Open Access Journal of Surgery
ISSN: 2476-1346

Juniper Publishers
key to the Researchers

Review Article
Volume 13 Issue 5 - May 2022
DOI: 10.19080/OAJS.2022.13.555874

Open Access J Surg

Congenital Duplications of the Jejunum: An Appraisal of Anatomical, Clinical and Therapeutic Aspects

Zoi Lamprinou*, Elisavet Kanna, Jonida Mene, Ioannis Skondras, Poulcheria Rousou and Alexander Passalides

Second Department of Pediatric Surgery, P & A Kyriakou Childrens' Hospital, Greece

Received: May 05, 2022; **Published:** May 25, 2022

***Corresponding author:** Zoi Lamprinou, Department of Pediatric Surgery, P & A Kyriakou Childrens' Hospital, Greece

Abstrcat

Duplications of the alimentary tract are one of the rarest congenital malformations, and the jejunum represents the second most common site of duplication after the ileum. The clinical manifestations of this condition can vary but can also remain asymptomatic and elude diagnosis. Enteric duplications often require operative intervention, through well tolerated procedures, with preservation of the native blood supply and intestine.

Keywords: Alimentary tract duplication; Congenital malformations; Jejunal cysts; Jejunal duplication

Introduction

Gastrointestinal tract duplications are uncommon congenital abnormalities, with a reported incidence of 1:4500 births15. They can occur along the entire gastrointestinal tract, with most cases occurring in the small bowel, in any of its sections. The ileum represents the majority of cases, followed by the jejunum and they are typically located on the mesenteric border of the intestine. The clinical manifestation can greatly vary from asymptomatic to acute abdomen and the diagnostic approach depends on age, location and clinical presentation. The way of treatment is surgical excision but asymptomatic cases' management can be controversial.

Embryology

Embryogenesis of gastrointestinal tract duplications, in general, is still unclear. They are believed to occur between the fourth and eighth weeks of gestation. Several hypotheses have been postulated but no single hypothesis can provide adequate explanation about their formation. The aberrant luminal recanalization theory can only be applied on the gastrointestinal segments that go through a solid stage such as the esophagus, the small bowel and the colon and cannot explain duplications at other levels. According to this, rapid proliferation of cells leads to the "solid stage" of development. Shortly thereafter, spaces or "vacuoles" appear within the solid lumen and it normally returns to patency. Persistence of a vacuole can result in the development of a cystic or tubular duplication. The exceptional overgrowth of diverticula in the embryonic intestine has also been proposed as well as the abortive twinning theory which claims at duplications represent incomplete twinning but they have both been discredited. Another hypothesis which is also in doubt is the intrauterine vascular accident theory. This one suggests that duplications arise from an intrauterine vascular accident during the early stages of fetal development [1,2]. The most widely accepted theory is the split notochord theory. During the third week of development the endoderm grows dorsally and part of it is pinched off as the notochord, which induces the development of the vertebral column from mesoderm. If this separation is incomplete, remnants of endoderm may be left behind as tubules, cysts, or cords that eventually develop into duplications. Due to these remnants, ventral closing of the vertebral column may be disturbed and may result in a cleft. This theory can also explain why congenital duplications are often associated with vertebral anomalies.

Anatomy

Jejunum begins at the duodenojejunal flexure and has a length of approximately 2,5m in an adult. The arterial supply of jejunum is from the superior mesenteric artery. Jejunal branches pass

through the mesenterium and anastomose with each other and form arterial arcades. The final direct supply is provided by the straight vessels (vasa recta) which are an extension of the distal arcades [3]. Venous drainage is from the superior mesenteric vein and lymphatic drainage is conducted into the superior mesenteric lymph nodes. Blood supply of the duplications derives usually from the surrounding mesenteric vessels and is shared with the native bowel. Nevertheless, in extremely rare cases, can duplications be isolated and have their own blood supply.

Clinical Presentation and Implications

Jejunal duplications may be asymptomatic or present with vague symptoms, mimicking other common pathologies or may accompany other congenital anomalies as well. The mode of presentation usually depends on age, size, presence of heterotopic mucosa within the duplication and communication with the adjacent lumen. In jejunal duplications most common symptoms are bilious vomiting at birth, palpable mass, abdominal bloating, constipation, pain and bleeding. Low gastrointestinal bleeding due to ectopic gastric mucosa can present with varying severity from occult blood loss with anemia to overt symptoms as enterorrhagia and it is more common in communicating tubular duplications. Although, noncommunicating duplications can also cause bleeding (by causing pressure necrosis of adherent mucosa), intussusception or localized volvulus and gangrene [4,5]. Even if the most duplications are identified during childhood, there are some which remain silent and present later on in life. Malignant change is a rare complication of jejunal duplications, presenting during adulthood and only few cases have been reported. The most common histological type observed is adenocarcinoma but other types as squamous cell carcinoma, neuroendocrine tumors and papillary adenocarcinoma have been described in literature [6,7].

Various congenital malformations have been found to be associated with alimentary tract duplications mainly vertebral and genitourinary anomalies. But as it concerns jejunal duplications in particular there are some specific anomalies that have been described such as jejunal atresia [8], intestinal malrotation [9], amelia and congenital diaphragmatic eventration [10]. Complications caused by jejunal duplications include, as mentioned before, perforation, intussusception, bowel obstruction [11,12] and volvulus [13]. It has also been indicated at jejunal duplication may induce the growth of lipoma hence causing symptoms of intestinal obstruction23. In the case of duplication cysts complications include bleeding into the cyst, cystic torsion or rupture or infection of the cyst [12,7,14,13]. Jejunal duplications can be detected either prenatally or postnatally. The differential diagnosis to be considered in a fetus includes neuroenteric cyst, mesenteric cyst, omental cyst and ovarian cyst [15,14]. In neonates and children can mimic other common pathologies such as appendicitis, Meckel's diverticulum [16] and intestinal atresia [17,12,1].

Diagnostic imaging

Detection of jejunal duplications can be made prenatally during the standard detailed second trimester scan at 18 weeks of gestational age with the help of ultrasonography [15]. Laje et al. [16] consider at when an enteric duplication is found on a pregnancy surveillance ultrasound, a prenatal MRI and echocardiogram are indicated to help rule out other associated malformations [16]. In adults, duplications are usually diagnosed incidentally either on routine examination or during the clinical investigation of vague symptoms. At early ages though, they are commonly diagnosed when complications like bowel obstruction, bleeding or perforation occur. The main preoperative diagnostic modalities consist on ultrasonography, barium swallow, computed tomography (CT) and magnetic resonance (MRI). Ultrasonography is considered to be the first-choice imaging modality18and the diagnosis is suggested by the characteristic double-wall sign consisting of a hyperechoic inner layer that is produced by the mucosa and a hypoechoic outer layer representing the muscularis propria [1]. Other characteristics that indicate enteric duplications are peristaltic muscular contractions of the wall and close contact with the mesenteric border. Doppler imaging showing Whirlpool-sign with superior mesenteric vein around superior mesenteric artery is suggestive of midgut malrotation [9].

On CT, jejunal duplications appear as well-defined, rounded, hypodense lesions with a thick, enhancing wall9and on MRI they present with heterogeneous signal intensity on T1-weighted images and homogenous signal intensity on T2-weighted images [17]. In pediatric patients MRI is preferable to the CT scan, although anesthesia or sedation may be needed. Barium examination can be helpful with the diagnosis of tubular duplications, if not contraindicated. If all other investigations are negative, small-bowel evaluation with capsule and balloon enteroscopy can be used but due to their size, have limited application in infants [18]. In cases with suspicion of presence of ectopic gastric mucosa, can radionuclide scanning with Technetium-99m pertechnetate be helpful. EUS-FNA can offer an accurate diagnosis of duplication cysts but also carries an increased risk of complications such as infection. No data has been published about the use of EUS-FNA in diagnosing jejunal duplications [19].

Management

The definitive management of all gastrointestinal duplications is surgical excision. The location, size, presence of ectopic mucosa and benign nature of the lesion must be taken into account. Surgical intervention is generally performed at the age of diagnosis, even in asymptomatic patients, so as to avoid future complications and due to the risk of malignant degeneration [16,8]. However, in asymptomatic newborns and infants diagnosed with jejunal duplications can surgery be delayed until later in childhood according to some authors [15,18]. Delayed intervention may offer a smoother perioperative course and a greater possibility of laparoscopic resection. The operation performed is a complete

How to cite this article: Zoi Lamprinou, Elisavet K, Jonida M, Ioannis S, Rousou P, et al. Congenital Duplications of the Jejunum: An Appraisal of Anatomical, Clinical and Therapeutic Aspects. Open Access J Surg. 2022; 13(5): 555874. DOI: 10.19080/OAJS.2022.13.555874

resection of the duplication as partial excision is associated with high risk of recurrence [11]. Cystic duplications can be definitely managed with simple cystectomy if there is no communication with the adjacent bowel. Nevertheless, in cases with broad-based cysts or tubular duplications it may, additionally, necessitate resection of the native intestine with end-to-end jejuno-jejunal anastomosis due to common blood supply. Partial resection with mucosal stripping is an alternative surgical option in longer tubular duplications or large duplication cysts to avoid short bowel syndrome [15,20]. Laparoscopic excision must be considered as an option when it is feasible as it is associated with low rate of complications, improved postoperative pain control and shorter recovery time. Decision must be taken relied on patient acuity and surgeon's familiarity with laparoscopic techniques [15,1].

Discussion

Jejunal duplications comprise a rare congenital entity and as other gastrointestinal duplications can be life-threatening. After ileum, jejunum is the second most common site of these anomalies but some authors as Liu and Adler [19] claim the reverse. There are two types of duplication, cystic and tubular, but the majority of them have a cystic nature as all authors agree and 25-39% of duplications contain ectopic gastric or pancreatic mucosa [1]. There are many theories about their embryological origin but the split notochord one is the most widely dominant. Duplications can remain asymptomatic for a long period but they usually present with vague symptoms or acute abdomen due to complications. Many authors describe recurrent abdominal pain as a usual symptom of jejunal duplications. The cornerstone of diagnostic approach is ultrasound; however, other imaging techniques can be used in difficult cases. Prenatal diagnosis by US imaging can occur even at the 18 weeks of gestation as Fahy and Pierro [15] support and the rate of prenatally diagnosed duplications can reach 37% in some centers as in Children's Mercy Hospital in Kansas [21]. Treatment of choice is radical excision of jejunal duplication, with or without the adjacent bowel, either through an open surgery or laparoscopically. Nevertheless, excision of asymptomatic duplications is a controversial issue and many authors suggest elective excision or monitoring of them. Fahy and Pierro [15] propose elective excision of the duplications, especially in infancy, as there are concerns regarding neurotoxicity and abnormal neurodevelopment from the anesthetics. Scheduled surgical intervention may be performed after the 6th month of age for antenatally diagnosed duplications, as Okur et al. [18] suggest [18,22,23] (Figure 1).

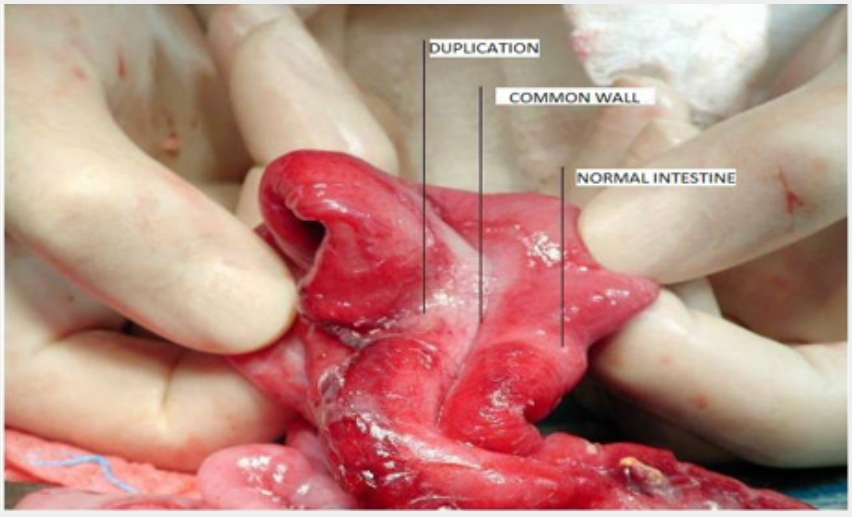

Figure 1: Jejunal duplication as an intraoperative finding during diagnostic laparotomy due to acute abdominal pain.

Conclusion

Alimentary tract duplications and especially those of the jejunum should be included in the differential diagnosis for all patients under 2 years old presenting with abdominal pain or other symptoms from the lower gastrointestinal tract. Our study depicted that the diagnosis is rare in asymptomatic patients and for this reason, clinical doctors as pediatricians and pediatric surgeons must have high suspicion level and proceed to imaging techniques when the diagnosis is doubtful. Surgical excision is the definitive treatment in these cases so we suggest removal of the duplication after diagnosis.

References

1. Patiño Mayer J, Bettolli M (2014) Alimentary tract duplications in newborns and children: diagnostic aspects and the role of laparoscopic treatment. World J Gastroenterol 20(39): 14263-14271.

2. Sangüesa Nebot C, Llorens Salvador R, Carazo Palacios E, PicóAliaga S, IbañezPradas V (2018) Enteric duplication cysts in children: varied presentations, varied imaging findings. Insights Imaging 9(6): 1097-1106.

How to cite this article: Zoi Lamprinou, Elisavet K, Jonida M, Ioannis S, Rousou P, et al. Congenital Duplications of the Jejunum: An Appraisal of Anatomical, Clinical and Therapeutic Aspects. Open Access J Surg. 2022; 13(5): 555874. DOI: 10.19080/OAJS.2022.13.555874

3. Barlow Te (1956) Variations in the blood-supply of the upper jejunum. Br J Surg 43(181): 473-475.
4. Pal K, Al-Dajani A, Mitra DK (2010) Occult enterorrhagia from jejunal duplication causing diagnostic dilemma in an infant. Afr J PaediatrSurg 7(1): 22-24.
5. Pandey S, Srivastava A, Lal R, Yachha SK, Poddar U (2014) Enteric duplication cysts in children: a target in algorithm for evaluation of lower gastrointestinal bleeding. Indian J Gastroenterol 33(3): 285-288.
6. Kim TH, Kim JK, Jang EH, Lee JH, Kim YB (2010) Papillary adenocarcinoma arising in a tubular duplication of the jejunum. Br J Radiol 83(987): e61-64.
7. Ma H, Xiao W, Li J, Li Y (2012) Clinical and pathological analysis of malignancies arising from alimentary tract duplications. SurgOncol 21(4): 324-330.
8. Shakya VC, Agrawal CS, Khaniya S, Koirala R, Pandey SR, et al (2011) Type IV jejunal atresia with an unusual variation of enteric duplication: report of a case. Surg Today 41(1): 130-132.
9. Tripathy PK, Jena PK, Mohanty HK (2016) Isolated Jejunal Duplication Cyst Associated with Intestinal Malrotation in a Newborn. J Neonatal Surg 5(4): 63.
10. Lai CY, Hsu WM, Peng SS, Chou HC, Chen CY, et al (2010) Tubular jejunal duplication, amelia and congenital diaphragmatic eventration in a neonate. ClinDysmorphol 19(3):169-171.
11. Rattan KN, Bansal S, Dhamija A (2017) Gastrointestinal Duplication Presenting as Neonatal Intestinal Obstruction: An Experience of 15 Years at Tertiary Care Centre. J Neonatal Surg 6(1): 5.
12. Khan RA, Wahab S, Ghani I (2016) Neonatal Intestinal Obstruction: When to Suspect Duplication Cyst of Bowel as the Cause. J Neonatal Surg 5(4): 52.
13. Rangaswamy R, Nyuwi KT, Singh CG, Sangtam TA, Varte L (2016) Enteric Duplication Cyst Leading to Volvulus: An Unusual Cause of Acute Intestinal Obstruction - A Case Report. J ClinDiagn Res 10(12): PD20-PD21.
14. Rahul SK, Upadhyaya VD, Kumar B (2016) Malrotation and Midgut Volvulus associated with Asymptomatic Duplication Cyst of Jejunum. APSP J Case Rep 7(4): 33.
15. Fahy AS, Pierro A (2019) A Systematic Review of Prenatally Diagnosed Intra-abdominal Enteric Duplication Cysts. Eur J Pediatr Surg 29(1): 68-74.
16. Laje P, Flake AW, Adzick NS (2010) Prenatal diagnosis and postnatal resection of intraabdominal enteric duplications. J PediatrSurg 45(7): 1554-1558.
17. Blank G, Königsrainer A, Sipos B, Ladurner R (2012) Adenocarcinoma arising in a cystic duplication of the small bowel: case report and review of literature. World J Surg Oncol 10: 55.
18. Okur MH, Arslan MS, Arslan S, Aydogdu B, Türkçü G, et al. (2014) Gastrointestinal tract duplications in children. Eur Rev Med Pharmacol Sci 18(10): 1507-1512.
19. Liu R, Adler DG (2014) Duplication cysts: Diagnosis, management, and the role of endoscopic ultrasound. Endosc Ultrasound 3(3): 152-160.
20. Kachi A, Haddad F, Geagea A, Tohmeh MJ (2016) UNUSUAL CASE OF JEJUNAL DUPLICATION IN A 64-YEAR-OLD FEMALE. J Med Liban 64(1): 50-53.
21. Sujka JA, Sobrino J, Benedict LA, Alemayehu H, Peter SS, et al. (2018) Enteric duplication in children. Pediatr Surg Int 34(12): 1329-1332.
22. Gourishankar A, Tsao K (2019) Acute Painless Lower Gastrointestinal Bleed That Mimics Meckel Diverticulum. Pediatr Emerg Care35(10): e188-e189.
23. Wan XY, Deng T, Luo HS (2010) Partial intestinal obstruction secondary to multiple lipomas within jejunal duplication cyst: a case report. World J Gastroenterol 16(17): 2190-2192.

DOI:10.19080/OAJS.2022.13.555874

How to cite this article: Zoi Lamprinou, Elisavet K, Jonida M, Ioannis S, Rousou P, et al. Congenital Duplications of the Jejunum: An Appraisal of Anatomical, Clinical and Therapeutic Aspects. Open Access J Surg. 2022; 13(5): 555874. DOI: 10.19080/OAJS.2022.13.555874