

# Review. Gastrointestinal Surgery in Neonates: Practice, Facts, and Trends

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## Abstract

The author presents an overview of the current trends in acute neonatal gastrointestinal surgery. Necrotizing enterocolitis and focal intestinal perforation are disorders of different etiology, appearance, and prognosis. In neonates with focal perforation, a good prognosis can be expected. Primary anastomosis is a valid option for primary surgery. Meconium ileus is based on obstruction of the terminal ileum. Medical treatment is the primary therapeutic option, although surgery may be required. In contrast, meconium plug syndrome is based on stool plugging in the left colon, and surgery is not appropriate. Hirschsprung's disease is confirmed on the basis of representative biopsy of the rectum just above the dentate line. There is a trend toward a single-step pull-through operation without protective stoma. Open hernia repair in small neonates can be performed via a mini-incision and without opening the external inguinal ring. Laparoscopic hernia repair has some advantages, especially in girls. In infants with pyloric stenosis, laparoscopic pyloromyotomy is a first line option for treatment. There is a general trend toward single-stage procedures and laparoscopic reconstructive surgery for gastrointestinal malformations. With the exception of tracheoesophageal fistula and low bowel obstruction, the vast majority of upper gastrointestinal malformations can be identified using prenatal ultrasound examination. One-stage repair with excellent results is even possible in neonates below 1000 g. Surgery of duodenal or small bowel atresia can be performed following the postnatal adaptation period. For some forms of anorectal malformation, the need for a protective stoma has been questioned in the last decade. Posterior sagittal anorectoplasty remains the standard procedure for the majority of cases. Midgut or segmental volvulus affects different groups of infants. If a neonate is suspected of having any type of volvulus, urgent surgery is required. Intestinal duplication carries a high risk for complications, and surgery should be performed on an elective basis.

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## Keywords

Neonatal Surgery, Acute Abdomen, Gastrointestinal Atresia, Necrotizing Enterocolitis, Bowel Obstruction

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## 1. Introduction

Congenital and acquired diseases of the gastrointestinal tract are predominantly rare diseases, with prevalences between 1:300 and 1:10,000 or less. The overview presented is based on the literature and experience. In this respect, meta-analyses and international surveys provide important information. Prospective randomized multi-institutional studies promise the highest-level evidence but are not feasible to address many of these questions for ethical reasons. Summary presentations in recent years have shown substantial progress in pediatric surgery, and they form the basis of this review [1] [2] [3] [4]. The present study is predominantly in line with evidence levels 3 and 4 and, in the case of empirical values or expert opinions, with level 5.

### I.1. Acute Abdomen

Acute abdomen in newborn babies is always a life-threatening condition. Generally, bilious vomiting is a red-flag that requires urgent imaging. A nasogastric tube for decompression is inserted. It does not, however, represent a form of aspiration protection [5].

Mature newborns first pass meconium within 24 hours of birth. No meconium is passed 24 to 48 hours after the first breast milk meal is an indication of an intestinal transit disorder.

Preterm neonates, and particularly children with developmental delays (small for gestational age), are at high risk of gastrointestinal complications if meconium retention presents. Timely detection of delayed passing of the meconium opens up the gate of early treatment. Opioids can represent an iatrogenic cause for the delay in passing meconium [6].

Important signs of a disruption to the passage of meconium are bilious stomach contents, increase in abdominal circumference, guarding and pain in the abdominal wall, intestinal rigidity and vascular proliferation and erythema of the abdominal wall (**Figure 1**). In cases where therapeutic rectal enemas are administered in newborns, phosphate-containing enemas must be strictly avoided, as life-threatening hyperphosphatemia, hypocalcemia and metabolic acidosis may occur [7].

Diagnostic imaging of acute abdomen primarily involves ultrasound examination in addition to conventional X-ray examinations, with or without administration of a water-soluble contrast medium [8]. On radiographic examination, air filling of the entire small intestine can be seen 3 hours after birth; the same applies for the sigmoid colon after 8 to 9 hours. As such, a latency period of a



**Figure 1.** Protrusion and redness of the anterior abdominal wall in a neonate with meconium peritonitis due to intrauterine intestinal perforation. Female neonate at the second day of life.

few hours is allowed to pass prior to X-ray assessment of the intestinal air distribution after birth.

In acute abdomen or ileus, there is an intravascular fluid deficit caused by vomiting, diarrhea and third-spacing. If hypovolemia is pronounced, bolus delivery of an isotonic glucose-free infusion solution (10 to 20 ml/kg body weight) is effective. The role of albumin in the case of hypovolemic shock is under discussion. Cases of perforation or ischemia-related pain are particularly serious and should be managed using opioids.

In cases of systemic inflammatory response syndrome (SIRS), suspected migration or perforation of the intestinal wall, calculated antibiotic therapy (gram-negative, gram-positive and anaerobic spectrum) is necessary.

Bowel resections should be performed as sparingly as possible. The aim is to preserve enteral autonomy, *i.e.*, to preserve the capacity of an oral diet without additional parenteral fluid or electrolyte and nutrient substitution. The length of the remaining small intestine is measured intraoperatively, starting at the flexuraduodenojejunalis (Treitz's band at the duodenojejunal flexure). If possible, the ileocecal valve (Bauhin's ileocecal valve) should be preserved.

From a functional point of view, short bowel results if the preserved intestine is not capable of absorbing sufficient nutrients, water and electrolytes. Highly critical intestinal lengths are below 40 cm in the residual small intestine. Following an acute event, those affected initially require longer-term parenteral nutrition, but a significant proportion of infants can be weaned from the parenteral diet. Functional adaptation (enteral adaptation) of the remaining intestine takes up to 4 years [9].

Causes of short bowel include, in particular, volvulus, small-intestinal atresia, necrotizing enterocolitis, and chronic intestinal pseudoobstruction. Special problems arise from the pathogenic microbiological colonization (microbiome)

of the remaining intestine. Limiting factors for body growth and development, and with reference to patient survival, include short-intestine-associated chronic liver disease as well as enterogenic and catheter-associated sepsis.

## 2. Acquired, Multifactorial, and Neuromuscular Diseases of the Gastrointestinal Tract

### 2.1. Necrotizing Enterocolitis and Focal (Spontaneous) Intestinal Perforation

With increasing control of lung maturity in newborns (surfactant supplementation), focal intestinal perforation (FIP) and necrotizing enterocolitis (NEC) are the most common causes of death in preterm neonates with a birth weight < 1500 g. Lethality rates are between 15% and 30% [10]. In a Swedish study mortality rates declined from 45% to 29% over a forty-year period [11].

FIP and NEC belong to the group of acquired intestinal diseases of extremely immature preterm neonates (*acquired neonatal intestinal diseases*, ANID). Abnormal postnatal microbiota plays a major role in the development of ANID. In addition, viral enteritis in preterm neonates (enterovirus, rotavirus) and cow's milk protein allergy (age > 6 weeks) may present under the clinical picture of NEC [12].

The classic NEC of the mature neonate in the presurfactant era was predominantly triggered by hypoxic-ischemic events occurring at birth; it was often associated with a severe heart defect and almost exclusively affected neonates not being breast fed. It manifested primarily in the ileocecal valve and the ascending colon.

The primary pathoanatomical substrate is ischemic hemorrhagic mucosal necrosis with edema and inflammation.

Contrasting with this, NEC of preterm neonates with a birth weight < 1500 g occurs more frequently in the middle and terminal ileum (Figure 2). It is the result of immaturity of the small intestinal mucosa, and microbial dysbiosis as well as disrupted intestinal transport function, with stasis in the middle and terminal small intestine. In these regions, there is an overgrowth of intestinal bacteria in the mucosa, which is followed by systemic bacterial invasion. Exaggerated toll-like receptor 4 (TLR4) signaling in gut mucosa cells results in an uncontrolled immune response and destruction of the mucosal barrier. This is a transmembrane protein, well known for recognizing lipopolysaccharide (LPS), a component present in many Gram negative bacteria [13].

Bell's classification can be used for the clinical evaluation of acute abdomen in preterm neonates. The clinical signs in cases of focal perforation do not strictly follow Bell's criteria, as perforation of the terminal ileum rapidly develops into a pneumoperitoneum. Bloody stools tend to indicate NEC. They can be symptoms of NEC, viral enteritis or cow's milk protein allergy (Figure 3).

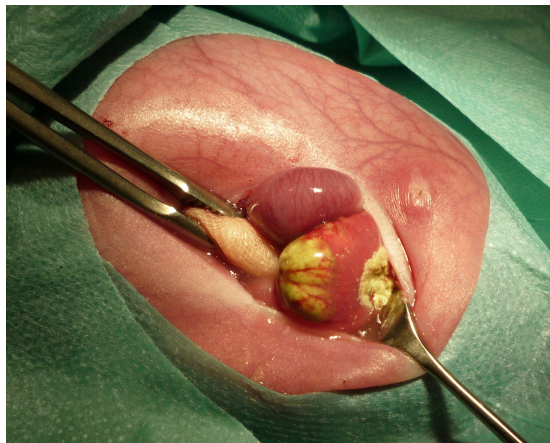
In contrast to NEC, FIP is never accompanied by pneumatosis (Bell stage II). On the other hand, one can find severe forms of NEC with sudden and almost complete necrosis of the intestine; in these cases, this most frequently occurs

without perforation.

Premature neonates of stages I (to II) are treated conservatively, *i.e.*, dietary restriction, antibiotics, fluid substitution and correction of the acid-base balance, treatment of coagulopathy/thrombocytopenia, and, if required, ventilation and administration of inotropes. In cases showing clinical deterioration and perforation, a surgical revision is necessary.

Follow-up diagnostic examinations for NEC cases include the following:

- Monitoring of clinical progress
- Monitoring of laboratory parameters (leukocyte and platelet counts, interleukin-6, C-reactive protein, blood gas analysis/lactate)
- Ultrasound including Doppler
- X-ray imaging (no contrast medium).



**Figure 2.** NEC of the terminal Ileum in very low-birth-weight infants. Very low birth weight infant at the age of 5 days.



**Figure 3.** Bloody nappies in a preterm male neonate suffering from NEC.

Significant findings during ultrasound examinations include localized edema, increased echogenicity of the intestinal wall, impaired intestinal perfusion, intramural or intravascular portal gas bubbles and “salt and pepper” signs on liver (pneumatosis of the liver) imaging.

To confirm the presence of free air, X-rays are carried out in lateral projection, with the patient lying in the left lateral side position. In the supine position, the *football* sign can be observed, whereby air accumulates at the most superior point of the peritoneal cavity.

The surgical approach consists of the resection of necroses, rinsing out of debris and decompression of the intestine. Alternatively, drainage of the abdominal cavity is suitable as a bridging measure in unstable preterm neonates. In prospective randomized trials, laparotomy or isolated drainage showed the same mortality. However, drainage operation as an individual measure usually requires an early-intervention operation [14].

Various methods for intestinal preservation exist, including the creation of enterostoma, resection and anastomosis, wedge-excision, and upstream protective stoma.

The stoma can usually be created as the laparotomy wound without the need to create a separate abdominal wall incision. Double-barreled stomata have the advantage that feces can be transferred into the lower intestine at a later point in time. According to the literature, stomata are closed at the earliest after 6 weeks. Compared to late closure, there is neither an increased complication rate nor a shortening of the parenteral nutrition period or duration of hospital stay [15]. The possibility of secondary intestinal stenosis subsequent to NEC must always be taken into account. For this reason, antegrade contrast imaging of the aboral intestine should be carried out before stoma closure.

Over the past few decades, pediatric surgeons have broken through the dogma of stoma creation. Especially in cases of focal perforation, a primary anastomosis even in patients under 1000 g body weight is feasible and shows promising results [16].

Pan-NEC is particularly associated with lethality. For these cases, high jejunostomy, second-look laparotomy or the clip-and-drop technique are appropriate treatment approaches.

Long-term effects to be expected following NEC include cognitive deficits as well as intestinal stenosis and adhesions. It has been shown that both, disturbance of the intestinal microbiota and the manifestation of NEC have a direct effect on brain functioning and development (brain-gut axis) [13]. Feeding with mother's own milk can reduce the incidence of NEC. There exists a good evidence that standardized feeding protocols and administration of probiotics to premature infants decrease the risk of NEC and death [17]. However, there are still some concerns regarding the safety of probiotic preparations.

## 2.2. Meconium Ileus, Obstruction

In meconium ileus, there is an accumulation of solidified meconium in the ter-



minal ileum that leads to the manifestation of deep small-intestinal ileus. Especially in mature neonates, the possibility of mucoviscidosis (cystic fibrosis) should always be considered. Clinically, signs may include resistance in the right middle and lower abdomen on palpation, and native X-ray imaging of the abdomen shows a fine-grained pixel-like densification on the right side, with dilated intestinal loops in the upstream bowel (Neuhauser sign). In advanced cases, “soap bubble” imaging can also be seen, reflecting a mixture of meconium and gas bubbles. Liquid levels, on the other hand, are usually not visible. Antegrade diagnostic-therapeutic administration of a hypertonic ionic contrast agent (Gastrografin®) has proven successful for uncomplicated forms of meconium ileus (Noblett’s method). In more than 50% of cases, surgery can be avoided [18]. Particular caution is required for hemodynamically unstable newborns, as the osmotic effect increases hypovolemia. Alternatively, the antegrade administration of a diluted solution of N-acetylcysteine is recommended, allowing the stool viscosity to be drastically reduced experimentally within 6 hours.

If a conservative approach does not lead to the desired result, surgical relief is necessary. The retained meconium often “plasters” the ileum, acting as a putty-like material. The surgery includes an enterotomy and rinsing maneuver. Furthermore, relief can be achieved by creating a stoma with postoperative rinsing (double-barreled ileostomy or Y-anastomosis) [19].

It is important to distinguish meconium ileus from meconium plug syndrome. The main symptom is the absence of passing meconium. Neonates are affected. A connection with maternal diabetes and magnesium or opioid administration was established. In cases of exclusive stool retention in the colon, a connection with cystic fibrosis is considered unlikely. Retrograde contrast imaging shows a slightly dilated right and transverse colon and a narrow descending colon, with abrupt transition at the left flexure. Meconium plugs are seen as multiple elongated filling defects in the dilated colon. Contrast enema using a water-soluble contrast agent is both diagnostically and therapeutically effective. If there is no improvement, Hirschsprung’s disease can be ruled out.

A special disease class is reserved for *meconium obstruction of low-birth-weight preterm neonates*.

These cases involve stool retention in the terminal ileum, stopping passing of meconium. Patients are at risk of migration and ultimately perforation. Diagnostic criteria for meconium obstruction in immature preterm neonates are as follows [20]:

- High-grade preterm birth (birth weight < 1500 g)
- Maternal risk factors (high-risk pregnancy, Cesarean section, tocolysis with magnesium)
- Mild obstruction signs (no systemic acidosis)
- Dilated loops without liquid level on X-ray examination.

Treatment of meconium obstruction is primarily conservative, but a laparotomy may be necessary. In most cases, an enterotomy is sufficient to relieve the

strain on the intestine. Depending on the findings, a decision must be made as to whether conservative treatment should be maintained or a temporary intestinal drainage (ileostomy) should be considered.

### 2.3. Hirschsprung's Disease, Congenital Aganglionic Colon

The main symptom of Hirschsprung's disease (HD) is the absence or inadequate passing of meconium, with increased bloating of the abdomen and increased bilious vomiting.

Embryologically, Hirschsprung's disease is based on a disorder of embryonic migration and maturation of neuroblasts from the neural crest into the gastrointestinal tract. Approximately half of patients with familial aganglionic exhibit mutations of the RET proto-oncogene.

The name *congenital megacolon* can be traced back to the phenomenological description by *Hirschsprung*.

The fundamental morphological criteria for aganglionic are the absence of ganglion cells in the submucosal plexus and the myenteric plexus of the rectosigmoid and hyperplasia of the parasympathetic nerve fibers, which is diagnosed in the mucosa and submucosa. Functionally, the innervation disorder of the rectosigmoid manifests itself as an absent rectoanal relaxation reflex (rectal manometry). Complications threatening sufferers of Hirschsprung's disease include Hirschsprung's enterocolitis with bacterial translocation and toxic megacolon as well as septic shock.

The presence of a narrow rectosigmoid (narrow segment) can be confirmed by a retrograde colonic-contrast enema. Diagnostic confirmation of HD is based on rectal mucosal biopsies using conventional, enzyme and immunohistochemical staining (conservative histological investigation and enzymatic or immunohistochemical staining). "Bed-side" suction biopsies are widespread in the form of harvesting mucosa or, alternatively, incision biopsy under general anesthesia. At least one biopsy is taken (if required, a full-wall-thickness biopsy including the myenteric plexus), or 2 to 3 biopsies of mucosa and submucosa (mucosa including the submucous tissue layer) are obtained [21] [22].

We prefer incision biopsies of the posterior rectal wall, 1 to 3 cm above the dentate line. The samples should be 3 to 5 mm in size and exhibit gray-white tissue of the lamina propria mucosae beneath the mucosa layer.

Pronounced hypertrophy of acetylcholinesterase (ACHE)-positive nerve fibers in the mucosa provides evidence of an HD diagnosis. In the first 3 weeks after birth, acetylcholinesterase detection in a mucosal biopsy may lead to false-negative results.

The symptomatic treatment for HD is intestinal relief by means of enemas ("wash out"). Alternatively, creation of a colostoma or ileostoma should be considered. Double-tube enterostomy (right transverse colon or terminal ileum) allows for the use of the aboral section for irrigation. Extramucosal colon biopsies can also be taken during the stoma procedure. The definitive determination of

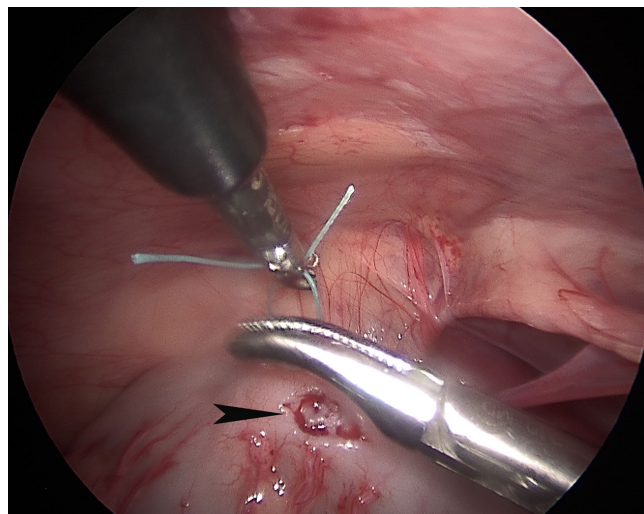


the length of the intestinal section to be resected is carried out visually as well as by means of intraoperative frozen sections as part of the pull-through operation (**Figure 4**).

At surgery, the narrow aganglionic segment including the transition zone has to be resected. Various surgical procedures are available for definitive resection and colorectal or coloanal anastomosis (pull-through). In many clinics, transanal pull-through following the *Soave/de la Torre* technique (transanal anorectal pull-through, TERPT) has become the method of choice. Both biopsies as well as skeletonization of the rectosigmoid can be performed efficiently using laparoscopic techniques.

Depending on the center, other surgical procedures (*Duhamel—OP*) lead to equivalent results. Irrespective of the method used, it can be assumed a priori that the colon will continue to exhibit a disorder with transport and continence problems, even after successful resection. The recipe for success for a continence-preserving pull-through surgery is as follows: a regular ganglionic colon, a well-executed pull-through (cave overstretching the anal canal) and a correctly positioned anastomosis 2 cm above the dentate line [23]. Instances of enterocolitis after pull-through surgery are not uncommon and may be associated with continence at a later point in time. As an alternative to the traditional three-step procedure (protective stoma, resection and pull-through, take-down of the stoma in patients 4 - 5 months of age with a body weight > 5 kg), a primary pull-through with or without a temporary stoma can also be performed [24]. The primary time-bridging treatment until pull-through can be performed is often by means of rectal wash outs.

After occlusion of the colostoma, patients should expect a very high stool frequency, which stabilizes over the course of a few months to a few evacuations per day.



**Figure 4.** Laparoscopic extramucosal biopsy is taken during pull-through surgery. The arrow indicates the intact mucosa at the biopsy site. Male infant with Hirschsprung's disease at the age of 3 months.

We are faced with a particular problem where there is aganglionia associated with *type-B intestinal neural dysplasia (IND)*, occurring in up to 40% of cases [25]. IND-B is associated with obstipation, defecation disorders and enterocolitis. The main morphological criterion is a hyperganglionia of the submucosal plexus, with a proportion of >20% giant ganglia (at least 8 neurons each) in 25 counted ganglia.

The clinical significance of a relevant finding is controversial. A maturation variant or disorder, a normal variant or a reactive change of the enteric nervous system prior to stenosis are discussed in addition to an independent disease class. In cases of an IND-B, conservative treatment measures are preferable as first-line options. In cases of treatment-resistant intestinal transport disorders of unclear origin, it is important to consider the possibility of *chronic intestinal pseudoobstruction (CIPO)*. CIPO comprises a heterogeneous group of intestinal transport disorders without evidence of anatomic or neurologic bowel wall pathology. A diagnosis can be made in neonates if the symptoms persist for more than 2 months after birth and defined causes have been ruled out. Since the exact pathogenesis remains unclear, a causal treatment is not possible. Due to the seemingly inexplicable cause of the disease, affected patients are often subjected to repeated procedures with only marginal success [26]. The aim of the treatment is to improve the quality of life on the basis of adapted enteral or parenteral nutrition, usually including jejunal tube feeding.

*Megacystis, microcolon, intestinal hypoperistalsis syndrome* is characterized by protrusion of the abdomen due to enlargement of the urinary bladder (absence of subvesical obstruction) and dilated small intestine in cases of microcolon [27], which is caused by smooth muscle myopathy. Recently, severe esophageal dysmotility (megaesophagus, candidiasis, eosinophilic esophagitis) has been found as an additional feature of this syndrome [28]. Antegrade contrast medium imaging shows hypomotility of the small intestine with partially retrograde peristalsis. The colon contrast inflow provides evidence of microcolon. Since 3/4 of patients are girls, MMIH should be considered during prenatal ultrasound in all female cases. There is no curative therapy. The bladder is emptied by intermittent catheterization or cystostomy. Parenteral nutrition and gastrostomy or ileostomy are often necessary.

## 2.4. Malrotation, Volvulus

Impaired herniation and subsequent return of the embryonic midgut into the embryonic coelom are basic mechanisms for the development of malrotation of the gut. In addition, *malrotation* or *nonrotation* is accompanied by inadequate fixation of the intestine, known as *Mesenteriumileocolicum commune*. The pendulum-like midgut attached to a narrow mesenteric stalk predisposes patients to future volvulation around the axis of rotation of the upper mesenteric vessels. In cases of midgut volvulus, subtotal hemorrhagic intestinal infarction can occur within just a few hours.

The variety of forms of intestinal fixation and rotation disorders is vast. In addition to a lack of rotation (nonrotation), there are cases with incomplete or, very rarely, even inverse rotation, as well as internal hernias (mesocolicparaduodenal hernias).

The main symptom of volvulus is unexpected bilious vomiting accompanied by acute abdominal pain. Chronic courses of the disease with failure to thrive are also possible.

The pain symptoms in preterm neonates and newborns are usually minimal. In contrast, older children show severe ischemic pain, which can occur cyclically in the course of progressive volvulation. In this context, physicians should be warned against diagnoses such as “cyclic vomiting, abdominal migraines” or “psychogenic, hysterical fits” [29].

In cases of volvulus, the time factor plays a decisive role, and clinical suspicion is critical in the case of decision-making. Importantly, native X-ray is inconspicuous in approximately 1/5 of cases with volvulus. With each “alleged” duodenal obstruction in neonates and infants, volvulus must be ruled out.

Color-Doppler ultrasound represents a fast and sensitive procedure. Normally, the *superior mesenteric vein* lies on the right side of the *superior mesenteric artery* (in cases of volvulus, the vein lies on the left side or in front of the artery). The classic ultrasound sign of midgut volvulus is the *whirlpool sign*, i.e., the clockwise twisting of the vessels of the mesenteric stalk. With oral contrast administration, a right-sided position of the duodenojejunal flexure (normally on the left side of the spinal column) indicates a rotational disorder. A high duodenal level or contrast agent not penetrating beyond the level of the third part of the duodenum are signs of a volvulus. In cases of chronic progression, sectional imaging techniques with contrast of the vessels provide the diagnosis. A retrograde X-ray colonic-contrast enema provides a sign of the volvulus in the form of a high-standing cecum and contrast medium not penetrating beyond the terminal ileum.

Isolated twisting of an intestinal loop in the form of a *segmental or compound volvulus* is typical in preterm neonates. This is possible due to lax and narrow intestinal suspension as a result of development. Iatrogenic triggering, e.g., by continuous positive airway pressure respiratory support, abdominal massage or pelvic rotation, has been discussed [30] [31]. Bilious reflux and a sunken abdomen that appears as almost airless on X-ray examination, as well as lactic acidosis (although not necessarily) are indicative signs. Since the twisted intestinal loop is often necrotic, resection and anastomosis are often required. The intestinal loss is less than that observed in cases of classic midgut volvulus.

Late-onset symptoms of volvulus are [elevated] hematin, hematochezia and intestinal paralysis. Multiple X-ray levels indicate an advanced infarction.

If volvulus is suspected, immediate surgery (laparoscopy if necessary, usually open) is vital. Laparoscopic manipulations (LADD maneuvers) are often difficult due to overstretched and rigid intestinal loops, such that laparotomies must be

performed. In cases of *strangulation ileus* due to congenital adhesions (e.g., residual material from vitellin arteries, omphaloenteric duct), laparoscopy is the method of choice.

In cases of midgut volvulus and hemorrhagic infarction, a *second-look operation* after 24 to 36 hours is an option for preserving the intestine (if necessary, also use of the clip-and-drop technique).

Discussion surrounding the removal of the appendix in cases of rotation anomalies of the intestine has proven to be controversial. On the basis of infectious-immunological reasons, we prefer *not* to remove the appendix, as it is a space for the normal intestinal flora to reside during enteritis [32] [33]. Nevertheless, parents and, depending on the case, an intelligent child should be made aware of the possibility of left-sided appendicitis [34].

## 2.5. Inguinal Hernia

The anatomical substrate is the absence of obliteration of the processus vaginalis peritonei (vaginal process; PVP). In more than 99% of cases, the child has an indirect (lateral) inguinal hernia, and patients are male in 9 out of 10 cases. The herniation of the greater omentum, bowel or adnexa and open PVP occurs more frequently in cases of preterm births and increased abdominal pressure as a result of ventilation or respiratory assistance. A right-sided inguinal hernia occurs in 60% of cases, a left-sided inguinal hernia in 25%, and a bilateral inguinal hernia in 15%.

Palpation, diaphanoscopy and especially ultrasound examinations are suitable for identifying the contents of the hernial sac. Important differential diagnoses are hydrocele, testicular torsion and, very rarely, meconium periorchitis.

In the first 6 months of life, the risk of herniation is up to 60% [35]. Inguinal hernias represent an emergency case and cause pain. Herniation in male patients is accompanied by a testicular circulation impairment and a risk of subsequent testicular atrophy. In female patients, there seems to be an increased tendency of torsion of the prolapsed ovary.

Nonreducible hernias require immediate surgery. A funiculocoele or undescended testis has to be excluded during differential diagnosis in male patients.

Preterm neonates are therefore advised to undergo surgery before discharge from the neonatological intensive care unit or, if diagnosed at a later point in time, within 2 weeks. The risk of postoperative apnea must be taken into account in preterm neonates. This is particularly high in former preterm neonates with a corrected (postnatal) age below 60 weeks, which justifies close postoperative monitoring [36].

In open surgery, the asymptomatic contralateral side is not routinely explored. In laparoscopic herniotomy in female patients, however, it is recommended to also close the contralateral PVP.

In contrast to the classical method according to Ferguson/Gross (opening of the external inguinal ring), herniorrhaphy without opening of the inguinal canal

has proven to be a fast and equally valid procedure for neonates and infants (extrainguinal method) [37].

In addition, the selective sac extraction method allows for minimizing the open approach [38]. Laparoscopic procedures have also been well validated, but there is an increased risk of retractile testes in 10% - 15% of cases in male patients [35] [39].

Depending on the patient group and the method in question, recurrence rates of 0% to 6 % following open correction and 0% to 5% after a laparoscopic procedure have been described. The cause for this may be the overlooking of a simultaneously present direct hernia (pantaloon hernia) during the initial open surgery. This is where laparoscopy shows some advantages, allowing for targeted diagnosis of unusual hernia forms (direct hernia or femoral hernia) [40].

## **2.6. Infantile Hypertrophic Pyloric Stenosis**

With a prevalence of 1:300, pyloric stenosis is 10 to 15 times more common than other congenital gastrointestinal malformations. Symptoms of pyloric stenosis appear 4 to 6 weeks after birth. Both genetic and external factors contribute to the development of pyloric hypertrophy [41]. Hypergastrinemia and gastric acid hypersecretion are mainly involved in the pathogenesis of pyloric stenosis [42]. The main symptom is the projectile, nonbilious vomiting of partially digested milk shortly after a meal. This leads to discontinuation of weight gain or weight loss. Differential diagnoses include gastroesophageal reflux, regurgitation and, in rare cases, pylorospasm without hypertrophy. Diagnosis is confirmed by ultrasound, which shows an elongated pylorus exhibiting muscular hypertrophy (length > 17 - 18 mm; muscle thickness > 3 mm). After a trial feed, the pyloric canal cannot be crossed, and a strong but unsuccessful antral peristalsis appears on ultrasound examination.

Preoperatively, both fluid loss and alkalosis are compensated by infusion of a balanced isotonic infusion solution with 5% glucose [43]. It should be noted that some neonates suffer from respiratory depression as a result of alkalosis (pulse oximetry) [44]. The preoperative insertion of a nasogastric probe has been the subject of some controversy, as an additional loss of electrolytes occurs. Pyloromyotomy allows oral fluid intake at just 4 hours after surgery. The laparoscopic procedure has become the standard approach in many clinics.

## **3. Congenital Esophageal and Gastrointestinal Malformations**

Prenatal ultrasound examination allows for the detection of approximately 60% of gastrointestinal malformations. Atresias play a special role here (Greek *atresia*, without opening or lumen), which is characterized by a congenital occlusion or a severe constriction of the esophagus or intestinal lumen. An exception here is esophageal atresia with tracheoesophageal fistula, where detection rates are 10% to 25% [45] [46]. In contrast, detection rates for isolated esophageal atresia

can be up to 85% [47]. In approximately 50% of cases, small intestinal atresia is confirmed by prenatal ultrasound, whereby there is a higher detection rate in jejunal atresia than in deeper ileal atresia [48].

The advantages of prenatal diagnosis include the possibility of preparing the parents and the avoidance of any postnatal transport, as well as providing preliminary information to neonatological, surgical and pediatric-anesthesiologic treatment teams. In-depth informing of the parents and parental decision-making should be supported on an interdisciplinary basis with cooperation of the participating disciplines. There must be regard for ethical aspects and the risks of intrauterine exposure compared to a preterm neonate. Gastrointestinal malformations in themselves are compatible with bearing a child. Prenatal interventions for the treatment of gastrointestinal malformations have not become established. Parents often fear a later limitation to learning, concentration and memory as a result of anesthesia and surgery. Here, efforts should be made for discussion with a pediatric anesthetist. At present, the data are not sufficient to rule out damage to the early infant brain due to stress and anesthetics [49]. Regional anesthesiologic procedures have been less criticized (spinal or caudal “neuroaxial” anesthesia).

### 3.1. Tracheoesophageal Fistula and Esophageal Atresia

These represent a group of malformations of the esophagus and the trachea with a prevalence of 1:4500 live births.

Tracheoesophageal fistula (TEF) and esophageal atresia (EA) result from an embryonic differentiation disorder of the foregut in the area of the esophago-tracheal septum. By far the most common combination is the *blind-ending upper pouch* and *lower TEF* (Gross, type C) [50]. Prenatal signs are polyhydramnios and a small or missing stomach. In some cases, the upper-pouch “sign” can be seen. Fetuses with EA without TEF are diagnosed prenatally more often than those with the classic form of EA.

Postnatally, the newborn’s inability to swallow saliva and colostrum as well as coughing and bouts of cyanosis are remarkable signs. In cases of clinical suspicion, an orogastric tube (6 to 8 Charrière) is used to form a clinical diagnosis (elastic resistance after 12 to 14 cm from the mouth opening). The contrast-based imaging of the blind-ending upper pouch is not a routine method as it can trigger a dangerous aspiration event. A further hazard is overinflation of the upper gastrointestinal tract due to respiratory synchronous inflation via the TEF, with resulting diaphragmatic elevation and risk of rupture of the stomach and intestines. Native X-ray examination of the thorax and upper abdomen with an inserted radiopaque probe provides conclusive evidence. Echocardiography and exclusion of relevant accompanying malformations (duodenal atresia, malformations of vertebrae, anus, kidneys and extremities; acronym VACTERL) are obligatory.

Infants with CHARGE syndrome (coloboma, heart anomaly, atresia choanae,



retardation of growth, genital and ear abnormality) can present in the same manner as EA.

Preoperative tracheobronchoscopy increases the safety of the procedure by ruling out unusual variants of the TEF (presence of a proximal fistula, presence of H-fistula) and *laryngotracheal fissure* [51].

For prognosis assessment, the criteria according to Spitz are accepted, taking into account the birth weight (>1500 g) and the presence of a major cardiac defect. The latter group includes heart defects that require either surgical intervention or treatment for heart failure. For “uncomplicated” neonates > 1500 g birth weight and without heart defects, a survival rate of 95% is assumed. There appears to be a survival threshold for preterm neonates with a birth weight below 800 g due to immaturity of the brain, lungs and intestines [52].

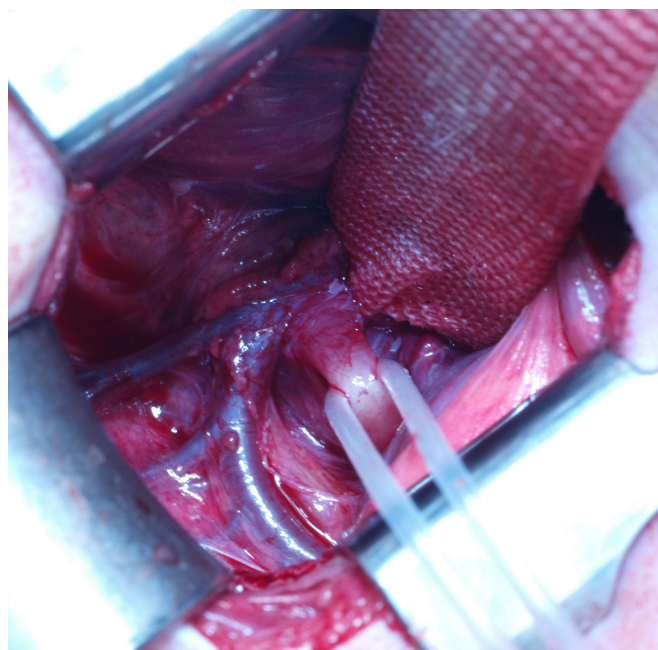
Surgery on neonates with EA/TEF is carried out with deferred urgency with the involvement of an experienced team. Perioperative care of the neonate is particularly important. After diagnosis, a two-lumen probe (*Replogle* tube) is inserted into the blind-ending upper pouch as gastric juices. In addition, regular aspiration is carried out in the mouth and throat. To avoid forced overinflation of the gastrointestinal tract, preoperative tracheal intubation should be avoided as much as possible in cases of EA and TEF.

In the standard approach, the TEF is dissected (**Figure 5**) and esophageal continuity is established by anastomosis.

Open access has traditionally been performed by retropleural preparation. Direct access through the pleural cavity is the standard procedure in the case of thoracoscopic approaches. Various techniques are recommended for overcoming large distances between segments. The flap technique according to Gogh and Bianchi or Bar Maor is suitable in cases of doubt for avoiding a maximum tension of the anastomosis in cases of “long gap.” Our preferred technique for anastomosis under tension is based on the creation of a dorsal flap of the upper esophageal pouch and insertion into the spatulated lower esophageal segment. Subsequently, the first sutures of the posterior wall can be accomplished with reduced tension [53]. The traction method according to *Foker* (open or thoracoscopic) allows for earlier anastomosis but requires two consecutive procedures (two-step).

During surgery, the transnasally inserted transanastomotic feeding tube has proven to be effective as it allows early enteral feeding. A local drain or chest tube is no longer standard in most centers [54] [55]. Immediately after surgery, hyperextension of the neck of the recently operated child should be avoided. When the neck is slightly flexed, the esophagus relaxes, as was also demonstrated in animal experiments [56].

Unstable preterm neonates, usually under 1000 g, require a risk-adapted procedure, whereby a two-stage operation with initial transpleural fistula transfixation, dissection, and subsequent anastomosis should be considered. The sole ligature with preserved continuity of the fistula is characterized by a significant early-recurrence rate, which even nonresorbable sutures cannot prevent.



**Figure 5.** The tracheoesophageal fistula is identified behind the azygos vein (loop). Surgery was performed at the second day of life.

As the newborn's ability to swallow its own saliva increases, the postoperative build-up of food intake can begin. Depending on the clinic, an X-ray examination of the anastomosis is carried out with a water-soluble contrast medium a few days after the operation.

Important variants of the malformation group are as follows: 1) isolated EA without fistula (isolated EA, Type A), and 2) isolated TEF (H-fistula) with continuous esophagus. The pathognomonic radiographic sign of *pure esophageal atresia, Gross A* (or rarely atresia with only upper fistula), is a gasless abdomen with a milky-white homogeneous turbidity (gastric bubble and intestine are free of air). Isolated atresia is, a priori, associated with a large distance between the two segments of the esophagus. A primary esophageal anastomosis is therefore usually ruled out. A temporary gastrostomy is used to bridge the time gap and allows for early nutrition. The final correction is carried out according to different temporal approaches. Depending on the clinic, an elongation treatment of the upper segment can be carried out. Cervical cutaneous esophagostomy leads to the loss of valuable esophageal length and thus should be avoided.

*Isolated TEF, Gross E*, usually presents days to weeks after birth due to respiratory problems and pneumonia. The fistula can be closed in open surgery via a cervical access, whereby the protection of the vocal cord nerves (recurrent laryngeal nerve) must be taken into consideration. As a practical hint for fistula localization during tracheoscopy, consider the possibility of transillumination by means of a tracheoscope light source while keeping the surgical room dark. In cases of deep fistulas, there would therefore be an argument for thoracic access to the TEF.

### 3.2. Pyloric Atresia and Membrane

The occlusion of the pyloric orifice occurs sporadically or in connection with an epidermolysis bullosa [57]. X-ray imaging shows an overinflated stomach bubble with a singular fluid level. Distally to this region, there is no air (with the exception of incomplete stomach membrane). Dangers result due to impending perforation of the stomach. Treatment may consist of pyloric resection and end-to-end anastomosis or side-to-side gastroduodenostomy. Prognosis is particularly critical where epidermolysis bullosa presents; sporadic cases, on the other hand, show a partially favorable outcome.

### 3.3. Duodenal Atresia, Stenosis and Annular pancreas

The frequency of these disorders is 1.5 to 2 per 10,000 births. Embryologically, this is the result of a disrupted epithelial-mesenchymal interaction during the recanalization of the originally epithelially occluded duodenum (*Tandler's* theory) [58]. Prenatally, there is polyhydramnios and a widening of the stomach and upper duodenum. In addition, prenatally diagnosed cases have a higher proportion of concomitant malformations [59]. The increased prevalence of vitium cordis and trisomy 21 (Down's syndrome) should be considered. In addition, a broad spectrum of associated malformations may be considered [60]. Postnatally, bilious vomiting and a protrusion of the upper abdomen are the main symptoms. A particularly rare case of atresia in the oral direction of the major duodenal papillae is associated with clear reflux. The native X-ray image of the stomach reveals the *double-double* mirrored formation of stomach and duodenum. For better visualization, a few milliliters of air can be instilled via the nasogastric probe before performing the X-ray examination. In unclear cases, a water-soluble contrast material provides additional diagnostic benefits. Morphological variants of duodenal atresia are membranes (with or without opening) or lumen/continuity interruptions. The standard method in surgical treatment is duodenoduodenostomy in the form of diamond-shaped anastomosis. It can be performed openly or laparoscopically where specialist expertise is available. A transanastomotic tube inserted transnasally allows for early postoperative enteral nutrition. Alternatively, membrane excision is possible (by enterotomy or, in larger children, using an endoscopic endoluminal technique). Great care must be taken when doing so to preserve the papillae. A tapering of a megaduodenum is necessary in rare cases. A preduodenal portal vein (persistent left vitelline vein) occurs in cases exhibiting heterotaxy or situs asymmetry. It runs anterior to the pancreas and is usually accompanied by an intrinsic obstruction.

*Annular pancreas* as a cause of duodenal stenosis results from a defective fusion of both embryonic pancreas anlagen (ventral and posterior). The pancreas surrounds the duodenum in a ring shape. Treatment consists of a bridging in the form of duodenoduodenostomy.

In the *Ladd* procedure, embryonic peritoneal ligaments cause external duodenal compression, starting from a cecum in a position superior to normal

(*coecum altum*). The additional narrow-base adhesion of the midgut predisposes these patients to developing volvulus. In addition to severing the Ladd bands, the operation involves mobilization of the mesenteric adhesions and positioning the intestine to nonrotation. In 2% to 7% of cases, a volvulus relapse is possible. The Ladd procedure gives patients the greatest possible chance of volvulus not recurring, but information about possible symptoms and urgency is required in the event of suspected revolution.

### 3.4. Jejunio-Ileal and Colonic Atresia

These are secondary atresias subsequent to fetal catastrophic events, leading to vascular occlusion (embolism, strangulation, segmental twisting) of the intestine. Special causalities are found in children with severe heart defects or after twin transfusion syndrome. Even at the intrauterine stage, intestinal dilatation can be seen on ultrasound examination.

The main symptoms are a constantly increasing bulging of the abdomen and bilious vomiting. Native X-ray examination usually shows several levels and an airless lower abdomen. In jejunal atresia, the filled, severely dilated, blind-ending, upper small intestine segment with wall thickening is often palpable as sausage-shaped resistance.

Morphologically, a distinction is made between membranous, strand-like or complete forms of atresia [60].

- Type I—intraluminal membrane;
- Type II—connecting myofibrous filum between both atretic ends, preserving the mesentery (**Figure 6**);
- Type III a—atretic ends are not connected in cases with mesenteric defect;
- Type III b—“Christmas tree” or “apple peel”: Occlusion of the superior mesenteric artery causes absence of part of the jejunum, and the distal ileum is maintained by collateral vessels and has an appearance suggestive of an apple peeling.

Type III atresias are pathogenetically attributed to an intrauterine volvulus or vascular occlusion of the distal superior mesenteric artery. The middle small intestine is lost and, a priori, a short-bowel situation exists. The blood supply to the remaining terminal ileum is provided by a branch of the ileocolic artery.

The preferred treatment option in cases of small-intestinal atresia is primary restoration of intestinal continuity by end-to-back anastomosis. It is almost always necessary to resect the enlarged preatretic segment sparingly. Tapering additionally allows for narrowing of the preatretic intestine. Furthermore, antimesenteric spatulation of the postatretic intestine can occur. A transnasally inserted transanastomotic probe or a temporary catheter-jejunostomy following the Witzel principle allows for early enteral nutrition. However, studies have questioned the effectiveness of this approach.

*Colonic atresias* are particularly rare (**Figure 7**). A swelling of the abdomen and vomit containing bile and meconium are the main symptoms. Treatment

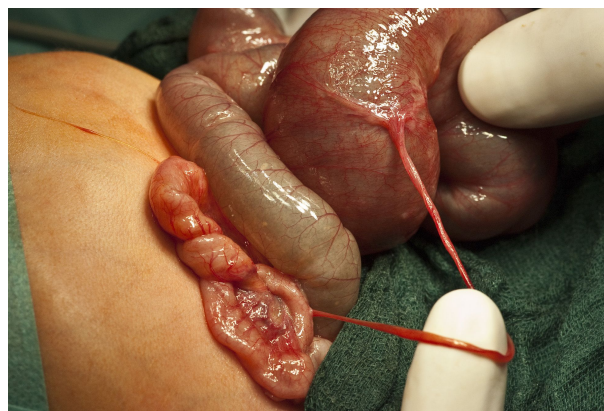
approaches correspond to the aforementioned. If necessary, a temporary stoma is created. Whenever possible, the ileocolic transition should be maintained with the ileocecal valve.

Subsequently, to surgery for a high-located atresia, a jejunal probe for early build-up of enteral food intake has proven effective.

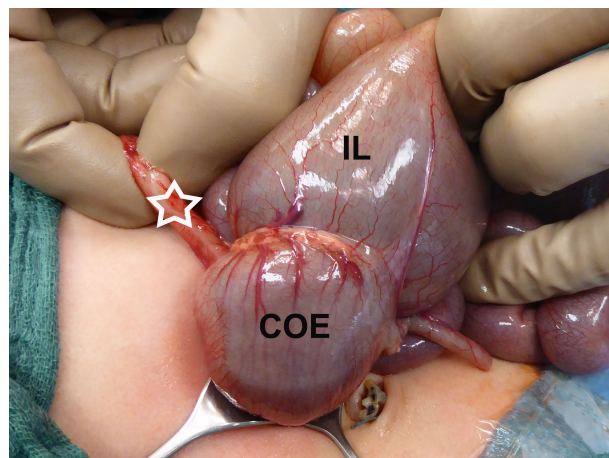
Food intake is started orally in consultation with the pediatric surgeon after a few days, usually after evacuation of the first instances of fresh stool.

### 3.5. Anorectal Malformation

This is a broad spectrum of malformations that affect the rectum as well as the anatomy of the pelvic floor and urogenital tract. If the anal opening is absent or narrowed, the diagnosis becomes apparent immediately after birth. Depending on the complex embryology of the urorectal septum or cloaca, very different gender-dependent malformation patterns occur. In approximately 2/3 of cases, there are accompanying malformations of the urogenital system, the heart and as part of a VACTERL association.



**Figure 6.** Type II atresia of the jejunum with a fibrous band between both segments. Male neonate at the second day of life.



**Figure 7.** Colonic atresia type I distally to the cecum. Both terminal ileum (IL) and cecum (COE) are dilated. Tiny, postatretic colon ascendens (star). Surgery four days after birth.



The traditional wingspread classification (1984) divided malformations into deep, translevatoric, intermediate and high, and supralelevatoric. **LITERATUR [61]** These refer to X-ray imaging taken in a hanging position (*Waterstone* image). An imaginary reference line is drawn between the pubic bone and the tip of the coccyx. This line represents the attachment of the levator muscle to the bony pelvis.

The *Krickenbeck* classification replaced the wingspread classification, particularly with regard to the standardized recording and comparability of surgical results. The main clinical groups of the Krickenbeck classification are follows [62]:

- Perineal, cutaneous fistula (male and female) (**Figure 8**)
- Recto-urethral (prostatic, bulbous)
- Recto-vesical fistula
- Vestibular (female)
- Cloaca (female)
- Blind-ending rectum without fistula
- Anal stenosis

There are also rare or local variants (pouch colon, H-fistula, rectal atresia or stenosis, recto-vaginal fistula).

For external evaluation, the localization, width and elasticity of the opening, the length of the perineum, the presence of an anal dimple, and an anal rosette are assessed.

In male patients with no anus, a confluence between rectum and urethra (prostatic or bulbar fistula) must always be considered. Clinically, dark-green meconium is found in the urine (assess diaper). Intermediate forms result as tiny stool fistulas along the raphe.

In female patients, recto-vestibular fistulas are most common. In these cases, the opening is hidden in the posterior vaginal commissure. If there is only one opening in the neonate girl, this is the opening of a cloacal malformation.



**Figure 8.** Female perineal fistula (arrow head) just behind the vestibulum vaginae. Newborn infant. Supine position.



Postnatal standards particularly include echocardiography and ultrasound of the urogenital system and lower spine.

If the anal opening is missing, it is possible to perform an X-ray the day after birth using the Columbia technique, *i.e.*, elevation of the pelvis using rollers, horizontal projection, marking of the anal dimple using lead plates and evaluation of the height of the air-filled rectum.

A subsequent micturition cystourethrography (MCU) is indicated in cases of sonographic abnormalities in the upper urinary tract. In cases of a stoma creation, the Columbia imaging can usually be replaced by a subsequent contrast-medium-pressure-cologram (“loopogram”).

To perform this procedure, balloon catheters are used to infuse water-soluble contrast medium into the aboral stoma section, and a fistula of the rectum to the urogenital tract is visualized or ruled out.

The limiting factor during correction with relation to achievable continence is the formation of the muscular levator complex. As a rule of thumb, in the case of flat perineum, missing anal dimple, and hypoplasia of the coccyx and sacrum, a reduced continence apparatus is to be expected.

The diversity of deep perineal variants deserves special attention [63] as follows:

- *Anterior anus.* Forward displacement of a “perfect” anus, with opening and anal rosette located in the anal funnel. In cases of symptoms, the relationship of the anal funnel to the sphincter complex must be checked. Sometimes there is a mucosal groove to the vestibule/rear vaginal commissure (*perineal groove*).
- *Anal stenosis or membrane.* There is a constriction or membrane at the level of the dentate line. The membrane can be treated by excision and/or dilatation treatment. A skin bridge (“bar”) covering the anal opening may be resected (depending on each case).
- *Funnel-shaped anus* with very deep anal funnel and stenosis at the skin-rectum transition. This rare form is usually associated with a pronounced megarectum. In addition, *Currarrino’s triad* (anal stenosis, defect of sacral bone, presacral tumor) should be expected. Anal stenosis can be dilated with a bougie. If the megarectum is pronounced, resection may be necessary.
- *Perineal fistula.* Morphologically, the regular structures of an anal canal are formed above the stenotic anus.

The pivotal point in the surgical treatment of these malformations is the concentric embedding of the newly placed intestine in the muscular levator-sphincter complex. The standard procedure is posterior sagittal anorectoplasty (PSARP). This procedure was performed for the first time in 1980 by A. Pena.

The PSARP is based on some very important anatomical findings. Pena and De Vries described a functionally important muscle continuity from the skin to

the sacral insertion of the levator ani [64]. The electrophysiological and visual localization of this continence apparatus is the supporting element of an anatomically based, continence-preserving procedure. The method is based on a splitting of the levator muscle complex in the midline, with the aim of fistula transection and mobilization of the blind sack and reimplantation of the anus at the target site.

Perineal fistulas can initially be treated with a bouginage treatment using Hegars dilators. A correction using mini-PSARP can then be performed electively and without stoma.

The definitive correction of high-grade malformations takes place at the age of approximately 8 weeks with protective colostomy creation. The perioperative insertion of a urinary bladder catheter is obligatory for several days (girls 3 days; boys 5 - 7 days).

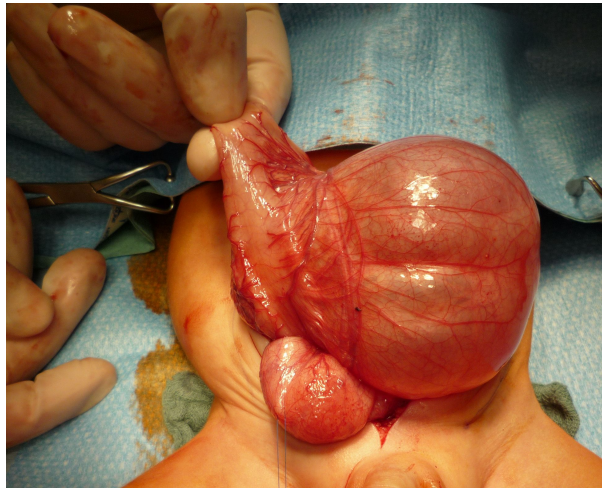
A laparotomy is only necessary for high anorectal malformations, with accessibility of the rectum via the abdominal cavity (e.g., bladder neck or vesical fistula). New possibilities for correction have arisen by means of laparoscopic fistula dissection and laparoscopically assisted pull-through [65]. The puborectal slings are made visible from inside and outside by means of electromyostimulation. Endoscopically assisted techniques should also be considered as a therapeutic option [66]. In this procedure, endoscopic transillumination is used to make the blind sac of the rectum externally visible and to perform a proctoplasty via a limited access via the anal funnel.

It is unclear whether a one-step surgery without protective stoma in female patients with vestibular fistula will produce results equivalent to those of a classic two-step surgery. A recent meta-analysis found that this procedure is associated with an increased rate of postoperative complications, without the continence rate (soiling, constipation) ultimately deteriorating [67].

### 3.6. Esophageal and Gastrointestinal Duplications

Duplications occur along the entire intestinal tube. These are spherical or cylindrical duplications of the lumen whose wall contains both mucous membrane and musculature. Duplicates are usually separated from the intestinal lumen or can be in an open connection with the lumen.

Embryology cannot be explained conclusively, such that a number of embryological theories have been put forward, including, split-notochord theory, persistence of embryonic diverticula, and disturbance of intestinal recanalization. Prenatal ultrasound diagnosis is often performed. Prenatal ultrasound most frequently detects ovarian cysts; less frequently, it detects gastrointestinal duplications and mesenteric cysts, meconium pseudocysts and other entities [68]. Usually, an organ allocation can already be determined prenatally. Cystic structures may also indicate an anorectal malformation, *i.e.*, a malformation group whose prenatal diagnosis is often unsuccessful. In approximately 1/4 to 1/3 of fetuses with cysts found prenatally, surgical intervention is indicated [69].



**Figure 9.** Large duplication cyst of the sigmoid colon containing intestinal mucosa. Male newborn infant with congenital protrusion of the lower abdominal wall.

Duplications most frequently occur in the ileum and jejunum, followed by the esophagus and colon (**Figure 9**). After birth, duplications initially remain asymptomatic. Increases in size, secondary infection and or as a result of disseminated dystopic gastric or pancreatic tissue, complications such as intestinal transit disorder, rupture and peritonitis occur. Clinically, physicians often feel a mobile mass in the abdomen. Usually, an ultrasound is sufficient for further identification. An elective indication for surgery exists due to the complications that might arise. The method of choice is resection with maintenance of continuity. However, resection of the affected intestinal section cannot always be avoided. Differential diagnoses are, in particular, mesenteric cysts, lymphangioma, and ovarian cysts.

#### 4. Conclusion

There is a clear trend toward less invasive approaches and the application of one-stage reconstructive surgery. The increasing rate of very immature neonates is a challenge for neonatologists and pediatric surgeons as well. Immaturity of the intestinal barrier function and intestinal microbiota play a major role in this group of patients. Reduction of exposure to invasive procedures, surgical strategies to reduce the amount of antibiotics, and immunonutrition offer the chance to reduce the burden of surgery in very preterm infants.

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#### Conflicts of Interest

The author declares no conflicts of interest regarding the publication of this paper.

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## Abbreviations

ACHE: acetylcholine esterase  
ANID: acquired neonatal intestinal diseases  
CIPO: chronic intestinal pseudoobstruction  
CPAP: continuous positive airway pressure  
EA + TEF: esophageal atresia + tracheoesophageal fistula  
FIP: focal intestinal perforation  
HD: Hirschsprung's disease, aganglionic colon  
IND: intestinal neuronal dysplasia  
NEC: necrotizing enterocolitis  
PVP: processus vaginalis peritonei  
TERPT: transanal endorectal pull-through