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Alimentary Tract Duplication in Pediatric Patients: Its Distinct Clinical Features and Managements

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Conflict of Interest
The authors have no financial conflicts of interest.

ABSTRACT

Purpose: Alimentary tract duplication (ATD) is a rare congenital condition that may occur throughout the intestinal tract. Clinical symptoms are generally related to the involved site, size of duplication, or associated ectopic mucosa. This study aimed to identify clinical implications by anatomical locations and age group and then suggest a relevant management according to its distinct features.
Methods: We retrospectively reviewed the clinical data of pediatric patients who received a surgical management due to ATD. Furthermore, data including patients' demographics, anatomical distribution of the duplication, clinical features according to anatomical variants, and outcomes were compared.
Results: A total of 25 patients were included in this study. ATD developed most commonly in the midgut, especially at the ileocecal region. The most common clinical presentation was abdominal pain, a sign resulting from intestinal obstruction, gastrointestinal bleeding, and intussusception. The non-communicating cystic type was the most common pathological feature in all age groups. Clinically, prenatal detection was relatively low; however, it usually manifested before the infantile period. A laparoscopic procedure was performed in most cases (18/25, 72.0%), significantly in the midgut lesion (p=0.012).
Conclusion: ATD occurs most commonly at the ileocecal region, and a symptomatic one may usually be detected before the early childhood period. Surgical management should be considered whether symptom or not regarding its symptomatic progression, and a minimal invasive procedure is the preferred method, especially for the midgut lesion.

Keywords: Alimentary tract; Duplication; Pediatric; Ileocecal; Minimal invasive; Midgut

INTRODUCTION

Alimentary tract duplication (ATD) is a rare congenital condition that may occur throughout the whole intestinal tract [1-3], with a reported incidence of one in 4,500 live births and a slight predominance among the male sex [4-6]. It was described first by Ladd in 1937 [4]. It is usually identified as a cystic or tubular structure attaching to a certain portion of the alimentary tract and thus could be manifested as various signs or symptoms according to the location and appearance of lesion. It is characterized by well-defined muscular and mucosal layers like those of a normal gastrointestinal tract. Duplication is often diagnosed during

the antenatal period or incidentally found during studies for unrelated conditions. In the majority of the cases, the symptoms are noted in early childhood. Approximately 70% of the patients are less than 2 years old; however, the condition may remain asymptomatic until adulthood [7,8].

There are several postulations to explain the development of duplications, including the split notochord, luminal recanalization, incomplete partial twinning, persistent embryonic diverticula, and intrauterine vascular accident theories [5,9-11]. The clinical symptoms are generally related to the involved site and size of duplication. The presence of ectopic mucosa in the lesion could cause associated symptoms secondary to mucosal ulceration and bleeding. Complete surgical removal of the duplication is required in most cases with the goals of not only symptomatic alleviation but also reduction in the risk of symptomatic complications and possibility of malignant degeneration.

There have been no recent published series of ATD that may offer guidance regarding appropriate therapy. Therefore, in this study, we retrospectively reviewed cases of ATD at Pusan National University Children's Hospital to identify the clinical implications and suggest appropriate management.

MATERIALS AND METHODS

Subject selection

This was a retrospective review of clinical data from pediatric patients (before the age of adolescence [below 16 years old]) who received management of ATD between January 2008 and December 2018 at Pusan National University Children's Hospital. There were 32 patients with ATD based on the electronic medical record; among these, 25 underwent surgical management and were with pathological confirmation. This study was approved by the Pusan National University Yangsan Hospital Istitutional Review Board (IRB No. 05-2019-160), and the data were managed with personal information protection.

Data extraction and analysis

We collected data regarding the patients' demographic characteristics, anatomical distribution of the duplication, clinical features according to anatomical variants, and postoperative outcomes.

Statistical analyses were performed using IBM SPSS Statistics ver. 23.0 software (IBM Co., Armonk, NY, USA). Fisher's exact tests were performed to identify significant associations. A p-value <0.05 was considered statistically significant.

RESULTS

Demographic findings

No difference in sex was noted. Most patients were infants younger than 12 months (mean postnatal age, 5.5 months old; 36.0%), followed by neonate (mean postnatal age, 6.1 days old; 28.0%), early childhood (mean, 25.9 months old; 28.0%), and late childhood (mean, 92.8 months old; 8.0%) (**Table 1**).

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Table 1. Demographic findings

Variable	Patients (n=25)
Sex	
Male	12 (48.0)
Female	13 (52.0)
Postnatal age at initial diagnosis	
Neonate (1–30 d)	7 (28.0)/ 6.1±3.9
Infant (1–12 mo)	9 (36.0)/ 5.5±2.1
Early childhood (12–60 mo)	7 (28.0)/25.9±12.7
Late childhood (>60 mo)	2 (8.0)/92.8±55.7

Values are presented as number (%) or mean±standard deviation.
d: days, mo: months.

Table 2. Distribution of duplication according to the location and age groups

Location	Patient (n=25)	Age groups			
		Neonate	Infant	Early C	Late C
Foregut	2 (8.0)				
Esophagus	-				
Stomach	2 (8.0)	2			
Midgut	20 (80.0)				
Duodenum	2 (8.0)	1			1
Jejunum	-				
Ileum	13 (52.0)	2	6	5	
Cecum	5 (20.0)	2	1	2	
Hindgut	3 (12.0)				
Rectum	1 (4.0)				1
Anal canal	2 (8.0)		2		

Values are presented as number (%) or number only.
C: childhood.

Distribution of duplication

Duplication developed most commonly in the midgut in all age groups, especially in the ileocecal region (72.0%). It also occurred in the foregut and hindgut. Moreover, foregut lesions were identified only in neonates. Although it is extremely rare, two cases of anal canal duplication were identified only in infants (**Table 2**).

Clinical features and its outcomes

The most common clinical presentation was abdominal pain, with signs and symptoms resulting from intestinal obstruction (7/25, 28.0%). Presentations also included gastrointestinal bleeding, intussusception, and incidental findings. Some cases were detected during the prenatal period (5/25, 25.0%); however, most cases were detected after birth. A laparoscopic procedure was performed in most cases (18/25, 72.0%), especially for lesions in the midgut. In gross appearance, a cystic form of duplication was common (20/25, 80%); however, lesions in the hindgut were tubular forms only. Pathologically, communication between the duplication and adjacent normal bowel was identified in six cases (24.0%), and an ectopic mucosa in duplication was noted in four cases (16.0%). Associated anomaly, such as congenital diaphragmatic hernia, atrial septal defect, and malrotation, were combined. There were no sequelae after surgery (**Table 3**).

DISCUSSION

This study was conducted based on patients with pathological confirmation and revealed its clinical features of ATD according to the anatomical location and age groups. Although its etiology is currently unknown, it is believed to occur between the fourth and eighth

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Table 3. Clinical features of alimentary tract duplication

Clinical variable	Foregut (n=2)	Midgut (n=20)	Hindgut (n=3)	Total (n=25)
Symptom and sign				
Abdominal pain	1	5	-	6 (24.0)
Vomiting	-	1	-	1 (4.0)
Gastrointestinal bleeding	-	2	-	2 (8.0)
Intestinal obstruction	-	7	-	7 (28.0)
Abdominal mass	-	-	1	1 (4.0)
Intussusception	-	3	-	3 (12.0)
Others	1 (incidental)	2 (prenatal cyst)	2 (ectopic anal opening)	5 (20.0)
Prenatal diagnosis				
No	2	15	3	20 (80.0)
Yes	-	5 (3 cystic masses, 1 mesenteric cyst, 1 renal cyst)	-	5 (20.0)
Surgical procedure*				
Conventional open Laparoscopic	2	3	2	7 (28.0)
Gross type	-	17	1	18 (72.0)
Cystic	2	18	-	20 (80.0)
Tubular	-	2	3	5 (20.0)
Pathology				
Communication	1	3	2	6 (24.0)
Ectopic mucosa	2	2	-	4 (16.0)
Associated anomaly	1 CDH 1 ASD	1 malrotation	-	3

Values are presented as number only or number (%).
CDH: congenital diaphragmatic hernia, ASD: atrial septal defect.
*p=0.012.

weeks during development. It may present clinically at any age with a high incidence during the infantile period [7,8,12]. This study showed also similar findings with a high incidence before the infantile period with a median age of 5.5 months (64.0%). Prenatal diagnosis using ultrasonography has been commonly widespread since the mid-1990s [13-15] and has shown a result between 13.8% and 31.8% [16]. It was not so different in the present study (5/25, 25.0%) although there is a possibility of improvement in diagnosis by prenatal ultrasonography. Considering its congenital origin and relatively low detection rate during the prenatal period, this lesion is assumed to have characteristics of progressive disease. In our study, the cystic type was common (80.0%), and communication was relatively not common (24.0%). This implies that the ATD lesion, especially the cystic type with non-communication to the adjacent normal bowel, generally tends to increase in size over time and cause related symptoms. This may affect the occurrence of clinical manifestation in the early stage, especially before the infantile period. Our finding of the most common location of duplication being the ileocecal region (72.0%) was similar to the results from previous reports [3-5,17-19].

Clinically, patients with duplications present with nonspecific gastrointestinal symptoms varying from abdominal pain to gastrointestinal bleeding; however, symptoms associated with intestinal obstruction were most common. This may depend on the size, location, type, and presence of ectopic mucosa. Symptoms associated with intestinal obstruction, intussusception, and gastrointestinal bleeding were also identified in this study. They were cystic lesions of the midgut that were confirmed pathologically to contain ectopic gastric mucosa. By contrast, the duplication could be discovered incidentally in the absence of symptoms. For this reason, the inaccuracy of preoperative diagnosis is not uncommon.

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Nevertheless, radiologic examinations such as ultrasonography and computed tomography may be useful for detection [20-22]. These were the primary imaging modalities, and ultrasonography is also useful in the prenatal period. Contrary to expectations, we found that prenatal diagnosis is not so common (25.0%) in the present study; nevertheless, a continuous upward trend in the prenatal diagnosis rate may be occurring [23,24]. In any case of asymptomatic ATD, there is a controversy regarding surgical versus nonsurgical management. However, it appears that surgical management may be appropriate because of the clinical characteristics we reviewed and a possibility of developing symptoms during the early period. In particular, duplications carry potential for malignant change although this is usually relevant to adult, not to pediatric, patients [25].

Surgical treatment was usually required for ATD in this study according to its clinical presentation. Conventional surgical procedures were commonly performed for the foregut and hindgut lesions, and laparoscopic procedures were used for the midgut lesions. This usually depends on the location and type of lesion and show difference between previous studies [17,18]. Minimally invasive techniques are recent strategies for management [26]. The procedures usually involve bowel resection; otherwise, simple enucleation or removal of cyst is available for the foregut lesions [26-28]. For all cases in this study, a segmental resection including the lesion was performed. The postoperative course was uneventful. This suggests that a laparoscopic procedure may be the preferred approach if there are no absolute contraindications.

There are a few limitations of this study. It is a retrospective study conducted at a single institution, with a small population of patients during a limited period. Although our study did not reflect all conditions of ATD, we report the clinical implications of disease-specific progression and may suggest a proper surgical method with regard to the change of trend as well as directions for studies in the future.

In conclusion, ATD occurs most commonly at the ileocecal region despite at any site of the gastrointestinal tract. Symptomatic duplications are usually detected before the early childhood period, especially in infants. We should consider a surgical management irrespective of the presence of symptoms because its main pathological feature is a non-communicating enteric cyst and there is the risk of its symptomatic progression after. If possible, minimally invasive procedures are preferred because of its efficacy, especially for the midgut lesion.

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REFERENCES

1. Lund DP. Alimentary tract duplications. In: Grosfeld J, O'Neill J, Coran A, Fonkalsrud E, eds. Pediatric surgery. 6th ed. Philadelphia: Mosby, 2006:1389-98.
CROSSREF
2. Patiño Mayer J, Bettolli M. Alimentary tract duplications in newborns and children: diagnostic aspects and the role of laparoscopic treatment. World J Gastroenterol 2014;20:14263-71.
PUBMED | CROSSREF

3. Schalamon J, Schleef J, Höllwarth ME. Experience with gastro-intestinal duplications in childhood. Langenbecks Arch Surg 2000;385:402-5.
PUBMED | CROSSREF
4. Ladd WE. Duplications of the alimentary tract. South Med J 1937;30:363-71.
CROSSREF
5. Sharma S, Yadav AK, Mandal AK, Zaheer S, Yadav DK, Samie A. Enteric duplication cysts in children: a clinicopathological dilemma. J Clin Diagn Res 2015;9:EC08-11.
PUBMED | CROSSREF
6. Okur MH, Arslan MS, Arslan S, Aydogdu B, Türkçü G, Goya C, et al. Gastrointestinal tract duplications in children. Eur Rev Med Pharmacol Sci 2014;18:1507-12.
PUBMED
7. Murty TV, Bhargava RK, Rakas FS. Gastroduodenal duplication. J Pediatr Surg 1992;27:515-7.
PUBMED | CROSSREF
8. Uzun MA, Koksal N, Kayahan M, Celik A, Kilicoglu G, Ozkara S. A rare case of duodenal duplication treated surgically. World J Gastroenterol 2009;15:882-4.
PUBMED | CROSSREF
9. Qi BQ, Beasley SW, Williams AK. Evidence of a common pathogenesis for foregut duplications and esophageal atresia with tracheo-esophageal fistula. Anat Rec 2001;264:93-100.
PUBMED | CROSSREF
10. Macpherson RI. Gastrointestinal tract duplications: clinical, pathologic, etiologic, and radiologic considerations. Radiographics 1993;13:1063-80.
PUBMED | CROSSREF
11. Bremer IL. Diverticula and duplication of intestinal tract. Arch Pathol Lab Med 1944;38:132-40.
12. Karnak I, Ocal T, Senocak ME, Tanyel FC, Büyükpamukçu N. Alimentary tract duplications in children: report of 26 years' experience. Turk J Pediatr 2000;42:118-25.
PUBMED
13. Richards DS, Langham MR, Anderson CD. The prenatal sonographic appearance of enteric duplication cysts. Ultrasound Obstet Gynecol 1996;7:17-20.
PUBMED | CROSSREF
14. Gerscovich EO, Sekhon S, Loehfelm TW, Wootton-Gorges SL, Greenspan A. A reminder of peristalsis as a useful tool in the prenatal differential diagnosis of abdominal cystic masses. J Ultrason 2017;17:129-32.
PUBMED | CROSSREF
15. Laje P, Flake AW, Adzick NS. Prenatal diagnosis and postnatal resection of intraabdominal enteric duplications. J Pediatr Surg 2010;45:1554-8.
PUBMED | CROSSREF
16. Puligandla PS, Nguyen LT, St-Vil D, Flageole H, Bensoussan AL, Nguyen VH, et al. Gastrointestinal duplications. J Pediatr Surg 2003;38:740-4.
PUBMED | CROSSREF
17. Kim DY, Kim SC, Kim IK. Gastrointestinal duplications in childhood. J Korean Assoc Pediatr Surg 2001;7:26-30.
CROSSREF
18. Kim TW, Jung PM. A clinical study of intestinal duplication. J Korean Assoc Pediatr Surg 2004;10:9-16.
CROSSREF
19. Stringer MD, Spitz L, Abel R, Kiely E, Drake DP, Agrawal M, et al. Management of alimentary tract duplication in children. Br J Surg 1995;82:74-8.
PUBMED | CROSSREF
20. Xiang L, Lan J, Chen B, Li P, Guo C. Clinical characteristics of gastrointestinal tract duplications in children: a single-institution series review. Medicine (Baltimore) 2019;98:e17682.
PUBMED | CROSSREF
21. Cavar S, Bogović M, Luetić T, Antabak A, Batinica S. Intestinal duplications--experience in 6 cases. Eur Surg Res 2006;38:329-32.
PUBMED | CROSSREF
22. Lee NK, Kim S, Jeon TY, Kim HS, Kim DH, Seo HI, et al. Complications of congenital and developmental abnormalities of the gastrointestinal tract in adolescents and adults: evaluation with multimodality imaging. Radiographics 2010;30:1489-507.
PUBMED | CROSSREF
23. Kumar K, Dhull VS, Karunanithi S, Chakraborty PS, Roy SG, Ghosh S, et al. Synchronous thoracic and abdominal enteric duplication cysts: accurate detection with (99m)Tc-pertechnetate scintigraphy. Indian J Nucl Med 2015;30:59-61.
PUBMED | CROSSREF

24. Sujka JA, Sobrino J, Benedict LA, Alemayehu H, Peter SS, Hendrickson R. Enteric duplication in children. Pediatr Surg Int 2018;34:1329-32.
PUBMED | CROSSREF

25. Holcomb GW 3rd, Gheissari A, O'Neill JA Jr, Shorter NA, Bishop HC. Surgical management of alimentary tract duplications. Ann Surg 1989;209:167-74.
PUBMED | CROSSREF

26. Guérin F, Podevin G, Petit T, Lopez M, de Lagausie P, Lardy H, et al. Outcome of alimentary tract duplications operated on by minimally invasive surgery: a retrospective multicenter study by the GECI (Groupe d'Etude en Coeliochirurgie Infantile). Surg Endosc 2012;26:2848-55.
PUBMED | CROSSREF

27. Erginel B, Soysal FG, Ozbey H, Keskin E, Celik A, Karadag A, et al. Enteric duplication cysts in children: a single-institution series with forty patients in twenty-six years. World J Surg 2017;41:620-4.
PUBMED | CROSSREF

28. Górecki W, Bogusz B, Zając A, Sołtysiak P. Laparoscopic and laparoscopy-assisted resection of enteric duplication cysts in children. J Laparoendosc Adv Surg Tech A 2015;25:838-40.
PUBMED | CROSSREF