Exomphalos Defects : A Review of 15 Cases

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Abstract

Background : The outcome after repair of exomphalos defects has improved significantly with refinements in surgical techniques, multidisciplinary management and monitoring of intra-abdominal pressures.

Methods : A retrospective study of medical records of 15 cases with omphalocele was done.

Results : Antenatal diagnosis was available in six cases. There were eight females and seven males with a mean birth weight of 2.2 kg. Nine babies had associated anomalies. There were nine major (defect size > 5 cm) and six minor defects. Immediate closure in neonatal period was carried out in 12 cases. Urinary bladder pressure (UBP) was measured to assess intra-abdominal pressure in cases where primary closure was difficult. Primary closure was omitted in the event of intravesical pressures exceeding 20 mm Hg (~ 25 cms of water). Primary surgical closure was possible in five (56%) major cases. Two cases were subjected to silo repair followed by delayed primary closure whereas the other two required a Goretex mesh closure. Three minor defects could be repaired primarily whereas the remaining three were managed conservatively and closed at age of 9 to 12 months. There were no significant anaesthetic complications. Elective postoperative ventilation was required in one baby. There were three deaths at ages five, nine and ten months due to unrelated causes.

Conclusion : Satisfactory outcome is possible in cases with exomphalos defects with intra-operative intravesical pressure assessment forming a convenient method for excluding abdominal compartment syndrome.

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Key Words : Abdominal wall defects; Omphalocele; Intra-abdominal pressure; Intravesical pressure measurement.

Introduction

The abdominal wall defects (AWD) are a spectrum of defects that include omphalocele, gastroschisis and other conditions such as umbilical hernia, prune belly and bladder exstrophy. These defects can be diagnosed by ultrasound (USS) in the first trimester [1] and are characterized by protrusion of varying amounts of abdominal viscera.

Surgical treatment in the perinatal period has often been catastrophic due to the resulting abdominal compartment syndrome. In recent years, antenatal diagnosis, advances in neonatal anaesthetic techniques, monitoring of intraoperative intra abdominal pressures and the availability of Goretex dual meshes has improved the survival outcome in these cases. The aim of this paper is to present our experience of managing omphalocele with a brief review of literature.

Material and Methods

The medical records of omphalocele cases managed in our centre over a period of three years were reviewed. The information was grouped into three categories. The availability of antenatal diagnosis, clinical presentation, period of gestation, birth weight, presence of infection of sac, congenital anomalies and any biochemical abnormalities

were noted in the first category. The second category included requirement of preoperative resuscitation and antibiotics, technique of anaesthesia, extent of monitoring and haemodynamic instability. Details regarding, surgical closure whether primary or delayed and use of any objective methods to guide the surgical repair were also sought. The last category comprised of recordings of extubation related events such as laryngospasm, apneic episodes, hypothermia, hypoventilation and requirement of elective or emergency ventilation, status of urine output, oxygen saturation in lower limbs and mortality.

The anaesthetic technique consisted of induction with thiopentone and intubation with atracurium with cricoid pressure. Fentanyl was used for analgesia whereas inhalational agents were chosen from sevoflurane, halothane or isoflurane. Intra-operative monitoring consisted of pulse oximetry, non-invasive blood pressure (NIBP), electrocardiogram and nasopharyngeal temperature. Two pulse oximeter probes, one each on the upper and lower limb were placed while central venous pressure (CVP) was measured via a peripheral indwelling catheter. Postoperative analgesia was provided with aliquots of fentanyl, rectal paracetamol and caudal epidural in three cases.

The surgical management included primary closure, silo reduction with delayed closure with or without Gore-Tex dual mesh. The choice of surgical option was made after

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peroperative assessment of intra-abdominal pressure, clinical assessments and measurement of intravesical pressures with a water column manometer. In order to measure urinary bladder pressure (UBP), the bladder was catheterized with Foley's catheter and filled with a saline so that the bladder protrudes in the abdominal cavity. The urobag was then clamped and a needle attached to a pressure monitoring line was inserted in the Foley's catheter (Fig. 1). The other end of the line was held against a scale to measure the pressure in "centimeters of water" or a pressure transducer. Primary fascial sheath closure was omitted in the event of intravesical pressures exceeding 20 mm Hg (~ 25 cms of water). After closure, the neonates were shifted to neonatal intensive care (NICU) unit for further management.

Results

A total of 15 cases of omphalocele were managed at our centre and antenatal ultrasound diagnosis was available in six cases. There were eight females and seven males. Nine of them were born at term, while others were premature at 32 - 37 weeks of gestation. Birth weight varied between 1.5 - 2.9 kg (mean 2.2 kg). There were associated anomalies in 12 babies (Table 1). Meckel's diverticulum was the commonest anomaly. Chromosomal defects were present in two cases. There were nine major defects (defect size > 5 cm) and six minor. Three cases required preoperative volume resuscitation as they were referred cases with dehydration. Two babies had features of sepsis. One case had suffered birth asphyxia and two episodes of seizures. These infants were started on phenobarbitone in the pre-operative period. Antibiotics and intravenous fluids with calcium in appropriate amounts was given as a routine to all the babies. There were no significant biochemical or electrolyte abnormalities in any of the cases.

Twelve cases were taken up for closure within 12 - 36 hours of presentation. Amongst neonates with major defects, primary surgical closure was possible in five (56%) cases (Fig. 2). The bladder pressures in these case was less than 15 cm of water whereas the remaining four cases had pressures ranging from 30 - 34 cm of water and hence a decision had to be made against primary closure. Two cases with very large defects (>7cms) were subjected to delayed closure with initial use of a silo. Subsequently, both underwent delayed primary closure on Day 10 and 13 after placement of silo (Fig. 3). Two others required a Goretex dual mesh closure where defect size was 5 and 5.5 cms respectively and sufficient skin was available after undermining for primary skin closure. Three cases of minor defects could be repaired primarily whereas

the remaining three were managed conservatively and closed at age of 9 to 12 months. Urine output remained adequate in all the cases. Two cases had Beckwith-Weidemann syndrome (BWS) (Fig. 4).

There were no significant anaesthetic complications. Brief periods of haemodynamic instability during reduction of the viscera and liver in case of major AWDs (Fig. 5), were managed with additional infusion of crystalloid or colloid (haemaccel @ 10 ml/kg body weight). Single shot caudal epidural blocks with 0.2% bupivacaine @ 1ml/kg were given in three cases. Nasopharyngeal temperature dropped to 34.5°C in three cases, however in most it was normal. Elective post operative ventilation was required in one case with ventricular septal defect (VSD). There were no adverse events related to extubation except in one where extubation was delayed till late evening as the closure was felt to be tight and intravesical pressure was 25 cms of saline. Three babies with less than 2 kg body weight had recurrent episodes of apnea. Three cases in the age group of less than 10 months died due to unrelated causes.

Discussion

The estimated birth prevalence of omphalocele is 2.5 in 10,000 births in western countries [2]. In omphalocele, abdominal viscera are contained within a translucent sac consisting of amnion, Wharton jelly and peritoneum. In 50% of cases, the liver, spleen, and ovaries or testes accompany the extruded midgut. The exposed viscera covered with thin sac leads to complications like rupture of sac, sepsis, hypothermia besides fluid and electrolyte losses. At least 30-50% of babies have associated anomalies which are important prognosticators of the final outcome. Beckwith-Weidemann syndrome (BWS) is a common genetic overgrowth syndrome that is associated with visceromegaly, macroglossia (Fig.4), abdominal wall defects, pre- and postnatal overgrowth, and neonatal hypoglycaemia. We had two babies with BWS and both of them made an uneventful recovery.

Use of prenatal diagnostic techniques including USS and maternal serum alpha-fetoprotein measurements has led to detection of abdominal wall defects before birth. In the present series, though mothers of 12 babies underwent a level III antenatal USS, diagnosis of abdominal wall defects was possible only in six (40%) cases. Omphalocele and gastroschisis are associated with elevation of maternal serum alpha-fetoprotein [3]. This data was not available for our cases.

The presence of congenital anomalies, size abdominal wall defect, respiratory failure at birth in infants with omphalocele is a significant predictor of mortality [4]. Anaesthetic management is directed at minimising fluid and heat loss which is compounded by the problems of prematurity and associated anomalies [5]. Epidural analgesia has been suggested as the best method as it also reduces abdominal and vascular pressures and

Table 1
Associated anomalies

Anomaly	Number of patients
Meckel's diverticulum	03
Beckwith Weidmann syndrome	02
Chromosomal anomalies (Down's)	02
Bilateral undescended testis	02
Hypospadias	01
Unilateral cleft lip and palate	01
Ventricular septal defect	01

allows better respiratory function [6]. We used caudal epidural in three cases. The fluid requirements can be as high as 10 - 15ml/kg/hour in addition to boluses. In fact in our series, three cases received fluids at an average rate of 20 ml/kg/hour without any adverse events. However acute fall in blood pressure is often attributable to compression and kinking of inferior vena cava while handling liver in omphalocele major cases. Obstruction of the suprahepatic, subdiaphragmatic vena cava leading to the development of acute Budd-Chiari

Fig. 1 : Urinary bladder pressure measurement set-up

Fig. 2 : Omphalocele

Fig. 3 : Silo closure

syndrome has been reported following repair of a giant omphalocele [7].

The most important issue in the management of AWDs is the choice of closure methods without causing elevation in Intra Abdominal Pressure (IAP). There is evidence that staged repairs of major AWDs are associated with lower incidence of abdominal compartment syndromes and infections even though they are associated with longer hospital stay [8]. IAP (normal ~15 mm Hg) can rise during primary closure of abdominal wall leading to intra-abdominal hypertension and subsequent development of abdominal compartment syndrome (ACS) characterised by compromised venous return, hypotension, reduced organ perfusion, elevated intrathoracic pressures with decreased pulmonary compliance, anuria and intracranial hypertension [9]. Swartz et al [10], in a retrospective study concluded that the “degree of viscerο-abdominal disproportion” and the physiologic response of the newborn to closure must guide the surgeon in the choice of repair.

IAP can be measured directly with a needle in the peritoneal cavity. The indirect methods include measurement of UBP, intra-gastric pressure, airway pressure, central venous pressure, or cardiac index [9]. Certain clinical indications such as difficulty with

Fig. 4 : BWS with macroglossia

Fig. 5 : Large omphalocele with liver (marked with arrow)

ventilation after closure, lower limb oedema, difference in upper limb and lower limb blood pressure, failure of lower limb pulse oximetry recordings and dusky wound margins, inability to palpate the femoral pulses, cyanosis of the lower extremities, progressive oliguria may suggest changing to a staged approach. Yaster et al [11], demonstrated that increases in intra-gastric pressures beyond 20 mm Hg were frequently associated with reductions in venous return and cardiac index, requiring surgical decompression of the abdomen.

However UBP is the gold standard of IAP measurement [9]. Chin et al [12], demonstrated a higher rate of complications in seven newborns with UBP exceeding 20 mmHg following repair of AWDs. Lacey et al [13], found that UBP altered approach to patient management in 64% of cases. Beck et al [14], suggested that the UBP should be less than 20 mm Hg. Rizzo et al [15], reviewed 40 neonates with AWDs and demonstrated significantly less morbidity and mortality in those with real-time intravesical pressure measurements. We used UBP measurements particularly in major defects to decide the surgical approach, which was changed in four (44%) cases. Recently, strategies such as vacuum-assisted closure, active enlargement of the abdominal cavity, use of human acellular dermis, tissue expanders etc have been employed as an alternative to primary closure to avoid mechanical complications [16,17].

In the postoperative period, respiratory function and IAP should be monitored. Re-exploration in adults is indicated if UBP exceeds 35 mm Hg [9], but children may develop ACS at a IAPs of 16 mm Hg and clinical signs of ACS are sufficient to justify abdominal decompression [15,18].

To conclude, it is possible to achieve a satisfactory outcome in cases with AWDs after surgical treatment. The intra-operative intravesical pressure assessment forms a convenient evidence based method for excluding abdominal compartment syndrome and to decide upon the choice of closure and ventilatory management.

Conflicts of Interest

None identified

Intellectual Contribution of Authors

Study Concept : Col B Puri

Drafting & Manuscript Revision : Col B Puri, Lt Col DK Sreevastava

Statistical Analysis : Col B Puri, Lt Col DK Sreevastava

Technical Support : Col B Puri

Study Supervision : Col B Puri

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