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CLINICAL STUDY

Prenatal and postnatal peculiarities and consequences of intestinal loop patency disorder with expansive giant cystic abdominal mass in a preterm newborn

Ingrid BRUCKNEROVA[1], Dusan DOBOS[1], Jaroslav BIBZA[2], Jozef BABALA[2], Jana BRUCKNEROVA[3]

Neonatal Department of Intensive Medicine, Faculty of Medicine, Comenius University in Bratislava and National Institute of Children's Diseases, Bratislava, Slovakia. **ingrid.brucknerova@fmed.uniba.sk**

ABSTRACT

AIM: To analyse the findings in a newborn (36 gestational weeks, birth weight: 4,030 grams, birth length: 48 cm, Apgar score 7/8/8 points) with prenatal suspicion of intestinal obstruction at the duodenum/jejunum level. The patient required urgent surgery on the first day of life.

RESULTS: Examination of the abdominal cavity confirmed the presence of a cystic mass with a volume of approximately 800 ml, which was located at the site of jejunal atresia. As part of the surgical solution, the cystic formation and the atretic segment of the intestine were resected with subsequent end-to-end jejuno-jejunal anastomosis and Bishop–Koop ileostomy. The histological examination of 3 samples collected confirmed the presence of mucous membrane and smooth muscle.

CONCLUSION: The cyst anatomically communicated with the aboral section of the jejunum, but the lumen of the jejunum was functionally obturated by solid whitish masses. The histological examination confirmed the diagnostic features of a cyst of intestinal origin. The ileum and colon were patent throughout, but of smaller diameter, so a Bishop-Koop relieving anastomosis was indicated. The condition of the child at the age of 9 months was stabilised and surgical closure of the stoma was carried out *(Tab. 1, Fig. 8, Ref. 21)*.

Text in PDF *www.elis.sk*

KEY WORDS: newborn, jejunal atresia, intestinal cyst.

Abbreviations: AS – alimentary system, USG – sonography, EUS – endoscopic ultrasonography

Introduction

Duplications of the alimentary system (AS) are rare congenital lesions that can occur at any level of the gastrointestinal tract from the oral cavity to the rectum. They are usually diagnosed in newborns and children with an incidence of approximately 1 in 4,500 (1). Nevertheless, according to literary sources, to a significant extent, the diagnosis is carried out often in adulthood (2).

The thoracic localisation occurs with a frequency of approximately 4 %, particularly in the right posterior mediastinum (3) which is most often affected. Gastric duplications account for 7 % of the total incidence, and the large intestine is affected in 15 % of cases. Most often, in up to 44 % of cases, duplications of the gastrointestinal tract are located on the small intestine, of which up to half of the cases are located in the area of the jejunoileal transition (2).

According to the relation to the lumen of the alimentary system they are divided into cystic (without communication with the AS) and tubular (communicating with the lumen of the AS) (4).

Several theories about the formation and origin of duplicating cysts are presented in literature. One of the most common is the lumen-recanalizing theory. It explains the origin of duplication cysts before the 12th gestational week, located only on the oesophagus, and small and large intestine, not at other levels of the gastrointestinal tract (5).

The vascular theory assumes the emergence of gastrointestinal duplication to be associated with atresia of the small intestine as a result of an intrauterine vascular accident during early prenatal development (6).

The theory of abortive twinning may explain the occurrence of colorectal tubular duplications and hindgut duplications, which are associated with genitourinary malformations (7).

The split notochord theory is based on the abnormal separation of the growing notochord from the endodermal cells (8). Abnormal

[1]Neonatal Department of Intensive Medicine, Faculty of Medicine, Comenius University in Bratislava and National Institute of Children's Diseases, Bratislava, Slovakia, [2]Department of Paediatric Surgery, Faculty of Medicine, Comenius University in Bratislava and National Institute of Children's Diseases, Bratislava, Slovakia, and [3]National Institute for Cardiovascular Diseases, Bratislava, Slovakia

Address for correspondence: I. BRUCKNEROVA, MD, PhD, Prof, Faculty of Medicine, Comenius University in Bratislava and National Institute of Children's Diseases, Limbova 1, SK-833 40 Bratislava, Slovakia. Phone: +421.2.59371232

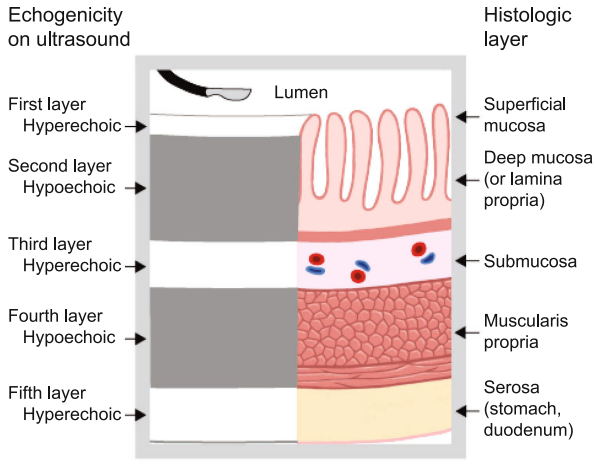

Fig. 1. Sonographic diagnosis of intestinal duplication – classic five-layer cyst wall with alternating hyperechoic and hypoechoic layers – "gut signature sign", which is pathognomonic for the duplication of the digestive tract (13).

splitting or deviation of the notochord is the cause of the persistent connection of the intestine with the dorsal skin and can be clinically manifested by the formation of intestinal duplications associated with enteric fistulas or vertebral/spinal anomalies, which in various combinations are diagnosed as a split notochord syndrome (9).

Intestinal duplications are characterized by a close contact with some part of the gastrointestinal tract they also share a common vascular supply with. Their boundary is formed by a layer of smooth muscle. The epithelial lining consists of a mucous membrane usually belonging to the given part of the AS. However, up to 35 % of duplications may contain ectopic mucosa, most often that of the stomach or pancreas, which significantly increases the patient's risk of ulceration, bleeding and even perforation (10). The diagnosis will be definitively confirmed solely by the histological finding of AS mucosa and muscle layer in the cyst wall (3).

Sonography (USG) has a prominent place with a typical image of a regular multilayer wall of a duplication cyst with a relatively hyperechoic inner layer of the mucosa surrounded by a relatively hypoechoic outer layer of smooth muscle (Fig. 1) (11). However, USG findings can also be irregular and atypical (12).

A modification of the classic sonographic examination, endoscopic ultrasonography (EUS), may have a higher ability to differentiate between solid and cystic lesions (4). EUS can be combined with aspiration diagnostics (fine needle aspiration), which however, increases the risk of iatrogenic infection of the lesion with serious consequences (4). To accurately determine the extent and location of the cyst, especially in the case of intrathoracic localisation, a CT scan (3) or MRI is required.

Duplications of the small intestine are the most common duplications of the gastrointestinal tract. Most of them occur in the ileum. A cystic duplication in close contact but without communication with the lumen of the related intestine is a typical finding. They may be associated with small-bowel atresia and almost a quarter of them contain ectopic gastric mucosa (1). Duplications of the small intestine can remain asymptomatic, or they can present clinically with painful abdominal distension and obstruction of the passage due to volvulus or intussusception (2). In very rare cases, an acute abdomen can be caused by torsion of the intestinal duplication itself, without involvement of the mesentery and without signs of a passage disorder in the sense of an ileus condition (Tab. 1) (14).

The treatment of symptomatic AS duplication consists of surgical resection of the duplication together with the adjacent normal intestine, followed by primary anastomosis. The surgical approach can be open (laparotomy or thoracotomy) or minimally invasive (laparoscopic or thoracoscopic). Asymptomatic lesions diagnosed prenatally or postnatally also require early surgery, due to the high risk of secondary complications (intussusception, volvulus, bleeding, malignant transformation) (15).

Tab. 1. Clinical presentation and differential diagnosis of alimentary tract duplications (2, 8).

Location	Clinical presentation	Differential diagnosis
Foregut	Respiratory distress Obstruction	Tracheoesophageal fistula, Intrathoracic mass
Gastric and duodenal duplications	Haemorrhage Obstruction (ileus)	Pyloric stenosis Gastroesophageal reflux
Gall bladder duplications	Cholecystitis: peptic acid secretion into the biliary system causing signs and symptoms of inflammation, ulceration, obstruction, icterus	Gall bladder atresia and cyst Pre-hepatic icterus
Duplications of the small intestine	Haemorrhage Obstruction (ileus) Intermittent vomiting	Meckel's diverticulum Intussusception Pseudocyst Volvulus Congenital segmental intestinal dilatation Cysts: mesenteric, omental, choledochal, ovarian Cystic teratoma
Colonic and rectal duplications	Constipation Obstruction	Constipation Hirschsprung´s disease Volvulus

Aim of the work

The aim of the presented work is to analyse the findings in a borderline mature newborn (36 gestational weeks, birth weight: 4,030 grams, birth length: 48 cm, Apgar score 7/8/8 points) with prenatal suspicion of intestinal obstruction at the duodenum/jejunum level. The patient required urgent surgery on the first day of life due to a congenital developmental defect of the alimentary system of unclear aetiology.

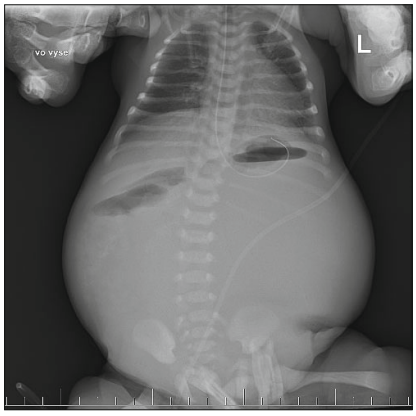

Fig. 2. Native x-ray of the abdomen in anterior-posterior view.

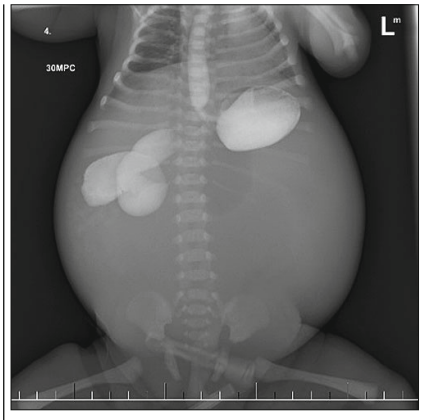

Fig. 3. The passage of alimentary system with stopping of the contrast substance.

Case report

Our patient was the 1st child from 1st pregnancy after *in vitro* fertilization. The suspicion of atresia at the level of the duodenum/jejunum and brain haemorrhage was made at 25th week of gestation. Magnetic resonance imaging confirmed an intestinal loop patency disorder – dilatation of the stomach, cystoid formation in the abdominal cavity ventrally in the mesogastrium with a mixed signal intensity (size: 4.5 x 3.9 x 3.1 cm), no meconium signal in the colon and a focus of haemorrhage in the basal ganglia in the left periventricular *caput nuclei caudati*, without ventriculomegaly, without intraventricular haemorrhage and mildly increased volume of amniotic fluid. Prenatal echocardiography at 25th gestational week was with physiological findings. The delivery was in 36th gestational week using caesarean section because of congenital anomaly of the alimentary system. Amniotic fluid was clear. Group B streptococcus screening was negative. Birth weight was 4,030 g, birth length 48 cm, value of Apgar score 7/8/8 points. A large amount of yellow-green amniotic fluid was aspirated from the oral cavity and upper respiratory tract immediately after birth. Bag-mask ventilation for 30 seconds was followed by non-invasive ventilation with 40 % oxygen for 20 minutes. Subsequently, the child was eupnoeic without the need for oxygenation and ventilation support. Orogastric tube drained a larger amount of clear yellow content. A significant distension of the abdomen was found as part of objective findings. He was normo-cardic, normo-saturated, did not urinate and did not have stool before transport.

A borderline mature newborn was admitted to our department for a suspected congenital abnormality of the alimentary tract at the age of 1.5 hour. The newborn was breathing spontaneously without dependence on oxygen. Neurological status and reflexes were equipped. Abdomen was significantly distended, spilled to the sides, soft on the surface, without palpable resistance and audible peristalsis. The liver and spleen were not enlarged.

Native x-ray of the abdomen revealed poor gas filling of intestinal loops, three bubbles shown in the right *hypochondrium*, without *pneumoperitoneum* and the nasogastric probe ending in the stomach was pushed cranially by a cystic formation in correlation with USG (Fig. 2).

On native x-ray of the abdomen as well as on sonography. an image of homogeneous shading (size: 14 x 12 x 9 cm with a volume of 756 ml) dominated the whole abdominal cavity. The borders of the cystic formation in the caudal areas with an air bubble were detected. This air bubble changes its position, without changing its size as compared to the x-ray. The radiologist concluded this finding to be an air collection in the cystic formation. Collapsed intestinal loops without content were detected.

Subsequently we carried out the imaging contrast study of the alimentary system. Through the nasogastric probe, the stomach was filled with a diluted aqueous contrast substance in a volume of 25 ml. On initial image the contrast substance filled the stomach and part of the duodenum. It took five minutes for the substance to progress to the duodenum/jejunum transition and fill it partially, Loops in the location of the right *hypochondrium* without distension were confirmed. Subse-

Fig. 4. Enlarged abdomen.

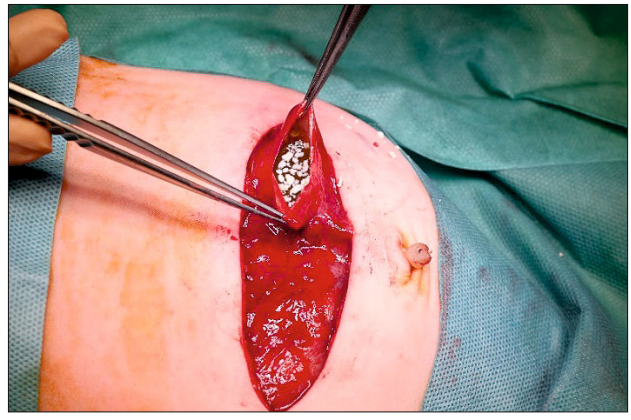

Fig. 5. Opening and evacuation of the cystic mass.

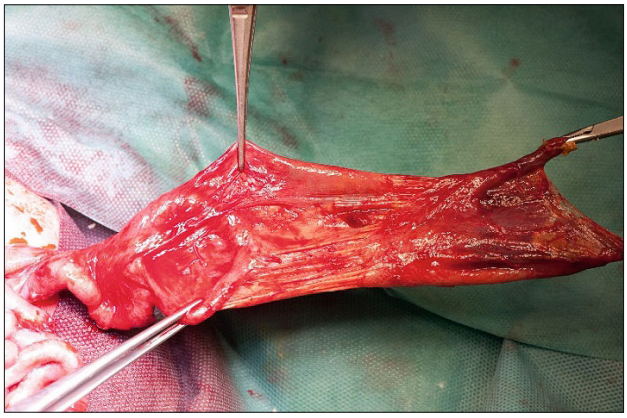

Fig. 6. Dissected and emptied cyst with entering of an atretic oral part of the jejunum and an impassable aboral jejunum.

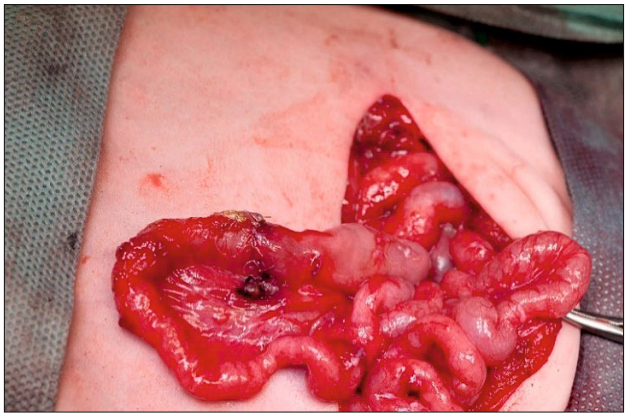

Fig. 7. Jejuno-jejuno end-to-end anastomosis.

quently, after 10 minutes, the duodenum/jejunum transition was completely filled without further passage of the contrast substance into the more aboral parts of the jejunum. The image persisted for 30 minutes and was followed by a reflux of the contrast substance into the oesophagus. The passage was blocked (Fig. 3). A complete obstruction of the passage at the level of the duodenum/jejunum junction as well as malrotation with a leading point (mesenteric cyst or meconium pseudocyst) were suspected.

Sonographic findings of the heart and great vessels, liver, spleen, kidneys, urinary bladder as well as biliary system and brain were negative.

Early surgery was indicated (Fig. 4). The surgeon found a large cystic mass extending from the stomach to the small pelvis. It contained 800 ml of intestinal contents with fatty white particles and bilirubin flakes (Fig. 5). The cystic mass was suppressing the intestinal loops. The oral part of the jejunum entered the cyst. The jejunum was atretic in the site of the origin of the cyst. The aboral part of the jejunum was obturated with solid white masses (Fig. 6). The loops of the small intestine and the colon were compressed by the cyst. The rotation and fixation of intestines was physiological. The cystic mass together with the atretic part of jejunum were resected and a jejuno-jejunal anastomosis was performed (Fig. 7).

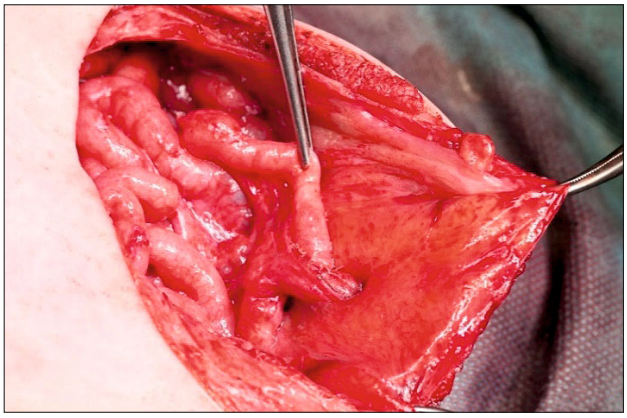

Fig. 8. Bishop-Koop ileo-ileo-anastomosis.

Distal parts of the small intestine as well as the colon were passable with minimal reduction in size. Based on these findings, Bishop-Koop ileo-ileo-anastomosis was carried out (Fig. 8).

Discussion

There are some hypotheses explaining the origin of jejunal cysts, namely the lumen-recanalizing theory, vascular theory, theory of abortive twinning and split notochord theory (5, 6, 7, 8).

In the case of our patient, the jejunum was atretic in the site the origin of the cystic mass, but the aboral part of the jejunum was obturated with solid white masses. The distal part of the small intestine as well as the colon were passable with minimal reduction in size. The wall of the cystic mass had a typical structure of the intestinal wall on histological examination. These findings can be explained in several ways.

Since the aboral parts of the small and large intestine were developed and freely passable, we assume that a perforation of the jejunum could have occurred prenatally before the 25th gestational week, which then healed and gave an image of secondary atresia, and as a result, the weakening of the intestinal wall gave way for cystic formation to develop and start blocking the patency at the site of its origin. This explanation is supported by the result of histological analysis of samples.

Our second hypothesis was based on the colour of the content of the cystic mass, and on assumption that the cystic mass was connected with the opening of the bile and pancreatic ducts in the duodenum. The atresia of the jejunum (oral to the cystic formation) is secondary and initially, the oral intestine was connected to the cystic mass. Atresia probably arose in a short period of time before birth by dislocation of the dilated sac, which exerted pressure on the jejunum. The volume of the amniotic fluid was mildly increased at 25th gestational week and was coloured green and yellow by bile (16). The oral part of the small intestine´s lumen was not dilated on the x-ray on the day of birth. Should atresia occur earlier, the jejunum would have been dilated. This hypothesis assumed that jejunal atresia was secondary to the dislocation of the dilatated cystic sac due to which the intestine became kinked.

Volvulus as a result of a small-sized cyst´s torsion around the mesentery, with a subsequent development of a sudden abdominal event is one of the most serious postnatal complications (17). However, volvulus on the basis of a duplication cyst of the small intestine can also occur in a foetus *in utero* and result in interrupted mesenteric blood supply leading to local ischaemia followed by intestinal stenosis or atresia with postnatal presentation (18). According to authors (18), it can be speculated that early *in-utero* volvulus can lead to the development of intestinal atresia, while volvulus in the later period of pregnancy can cause "only" an intestinal stenosis.

Based on the findings of the width of the intestinal lumen below the atretic part, we can assume that the total intestinal obstruction in our patient could be a consequence of a volvulus developed in an early prenatal period at the time of the intestinal lumen being adequate. The aboral parts of the intestine were optimally passable. However, the findings of fatty druses in the sac of the cystic formation as well as in the aboral part of the intestinal duplication and minimally reduced lumen of the small intestine behind the obstruction support the hypothesis of gradual development of atresia.

Tubular duplications can also be a cause of atresia. Long tubular duplicates with proximal communication with the intestinal lumen are usually poorly drained and a progressive retention of intestinal contents can occlude adjacent segments of the intestine. Distal communication is more common in tubular duplications and more difficult to diagnose than the proximal communication (19).

Intestinal duplication in our patient was atypical in several aspects. It showed several similarities with mesenteric cysts (20). In terms of size, enteral duplication cysts in newborns typically reach 2–5 cm in diameter (21). In our case, the volume of the cyst was up to 800 ml with thick contents mixed with fatty particles. Histological examination confirmed the presence of the intestinal wall in all examined samples. We thereby ruled out the possibility of the cystic mass being a mesenteric cyst.

Conclusion

Despite comprehensive examination of the patient, we were unable to identify the cause of the serious congenital abnormality of the AS. In our patient, it was a condition that required early surgical intervention at the age of 1 day in order to restore the continuity of the small intestine and to remove a huge cystic mass from the abdominal cavity. The patient's clinical condition was stabilized after being discharged from our department at the age of 1 month. The closure of Bishop-Koop ileo-ileo-anastomosis was carried out at the age of 9 months owing to good clinical condition of the patient.

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