Alimentary Tract Duplications

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18.1 Introduction

Enteric duplications are rare development malformations that can occur anywhere along the alimentary tract from the tongue to the anus. To date, there is no unique classification that can explain their extremely variable onset, size, and location. Calder was the first to publish a report of alimentary tract duplications, back in 1773, describing a duodenal duplication. The term "intestinal duplication" was used for the first time by Fitz in 1884 but was not widely used until it became commonly used in 1937, thanks to Ladd and a further classification by Gross in 1953 [1–3]. Several terms had been used before then to describe such anomalies such as giant diverticula, enterogenous cyst, or unusual Meckel's diverticula. The current nomenclature, as suggested by Ladd and Gross, depends on the anatomic location of the duplication in relation to the normal gastrointestinal tract.

18.2 Epidemiology

Gastrointestinal duplications are observed in 1 every 4500 births, representing 0.1–0.3 % of all congenital malformations, with a slight predominance in white males [1, 4]. Most duplications are diagnosed in children, prenatally or within the first two years of life in approximately two thirds of the cases; less than 30 % of all lesions are detected in adults [1,5]. They are generally single, but in 10–20 % of cases, they are multiple; if one duplication is diagnosed, it is therefore advisable to search for other ones. About 75 % of the lesions are found in the abdomen and 20 % in the thoracic cavity, and in a small percentage, they are located in the thoracoabdominal region. The small bowel, in particular the ileum, is the site where they mostly occur; jejunal and ileal lesions are most common (50–53 %), followed by esophageal and mediastinal (15–21 %), colonic (12–15 %), gastric (6–9 %), duodenal (4–7 %), rectal (4–5 %), thoracoabdominal (2–4 %), and cervical (1 %) lesions (Fig. 18.1 and Table 18.1). Although the embryologic site of origin is uncertain, duplication is commonly referred to as foregut, midgut, or hindgut derived, depending on its location; almost half of all duplications occur in the midgut, while a third is located in the foregut [1,6,7]. Associated anomalies have been observed in about 30–50 % of patients: spinal and vertebral malformation in thoracic or thoracoabdominal duplications, intestinal malrotation, atresia and anorectal malformation in abdominal lesions, and urogenital tract anomalies in midgut and hindgut malformations. Congenital heart diseases and pulmonary airway malformations are rarely associated with foregut duplications [5,6].

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18.3 Embryology

The embryologic site of origin and the pathophysiology of these anomalies are unknown. Several theories have been proposed, but none of them, if considered individually, can explain all the different combinations of type, location, and associated anomalies of duplications.

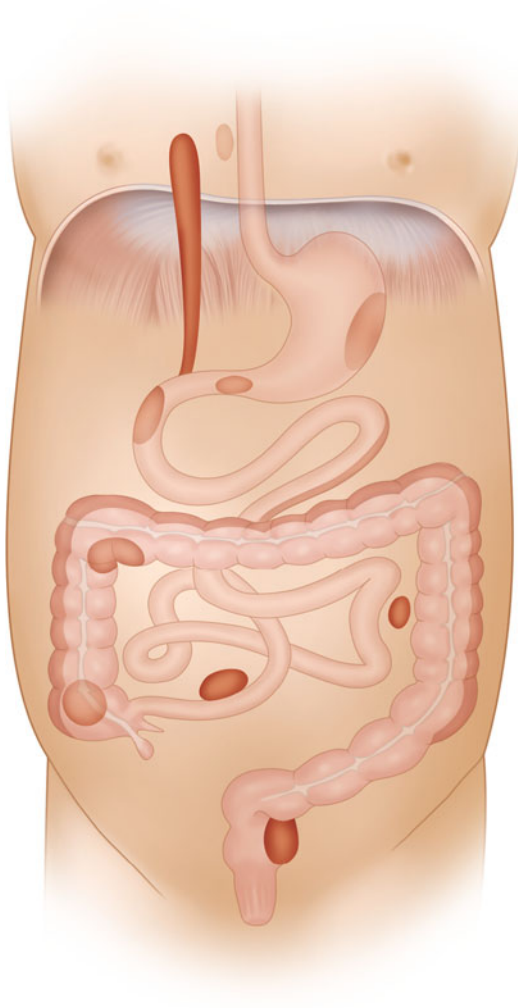

Fig. 18.1 Distribution of duplications by location and frequency

There are four major theories concerning the origin of enteric duplications:

The partial or abortive twinning theory states that alimentary tract duplications are the result of incomplete twinning. This hypothesis can apply in particular for the foregut and hindgut duplications that are associated with doubling of the mouth, genitourinary tract, and lower bowel. The extent of the twinning depends on the moment the process starts [8].

The aberrant luminal recanalization theory, postulated by Bremer in 1944, proposed that duplication is due to the persistence of outpouching of the fetal bowel. This theory may apply to those duplications of the segment of the gastrointestinal tract that goes through a "solid stage," such as the esophagus, small bowel, and colon. After this phase, which occurs between 6 and 8 weeks of intrauterine life, the progressive craniocaudal growth allows the development of a lumen. According to Bremer's theory, duplication may be the result of an incomplete or defective vacuolization of the intestine and/or the persistent embryological diverticula with the formation of two channels, either communicating with each other or not. This theory can apply to simple duplications, i.e., not associated with other anomalies, and provides a functional explication of the highest occurrence of duplications in the ileal tract, which is the main site of diverticula; however, it does not provide proper explanation for the heterotopic mucosa found in some duplications and why the lesions are located more frequently on the mesenteric side while most of diverticula are found instead on the antimesenteric side [9].

Bentley and Smith in 1960 proposed *the split notochord theory* to describe the duplications that develop in the chest and are associated with spinal defects and skin anomalies. During the third to fourth week of gestation, the notochord starts to

Table 18.1 Distribution of duplications by location and frequency

Foregut duplications	Midgut duplications	Hindgut duplications
Esophageal: 15–21 %	Jejunal: 10–18 %	Colonic: 12–15 %
Thoracoabdominal: 2–4 %	Ileal: 35–40 %	Rectal: 4–5 %
Gastric: 6–9 %	Cecal: 3–5 %	
Duodenal: 4–7 %		

close and separate from the endoderm. If an error occurs during the separating phase, an abnormal adhesion between the neural tube ectoderm and the gut endoderm forms, with the development of a gap and a secondary herniation of endodermal cells; furthermore, endodermic tissue can act as a barrier to the anterior fusion of the vertebral mesoderm resulting in vertebral defect. This mechanism may explain the long duplication cysts and foregut duplication, their dorsal location, and the association with spinal malformation (15 % of the cases). It does not explain, however, the entire range of abnormalities (such as heterotopic gastric mucosa) [10].

The environmental factors theory suggests that stress, hypoxia, and trauma can be involved as cause factors of the malformation as described by Mellish and Koop in 1961. Although the actual mechanism that induces the malformation is not clear, an intrauterine vascular accident and/or a compression from nearby organs could explain the anomalies and the association with other malformations as intestinal atresia [11].

Foregut duplications need to be discussed separately. Foregut duplications include a wide spectrum of anomalies, the esophageal, the bron-chogenic, and the neurenteric cysts, subdivided according to their embryologic origin, the anatomopathological characteristics, and the anatomical district concerned. It is believed that bronchogenic and the esophageal duplication cysts result from an altered budding of the embryonic foregut between the fifth and the eighth week of gestation; notochord subdivision alterations can explain the origin of the neurenteric cysts. About 50–60 % of foregut duplications are bronchogenic; they are usually located close to the trachea but can be found in many locations (mediastinum, intraparenchymal, paraesophageal, paratracheal, perihilar) and are frequently associated with congenital pulmonary airway malformations (congenital cystic adenomatoid malformations, pulmonary sequestration), forming hybrid lesions. An enteric cyst may be lined by ciliated respiratory epithelium, but the presence of bronchial wall structures, particularly cartilage, but also smooth muscle and glands, is necessary for diagnosis of bronchogenic cyst; a neurenteric cyst can also be lined by enteric-type mucosa and has a pedicle that extends to the spinal canal [12, 13].

18.4 Anatomical Pathology

As described by Ladd, enteric duplications have three characteristics: (1) an intimate anatomical connection with any part of the gastrointestinal tract, (2) an epithelial lining representing some portion of the alimentary tract, and (3) a well-developed coat of smooth muscle [3]. The lesion tends to locate on the antimesenteric side of the alimentary tract with which it frequently shares the muscular coat and blood supply. The epithelial lining is usually the same as the mucosa native to the lesion, but in 35 % of the cases, an ectopic tissue is present, most commonly gastric followed by pancreatic mucosa, which predisposes to complication as ulceration, hemorrhage, and perforation; rarely, in the thoracic duplication, a respiratory epithelial lining can be present. Generally duplications are classified as two entities, the tubular and the cystic type. Cystic lesions are more common (65–90 %); they are more frequently found in the small intestine and can have big size; and they are closed at their two ends and normally covered with the same mucosa as the native intestine. They do not usually communicate with the intestinal lumen. The tubular type (10–35 %) can often be remarkably long; it may communicate with the adjacent alimentary tract, usually in the caudal end or at both ends, and can contain heterotopic mucosa more frequently than the cystic type [1, 5, 14].

Li et al. have classified small intestinal duplications in two types based on the vascular pattern (Fig. 18.2).

Type 1 lesion or parallel type (74.4 %): the duplication develops in one side of the mesentery, and there are two separated blood vessels, one perfusing duplication and one native bowel.

Type 2 lesion or intramesenteric type (24.6 %): the duplication is located between the two layers of mesentery and vessels from both sides of the mesentery cross the duplication to reach the native bowel.

In their study, Li and colleagues found that vertebral defects were more frequently associated with the type 2 lesion (91.6 %) than the type 1 lesion (5.5 %), hypothesizing a different embry-

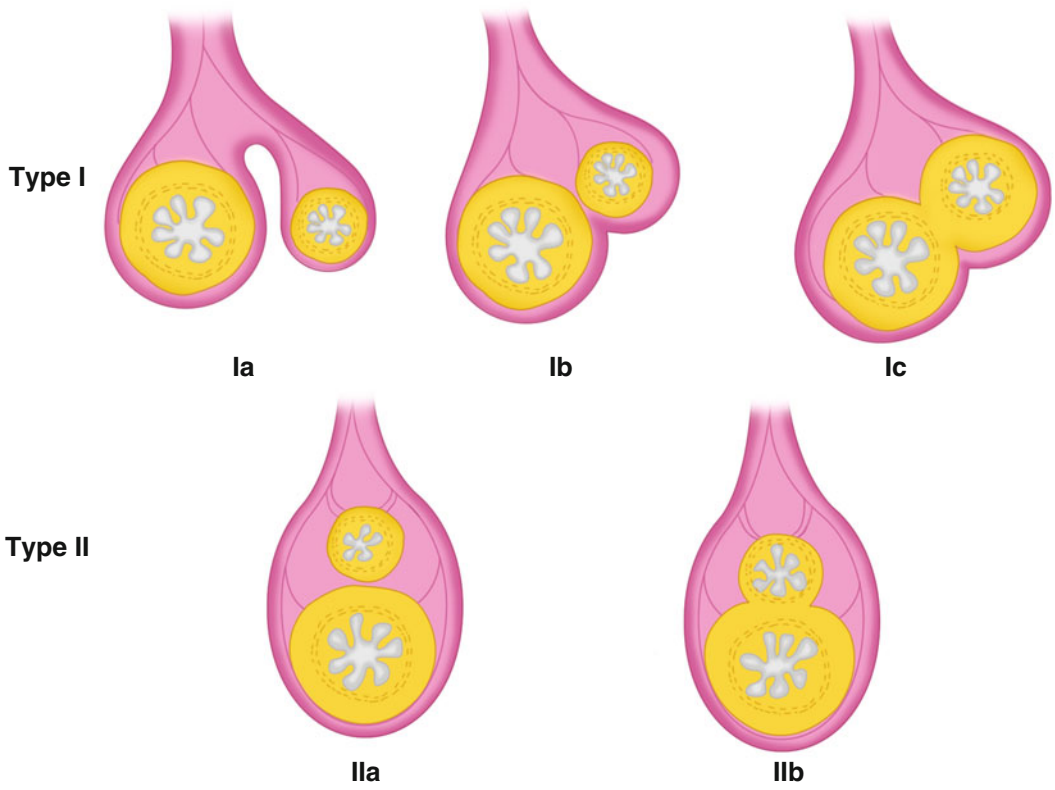

Fig. 18.2 Classification of small intestinal duplications based on their blood supply. In *type 1*, the duplication is located on one side of the mesentery, and the main artery of the duplication is parallel to the main artery of the bowel. *Type 1a*, the duplication has a separate mesentery; *type 1b*, the duplication shares common mesentery with the gut; and *type 1c*, the duplication shares common muscular coat with the gut. In *type 2*, the lesion is located between the two layers of the mesentery and supplied by vessels from both sides of the mesentery that pass over from both surfaces of the duplication to reach the native bowel. *Type 2a*, the duplication is separate from the bowel, and *type 2b*, the duplication shares common muscular coat with the bowel

ologic cause as the origin of the two anomalies. Furthermore, their study shows how the knowledge of the vascular anatomy of small intestinal duplications may have a surgical implication, allowing excision of these lesions without resection of the adjacent bowel. The type 1 duplication could be excised by dividing the mesentery; the type 2 duplication may be enucleated by tying off the short branches from the main vessels [15].

18.5 Clinical Manifestations

Clinical manifestations are extremely variable, depending on the site, type, and size of the duplication and if it contains gastric mucosa. Most duplications (80%) are detected in the first two years of age, in particular in half of the cases in the first six months of age. They are frequently asymptomatic and diagnosed incidentally on routine X-ray or ultrasound examination performed for other malformations or rarely may be an incidental intraoperative finding. In recent years the diagnosis has been increasingly made with prenatal ultrasonography.

When located in the mediastinum, they could lead to pneumonia, wheezing, cough, or dysphagia. Symptoms such as respiratory distress and failure to thrive are more common in small infants, whereas chest pain occurs more frequently in older children.

The most common presentations of an abdominal duplication are abdominal pain and

distension, vomiting, and abdominal mass. Complications include bleeding and perforation due to an ectopic gastric mucosa, with peptic ulceration or intestinal occlusion due to the development of volvulus, intussusception, or extrinsic compression frequently for an acutely enlarged cystic mass. Other rare complications are pancreatitis and cholecystitis in the gallbladder, duodenal and pancreatic duplication, cyst infection, and malignancy, the latter more frequent in adulthood and in hindgut duplications. Volvulus and intussusceptions are more frequent in midgut duplications, whereas in hindgut duplications symptoms by mass effect with obstruction of both the urinary tract and the bowel tract prevail. Bleeding, hemorrhage with melena, and perforation are frequent both in midgut and hindgut duplications [4, 14, 16].

18.6 Diagnosis

The high resolution modern imaging techniques allow physicians to identify enteric duplications in the prenatal age, in particular those located in the chest and in the upper abdomen, in approximately 30 % of cases. Since duplications are often associated with other malformations, if an enteric duplication is found during a prenatal ultrasound, a fetal magnetic resonance imaging (MRI) is advisable. The fetal MRI also allows physicians to identify fetuses at risk who might require invasive procedures (i.e., thoraco-amniotic shunt for fetal hydrops or mediastinal shift) and to establish the best therapeutic strategy in the postpartum so as to avoid complications [14, 16].

The clinical history and the physical examination are the first step for the diagnosis of a duplication, whereas the laboratory exams may detect only anemia in case of bleeding due to heterotopic mucosa or higher serum amylase and lipase levels in the rare case of pancreatitis.

Postnatally ultrasound can be useful to demonstrate the nature (solid or cystic) and the location of the mass as well as to evaluate any connection with the adjacent intestine. The cystic lesion appears as an anechoic structure (in case of no bleeding) surrounded by a 2–3 mm thick wall that determines a characteristic and pathognomonic echogenic signal defined as "gut signature" or "double layer," which is made up of an hyperechoic inner mucus layer and a hypoechoic muscular outer layer.

An abdominal X-ray can show a mass effect in the event of a large cyst or signs of intestinal obstruction or perforation in complicated cases. A thorax X-ray can show a mass usually located in the medium or posterior mediastinum and eventually associated vertebral anomalies.

Computerized axial tomography scan (CT scan) and MRI are more accurate in showing the anatomical features and the relationship with nearby organs; MRI is advisable in the childhood and useful to detect any spinal involvement.

Gastrointestinal contrast studies can demonstrate a filling defect as the duplication does not usually communicate with the intestinal lumen. The endoscopic ultrasonography, the esophagogastroduodenoscopy, and the wireless capsule endoscopy can show ulcers or stenosis and can help defining better the anatomy before a surgery. The endoscopic ultrasound (EUS) can provide further information, detecting the cystic nature of the lesion with its characteristic wall, its location, and its anatomical relationships with adjacent structures; it can also be useful as a guide for fine needle aspiration, which is used only in selected cases.

In the case of a patient presenting anemia and lower gastrointestinal bleeding, a scintigraphy with technetium-99m pertechnetate can be useful to identify ectopic gastric mucosa and make a differential diagnosis with the Meckel's diverticulum.

Definitive diagnosis is based on histopathological findings after surgical excision.

Differential diagnosis includes intrathoracic mass or tracheoesophageal fistula in the foregut duplications, appendicitis, Meckel's diverticulum and other causes of intussusception in the small bowel duplications, and constipation or Hirschsprung's disease in colonic and hindgut duplication. In the rare case of a gastric or duodenal duplication, the sign and symptoms can mimic a hypertrophic pyloric stenosis or a gastroesophageal reflux [1, 4, 8, 14].

18.7 Management

Management depends on the presentation. Intestinal duplications often require an urgent surgical intervention due to the onset of complications such as perforation, intestinal occlusion, or severe bleeding. This is why, although the treatment of asymptomatic lesions remains controversial, most authors currently recommend surgery even with no symptoms in order to avoid any complication at a later moment [16, 17]. The type of surgery varies depending on the cases [6, 7]:

- Enucleation of the intestinal duplication only where possible.
- Resection of the duplication along with the adjacent intestinal segment and end-to-end anastomosis in the event of small cystic or short tubular duplications.
- Total excision of the duplication through extramucosal dissection according to Wrenn procedure, which includes stripping of the entire mucosa through a series of multiple incisions; this technique applies to extended tubular duplications or large cystic ones where an intestinal resection would cause a short bowel syndrome [18, 19].
- Drainage procedure, like fenestration or marsupialization of the duplication into the lumen after total or partial resection or puncture of the lesion, in the cases where removal is difficult, i.e., in the duodenal duplication, mediastinal duplication, or duplication near biliary or pancreatic tracts; this intervention has a high risk of recurrence.

18.8 Classification and Treatment by Location

18.8.1 Oropharyngeal Duplications

Oropharyngeal duplications are very rare, being 1% of all duplications. The most common location is the floor of the mouth and they may contain gastric or colonic mucosa. In most cases they are asymptomatic; sometimes, however, failure to thrive may be the presenting symptom. The treatment consists in an oral approach with cyst resection and oral mucosa reposition [1, 14].

18.8.2 Cervical Duplications

Esophageal duplications starting from the cervical area are extremely rare, are usually cystic, and are strictly adherent to the esophagus. Symptoms include a palpable mass and/or symptoms due to compression of the adjacent structures; this is why the diagnosis is usually made during the first months of life. The CT scan is the gold standard for diagnosis to evaluate any anatomical connections. The differential diagnosis includes lymphatic malformations, cysts of the airway, branchial apparatus cysts, thyroglossal cysts, cervical lymphadenopathies, and esophageal diverticula. Treatment consists in complete excision, if possible, or partial excision with mucosa removal through a supraclavicular approach [1, 14].

18.8.3 Thoracic Duplications

Thoracic duplications are the second most common site of duplications, accounting for approximately 15–21% of alimentary tract duplications; the foregut duplication is the second cause of mediastinal mass after neurogenic tumors. Although esophageal duplications may develop throughout the length of the esophagus, two thirds of these lesions are found in its lower third and one third in the upper/middle third of the esophagus. They are typically located in posterior mediastinum, more commonly in the right side. They are usually cystic and do not share a muscular wall or communicate with the esophageal lumen. In 80% of the cases, they are diagnosed in childhood and are more frequent in male patients. The clinical presentation includes symptoms secondary to extrinsic tracheal and esophageal compression up to respiratory distress; the upper esophageal duplication can cause stridor, tirage, and cough, while duplication in the middle or lower esophagus can cause dysphagia, epigastralgia, chest pain, or vomiting. Furthermore, gastric mucosa is present in about 30–50% of all esophageal duplications,

with possible bleeding and secondary hematemesis, melena, or hemoptysis as a manifestation of fistula formation with bronchial tree. Other rare manifestations include cardiac arrhythmia, retrosternal and thoracic back pain, and cyst rupture with mediastinitis. Additionally, they are very frequently associated with vertebral anomalies (20–50%) and thus defined neurenteric cyst, including hemivertebra or spinal dysraphism, with possible neurological problems (in 20% of cases, a communication with the spinal cord is described); other anomalies found in a lower percentage of cases include diaphragmatic hernia and esophageal atresia. A clinical suspicion, if not raised prenatally, may arise with a thorax X-ray, a contrast study of the upper gastrointestinal tract, and eventually an endoscopy. Endoscopically they cannot be distinguished from a lipoma, a leiomyoma, a gastrointestinal stromal tumor, or a submucosal lesion. With EUS the duplication appears as an anechoic cyst with the characteristic multilayered wall structure; the EUS-guided fine needle aspiration is possible but usually reserved for lesions of indeterminate appearance, atypical or suspected for malignancy, also due to the high risk of post-aspiration infections. The best imaging technique to define anatomic details is CT scan or MRI; the latter is the gold standard for defining neurenteric cysts. An abdominal ultrasound is required to exclude any associated intestinal duplications which are found up to a third of the cases [4, 14].

The treatment of choice consists of the exeresis or enucleation, if possible, through a thoracotomy or a thoracoscopic procedure; other authors suggested the surveillance with EUS of some asymptomatic lesions.

The approach used in the intrathoracic cysts is a posterolateral thoracotomy with the patient positioned on the opposite site with respect to the lesion location. The muscle-sparing technique ensures a gentle separation and a rapid cicatrization. Resection of the cyst must be close to the esophagus and must include the removal of the residual mucosa. If the cyst is in the contest of the esophagus wall and does not communicate with the esophageal lumen, the duplication must be removed by opening the wall of the esophagus and performing an extramucosal excision, leaving the mucosa intact. Any communication with the esophageal lumen must be closed, repairing the muscular defect also with an edge of duplication and checking the mucosal integrity by air or fluid insufflation through a nasogastric tube. In some cases it might be useful to perform the surgery under the guide of a flexible endoscope.

Video-assisted thoracoscopic resection is possible in many cases (Fig. 18.3). The patient is placed in a lateral position on the healthy flank. The homolateral arm is raised above the head, the hemithorax is elevated by axillary roll, and the body is rotated into a near-prone position to expose the posterior part of the thorax. The optic (5–10 mm/0–30°) is inserted with an open technique in the V intercostal space, along the middle axillary line, inferior to the tip of the scapula. Two or more 3–5 mm instrumental ports are introduced to create a triangulation of the instruments. Single-lung ventilation with collapse of the ipsilateral lung can help the surgical removal of the cyst. The mass is exposed with blunt dissection and cauterization; after dissection, the mass is aspirated for decompression, if required, and extracted through a trocar site. If the cyst cannot be removed completely due to the risk of damaging the esophagus or the airways, the borders of the cyst can be left and ablated using electrocauterization. At the end of the procedure, it is important to check and close any leak in the esophageal wall which might pose the risk of an esophageal perforation or a pseudodiverticulum formation. Usually an apical chest drainage is left.

In the case of neurenteric cysts, a surgery with the aid of neurosurgeons is required; resection of the bone and laminectomy, if required, are usually the first step, with subsequent exeresis of the thoracic mass. Mortality in these cases may be high [12, 13].

18.8.4 Thoracoabdominal Duplications

Thoracoabdominal duplications are 2–4% of all duplications. They are generally the tubular type, can communicate with the intestinal lumen, and frequently have ectopic gastric mucosa. They go down to the right of the esophagus, go over the

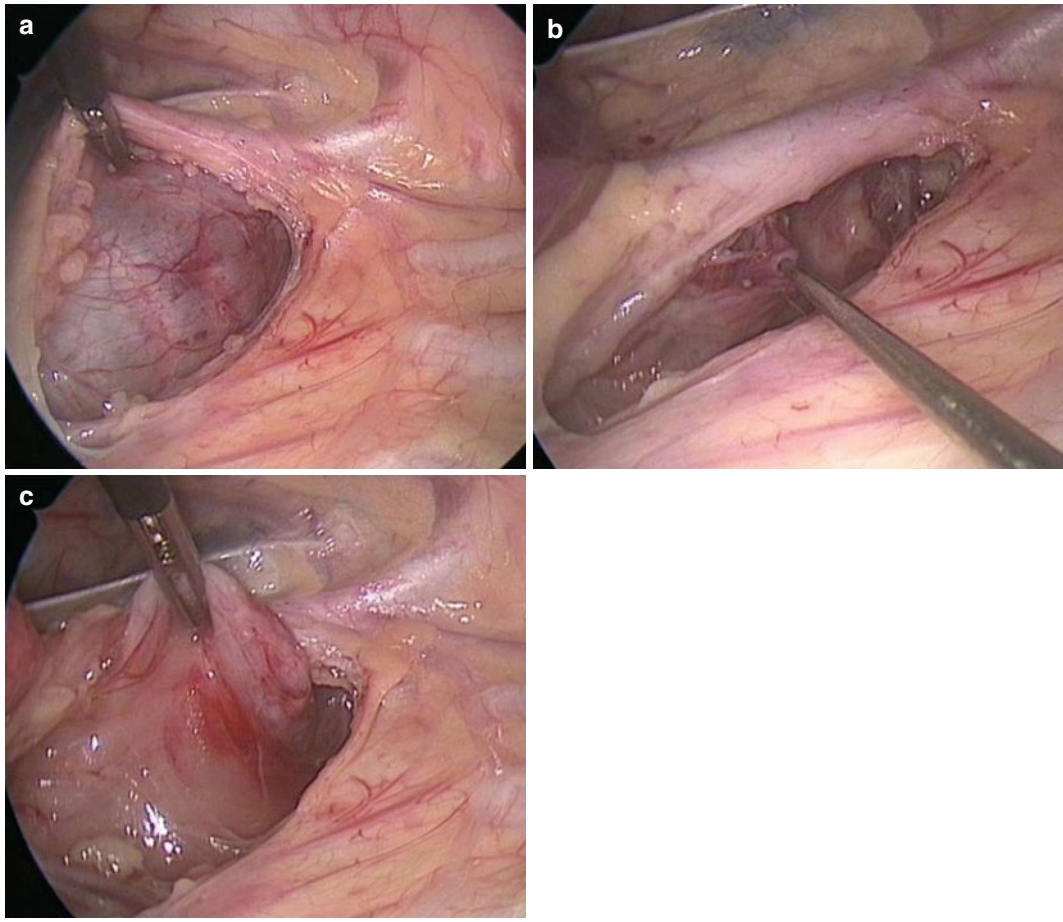

Fig. 18.3 Thoracoscopic excision of an esophageal duplication. (**a**) Thoracoscopic identification of the duplication. (**b**) The lesion is aspirated to facilitate the removal. (**c**) After blunt dissection, the mass is extracted through a trocar site by an endobag or a finger glove

diaphragm near the right pillar or through the esophageal or aortic orifice, and extend along the greater curvature of the stomach and the mesenteric side of the duodenum and jejunum. Clinical picture is similar to that of the thoracic duplications, with the association of abdominal symptoms such as recurrent abdominal pain or palpable mass; neurological symptoms may also be present as they are frequently associated with rachis malformations in the thoracic area. The diagnostic investigations required are the same as those for the thoracic duplications: thoracic and abdominal CT scan and MRI are required before performing any surgery. The treatment consists in one-stage combined thoracoabdominal approach; occasionally a laminectomy is also necessary in the case of intraspinal lesions. After removing the rachis component, the duplication is separated from the esophagus and is removed up to the diaphragmatic defect and then sutured; alternatively, the thoracic portion is pulled into the abdomen through the diaphragm and then removed. The duplication is finally removed through an abdominal approach, by means of an excision or following Wrenn's surgical principles [1, 5].

18.8.5 Gastric Duplications

Gastric duplications account for 5–9% of all duplications. Unlike other duplications, they appear to be more frequent in females. They are

more often cystic and noncommunicating lesion and are located along the greater curvature, although they can be found in any part of the stomach. The lining mucosa is usually the gastric type, although intestinal or colonic mucosa may be found; gastric duplications may also contain pancreatic mucosa or ciliated cells and can rarely communicate with the pancreatic duct system. These patients usually become symptomatic in the first months of life. The symptoms are usually related to the mass effect and include vomiting, abdominal pain, epigastric pain, weight loss and failure to thrive, and sometimes pancreatitis; if a communication with the lumen is present, a peptic ulceration can result in a hemorrhage with hematemesis and melena or perforation.

Diagnostic evaluation includes an abdominal ultrasound in order to differentiate a gastric duplication from a hypertrophic pyloric stenosis, of which it can mimic the symptoms, and from pancreatic pseudocysts and choledochal cysts which are included in the differential diagnosis of these duplications as well. A barium swallow, an endoscopy, or an EUS can provide further information, but an abdominal CT scan is usually necessary to define better the anatomy and identify any synchronous lesions [15, 16].

The treatment of the gastric duplication should be complete resection. Most duplications can be easily removed by an extramucosal dissection, suturing the muscular defect, and checking, at the end of the procedure, the integrity of the gastric wall by insufflating air through a nasogastric tube; the excision can be performed via laparoscopy (Fig. 18.4). In other cases, the duplication is removed via a partial gastrectomy (wedge resection)

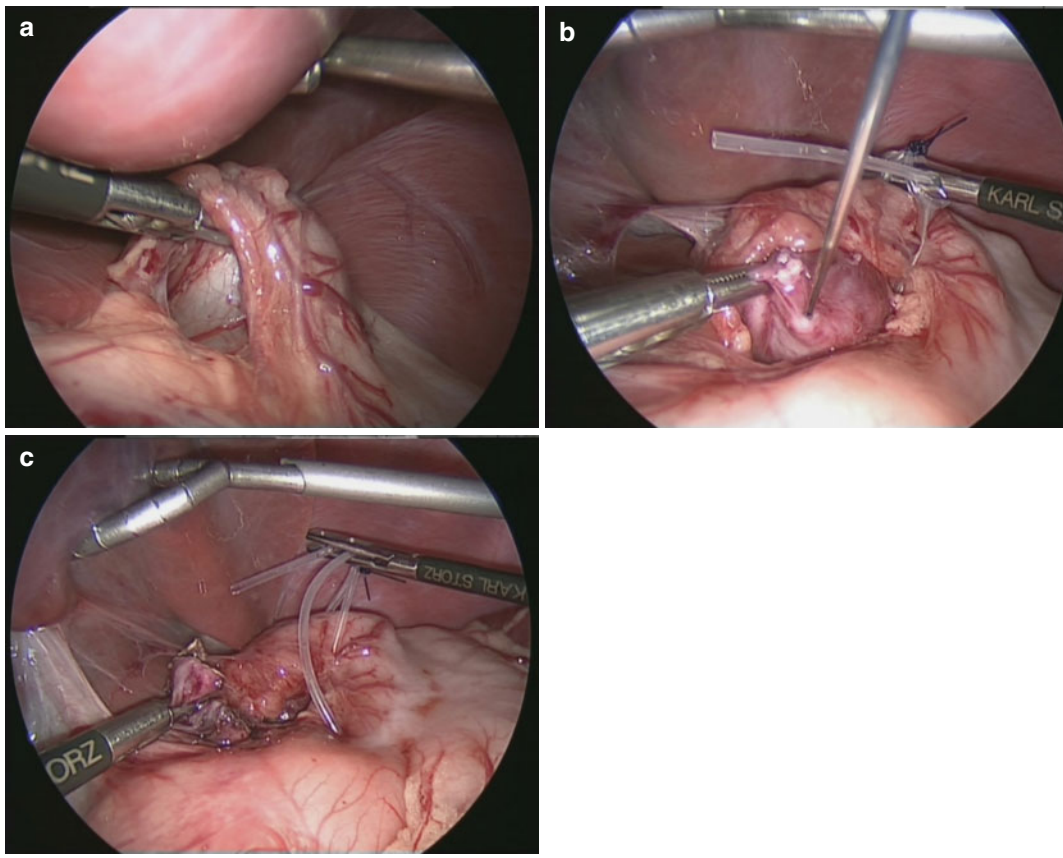

Fig. 18.4 Laparoscopic removal of a gastric duplication. (**a**) Laparoscopic identification of the gastric duplication. (**b**) The cystic lesion is emptied before the enucleation. (**c**) The lesion is removed by an endobag. Gastric duplications, as well as duodenal and colonic duplications, need a purely laparoscopic approach

or, in case of a long duplication, by partial resection and stripping of the residual mucosa. An alternative procedure is to divide the septum between the gastric duplication and the gastric lumen with a linear stapler, with a high risk of complication if the ectopic mucosa is left in situ [6, 20, 21].

18.8.6 Duodenal Duplications

The duodenum is a rare location for duplications and accounts for 4–7 % of all duplications. They are frequently the cystic type, may occasionally communicate with the duodenal lumen, and are covered by duodenal mucosa in most cases or more rarely by pancreatic, gastric, or intestinal mucosa. They are located in the medial or posterior portions of the first or the second part of the duodenum with a possible communication with the pancreatic or common bile duct. The clinical picture is usually not specific, with abdominal pain or distension or failure to thrive, up to an intestinal obstruction with a palpable mass; pancreatitis, jaundice, and hemorrhage due to peptic ulceration may be the onset. An abdominal ultrasound can show a cystic mass, the position of which, in association with the clinical picture, often creates difficulties with the differential diagnosis with a pancreatic cyst and pseudocyst and choledochal cysts; an upper gastrointestinal contrast study can show an extrinsic compression of the duodenum with alterations of the C-shaped duodenal convexity. A CT scan, an endoscopic retrograde cholangiopancreatography (ERCP), and an MRI cholangiography are useful to evaluate the connections with the adjacent structure, in particular with the pancreas and biliary tree. Due to their location and close anatomical relations with the duodenum, biliary tree, and pancreas, the complete exeresis with division of any ductal communication can be very difficult; if an exeresis cannot be performed, partial excision with mucosectomy or endoscopic or percutaneous drainage is possible, although internal drainage is preferable. Marsupializations of the cyst to the duodenum or to a Roux-en-Y loop of the intestine are possible techniques; rarely a pancreaticoduodenectomy is required (Whipple procedure). In any case it is advisable to perform an intraoperative cholangiography to evaluate any connection with the biliary tree; in the case of gastric mucosa diagnosed through an intraoperative biopsy, it is highly recommended to remove it completely even with mucosal stripping only [7, 21].

18.8.7 Gallbladder and Pancreatic Duplications

These types of duplications are very rare.

Pancreatic duplications are more frequently located in the head of the pancreas (51 %), whereas the remaining ones are placed with the same frequency in the body or the tail. Symptoms and radiologic examinations are the same as with duodenal duplications. Treatments include duplication excision, cystojejunostomy, pancreaticoduodenectomy, or partial pancreatectomy; the differential diagnosis between pancreatic duplications and pancreatic pseudocysts can be made with a histological examination [8, 14].

Gallbladder duplications look like double gallbladders and duplicated cystic ducts. According to Boyden's classification, gallbladders may share a common cystic duct (vesica fellea divisa or bilobed gallbladder) or more frequently may have two cystic ducts (vesica fellea duplex or true duplication); the true duplication is subclassified into "H-shaped type," in which two separate gallbladders and cystic ducts enter separately into the common bile duct, and "Y-shaped type," where the two cystic ducts unite before entering into the common bile duct [22]. The clinical presentation can be like an acute cholecystitis or may include bleeding due to the presence of heterotopic gastric mucosa. The diagnosis of gallbladder duplication is often made intraoperatively. ERCP and intraoperative cholangiography are the most accurate tests in displaying the biliary tract anatomy of gallbladder duplications and have been recommended to define the biliary tract anatomy clearly before surgical intervention. Simultaneous removal of both gallbladders by both open and laparoscopic techniques is recommended to avoid cholecystitis and symptomatic gallstones in the remaining organ [6, 7].

18.8.8 Small Bowel Duplications

They are the most common intestinal duplications (50–53 % of all duplications). The most common location is the ileum. They may be the noncommunicating cystic type or the tubular type, frequently communicating, and are located on the mesenteric side. A third of the duplications is diagnosed in the neonatal period, and in about 70 % of the cases, the onset is within the first two years of life. The clinical picture can include a palpable mass, recurrent abdominal pain, or symptoms secondary to a complication such as intussusception, volvulus, small bowel obstruction, peptic ulcer, and perforation due to the presence of ectopic gastric mucosa. If not diagnosed prenatally, radiological investigations to be performed postnatally include an abdominal ultrasound, which can show the typical cystic aspect of duplications with "double layer" and can be sufficient to diagnose a duplication; the technetium-99m pertechnetate scintigraphy in patients who have bleeding, which ensures a differential diagnosis with Meckel's diverticulum; and CT scan and MRI in case of diagnostic doubts. The differential diagnosis includes mesenteric or omental cyst, ovarian cyst, and Meckel's diverticulum, which is conversely located on the antimesenteric side of the bowel [5, 6, 8, 14].

The diagnosis is frequently intraoperative. Recently, laparoscopy has been suggested as a diagnostic and therapeutic method as it reduces hospitalization and post-surgery pain; in small intestinal duplications, a video-assisted procedure may be performed because the bowel is easily extracted from the umbilicus (Fig. 18.5). In laparoscopic procedure, the patient is in a supine lithotomic position. An umbilical trocar for the optic is inserted; a 5 mm is usually used for small bowel and cecal duplications, while a 10 mm Hasson-type trocar is recommended for video-assisted procedure. Two or more 3–5 mm instrumental ports are introduced to create a triangulation of the instruments; in the video-assisted procedure, only one instrumental trocar is necessary, usually positioned in the left flank. Pneumoperitoneum is created (8–10 mmHg of pressure, 0.5–1 lt/min of flow). The procedure is performed in a laparoscopic way in the gastric, duodenal, and colonic duplications, while in the small intestinal duplication, a video-assisted procedure may be used. Small intestinal cystic duplications can be enucleated without resection of the adjacent bowel, according to the principles of Li and colleagues (see above). If it is not possible, a resection of both duplications and native bowel after ligation and division of associated mesenteric vessels with primary end-to-end anastomosis can be performed [16, 17, 20, 21].

Very long tubular duplications represent a greater surgical challenge for the high risk of short bowel syndrome secondary to massive resection. The extramucosal dissection envisages the stripping of the mucosal lining through a series of longitudinal seromuscular incisions and the resection of the two ends of the duplication, which usually communicate and are possible sites of bleeding ulcers due to the presence of ectopic gastric mucosa [18, 19]; this operation may be performed by video-assisted procedure (Fig. 18.6) [21]. Alternatively, a marsupialization with creation of a large window proximally and distally between the duplication and the adjacent intestinal lumen may be performed to allow the drainage of the duplication, however, with the risk of retained gastric mucosa. During surgery it is important to check the whole bowel in order to exclude the presence of multiple lesions and any association in the newborn period with intestinal atresia or malrotation.

18.8.9 Hindgut Duplications

Colonic and rectal duplications account for about 16–20 % of all duplications. The hindgut duplication may be classified in three types. The first type is the cystic or short tubular duplication, frequently located in the mesenteric side, with clinical and imaging features similar to the small bowel duplications. The second type includes the mass located in the midline, in front of the sacrum or coccyx, and behind the rectum; they are usually cystic masses that frequently share the rectum vascularization and can be difficult to remove. The last type is the side-to-side rectal and

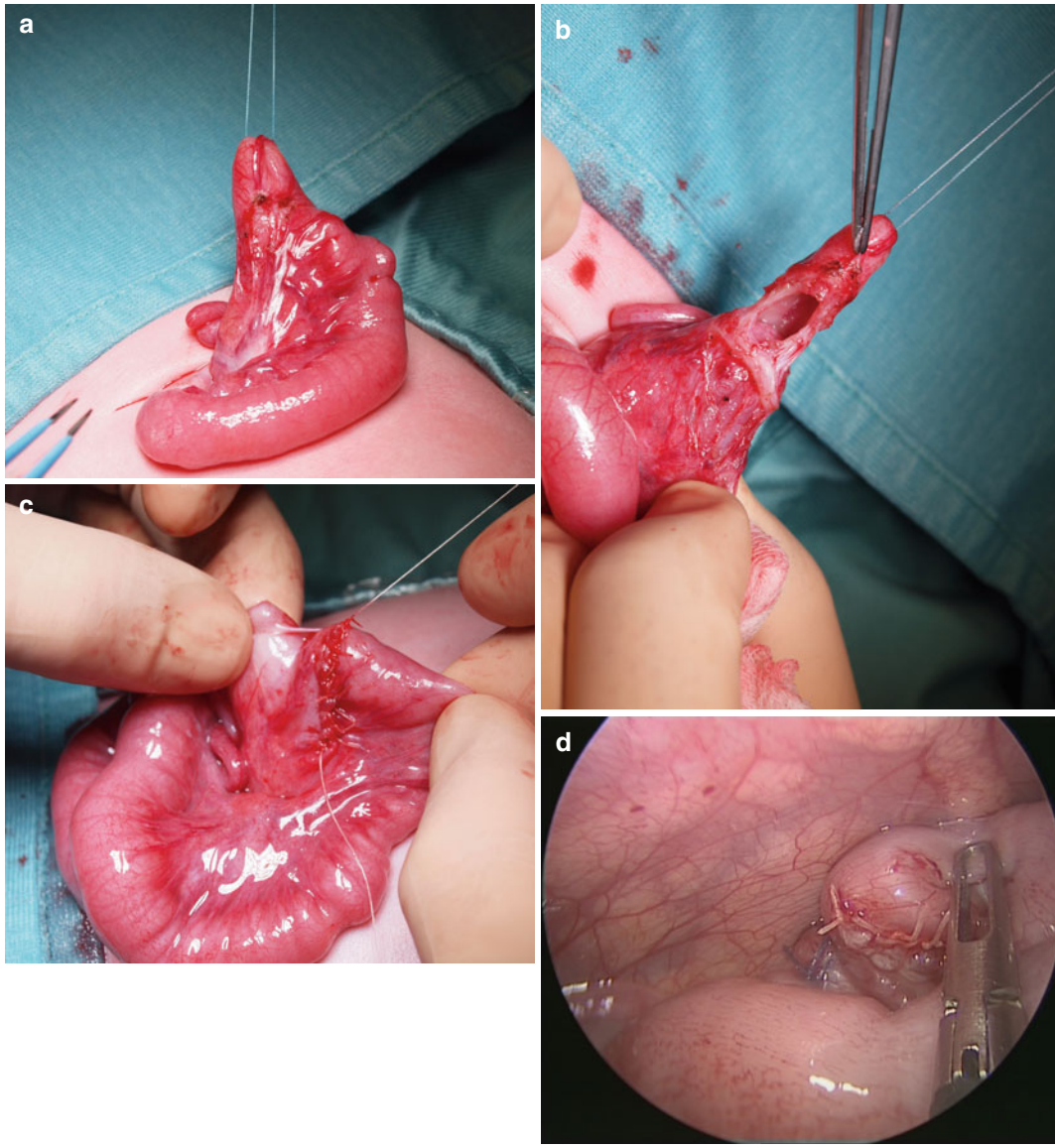

Fig. 18.5 Video-assisted procedure for small intestinal duplications. (**a**) After laparoscopic identification of the ileal duplication, the ileum is exteriorized through the umbilical wound. (**b**) The small intestinal cystic duplication is enucleated in an open way without a need of resection of the adjacent bowel. (**c**) The residual muscular defect is repaired. (**d**) At the end of the procedure, a laparoscopic control is performed to check the suture or anastomosis and to exclude bleeding

colonic duplication; they are tubular, located on the mesenteric or antimesenteric side, and usually communicate with the intestinal lumen; they can develop throughout the entire colon and open up in the perineum. The last type is twice more frequent in females and may be associated with rectogenital or rectourinary fistula, duplication of internal or external genitalia, or vertebral anomalies (abortive twinning anomalies). The clinical picture varies and includes abdominal pain, constipation, and obstruction symptoms; in the case of urogenital fistula, gas or stool can pass through the vagina or with urine; urinary obstruction or retention and bleeding are very rare symptoms. Rectal duplications are typically the cystic type and located in the retrorectal space. They can

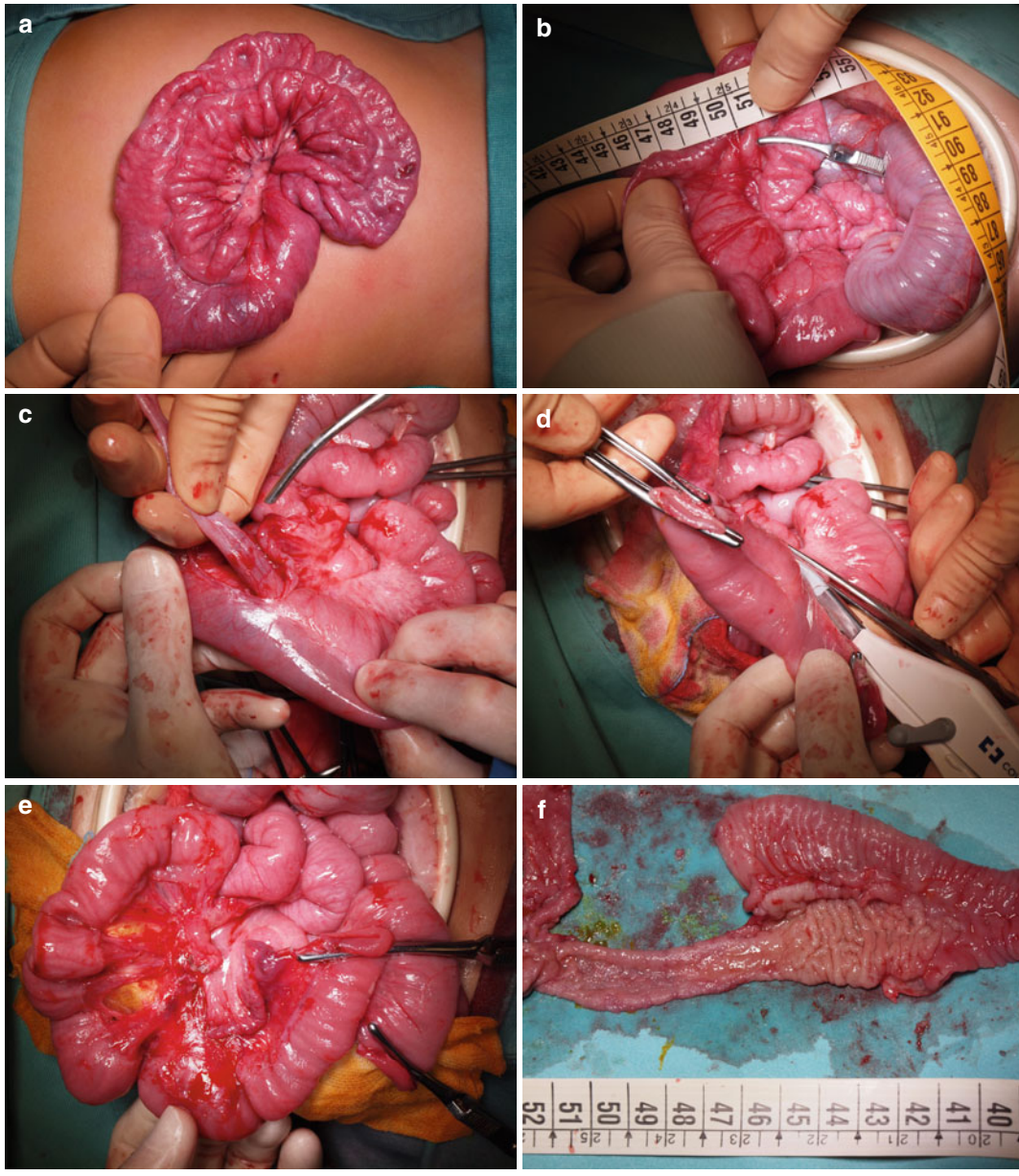

Fig. 18.6 Surgical approach for long tubular duplications. (**a**) After laparoscopic identification of the long tubular duplication, the ileum is extracted through the umbilical wound, using an Alexis retractor. (**b**) The resection of the entire tubular duplication can lead to short bowel syndrome. (**c**) A mucosal stripping is performed. (**d**) The junctions of duplicated and normal bowel are resected, due to the frequent presence of heterotopic gastric mucosa in these sites. (**e**) The ileum aspect at the end of the procedure. (**f**) The duplication aspect after resection (mucosa lining and distal communication segment)

dislocate or compress the rectum, the bladder, or the ureters, thus causing stypsis or fecal incontinence, hydronephrosis, or perianal fistula in the case of communication with the rectal lumen; prolapse is very rare. Other congenital anomalies are rarely associated; the differential diagnosis must be made with ovarian cyst, rhabdomyosarcoma, or sacrococcygeal teratoma. Pelvic ultrasound, barium enema, fistulography, and CT scan or MRI are the investigations that can be performed in

order to diagnose hindgut duplications; a voiding cystourethrogram must be performed in the forms with genitourinary fistula [1, 7, 8, 14].

Treatment of colon duplications varies according to their type, extension, and form. Cystic duplications can be removed by enucleation or resection. For tubular duplications the approach consists of resection, if possible, or marsupialization or fenestration, creating a large communication between the duplication and the colon, both proximally and distally, with dissection of the distal part of the duplication, if there is an opening into the perineum or in the urogenital system.

Treatment of rectal duplications varies from exeresis via a sagittal posterior approach or endorectally to marsupialization via a transanal approach up to separation of the septum between the duplication and rectum.

As colon and rectal duplications rarely contain ectopic mucosa, stripping of the mucosa is not usually performed. The incidence of neoplastic changes (mainly adenocarcinoma) in hindgut duplications is anyway higher than in other locations [1, 8, 21].

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