ORIGINAL ARTICLE

Omphalocele and Hernia of Umbilical Cord: An Early Outcome Analysis from a Tertiary Care Hospital

Prameshwar Lal[1], Neeraj Tuteja[1], Dinesh Kumar Barolia[2], Vinita Chaturvedi[1], Rahul Gupta[1], Gurudatt Raipuria[1], Vikas Joshi[1], Arun Gupta[1] and Ajay Kumar[1]

[1,2]Department of Paediatric Surgery, [1]Sir Padampat Mother and Child health Institute, S.M.S. Medical College, Jaipur-302004, Rajasthan, India. [2]Jawaharlal Nehru Medical College, Ajmer-305001, Rajasthan, India.

Abstract:

Background: Omphalocele and hernia of umbilical cord (HUC) are frequent types of abdominal wall defect dealt by paediatric surgeon. We aimed to study the early outcome analysis of omphalocele and HUC in our tertiary care hospital. *Material and Methods:* A prospective observational study was carried out in our tertiary care teaching institute from 16 October 2018 to 15 April 2020. A detailed history, clinical examination, baseline blood investigations and radiological evaluation were performed in each case. Cases of omphalocele major were subjected to initial conservative management while all neonates with omphalocele minor and hernia of umbilical cord were managed surgically. *Results* Sixty-nine neonates with omphalocele major (17), minor (29) and HUC (23) were included in the study. Male to female ratio was 2.83: 1 in both omphalocele and HUC. Out of 17 patients of omphalocele major, 10 (58.82 %) were managed conservatively. Emergency surgery was performed in 4 patients (23.52%) while 3 patients (17.64%) died before any intervention. All patients of omphalocele minor and HUC were operated immediately except 1 patient (3.44%) of omphalocele minor who died before surgery. Mortality was 4 (23.52%), 8 (27.58%) and 4 (17.39 %) in omphalocele major, omphalocele minor and HUC respectively. *Conclusion:* There was male predominance in both of these defects. Early admission and timely management improve the prognosis. Primary anatomical repair is best in omphalocele minor and hernia of umbilical cord. In omphalocele major, primary anatomical repair is often not feasible, so "escharotic therapy" is a good alternative. We found that primary anatomical repair is better choice for omphalocele minor and conservative scar therapy is good for omphalocele major, until unless sac was ruptured.

Keywords: Abdominal wall defect, Hernia of umbilical cord, Omphalocele, Omphalocele major, Omphalocele minor, Ventral hernia.

Introduction:

Abdominal wall defects are a complex group of congenital abnormalities with a broad spectrum of manifestations. Abdominal wall defect includes mainly omphalocele, gastroschisis and hernia of umbilical cord (HUC) [1]. Omphalocele and hernia of umbilical cord are the common abdominal wall defects and are the focus of this study. Although both of these conditions affect the umbilical area, but their management and outcomes are quite different. The present study was undertaken to evaluate the early outcome analysis of omphalocele and HUC in our tertiary care teaching hospital.

Material and Methods:

This is a single centre, prospective study; conducted in the Department of Paediatric Surgery of a tertiary care teaching hospital of India, over a period of 18 months from October, 2018 to April, 2020. The follow up period was three months to one year. In our study we used povidine iodine 5% solution as escharotic agent for omphalocele major. All neonates presenting with omphalocele minor, omphalocele major, and HUC were included in the study. Cases of other abdominal wall defects like gastroschisis, bladder exstrophy, cloacal exstrophy and prune belly syndrome were excluded from this study.

A detailed history (ante-natal and post-natal), birth weight and mode of delivery were recorded. Detailed clinical examination and baseline investigations were

performed in each case. All neonates were assessed with regards to their age, gender, weight, size of defect, anal opening, and other associated anomalies. Vital signs, hydration status, visible bowel loops, faecal discharge were noted. Systemic examination of the cardiovascular system, respiratory system, and central nervous system was done.

All neonates were resuscitated and optimised; attention was given to keep the baby dry, prevent dehydration & hypothermia, prevent heat loss and maintain a warm environment. Serum glucose levels were checked in neonates. Orogastric decompression and bladder catheterisation was done. Intravenous access was obtained in upper extremities for fluids and broad-spectrum prophylactic antibiotics.

Surgical procedure for patient was decided according to size of defect, and content of the sac. Omphalocele was categorised as major if size of defect was more than 4 cm or sac contains part of liver. Neonates with defect size smaller than 4 cm were included as omphalocele minor. Anatomical closure is possible undue raising intra-abdominal pressure, than anatomical closure was done. If not then we form ventral hernia or silo repair. All surgery was done under general anaesthesia with endotracheal intubation. There was no detectable post-operative complication in this study.

Screening USG and 2D echo was not done in all neonates due to high patient loads at our centre. Only clinically unstable neonates or neonates having murmur were posted for 2D echo. USG abdomen was done only in patients who did not pass meconium.

Results:

There were 69 neonates with omphalocele major (17), minor (29) and HUC (23). Male to female ratio was 2.83:1 in both omphalocele and hernia of umbilical cord, suggesting male predominance in each type of defect [Table 1]. Average gestational age of neonate was 39.25±1.49 and 39.35±2.01 weeks in omphalocele and hernia of umbilical cord respectively. Average weight of neonates was 2.33±0.50, and 2.13±1.17 kg in omphalocele and hernia of umbilical cord respectively. Average age of mothers was 23.64±2.16, and 23.57±1.42 years in omphalocele and hernia of umbilical cord respectively. Average size of defect was 3.42±0.83cm and 1.57±.87 cm in omphalocele and hernia of umbilical cord respectively. Number of caesarean deliveries in omphalocele and HUC was 2 in each case. History of Polyhydramnios was present in 2 (4.34 %) cases of omphalocele and 1 (4.34 %) case of HUC in our study. Out of all 46 cases of omphalocele, one case (2.17%) presented with ruptured sac. Remaining all cases including HUC were admitted with intact sac.

Maximum number of neonates were admitted on first live day (3 to 24 hours) after birth. It was observed that 43 neonates (62.31 %) were admitted on day first, 16 neonates (23.18 %) on day second, 5 neonates (7.24 %) on day third, 1 neonate (1.44 %) on day fourth and rest 4 (5.79 %) neonates on day 5 or even later. In day first admitted neonates, survival rate was 83.72% (36) followed by 81.25% (13),60% (3),zero (nil) and 25 % (1) in neonates admitted on day 2,3,4,and 5 or later respectively [Table 2].

Contents in sac of all type of abdominal wall defect were identified either pre operatively or intra operatively [Table 3]. In 17 cases of omphalocele major [Figure 1], 10 were managed conservatively [Figure 2] and 3 patients died before any surgical intervention. Because of coarse and opaque sac, contents could not be differentiated in all these patients (76.47 %). Small bowel and large bowel loop were identified in two cases of omphalocele major. Remaining 2 cases of omphalocele showed loop of small bowel, large bowel and liver in sac.

There were 29 patients of omphalocele minor [Figure 3] in our study. Only small bowel loops were present in 15 neonates (51.72 %). In 10 patients sac contents were loops of small and large bowel. The sac of 3 omphalocele minor patients contained small bowel loops along with Meckel's diverticulum. One patient of omphalocele minor expired before examination of the sac contents.

In HUC [Figure 4], contents of sac in 18 neonates (78.26 %) were single loop of small intestine, loops of small and large intestine in 3 neonates (13.4%) and small intestine with Meckel's diverticulum in 2

neonates (8.69 %).

Out of 17 patients of omphalocele major, 10 (58.82 %) were treated conservatively [Table 4]. All 10 patients were managed successfully and discharged. In 4 patients, surgical intervention was contemplated. Anatomical repair was done, in 2 neonates. In one patient, contents were loops of small bowel, large bowel and liver; anatomical repair was not possible, so silo repair was done. In one patient admitted with ruptured sac, skin flap closure was performed to form ventral hernia. Two patients were admitted with poor general condition with septicaemia and died before any surgical intervention. Another patient with omphalocele major and "ectopia cordis" admitted with poor general condition died on the day of admission. Mortality for omphalocele major was 23.52 % (4 patients) in our study. We did not found the case of beckwith-wiedemann syndrome (BWS) or visceromegaly in my study.

Out of 29 neonates with omphalocele minor, one died before surgery and remaining 28 patients were operated. In 24 patients, primary repair of defect was done successfully. Two patients had Meckel's diverticulum; excision of ileal segment with diverticulum and ileo-ileal anastomosis with repair of defect was done. One patient presented terminal ileal atresia with proximal segment dilatation. Resection of ileo-cecal junction including ileal atresia and ileo-ascending anastomosis was done with primary repair of wall defect. Patent urachus was also present in one patient; ligation of urachus and repair of defect was done. Total mortality in omphalocele minor patients was 27.58 % (8 patients).

All 23 neonates with HUC [Figure 4] were operated. In 3 patients, minimal surgery with reduction of contents and ligation of cord was done. In 14 patients, we did primary repair with reduction of contents and umbilicoplasty [Figure 4]. Out of 23 patients, HUC was associated with perforated ileum in 2 and with ileal atresia in 2 patients. Resection of ileal segment and ileo-ileal anastomosis was done in these 4 patients. In two patients, small intestine with Meckel's diverticulum was found as content of the sac.

Overall mortality was 17.39% (4 patients) in this group [Table 4].

Associated renal and cardiac anomalies were not reported in this study due to overburden of patients in 2D echo and USG centre at our institution.

Figure 1: (A) Omphalocele major with intact sac at the time of admission; (B) initiation of escharotic therapy with povidone iodine solution (5%) application; (C) eschar formation over the omphalocele sac.

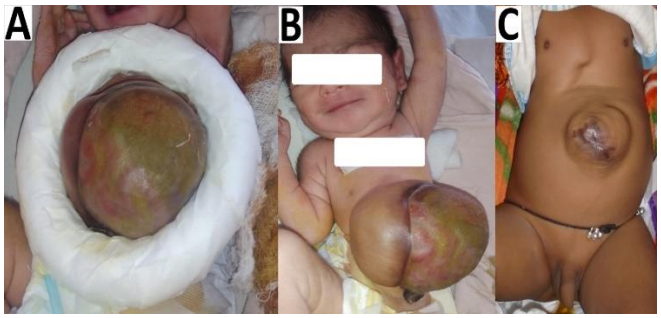

Figure 2: (A) Bandage ring supporting omphalocele major and helping escharotic therapy; (B) epithelialisation of omphalocele sac; (C) skin cover omphalocele.

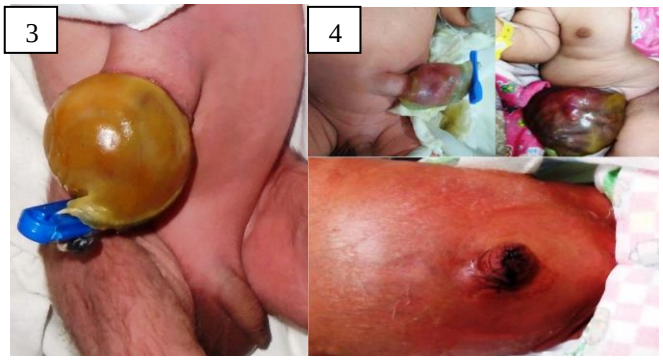

Figure 3: Omphalocele minor. Figure 4: Hernia of umbilical cord (above); and umbilicoplasty (below).

Table No. 1: Gender distribution of the patients

Type of defect	Male	Female
Omphalocele major n=17(24.73%)	11(15.94%)	6(8.69%)
Omphalocele minor n=29 (42.02%)	23(33.33%)	6(8.69%)
Hernia of umbilical cord n=23 (33.33%)	17(24.73%)	6(8.69%)
Total n = 69(100%)	51(73.9%)	18(26.08%)

Table No. 2: Age wise distribution of the patients (at the time of admission)

Age	Omphalocele major	Omphalocele minor	HUC	Survival
1day n=43 (62.31%)	9 (13.0 %)	20 (28.98%)	14 (20.28%)	36 (83.72%)
2 day n=16 (23.18%)	6 (8.69%)	5 (7.24%)	5 (7.24%)	13 (81.2%)
3 day n=5 (7.24%)	1 (1.44%)	1 (1.44%)	3 (4.34%)	3 (60%)
4 day n=1 (1.44%)	0	1 (1.44%)	0	0
5 day n=4 (5.79%)	1 (1.44%)	2 (2.89%)	1 (1.44%)	1 (25%)
Total n=69 (100%)	17 (24.73%)	29 (42.02%)	23 (33.33%)	53 (76.8%)

Table No. 3: Contents present in the sac

Type of defect	Contents	No. of patients
Omphalocele major n=17(24.73%)	Small bowel + large bowel	2 (11.76%)
	Small bowel + large bowel + liver	2 (11.76%)
	Contents could not be differentiated	13 (76.47%)
Omphalocele minor n=29(42.02%)	Small bowel	15 (51.72%)
	Small bowel + large bowel	10 (34.48%)
	Small bowel containing Meckel's diverticulum	3 (10.34%)
	Contents could not be differentiated	1 (3.44%)
HUC n=23(33.33%)	Small bowel	18 (78.26%)
	Small bowel + large bowel	3 (13.04%)
	Small bowel containing Meckel's diverticulum	2 (8.69%)
Total n=69(100%)		69 (100%)

Table No.4: Management of omphalocele and hernia of umbilical cord

Type of defect	Treatment	Frequency	Procedure	No. of Discharge	No of deaths
Omphalocele major n=17(24.73%)	Conservative	10	-	10	0
	Surgery	4	Anatomical repair -2	2	0
			Skin closure (ruptured sac)-1	1	0
			Silo repair-1	0	1
	Death before intervention	3	-	0	3
Omphalocele minor n=29 (42.02%)	Conservative	0	-	0	0
	Surgery	28	Primary anatomical repair-24	17	7
			Meckel's excision, resection anastomosis and anatomical repair-2	2	0
			Resection of Ileo-cecal Junction (ileal atresia) with ileo ascending anastomosis and repair-1	1	0
			Urachus ligation and anatomical repair-1	1	0
	Death before intervention	1	-	0	1
HUC n=23 (33.33%)	Minimal surgery	3	Reduction of content and ligation of cord-3	3	0
	Surgery	20	Primary repair-14	12	2
			Resection of gut, anastomosis (atresia or gangrenous gut) and repair-4	2	2
			Meckel's excision, resection anastomosis and repair-2	2	0

There were 69 neonates with omphalocele major (17), minor (29) and HUC (23). Male to female ratio was 2.83:1 in both omphalocele and hernia of umbilical cord, suggesting male predominance in each type of defect [Table 1]. Average gestational age of neonate was 39.25±1.49 and 39.35±2.01 weeks in omphalocele and hernia of umbilical cord respectively. Average weight of neonates was 2.33±0.50, and 2.13±1.17 kg in omphalocele and hernia of umbilical cord respectively. Average age of mothers was 23.64±2.16, and 23.57±1.42 years in omphalocele and hernia of umbilical cord respectively. Average size of defect was 3.42±0.83cm and 1.57±.87 cm in omphalocele and hernia of umbilical cord respectively. Number of caesarean deliveries in omphalocele and HUC was 2 in each case. History of Polyhydramnios was present in 2 (4.34 %) cases of omphalocele and 1 (4.34 %) case of HUC in our study. Out of all 46 cases of omphalocele, one case (2.17%) presented with ruptured sac. Remaining all cases including HUC were admitted with intact sac.

Maximum number of neonates were admitted on first live day (3 to 24 hours) after birth. It was observed that 43 neonates (62.31 %) were admitted on day first, 16 neonates (23.18 %) on day second, 5 neonates (7.24 %) on day third, 1 neonate (1.44 %) on day fourth and rest 4 (5.79 %) neonates on day 5 or even later. In day first admitted neonates, survival rate was 83.72% (36) followed by 81.25% (13),60% (3),zero (nil) and 25 % (1) in neonates admitted on day 2,3,4,and 5 or later respectively [Table 2].

Contents in sac of all type of abdominal wall defect were identified either pre operatively or intra operatively [Table 3]. In 17 cases of omphalocele major [Figure 1], 10 were managed conservatively [Figure 2] and 3 patients died before any surgical intervention. Because of coarse and opaque sac, contents could not be differentiated in all these patients (76.47 %). Small bowel and large bowel loop were identified in two cases of omphalocele major. Remaining 2 cases of omphalocele showed loop of small bowel, large bowel and liver in sac.

There were 29 patients of omphalocele minor [Figure 3] in our study. Only small bowel loops were present in 15 neonates (51.72 %). In 10 patients sac contents were loops of small and large bowel. The sac of 3 omphalocele minor patients contained small bowel loops along with Meckel's diverticulum. One patient of omphalocele minor expired before examination of the sac contents.

In HUC [Figure 4], contents of sac in 18 neonates (78.26 %) were single loop of small intestine, loops of small and large intestine in 3 neonates (13.4%) and small intestine with Meckel's diverticulum in 2 neonates (8.69 %).

Out of 17 patients of omphalocele major, 10 (58.82 %) were treated conservatively [Table 4]. All 10 patients were managed successfully and discharged. In 4 patients, surgical intervention was contemplated. Anatomical repair was done, in 2 neonates. In one patient, contents were loops of small bowel, large bowel and liver; anatomical repair was not possible, so silo repair was done. In one patient admitted with ruptured sac, skin flap closure was performed to form ventral hernia. Two patients were admitted with poor general condition with septicaemia and died before any surgical intervention. Another patient with omphalocele major and "ectopia cordis" admitted with poor general condition died on the day of admission. Mortality for omphalocele major was 23.52 % (4 patients) in our study. We did not found the case of beckwith-wiedemann syndrome (BWS) or visceromegaly in my study.

Out of 29 neonates with omphalocele minor, one died before surgery and remaining 28 patients were operated. In 24 patients, primary repair of defect was done successfully. Two patients had Meckel's diverticulum; excision of ileal segment with diverticulum and ileo-ileal anastomosis with repair of defect was done. One patient presented terminal ileal atresia with proximal segment dilatation. Resection of ileo-cecal junction including ileal atresia and ileo-ascending anastomosis was done with primary repair of wall defect. Patent urachus was also present in one patient; ligation of urachus and repair of defect was done. Total mortality in omphalocele minor patients was 27.58 % (8 patients).

All 23 neonates with HUC [Figure 4] were operated. In 3 patients, minimal surgery with reduction of contents and ligation of cord was done. In 14 patients, we did primary repair with reduction of contents and umbilicoplasty [Figure4]. Out of 23 patients, HUC was associated with perforated ileum in 2 and with ileal atresia in 2 patients. Resection of ileal segment and ileo-ileal anastomosis was done in these 4 patients. In two patients, small intestine with Meckel's diverticulum was found as content of the sac. Overall mortality was 17.39% (4 patients) in this group [Table 4]. Associated renal and cardiac anomalies were not reported in this study due to overburden of patients in 2D echo and USG centre at our institution.

Discussion:

Abdominal wall defects include mainly omphalocele, gastroschisis and HUC. The reported prevalence of omphalocele is 0.9–3.8 per 10,000 live births [1].Congenital hernia of the umbilical cord (HUC) is a different type of ventral abdominal wall defect with incidence of 1 per 5000 live birth. Unlike omphalocele, HUC is not associated with chromosomal anomalies, and association with other anomalies is also unusual [2]. Management of omphalocele is tedious because of its association with cardiac, pulmonary, various chromosomal anomalies and syndromes [3-8].Careful clinical examination is the best way to differentiate HUC and omphalocele [9].In case of HUC,the bowel usually herniates into the base of normally inserted umbilical cord through a patent umbilical ring. But in omphalocele there is a large defect of the anterior abdominal wall involving muscle and skin [10]. Although both of these conditions affect the umbilical area, their underlying pathology, genetics, and associated disorders are different. Consequently, the management techniques and outcomes are quite different [10,11].

The abdominal wall forms during the fourth week of intra-uterine life (IUL) when differential growth of the embryo causes infolding in the cranio-caudal and medio-lateral directions. During the sixth week of IUL, there is physiologic herniation of the gut in the umbilical coelom i.e. rapid intestinal and liver growth leads to herniation of the midgut into the umbilical cord. Elongation and rotation of midgut occurs over the ensuing 4 weeks. By week 10, the midgut returns to the abdominal cavity. An abdominal wall defect occurs due to interruption of these embryological processes [2,3,12,13]. Umbilical ring is formed by apex of four

body folds. Umbilical ring separates the body wall from the amnion [13].By 10th week of IUL, the epithelium on either side of the embryo fuses in the midline and only umbilical vessels are left in the region of the umbilical ring [14]. Gross and Blodgett proposed that omphalocele develops as a result of developmental arrest of the body cavity between 8th and 12th week of IUL, and herniated midgut fails to return back [15]. Margulies proposed that omphalocele develops before 3rd week of IUL, as a result of failure of either union of the mesodermal transverse septum with its amniotic covering or failure in proliferation of embryonal connective tissue in the transverse septum [16]. According to Gray and Skandalakis omphalocele develop due to arrest physiologic herniation of the gut in the umbilical coelom. The presence of liver (common) and other abdominal organs in sac is due to secondary herniation [17]. According to embryonic dysplasia theory, presence of several malformations occurs due to early germinal disc defects [18,19,20].

As such, the prognosis in these patients is affected by the severity and number of other anomalies and, amount and function of the bowel. The goal of surgical treatment for both conditions consists of reduction of the abdominal viscera and closure of the abdominal wall defect. Primary closure or a variety of staged approaches can be used avoiding injury to the intra-abdominal contents or developing abdominal compartment syndrome.

In this study, male to female ratio was 2.83:1 in both omphalocele and hernia of umbilical cord, suggesting male predominance in each type of defect. Male predominance with male to female ratio of 1.6:1 was also present in a recent retrospective study of omphalocele on 65 neonates [21]. In contrast, in one study for omphalocele and gastroschisis in 103 patients, female predominance in omphalocele was seen with male to female ratio 1:1.4 [22].

In our study, most of the neonates were admitted on first live day (3 to 24 hours) after birth. We observed that symptoms and signs of septicaemia and dehydration were more common in neonates with delayed presentation. Survival rate was also higher (83.72%) in neonates who were admitted on day first of life than the overall survival rate (76.81%) observed in the present study [Table 2].

Contents of sac in most of the abdominal wall defects were identified either pre operatively or intra operatively. In omphalocele major, most of the cases were managed conservatively, so contents could not be identified because of coarse and opaque sac. In 3 patients, contents could not be identified as they died before any intervention could be done. Among the remaining 4 neonates who were operated, small bowel and large bowel loop were identified as contents in two cases of omphalocele major during operation and small bowel, large bowel and liver in two cases. All cases of omphalocele minor patients were operated except one who died before operation. Contents in most of minor cases were small bowel loops followed by loops of both small and large bowel. In hernia of umbilical cord, contents of sac in most neonates were single loop of small intestine [Table 3].

The initial management of newborn with abdominal wall defects begins with the resuscitation and stabilization. Serum glucose levels needs to be checked and optimised in neonates with abdominal wall defects as they are associated with prematurity, especially in Omphalocele because of the possibility of Beckwith-Wiedemann Syndrome [23]. Oro(naso)gastric decompression is done to prevent gastrointestinal distension and risk of aspiration; bladder catheterisation is done to closely monitor urine output and direct on-going resuscitation. Intravenous access is obtained in upper extremities for fluids and broad-spectrum prophylactic antibiotics [23].

Surgical management of patients was planned according to their type, size and content of sac after thorough clinical examination and optimization of neonates. The defect should be inspected to make sure that the covering membrane is intact, andnon-adherent dressings are applied and stabilized to prevent trauma to the sac. If the omphalocele sac is ruptured, then exposed bowel should treated as per protocol for gastroschisis. If the covering sac is complete, then there is no urgency to accomplish operative closure [23,24]. The primary treatment of choice was anatomical closure of defect. A primary repair was done by

excising the omphalocele membrane, reducing the herniated viscera, and closing the fascia and skin. Membrane overlying the liver that might be injured during excision was left in place [23].

Primary closure was difficult in large size defect. In such situation staged repair was done to enhance size of abdominal cavity. In 1948, Gross gave the concept of ventral hernia repair and this technique has evolved considerably [25,26]. Various other methods are used by applying prosthetic material such as silastic silo, Schuster bag, tissue expander, vicrylmesh with variable results [27,28].

Another form of staged closure is "Escharotic therapy" with gradual epithelialization of the sac over the ensuing weeks to months [Figure 2]. It is contemplated when neonate cannot tolerate operative intervention in conditions like prematurity, pulmonary hypoplasia, congenital heart disease or other anomalies. Escharotic agents that are commonly used are mercurochrome, povidone iodine solution and silver sulfadiazine. Due to associatedmercury poisoning of Mmercurochrome silver sulfadiazine is preferred escharotic agent [29]. The remaining defect can be repaired either by anatomical closure or skin flap coverage along with prosthetic mesh if required. Tissue expanders can be used at this stage as well as in neonatal period to enhance abdominal cavity [27].

Most of patients with omphalocele major were managed by conservative therapy in our study. Povidone iodine solution (5%) and antiseptic dressing was used as escharotic agent since it is easily available. We use bandage rings to support and apply povidone iodine-soaked antiseptic dressing for escharotic therapy in our institute [Figure 2].Out of 17 patients of omphalocele major, 10 (58.82 %) were treated conservatively. All 10 patients were managed successfully and discharged. Application of Povidone iodine has been associated with transient hypothyroidism so regular thyroid monitoring should be done [30].

In 4 patients, emergency surgery was done. Out of 4, contents of 2 patients (11.76 %) were small bowel and large bowel loops, so anatomical repair done successfully after reduction of contents. In one patient, contents were loops of small bowel, large bowel and liver. Anatomical repair was not possible in this patient because of discrepancy between abdominal cavity and contents, so silo repair was done. But despite of best possible effort, patient died on third post-operative day due to septic shock. In one patient admitted with ruptured sac, contents were parts of small bowel large bowel and liver. So skin closure was done to form ventral hernia. Two patients were admitted with poor general condition with septicaemia and died before any surgical intervention. One another patient with omphalocele major and ectopia cordis also came with poor general condition and was diagnosed as a case of Cantrell's pentalogy. It was planned for emergency surgery but patient died on the day of admission. Mortality for omphalocele major was 23.52 % in our study.

Total 29 neonates were admitted with omphalocele minor. All were planned for surgery but one patient admitted with very poor general condition and died before surgery whereas 28 patients were operated. In 24 patients, primary repair of defect was done successfully. Out of these 24 patients 7 patients expired in post-operative period. Total mortality in omphalocele minor patients was 27.58 % (8), because of associated anomalies.

Out of 46 patients of omphalocele (major and minor), overall mortality in the study was 26.08% (12). In 2018, Rattan KN et al reported 7.69% mortality in their study of 65 cases of omphalocele [21]. In a population based-based study done by Marshall et al, survival rate of omphalocele was reported at 71.3%. They recognized chromosomal anomalies and very low birth weight as consistent determinants of 1year survival [31].

A total 23 neonates were admitted with HUC and all were operated. Out of these 23 patients, two patient of perforated ileum expired after surgery. Delayed admission and septicaemia could be the possible reason. Two patients expired after primary repair. Overall mortality was 17.39% (4 patients) in this group.

Conclusion:

Omphalocele is more common than hernia of umbilical cord. Male predominance was observed in our study,

both in omphalocele and HUC. Treatment of omphalocele especially the major type remains a challenge for pediatric surgeon. Early admission and early management improves the prognosis. Primary anatomical repair is most often done for omphalocele minor and HUC. In omphalocele major primary anatomical repair is often not feasible so "escharotic therapy" followed by definitive surgery is done.We found that primary anatomical repair is better choice for omphalocele minor and conservative scar therapy is good for omphalocele major, until unless the sac was ruptured.

Conflict of Interest - Nil
Sources of Support - Nil

References

1. Gong TT, Wu QJ, Chen YL, Jiang CZ, Li J, Li LL, Liu CX, Li D, Zhou C, Huang YH. Evaluating the time trends in prevalence of exomphalos in 14 cities of Liaoning province, 2006 to 2015. *Scientific Reports* 2016; 6:32901.
2. Achiron R, Soriano D, Lipitz S, Mashiach S, Goldman B, Seidman DS. Fetal midgut herniation into the umbilical cord: Improved definition of ventral abdominal anomaly with the use of transvaginal sonography. *Ultrasound in Obstetrics and Gynecology* 1995; 6:256-260.
3. Lap CC, Brizot ML, Pistorius LR, Kramer WL, Teeuwen IB, Eijkemans MJ, et al. Outcome of isolated gastroschisis; an international study, systematic review and meta-analysis. *Early Human Development* 2016; 103:209-218.
4. Feldkamp ML, Carey JC, Sadler TW. Development of gastroschisis: review of hypotheses, a novel hypothesis, and implications for research. *American Journal of Medical Genetics* 2007; 143A:639-652.
5. Feldkamp ML, Alder SC, Carey JC. A case control population-based study investigating smoking as a risk factor for gastroschisis in Utah, 1997-2005. *Birth Defects Research A Clinical and Molecular Teratology* 2008; 82:768-775.
6. Feldkamp ML, Carmichael SL, Shaw GM, Panichello JD, Moore CA, Botto LD, et al. Maternal nutrition and gastroschisis: findings from the National Birth Defects Prevention Study. *American Journal of Obstetrics and Gynecology* 2011; 204:404.e1–404.e10.
7. Browne ML, Hoyt AT, Feldkamp ML, Rasmussen SA, Marshall EG, Druschel CM, Romitti PA. Maternal caffeine intake and risk of selected birth defects in the national birth defects prevention study. *Birth Defects Research A Clinical and Molecular Teratology* 2011;91:93-101.
8. Frolov P, Alali J, Klein MD. Clinical risk factors for gastroschisis and omphalocele in humans: a review of the literature. *Pediatric Surgery International* 2010;26:1135-1148.
9. Axt R, Quijano F, Boos R, Hendrik HJ, Jessberger HJ, Schwaiger C, Schmidt W. Omphalocele and gastroschisis: prenatal diagnosis and peripartal management. A case analysis of the years 1989-1997 at the Department of Obstetrics and Gynecology, University of Homburg/Saar. Europian *Journal of Obstetrics & Gynecology and Reproduction Biology* 1999 Nov; 87(1):47-54.
10. Brantberg A, Blaas HGK, Salvesen KA, Haugen SE, Eik-Nes SH. Surveillance and outcome of fetusses with gastroschisis. *Ultrasound in Obstetrics and Gynecology* 2004;23:4-13.
11. Fraga MV, Laje P, Peranteau WH, Hedrick HL, Khalek N, Gebb JS, Moldenhauer JS, Johnson MP, Flake AW, Adzick NS. The influence of gestational age, mode of delivery and abdominal wall closure method on the surgical outcome of neonates with uncomplicated gastroschisis. *Pediatric Surgery International* 2018 Apr;34(4):415-419.
12. Brantberg A, Blaas HGK, Salvesen KA, Haugen SE, Eik-Nes SH. Surveillance and outcome of fetusses with gastroschisis. *Ultrasound in Obstetrics and Gynecology* 2004;23:4-13.
13. Brewer S, Williams T. Finally, a sense of closure? Animal models of human ven- tral body wall defects. *Bioessays* 2004;26:1307-1321.
14. Moore KL, Persaud TV, Torchia MG. The developing human E-book. Elsevier Health Sciences, 11th edition; 2011.
15. Gross RE, Blodgett JB. Omphalocele (umbilical eventration) in the newly born. *Surgery, Gynecology and Obstetrics* 1940;71:520.
16. Margulies L. Omphalocele (amniocele): its anatomy and etiology in relation to hernias of umbilicus and the umbilical cord. *American Journal of Obstetrics and Gynecology* 1945;49:695-699.
17. Skandalakis, JE, Gray SW. (1972) Embryology for Surgeons. The Embryological Basis for the Treatment of Congenital Anomalies. 2nd Edition, WB Saunders, Philadelphia, 414-415.
18. Streeter G. Focal deficiencies in fetal tissues and their relation to intrauterine amputations. *Contributions to Embryology Carnegie Institution* 1930;22:1-46.
19. Herva R, Karkinen-Jääskeläinen M. Amniotic adhesion malformation syndrome: fetal and placental pathology. *Teratology* 1984;29:11-19.

20. Khan FA, Hashmi A, Islam S. Insights into embryology and development of omphalocele. *Seminars in Pediatric Surgery* 28:80-83.
21. Rattan KN, Singh J, Jakhar R, Dalal P, Sonika P. Omphalocele: 15-years experience from a single center in developing country. *Journal of Clinical Neonatology* 2018;7:125-129.
22. Jwa E, Kim SC, Kim DY, Hwang JH, Namgoong JM, Kim IK. The prognosis of gastroschisis and omphalocele. *Journal of Korean Association of Pediatric Surgery* 2014;20:38-42.
23. Ledbetter DJ. Gastroschisis and omphalocele. *Surgical Clinics of North America* 2006;86:249-260.
24. Nuchtern JG, Baxter R, Hatch EI Jr. Nonoperative initial management versus silonchimneyfor treatment of giant omphalocele. *Journal of Pediatric Surgery* 1995;30(6):771-776.
25. Gross RE. A new method for surgical treatment of large omphaloceles. *Surgery* 1948; 24:277-292.
26. Bax NM, Vander Zee DC, Pullter Gunne AJ, Rövekamp MH. Treatment of giant omphalocele by enlargement of the abdominal cavity with a tissue expander. *Journal of Pediatric Surgery* 1993;28:1181-1184.
27. Foglia R, Kane A, Becker D, Asz-Sigall J, Mychaliska G. Management of giant omphalocele with rapid creation of abdominal domain. *Journal of Pediatric Surgery* 2006;41:704-709
28. Schuster SR. A new method for the staged repair of large omphaloceles. *Surgery, Gynecology and Obstetrics* 1967; 125:837-850.
29. Yeh TF, Pildes RS, Firor HV. Mercury poisoning from mercurochrome therapy of an infected omphalocele. *Clinical Toxicology* 1978; 13:463-467.
30. Whitehouse JS, Gourlay DM, Masonbrink AR, Aiken JJ, Calkins CM, Sato TT, Arca MJ. Conservative management of giant omphalocele with topical povidone-iodine and its effect on thyroid function. *Journal of Pediatric Surgery* 2010 Jun;45 (6):1192-1197.
31. Marshall J, Salemi JL, Tanner JP, Ramakrishnan R, Feldkamp ML, Marengo LK, Meyer RE, Druschel CM, Rickard R, Kirby RS; National Birth Defects Prevention Network. Prevalence, Correlates, and Outcomes of Omphalocele in the United States, 1995-2005. *Obstetrics and Gynecology* 2015 Aug;126 (2):284-293.

Address for correspondence: *Dr. Prameshwar Lal, Senior Resident, Department of Paediatric Surgery, Sir Padampat Mother and Child health Institute, S.M.S. Medical College, Jaipur, Rajasthan, India. Email:drparmesh.kumawat@gmail.com, Mobile: +91 9571452010.*

How to cite this article: Prameshwar Lal, Neeraj Tuteja, Dinesh Kumar Barolia, Vinita Chaturvedi, Rahul Gupta, Gurudatt Raipuria, Vikas Joshi, Arun Gupta and Ajay Kumar. Omphalocele and Hernia of Umbilical Cord: An Early Outcome Analysis from a Tertiary Care Hospital. Walawalkar International Medical Journal 2021; 8(1):42-51. http://www.wimjournal.com

Received date: 08/02/2021 Revised date: 04/06/2021 Accepted date: 05/06/2021