RUPTURATED CONGENITAL UMBILICAL CORD HERNIA (CHUC) IN NEWBORN OPERATIVELY TREATED 4 HOURS AFTER DELIVERING

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ABSTRACT

Umbilicus is a site of numerous embryopathies involving vessels, urachus, midgut herniation, anterior abdominal wall defects and congenital cysts. Congenital hernia of the umbilical cord (CHUC) is a rare congenital midline (incidence 1:5000) abdominal defect as compared to the umbilical hernia which is much more common and occurs post-natally. Despite its recognition as a distinct entity since 1920s, CHUC is often misdiagnosed as a small omphalocele, resulting in its under-reporting. These masses may be easily overlooked at birth.
In this case, we will present new-born that had congenital hernia of the umbilical cord (CHUC) which was wrapped around its neck, shoulders and upper belly and it ruptured during delivery, exposing in this way small intestines to the external environment. Newborn was operated after admission in Clinical University Centre in Sarajevo (KCUS), four hours after birth. The operation was performed fast and well after quick preoperative preparation (enemas, nasogastric tube, administration of antibiotics and laboratory findings, belly ultrasound, X rays of pulmo and cor) and postoperative course was uneventful.

This is rare case and can be easily be mistaken for ruptured omphalocele or even gastroschisis, but care must be taken in consideration during ligation of the umbilical cord not to clamp intestines that can be present in CHUC that did not rupture. After the rupture, operative treatment must be performed as soon as possible due to exteriorisation of the intestines to external environment.

Keywords: CHUC, Umbilical hernia, Omphalocele.

INTRODUCTION

There are several embryopathies involve umbilicus including midgut herniation, omphaloceles, urachal and vascular anomalies (Pal et al., 2009). During early fetal life there is physiological herniation of a greater portion of the intestines into the proximal part of the umbilical cord, which is called extracelomic cavity. At about 10-12 weeks gestation, intestines withdraw into the abdominal cavity, the umbilical ring mostly closes and the extracelomic cavity disappears leaving behind Wharton’s jelly and umbilical vessels in the cord. In rare instances, the umbilical ring does not close and variable portions of the intestines remain in the extracelomic cavity which present at birth as congenital hernia into the umbilical cord (CHUC) (Pal, 2014).

Unlike omphaloceles and gastroschisis, CHUC has an intact abdominal wall, a complete umbilical ring, a sac comprising of outer amnion and inner peritoneal lining and contains contents varying from loops of intestines to any movable intraperitoneal organs. Also, a cuff of skin extends from abdominal wall onto the neck of the sac.

Congenital hernia of the umbilical cord (CHUC) is a rare congenital midline abdominal defect as compared to the umbilical hernia which is much more common and occurs post-natally. Despite its recognition as a distinct entity since 1920s, CHUC is often misdiagnosed as a small omphalocele, resulting in its under-reporting. These masses may be easily overlooked at birth, which may result in complications like infections and an intestinal injury due to careless proximal application of the cord clamp (Prakash et al., 2006). At the time of birth, anterior abdominal wall defects can present either as gastroschisis or omphalocele. Gastroschisis presents with herniation of the fetal abdominal contents into the amniotic fluid without any covering. Omphalocele, on the other hand, is a congenital anterior abdominal wall defect at the base of an umbilical cord with herniation of the abdominal contents covered by the parietal peritoneum, the amnion and Wharton’s jelly (Pal et al., 2009). Mild forms of omphalocele can present as congenital hernia of the umbilical cord.
(CHUC) with herniation of either the solitary intestinal loop or persistent omphaloenteric duct (Sabzehei et al., 2017). Like all abdominal wall defects in which midgut has not returned to the abdominal cavity before birth to allow rotation and fixation, these patients present with malrotation but it usually does not cause intestinal obstruction. It is quite different from the post-natally diagnosed umbilical hernia and is believed to arise from persistent physiological mid-gut herniation. The content of CHUC can also be Meckel's diverticle, and, rarely, liver or gallbladder.

The incidence of CHUC is estimated to be 1 in 5000. Unlike an omphalocele, it is not linked with chromosomal anomalies but has been loosely associated with intestinal anomalies, suggesting the need for a complete fetal anatomical ultrasound evaluation (Raju et al., 2015).

Very often this anomaly is followed by Meckel's diverticle or anomalies of the gut including ileal or colon atresia that must be excluded during preoperative management, operative treatment and postoperative care. To the worst, inadvertent clamping of cord leads to iatrogenic gut injury in a situation of hernia of cord (Pal et al., 2009).

CASE REPORT

On the night of the August 5th, 2019. After admission of the female newborn from city Travnik, borned around 19:50h, pediatric surgeon was called to consilliary examination. The child was fourth child from third pregnancy, borned by mother aged 37 and APGAR was 9/9. Pregnancy was ordinary, and the child was born in 41st week of gestation. In discharge documentation from hospital in Nova Bila where child was delivered before urgent transport to Sarajevo’s Neonatal Intensive Care Unit in Clinical University Centre in Sarajevo (NICU in KCUS), it was written that umbilical cord was wrapped around its neck, shoulders and upper belly and it ruptured during delivery, exposing in this way small intestines to the external environment (Figure 1).

All other findings were in reference values, so the child was urgently transported to NICU and paediatric surgeon was called to make an examination due to suspicion of the Gastroscisis. The intestines did not seemed as those found in the Gastroschisis cases and we had data about ruptured umbilical cord, so, after immediate preparation (blood laboratory findings, examination made by paediatrician and anaesthesiologist and ultrasound of the belly

**FIGURE 1**
CHILD AFTER ADMINISTRATION TO NICU (NEONATAL INTENSIVE CARE UNIT OF CLINIC FOR PEDIATRIC SURGERY KCUS)

All other findings were in reference values, so the child was urgently transported to NICU and paediatric surgeon was called to make an examination due to suspicion of the Gastroscisis. The intestines did not seemed as those found in the Gastroschisis cases and we had data about ruptured umbilical cord, so, after immediate preparation (blood laboratory findings, examination made by paediatrician and anaesthesiologist and ultrasound of the belly
and X rays of heart and lungs, in order to exclude any other following congenital and chromosomal anomalies), nasogastric tube was inserted, as well as urinary catheter, we checked possibility of bowel atresia by enema where which we got stool, and antibiotics were administrated, and, around midnight, we entered operating theatre and started exploration. We found majority of small bowels out of umbilical ring, but they appeared vital (Figure 2).

![FIGURE 2](image1)

**OPERATION THEATRE - PREPARATIONS FOR THE OPERATIVE TREATMENT**

After making incision in proximal part of umbilicus, we made gentle milking of the content from bowels and returned them into abdominal cavity without any pressure or changes in respiratory status of the child (Figure 3). After that, we closed the defect and created new umbilicus for the child (Figure 4). After the operation, child was returned to NICU due to continuation of the monitoring and reanimation. Next day, child was extubated, conscious, on oxygen, was not febrile, had normal urine output, had several stools on enema and slowly, per oral intake through nasogastric tube (glucoses) was begun which child tolerated well.

![FIGURE 3](image2)

**LOOK OF THE UMBILICUS AFTER PUTTING SMALL INTESTINES BACK INTO ABDOMINAL CAVITY AND THE DEFECT THAT LEFT**

Next day child was administered to our ward (Clinic for paediatric surgery) and mother started breastfeeding the child who tolerated this intake well and had normal stools. Operatioal wound was also looking satisfying all the time of the hospitalisation. During time on our Clinic, recovery was uneventful and all control examinations were good as well as laboratory findings. Child had no other congenital or chromosomal anomalies so we expected good outcome. After 12 days, we took out skin sutures and child was discharged home with meticulous advises we gave to the parents. (This case report was written after informed
consent of the parents who were explained what it was about and approved that these information could be used for this article.

DISCUSSION

Due to the fact that this entity is rare comparing to other umbilical defects, there are no many published papers regarding this issue. During the third week of intrauterine life, there is a link between fetus intestine and yolk sac through a narrow tube called as Omphalomesenteric duct, which it disappears at end of the seventh week. There is a midgut physiological herniation to cord during fifth and seventh week of pregnancy. At about 10-12 weeks gestation, intestines withdraw into the abdominal cavity, the umbilical ring mostly closes and the extracelomic cavity disappears leaving behind Wharton’s jelly and umbilical vessels in the cord. In rare instances, the umbilical ring does not close and variable portions of the intestines remain in the extracelomic cavity (a small part of the intestine cannot fully return the abdomen and stay in umbilical cord) which present at birth as congenital hernia into the umbilical cord (CHUC) (Pal, 2014; Raicevic et al., 2017; Mirza & Saleem, 2014).

Unlike omphaloceles and gastroschisis, CHUC has an intact abdominal wall, a complete umbilical ring, a sac comprising of outer amnion and inner peritoneal lining and contains contents varying from loops of intestines to any movable intraperitoneal organs. Distinctively a cuff of skin is seen extending from abdominal wall onto the neck of the sac.

FIGURE 4
LOOK OF THE UMBILICUS AFTER PERFORMED OPERATION TREATMENT

Hernia of small intestine and sometimes other viscera to umbilical cord because of lack of full return to intestine is one of the uncommon diseases in infants, and it is less explained in scientific textbooks and sometimes it is confused with small omphalocele; while, there is real defect of abdomen wall in omphalocele, in the hernia of umbilical cord, the anterior abdominal wall is usually normal and umbilical cord base skin and umbilical ring are normal. Herniation to umbilical cord may include a small portion to entire small intestine or associated with a portion of large intestine (Mirza et al., 2015; Haas et al., 2011; Gajdhar et al., 2013; Pal et al., 2009). Rarely other abdominal viscera herniated into the umbilical cord. In one study, even one case of liver and gallbladder herniation to the umbilical cord has been reported (Jona, 1996). Sometimes, loop of small intestine is herniated into the umbilical cord ring and Meckel's diverticulum and Vitelline duct patency can be seen and resection and restoration of umbilical cord must be performed.
Historically, in 1929, for the first time Hempel-Jorgensen (Hempel-Jorgensen, 1929) had reported two cases of this entity in a family and had coined the term ‘familial congenital umbilical hernia’. Subsequently (Tow, 1937) and (Burnin, 1938) described the characteristics and embryogenesis of this entity in couple of cases. Ever since only few case reports (Raju et al., 2015; Pal & Nofal, 2007; Achiron et al., 1995) have been published clearly describing this entity. Possibly poor understanding of CHUC has led to its under reporting. It seems there is a male preponderance (3:1) and association of prematurity (3 out of 4 cases). (Hempel-Jorgensen, 1929; Tow, 1937). Unlike omphaloceles, CHUC is believed to be a simple anomaly without any associated chromosomal or other organ involvements.

There were found persistent vitello intestinal duct (PVID) in one 5 and cloacal anomaly in another case that is the only reported associations so far. There is also found two cases of Meckel’s diverticulum, one case of type I colonic atresia and one case of type IIIb ileal atresia as the associated GI anomalies in the series. Surgical exploration was done in all cases (Pal et al., 2009; Jona, 1996). CHUC occurs at a specific embryological stage and the cause of failure of return of gut into celomic cavity is still obscure.

Animal experiments and clinical scenarios have clearly described the etiopathogenesis of intestinal atresias. Intrauterine mesenteric vascular accidents due to volvulus, intussusception, and internal hernia, constriction of the mesentery in a tight gastrochisis or omphalocele defect have been observed (Louw & Barnard, 1955; Abrams, 1968; Santulli & Blanc, 1961; Nixon & Tawes, 1971; Okmian & Kövamees, 1964).

It is quite possible that in two of cases described in one of the articles, such a vascular accident could have led to intestinal atresia and persistence of intestinal loops in the extracelomic cavity as CHUC. In other two cases a persistent vitello intestinal duct structure (e.g., Meckel’s diverticulum) could have been the cause of CHUC. Therefore we hypothesise that the varied clinical presentation of CHUC may be multifactorial in causation (of arrested withdrawal of physiological herniation of gut).

Achiron et al., 1995 has demonstrated that CHUC occurs at early embryological stage and can be detectable at early 2nd trimester on antenatal USG. The present study did not have any of our cases being diagnosed by antenatal USG (Pal et al., 2009).

Cord hematoma, cyst, omphaloceles and gastrochisis etc. form the differential diagnoses for antenatally detected hernia of the cord. Few of these might disappear before term and rest may persist as CHUC. No specific antenatal intervention has been advocated although a regular follow up is required as cord hematoma carries increased risk of fetal death (Pal et al., 2009).

Therefore CHUC is a distinct entity with varying clinical presentations. Clinically it can be easily distinguished from omphaloceles and gastrochisis. Except in spontaneously reducing varieties, rest would require surgical exploration to rule out atresia or remnants of vitello-intestinal duct. Outcome is excellent due to lack of association with other congenital or chromosomal anomalies (Pal et al., 2009).

The incidence of CHUC is low. Tow had reported incidence of 1 in 5000 births compared to postnatal umbilical hernia, partly because most of these are misdiagnosed as "omphalocele minors" (Raju et al., 2015). Few early reports had shown male preponderance, association with prematurity and familial occurrence. (2) CHUC is usually a benign pathology, where, meconium discharge from the sac through an associated PVID may be
found occasionally (Jona, 1996). Cases with meconium stained liquor and meconium aspiration syndrome in a newborn with CHUC associated with type III A ileal atresia and perinatal gut perforation have been reported. (Pal & Nofal, 2007).

Prenatal ultrasound examinations play a vital role in the diagnosis of fetal anomalies and have thus influenced the management of new-borns at the time of delivery. Accurate prenatal diagnosis of anterior abdominal wall defects is generally possible during routine prenatal ultrasound examinations and these anomalies. Knowledge of characteristic ultrasound features facilitates therapy and prognosis (Van Tuil et al., 2006; Fogata et al., 1999; Hata et al., 1998). Congenital hernia into the umbilical cord occurs at an early embryological stage and can be detectable by fetal ultrasonography as early as the second trimester (Achiron et al., 1995). Despite advances made in prenatal ultrasound diagnostics, CHUC, which is the minimal form of umbilical cord herniation, can go unnoticed. The presence of a single loop of intestine or POD may be quite challenging at the time of delivery (Van Tuil et al., 2006).

Delivery of uncomplicated pregnancies by midwives is common, and new-borns with CHUC pose a high-risk group for herniated structure injuries. These injuries may also go primarily unnoticed. It is only later, after careful evaluation, that their presence may be discovered.

Any unusual thickening of the base of the cord along with the minute fistulous opening to its side should alert the physician to the existence of this anomalies (Jona, 1996). The cord in such patients must be clamped a safe distance away and an early paediatric surgical consultation must be contemplated. Further medical treatment or surgical intervention should be left to the discretion of paediatric surgeon. Neonatal surgical exploration is indicated to rule out associated intestinal atresia.

It is noteworthy that timely surgical exploration and intervention in suspected cases helps to avoid complications and reduce morbidity. Restoration of umbilical cord hernia is usually a simple operation with good result, and its complications are rarely seen.

In terms of taking propylthiouracil drug and association with abnormalities in the fetus, hernia of umbilical cord or omphalocele with methimazole has been reported in some studies and taking the propylthiouracil has been reported safe in the first three months of pregnancy (Hackmon et al., 2012; Laurberg et al., 2014). In another study, there is a risk of birth defect in both of them, but it is severe with methimazole (Carlisle et al., 2007; Bilderback & Rosenblatt, 1946; Mirza & Ali, 2016; Hasaniya et al., 2013).

CONCLUSION

Special care must be taken not to strangulate herniated bowel by clamping the umbilical cord in this cases. Careful inspection of the umbilical cord of every new-born at the time of delivery is essential. Obstetricians, paediatricians and nurses who customarily clamp, manipulate, or shorten the cord in the delivery room or upon arrival in the nursery should be cognizant of this anomaly. Increased awareness and knowledge of such an entity among health professionals is of paramount important in preventing misdiagnosis, inadvertent bowel injury during cord clamping at delivery and associated morbidity and mortality.

Prompt reaction of paediatricians from neonatal intensive care unit and paediatric surgeons is very important in these cases. All other bowel anomalies must be excluded (during preoperative management by ultrasound and by giving enema to see if baby has
meconial stool, and, of course, by the examination of the bowels itself during the operation). As always, good preparation of the child ensures better outcome (preoperative examinations excluding other anomalies, administration of the antibiotics, and insertion of the nasogastric tube). In simple cases (where there is not bowel resection or sepsis included in postoperative period) recovering is rather fast and uneventful – children very quickly restore peroral intake and normal stool output and can be discharged home. In all the cases without other congenital or chromosomal anomalies good outcome is expected.

REFERENCES


