Case Series	
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Title	Umbilical Cord Case Series Hernia in Neonates: A Retrospective and Diagnostic Challenge
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Keywords

Umbilical cord hernia
Neonatal abdominal wall defects

Abbreviations
UCH- Umbilical Cord Hernia

Abstract

Umbilical cord hernia (UCH) is an uncommon congenital defect that is frequently misdiagnosed as omphalocele minor, leading to underrecognition in clinical practice. In this retrospective case series, we reviewed seven neonates treated over four years at a tertiary paediatric surgical unit in Sri Lanka. All underwent surgical repair via an umbilical approach, with no recurrences noted during follow-up. Meckel's diverticulum was the most common sac content, and unusual findings included liver and appendix. Two patients experienced postoperative complications requiring intensive care and ileostomy formation. Most cases were isolated anomalies, though one neonate presented with a previously unreported association of tracheoesophageal fistula. Notably, none of the cases were identified antenatally, underscoring the value of prenatal detection to avoid potential visceral injury at birth. Our experience highlights the diverse presentations of UCH and the importance of distinguishing it from omphalocele for appropriate management and prognosis.

Introduction

Hernia of the umbilical cord is a rare benign congenital defect of the umbilicus resulting in herniation of the abdominal contents, with an incidence of 1 in 5000 live births[1]. It is frequently misdiagnosed as a minor omphalocele and is hence underreported. Pathology of the neonatal umbilicus varies widely and includes anterior abdominal wall defects such as omphalocele and gastroschisis, umbilical cord hernia, urachal abnormalities, and pathologies related to the omphalomesenteric duct, such as Meckel diverticulum.

During embryogenesis, physiological herniation of intestine into the extracelomic cavity occurs at 5–6th week of gestation and returns to the abdominal cavity after intestinal rotation at 10–12 weeks of gestation. During this process, failure of umbilical ring closure could lead to umbilical cord hernia, which presents as a cystic swelling at the base of the cord with peritoneum and amnion covering the readily visible intestines[2].

Unlike omphalocele, umbilical cord hernia is not associated with chromosomal abnormalities or other congenital abnormalities, except for sporadic anomalies like ileal atresia, malrotation, and omphalomesenteric duct anomalies[2,3,4]. Iatrogenic visceral injury following inadvertent clamping of the umbilical cord with the hernia is a well-recognized complication of this anomaly, if it's not antenatally detected[1,5]. It has a very good prognosis after surgical intervention.

Method

This is a retrospective review over a period of 4 years in the Department of Paediatric Surgery, Sirimavo Bandaranaike Specialized Children's Hospital, Sri Lanka, which is a tertiary care children's hospital. Patient data were retrospectively collected from a prospectively maintained electronic health record system. Demographic details of the patients, intraoperative findings, complications, and postoperative outcomes were tabulated. All the patients were operated on by a single surgeon. Surgical exploration was done through umbilical access in all patients, with extension of the incision if required for extended access. Adhesions to the viscera in the sac were released before the repair of the defect. Reduction of the viscera and ligation of the sac were not performed in any case.

Observations/ Results

There were 7 neonates over the study period, out of which 4 were males and 3 were females. Age ranged from day 1 to day 3 on presentation to the children's hospital. Associated abnormalities included oesophageal atresia with tracheoesophageal fistula in one baby, a congenital cardiac abnormality in another, and duodenal atresia with midgut malrotation and annular pancreas in a third. In all other babies, the umbilical cord hernia was the only congenital abnormality.

On presentation, all the babies had amniotic membrane-covered contents with skin circumferentially covering like a sleeve on the hernial sac. None of the babies were detected to have UCH antenatally.

Four babies were born prematurely, and the other three babies were term. One baby presented with intestinal obstruction and was found to have an ileal perforation on exploration of UCH. He had a complicated postoperative course with an ileostomy, which was closed subsequently. Another baby had Meckel's diverticulum and developed NEC and an ileal perforation. He had an ileostomy, which was closed on the same admission due to severe diarrhea. These two patients and the baby with duodenal atresia and midgut malrotation needed TPN and ventilation with ICU care.

Contents of the hernia:
Three babies had Meckel's diverticulum in the sac, which was adhered to the sac. Two babies had a segment of liver in the sac without any hollow viscera. Another baby had an appendix in the sac, which was adhered to the margin. The estimated diameter of the defect was around 1.5cm in all cases. All babies had primary repair of the hernia, leaving the umbilical cord attached. On follow-up, none of them presented again with an umbilical hernia.

Case No	Age	Sex	Gestation	Antenatal Diagnosis	Birth weight	Content of Sac	Associated anomalies	Post op Complications
01	Day 01	Male	36+5	None	3.3	Small bowel, large bowel, appendix	None	None
02	Day 03	Male	37	None	2.34	Meckel's diverticulum	None	NEC with ileal perforation. Ileostomy created & closed before discharging due to high output
03	Day 01	Female	Preterm	None	2.1	Part of the liver	None	None
04	Day 01	Male	35+3	None	2.2	Meckel's diverticulum	Type C TOF with esophageal atresia	None

05	Day 03	Male	Term	None	3.2	Obstructed cord hernia containing Meckel's diverticulum with an ileal perforation	None	No immediate postoperative complications. Following ileostomy closure, developed an anastomotic leak requiring a right hemicolectomy
06	Day 01	Female	37+4	None	2	Caecum, terminal ileum & appendix	None	None
07	Day 01	Female	32+5	Duodenal atresia	1.41	Part of the right lobe of the liver	Duodenal atresia Midgut malrotation Annular pancreas	None

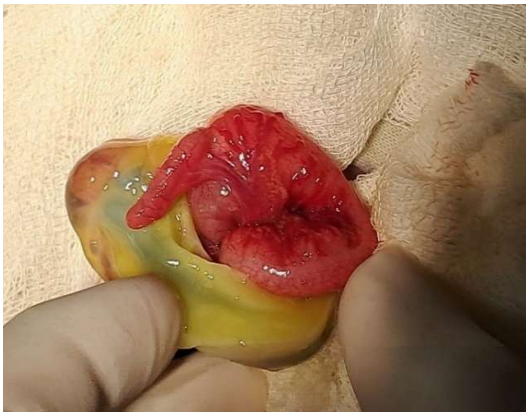

Figure 1- Case number 06 (umbilical cord hernia containing appendix, caecum & terminal ileum)

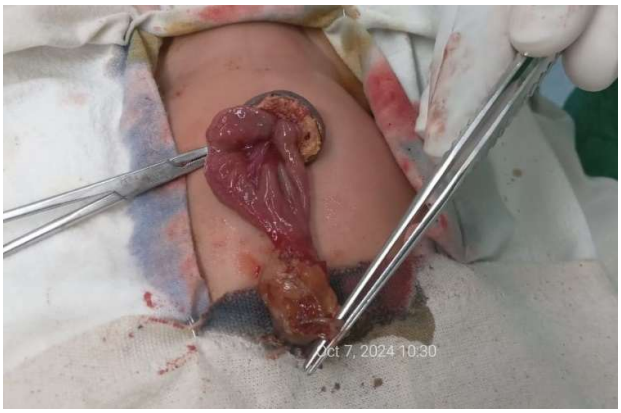

Figure 2- Case number 02 (Umbilical cord hernia containing Meckel's diverticulum)

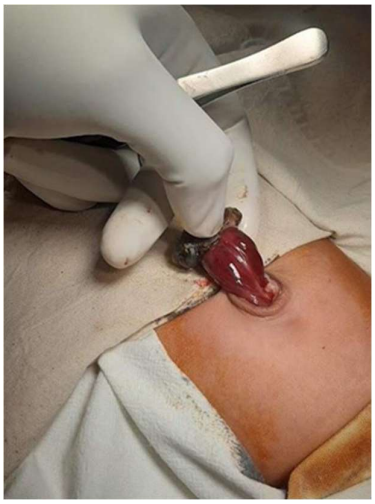

Figure 3- Case number 03 (umbilical cord hernia containing part of the liver)

Discussion

Umbilical cord hernia was first described in the literature by Hempel-Jorgensen in his 2 cases [1]. Subsequently, Tow and Burn in 1937 and 1938 reported a few cases with the embryological background [6]. Due to the morphological similarity with omphalocele minor, many of these cases were underreported, and due to the rarity of the pathology, only a few case reports and series have been published to date.

During embryogenesis, the midgut herniates into the extracelomic cavity formed by the umbilical cord and returns to the peritoneal cavity at the 10–12th week of gestation. Failure of return of the midgut contents back to the peritoneal cavity during this process is considered the aetiology of UCH. The failure of return of contents is thought to be due to adhesions of the contents to the sac. Unlike omphalocele and gastroschisis, UCH has a normal anterior abdominal wall with rectus muscles aligned in the midline and attached normally to the xiphoid process, with an enlarged umbilical ring[1,²]. Classically, skin is extended onto the cord like a sleeve. In contrast, the defect of an omphalocele is due to the failure of fusion of the lateral body folds, leading to a defect above the umbilicus.

Omphalocele is classically associated with other congenital and chromosomal anomalies, whereas UCH is considered a simple anomaly.

UCH is classified into 4 categories according to the presentation and complications[4].

Type 1 – Simple UCH
Type 2 – Associated with intestinal obstruction
Type 3 – Associated with mucosal prolapse when the omphalomesenteric duct anomaly is present
Type 4 – Associated with evisceration due to in utero rupture of the umbilical cord

In our series, only one baby presented with Type 2 UCH, while all others had Type 1 defect.

The contents of the UCH vary widely but primarily comprise derivatives of the midgut. Additionally, the presence of a patent urachus[7] and a segment of liver[8,9] in the UCH has been described in the literature. Some authors define UCH as having midgut derivatives in the sac solely, while others accept any intra-abdominal content.

In our series, two babies had a segment of liver as the sole content in the sac — an extremely rare finding according to the literature[8,9].

Antenatal detection of cord hernia could prevent potential complications of visceral damage caused by inadvertent clamping of the cord after birth. Visceral damage from clamping is a potentially lethal complication of UCH[1]. In our series, none of the babies had antenatal detection, hence all were diagnosed postnatally.

Presentation of UCH varies from an asymptomatic hernia to complications like intestinal obstruction. In our series, one baby presented with obstruction caused by Meckel's diverticulum in the sac. In all other cases, the contents were reduced after releasing the adhesions.

Meckel's diverticulum in the UCH is a well-documented occurrence[10]. In our series, 3 out of 7 patients had Meckel's diverticulum in the sac. In our series, one baby had UCH with esophageal atresia and tracheoesophageal fistula — an association not previously reported in the literature.
We also observed one case with the appendix adhered to the sac — a rare variant.

Unlike omphalocele, UCH is associated with fewer congenital malformations, usually gastrointestinal, such as small and large bowel atresia or stenosis, cloacal malformations, short bowel syndrome, Meckel's diverticulum, and other omphalomesenteric duct anomalies[1,2,10]. Other associations include cleft lip and palate, congenital glaucoma, pulmonary stenosis, and tetralogy of Fallot.

Conclusion

Umbilical cord hernia is a rare congenital defect that can be easily mistaken for

omphalocele minor. Most cases are isolated, with variable sac contents including Meckel's diverticulum, liver, or appendix. Timely surgical repair leads to excellent outcomes, and antenatal recognition is essential to avoid potentially serious iatrogenic injury.

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	1964 Helsinki declaration and its later amendments. Institutional approval was obtained prior to data collection.
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