Abstract
Alimentary tract duplications are rare congenital cystic or tubular malformations that are usually found adjacent to a segment of the gut, sharing a common wall but are occasionally located at a more distant site. They are usually single, variable in size, and characterized by a well-defined coat of smooth muscle and an epithelial lining consisting of some type of gut mucosa. They may be asymptomatic and discovered incidentally on physical examination or during imaging for unrelated conditions, e.g., at prenatal sonography. However, the majority cause symptoms, most often in early childhood, but sometimes not until adult life. Presenting symptoms and signs are related to the site and size of the duplication and whether it contains ectopic gastric mucosa, which predisposes to mucosal ulceration and bleeding. Various associated congenital anomalies have been reported, particularly vertebral anomalies. Duplication cysts should be excised completely to relieve symptoms and avoid the risk of complications, including malignant degeneration in adulthood. After complete excision, long-term outcome is generally excellent.

Keywords
Duplication cyst • Enteric cyst • Neurenteric cyst

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Introduction

Alimentary tract duplications are rare and diverse congenital malformations found anywhere from the mouth to the anus (Ladd and Gross 1940; Patiño Mayer and Bettolli 2014). They are also known as enteric cysts, enterogenous cysts, or reduplication cysts. When associated with intradural spinal pathology, they are frequently referred to as neurenteric cysts (Alrabeeah et al. 1988).

Although most enteric duplications are cystic, some are tubular in shape. They vary widely in size. Enteric duplications have two characteristic features: (i) the presence of a well-defined coat of smooth muscle and (ii) an epithelial lining consisting of some type of gut mucosa. Most but not all duplications share a common muscular wall and blood supply with the adjacent part of the gut. They are usually single but are multiple in 5–15% of cases, e.g., an individual with an esophageal and small bowel duplication cyst. The epithelial lining of a duplication cyst is usually similar to that of the adjacent gut but ectopic mucosa (e.g., gastric, respiratory, and/or pancreatic ductal epithelium) may be present. Ectopic gastric mucosa is found in 20–30% of duplications (Bower et al. 1978; Holcomb et al. 1989; Stringer et al. 1995) (Fig. 1). Ectopic respiratory epithelium has been documented in enteric duplication cysts as far distally as the ileum (De Roeck et al. 2008). While enteric duplications may communicate with the lumen of the gut, most are noncommunicating. Tubular lesions are more likely to be communicating and contain ectopic gastric mucosa. Occasionally, enteric duplications are found separate from the alimentary tract, e.g., in the retroperitoneum or vertebral canal. Enteric duplications are generally classified according to their location rather than their mucosal lining.

Thus, a gastric mucosa-lined cyst lying adjacent to the esophagus in the posterior mediastinum is termed an esophageal duplication cyst.

The incidence of enteric duplications is unknown but a figure of 1 in 4,500 based on fetal and neonatal autopsy data is often quoted. Overall, there is a slight male predominance but colonic duplications may be more common in girls (Temiz et al. 2013).

Pathogenesis

There is no satisfactory unifying explanation to account for the pathogenesis of all gastrointestinal duplications. They are understood to arise from defective embryogenesis. Several mechanisms have been proposed:

(i) Partial twinning – some hindgut duplications, particularly those with complete doubling of the genitalia, bladder, and colon (Ravitch 1953) are probably an expression of caudal twinning.

(ii) Split notochord theory – abnormal adherence of the endoderm of the developing gut tube to the notochord may account for enteric duplications associated with vertebral anomalies (Bentley and Smith 1960).

(iii) Intrauterine mesenteric vascular accident – this might explain the origin of duplication cysts associated with intestinal atresia (Favara et al. 1971).

(iv) Disordered foregut budding – this could account for some foregut duplication cysts.

(v) Disordered recanalization of the developing gut – this may explain enteric duplications arising at sites where the developing gut is temporarily occluded by epithelial proliferation (Gross et al. 1952). This mechanism could also explain rare cases of triplication or even quadruplication of segments of the intestine.

Distribution and Presentation

Most enteric duplications cause symptoms in infancy or early childhood, although they may remain asymptomatic until adult life. An increasing proportion is now detected before the onset of symptoms, presenting either as an incidental mass on physical examination or during imaging for unrelated symptoms or another condition. The
latter includes duplications that are detected by routine prenatal ultrasound scans.

The distribution of alimentary tract duplications based on the four largest single institution series (to reduce publication bias) is shown in Table 1 and schematically in Fig. 2. Almost 80% of all enteric duplications are intra-abdominal and approximately 50% are related to the midgut, with ileal being the commonest.

Presenting symptoms and signs depend on the location and size of the duplication, and whether it contains ectopic gastric mucosa. Duplications commonly cause obstruction – of the airway (coughing, wheezing, and/or pneumonia), esophagus (dysphagia), or intestine (abdominal pain and vomiting) (Erginel et al. 2017). Those lined by ectopic gastric mucosa are prone to ulceration, hemorrhage, or perforation. Intussusception and segmental volvulus are additional potential complications of small bowel duplications. Occasionally, duplications are complicated by infection.

In adults, carcinoma (most often adenocarcinoma) has been reported as a complication of duplication cysts in the esophagus (Singh et al. 2001), stomach (Zheng and Jing 2012), duodenum (Chen et al. 2010), small bowel (Blank et al. 2012), colon (Hsu et al. 2011), and rectum (Michael et al. 1999). Many of these tumors are advanced and associated with metastases at presentation. To date, there have been no recorded instances of malignant degeneration in children under 16 years of age.

Clinical Features

Clinical features of alimentary tract duplications are best considered according to their location.

Lingual

Most are located in the anterior two-thirds of the tongue (Fig. 3). The differential diagnosis includes lymphatic malformation, ranula, and hemangioma. Duplication cysts in the tongue may be asymptomatic or present with feeding or breathing difficulties in the newborn (Hambarde et al. 2011). If detected prenatally, the potential for neonatal airway obstruction should be considered; these cases need to be followed closely with serial ultrasound scans to anticipate problems at or after delivery, including the small possibility of an ex-utero intrapartum treatment procedure. Dynamic magnetic resonance imaging of the fetus may provide additional information (Houshmand et al. 2011). Duplication cysts of the tongue may be lined by gastric mucosa (Hambarde et al. 2011). Late malignant degeneration has been described (Volchok et al. 2007). They are best treated by complete surgical excision.

Esophageal

Esophageal duplication cysts are typically located in the posterior mediastinum, more often on the right side, but may also be found in the neck (Fig. 4). Most do not communicate with the esophageal lumen. Esophageal duplications may cause respiratory problems such as stridor, tachypnea, chest infection, or feeding difficulties in infants (Nayan et al. 2010) and dysphagia in older children. There may be palpable neck swelling. If the duplication contains ectopic gastric mucosa and communicates with the esophageal lumen, peptic esophageal stricture is a potential complication (Stringer et al. 1995).

Associated thoracic or cervical vertebral anomalies should be excluded; if present, a potential intraspinal extension should be considered.
Table 1  Large single institution series of children with alimentary tract duplication cysts

| Author                  | No. of duplications (no. of patients) | Cervical (oropharyngeal) | Mediastinal/ esophageal | Thoracoabdominal | Gastric | Duodenal | Jejunal + Ileal<sup>a</sup> | Colonic | Appendix | Distal colonic | Rectal | Other sites |
|-------------------------|---------------------------------------|---------------------------|-------------------------|------------------|---------|----------|-----------------------------|---------|----------|-----------------|--------|-------------|
| Gross et al. (1952) (Boston) | 68 (67)                               | 1                         | 13                      | 3                | 2       | 4        | 4 + 28                       | 5       | 3         | 5               |        |             |
| Bower et al. (1978) (Pittsburgh) | 74 (64)                               | 15                        | 1                       | 7                | 3       | 6 + 28   | 8                            | 2       | 2         | 2               |        |             |
| Holcomb et al. (1989) (Philadelphia) | 101 (96)                              | 21                        | 3                       | 8                | 2       | 12 + 35  | 13                           | 1       | 1         | 5               |        |             |
| Stringer et al. (1995) (London)   | 77 (72)                               | 2                         | 15                      | 6                | 10      | 3        | 5 + 16                       | 6       | 3         | 1               | 6      | 3 Retroperitoneal |
| Total                    | 320                                   | 3 (1%)                    | 64 (20%)                | 13 (4%)          | 27 (8%) | 12 (4%)  | 27 (8%) + 107 (33%)         | 32 (10%)| 6 (2%)    | 5 (2%)          | 18 (6%)| 6 (2%)     |

<sup>a</sup>Including ileocecal
Esophageal duplications must be distinguished from neurogenic tumors (neuroblastoma and ganglioneuroma) and bronchogenic cysts (Fig. 5). Bronchogenic cysts are similar to esophageal duplications but are understood to arise from parts of the foregut destined for respiratory differentiation. They are lined by ciliated columnar epithelium and their walls contain bronchial mucous glands, smooth muscle, and hyaline cartilage (Tireli et al. 2004; Jiang et al. 2015). Bronchogenic cysts are usually unilocular and filled with clear fluid but they may contain air or pus if they have a patent connection with the airway. Most bronchogenic cysts are located in the mediastinum, although they may be found in the lung parenchyma, neck, or at more distant sites.

Thoracoabdominal

A thoracoabdominal duplication usually descends as a tubular structure to the right of the esophagus (but slightly separate from it) in the posterior mediastinum. It communicates through the diaphragm with the stomach, duodenum, pancreas, jejunum, or ileum (Stringer et al. 1995). Thoracic and/or cervical vertebral anomalies may be
present and the cyst may have an intradural connection. They are frequently lined by ectopic gastric mucosa. Thoracoabdominal duplications can therefore present with respiratory distress, gastrointestinal bleeding, vomiting, and even meningitis.

Gastric

Gastric duplication cysts are most often located on the greater curvature of the stomach or in the pyloric region. Cystic varieties are more common and, unlike tubular lesions, these rarely communicate with the stomach lumen. Very occasionally, a gastric duplication communicates with a pancreatic duct (Hoffman et al. 1987; Moss et al. 1996). Gastric duplications may present as an asymptomatic mass detected on abdominal examination or on ultrasound scan (including prenatal) or with vomiting and/or bleeding. In infants, they may cause pyloric obstruction and mimic infantile hypertrophic pyloric stenosis. Rarely, they present with perforation.

Duodenal

Duodenal duplications are variable in size but frequently measure 2–5 cm in diameter; they may be dominantly intraluminal or extraluminal (Lopez-Fernandez et al. 2013). Most are located medial or posterior to the second or third part of duodenum and can therefore be confused with type III choledochal cysts (choledochoceles) (“Congenital Biliary Dilatation” chapter). Almost 50% communicate with the pancreatic and/or bile duct or with a smaller pancreatic duct or the duodenal lumen (Chen et al. 2010). There is one report of a duodenal duplication cyst communicating with a duplicated gallbladder (Menon et al. 2013).

Presentation is often with nonspecific symptoms such as upper abdominal pain and vomiting, making delays in diagnosis common (“Duodenal Obstruction” chapter). Acute pancreatitis...
(which may be recurrent) is a presenting feature in more than half the cases (Chen et al. 2010) (“▶ Pancreatic Disorders” chapter). Other potential clinical manifestations include biliary obstruction and intestinal bleeding; the latter is related to ectopic gastric mucosa which is uncommon according to a meta-analysis (Chen et al. 2010) but was present in 6 of 11 cases in one series (Lopez-Fernandez et al. 2013).

**Pancreatic**

There is some confusion about the classification of duplication cysts within the pancreas. They are variably reported as pancreatic, gastric, or duodenal duplication cysts. Some are clearly intrapancreatic while others are in continuity with the stomach or duodenum. The presence of gastric epithelium may signify a gastric origin or may represent ectopic gastric mucosa. Since enteric duplication cysts are classified by their location rather than their mucosal lining, those primarily associated with the stomach or duodenum are gastric and duodenal duplications, respectively even though they may communicate with a pancreatic duct. In contrast, duplication cysts within the pancreas should be regarded as pancreatic duplication cysts, irrespective of their mucosal lining. About half are located in the head of the gland and they may communicate with the pancreatic duct (Fujishiro et al. 2011).

Pancreatic duplication cysts most commonly present with abdominal pain and vomiting, with or without evidence of acute pancreatitis (“▶ Pancreatic Disorders” chapter). They may be confused with a pancreatic pseudocyst or cystic neoplasm (Hunter et al. 2008).

**Small Bowel**

This is the commonest site for an enteric duplication (Table 1); most are ileal or ileocecal. They are usually cystic but can be tubular (when they are more likely to communicate with the bowel lumen) and typically lie on the mesenteric aspect of the intestine (Fig. 6). Cystic duplications may be detected incidentally as a mobile mass on abdominal examination or as a cystic mass on ultrasound scan (or other cross-sectional imaging), particularly in the fetus. Others can present acutely with intestinal obstruction due to luminal compression by the mass or from a volvulus or intussusception; such cases may be confused with acute appendicitis (Fig. 7). Tubular small bowel duplications are more likely to contain ectopic gastric mucosa and therefore may present with intestinal bleeding (“▶ Gastrointestinal Bleeding” chapter) or perforation.

**Colonic**

Hindgut duplications largely fall into three groups: duplication cysts of the colon, tubular duplications of the rectum and/or colon, and duplication of the appendix.

Colonic duplications may be cystic or tubular. Ectopic gastric mucosa is uncommon (Stringer et al. 1995; Temiz et al. 2013). In the rare total colonic duplication, the duplicated bowel is located lateral or medial to the normal colon and usually has a proximal communication (Ravitch 1953). Colonic duplications tend to present with abdominal pain, constipation, and/or bleeding. Cecal duplication cysts may mimic an appendix mass (Temiz et al. 2013). Extensive tubular duplications of the colon and rectum may be discovered during the investigation of associated genitourinary and spinal anomalies.

**Rectal**

As with other duplications, these may be cystic or tubular. They may manifest as a perineal mucosal swelling and/or a perianal or perineal fistula, causing confusion with perianal sepsis (Flint et al. 2004; La Quaglia et al. 1990). Alternatively, they may obstruct defecation or cause rectal prolapse or bleeding. Cystic rectal duplications must be distinguished from cystic sacrococcygeal teratoma, tailgut cyst, and anterior meningocele. When associated with an anorectal malformation
and sacral vertebral anomalies, the cyst may be part of the Currarino triad (Currarino et al. 1981).

**Prenatal Diagnosis**

Enteric duplications may be identifiable as early as 12 weeks’ gestation (Chen et al. 2002) but most of those that are detected prenatally are first observed at or after 20 weeks’ gestation (“Antenatal Diagnosis” and “Prenatal Diagnosis of Congenital Malformations” chapters). Prenatal complications are rare (Laje et al. 2010): There is one report of successful thoracoamniotic shunting in a fetus with a thoracic duplication cyst causing mediastinal shift and hydrops fetalis (Martínez Ferro et al. 1998) and another of a large ileal duplication cyst in a fetus that was drained percutaneously to relieve umbilical vein obstruction (Ness et al. 2006). A localized volvulus of an intestinal duplication cyst in the fetus may be a rare cause of intestinal atresia (Sinha et al. 1992).

Enteric duplications account for approximately 10% of prenatally diagnosed intra-abdominal cysts (Thakkar et al. 2015). Distinguishing these cysts from other fetal intra-abdominal cysts such as ovarian, choledochal, and mesenteric cysts can be difficult. Some centers use prenatal magnetic resonance imaging to characterize these cysts (Laje et al. 2010) but this is unlikely to influence postnatal management. If postnatal imaging confirms an enteric duplication, symptomatic cases require prompt surgery. Asymptomatic lesions need close postnatal follow-up and surgery is advisable during the first 6 months of life because of the potential for serious complications (Laje et al. 2010).

**Associated Anomalies**

Associated anomalies are present in 30% to 50% of patients with enteric duplication cysts (Stringer et al. 1995).

*Vertebral anomalies* are more likely with thoracic and hindgut duplications (Bower et al. 1978; Stringer et al. 1995) but may be found with duplications at other sites, including the small bowel (Li et al. 1998). Between 20% and 30% of thoracic duplications have associated vertebral anomalies (Bower et al. 1978; Holcomb et al. 2006). A localized volvulus of an intestinal duplication cyst in the fetus may be a rare cause of intestinal atresia (Sinha et al. 1992).
1989; Stringer et al. 1995). These include cleft vertebrae, hemivertebrae, spina bifida, vertebral fusion, and scoliosis (Fig. 8). There may be associated intradural spinal pathology including a direct communication between the cyst and the dural sac (Alrabeeah et al. 1988).

Numerous other malformations have been reported infrequently in association with enteric duplications. They include congenital cardiac disease, esophageal atresia, congenital diaphragmatic hernia, congenital pulmonary malformations, and myelomeningocele with foregut duplications (Stringer et al. 1995; Iyer and Mahour 1995); intestinal malrotation or less commonly intestinal atresia with midgut duplications; and genitourinary duplication, bladder extrophy, and imperforate anus with hindgut duplications (Bower et al. 1978; Holcomb et al. 1989; Stringer et al. 1995; Gross et al. 1952).

**Imaging**

The choice of imaging is dictated by the site of the duplication, clinical urgency, and the potential for associated anomalies. Isolated small bowel duplications require few preoperative investigations other than abdominal sonography and plain radiography.

- Plain radiography: a chest radiograph may show a posterior mediastinal mass with an esophageal duplication cyst. Plain radiographs also demonstrate vertebral anomalies.
- Ultrasound is especially useful for intra-abdominal enteric duplications. Most duplication cysts have an anechoic center unless there has been bleeding into the cyst. Two sonographic features strongly support the diagnosis of a duplication cyst: the “double-wall sign” (an inner hyperechoic rim corresponding to the mucosa/submucosa and an outer hypoechoic layer corresponding to the muscle coat) and the presence of peristalsis (Fig. 9). This “gut signature” is highly suggestive but not pathognomonic of a duplication cyst since it is occasionally seen with other intra-abdominal cysts such as ovarian and mesenteric cysts (Cheng et al. 2005). Urinary tract ultrasound is often advisable in the evaluation of pelvic duplication cysts. Endoscopic ultrasound may be advantageous in duodenal and pancreatic duplications (Liu and Adler 2014).
- Contrast studies such as a contrast swallow, meal, or enema may be helpful when evaluating esophageal and gastrointestinal duplication cysts (Fig. 10). A “fistulogram” is valuable in investigating rectal duplications with a perineal opening (Fig. 11).
- Magnetic resonance or CT cross-sectional imaging offers the best method of delineating thoracic and pelvic duplications in particular. The precise location and extent of a thoracic duplication cyst can be mapped together with
associated vertebral and spinal anomalies. Most duplications have low signal intensity on T1-weighted MR images and high signal intensity on T2-weighted images (Hur et al. 2007). Magnetic resonance cholangiopancreatography (MRCP) can provide useful information when evaluating a duodenal or pancreatic duplication cyst and MR angiography in large retroperitoneal lesions. On CT, enteric duplications often show peripheral enhancement and may need to be distinguished from an abscess, although this is usually evident clinically.

- A technetium-99m pertechnetate radionuclide scan to detect ectopic gastric mucosa (Kiratli et al. 2009) is useful in selected duplications: (i) in when evaluating a child with abdominal pain and GI bleeding, (ii) when deciding the urgency of surgery in a child with an asymptomatic duplication cyst, and (iii) when indicating the approximate extent of a tubular duplication (Fig. 12).
- Endoscopy: endoscopic retrograde cholangiopancreatography (ERCP) is helpful in defining the relationship of a duodenal duplication cyst to the pancreatic and bile ducts. Flexible upper and lower gastrointestinal endoscopy may

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**Fig. 9** Ultrasound scan of an ileal duplication cyst showing the “double-wall sign”: an inner hyperechoic rim (*thick arrows*) corresponds to the mucosa/submucosa and an outer hypoechoic layer (*thin arrows*) corresponds to the muscle coat.

**Fig. 10** A contrast swallow in a child with a tubular esophageal duplication that was lined by ectopic gastric mucosa (*thin arrow*) and communicated with the distal native esophagus (*thick arrow*).

**Fig. 11** A contrast study in a child with a rectal duplication associated with a perineal opening.
Surgical Treatment

The optimum treatment of alimentary tract duplications is complete excision. This avoids future complications including the risk of malignant degeneration. Occasionally, complete resection is deemed to be too hazardous when the aim should be to remove the entire mucosal lining of the cyst, especially if it contains ectopic gastric mucosa. Only rarely is it necessary to leave an intact part of the duplication in situ (see below).

If the expertise is available and the same result can be achieved with minimally invasive surgery, then a thoroscopic or laparoscopic approach is preferable because it is associated with reduced postoperative discomfort and improved cosmesis. However, the emphasis should be on complete excision rather than the surgical technique.

All patients should receive prophylactic intravenous broad spectrum antibiotics at induction of anesthesia.

Esophageal

Cervical esophageal duplications can be removed via a cervical incision (Fig. 13), care being taken to avoid injury to structures such as the recurrent laryngeal nerve. A nasogastric tube in the esophagus may assist with identification.

Thoracic esophageal duplications can be excised via a transpleural posterolateral thoracotomy (Fig. 14). Cystic duplications are usually intramural and noncommunicating and more often related to the right side of the esophagus. It is usually possible to excise the cyst leaving the esophageal mucosa intact. If the cyst cannot be removed intact, then it can either be transected close to the esophagus with stripping of the residual mucosa or a short segment of esophagus can be excised in continuity. Laser ablation of residual cyst mucosa does not reliably prevent recurrence (Merry et al. 1999). The proximity of the vagus nerve and the thoracic duct should be noted. Any communication with the esophageal lumen or breach of the esophageal mucosa must be closed; leak testing with air injected via a nasogastric tube may be helpful. The esophageal repair can be buttressed with the muscular fringe of the duplication. Closing the muscular defect in the esophageal wall probably helps to avoid the development of an esophageal diverticulum (Merry et al. 1999). The chest is closed and pleural drainage is not usually necessary.

Video-assisted thoracoscopic resection is an effective technique for cystic (rather than tubular) duplication cysts (Hirose et al. 2006; Perger et al. 2006). Intraoperative cyst decompression and single lung ventilation may assist with thoracoscopic resection by creating more working space. A flexible endoscope within the esophagus may help to monitor the integrity of the esophageal mucosa intraoperatively (Perger et al. 2006).
Thoracoabdominal

These are potentially very challenging resections and detailed preoperative assessment of the mediastinal, abdominal, and potential spinal components of the duplication is critical. The duplication cyst may be adherent to thoracic or cervical vertebrae and can have an intradural connection (Stringer et al. 1995). The duplication may extend distally to the stomach, duodenum, pancreas, or ileum but often appears tenuous as it passes behind the diaphragm. The abdominal portion most often appears as a tubular lesion communicating with the small bowel but may occasionally end blindly along the greater curve of the stomach. These duplications are generally best excised through separate thoracic (right-sided posterolateral thoracotomy) and abdominal incisions, rather than a single oblique thoracoabdominal incision (Fig. 15). When the cyst communicates with the vertebral canal, neurosurgical input is advisable. A staged approach, excising each component sequentially, should be avoided (Stringer et al. 1995).

Incomplete excision is a particular risk with thoracoabdominal duplications because of their size and complexity; the potential consequences of this include meningitis, gastrointestinal bleeding and perforation, and respiratory complications.
Greater curvature or pyloric duplication cysts can usually be completely excised by dissecting the cyst off the gastric submucosa and repairing the residual seromuscular defect. Gastric mucosal integrity is checked prior to seromuscular repair by insufflating air via a nasogastric tube. This can be achieved by open or laparoscopic surgery.

Small gastric duplications are occasionally more simply excised by a wedge resection of the cyst and a segment of stomach followed by a two layer gastric closure. Extensive duplications of the greater curvature of the stomach can be treated by partial resection and stripping of the residual mucosal lining followed by repair. A less ideal approach is to divide the septum separating the tubular duplication from the gastric lumen with linear staplers introduced via proximal and distal gastrotomies. The division should be as complete as possible but since the mucosal lining of the duplication has not been removed, there remains a risk of long-term mucosal complications.

Some duodenal duplications are simple cystic lesions that can be excised easily without risk of injury to pancreatic or biliary ducts. Those on the medial aspect of the second or third part of the duodenum may be complex and communicate with the pancreatic and/or common bile duct. Previous pancreatitis or inflammation secondary to ectopic gastric mucosa may make surgical dissection more difficult. Facilities for intraoperative cholangiopancreatography should be available with prior positioning of the child on the operating table allowing for image intensifier access.

In open surgery, the duodenum is approached via a right upper quadrant incision and “Kocherised” to elevate it into the wound. The adjacent peritoneal cavity is isolated with swabs and an oblique incision is made in the lateral wall of the descending duodenum to expose the medially situated duplication cyst (Fig. 16). If the opening of the common bile duct or pancreatic duct is uncertain, intraoperative cholangiography may be helpful (Fig. 17). Alternatively, a cholecystectomy...
can be performed and a fine probe passed distally through the cystic and common bile duct into the duodenum.

Once the duplication cyst is exposed, surgical alternatives are dictated by the anatomy. They include (i) complete excision of the cyst with division of any ductal communication (optimum); (ii) partial excision and mucosectomy of the remaining part of the cyst (Lopez-Fernandez et al. 2013), which can be achieved laparoscopically in selected cases (Byun et al. 2014); and (iii) if the duplication is adjacent to the ampulla of Vater, it can be fenestrated into the duodenal lumen, cautiously oversewing the edges of the window to achieve hemostasis. If the latter option is selected, the presence of ectopic gastric mucosa must be excluded by intraoperative biopsy and the fenestration must be of sufficient size to permit free dependent drainage without forming a cul-de-sac. Pancreatoduodenectomy is rarely required (Chen et al. 2010).

There are recent reports of endoscopic marsupialization of duodenal duplication cysts, mostly in adults (Chen et al. 2010). However, the paucity of long-term results of this technique is a particular concern in children because malignant degeneration has been described in adults (Seeliger et al. 2012).

**Pancreatic**

Surgical options include complete local excision with or without Roux loop drainage of the residual cavity or a Whipple-type pancreatoduodenectomy. Roux loop drainage of the cyst alone is inadequate because the mucosal lining of the cyst must be completely excised if pancreatitis is to be prevented. When the duplication is in the tail of the pancreas, distal pancreatectomy with splenic preservation (laparoscopic or open) can be performed.

**Small Bowel**

Cystic lesions of the ileum or jejunum can usually be excised without difficulty using open or laparoscopic techniques. The continuous muscular coats of the normal and duplicated bowel make excision of the cyst alone more demanding than simple segmental resection of the bowel (Fig. 18) but both are acceptable methods. Excising the cyst alone is achievable in some cases by laparoscopy (Laje et al. 2010), although a laparoscopic-assisted resection after preliminary decompression of the cyst and exteriorization of the affected segment of the bowel is often easier. In ileocecal duplication cysts, it may be possible to preserve the ileocecal valve by separately resecting the ileal and cecal components of the cyst (Catalano et al. 2014).
Short tubular small bowel duplications may be excised in continuity with the adjacent bowel. Care should be taken to obtain complete excision of the proximal and distal margins of the lesion where the normal and duplicated bowel merge; distinguishing the two can be difficult. Long tubular duplications, where remaining intestinal length is a potential problem, pose a more difficult problem. The tubular duplication runs parallel to the native bowel between the leaves of the adjacent mesentery risking ischemia of the native intestine with complete resection of the duplication. In these cases, submucosal resection is an alternative but difficult option. The mucosal lining is stripped out using a series of longitudinal seromuscular incisions in the duplication. The residual seromuscular sleeve of the duplication may be safely left in situ. Bleeding within the sleeve almost always stops spontaneously. For duplications within the mesentery but separate from the intestine, careful separation of the two leaves of the mesentery and division of vessels in one leaf only may enable excision of the duplication without jeopardizing the blood supply of the adjacent bowel (Norris et al. 1986) (Fig. 19). Whichever technique is used, the junction between the duplicated and normal bowel must be resected since ectopic gastric mucosa is frequently present at this site.

Associated intestinal malrotation requires a Ladd’s procedure.

**Colonic and Appendiceal**

All cystic and most tubular duplications of the colon can be excised in continuity with an adjacent segment of bowel. In long tubular duplications, distal fenestration is possible provided there is no ectopic gastric mucosa (which is uncommon with colonic duplications); this is conveniently done with a linear stapler introduced through an enterotomy near the distal margin of the duplication (Fig. 20). The distal end of the septum must be divided completely to avoid leaving a spur. If both colons reach the perineum, then a preliminary double defunctioning colostomy may be necessary. The duplicated bowel can later be anastomosed to the native rectum and the mucosa of the redundant distal duplicated colorectal segment excised.

Asymptomatic duplications of the appendix found in association with cloacal or bladder extrophy can be retained for later use in reconstructive surgery.

**Rectal**

Rectal duplications may be excised using one of several approaches. Small submucosal cysts can
be excised via an endorectal route; leaving the cyst intact facilitates dissection (Fig. 21). After excision, the rectal mucosa is repaired. The posterior sagittal approach provides excellent exposure of the retrorectal space for excision of tubular and larger cystic duplications (La Quaglia et al. 1990). With attention to bowel preparation and antibiotic prophylaxis, a covering colostomy may be avoided. Infected rectal duplication cysts are best treated by preliminary perineal drainage followed by resection once the inflammation has settled. Rectal duplications should be completely excised because of the risk of late malignant degeneration (see above).

Retroperitoneal duplication cysts may be very large and are frequently adherent to retroperitoneal structures including the pancreas. Care is required during surgery to avoid injury to mesenteric and renal vessels and nearby viscera. Duplication cysts in the tongue tend to be small and are usually removed via an intraoral, sublingual approach. Intra- or extradural duplication cysts
in the vertebral canal are best managed in conjunction with neurosurgeons.

All excised duplications should be examined histologically.

**Conclusion and Future Directions**

The key steps in the successful surgical management of gastrointestinal duplications include a thorough understanding of the spectrum of these congenital malformations; careful preoperative assessment of the cyst and potential associated pathology; appropriate operative planning; complete excision whenever possible (using open or minimally invasive approaches); and an awareness of alternative techniques and potential pitfalls in complex cases where complete excision may be unduly hazardous. After complete excision, long-term outcome is generally excellent.

**References**

Alrabeeah A, Gillis DA, Giacomantonio M, Lau H. Neuenteric cysts—a spectrum. J Pediatr Surg. 1988;23(8):752–4.

Bentley JF, Smith JR. Developmental posterior enteric remnants and spinal malformations: the split notochord syndrome. Arch Dis Child. 1960;35:76–86.

Blank G, Königsrainer A, Sipos B, Ladurner R. Adenocarcinoma arising in a cystic duplication of the small bowel: case report and review of literature. World J Surg Oncol. 2012;10:55.

Bower RJ, Sieber WK, Kiesewetter WB. Alimentary tract duplications in children. Ann Surg. 1978;188:669–74.

Byun J, Oh HM, Kim SH, Kim HY, Jung SE, Park KW, et al. Laparoscopic partial cystectomy with mucosal stripping of extraluminal duodenal duplication cysts. World J Gastroenterol. 2014;20(4):1123–6.

Catalano P, Di Pace MR, Caruso AM, De Grazia E, Cimador M. Ileocecal duplication cysts: is the loss of the valve always necessary? J Pediatr Surg. 2014;49(6):1049–51.

Chen M, Lam YH, Lin CL. Sonographic features of ileal duplication cyst at 12 weeks. Prenat Diagn. 2002;22(12):1067–70.

Chen JJ, Lee HC, Yeung CY, Chan WT, Jiang CB, Sheu JC. Meta-analysis: the clinical features of the duodenal duplication cyst. J Pediatr Surg. 2010;45(8):1598–606.

Cheng G, Soboleski D, Daneman A, Poenaru D, Hurlbut D. Sonographic pitfalls in the diagnosis of enteric duplication cysts. AJR Am J Roentgenol. 2005;184(2):521–5.

Currarino G, Coln D, Votteler T. Triad of anorectal, sacral, and presacral anomalies. AJR Am J Roentgenol. 1981;137(2):395–8.

De Roeck A, Vervoelsem D, Mattelaer C, Schwagten K. Isolated enteric duplication cyst with respiratory epithelium: case report and review of the literature. Eur J Pediatr Surg. 2008;18(5):337–9.

Erginel B, Soysal FG, Ozbey H, Keskin E, Celik A, Karadag A, et al. Enteric duplication cysts in children: a single-institution series with forty patients in twenty-six years. World J Surg. 2017 Feb;41(2):620–4.

Favara BE, Franciosi RA, Akers DR. Enteric duplications. Thirty-seven cases: a vascular theory of pathogenesis. Am J Dis Child. 1971;122(6):501–6.

Flint R, Strang J, Bissett I, Clark M, Neill M, Parry B. Rectal duplication cyst presenting as perianal sepsis: report of two cases and review of the literature. Dis Colon Rectum. 2004;47(12):2208–10.

Fujishiro J, Kaneko M, Urita Y, Hoshino N, Jinbo T, Sakamoto N, et al. Enteric duplication cyst of the pancreas with duplicated pancreatic duct. J Pediatr Surg. 2011;46(8):e13–6.

Gross RE, Holcomb GW Jr, Farber S. Duplications of the alimentary tract. Pediatrics. 1952;9:448–68.

Hambarde S, Bendre P, Taide D. Foregut duplication cyst presenting as lingual swelling: case report and review of literature. Natl J Maxillofac Surg. 2011;2(1):2–5.

Hirose S, Clifton MS, Bratton B, Harrison MR, Farmer DL, Nobuhara KK, et al. Thoracoscopic resection of foregut duplication cysts. J Laparoendosc Adv Surg Tech A. 2006;16(5):526–9.

Hoffman M, Sugerman HJ, Heuman D, Turner MA, Kisloft B. Gastric duplication cyst communicating with aberrant pancreatic duct: a rare cause of recurrent acute pancreatitis. Surgery. 1987;101(3):369–72.

Holcomb GW, Gheissari A, O’Neill JA, Shorter NA, Bishop HC. Surgical management of alimentary tract duplications. Ann Surg. 1989;209:167–74.

Houshmand G, Hosseinzadeh K, Ozolek J. Prenatal magnetic resonance imaging (MRI) findings of a foregut duplication cyst of the tongue: value of real-time MRI evaluation of the fetal swallowing mechanism. J Ultrasound Med. 2011;30(6):843–50.

Hsu H, Gueng MK, Tseng YH, Wu CC, Liu PH, Chen CC. Adenocarcinoma arising from colonic duplication cyst with metastasis to omentum: a case report. J Clin Ultrasound. 2011;39(1):41–3.

Hunter CJ, Connelly ME, Ghaffari N, Anselm D, Gonzalez I, Shin C. Enteric duplication cysts of the pancreas: a report of two cases and review of the literature. Pediatr Surg Int. 2008;24(2):227–33.

Hur J, Yoon CS, Kim MJ, Kim OH. Imaging features of gastrointestinal tract duplications in infants and children: from oesophagus to rectum. Pediatr Radiol. 2007;37(7):691–9.

Iyer CP, Mahour GH. Duplications of the alimentary tract in infants and children. J Pediatr Surg. 1995;30(9):1267–70.
Jiang JH, Yen SL, Lee SY, Chuang JH. Differences in the distribution and presentation of bronchogenic cysts between adults and children. J Pediatr Surg. 2015;50(3):399–401.

Kiratli PO, Aksoy T, Bozkurt MF, Orhan D. Detection of ectopic gastric mucosa using 99mTc pertechnetate: review of the literature. Ann Nucl Med. 2009;23(2):97–105.

Ladd WE, Gross RE. Surgical treatment of duplications of the alimentary tract: enterogenous cysts, enteric cysts, or ileum duplex. Surg Gynecol Obstet. 1940;70:295–307.

Laje P, Flake AW, Adzick NS. Prenatal diagnosis and postnatal resection of intraabdominal enteric duplications. J Pediatr Surg. 2010;45(7):1554–8.

La Quaglia MP, Feins N, Eraklis A, Hendren WH. Rectal duplications. J Pediatr Surg. 1990;25:980–4.

Li L, Zhang JZ, Wang YX. Vascular classification for small intestinal duplications: experience with 80 cases. J Pediatr Surg. 1998;33(8):1243–5.

Liu R, Adler DG. Duplication cysts: diagnosis, management, and the role of endoscopic ultrasound. Endosc Ultrasound. 2014;3(3):152–60.

Liu R, Adler DG. Duplication cysts: diagnosis, management, and the role of endoscopic ultrasound. Ann Pediatr Surg. 1998;33(8):1243–5.

Lopez-Fernandez S, Hernandez-Martin S, Ramirez M, Ortiz R, Martinez L, Tovar JA. Pyloroduodenal duplication cysts: treatment of 11 cases. Eur J Pediatr Surg. 2013;23(4):312–6.

Martinez Ferro M, Milner R, Voto L, Zapaterio J, Cannizzaro C, Rodriguez S, et al. Intrathoracic alimentary tract duplication cysts treated in utero by thoracoamniotic shunting. Fetal Diagn Ther. 1998;13(6):343–7.

Menon P, Rao KL, Thapa BR, Goyal R, Garge S, Rathore MK, et al. Duplicated gall bladder with duodenal duplication cyst. J Pediatr Surg. 2013;48(4):e25–8.

Merry C, Spurbeck W, Lobe TE. Resection of foregut-derived duplications by minimal-access surgery. Pediatr Surg Int. 1999;15(3–4):224–6.

Michael D, Cohen CR, Northover JM. Adenocarcinoma within a rectal duplication cyst: case report and literature review. Ann R Coll Surg Engl. 1999;81(3):205–6.

Moss RL, Ryan JA, Kozarek RA, Hatch EL. Pancreatitis caused by a gastric duplication communicating with an aberrant pancreatic lobe. J Pediatr Surg. 1996;31(5):733–6.

Nayan S, Nguyen LH, Nguyen VH, Daniel SJ, Emil S. Cervical esophageal duplication cyst: case report and review of the literature. J Pediatr Surg. 2010;45(9):e1–5.

Ness A, Bega G, Wood DC, et al. Massive fetal ileal duplication requiring antenatal intervention. J Ultrasound Med. 2006;25(6):785–90.

Norris RW, Brereton RJ, Wright VM, Cudmore RE. A new surgical approach to duplications of the intestine. J Pediatr Surg. 1986;21:167–70.

Patiño Mayer J, Bettolli M. Alimentary tract duplications in newborns and children: diagnostic aspects and the role of laparoscopic treatment. World J Gastroenterol. 2014 Oct 21;20(39):14263–71.

Perger L, Azzie G, Watch L, Weinsheimer R. Two cases of thoracoscopic resection of esophageal duplication in children. J Laparoendosc Adv Surg Tech A. 2006;16(4):418–21.

Ravitch MM. Hindgut duplications: doubling of the colon and genital urinary tracts. Ann Surg. 1953;137:588–601.

Seeliger B, Piardi T, Marzano E, Mutter D, Marescaux J, Pessaux P. Duodenal duplication cyst: a potentially malignant disease. Ann Surg Oncol. 2012;19(12):3753–4.

Singh S, Lal P, Sikora SS, Datta NR. Squamous cell carcinoma arising from a congenital duplication cyst of the esophagus in a young adult. Dis Esophagus. 2001;14(3–4):258–61.

Singh S, Gangopadhyay AN, Harshwardhan, Gopal SC. Ileal atresia with intestinal duplication. Indian Pediatr. 1992;29(12):1573–4.

Stringer MD, Spitz L, Abel R, Kiely E, Drake DP, Agrawal M, et al. Management of alimentary tract duplication in children. Br J Surg. 1995;82:74–8.

Temiz A, Oğuzkurt P, Ezer SS, İnce E, Gezer HÖ, Hiçşönmez A. Different clinical presentations, diagnostic difficulties, and management of cecal duplication. J Pediatr Surg. 2013;48(3):550–4.

Thakkar HS, Bradshaw C, Impey L, Lakho K. Post-natal outcomes of antenatally diagnosed intra-abdominal cysts: a 22-year single-institution series. Pediatr Surg Int. 2015;31(2):187–90.

Tireli GA, Ozhey H, Temiz A, Salman T, Celik A. Bronchogenic cysts: a rare congenital cystic malformation of the lung. Surg Today. 2004;34(7):573–6.

Volchok J, Jaffer A, Cooper T, Al-Sabbagh A, Cavalli G. Adenocarcinoma arising in a lingual foregut duplication cyst. Arch Otolaryngol Head Neck Surg. 2007;133(7):717–9.

Zheng J, Jing H. Adenocarcinoma arising from a gastric duplication cyst. Surg Oncol. 2012;21(2):e97–101.