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Review

Duodenal Duplication Cysts in Children: Clinical Features and Current Treatment Choices

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What Is It about?

- We reviewed English-language pediatric reports of duodenal duplication cysts published over the last 20 years. Duodenal duplication cysts are rare gastrointestinal tract malformations occurring during embryonic development. Most patients experience symptoms in the first decade of life. Clinical presentation is variable, depending on the size and location of the cyst and its relationship with nearby structures. Symptoms commonly include recurrent abdominal pain, nausea and vomiting. Pancreatitis is the most frequent complication. Treatment involves complete surgical resection even for asymptomatic patients. Endoscopic marsupialization is a valid conservative treatment, especially if the anatomical relation with the pancreaticobiliary tract is unclear.

Keywords
Duodenal duplication cyst · Imaging · Endoscopy · Surgery · Pediatrics

Abstract
Background: Duodenal duplication cysts are rare gastrointestinal tract malformations. Most patients experience symptom onset in the first decade of life. This review aims to examine clinical presentation, management strategies and outcomes of duodenal duplication cysts in childhood. ***Methods:*** A Pubmed/Medline (http://www.ncbi.nlm.nih.gov/pubmed/) search in October 2019 for articles published since 1999 using the keywords "duodenal duplication cyst," "child" and "newborn" was carried out. Clinical symptoms, complications, diagnostic examinations, treatment options and outcomes were analyzed and tabulated. ***Results:*** There were 41 citations in the literature providing adequate descriptions of 45 cases of duodenal

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duplication cysts. The age of presentation ranged from newborn to 18 years. The median interval between initial presentation and definitive diagnosis and treatment was 17 months (range: 2 months to 12 years). Overall, 67% of cases presented with abdominal pain, and 43% were complicated with pancreatitis. Different surgical and endoscopic therapeutic strategies were reported. ***Conclusions:*** Duodenal duplication cysts may be associated with life-threatening complications and/or recurrent symptoms, impairing quality of life. Early recognition of patients who demonstrate suggestive signs and symptoms is important to ensure success of treatment. This review may be useful to highlight the main diagnostic aspects and limit the risk of a delayed diagnosis.

Introduction

Gastrointestinal (GI) tract duplication cysts are rare congenital anomalies that occur during embryonic development, in young patients and adults [1]. Criteria for diagnosis are intimate attachment to the native GI tract, smooth muscle coat and alimentary mucosal lining [1, 2]. Several theories have been proposed to explain their development, including the split notochord syndrome (commonly used to explain thoracic duplications, probably due to the incomplete separation of the notochord from the GI endoderm), defects in recanalization involving the neonatal solid GI tract and embryonic diverticulum remnants [2, 3]. However, none of these theories alone is able to explain the heterogeneity of these lesions, and to date the cause of GI tract duplication cysts remains debated. The most common location of GI tract duplication cysts is the distal ileum, followed by the esophagus and ileocecal region, while the duodenal duplication cyst is extremely rare and accounts for only 4% of all GI tract duplications [4]. The estimated prevalence of duodenal duplication cysts is less than 1 per 100,000 live births [5]. A 2010 meta-analysis identified less than 50 published cases of duodenal duplication cysts in both adult and pediatric populations in 10 years [5]. The aim of this review is to report on clinical presentation, management strategies and outcomes of duodenal duplication cysts in children.

Methods

A Pubmed/Medline (http://www.ncbi.nlm.nih.gov/pubmed/) search in October 2019 for articles published since 1999 using the keywords "duodenal duplication cyst," "child" and "newborn" yielded 68 articles published in English-language journals (Fig. 1). These publications were all reviewed. The reference lists of all retrieved articles were manually reviewed to identify any further relevant studies. Articles that did not have a full text available online were excluded. There were 44 reports [5–49] that provided adequate descriptions of 53 cases of duodenal duplication cysts in children and adolescents (Table 1).

Results

Demographics and Clinical Presentation

Of 51 cases with a description of gender, 30 were female. The age of presentation ranged from newborn [9, 19] to 18 years [24, 29]. Most patients (n = 42, 79%) experienced symptom onset in the first decade of life. Outcome was described in 27 patients. There were no deaths or cases of relapsing symptoms during follow-up periods ranging from 3 months to 5 years.

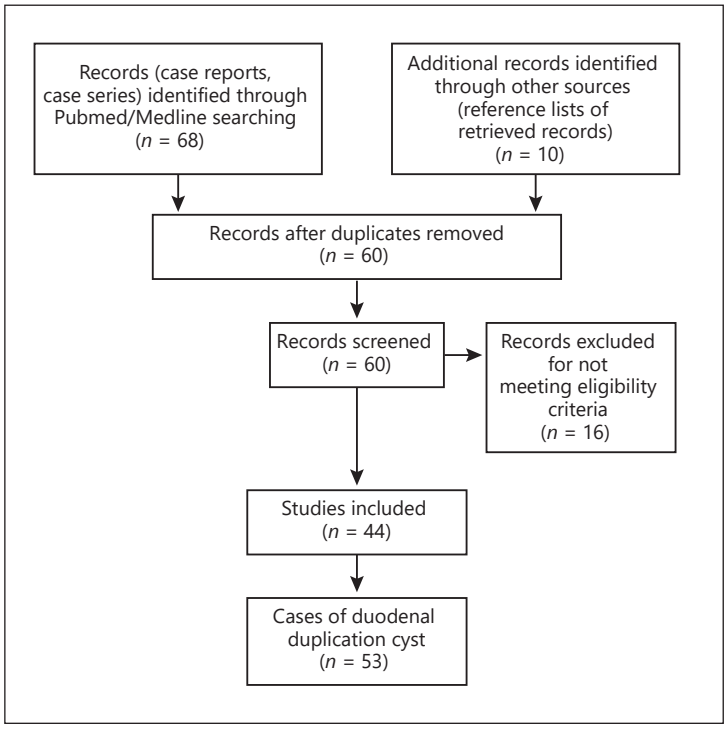

Fig. 1. Flowchart and results of literature review.

The initial presentation of duodenal duplication cyst was variable. The most frequently reported clinical manifestations were abdominal pain (*n* = 33), nausea/vomiting (*n* = 28) and pancreatitis (*n* = 20), either acute (*n* = 9), recurrent (*n* = 10) or chronic (*n* = 1), followed by cholestasis or hepatitis, failure to thrive or weight loss, GI bleeding, fever, cyst infection, gastroesophageal reflux, intussusceptions and stridor. Eight cases had a prenatal diagnosis made after ultrasound (US) [11, 16, 22, 28, 34, 36, 38], whereas one other was asymptomatic and was an incidental finding [24]. The most common complication was pancreatitis, which occurred in 38% of reported pediatric patients. Hepatobiliary involvement, including hepatitis or cholestasis, was also reported in 13% of patients.

Characteristics of Duodenal Duplication

The cyst size varied from 2.0 cm [38] to giant cysts [34, 35]. In 41 cases, the duodenal duplications were located within the duodenal wall (more commonly in the II portion). In the remaining 12 cases, the location of the duplication was variable. Seven were within the pancreas: 5 in the pancreatic head [5, 14, 21, 39, 44], 1 in the aberrant lobe connected with the main pancreatic lobe [45] and 1 in the pancreatic tail [37]. In 5 cases, the duplication cysts were not found within the pancreas. Twenty cysts communicated with the lumen of the native duodenum, and 11 were connected to the pancreaticobiliary ducts (common bile duct, main pancreatic duct). A pancreatic pseudocyst was associated with the duodenal duplication cyst in 3 patients [37, 43, 47]. Some cysts were examined and contained stones, bile, thrombi, blood, clear fluid or turbid fluid. No malignant lesions were found inside them.

Diagnostic Tools

In the reported cases, abdominal US, abdominal computed tomography (CT), magnetic resonance imaging (MRI) and magnetic resonance cholangiopancreatography (MRCP) were the most frequently used diagnostic tools, based on clinical suspicion. Upper GI series and endoscopic retrograde cholangiopancreatography (ERCP) were also performed. The false-

Table 1. Duodenal duplications in children and adolescents

Author, year	Sex	Age		Clinical symptoms	Duodenal duplication					US	Other diagnostic tests	Treatment	Follow-up
		initial presentation	diagnosis/ definitive treatment		location/ size, cm	continuity with duodenum	communication with pancreatic/ bile duct	association with pancreatic pseudocyst	cyst content				
Salazar [6], 2018	NR	15 m	3 years	Abdominal pain, vomiting, AP	Intraduodenal (II portion)/ 5.2×4×1.6	+	Common bile duct	–	Bile-stained fluid	+	MRCP (+), EUS (+)	Endoscopic marsupialization (with an insulated-tip knife)	22 months
Taghavi [7], 2017	M	8 years	17 years	RAP	Intraduodenal (II portion)/ 3×2	+	–	–	Mucus	–	MRCP (1st –, 2nd +), EUS (+), intraoperative cholangiopancreatography	Surgical resection, sphincteroplasty with stent implantation on the terminal pancreatic duct	16 months
Dogan [8], 2016	F	NR	10 years	RAP	Intaduodenal (II portion)/ 1.3×2.1×3.9	+	–	–	NR	+	CT, MRI	Surgical resection	NR
Župančić [9], 2015	M	At birth	16 days	Bilious vomiting, jaundice	Intraduodenal (III portion)/ 2.2×1.4	+	–	–	Fluid	+	Abdominal X-ray, UGI series, MRI, upper endoscopy	Surgical marsupialization	12 months
Thorpe [10], 2015	F	NR	16 years	Abdominal pain, vomiting, AP; duodenal intussusception	Intraduodenal (II portion)/NR	+	Common bile duct	–	NR	–	MRCP (–), CT (heterotaxy), upper endoscopy, intraoperative upper endoscopy	Surgical resection	6 months
Am [11], 2014	M	Prenatal diagnosis by US	At birth	Abdominal lump, jaundice	Intraduodenal (II portion)/ 10×7×7	+	–	–	Fluid, ectopic gastric mucosa	+	CT	Surgical resection	NR
Byun [12], 2014	F	NR	8 years	Intermittent abdominal pain, nausea, vomiting	Intraduodenal (II portion)/ 6×5	+	–	–	NR	+	UGI series, MRI, CT	Surgical marsupialization	NR
Callahan [13], 2013	M	1 year	1 year	Vomiting, weight loss, failure to thrive	Surrounding the pyloric sphincter/NR	–	–	–	Blood	– (intrinsic pyloric obstruction)	Upper endoscopy, UGI series, CT (+)	Surgical resection	NR
Menon [14], 2013	F	18 months	4 years	Intermittent abdominal pain, failure to thrive	Pancreatic head/ 3.4×3×3.3	–	–	–	Altered blood (ectopic gastric mucosa)	– (complex cystic mass in the right lobe of the liver)	CT (+), MRCP (+), duplicated gallbladder), HIDA scan	Surgical resection	NR

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Table 1 (continued)

Author, year	Sex	Age		Clinical symptoms	Duodenal duplication					US	Other diagnostic tests	Treatment	Follow-up
		initial presentation	diagnosis/ definitive treatment		location/ size, cm	continuity with duodenum	communication with pancreatic/ bile duct	association with pancreatic pseudocyst	cyst content				
Yang [15], 2013	F	11 years	11 years	Abdominal pain, hematemesis, dark stools, RAP	Intraduodenal (II portion)/ 2.5×4.8×2.8	–	–	–	NR	– (severe ascites)	Upper endoscopy, MRI, MRCP, CT	Surgical marsupialization	12 months
Palacios [16], 2013	F	Prenatal diagnosis by US	At birth	None	Intraduodenal (I portion)/NR	–	–	–	NR	+	UGI series	Surgical resection	NR
Koffie [17], 2012	M	13 years	13 years	RAP, nausea, vomiting, early satiety, AP	Intraduodenal (II portion)/ 2×2.7×2.9	–	Common bile duct	–	Clear viscous, bile	+	CT, MRCP, MRI	Surgical resection	4 months
Meier [18], 2012	F	9 years	9 years	Abdominal pain, nausea	Intraduodenal (II portion)/ 2.9x2.6	–	–	–	Fluid, white stones	NP	CT, HIDA scan, MRCP	Endoscopic resection	6 months
Mirza [19], 2012	F	At birth	1 year	Abdominal pain, epigastric lump, vomiting	Along the pylorus and duodenum (I portion)/5×5	–	–	–	Mucus, ectopic gastric mucosa	+	CT	Surgical marsupialization	12 months
Rai [20], 2012	F	4 months	1 year	Abdominal pain, abdominal distention, vomiting	Intraduodenal (II portion)/ 6.9×7.5	–	–	–	Mucus, ectopic gastric and pancreatic mucosa	+	Abdominal X-ray, CT, intraoperative cystogram	Surgical marsupialization	NR
Tantemsapya [21], 2010	F	9 years	10 years	Intermittent abdominal pain, vomiting, CP	Pancreatic head/2.7×4.6	+	Common bile duct	–	NR	NP	CT, MRCP, intraoperative cholangiogram	Pancreaticoduodenectomy (Whipple procedure)	2 years
Chen [5], 2010	F	8 years	8.3 years	Intermittent abdominal pain, vomiting, RAP	Pancreatic head/3.5×2.6	–	Main pancreatic duct	–	NR	+	MRI, MRCP, CT, PES, ERCP (failure)	Surgical marsupialization	NR
Chiang [22], 2009	F	Prenatal diagnosis by US	At birth	Abdominal lump	Intraduodenal (I–II portion)/ 4.2×6.8×5.9	–	–	–	Mucus, ectopic pancreatic mucosa	+	MRI	Surgical marsupialization	NR

Table 1 (continued)

Author, year	Sex	Age		Clinical symptoms	Duodenal duplication					US	Other diagnostic tests	Treatment	Follow-up
		initial presentation	diagnosis/ definitive treatment		location/ size, cm	continuity with duodenum	communication with pancreatic/ bile duct	association with pancreatic pseudocyst	cyst content				
Trobs [23], 2009	M	5 years	8 years	Recurrent abdominal pain, nausea, AP, hepatitis	Intraduodenal/3	+	Pancreas through aberrant pancreatic duct	–	Stones	+	MRI, MRCP, CT, intraoperative cholangiography	Surgical marsupialization with cholecystectomy	2 years
Tekin [24], 2009	F	NR	18 years	Recurrent abdominal pain, AP	Intraduodenal/3×2	–	Main pancreatic duct	–	Whitish viscous fluid	NP	MRI, MRCP, CT, PES, ERCP	Endoscopic sphincterotomy with stent implantation	4 months
Ozel [25], 2008	F	8 years	8 years	Abdominal pain, vomiting, AP	Intraduodenal (II portion)/NR	–	–	–	Stones	+	MRI, MRCP, CT	Surgical marsupialization	NR
Antaki [26], 2008	M	5 years	7 years	Abdominal pain, RAP	Intraduodenal/ NR	NR	NR	–	NR	+	UGI series, PES	Endoscopic marsupialization	NR
Antaki [26], 2008	F	15 years	17 years	Abdominal pain, RAP	Intraduodenal/ NR	NR	NR	–	NR	NP	MRCP, PES	Endoscopic marsupialization	NR
Koh [27], 2007	M	18 months	18 months	Asymptomatic (incident finding when surgery for splenic cyst)	Intraduodenal (III portion)/ 5×2×2	NR	–	–	NR	NP	Abdominal X-ray, CT	Surgical resection	NR
Merrot [28], 2006	F	NR	4 years	Hematemesis, bloody stools	Intraduodenal (II portion)	–	–	–	Ectopic gastric mucosa	+	UGI series	Surgical intraduodenal derivation	NR
Merrot [28], 2006	M	Prenatal diagnosis by US	9 months	None	Intraduodenal (II portion)	–	–	–	NR	+	UGI series	Surgical intraduodenal derivation	NR
Merrot [28], 2006	F	Prenatal diagnosis by US	4 days	High intestinal occlusion	Intraduodenal (II portion)	–	–	–	Ectopic gastric mucosa	+	None	Surgical intraduodenal derivation	NR
Merrot [28], 2006	F	NR	9 years	Anemia	Intraduodenal (III portion)	–	–	–	NR	+	UGI series	Surgical resection	NR
Merrot [28], 2006	M	NR	3 years	Abdominal mass	Intraduodenal (III portion)	–	–	–	NR	+	UGI series	Surgical resection and partial duodenectomy	NR

Table 1 (continued)

Author, year	Sex	Age		Clinical symptoms	Duodenal duplication					US	Other diagnostic tests	Treatment	Follow-up
		initial presentation	diagnosis/ definitive treatment		location/ size, cm	continuity with duodenum	communication with pancreatic/ bile duct	association with pancreatic pseudocyst	cyst content				
Guaries [29], 2006	M	NR	18 years	Recurrent abdominal pain, AP	Intraduodenal (III portion)/4	–	Common bile duct	–	Stones	–	UGI series, CT, EUS, MRI, MRCP, PES, ERCP (failure)	Surgical resection with cholecystectomy	NR
Cauchi [30], 2006	M	5 months	10	Fever, abdominal pain, gastrointestinal bleeding, failure to thrive; bilious vomiting, cyst infection	Intraduodenal (IV portion)/6.5	+	–	–	Ectopic gastric mucosa	–/+	UGI series, CT, PES (–)	Surgical resection	2.5 years
Yamauchi [31], 2005	M	5 years	8 years	Fever, recurrent abdominal pain, vomiting, hepatitis; cyst infection	Intraduodenal (II portion)/7×	+	–	–	Purulent	+	CT	Percutaneous cyst drainage and surgical resection	5 years
Prasad [32], 2005	F	2 years	2 years	Abdominal pain, vomiting	Intraduodenal (II portion)/ 6×4×4	–	Pancreas through aberrant pancreatic duct	–	Fluid	+	CT, intraoperative cholangiogram	Surgical resection	1 year
Niehues [33], 2005	M	5 years	17 years	Abdominal pain, vomiting, RAP, cholestasis	Intraduodenal/ 2.5×4	–	Common bile duct	–	NR	–/+	EUS, MRI (1st –, 2nd +), MRCP, PES, ERCP, intraoperative cholangiogram	Surgical resection with cholecystectomy	6 months
Martinez-Ferro [34], 2005	F	Prenatal diagnosis by US	At birth	None	Giant thoraco-abdominal duodenal cyst	–	–	–	Viscous brownish fluid	+	UGI series, CT	Surgical resection	19 months
Wakisaka [35], 2004	F	6 months	6 months	Cough, stridor	Giant thoraco-abdominal duodenal cyst	+	–	–	NR	NP	Chest X-ray, UGI series, MRI	Surgical resection	4 years

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		initial presentation	diagnosis/ definitive treatment		location/ size, cm	continuity with duodenum	communication with pancreatic/ bile duct	association with pancreatic pseudocyst	cyst content				
Khanna [36], 2004	M	Prenatal diagnosis by US (combined with duplication cyst of colon)	At birth	Abdominal lump	Paraduodenal/ NR	–	–	–	Mucus	+	NP	Surgical resection	NR
Kawahara [37], 2002	F	12 years	13 years	Intermittent abdominal pain	Pancreatic tail/ 3×2	–	Main pancreatic duct	+	NR	+	UGI series, CT, MRI, MRCP	Surgical resection	3 years
Messina [38], 2002	F	Prenatal diagnosis by US	8 days	Biliary vomiting, abdominal mass; intestinal occlusion	Intraduodenal (II portion)/2.3	NR	NR	–	Corpuscled	+	UGI series	Surgical resection with cholecystectomy (gallbladder sludge)	3 months
Wong [39], 2002	F	8 years	9 years	Intermittent abdominal pain, vomiting	Between the pancreatic head and the duodenum/2.5	NR	–	–	Fluid	NP	UGI series, MRI, MRCP, CT	Surgical resection	NR
Narlawar [40], 2002	M	5 months	5 months	Bilious vomiting, abdominal lump	Intraduodenal (near the pyloro-duodenal junction)/3×3	–	–	–	Debris, fluid, ectopic pancreatic tissue	+	CT	Surgical resection	NR
Arbell [41], 2002	F	5 months	5 months	Bloody stools, hematemesis, failure to thrive	Intraduodenal (I portion)/ 3.5×3×	–	–	–	Thrombi	+	Upper endoscopy, intraoperative cholangiography, PES (negative)	Surgical resection	3 years
Messina [42], 2001	NR	10 months	8 years	Abdominal pain, vomiting, RAP	Intraduo-denal/3.5	+	NR	–	NR	+	UGI series, MRI	Surgical marsupial-ization	6 months
Keller [43], 2001	F	9 years	9 years	Abdominal pain, AP	Intraduodenal (II portion)/2×4	+	Common bile duct	–	Debris	+	CT, intraoper-ative cholangi-ography	Surgical marsupial-ization	2 years
Keller [43], 2001	F	11 years	11 years	Abdominal pain, vomiting, AP	Intraduodenal (II portion)/ 3.3×.1×3	+	NR	–	Bile	NP	CT	Surgical marsupial-ization	6 months

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		initial presentation	diagnosis/ definitive treatment		location/ size, cm	continuity with duodenum	communication with pancreatic/ bile duct	association with pancreatic pseudocyst	cyst content				
Keller [43], 2001	F	18 months	18 months	Irritability, vomiting, AP, abdominal distension	Intraduodenal (II portion)/ 2.9×2.5	+	NR	+	Clear fluid (amylase, 290)	+	CT	Surgical marsupialization	6 months
Haliloglu [44], 2001	M	NR	18 months	Abdominal pain, intermittent vomiting	Pancreatic head/ NR	–	–	–	Ectopic gastric mucosa	NP	UGI series, upper endoscopy, CT, PES (negative)	Surgical resection	NR
Yang [45], 2000	M	8 years	8 years	Abdominal pain, vomiting, RAP	Adherent to pancreatic aberrant lobe/6×5×4	–	–	–	NR	– (moderate ascites)	UGI series, CT, ERCP	Surgical resection	NR
Lad [46], 2000	F	11 years	12 years	Abdominal pain, vomiting, RAP, hepatitis	Intraduodenal (II portion)/ 3×3×	+	–	–	NR	+	UGI series, CT, PES, ERCP	Surgical marsupialization	NR
Tillig [47], 2000	F	16 months	16 months	Acute abdominal pain, fever, vomiting	Intraduodenal/ 3×3	+	Main pancreatic duct	+	Bile-stained fluid	+	CT, intraoperative cholangiography	Surgical marsupialization	NR
Zamir [48], 1999	M	17 years	17 years	Abdominal pain, diarrhea, hepatitis, duodenojejunal intussusception	Intraduodenal (III portion)/ 4×5	NR	Gallbladder	–	Bile, stones	+	CT, UGI series	Surgical marsupialization	1 year
Mattioli [49], 1999	M	9 years	9 years	Abdominal pain, vomiting, GER, RAP, hepatitis, cholestasis	Intraduodenal/3	+	Pancreatic head	–	NR	–	UGI series, upper endoscopy, CT (+), PES (–, then +), ERCP	Surgical marsupialization	18 months
Mattioli [49], 1999	F	9 years	11 years	Abdominal pain, GER	Intraduodenal/2.5	+	Biliary ducts, pancreas	–	Biliary stones	+	UGI series, upper endoscopy, CT, PES (–), ERCP (failure), percutaneous cholangiography	Surgical marsupialization, sphincterotomy, sphincteroplasty	14 months

US, ultrasonography; NR, not reported; AP, acute pancreatitis; MRCP, magnetic resonance cholangiopancreatography; EUS, endoscopic ultrasonography; RAP, recurrent acute pancreatitis; UGI, upper gastrointestinal; MRI, magnetic resonance imaging; CT, computed tomography; HIDA, hepatobiliary iminodiacetic acid; ERCP, endoscopic retrograde cholangiopancreatography; PES, preoperative endoscopic sphincterotomy; NP, not performed; CP, chronic pancreatitis; GER, gastroesophageal reflux.

negative rates were 24% with ultrasound and 5% with CT scan or MRI/MRCP. The median interval between initial presentation and definitive diagnosis and/or treatment was 17 months (range: 2 months to 12 years). One patient had had other operations before definitive diagnosis [13].

Treatment

Forty patients underwent surgical treatment of the cyst, including 21 surgical resections, 18 surgical marsupializations, 4 cholecystectomy and 1 pancreaticoduodenectomy. Five patients were successfully treated by endoscopic marsupialization [6, 15, 21, 23]. No patients received medical therapy alone.

Discussion

Duodenal duplication cysts represent a minor part of all GI tract duplications. They were first described by Calder in 1733 [2]. The pathogenic mechanism seems to be related to a duodenal epithelial pinching during the outgrowth of the dorsal pancreatic bud or epithelial sequestration [3]. Clinical presentation of the duodenal duplication cyst is highly variable, depending on the size and location of the cyst and its relationship with nearby anatomical structures. The time interval between the onset of symptoms and the diagnosis is relatively long (median >1 year), consisting of diagnostic delay which may be explained by the scarce specificity of symptoms and knowledge of the entity. The most common presenting symptoms include abdominal pain, nausea and vomiting. Pancreatitis is the most frequently reported complication, due to different mechanisms: (a) transient, mobility-related duodenal obstruction of the major papilla outflow by the cyst; (b) compression of the pancreatic duct or hepatobiliary tree by a large cyst [29]; (c) obstruction of the pancreatic duct by migrating biliary sludge or microstones, viscid mucous secretions or shed cyst blood [33, 38]. Acute episodes of pancreatitis usually resolve after bowel rest and medical treatment. A high index of suspicion for anatomical etiologies is mandatory in cases of recurrent pancreatitis. In the case of ectopic gastric mucosa (up to 20% of cases) there could be intracystic hemorrhage or perforation of the cyst with GI bleeding and peritonitis [14, 19, 20, 44]. Intraluminal cysts are difficult to differentiate from choledochoceles (type III choledochal cyst) [17, 21]. Differential diagnosis is only possible on pathological examination, since the duodenal duplication cyst is covered both inside and out with duodenal mucosa containing a distinct layer of smooth muscle.

Diagnostic methods include US, MRI or CT, based on clinical suspicion. Two US signs which are highly suggestive of enteric duplication have been described: the presence of peristalsis and the pathognomonic "double-wall" sign, such as an inner hyperechoic rim correlating with the mucosa-submucosa and an outer hypoechoic layer representing the muscularis propria. US is the elective test also for prenatal diagnosis, which allows close neonatal surveillance [16]. Endoscopic US can be useful to assess the cyst, especially if there is biliary obstruction or pancreatitis [50]. Contrast-enhanced CT scans may demonstrate the location and size of the cyst as well as any accompanying lesions of the pancreatic head. An ERCP is useful to outline ductal anatomy and communication to the main pancreatic duct, especially in planning the surgical approach. MRCP is a valid, noninvasive alternative, especially in small children when ERCP is not appropriate [21]. An intraoperative cystogram may be useful to rule out communication of the cyst with biliary and alimentary tracts [20]. Because of its rarity and variety in terms of clinical presentation and radiological findings, preoperative diagnosis is not always correct [13, 25, 33].

The choice of the best treatment depends on the size and location of the cyst and its relationship with the nearby anatomical structures. Treatment of duodenal duplication cysts

classically involves complete surgical resection, either by local excision or by pancreaticoduodenectomy for the cysts that involve the pancreaticobiliary tract. The close proximity of the major papilla and the associated risk of surgical complications stimulated an interest in treating these cases endoscopically [6]. Endoscopic marsupialization using a needle-knife, sphincterotome, or polypectomy snare, is less invasive than a surgical approach and leads to faster recovery times but has only been used in few selected cases, especially cases in which the anatomical relation with the pancreaticobiliary tract is not clear.

Prognosis is excellent if total excision is achieved and there are no histological elements of malignancy. When complete resection is not feasible, the excision of as much as possible of the duplication and mucosal stripping of the rest are recommended, especially to remove ectopic gastric mucosa [20]. Although a very rare occurrence, duodenal duplication cysts may contain dysplasia mucosa or early malignancy tissue. Ma et al. [51] reported 67 cases of malignancies arising from alimentary tract duplication cysts from a 57-year literature review and identified 3 arising from the duodenum in adult patients. Complete surgical resection is the optimal treatment also for asymptomatic patients, to prevent any complication and the risk of dysplastic or malignant lesions [51]. However, timing of resection in asymptomatic patients is still controversial. Some authors suggest early removal (within the first 6 months of life) because of the high rate of complications that can occur in the first year of life, while others conclude that it is safe to wait until the asymptomatic child is older, when elective surgery is technically easier [16].

This review is limited by the small number of studies available in the literature investigating this relatively rare GI malformation in children and adolescents. Furthermore, the studies that were identified and included in this semiquantitative and narrative review are mainly case series and case reports.

In conclusion, duodenal duplication cysts are rare congenital anomalies but can be associated with life-threatening complications and/or recurrent symptoms impairing quality of life of young patients. Lack of specific signs and symptoms makes diagnosis a challenge even for expert clinicians. For repeated abdominal pain in children and adolescents, a duodenal duplication cyst needs to be ruled out. The choice of the best therapeutic technique depends on the size and location of the cyst and its relationship with the nearby anatomical structures. The ideal goal for the management of duodenal duplication cysts should be to diagnose and treat before the onset of pancreatic complications. This review can guide health care providers towards an early diagnosis and the best therapeutic choice.

Disclosure Statement

The authors have no conflicts of interest to declare. The authors have no financial interests.

Author Contributions

All 8 authors had full access to all study data and take responsibility for the integrity of the data and the accuracy of the data analysis. *Study concept and design:* V.D., P.B., S.F., V.B., P.D., F.M.D., L.D., C.R. *Acquisition, analysis and interpretation of data:* V.D., P.B., S.F., V.B., P.D., F.M.D. *Drafting of the manuscript:* V.D. *Critical revision of the manuscript for important intellectual content:* V.D., P.B., S.F., V.B., P.D., F.M.D., L.D., C.R. *Study supervision:* L.D., C.R.

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