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CLINICAL CASE REPORT

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Safety and Efficacy of Endoscopic Therapy for Nonmalignant Duodenal Duplication Cysts

Case Report and Comprehensive Review of 28 Cases Reported in the Literature

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Abstract: Analyze efficacy, safety of endoscopic therapy for duodenal duplication cysts (DDC) by comprehensively reviewing case reports.

Tandem, independent, systematic, computerized, literature searches were performed via PubMed using medical subject headings or Keywords ''cyst'' and ''duodenal'' and ''duplication''; or ''cyst'', and ''endoscopy'' or ''endoscopic'', and ''therapy'' or ''decompression''; with reconciliation of generated references by two experts. Case report followed CARE guidelines.

Literature review revealed 28 cases (mean = 1.3 ± 1.2 cases/report). Endoscopic therapy is increasingly reported recently (1984–1999: 3 cases, 2000–2015: 25 cases, $P = 0.003$, OR = 8.33, 95%-CI: 1.77–44.5). Fourteen (54%) of 26 patients were men (unknown-sex = 2). Mean age = 32.2 ± 18.3 years old. Procedure indications: acute pancreatitis-16, abdominal pain-8, jaundice-2, gastrointestinal (GI) obstruction-1, asymptomatic cyst-1. Mean maximal DDC dimension = 3.20 ± 1.53 cm (range, 1–6.5 cm). Endoscopic techniques included cyst puncture via needle knife papillotomy (NKP)/papillotome-18, snare resection of cyst-7, cystotome-2, and cyst needle aspiration/ligation-1. Endoscopic therapy was successful in all cases. Among 24 initially symptomatic patients, all remained asymptomatic post-therapy without relapses (mean follow-up = 36.5 ± 48.6 months, 3 others reported asymptomatic at follow-up of unknown duration; 1 initially asymptomatic patient remained asymptomatic 3 years post-therapy). Two complications occurred: mild intraprocedural duodenal bleeding related to NKP and treated locally endoscopically.

A patient is reported who presented with vomiting, 15-kg-weight-loss, and profound dehydration for 1 month from extrinsic compression of duodenum by 14 × 6 cm DDC, underwent successful endosonographic cyst decompression with large fenestration of cyst and endoscopic aspiration of 1 L of fluid from cyst with rapid relief of symptoms. At endoscopy the DDC was intubated and visualized and random endoscopic mucosal biopsies were obtained to help exclude malignant or dysplastic DDC.

Study limitations include retrospective literature review, potential reporting bias, limited patient number, variable follow-up.

In conclusion, endoscopic therapy for DDC was efficacious in all 29 reported patients including current case, including patients presenting acutely with acute pancreatitis, or GI obstruction. Complications were rare and minor, suggesting that endoscopic therapy may be a useful alternative to surgery for nonmalignant DDC when performed by expert endoscopists.

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Abbreviations: CI = confidence interval, CT = computerized tomography, DDC = duodenal duplication cyst, EGD = esophagogastroduodenoscopy, ERCP = endoscopic retrograde cholangiopancreatography, EUS = endoscopic ultrasonography, GI = gastrointestinal, IV = intravenous, MRCP = magnetic resonance cholangiopancreatography, MRI = magnetic resonance imaging, NKP = needle knife papillotomy, OR = odds ratio, RUQ = right upper quadrant, US = ultrasound.

INTRODUCTION

Symptomatic duodenal duplication cysts (DDC) were traditionally treated surgically, but have been increasingly treated endoscopically to avoid considerable morbidity from surgery.[1] This work comprehensively reviews this subject with review of 28 previous cases, and shows this technique is likely efficacious, relatively safe, and durable. A 29th patient is also reported who presented with severe recurrent vomiting for 1 month from duodenal compression by a huge DDC that was successfully treated by DDC decompression under endosonographic guidance with symptomatic resolution. During decompression at endosonography, the DDC was intubated and biopsied to exclude malignancy/dysplasia. This work suggests that endoscopic treatment is a viable and potentially preferable alternative to surgery for most DCC, provided endoscopic expertise is available and malignancy is excluded.

METHODS

Computerized literature searches independently performed in tandem via PubMed using the medical subject headings (MeSH) or key words of ''cyst'' and ''duodenal'' and

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''duplication''; or ''cyst'' and ''endoscopy'' or ''endoscopic'' or ''therapy'' and ''decompression''; with reconciliation of cited references. One article written in French,[2] and one article written in Spanish[3] were professionally translated. Case report followed CARE guidelines. This review received exemption/approval by William Beaumont Hospital IRB on August 14, 2015.

CASE REPORT

See Appendix 1.

RESULTS

Epidemiology and Anatomy

DDC are extremely rare congenital anomalies, with a prevalence of less than 1 per 100,000 live births.[4] DDC are the least common small bowel duplication cysts, preceded by ileal and jejunal duplication cysts.[5] In a comprehensive meta-analysis, 47 DDC were reported from 1999 to 2009.[4] About 60% of cases are diagnosed during infancy and childhood, and 40% are diagnosed in adulthood.[4–8]

By definition, a DDC must adhere to the duodenum, contain a smooth muscle layer in its walls, and be lined by duodenal epithelium.[5,6,9–11] DDC usually share the blood supply with the rest of the duodenum.[6] DDC are usually filled with clear fluid, but can contain bile, pancreatic fluid, or gallstones if they connect to the pancreaticobiliary ducts.[12–15] DDC can occur in any duodenal segment, but most commonly arise in the second or third parts of the duodenum. They usually arise on the mesenteric side.[5,13]

DDC can communicate directly with the true lumen via the duodenum, or via the pancreaticobiliary ducts, or have no communication.[4–6,9,16] About 30% of DDC communicate with the pancreaticobiliary ducts.[4] DDC are also classified according to shape as cystic or elongated/tubular.[13] DDC are most commonly cystic.[5,12]

Clinical Course

DDC are usually asymptomatic and incidentally discovered during radiologic imaging or esophagogastroduodenoscopy (EGD).[6,7] They can remain clinically silent for many years.[6] Symptoms, when present, most commonly include abdominal pain, followed by nausea in 80%, and vomiting in 40%.[4] The most common complication is acute pancreatitis.[4,12] Other complications include: jaundice, biliary obstruction, cyst infection, intussusception, cholestasis, and hepatitis.[4,14–20] DDC may contain ectopic pancreatic or gastric tissue that can cause duodenal ulcers, gastrointestinal (GI) bleeding, or rarely duodenal perforation.[6,11,21,22] DDC are usually benign, but are occasionally malignant.[7,23–25]

Diagnosis

DDC are strongly suggested by characteristic radiologic imaging findings, but imaging findings are occasionally misleading.[12,18] Twin major goals of radiologic imaging are to determine that the lesion is fluid-filled and to locate the cyst attachment site.[13] Contrast radiographs reveal a submucosal or extrinsic mass or filling defect.[4] Abdominal computerized tomography (CT), ultrasound (US), and magnetic resonance imaging (MRI) are commonly used to detect DDC. DDC appear as discrete, fluid-filled structures attached to the duodenal wall.[4,13] CT is useful to depict cyst location and dimensions, potential communications, and other anomalies.[26] On CT scan, the wall of duplication shows contrast enhancement, but the cyst content has a density of 0–20 Hounsfield units without contrast enhancement.[13] On US, DDC are highly suspected when an outer hypoechoic muscular layer and an inner echogenic mucosal layer is detected, a phenomenon called the ''double-wall'' or ''muscular rim'' sign.[5,13,26,27] Cysts may contain keratin and desquamated cells, which can appear solid on US.[17] MRI typically shows low attenuation signals in T1- and T2-weighted images.[13] A Meckel's technetium scan is used to exclude ectopic gastric tissue within DDC.[13] Magnetic resonance cholangiopancreatography (MRCP) or endoscopic retrograde cholangiopancreatography (ERCP) can also be used diagnostically, and to exclude cyst communication with the pancreaticobiliary ducts.[4,16,28,29] If the choledochus drains into the DDC, only the pancreatic duct fills with contrast during ERCP.[16] A meta-analysis revealed false negative rates for DDC of 23% with US and 9% with CT or MRI/MRCP.[4] Three-dimensional virtual cholangioscopy has been used to exclude communication between DDC and pancreaticobiliary ducts.[7,30]

Definitive diagnosis requires EGD with endosonographic confirmation, or surgery with pathologic analysis of resected tissue.[22,29,31–34] At EGD, DDC appear as a duodenal bulge with normal overlying mucosa, or as a variably long diverticulum. DDC usually have a smooth, regular, mucosal appearance. Other cystic masses in the differential diagnosis include: choledochal cyst, cystic dystrophy of duodenal wall, pancreatic pseudocyst, duodenal polyp, cystic pancreatic tumors, lymphatic malformation, mesenteric cyst, omental cyst, and gastrointestinal stromal tumor.[13,35–37] Endoscopic ultrasonography (EUS) can confirm the presence of muscular layer continuity between the duodenum and its duplication.[9,13,38] Fine needle aspiration can be performed during EUS if the diagnosis is uncertain, but aspiration occasionally results in cyst infection.[17,39–42]

Therapy

Asymptomatic cysts are usually managed expectantly, but some authorities recommend intervention based upon potential complications, including malignant transformation,[5] especially in patients who are unlikely to follow-up.[4,43] Prospective studies of the natural history of DDC are, however, unavailable. Symptomatic DDC generally mandate endoscopic or surgical therapy. Moreno et al,[44] however, reported one case of medical management of a symptomatic DDC with prokinetic therapy, and recommended annual follow-up with EGD or abdominal US, if a symptomatic cyst is managed medically. Some authorities recommend complete surgical resection of symptomatic DDC,[7,19,45,46] whereas others recommend therapeutic endoscopy to establish free drainage.[6,46–48] Surgical extirpation entails higher morbidity, and appreciable mortality as compared with endoscopic therapy, but eliminates the risk of malignant cyst degeneration.[6,7] Pancreaticoduodenectomy is sometimes necessary intraoperatively if the DDC is too close to the pancreaticobiliary ducts.[7] Less invasive, surgical techniques, including partial cyst resection, internal derivation,[19] and marsupialization,[4] have become obsolete.

Systematic Literature Review of Therapeutic EGD

Systematic literature review revealed 28 reported cases of endoscopic therapy (Table 1,[3,8,45–63] number of case reports per publication $= 1.3 \pm 1.2$). Endoscopic therapy is increasingly reported recently: 3 cases reported from 1984 to 1999, versus 25 cases reported from 2000 to 2015 ($P = 0.003$, OR = 8.33, 95%-CI: 1.77–44.5, Fisher exact test). Among 26 patients in whom the sex was reported, 14 (54%) were men. The mean

TABLE 1. Clinical Presentation and Endoscopic Therapy for Duodenal Duplication Cysts: Comprehensive Review of 28 Cases Reported in the Literature with Patient Outcomes

First Author, Year of Publication [reference #]	Age in years, Sex	Symptoms (Clinical Syndrome)	Physical Examination	Laboratory Findings	Abdominal Imaging Findings	Endoscopic Therapy	Follow-Up
Presenting as (de novo or recurrent) acute pancreatitis							
1. Traif and Khan, 1992,[47]	11, M	Recurrent, severe, sudden, epigastric pain (acute pancreatitis)	Epigastric tenderness, abdomen firm with voluntary guarding, T = 38.6°C.	WBC = 14,000/cumm, amylase = 766 IU (Nl: 23–85 IU), normal liver enzymes. Amylase declined to 160 IU after cyst drained	US: peristaltic wave passing through cystic structure. CT and barium swallow: structure in C loop of duodenum, compressing head of pancreas, and second and third portions of duodenum, cyst dimensions not reported	NKP used to make a 3-cm incision and opening on medial wall along longitudinal axis of descending duodenum	CT at 1 year and EGD at 2 years follow-up: collapsed cyst No further attacks of pancreatitis
2. Antaki et al, 2008,[48]	40, M	Acute pancreatitis	Not reported	Not reported	US, CT	Initial puncture with NKP; then inverted sphincterotome used; endoscopic biliary sphincterotomy (EBS) performed	MRCP and ERCP; asymptomatic at 120 months follow-up
3. Martínez-Alcalá García et al, 2014,[3]	24, F	Recurrent acute pancreatitis	Not reported	Not reported	US, CT, EUS, ERCP, MRCP, EUS: intramural anechoic lesion, separate from pancreatic and biliary ductal systems	Initial puncture with cystotome, then used sphincterotome	EGD, MRCP; asymptomatic at 26 months follow-up
4. ibid.[3]	47, M	Acute pancreatitis and chronic GI blood loss	Not reported	Not reported	EUS: intramural anechoic lesion, separate from the pancreatic and biliary ductal systems. Also underwent US, ERCP, MRCP	Initial puncture via NKP; endoscopic biliary and pancreatic sphincterotomies performed with placement of stents	MRCP; asymptomatic at 12 months follow-up
5. ibid.[3]	8, M	Recurrent acute pancreatitis	Not reported	Not reported	US, upper GI series	Initial puncture via NKP; then sphincterotome used, partial resection of cyst using snare	ERCP; asymptomatic at 152 months follow-up
6. ibid.[3]	43, M	Acute pancreatitis	Not reported	Not reported	CT, ERCP	Initial puncture using NKP; partial resection of cyst using snare	US, ERCP: asymptomatic at 160 months follow-up
7. ibid.[3]	18, F	Recurrent acute pancreatitis	Not reported	Not reported	MRCP	Partial resection of cyst using snare, without initial incision	MRCP: asymptomatic at 125 months follow-up
8. ibid.[3]	31, F	Recurrent acute pancreatitis	Not reported	Not reported	EUS: anechoic intraluminal lesion, separate from pancreatic and biliary ductal systems. MRCP	Partial resection of cyst using snare, without initial incision	EGD; asymptomatic at 5 months follow-up

First Author, Year of Publication [reference #]	Age in years, Sex	Symptoms (Clinical Syndrome)	Physical Examination	Laboratory Findings	Abdominal Imaging Findings	Endoscopic Therapy	Follow-Up
9. Chryssostalis et al, 2007,[49]	17, unknown sex	Recurrent acute pancreatitis and abdominal pain (acute pancreatitis)	Normal physical examination	Normal routine laboratory tests	EUS: 2-cm cystic lesion filled with hyperechoic stones surrounded by a thick wall near major papilla. ERCP: small opening on left edge of cyst, contrast reveals several stones, but no reflux of contrast into either CBD or MPD	Excision of cyst wall using papillotome and evacuation of stones	ERCP at 6 months follow-up: partially excised cyst with residual elevation of lateral walls; asymptomatic at 9 months follow-up
10. Rockx and McAlister, 2007,[50]	26, M	Persistent abdominal discomfort (from recurrent acute pancreatitis)	Normal physical examination	Reportedly normal	MRCP: cystic filling defect protruding into lumen of descending duodenum. ERCP: large protrusion into descending duodenum near papilla, contrast injected via a pinhole filled the cyst	Papillotome placed into cyst to widen opening into duodenum; fenestration done by cruciate extensions of initial incision	No further episodes of pancreatitis during 1 year follow-up; chronic abdominal pain resolved
11. Tekin et al, 2009,[51]	18, F	Recurrent abdominal pain and prior pancreatitis (recurrent abdominal pain)	Normal physical examination	Normal routine laboratory tests	CT: cystic duodenal polypoid lesion. ERCP: 3×2 cm cystic lesion; not communicating with CBD	NKP incision; guide wire placement and dilatation with 8-mm balloon; consecutive plastic stent implantation (10-Fr double-pigtail) with drainage; removal of pigtail stent after 1 month	Collapsed cyst cavity at 1 month follow-up at EGD; asymptomatic at 4 months follow-up
12. Redondo-Cerezo et al, 2010,[8]	37, M	(Recurrent acute pancreatitis)	Mild epigastric tenderness without peritoneal signs	Amylase = 861 U/L; no other abnormalities	US: normal. MRI: cystic dilatation of distal, periampullary CBD, protruding into duodenal lumen. ERCP: protruding duodenal mass from descending duodenum, proximal third of pancreatic duct blocked at pancreatography, suggestive of pancreas divisum. EUS: cystic cavity with debris and a wall with three identifiable layers	Drainage achieved by opening cyst using NKP	Repeat EUS at 1 month: cyst not detected; asymptomatic at 3 months follow-up
13. Criblez et al, 2011,[46]	17, M	Severe epigastric pain, nausea (acute epigastric pain)	Afebrile, no jaundice, upper abdominal tenderness, decreased bowel sounds	Lipase = 5400 U/L, liver function tests within normal limits, elevated CRP	US: unremarkable; CT: 2 cm wide cystic structure protruding into descending duodenum. ERCP: no communication with CBD or MPD	Decompression and marsupialization using needle knife and a standard biliary papillotomy with a semicircular incision; deroofing using a polypectomy snare	Asymptomatic during 4 years of follow-up

First Author, Year of Publication [reference #]	Age in years, Sex	Symptoms (Clinical Syndrome)	Physical Examination	Laboratory Findings	Abdominal Imaging Findings	Endoscopic Therapy	Follow-Up
14. Meier and Mellinger, 2012,[45]	9, M	Sharp epigastric pain and nausea for 1 day	Mild abdominal tenderness, no peritoneal signs	Amylase = 270 U/mL, lipase = 824 U/mL. Normal WBC, hemoglobin and electrolytes	CT: 2.9 × 2.6-cm cystic mass within descending duodenum near papilla. MRCP: DDC with intraluminal debris just caudad to major papilla and apparent separate duodenal opening for pancreatic duct	Cyst entered via NKP. Guidewire placed into cyst lumen and cyst opened with sphincterotome. Endoscopic snare used to resect portion of opened cyst wall to facilitate drainage	Asymptomatic at 6 months follow-up, repeat CT: small residual irregularity in area of prior cyst
15. Arantes et al, 2012,[52]	22, M	Recurrent acute pancreatitis	Normal physical examination	Not reported	MRCP: 2.3 × 1.7 cm cyst abutting duodenal lumen close to papilla; EUS: bulging anechoic duodenal cyst with distinct walls next to papilla, covered by normal mucosa ERCP: normal findings, no communication with CBD or pancreatic duct	Wide opening of cyst with NKP, with subsequent snare resection of margins using a rotatable hexagonal snare	Asymptomatic at 4 years follow-up, no recurrent acute pancreatitis; EGD at 2 years follow-up: wide communication between cyst and duodenal lumen
16. Antaki et al, 2012,[53]	21, M	Recurrent attacks of severe epigastric pain during 30 months	Normal physical examination	Elevated amylase 3–4 times during attack, otherwise normal blood tests including ALT, ALK P, CBC	CT, MRI, and MRCP: ovoid cystic mass in duodenum; ERCP: pancreas divisum with orifices of major and minor papilla surrounding cystic mass EUS: bulging duodenal 2.3 × 1.4 cm, anechoic cyst with distinct walls, situated adjacent to papilla margin	Partial resection of lower part of cyst using polypectomy snare and minor papilla sphincterotomy	Asymptomatic at 10 months follow-up
Presenting as epigastric or abdominal pain							
17. Johanson et al, 1992,[54]	62, M	Severe, sudden, epigastric pain	Normal physical examination	Serum amylase, lipase, and liver function tests all within normal limits	US: mildly dilated CBD (7.5 mm wide). Abdominal radiograph and CT scan: cystic mass at junction of second and third portions of duodenum. EGD: 4 × 5 cm peri-ampullary mass ERCP: cystic peri-ampullary mass, communicating with choledochus via small aperture adjacent to papilla	2-cm incision via NKP at dependent portion of cyst; epinephrine injected submucosally to control bleeding; nasobiliary catheter placed into cyst and removed after 48 hours	ERCP 2 months later: markedly diminished cyst size Asymptomatic at 1 year of follow-up

First Author, Year of Publication [reference #]	Age in years, Sex	Symptoms (Clinical Syndrome)	Physical Examination	Laboratory Findings	Abdominal Imaging Findings	Endoscopic Therapy	Follow-Up
18. Wada et al, 2001,[55]	30, F	Intermittent epigastric and back pain	Normal physical examination	Normal routine laboratory tests	US: 3.6 × 3.2 × 1.7 cm cystic mass, adjacent to pancreatic head. EUS: cyst connected to CBD and main pancreatic duct. ERCP: no reflux of injected contrast into CBD or MPD from cyst. 3D-CT with drip infusion cholangiography: cyst connected to CBD with retention of contrast medium	Cyst aspiration, then ligation and snare resection of most of cyst	Asymptomatic at 1 year, endoscopy at 2 months showing well-opened cystoduodenal fistula with normal drainage into duodenum by 3D-CT with drip infusion cholangiography
19. Gyökeres et al, 2001,[56]	19, F	Intermittent upper abdominal pain	Normal physical examination	Normal routine laboratory tests	US: 6.5 × 3.0 × 3.0 cm cystic mass in right hypochondrium. CT, EUS and hypotonic duodenography: support diagnosis of DDC	Cystoduodenostomy, injection of contrast medium into cyst, NKP used to enlarge incision on bulging wall; nasocystic catheter inserted (forming multiple loops) into cyst cavity for 3 days to stop cyst bleeding and prevent early cyst collapse; standard papillotome and NKP used to extend opening to create 2 cm wide aperture	Asymptomatic at 1-year follow-up, with open duodenocystic communication
20. Vandenbroucke et al, 2005,[57]	23, F	Epigastric pain and gastroesophageal reflux	Normal physical examination	Normal routine laboratory tests	CT and EUS: paraduodenal, hypoechoic, cystic lesion on antimesenteric side of descending duodenum, >6 cm in length, composed of multiple layers that are similar to layers of normal duodenal wall	Incision with NKP of proximal, dependent portion of cyst. Cannulation with sphincterotome to extend opening to 1.5 cm in length	Asymptomatic at 1-month of follow-up, with collapsed cyst on repeat EGD, and extension of prior incision by 1 cm
21. Jung et al, 2009,[58]	52, M	Sudden onset of RUQ pain	Mild abdominal tenderness without rebound tenderness in right upper quadrant	ALT = 51 IU/L (normal: 0–41); γ-GT = 85 U/L, normal amylase, lipase, CA 19–9, and CEA	US: CBD stone and gallbladder polyp. EUS:bulging cystic lesion containing echogenic material with posterior acoustic shadowing near papilla. 3D CT: presence of cyst with a stone at duodenal ampulla. ERCP: round filling defect without visualization of pancreaticobiliary ducts	Endoscopic resection using standard polypectomy snare. Gross pathology: cyst measured 2.5 × 2.5 × 1.2 cm	Asymptomatic with normal routine lab tests at 6 months follow-up
22. Martínez-Alcalá García et al, 2014,[3]	37, F	Acute and recurrent postprandial pain accompanied by vomiting	Not reported	Not reported	CT and MRCP: 5 cm DDC, not communicating with pancreaticobiliary ducts	Cyst punctured using NKP, guide-wire introduced and opening expanded using sphincterotome	Asymptomatic at 1year of follow-up

First Author, Year of Publication [reference #]	Age in years, Sex	Symptoms (Clinical Syndrome)	Physical Examination	Laboratory Findings	Abdominal Imaging Findings	Endoscopic Therapy	Follow-Up
23. Kurien et al, 2014,[59]	Young, F	Intermittent abdominal pain.	Normal physical exam except for mild abdominal tenderness	Not reported	MRCP: cystic lesion next to CBD and MPD. EUS: cystic lesion with layered appearance in descending duodenum	Partial deroofing using oval snare (without drainage), subsequent puncture of cyst wall with a cystotome. Guidewire placed into cyst and deroofing of cyst using sphincterotome. Further dilations with a 15-mm wide expansion balloon	Doing well at follow-up
24. Johnson and Gopal, 2015,[60]	21, F	Episodic epigastric abdominal pain	Normal physical examination, except for mild epigastric abdominal tenderness	Not reported	CT: cystic mass in medial wall of duodenum. EUS: 2.6 × 1.0 cm anechoic cyst arising from submucosa, with a well-defined double-layer wall. EUS and MRCP: normal CBD and pancreatic duct not communicating with cyst	Fenestration of cyst with NKP and extension of fenestration using a sphincterotome. Opening widened using 11.5 mm wide balloon	Improvement of abdominal symptoms Follow-up MRCP: cyst resolution
Jaundice (including obstructive jaundice)							
25. Sezgin et al, 2001,[61]	30, M	Intermittent RUQ pain remitting spontaneously for 3 years	Epigastric tenderness, jaundice (Obstructive jaundice)	AST = 57 IU/L, ALT = 147 IU/L, Direct bilirubin = 13.3 mg/dL, Indirect bilirubin = 4.1 mg/dL, normal WBC count	US: dilated CBD (14 mm wide) and intrahepatic bile ducts. CT: 25 mm wide cyst in duodenal wall. ERCP: 3 cm wide cyst that communicates with biliary tree	NKP: long incision with formation of artificial biliary orifice to cyst; additional 1 cm incision performed using standard sphincterotome. Removal of stones from cyst with basket and balloon	ERCP 3 months later: patent orifice of cystoduodenostomy, with diminished cyst size and no stones. Patient asymptomatic at follow-up
26. Antaki et al, 2008,[48]	72, M	Jaundice	Not reported	Not reported	MRCP	Initial puncture via NKP; then endoscopic biliary sphincterotomy performed using insulated-tip knife	ERCP; asymptomatic at 55 months follow-up
UGI obstruction							
27. Dave et al, 1984,[62]	73, F	Postprandial vomiting, epigastric fullness, and pain, 5 kg weight loss during 3 months	Mild epigastric tenderness (Gastric outlet obstruction)	Serum amylase, lipase, and liver functions tests all within normal limits	Normal chest and plain abdominal radiographs. EGD: apparent 2 × 2 × 1 cm sessile polyp	Snare removal (like polypectomy) of sessile duplication cyst at junction between apex of bulb and descending duodenum Pathology: cyst center of a DDC and not an ordinary polyp	Symptoms resolved after polypectomy

First Author, Year of Publication [reference #]	Age in years, Sex	Symptoms (Clinical Syndrome)	Physical Examination	Laboratory Findings	Abdominal Imaging Findings	Endoscopic Therapy	Follow-Up
28. CURRENT REPORT	40, M	Recurrent vomiting, 15 kg weight loss, and signs of dehydration for 1 month	Orthostatic changes in pulse, dry mucous membranes, absent axillary sweat, midepigastric tenderness without rebound tenderness	AST = 87 u/L, ALT = 46 u/L, total bilirubin = 1.9 mg/dL, lipase = 209 u/L, WBC = 10,000/mm^3, Hgb = 15.0 g/dL	CT: 6 × 14 cm, smooth-walled, intraluminal filling defect extending from proximal descending duodenum to ligament of Treitz compressing duodenum lumen MRI: 8 × 13 cm intramural cystic mass. EGD: slit-like, opening in descending duodenum, from extrinsic compression, with no intrinsic mucosal lesions Also underwent abdominal US, SBFT, and EUS	Cyst punctured using a 19 gauge needle; thick, dark fluid aspirated; a 0.035 in. Jagwire was advanced via the needle into cyst; NKP fed over the Jagwire; cyst wall incised using papillotome, then aperture serially dilated up to 18 mm. Copious fluid seeped out of cyst which was suctioned	Soon after procedure patient felt well and tolerated oral feedings without nausea and vomiting. Feeling well and asymptomatic at 1 month of follow-up with 7-kg-weight-gain. EUS at 1 month: 1.5 × 2.0 residual cyst
Asymptomatic							
29. You et al, 2012,[63]	62, F	Asymptomatic; polypoid duodenal lesion found during EGD	Normal physical examination	Normal routine laboratory values except Hg = 9.9 g/dL	EUS: anechoic cystic lesion located in third layer of duodenal wall, cyst wall contains three layers EGD: soft, 1-cm wide polypoid lesion at posterior wall of superior duodenal angle	Endoscopic mucosal resection using snare	No cyst recurrence during 3 years of follow-up

3D-CT = three dimensional computerized tomography, CBD = common bile duct, CT = computerized tomography, EGD = esophagogastroduodenoscopy, ERCP = endoscopic retrograde cholangiopancreatography, ESD = endoscopic submucosal dissection, EUS = endoscopic ultrasound; F = female, GI = gastrointestinal, M = male, MRCP = magnetic resonance cholangiopancreatography, MRI = magnetic resonance imaging, NKP = needle knife papillotomy, RUQ = right upper quadrant, US = ultrasound.

TABLE 2. Clinical Presentation, Endoscopic Therapy, and Patient Outcomes for Gastric or Esophageal Duplication Cysts: Comprehensive Review of the Literature

First Author, Year of Publication [Reference #]	Age in Years, Sex	Symptoms (Clinical Syndrome)	Physical Examination	Laboratory Findings	Abdominal Imaging Findings	Endoscopic Therapy	Follow-Up
A. For seven reported cases of gastric duplication cysts							
1. Kobara et al, 2016[64]	41, M	NR	NR	NR	EUS: anechoic mass exhibiting double-wall thickness and originating in submucosal layer, findings characteristic of a duplication cyst. CT: intraluminal, enhancing mass	Submucosal endoscopy with a mucosal flap method (SEMF): creating a submucosal tunnel towards cyst using a 10-mm opening flap, cutting cyst, and inserting endoscope	NR
2. Klair et al, 2015,[65]	67, M	Epigastric pain; intermittent nausea and vomiting, which continued despite taking ondansetron, promethazine, and omeprazole; anorexia; and 7 kg weight loss during prior 2 months	Mild epigastric tenderness, no peritoneal signs	Unremark-able	Contrast-enhanced CT: 2.9 × 2.3-cm round mass protruding into gastric lumen at gastroesophageal junction. Mass suspicious for malignancy. EUS: smooth 3 × 1-cm cystic anechoic lesion in third gastric layer (submucosa) with thin septations, suggestive of lymphangioma. EGD: polypoid subepithelial lesion distal to gastroesophageal junction, best visualized on retroflexion	Standard lift-and-cut EMR performed with complete cyst excision	No follow-up evaluation planned, unless symptoms recur
3. Deesomsak et al, 2013[66]	52, M	Progressive iron deficiency anemia without overt gastrointestinal bleeding for 1 month	NR	Hemoglobin = 9.0 mg/dL; borderline low serum ferritin = 18.0 ng/mL (normal:12–200 ng/mL)	EGD: 2.0 cm pinkish umbilicated submucosal lesion in antrum with overlying ulceration and blood clot	EUS: 1.8 cm hypoechoic mass arising from second and third layers of gastric wall, with a centralized anechoic area lacking vascular flow, consistent with small GIST with cystic changes. EMR performed to create submucosal cushion by injecting 4 mL mixture of dilute epinephrine. Cyst removed en-bloc by snare with cautery	NR
4. Lee et al, 2011,[67]	23, F	Intermittent epigastric pain for several years	NR	NR	EUS: 5.1 × 4.2-cm cyst with four distinct walls and anechoic content, but containing echogenic debris, suggestive of gastric duplication cyst	Unable to grasp lesion with snare because lesion too large. Small area of covering mucosa resected. Large crosswise incision in cyst made by hook-knife, leading to outpouring of yellow viscous fluid from cyst and cyst collapse. Cyst wall was then resected in piecemeal fashion using a snare	NR
5. Eom et al, 2011,[68]	28, M	Dyspepsia for 1 year	NR	NR	EUS: anechoic homogenous, oval cyst originating from submucosal layer. Cyst wall was five-layered	Saline solution with epinephrine first injected beneath cyst to decrease risk of bleeding, but the saline spread into surrounding tissue and flattened cyst making it impossible to remove cyst via injection-and-cut technique; cyst was impossible to grasp by snare because contour was dented. Cyst removed by ESD without complications	NR
6. Stecevic et al, 2003,[69]	51, M	Melena for 2 days	NR	Decrease in hemoglobin from 14.0 g/dL to 10.5 g/dL	EGD: 3.0 × 2.6-cm polyp in proximal stomach	Removed by snare with cautery, like for standard polypectomy	No further melena. Hemoglobin level gradually normalized. EGD 3 months later: small scar at prior site of cyst, and hyperplastic gastric polyps

First Author, Year of Publication [Reference #]	Age in Years, Sex	Symptoms (Clinical Syndrome)	Physical Examination	Laboratory Findings	Abdominal Imaging Findings	Endoscopic Therapy	Follow-Up
7. Woolfolk et al, 1998,[9]	33, M	Epigastric discomfort and pyrosis for 3 years	Unremarkable physical examination	NR	EUS: 6.5 × 4.5 cm mass comprised almost entirely by a large 3.8 × 3.7 cm round, homogeneous anechoic lesion consistent with gastric duplication cyst. Lesion entirely encapsulated within third hyperechoic layer of gastric wall (submucosa)	Under fluoroscopic guidance, cyst punctured using sclerotherapy needle. Contrast injection showed filling of cystic cavity. NKP used to perform a 4.0 cm endoscopic cystotomy incision. Serous and sebaceous material then poured out from cyst incision with collapse of cyst. EGD 2 weeks later: persistent, smaller cyst in gastric fundus, with a confluence of folds and central scar from prior cystotomy. EUS: lesion reduced to 3.0 × 3.0 × 5.0 cm mass with only 1.3 × 3.0 × 1.1 cm remnant cyst. After injecting base with dilute epinephrine, standard electrocautery snare used to resect piecemeal remaining intragastric tissue, leaving a large clean, ulcerated, base with no significant bleeding	EGD performed 4 months later: small 1.5 × 2.0 cm confluence of folds in gastric fundus at site of previous cyst. EUS: apparent confluence of folds involving gastric mucosa and submucosa with a cleft from previous incision leading down to a small central lumen. No residual remnant of cyst seen. Patient became asymptomatic
B. For seven reported cases of esophageal duplication cysts							
1. Mou et al, 2015,[70]	NR	Progressive dysphagia during 12 months, without weight loss	NR	NR	EUS: 5-cm hypoechoic, intraluminal, cystic mass with clear margins originating from muscularis propria. CT: well-defined, cystic mass without internal enhancement	Endoscopic submucosal dissection of esophageal cyst: Cyst raised by submucosal injection of 100 mL of 10% glycerol fructose. After incising cyst wall with a dual-knife (KD-650L; Olympus, Tokyo, Japan), yellowish fluid drained from cyst. Cyst resected using dual-knife and hook-knife. Anhydrous alcohol sprayed into cyst cavity	Repeat EGD 7 days later: anhydrous alcohol again sprayed into cyst cavity
2. Ivekovic et al, 2012,[71]	51, M	Recent dysphagia for solid foods	NR	NR	EGD and esophagography: submucosal lesion in lower third of esophagus protruding into lumen EUS: hypoechogenic 3.5 cm cystic lesion, surrounded by a multilayered wall, consistent with esophageal duplication cyst	After initial incision with NKP, anterior cyst wall fenestrated with IT-knife (Olympus, Tokyo, Japan), resulting in complete opening of cystic cavity into esophageal lumen. Lateral margins clipped to close gaps and prevent delayed bleeding	Follow-up EGD: epithelialization of posterior wall of esophageal duplication cyst
3. Ivekovic et al, 2012,[71]	32, F	Recent dysphagia for solid foods	NR	NR	EUS: hypoechogenic, 4.5 cm cystic lesion, surrounded by a multilayered wall, consistent with esophageal duplication cyst	After initial incision with NKP, anterior cyst wall fenestrated with IT-knife (Olympus, Tokyo, Japan), resulting in complete opening of cystic cavity into esophageal lumen. Lateral margins clipped to close gaps and prevent delayed bleeding	Follow-up EGD: epithelialization of posterior wall of esophageal duplication cyst

First Author, Year of Publication [Reference #]	Age in Years, Sex	Symptoms (Clinical Syndrome)	Physical Examination	Laboratory Findings	Abdominal Imaging Findings	Endoscopic Therapy	Follow-Up
4. Joyce et al, 2006,[72]	72, F	Progressive dysphagia for 3 months, 5.5 kg weight loss. No symptoms of reflux or anorexia	NR	NR	Chest CT and MRI: diffuse wall thickening of mid-to-distal esophagus, with proximal dilation of esophagus with an air-fluid level. EGD: esophageal mass, thought to represent mediastinal metastasis from known breast cancer. EUS: thickened esophageal wall with a 10.2 mm cystic lesion within the wall. Cyst had an incomplete wall-layer pattern	Three milliliter of saline solution injected into base of lesion stalk. Lesion then resected by snare with electrocautery, and retrieved with a Roth net	On follow-up, denied dysphagia and regained premorbid weight
5. Coumaros et al, 2010,[73]	14, M	Acute dysphagia for solids and liquids, acute retrosternal pain	NR	NR	Barium esophagram: 7-cm-long esophageal diverticulum with narrowing of upper third of esophageal lumen, compatible with cystic esophageal duplication CT and MRI: suggested esophageal duplication cyst, but communication with native esophagus was unclear	EGD under general anesthesia: Twin esophageal lumens encountered at 25 cm from incisors with thick bridge extending from 26 to 31 cm separating the lumens. Pediatric endoscope then advanced through main esophageal lumen into stomach. Endoscope passed through narrowed duplicated lumen with aid of a 0.035-inch guidewire, revealing that distal end of narrowed lumen was open to esophagus, compatible with tubular esophageal duplication. Endoscope was reintroduced into main esophagus and intraluminal bridge was incised longitudinally using a needle-knife starting from upper end. Upper esophageal stricture was dilated using a wire-guided balloon with expansion up to 15 mm. Complication: mucosal laceration by standard endoscope just above entrance of duplication cyst without significant bleeding	Eleven months later patient tolerated normal diet without symptoms, and was gaining weight
6. Kochhar et al, 2006,[74]	35, F	Chest pain, with spiking fever 10 days after surgery for esophageal duplication cyst with partial excision and partial suture closure due to extensive adhesions	Unremarkable except for toxemia	NR	Gastrografin esophagogram: esophageal leak into posterior mediastinum EGD: 8 mm long mid-esophageal tear	Three metallic clips successfully applied to close esophageal leak	Repeat contrast study on day 15 and endoscopic examinations on days 7 and 21: complete closure of leak

First Author, Year of Publication [Reference #]	Age in Years, Sex	Symptoms (Clinical Syndrome)	Physical Examination	Laboratory Findings	Abdominal Imaging Findings	Endoscopic Therapy	Follow-Up
7. Will et al, 2005,[75]	25, M	Dysphagia for several weeks	NR	NR	EGD: extrinsic impression on the lower esophagus EUS: 4 × 5 cm cystic lesion within esophageal wall, with an echo-dense wall surrounding an echo-poor cystic lumen. Cystic wall was 3 mm thick. The interlaying wall between cyst and esophagus consisted of four single layers. The esophageal muscularis propria was the bordering layer to the cystic lesion	Under EUS guidance needle punctured cyst wall. Then fenestration extended to 1 cm using a needle-knife and cyst completely drained. Incision then extended to 2 cm using a papillotomy knife. Endoscopic inspection of cyst cavity revealed a prominent cyst wall with a smooth mucosa. Mucosal biopsies revealed esophageal squamous epithelium. Patient was asymptomatic during the next 6 weeks, but then developed recurrent dysphagia. Repeat EGD revealed scarring of cyst stoma with adjacent pus. Mucosectomy of cyst wall using a diathermy snare was followed by removal of lamina propria by papillotomy knife. Opening was extending to 4 cm allowing pus to drain out of cyst. Severe inflammation and aphthous, ulcers were present on cystic mucosa. Cyst cavity then irrigated	Patient's condition subsequently improved, with no further complaints. Follow-up EGD 1 year later: normal esophagus with a diverticulum-like extension at the former cavity site. Pathologic examination of mucosal biopsies showed no malignant transformation

CBD = common bile duct, CT = computerized tomography, EGD = esophagogastroduodenoscopy, ERCP = endoscopic retrograde cholangiopancreatography, ESD = endoscopic submucosal dissection, EUS = endoscopic ultrasound, F = female, M = male, MRCP = magnetic resonance cholangiopancreatography, MRI = magnetic resonance imaging, NKP = needle knife papillotomy, US = ultrasound.

patient age = 32.2 ± 18.3 years old. Procedure indications included: acute pancreatitis-16 (first episode-10, recurrent-6), abdominal pain-8, jaundice-2, upper GI obstruction-1, and asymptomatic cyst-1 (Table 1). Two patients had two simultaneous clinical syndromes: including one patient presenting with acute pancreatitis and chronic GI blood loss (patient-4[48]), and one patient presenting with obstructive jaundice and RUQ abdominal pain (patient-25[61]). Only 1 patient had chronic GI blood loss attributed to DDC (patient-4[48]). The rarity of this presentation is likely because DDC are uncommonly inflammatory, invasive, or malignant. Patients typically presented with symptoms, signs, and laboratory abnormalities suggestive of the presenting syndrome (e.g., acute pancreatitis), but without clinical features suggesting that the underlying cause was DDC. DDC was, however, generally diagnosed by radiologic imaging, including EUS, CT, ERCP, or MRCP. The mean maximal diameter of the reported DDC = 3.20 ± 1.53 cm.

Systematic literature review demonstrates that endoscopic treatment is safe and effective, avoiding the morbidity associated with laparotomy[57]. Endosonographic therapeutic techniques included cyst puncture via needle knife papillotomy (NKP) or papilotome-18, snare resection of cyst-7, cystotome-2, and cyst needle aspiration and ligation-1. Endosonographic therapy was successful in all 28 prior cases (Table 1). Only 2 complications (7.1%) were reported. Both complications were relatively mild: GI bleeding, related to NKP, that were easily managed without any permanent clinical sequelae by dilute subcutaneous epinephrine injection and placement of a cyst catheter for drainage that was removed after 48 hour (patient-17[54]), or by leaving a cyst drain for 3 days for tamponade and monitoring the bleeding (patient-19[56]). All patients remained asymptomatic during variable follow-up. Among 25 patients in whom the length of follow-up was reported, all were asymptomatic during mean follow-up of 35.1 ± 48.1 months. Three other patients were reported as asymptomatic at follow-up, but follow-up duration was not reported (patients 23,24,27[59–61]). One patient who had presented with an asymptomatic DDC remained asymptomatic for 3 years after endoscopic therapy (patient 29[63]). These data suggest that endoscopic therapy may be durable without symptomatic recurrences. Table 3 summarizes the key findings of this comprehensive review of endosonographic therapy for DDC.

Advantages of endoscopic therapy include no visible abdominal scars, decreased post-operative pain, and shorter hospitalization.[45] Surgery is necessary if a symptomatic cyst cannot be approached endoscopically.[4] Before therapeutic endosonography, the patient should be advised of a potential need for traditional surgery if therapeutic endoscopy fails.[45]

Comprehensive review of 93 gastric duplication cysts reported in the literature revealed 7 cases treated endoscopically. In all these cases the endsocopic therapy was successful without complications (Table 2A).[9,64–69] Comprehensive review of 80 esophageal duplication cysts reported in the literature revealed 7 cases treated endoscopically. In all these 7 cases the endoscopic therapy was successful (Table 2B).[70–75] Two minor endoscopic complications occurred: minor

TABLE 3. Key Findings in Endoscopic Therapy of Duodenal Duplication Cysts: 28 Previously Reported Cases vs. Currently Reported Case

Parameter	Previously Reported Cases (N = 28)	Currently Reported Case (N = 1)
Mean ± S.D. number of reported cases per publication	1.3 ± 1.2	1
Number of cases reported 2000–2015 vs. number of cases reported 1984–1999	25 vs. 3	1 vs. 0
Number (%) male	14 (54%)*	1
Mean ± S.D. patient age	32.3 ± 18.3 years	40 years
Clinical syndrome: acute pancreatitis vs. other	16 vs. 12†	0 vs. 1 (gastrointestinal obstruction)
Mean ± S.D. maximal cyst diameter	3.2 ± 1.5 cm	14.5 cm (N = 1)
Primary technique of endoscopic fenestration: NKP or papillotomy vs. other	18 vs. 10‡	1 vs. 0
Number (%) successful endoscopic therapy§	28 (100%)	1
Number (%) of patients with complications from endoscopic therapy	2 (7.1%) both complications were minor	0
Follow-up after endoscopic therapy	35.1 ± 48.1 months (N = 25 patients)	1 month
Number remaining asymptomatic during follow-up	28 (100%) (3 with duration of follow-up unspecified)	1

CT = computerized tomography, N = number, NKP = needle knife papillotomy, S.D. = standard deviation, vs. = versus.
*Sex reported in only 26 patients.
†The other syndromes in 12 patients included: abdominal pain-8, jaundice-2, upper gastrointestinal obstruction-1, and asymptomatic cyst-1.
‡Other endoscopic techniques included: snare resection of cyst-7, cystotome-2, and cyst needle aspiration and ligation-1.
§Successful endoscopic therapy defined as successful endoscopic drainage of cyst, relief of symptoms, signs, and laboratory abnormalities, and no or only minor complications of endoscopic therapy.

esophageal mucosal laceration without GI bleeding in 1, and minor infection at the cyst incision site successfully treated by irrigation and cyst opening extension at repeat EGD in 1. The number of gastric or esophageal duplication cysts treated endoscopically is relatively small. However, these literature reviews on gastric or esophageal duplication cysts further support the efficacy and safety of therapeutic endoscopy for upper gastrointestinal duplication cysts.

DISCUSSION

The currently reported patient was unusual in that the DDC was 14 cm long, more than twice as long as the previously reported longest DDC of 6.5 cm, and that the DDC caused GI obstruction by compressing the duodenal lumen, as demonstrated by abdominal CT and EGD. Only 1 previously reported patient treated endoscopically had presented with GI obstruction (patient-27: gastric outlet obstruction[62]). However, two children treated surgically had presented with gastric outlet obstruction from compression by pyloroduodenal duplication cysts.[76,77]

This current case illustrates that even an extremely long DDC can be adequately drained by therapeutic endosonography, without recurrence at least during 1 month of follow-up, and that adequate DDC decompression can reverse nausea, vomiting, and weight loss from the DDC. The key requirement is adequate DDC drainage and decompression to relieve extrinsic compression of the duodenal lumen causing the severe vomiting. The currently reported patient also had acute pancreatitis, likely from pancreatic ductular hypertension from the DDC. The key findings in the currently reported DDC are compared with those in the previously reported DDCs in Table 3.

Keratinized cysts are lined by keratinized squamous mucosa and tend to have thicker and more viscous fluid within individual cysts than non-keratinized cysts. DDC are generally non-keratinized because they are lined by columnar epithelium and tend to have thinner and less viscous fluid within the cyst. The currently reported cyst was non-keratinized histologically and the cyst fluid was thin and, therefore, relatively easy to aspirate.

The current case report adds to the literature by reporting endoscopic intubation and visualization of the DDC with random mucosal endoscopic biopsies. We speculate that this endoscopic maneuver might prove helpful to exclude potential malignancy or dysplasia in a DDC, which would preclude endoscopic therapy.

Limitations of this review include incorporation of retrospectively reported case reports, potential reporting bias in that successful therapeutic endoscopy might have been preferentially reported, variable follow-up, and review of only 29 reported cases of DDC. Review strengths include performance of a systematic literature review; use of tandem, independent, investigators for the literature search with subsequent reconciliation of references to minimize omissions or biases; relatively long mean follow-up of 35.1 months in 25 cases; and illustration of endosonographic therapy to successfully reverse severely symptomatic duodenal compression from one extremely long DDC.

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APPENDIX 1: CASE REPORT

A 40-year-old man with prior mild gastroesophageal reflux and no prior gastrointestinal (GI) surgery presented with moderate epigastric pain, and recurrent nausea and vomiting of partly digested food 1 hour post-cibum, associated with 15-kg-weight-loss during the prior month. Physical examination revealed a blood pressure of 106/71 mmHg, pulse of 70 beats/min with orthostasis, dry mucous membranes, poor skin turgor, and absent axillary sweat. Abdominal examination revealed mild epigastric tenderness, without rebound or guarding, and normoactive bowel sounds. Rectal examination revealed no fecal occult blood. Laboratory analysis revealed 10,000 leukocytes/mm^3, and hematocrit of 44.9. The serum level of sodium = 131 mmol/L, potassium = 3.6 mmol/L, chloride = 73 mmol/L, bicarbonate = 39 mmol/L, blood urea nitrogen = 80 mg/dL, and creatinine = 2.4 mg/dL. The alkaline phosphatase = 140 U/L, AST = 87 U/L, ALT = 46 U/L, total bilirubin = 1.9 mg/dL, and lipase = 209 U/L. International normalized ratio and platelet count were within normal limits.

The patient was aggressively administered IV fluids with electrolytes for volume resuscitation and electrolyte repletion, with prompt improvement in electrolyte abnormalities. Abdominal CT administered with oral, but not IV, contrast, due to prerenal azotemia, revealed a 6 × 14 cm, smooth-walled, intraluminal filling defect extending from proximal descending duodenum to nearly the ligament of Treitz which compressed the duodenum lumen, but some contrast traversed through the compressed segment to distal small bowel, findings consistent with DDC. CT revealed no pancreatic or peripancreatic inflammation. Abdominal US confirmed a large, simple, retroperitoneal cyst was present. The patient was administered total parenteral nutrition because of intolerance of oral feedings and was administered chlorpromazine for intractable hiccups.

EGD revealed a slit-like, opening in descending duodenum, from extrinsic compression, with no intrinsic mucosal lesions. Histopathologic analysis of biopsies from the slit-like opening revealed mild chronic inflammation without dysplasia.

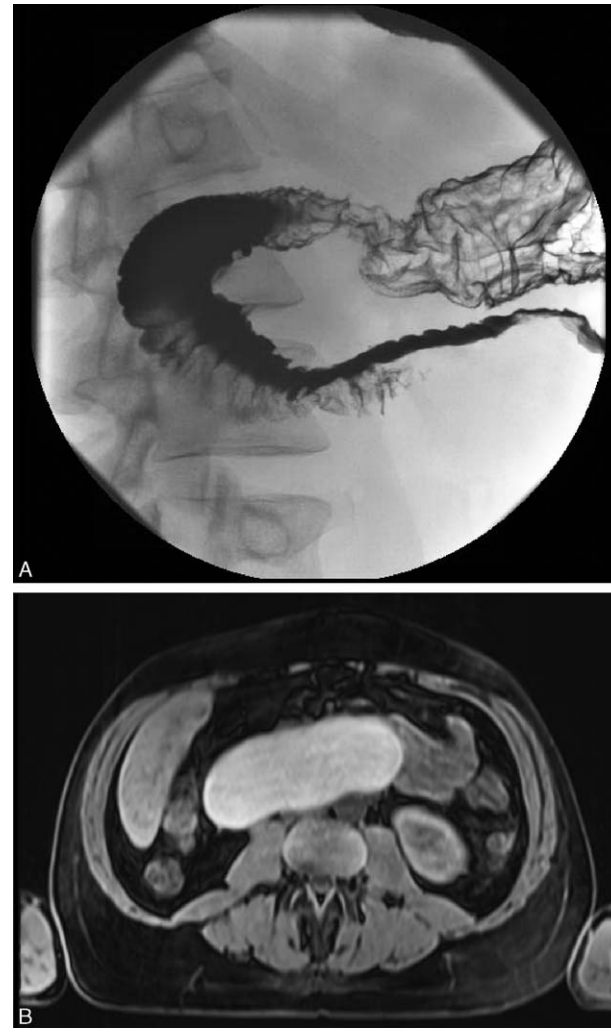

FIGURE 1. A, Upper gastrointestinal series with small bowel follow-through in a 40-year-old man presenting with severe nausea and vomiting post cibum associated with 15 kg weight loss during the prior month reveals a smoothly contoured deformity along the third portion of the duodenum caused by a central mass displacing and compressing this part of the duodenum. The duodenum is mostly but not completely obstructed by the central mass. B, Abdominal magnetic resonance imaging (MRI) without IV gadolinium contrast reveals an 8 × 13 cm intramural cystic mass arising along wall of the descending and transverse duodenum that demonstrated T1 and T2 hyperintense signals, and an irregularly thickened posterior cyst wall, findings consistent with a duodenal duplication cyst containing complex fluid, such as hemorrhagic or proteinaceous material.

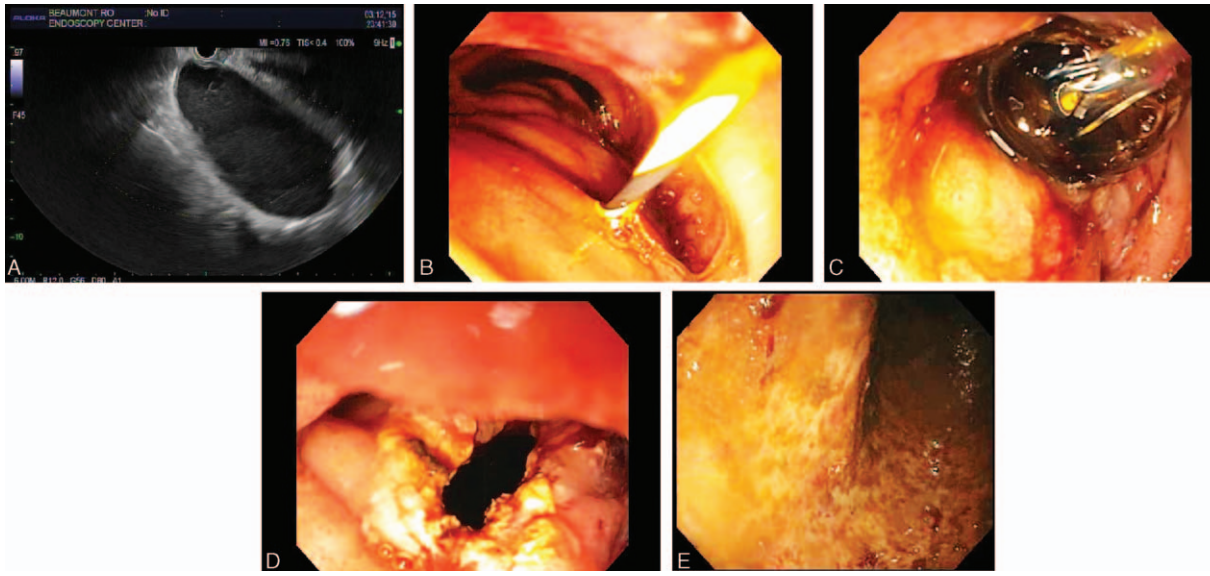

FIGURE 2. Progressive stages of endosonographic therapy to decompress a highly symptomatic and very long duodenal duplication cyst. A, Endoscopic ultrasound (EUS) revealed an oval, intraluminal (subepithelial), anechoic lesion, that endosonographically originated from within the submucosa (layer 3), consistent with duodenal duplication cyst. The cyst arose from muscularis propria of descending duodenum just distal to the papilla, extended deeply into the third portion of duodenum, and contained significant debris within it. B, The duodenal cyst was punctured using a 19 gauge needle; thick, dark fluid was aspirated; a 0.035 in. Jagwire was advanced via the needle into the cyst; the needle was withdrawn; a needle-knife papillotome was fed over the Jagwire; and the cyst wall was incised using the papillotome. C, The papillotome was exchanged with a pyloric balloon dilator, and the aperture was serially dilated up to 18 mm using increasingly larger balloons. D, EGD reveals a wide aperture after progressive balloon dilatation. E, The echoendoscope was withdrawn and a pediatric colonoscope was intubated and advanced into the cyst, and 800 cc of dark, brown fluid was aspirated via the colonoscope This endoscopic photograph shows the mucosa within the cyst has no evident lesions after cyst aspiration.

Upper gastrointestinal series with small bowel follow-through revealed a smoothly contoured deformity along the third portion of the duodenum caused by a central mass displacing adjacent bowel loops (Figure 1A). Most of the contrast remained in the stomach, but some contrast traversed through the small bowel with a normal transit time. Abdominal magnetic resonance imaging (MRI), without IV gadolinium contrast, revealed an 8×13 cm intramural cystic mass arising along wall of the descending and transverse duodenum that demonstrated T1 and T2 hyperintense signals, and an irregularly thickened posterior cyst wall, findings consistent with a DDC containing complex fluid, such as hemorrhagic or proteinaceous material (Figure 1B). The pancreaticobiliary ducts were not dilated. A Meckel's technetium scan revealed no scintigraphic evidence of ectopic gastric mucosa within the cyst.

For endoscopic therapy, the patient underwent endotracheal intubation and a linear echoendoscope was advanced into descending duodenum with some difficulty due to the mostly obstructed duodenal lumen from extrinsic compression. Limited EUS revealed an oval, intraluminal (subepithelial), anechoic cystic lesion, that endosonographically had a muscular wall that arose from the muscular wall of the native duodenum, sonographic findings consistent with DDC (Figure 2A). The cyst arose from muscularis propria of descending duodenum just distal to the papilla, extended at least 6 cm into the third portion of duodenum, and contained significant debris within it. Doppler ultrasound excluded pericystic vessels. The cyst was punctured using a 19 gauge needle; thick, dark fluid was aspirated; a 0.035 in. Jagwire was advanced via the needle into the cyst; the needle was withdrawn; a 7 French triple lumen needle-knife papillotome (Wilson Cook Medical, Winston-Salem, USA) was fed over the Jagwire; the cyst wall was incised using the papillotome (Figure 2B), the papillotome was exchanged with a pyloric balloon dilator, and the aperture was serially dilated up to 18 mm using increasingly larger balloons (Figure 2C), creating a large aperture (Figure 2D). Copious dark, thin, fluid seeped out of the cyst and was suctioned. The echoendoscope was withdrawn and a pediatric colonoscope was intubated and advanced into the cyst, and 800 cc of dark, brown fluid was aspirated via the colonoscope (Figure 2E, after cyst aspiration). After confirming that no lesions were endoscopically visible within the DDC, random biopsies were taken of DDC mucosa, which demonstrated no dysplasia or tumor within the DDC by subsequent histological analysis. Insertion of a double pig tail catheter within the cyst was considered to prevent fenestration closure, but was not performed due to a difficult cyst position for this cannulation. No bleeding occurred during the procedure.

The patient was administered IV ciprofloxacin for 3 days after the procedure. The patient was discharged 3 days after the procedure feeling well, and tolerating oral feedings without nausea and vomiting. One month later the patient was asymptomatic, was tolerating oral feedings without nausea and vomiting, and had regained 7 kg in weight. Repeat EGD revealed minimal extrinsic compression of duodenum, and a wide aperture to the cyst. EUS revealed a residual 1.5×2.0 cm cyst. The orifice of the prior cyst-duodenostomy could not be identified using either straight-viewing or side-viewing (ERCP) endoscopes. A needle was not passed to aspirate the residual cyst under endosonographic guidance to avoid traversing pancreatic parenchyma.