

**Clinical Case Seminar**

**A5(1-7)**

# Incidentally discovered enteric duplication cyst: a case report

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## Abstract

Enteric duplications are rare congenital diseases with heterogeneous clinical pictures ranging from an asymptomatic course to life-threatening consequences, most commonly arising at the ileal and ileocecal region. The antenatal discovery is possible when it concerns a voluminous cystic form enabling an early management. The radiologic examinations are nonspecific and no diagnostic tools can allow a certain diagnosis on its own. Sometimes, the diagnosis of intestinal duplication is only made during the surgical exploration and confirmed after a histopathological examination.

We report a 4 years old girl with antenatal diagnosed ovarian cystic mass of about 4 mm. She was admitted to our unit for abdominal pain and constipation. Abdominal ultrasonography showed a cystic mass in the right iliac fossa. MRI revealed a well-defined cystic mass (6 x 4.2 x 5.4 cm) in the right mid abdomen displacing the bowel to the left, likely to be strongly adherent to the last part of the ileum. 99mTc pertechnetate scan was negative for ectopic gastric mucosa. A laparoscopic approach was eventually necessary and the diagnosis of duplication cyst was confirmed.

Children with antenatal diagnosis of abdominal mass need a close follow up and enteric duplication should be considered as potential diagnosis. The laparoscopic approach has an important role in differential diagnosis between intestinal duplications and other mass.

**KeyWords:** enteric duplication, ileocecal resection, ileocecal valve/junction, abdominal mass

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## Introduction

Enteric duplications (ED) are rare congenital diseases occurring in 4500-12.500 live births (1), more frequent in males (2).

They have heterogeneous clinical presentations ranging from an asymptomatic course to life-threatening consequences (3, 4).

## Clinical Case

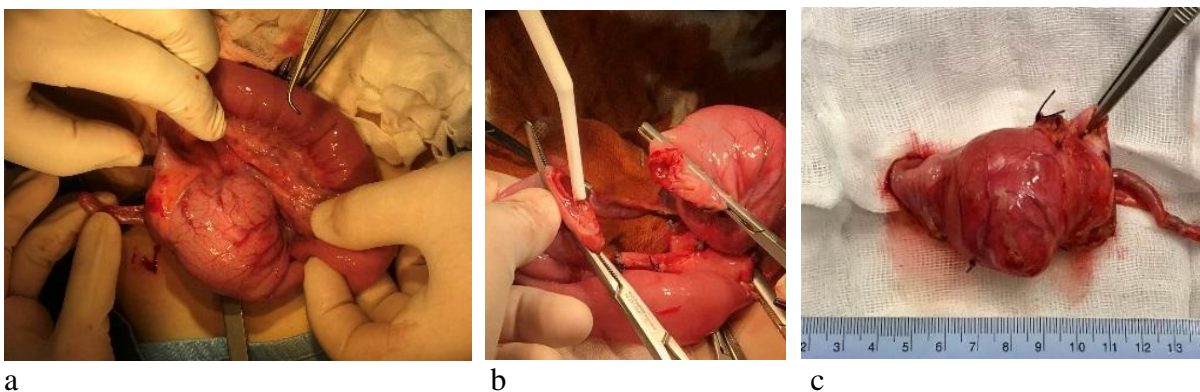
A 4 years old girl was referred at our Institution with the ultrasonographic diagnosis of a 70mm cystic mass in right iliac fossa and an history of constipation. She had an antenatal diagnosis of

ovarian cyst of about 4 mm and no other relevant symptoms in the postnatal period. Because of referred poor weight gain, constipation and recurrent abdominal pain at age of one she was been investigated for celiac disease and cystic fibrosis with negative results. During a routinary clinical evaluation by General Practioner, an abdominal mass was found in the right hypochondrium and she was referred to the Pediatric Emergency Department where she underwent an ultrasonographic evaluation and was admitted at Pediatric Surgery Department. During the hospitalization, additional investigations were performed.

A MRI revealed a well-defined mass (6 x 4.2 x 5.4 cm) in the right mid abdomen. The mass displaced the bowel to the left and anteriorly and appeared to be strongly adherent to the last part of the ileum. 99 Tcm-perechnetate scanning showed no ectopic gastric mucosa.

A diagnostic laparoscopy was planned: a large cystic mass occupied the inferior-right quadrant of the abdomen. Pelvic exploration allowed excluding adnexal mass. The cyst was laparoscopically mobilized and a median sub-umbilical incision (2 cm) was made to bring out the mass. The mass shared a common muscular wall with the terminal ileum, close to the valve and compressed the cecum. The cecal rim was not clearly evident so a segmental bowel resection was necessary with the loss of ileo-colic valve (Fig. 1)

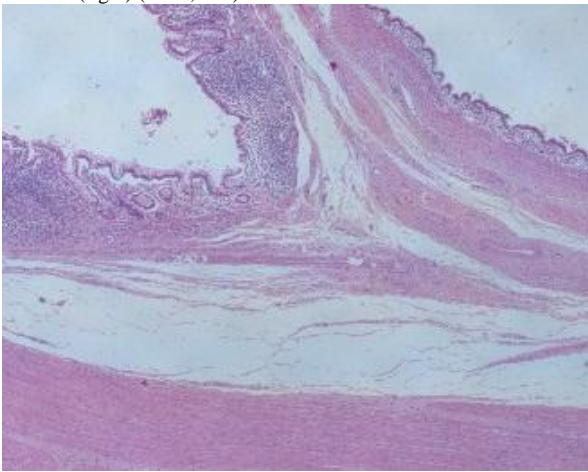
**Fig. 1** Intraoperative view (a and b) and view of the excised mass (c)



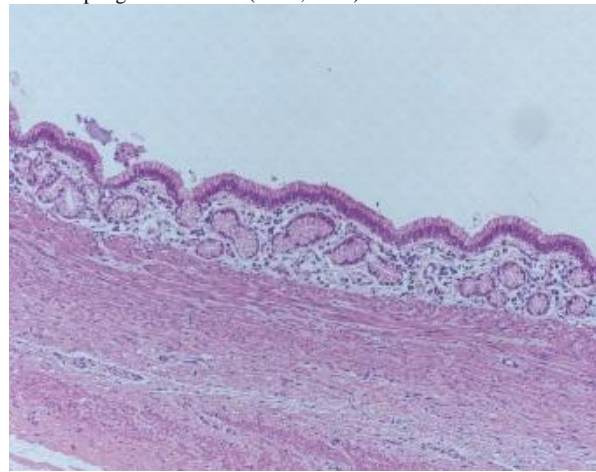
The histopathology examination revealed a cystic mass contained stained fluid, compatible with a duplication cystic mass. The mucosa of the cyst included heterotopic immature gastric mucosa(Fig. 2) better evidenced at magnification (Fig. 3).

The post-operative course was uneventful .

**Fig.2** Double lumen sharing a common muscular wall. Columnar ileal epithelium with goblet cells (left), immature heterotopic gastric mucosa (right) (H&E, 50x)



**Fig 3.** A higher-power magnification shows immature heterotopic gastric mucosa (H&E, 100x)



## Discussion

The term “digestive duplications” was firstly used by Calder in 1733s and later by Fitz in 1884 even if it was not widely used until it was popularized by Ladd in 1937 (5, 6) . It refers to cystic or tubular formations in intimate contact with the various segments of the alimentary canal (3) (Table 1). They can be located at any level of the gastrointestinal tract, but most commonly at the mid or terminal ileum (2, 7). They can present at any age but about 80% of cases present within first two years, mostly during the first three months of life (8, 9). They are thought to arise from disturbances in embryologic development. Multiple theories have been postulated to account for their development even if to date no single theory has explained the origin, various locations involved, and the associated anomalies of the duplication cyst. Bentley and Smith (10) proposed that the primary defect was the development of a split notochord that connect the yolk sac endoderm to the ectoderm and that subsequent duplication of the gut resulted from eventration of the yolk sac between the halves of the vertebra. Other authors suggested that a abnormal recanalization of the gut could explain the pathogenesis of ED (11).

Parker et al described duplication of the alimentary tract to form a cystic or spherical structure attached to a part of the bowel, sharing a wall of smooth muscle and lined by a mucous membrane similar to some part of the alimentary canal. ED symptoms varies according to size, location, type of duplication, and presence of heterotopic mucosa. Prenatal ultrasonography can detect ED as early as 16 weeks of gestational age (12). In some case ED may be completely asymptomatic and they are identified on routine physical examination or during incidental investigations (13).

**Table 1.** Prevalence in % of duplications for morphology, sex and localization.

TOTAL CASES	% CISTIC	% TUBULAR	Morphology Nd
220	80 %	14 %	6 %

TOTAL CASES	F	M	Sex Nd
220	40 %	52 %	8 %

TOTAL CASES	FOREGUT	MIDGUT	HINDGUT
220	25,45 %	64,10 %	10,45 %

Nd = not described

In our case the localization of ED was at IC region. ED at the IC junction is considered a particular entity, to be distinguished by other ileal ED as such. Only few Authors described the IC type, for which the operative approach consisted of segmental resection with primary anastomosis (14). In particular Puligandla et al. examined the duplications found in the IC valve region as an independent entity from the duplications found in the ileum and described a frequency of 30,2% and 31% respectively, reporting the largest series (15). In our case pain and constipation were the only symptoms and the mass was incidentally found during a routine medical examination. The preoperative diagnosis duplication of cysts are often inaccurate (16). USG is the imaging modality of choice for the evaluation of an abdominal mass in the neonate which can demonstrate nature and location of the mass. Using scans during pregnancy result in a higher rate of antenatal detection of duplications which allow early treatment and avoidance of possible complications (17). Ultrasound plays a crucial role in the diagnosis of intestinal duplication because it identifies the cyst and its anatomical location (18). CT scans are more useful in demonstrating the precise anatomical relationship between the cysts and surrounding structures (19). These cysts can manifest as smooth, rounded, fluid filled cysts or tubular structure with thin slightly enhancing wall on CT scan. Magnetic resonance imaging (MRI) and endoscopic ultrasonography are other diagnostic modalities (20). Radioisotope scanning is useful for evaluation of bleeding from these cysts. However, all these modalities allow us only to suspect the presence of an abnormal lesion and diagnostic confirmation is possible only after resection. In our case there was an antenatal diagnosis of presumptive ovarian cyst. Abdominal ultrasound and MRI revealed the presence of the mass but the differential diagnosis between ED

and other mass was not reliable. Also Technetium-99m pertechnetate scanning was negative for heterotopic gastric mucosa. Histopathological examination enabled us to confirm the diagnosis and revealed the presence of immature heterotopic gastric mucosa. Usually, the treatment of choice for cystic masses in children is complete surgical resection, through laparotomy, laparoscopy or laparo-assisted surgery (11, 21). Total resection when possible should be the aim of the intervention because the partial excision contains high risk of recurrence (22). Due the risk of potential complications, ED should be always excised, even if they are asymptomatic (22,23,24).

Complications include intussusception, intestinal obstruction and ulceration, perforation, and hemorrhage, due to the presence of gastric mucosa.

Malignant changes can occur in the mucosa of an ED (25). In the literature, from 1955 to 2012 have been reported 67 cases of malignancies arising from ED most involving the large intestine and rectum. The age of presentation ranged from 12 to 88 years old, but most patients were between the ages of 40 and 60. Female predominance of 3:1 is found in the colorectum site. Malignant transformation should be suspected if any abnormal solid component is found within the duplication or serum CEA or CA19e9 level is elevated. Indeed, although CA19e9 is not clearly associated with malignancy, the prognostic value of CA19e9 levels in colorectal cancer has been reported and could be of interest in the diagnosis and management of intestinal duplications. More authors think that serum levels of CEA may serve as a valuable index for predicting tumour progress arising from gastrointestinal duplication (26). If malignant change is found in small bowel duplications, the high rate of lymph node metastases should be considered. Curative resections could hardly be performed. The prognosis is generally poor once malignant change has occurred (27).

## **Conclusions**

ED are rare congenital lesions, which may present with nonspecific symptoms. It is important to make a definitive diagnosis of this rare congenital anomalies since they can lead to significant complications as perforation, obstruction, hemorrhage and malignancy.

In children with an antenatal diagnosis of mass of unknown origin, ED should be considered. These children should be followed-up and the persistent of the mass is indication for adjunctive diagnostic evaluations. The radiologic examinations are nonspecific and none diagnostic tools can allow a certain diagnosis on its own. Technetium-99m pertechnetate scanning can result negative if heterotopic gastric mucosa shows immature features. For this reason, the utility of

scintigraphic scanning should be reviewed in the diagnostic course. Surgical approach should be achieved in order to localize the mass and to clarify the diagnosis even in asymptomatic case, to avoid eventual future severe complications.

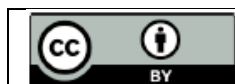
**Conflicts of Interest:** All the authors have no conflict of interest to declare.

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