ORIGINAL ARTICLE

Congenital Hernia of the Umbilical Cord: A Retrospective Case Study

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ABSTRACT

Background. Congenital hernia of the umbilical cord (CHUC) is the rarest type of anterior abdominal wall defect, in which an intact umbilical ring is always present and viscera pass through the base of normal-looking umbilicus.

Objectives. This study was conducted to document the intraoperative findings and postoperative outcomes of patients with congenital hernia of the umbilical cord up to discharge from a tertiary care center.

Methods. This study was a retrospective observational study conducted for two years (August 2020 to July 2022) in the Department of Pediatric Surgery, at the tertiary health care center of UP, India.

Results. During this two-year duration, a total of 10 cases with CHUC were seen in our department and were surgically managed. In this study, out of these 10 patients (male 7 and female 3), eight had normal gastrointestinal tract, one had accessory liver tissue on thin pedicle, and one had features of gangrenous bowel. Of these 10 cases, three patients developed postsurgical complications in which two patients developed superficial wound infection while one developed wound dehiscence. No mortality was noted.

Conclusions. Congenital hernia of the umbilical cord induces stress on parents and relatives. In this study, we conclude that the majority of cases had normal gastrointestinal tract and had no serious postoperative complications up to discharge.

Keywords: abdominal wall defect, umbilical cord hernia, congenital, umbilical cord

INTRODUCTION

Umbilicus is considered the mirror of the abdomen, providing the necessary passage for vessels and structure required for the developing fetal gastrointestinal and urinary systems.[1] Umbilicus is a site of numerous embryopathies involving vessels, urachus, midgut herniation, anterior abdominal wall defects, and congenital cysts. The incidence of congenital hernia of the umbilical cord (CHUC) was reported to be about 1 in 5000.[2] Congenital abdominal wall defect includes omphalocele, gastroschisis, and umbilical cord hernia.[3] In comparison with omphalocele and gastroschisis, CHUC is rare. CHUC is embryologically and anatomically different from omphalocele, umbilical hernia, and gastroschisis.[4] CHUC is categorized as a benign and isolated condition.[2] Variation in the size of herniating mass in CHUC makes it difficult to differentiate it from other conditions like umbilical cord cyst, hematoma, etc.[1] Small

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and reducible congenital hernias of the cord are left intact without surgical intervention by some clinicians which finally get epithelialized as 'cutis navel'. Inadvertent clamping of congenital hernia of cord containing viscera leads to iatrogenic visceral injury.[5] Congenital umbilical cord hernia is often mistaken as 'omphalocele minor'. Owing to the lack of exposure and awareness about this entity, it is often misdiagnosed and under-reported, with limited data in the literature.[6] Antenatal diagnosis is important for postnatal management which may differ widely. Accurate diagnosis of disease entity is important to achieve appropriate and accurate prenatal counseling and postnatal management. This study was conducted to document the outcome of operated cases of congenital hernia of the umbilical cord at a rural tertiary care center.

MATERIAL AND METHODS

This study was a retrospective observational study, conducted for a 2-year duration from August 2020 to July 2022, in the Department of Pediatric Surgery, at the tertiary health care center of UP, India. Our tertiary care center is in the rural part of western U.P in northern India and primarily caters to the rural population. Data of patients presenting with congenital hernia of the umbilical cord were collected from hospital record registers and case sheets after taking ethical clearance from the Institutional Ethical Committee (1/2021-22). Demographic profile, radiological image, intraoperative findings, and postoperative complications were tabulated in Microsoft Office Excel sheet. Patients with intact hernial sacs were included in this study while patients with ruptured sacs and incomplete data were excluded in this study. All patients were surgically treated by trained Pediatric surgeons after resuscitation and preoperative necessary consent was taken from the parents of patients. Umbilicus was explored through the right lateral side of the umbilicus in all neonates however, extension of incision was made on the lateral abdominal wall, if needed. Patients were followed-up for a period of two months.

RESULTS

In this study, the age of neonates ranged from 1 day to 8 days old, in which seven patients were males and three patients were females, Mean age was 2.4 days. All neonates presented with mass protruding through the normal-looking umbilicus covered with amniotic membrane since birth. All neonates were delivered at full term and no visible congenital anomalies other than CHUC were present (Table 1). Routine investigations e.g., complete blood count, liver function test, kidney function test, and electrolytes were within normal limits in all neonates. All neonates with antenatal ultrasonography images did not show a positive finding of a hernia of the umbilical cord. Postnatal echocardiograms were normal in all neonates.

Eight neonates (Cases 1, 2, 4, 5, 7, 8, 9, and 10) had afferent and efferent bowel loop inside the amniotic sac that was side to side, circular pattern, and gut mesentery of the same loop was narrow (Figures 1A and B). No other significant intraabdominal pathology was found. For these eight cases, minor widening of bowel mesentery inside the amniotic sac followed by umbilicoplasty were done. Feeding was started at 2nd postoperative day after bowel movement. The postoperative period of these neonates except for case 1 was uneventful and they were discharged on the 5th postoperative day. Case 1 developed superficial wound infection most probably due to endogenous flora on the 3rd post-operative day for which saline irrigation and dressing were done. The wound swab culture was found sterile and the patient was discharged on the 8th postoperative day.

Distended and gangrenous midgut was present inside the hernial sac in one neonate (Case 3). Resection and anastomosis followed by umbilicoplasty were done (Figure 2). Feeding was started on the 4th postoperative day after bowel

Table 1. Patient profile, antenatal diagnosis, visible congenital anomalies, intraoperative findings, and postoperative complications in congenital hernia of umbilical cord

Case No.	Age (days)	Sex	Antenatal diagnosis	Associated congenital anomalies	Intraoperative findings	Postoperative complication
1	1	M	none	none	Mid gut with narrow mesentery inside the amniotic membrane	Superficial wound infection
2	2	M	none	none	Mid gut with narrow mesentery inside the amniotic membrane	None
3	5	M	none	none	Gangrenous small bowel	Superficial wound infection
4	1	M	none	none	Mid gut with narrow mesentery inside the amniotic membrane	None
5	2	F	none	none	Mid gut with narrow mesentery inside the amniotic membrane	None
6	8	M	none	none	Liver tissue attached by a thin pedicle	Wound dehiscence
7	1	M	none	none	Mid gut with narrow mesentery inside the amniotic membrane	None
8	1	F	none	none	Mid gut with narrow mesentery inside the amniotic membrane	None
9	1	F	none	none	Mid gut with narrow mesentery inside the amniotic membrane	None
10	2	M	none	none	Mid gut with narrow mesentery inside the amniotic membrane	None

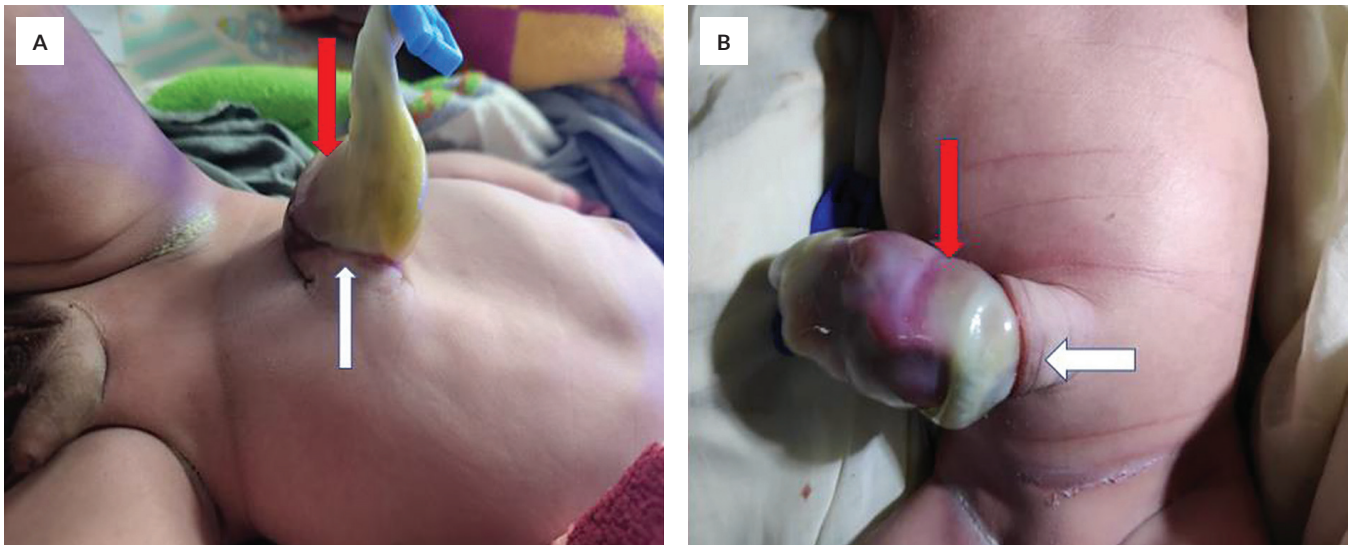

Figure 1. ***Preoperative images of patients with CHUC.*** **(A)** Case 1 shows normal umbilical ring *(white arrow)* and umbilical cord *(red arrow)*. **(B)** Case 7 shows a normal-looking umbilical ring (*white arrow)* with intestine inside the umbilical cord *(red arrow)*.

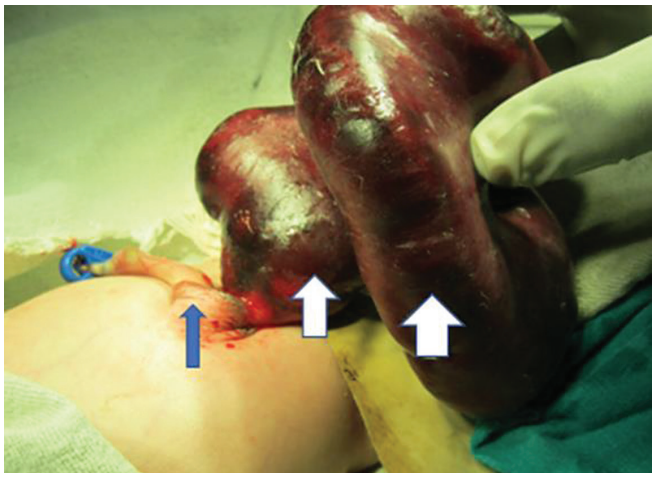

Figure 2. Intraoperative image of Case 3 patient showing normal umbilicus *(blue arrow)* with gangrenous intestinal loop *(white arrows)*.

movement. The patient developed superficial wound infections most probably due to intraoperative contamination during surgical procedure on the 3rd postoperative day for which saline irrigation and dressing were done. The wound swab culture was found sterile and the patient was discharged on the 8th postoperative day.

An accessory liver tissue with a thin pedicle attached to the inferior surface of the liver was found in the hernial sac in one neonate (Case 6, Figure 3A). Accessory liver tissue was resected from the inferior surface of the liver and umbilicoplasty was done (Figure 3B). Histopathology of the specimen showed liver tissue (Figure 3C). Feeding was started after bowel movement on the second postoperative day. The patient developed wound dehiscence probably due to endogenous flora and seroma formation on postoperative day 5, for which secondary suturing was done on the same day. Pus culture showed gram-negative E. coli for which antibiotics were given according to culture sensitivity.

All neonates were doing well in the follow-up period of two months. No mortality occurred during this study.

A patient with CHUC was operated two years back and was admitted for inguinal hernia repair having normal looking umbilicus (Figure 4).

DISCUSSION

CHUC is categorized as a benign and isolated condition.[2] It is sporadically associated with other congenital anomalies like intestinal anomalies, short gut, atresia, patient omphalomesenteric duct, malrotation, and volvulus.[2] Unlike omphalocele, CHUC is not linked with chromosomal anomalies. During the fifth to sixth week of embryonic development, the midgut physiologically protrudes into the extraembryonic coelom through proposed umbilics. Then, it returns to the abdominal cavity around the 10th to 12th week of gestational age. If any part of the midgut fails to return to the abdominal cavity, the umbilical cord hernia develops.[6] Mirza et al. classifies CHUC into four types based on clinical presentation.[7,8] Type 1 is a simple hernia into the cord without any associated complications, Type 2 is CHUC associated with intestinal obstruction, Type 3 is CHUC associated with mucosal prolapse, and Type 4 is CHUC associated with evisceration. By clinical examination, CHUC can be differentiated from omphalocele minor based on the location of the umbilical cord insertion. A regular cord insertion and undamaged skin surrounding the umbilical ring occur in an umbilical cord hernia. An omphalocele, on the other hand, is characterized by a significant herniation

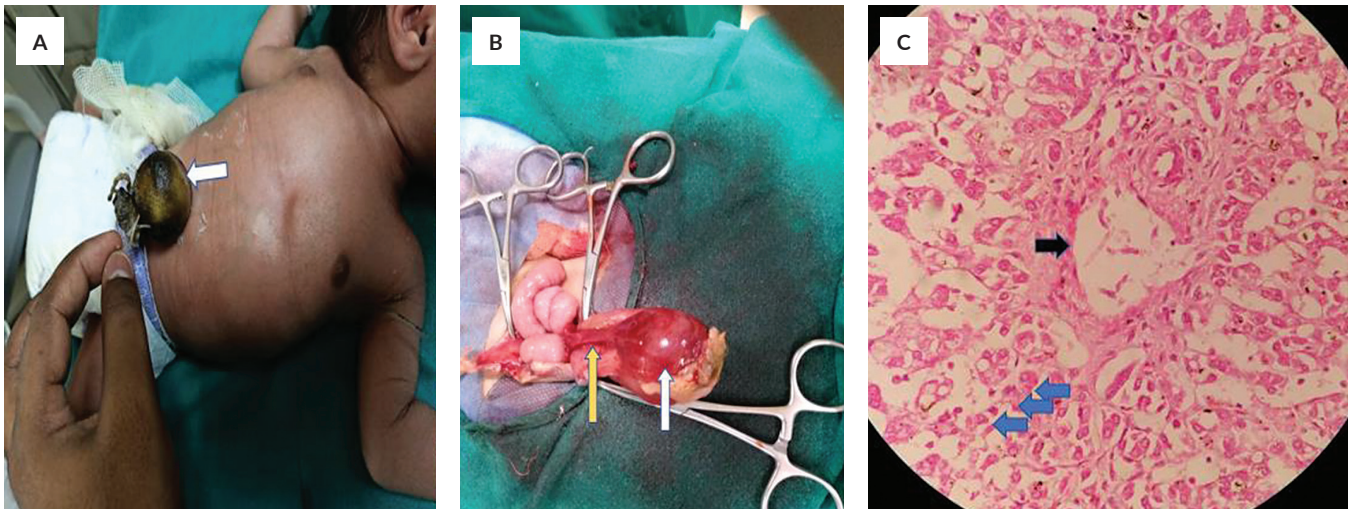

Figure 3. ***Patient (Case 6) with congenital hernia of the umbilical cord with accessory liver.*** **(A)** Preoperative case of CHUC with normal umbilical ring *(white arrow)*. **(B)** Accessory liver tissue *(white arrow)* with narrow liver tissue pedicle *(yellow arrow)*. **(C)** Micrograph showing strands of hepatocytes *(blue arrow)* and central vein *(black arrow)*. (H & E stain, 10x).

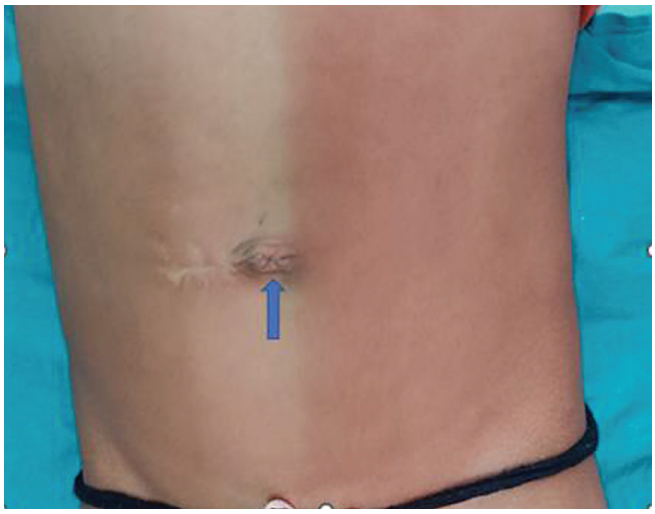

Figure 4. Showing a 2-year postsurgical CHUC patient with normal-looking umbilicus *(blue arrow)*.

of the sac, with an insertion of the umbilical cord on top of the herniated sac, and involves a big defect in the umbilical ring including the skin and muscle.[6,9] A thin strip of normal skin always enwraps the umbilical ring and proximal part of the cord in the case of CHUC.[10] CHUC with patent omphalomesenteric (OMD) anomaly was reported by Raicevic et al. in their case report on neonates.[2] In their study, they described the intraoperative identification of patent OMD in CHUC. In patent OMD with an opening on both sides where meconium has been passed, they resected widely open patent OMD, and end-to-end ileo-ileal anastomosis was performed. They concluded that any unusual thickening of the base of the cord with fistula opening should be an alert signal for clinicians for OMD anomaly in CHUC patients.

CHUC with Meckel's diverticulum was reported by Laezza et al. in their case report.[11] In their study, they found clinically a bulge with air and fluid content, palpable at the base of the umbilical stump. Herniated Meckel's diverticulum was found intraoperatively for which wedge diverticulectomy and umbilicoplasty were done. Gastroschisis presents as a right-sided paraumbilical defect with herniation of abdominal viscera, free-floating in the amniotic cavity.[6] Rupture of CHUC is a rare event and is often misdiagnosed as gastroschisis or ruptured omphalocele.[8] Other complications accompanying CHUC include bowel obstruction, ileal and colonic atresia, congenital short bowel, malrotation of the GI tract, and inadvertently trauma caused by clamping of the umbilical cord. Atresia in CHUC is probably caused by intrauterine vascular accidents, volvulus, and intussusception.[7] Uncommon associations were reported by different authors in CHUC like congenital heart disease, cleft lip, palate, and glaucoma.[12-14] In CHUC, no published data for the association of genetic disorders to date is available.[1] An umbilical hernia occurs due to incomplete closure or weakness at the umbilical ring during post-natal life by which intra-abdominal content protrudes. The fascia posterior to the umbilical cord is thin and weak which leads to an area of weakness causing the umbilical hernia.[15]

Differentiation between ventral abdominal wall defect and CHUC by ultrasonography is difficult and omphalocele minor may be misdiagnosed as CHUC because of similarity in USG findings.[6] An omphalocele is a more serious prenatal ultrasound finding. Prenatal ultrasound findings of omphaloceles are typically characterized by a sizable umbilical defect that develops when the ventral abdominal wall fails to close, and they are 30-40% linked to chromosomal defects, which worsen the prognosis. In CHUC, chromosomal anomalies have not been linked hence it has a better prognosis.

CHUC, omphalocele, and gastroschisis can develop at an early embryological stage and can be detected by ultrasonography in the second trimester.[16] CHUC can be accurately detected by high-resolution transvaginal ultrasonography (TVS) around 13-14 weeks of gestational age but this antenatal scan is operator dependent.[1] Three-dimensional USG adds the anatomical findings in case of anterior wall defect.

Recently, the role of Magnetic Resonance Imaging (MRI) has evolved in the diagnosis of CHUC and other ventral wall defect spectrums. Fetal MRI has an important role in the diagnosis of fetal ventral body wall defects like omphalocele, exstrophy, cloacal exstrophy, pentalogy of Cantrell (POC), etc.[17,18] In the study conducted by Victoria et al., they concluded that fetal MRI is an important tool to diagnose abdominal wall defects.[19] In their study, they elaborated on abdominal wall defects ranging from the mild umbilical cord hernia to the highly complex limb-body wall syndrome. Gastroschisis and omphalocele were the most common defects found in their study while exstrophy complex, pentalogy of Cantrell, and limb body wall syndrome were rare clinical entities. Viscera herniation through a defect in the anterior body wall was similar to the finding in our study.

CONCLUSIONS

In congenital hernia of the umbilical cord, early surgical exploration is mandated to prevent complications like congenital bowel atresia, perforation, gut gangrene, etc. The benign nature of the disease minimizes postnatal stress on parents. In this study, we conclude that the majority of cases had normal gastrointestinal tract, and no serious postoperative complications were seen up to patients' discharge from the hospital.

Statement of Authorship

All authors certified fulfillment of ICMJE authorship criteria.

Author Disclosure

All authors declared no conflicts of interest.

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