observed to be largely responsible for the mortality recorded in this study. The use of silastic silo for staged repair of the omphalocele may help to reduce the challenges of post-operative respiratory insufficiency.

**Conclusions**

Omphalocele repair is a challenge to both the surgeons and the anaesthetists in the developing country. These neonates may present with sepsis, hypothermia, fluid and electrolyte imbalance and hypoglycemia these had to be corrected before the surgery. General anaesthesia is a safe and effective technique for surgery but problems of difficult intubation, drug metabolism and postoperative respiratory distress should be anticipated. The addition of neonatal ventilator, neonatal intensive care and parental nutrition to the present protocol should reduce morbidity and mortality in this high-risk group in the future.
References


### Table 1: Demographic characteristics of the neonates

<table>
<thead>
<tr>
<th>Variables</th>
<th>Number =57</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender</td>
<td>Male</td>
<td>34</td>
</tr>
<tr>
<td></td>
<td>Female</td>
<td>23</td>
</tr>
<tr>
<td>Mean Age ± SD (days)</td>
<td>2.7±3.26</td>
<td></td>
</tr>
<tr>
<td>Mean birth weight ± SD (kg)</td>
<td>3.06±0.54</td>
<td></td>
</tr>
<tr>
<td>Types of Omphalocele</td>
<td>Major</td>
<td>33</td>
</tr>
<tr>
<td></td>
<td>Minor</td>
<td>24</td>
</tr>
<tr>
<td>ASA 1</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>ASA 2</td>
<td>40</td>
<td></td>
</tr>
<tr>
<td>ASA 3</td>
<td>11</td>
<td></td>
</tr>
<tr>
<td>ASA 4</td>
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<td></td>
</tr>
</tbody>
</table>

ASA – American Society of Anaesthesiologist’s classification

### Table 2: Preoperative associated comorbidities

<table>
<thead>
<tr>
<th>Variables</th>
<th>Number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital heart disease</td>
<td>2 (3.5)</td>
</tr>
<tr>
<td>Beckwith welderman syndrome</td>
<td>2 (3.5)</td>
</tr>
<tr>
<td>Intestinal atresia</td>
<td>2 (3.5)</td>
</tr>
<tr>
<td>Sepsis</td>
<td>10 (17.5)</td>
</tr>
<tr>
<td>Anaemia</td>
<td>3 (5.3)</td>
</tr>
<tr>
<td>Jaundice</td>
<td>2 (3.5)</td>
</tr>
</tbody>
</table>

### Table 3: Postoperative complications

<table>
<thead>
<tr>
<th>Variables</th>
<th>Number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Respiratory distress</td>
<td>6 (12.7)</td>
</tr>
<tr>
<td>Wound infection</td>
<td>6 (12.7)</td>
</tr>
<tr>
<td>Burst abdomen</td>
<td>2 (3.5)</td>
</tr>
<tr>
<td>Persistent hypoglycemia</td>
<td>1 (2.1)</td>
</tr>
<tr>
<td>Mortality</td>
<td>14 (16.9)</td>
</tr>
</tbody>
</table>