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 ± 4.6 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. 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...
“duplication”; or "cyst" and "endoscopy" or "endoscopic" or "therapy" and "decompression"; with reconciliation of cited references. One article written in French, and one article written in Spanish 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. DDC are the least common small bowel duplication cysts, preceded by ileal and jejunal duplication cysts. In a comprehensive meta-analysis, 47 DDC were reported from 1999 to 2009. About 60% of cases are diagnosed during infancy and childhood, and 40% are diagnosed in adulthood.

By definition, a DDC must adhere to the duodenum, contain a smooth muscle layer in its walls, and be lined by duodenal epithelium. DDC usually share the blood supply with the rest of the duodenum. DDC are usually filled with clear fluid, but can contain bile, pancreatic fluid, or gallstones if they connect to the pancreaticobiliary ducts.

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.

DDC can communicate directly with the true lumen via the duodenum, or via the pancreaticobiliary ducts, or have no communication. About 30% of DDC communicate with the pancreaticobiliary ducts. DDC are also classified according to shape as cystic or elongated/tubular. DDC are most commonly cystic.

Clinical Course

DDC are usually asymptomatic and incidentally discovered during radiologic imaging or esophagogastroduodenoscopy (EGD). They can remain clinically silent for many years. Symptoms, when present, most commonly include abdominal pain, followed by nausea in 80%, and vomiting in 40%. The most common complication is acute pancreatitis.

Other complications include: jaundice, biliary obstruction, cyst infection, intussusception, cholestasis, and hepatitis. DDC may contain ectopic pancreatic or gastric tissue that can cause duodenal ulcers, gastrointestinal (GI) bleeding, or rarely duodenal perforation.

Diagnosis

DDC are strongly suggested by characteristic radiologic imaging findings, but imaging findings are occasionally misleading. Twin major goals of radiologic imaging are to determine that the lesion is fluid-filled and to locate the cyst attachment site. Contrast radiographs reveal a submucosal or extrinsic mass or filling defect. 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. CT is useful to depict cyst location and dimensions, potential communications, and other anomalies. 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. 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. Cysts may contain keratin and desquamated cells, which can appear solid on US.

Definitive diagnosis requires EGD with endosonographic confirmation, or surgery with pathologic analysis of resected tissue. 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 dysplasia of duodenal wall, pancreatic pseudocyst, duodenal polyp, cystic pancreatic tumors, lymphatic malformation, mesenteric cyst, omental cyst, and gastrointestinal stromal tumor. Endoscopic ultrasonography (EUS) can confirm the presence of muscular layer continuity between the duodenum and its duplication.

Therapy

Asymptomatic cysts are usually managed expectantly, but some authorities recommend intervention based upon potential complications, including malignant transformation, especially in patients who are unlikely to follow-up. Prospective studies of the natural history of DDC are, however, unavailable. Symptomatic DDC generally mandate endoscopic or surgical therapy. Moreno et al., 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, whereas others recommend therapeutic endoscopy to establish free drainage. Surgical extirpation entails higher morbidity, and appreciable mortality as compared with endoscopic therapy, but eliminates the risk of malignant cyst degeneration.

Pancreaticoduodenectomy is sometimes necessary intraoperatively if the DDC is too close to the pancreaticobiliary ducts. Less invasive, surgical techniques, including partial cyst resection, internal derivation, and marsupialization, have become obsolete.

Systematic Literature Review of Therapeutic EGD

Systematic literature review revealed 28 reported cases of endoscopic therapy (Table 1). 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.
| First Author, Year of Publication | Age in years | Sex | Symptoms (Clinical Syndrome) | Physical Examination | Laboratory Findings | Abdominal Imaging Findings | Endoscopic Therapy | Follow-Up |
|----------------------------------|--------------|-----|------------------------------|----------------------|---------------------|---------------------------|-------------------|----------|
| 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 (NI: 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 |
| 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. MRCP performed | 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 |

Endoscopic Decompression of Duodenal Duplication Cyst
| First Author, Year of Publication | Age in years, Sex | Symptoms (Clinical Syndrome) | Physical Examination | Laboratory Findings | Abdominal Imaging Findings | Endoscopic Therapy | Follow-Up |
|----------------------------------|------------------|------------------------------|----------------------|---------------------|----------------------------|-------------------|-----------|
| 9. Chryssostalis et al, 2007, a | 49, 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, b | 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, c       | 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, d | 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, peripapillary 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, e    | 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: 2cm wide cystic structure protruding into descending duodenum. ERCP: no communication with CBD or MPD | Decompression and marsupialization using needle knife and a standard biliary papillotome with a semicircular incision; deroofing using a polypectomy snare | Asymptomatic during 4 years of follow-up |
| First Author, Year of Publication | Age | Sex | Symptoms (Clinical Syndrome) | Physical Examination | Laboratory Findings | Abdominal Imaging Findings | Endoscopic Therapy | Follow-Up |
|----------------------------------|-----|-----|------------------------------|---------------------|-------------------|--------------------------|--------------------|-----------|
| 14. Meier and Mellinger, 2012,25  | 45  | 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 caudal 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        | 52  | 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 | 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         | 53  | 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 | Partial resection of lower part of cyst using polypectomy snare and minor papilla sphincterotomy | Asymptomatic at 10 months follow-up |
| 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 | 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 | Asymptomatic at 1 year of follow-up |

Presenting as epigastric or abdominal pain
| First Author, Year of Publication | Age | Sex | Symptoms (Clinical Syndrome) | Physical Examination | Laboratory Findings | Abdominal Imaging Findings | Endoscopic Therapy | Follow-Up |
|----------------------------------|-----|-----|-------------------------------|----------------------|---------------------|--------------------------|-------------------|-----------|
| Wada et al, 2001<sup>55</sup>     | 55  | F   | Intermittent epigastric and back pain | Normal physical examination | Normal routine laboratory tests | US: 3.6 x 3.2 x 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 |
| Gyökeres et al, 2001<sup>56</sup> | 56  | F   | Intermittent upper abdominal pain | Normal physical examination | Normal routine laboratory tests | US: 6.5 x 3.0 x 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 |
| Vandenbroucke et al, 2005<sup>57</sup> | 57  | 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 |
| Jung et al, 2009<sup>58</sup>    | 58  | M   | Sudden onset of RUQ pain | Mild abdominal tenderness without rebound tenderness in right upper quadrant | ALT = 511 U/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 x 2.5 x 1.2 cm | Asymptomatic with normal routine lab tests at 6 months follow-up |
| Martínez-Alcalá García et al, 2014<sup>5</sup> | 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 1 year of follow-up |
| First Author, Year of Publication | 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 |
| 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. | Follow-up MRCP: cyst resolution |
| 26. Antaki et al, 2008,68       | 72, M            | Jaundice                     | Not reported | Not reported | MRCP | ERCP 3 months later: patent orifice of cystoduodenostomy, with diminished cyst size and no stones. Patient asymptomatic at follow-up | ERCP: asymptomatic at 55 months follow-up |
| 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, midepigastic 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/µm³, 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 | 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 | 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 | 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.
| 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 | 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 | 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 | Unremarkable | 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 | 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 hyperechoic 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 Unable to grasp lesion with snare because contour was dented. Cyst wall was then resected in piecemeal fashion using a snare 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 |
| 4. Lee et al, 2011 | 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 | NR |
| 5. Eom et al, 2011 | 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 | 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 | Age in Years, Sex | Symptoms (Clinical Syndrome) | Physical Examination | Laboratory Findings | Abdominal Imaging Findings | Endoscopic Therapy | Follow-Up |
|---------------------------------|-----------------|-------------------------------|----------------------|-------------------|---------------------------|------------------|----------|
| Woolfolk et al, 1998,          | 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, homogenous 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 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

| First Author, Year of Publication | Age in Years, Sex | Symptoms (Clinical Syndrome) | Physical Examination | Laboratory Findings | Abdominal Imaging Findings | Endoscopic Therapy | Follow-Up |
|---------------------------------|-----------------|-------------------------------|----------------------|-------------------|---------------------------|------------------|----------|
| Mou et al, 2015,               | 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 |
| Ivekovic et al, 2012,          | 51, M           | Recent dysphagia for solid foods | NR | NR | EGD and esophagography: submucosal lesion in lower third of esophagus protruding into lumen | 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 |
| Ivekovic et al, 2012,          | 32, F           | Recent dysphagia for solid foods | NR | NR | EUS: hypoechoic 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 |
| Age in Years, Sex | Symptoms (Clinical Syndrome) | Physical Examination | Laboratory Findings | Abdominal Imaging Findings | Endoscopic Therapy | Follow-Up |
|------------------|-----------------------------|---------------------|-------------------|--------------------------|-------------------|-----------|
| 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 | Three metallic clips successfully applied to close esophageal leak | On follow-up, denied dysphagia and regained premorbid weight |
| 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. 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 | CT and MRI: suggested esophageal duplication cyst, but communication with native esophagus was unclear | Eleven months later patient tolerated normal diet without symptoms, and was gaining weight |
| 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 | Gastrograffin esophagogram: esophageal leak into posterior mediastinum | 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 |

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www.md-journal.com
| First Author, Year of Publication | Age in Years, Sex | Symptoms (Clinical Syndrome) | Physical Examination | Laboratory Findings | Abdominal Imaging Findings | Endoscopic Therapy | Follow-Up |
|----------------------------------|------------------|-------------------------------|----------------------|--------------------|---------------------------|-------------------|-----------|
| Will et al, 2005                 | 75 M             | Dysphagia for several weeks  | NR                   | NR                 | 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. | EGD: extrinsic impression on the lower esophagus | 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. |

**Notes:**

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

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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), and one patient presenting with obstructive jaundice and RUQ abdominal pain (patient-25). Only 1 patient had chronic GI blood loss attributed to DDC (patient-47). 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. 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), or by leaving a cyst drain for 3 days for tamponade and monitoring the bleeding (patient-19). 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). One patient who had presented with an asymptomatic DDC remained asymptomatic for 3 years after endoscopic therapy (patient 29). 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. Surgery is necessary if a symptomatic cyst cannot be approached endoscopically. Before therapeutic endosonography, the patient should be advised of a potential need for traditional surgery if therapeutic endoscopy fails. Comprehensive review of 93 gastric duplication cysts reported in the literature revealed 7 cases treated endoscopically. In all these cases the endoscopic therapy was successful without complications (Table 2A). 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). 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                        |
| Mean ± S.D. maximal cyst diameter                | 3.2 ± 1.5 cm                     | 14.5 cm (N = 1)                |
| Primary technique of endoscopic fenestration: NKP or papilotomy 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.

§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 irradiation 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.[67]

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 ductal 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 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 chronic inflammation without dysplasia.

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.
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 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.

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 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. D, 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.

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.