Abstract
Pediatric anorectal disorders include a variety of relatively common functional and anatomical defects. Around 10% of children are brought to medical attention because of an anorectal disorder. Besides constipation, other typical complaints include blood in the stool, pain, and specific local anal findings. It is essential that pediatric surgeons are familiar with the pathophysiology and management of these diverse and usually relatively benign conditions, as these problems are not uncommon and are regularly seen in emergency room and outpatient clinics.
Introduction

Pediatric anorectal disorders are common, almost always benign processes and rarely associated with an underlying pathology (Jamshidi 2018). Pediatric anorectal disorders include a diverse group of functional and anatomical defects that may cause significant discomfort and disability. It has been estimated that 10% of children are brought to medical attention because of an anorectal disorder, constipation being the most common of these problems. Other typical complaints include blood in the stool or pain along with specific local anal findings. Many pediatric anorectal disorders are easily treated if the child receives prompt and adequate attention. However, children commonly present long after the onset of symptoms, which may render the condition much more difficult to treat. It is essential that pediatric surgeons are familiar with the pathophysiology and management of these diverse and usually relatively benign conditions, as these problems are not uncommon and are regularly seen in emergency room and outpatient clinics. Hirschsprung’s disease, anorectal malformations, and inflammatory bowel disease are addressed elsewhere in this book.

Constipation

Constipation in children is a common condition, especially in the Western World. In the majority of patients, the cause remains obscure; therefore, the condition is called functional constipation. According to the Rome III classification, functional constipation in children is defined as constipation not associated with congenital abnormalities, acquired diseases, or medication (Hyman et al. 2006). The following criteria apply for children of all ages; two or more of the following has to be fulfilled:

1. Two or fewer defecations in the toilet per week
2. At least one episode of fecal incontinence per week (after acquisition of toileting skills)
3. History of retentive posturing or excessive volitional stool retention
4. History of painful or hard bowel movements
5. Presence of a large fecal mass in the rectum
6. History of large diameter stools that may obstruct the toilet

Besides constipation, fecal soiling is also common. Soiling occurs in approximately 3% of children older than 4 years of age, and constipation accounts for at least 3–5% of all medical and 25% of pediatric gastroenterology referrals (Levine 1981; Kyrklund et al. 2012; Flemming 2019). Most studies suggest a male predominance. During the 1st year of life, organic causes for constipation should be ruled out, especially in the case of delayed passage of meconium.

Acute constipation. Acute constipation may be secondary to physical inactivity, changes of environment or diet, or an anal fissure. Presentation as acute abdominal pain is common. It is usually relieved by one single enema. The management of acute constipation is straightforward. Increasing dietary fluids and fiber and restriction of cow’s milk intake usually relieve the symptoms. A short course of bulk laxatives may prevent recurrence of symptoms.

Chronic constipation. Persistent constipation, which does not rapidly respond to dietary manipulation or simple laxative treatment, can be defined as chronic. A child with chronic constipation commonly presents with overflow fecal incontinence. Organic causes that should be taken into account in the diagnostic workout for chronic constipation are summarized in Table 1. Chronic constipation and associated soiling typically presents between the ages of 2 and 4 years, but symptoms may begin during the 1st year of life. The diagnosis of chronic constipation relies on history and clinical examination. The incidence of clinical findings in patients with chronic constipation is summarized in Table 2. On clinical examination, the abdomen is usually not tender
and rarely distended. Stool masses are often palpable. Perineal inspection should verify the position and condition of the anus and the perianal skin including soiling marks. The normal position of the anus must be defined, since anterior position of the anus may cause constipation. Perianal sensation should be also tested. It is advisable to perform a digital rectal examination at least once in a child with chronic constipation to rule out organic obstructing causes such as presacral tumors.

A small infant with a history of neonatal symptoms and early-onset constipation should undergo barium enema without bowel preparation. An abnormal barium enema should be followed by a rectal biopsy and anorectal manometry, if available, to rule out Hirschsprung’s disease and allied disorders. Diagnostic studies are not required in older children without any history of significant neonatal constipation, before initiation of medical management. Imaging studies such as barium enema, transit time studies or MRI, and anorectal manometry and biopsies may be indicated in patients who have a poor response to appropriate medical treatment or abnormal clinical findings. Slow-transit constipation that is common in adults, especially in young women, has been reported to occur in a significant proportion of children with recalcitrant constipation (Hutson et al. 2009).

**Management of chronic idiopathic constipation.** If the clinical history and physical examination do not suggest organic etiology, a trial of medical treatment can be initiated. The general approach to the management of chronic constipation includes the following steps (Clayden and Agnarsson 1991):

- Provide parental counseling and education.
- Determine whether a fecal impaction is present.
- Disimpact the fecaloma if present.
- Initiate oral medication.

The pathogenesis and prognosis of constipation need to be explained to the parents. The involuntary nature of associated overflow incontinence is clarified to the parents, and the family is informed that it may take months or even years before normal stooling pattern is achieved. The recommendation for the management of functional constipation includes a normal intake of fibers and fluids, normal physical activity, and
pharmacologic treatment of fecal impaction followed by pharmacologic maintenance therapy (Flemming 2019). If the patient has fecal impaction, disimpaction is necessary before the initiation of oral maintenance therapy. A typical symptom of fecal impaction is overflow incontinence. Disimpaction has been traditionally accomplished with bowel washouts, but oral medication is also effective. The goal of maintenance therapy is to produce one to two soft stools per day and prevent recurrent fecal impaction. The treatment consists of dietary interventions, laxatives, and behavioral modification. Dietary changes are universally advised, particularly increased intake of fluids and fiber. There is no clear evidence that any particular drug regimen is superior in the management of chronic constipation. Good compliance and commitment to the selected treatment are more essential. Initially, more effective medication with stimulant laxatives (senna, bisacodyl, sodium picosulfate) is often required. These should be used for as short a period as possible and should be replaced by less harmful therapies. Osmotic laxatives (lactulose, docusate, PEG) can be safely used for months or years. A recent meta-analysis has shown that PEG is superior to lactulose for childhood chronic constipation (Lee-Robichaud et al. 2010).

After a regular stooling pattern is achieved, medication is continued as long as necessary, usually for months and sometimes for years. A good rule of thumb is that the treatment continues at least as long as the patient has had symptoms prior to the management. There are a few studies of the long-term results of the management of chronic idiopathic constipation. The majority improve within 6–12 months following onset of appropriate therapy (Pijpers et al. 2010). On the other hand, recent longitudinal follow-up studies have shown that constipation continues beyond puberty in one fifth to one third of children with chronic constipation (Bongers et al. 2010). There are no controlled studies on the surgical treatment of pediatric constipation. The majority of children with chronic constipation improve with appropriate medical therapy in time. However, a few children remain, who despite maximal medical therapy have persistent constipation and soiling and may benefit from surgery. There are some surgical options for these patients. It is essential that organic causes of constipation are ruled out. The antegrade colonic enema (ACE) procedure has also been successfully used for idiopathic constipation (Youssef et al. 2002). The ACE procedure is easily reversible, which may make it an attractive alternative, especially for pubertal and adolescent patients. Resection of the megarectosigmoid has been performed for patients with functional constipation, but there is a lack of controlled studies and longer-term follow-up data (Levitt et al. 2009). Moreover, high failure rates have been reported following colon resections (Gladman et al. 2005). Long-term bowel diversion has been used in selected patients with significant patient and parent satisfaction (Woodward et al. 2004). Other options that have shown some promise include spinal neuromodulation and transcutaneous electrical colon stimulation (Clarke et al. 2012). Pediatric experiences from these modalities are based on small series.

**Functional Fecal Soiling Without Constipation**

Nonretentive fecal soiling is occasionally observed without associated constipation or any other organic or neuropsychiatric underlying cause for the incontinence (Pakarinen et al. 2006; Rajindrajith et al. 2013). The principal complaint is involuntary passage of variable amounts of stool into underwear one to several times a day. Most patients have regular bowel actions without a history of constipation, although mixed forms also exist. The most common age of presentation is between 4 and 8 years of age. A significant proportion (40%) of these children also demonstrates daytime or nighttime enuresis. Patients have often received a variety of unsuccessful therapies for constipation.

Although pathophysiology remains unclear, anorectal manometry may demonstrate an isolated impairment of rectal sensation (Pakarinen et al. 2006). The sphincter pressures and performance are well preserved. Colonic transit times are in the
normal range. No significant bowel dilatation or fecal impaction is seen in contrast enema, and spinal radiography is normal, and, when performed, magnetic resonance imaging indicates no signs of myelopathy. Treatment consists of counseling, toilet training, and dietary modifications. Some patients, especially those with accompanying enuresis, may benefit from anticholinergic treatment with oral oxybutynin hydrochloride (5–15 mg/day) by demonstrating a decreased frequency of soiling. Improvement of soiling may occur spontaneously by adolescence.

**Rectal Prolapse**

A complete rectal prolapse is a circumferential several centimeters long full-thickness anal outpouching of the rectum. The circular folds of the rectal mucosa are visible on the prolapse. A mucosal prolapse is usually shorter than 3 cm long and asymmetric. Peak incidence of rectal prolapse occurs in children between 1 and 3 years.

**Pathogenesis.** Rectal prolapse occurs most often in an otherwise healthy child. Constipation is uncommon and not a typical feature of prolapse. Prolapse can occur secondary to paralysis of the levator ani muscles, most often in association with myelomeningocele. In patients with bladder extrophy, wide separation of the symphysis pubis and puborectal muscle predisposes to prolapse, as well as misplaced anus and stretched levator ani muscle in patients with presacral teratoma. In addition, severe malnutrition, chronic diarrhea, cystic fibrosis, rectal polyps, behavioral disorders (e.g., Asperger), and connective tissue disorders (Ehlers-Danlos) predispose to rectal prolapses. After anorectal operations for anorectal malformations or other disorders, mucosal or complete prolapses can occur (Peña et al. 2007). The etiology of rectal prolapse in healthy children is thought to be associated with low pelvic location of the rectum, immature rectal fixation, and straight anorectal angle. According to another theory, a distally advancing sigmoid intussusception eventually reaches the rectum and pushes out of the anus, but radiological evidence for this is found only in some of the patients.

**Diagnosis.** Rectal prolapse occurs usually only during defecation, and it may be painless and reduce spontaneously. Often, however, manual reduction is needed. It may cause hemorrhage, mucus excretion, and pain. Occasionally, an edematous prolapse is not manually reducible, and reduction under anesthesia is required. Reduction of a rectal prolapse under anesthesia does not preclude continued conservative treatment. Rectal prolapses associated with malnutrition and infectious diarrhea may be of significant size, remain prolapsed, and cause incontinence (Ismail et al. 2010). Usually, prolapses are diagnosed by positive history. The diagnosis can be confirmed with the help of a photograph taken by the parents. It is exceptional that prolapse can be demonstrated during straining or rectal examination in an outpatient clinic. In the majority of patients (85%), the condition is self-limiting, and no surgical intervention is needed (Koivusalo et al. 2006). Surgery is indicated if recurrent prolapse causes bleeding and pain or reduction is difficult. In school-aged children, recurrent prolapse may cause problems in hygiene and discomfort associated with lavatory visits. For this reason, it is recommended that children with a rectal prolapse, without signs of attenuation of symptoms, should undergo surgery at 7 years of age or around the time school is begun.

Radiologic imaging of rectal prolapse is recommended when the symptoms are atypical or when there is associated abdominal or anal pain. Radiologic imaging is also recommended whenever surgical treatment is considered. A standard defecography is most widely used. The patient defecates the rectally injected contrast while sitting, and the descent of the pelvic floor with rectal prolapse is verified by cinefluoroscopy. Oral water-soluble contrast given half an hour before the study may demonstrate an enterocoele, i.e., the descent of small bowel loops into the pelvis and into the folds of the prolapse during defecation. Other radiologic features include sigmoid intussusception and anterior bulging of the rectal ampulla (rectocele), with or without prolapse, and rectal intussusception without prolapse (Koivusalo et al. 2012). Defecography may be performed to children
from the age of 4 years onward. In older and cooperative children, a MRI defecography may be used. Colonoscopy is indicated in patients in whom rectal polyp or solitary rectal ulcer syndrome (SRUS) is suspected.

**Treatment.** Surgical therapy aims to prevent the prolapse by surgical fixation of the rectum. Multiple operative and procedural approaches to rectal prolapse exist with variable recurrence rates and without a clearly superior operation (Rentea and St Peter 2018). These include sclerotherapy, Thiersch’s anal cerclage, Ekehorn’s rectoplasty, laparoscopic suture rectoplasty, and posterior sagittal rectoplasty. If a high sigmoid intussusception is verified, an anterior rectosigmoid resection may be added. Patients with rectocele, enterocele, or rectal intussusception without a verified clinical prolapse whose symptoms do not respond to conservative treatment may also benefit with rectopexy. Perineal operations, including perianal Thiersch sutures, Ekehorn operation, linear electrocaulation, mucosal ablation, phenol injection, or plication of the muscular wall, are not recommended in full-thickness rectal prolapse. Mucosal prolapses cannot be controlled with rectopexy, and local excision is the preferred method.

Laparoscopic rectopexy is a relatively simple procedure and may be used as a primary therapy (Koivusalo et al. 2006, 2012; Laituri et al. 2010). The mesorectum is mobilized posteriorly up to the sacral promontorium. Holding the rectum taut, both sides of the rectal wall are fixed with two strong non-absorbable sutures to the promontorium. In patients with enterocoele or rectocele, peritoneum between the rectum and vagina or urinary bladder is closed to prevent descent of small bowel into pelvis and to reinforce the anterior rectal tissue. Perineal operations, including perianal Thiersch sutures, Ekehorn operation, linear electrocoagulation, mucosal ablation, phenol injection, or plication of the muscular wall, are not recommended in full-thickness rectal prolapse. Mucosal prolapses cannot be controlled with rectopexy, and local excision is the preferred method.

Solitary Rectal Ulcer

Solitary rectal ulcer syndrome (SRUS) is a chronic, benign disorder characterized by hematochezia, mucous discharge, tenesmus, and local perianal pain. It is uncommon in children (Dehghani et al. 1999). The macroscopic finding at endoscopy is a thickened and edematous lesion that may have an ulcerative or polypoid appearance in the anterior rectal wall 3–10 cm above the anal verge. Rectal or internal rectosigmoid prolapse has been reported to occur in a significant percentage of cases. Conservative management with stool softeners and local steroid suppositories is successful in most children. Open or laparoscopic rectopexy has been suggested to correct the external or internal rectal prolapse that is often associated with SRUS.

Perianal Abscess and Fistula

Perianal sepsis including abscesses and fistulas is common in small infants. The majority of patients are male (Oh et al. 2001). Etiology of infant anal fistulas may be related to congenital androgen activity resulting in deep, epithelialized crypts explaining the predominant occurrence in males. A typical presentation is a tender, sometimes fluctuating, perianal mass with or without discharge in a child younger than 12 months, most commonly during the first 6 months of life. After the initial incision, the condition may progress to or recur as an anal fistula. The incidence of associated fistula tract in patients with perianal abscess may be as low as 10–20% (Macdonald et al. 2003), while fistulas may also present without a preceding perianal abscess. The fistula tract is typically subcutaneous and straight. It traverses from the affected crypt subcutaneously or through the lower part of the external sphincter to the perianal skin. The fistulas occur evenly around the anal circumference. Multiple lesions occur in 15–20% of cases.

The traditional management of perianal abscesses involves incision and drainage, which is associated with a significant recurrence rate (Murthi et al. 2002). Although an incision provides rapid pain relief, eventually most abscesses
are cured by expectant treatment only (Rosen et al. 2000). Anal fistula has been considered as an indication for surgical treatment. The traditional method has been fistulotomy, by incising the fistula tract after identification of the affected anal crypt. Recurrent fistulas occur in 10–20% of cases. Recent reports have advocated expectant treatment for asymptomatic anal fistulas. Most fistulas heal within 12–24 months without further sequelae.

Perianal fistulas in older children and adolescents are cryptoglandular in origin or associated with inflammatory bowel disease (Yamana 2018). Crohn’s disease should be considered in older children and ruled out with endoscopic examinations and mucosal biopsies in those with associated bowel symptoms or positive family history. Treatment of perianal fistulas in adolescents follows the same lines as in adults. The fistulous tract, once identified and probed, is either incised and left open to granulate or excised with primary closure of the defect. More complex fistulas are assessed by magnetic resonance imaging and treated with setons.

**Anal Fissure**

Anal fissure is a longitudinal ulcer in the squamous epithelium of the anus, located just distal to the mucocutaneous junction extending to the anal verge. Most acute fissures heal spontaneously within a few weeks, but some become chronic and require therapeutic measures. An anal fissure causes anal pain during defecation and is an important cause of hematochezia in children. Typical age of presentation is 2 years. The exact incidence is unknown.

**Etiology and pathogenesis.** The etiology of an anal fissure is not clear. In children, a fissure is often associated with secondary constipation and painful passage of stools, but it is unlikely that hard stools are the primary cause. A painful anal tear may lead to a fear of defecation and fecal retention, thus initiating a vicious circle. In adults, the underlying pathology includes hypertonia of the internal anal sphincter with the resulting ischemia causing a fissure. The fissure is most often located in the posterior midline corresponding with an area of the anal canal less vascularized than other areas. Decreasing the sphincter hypertension by surgical or pharmacological intervention improves perfusion and allows healing of the fissure. It is not certain whether an identical etiopathology applies in children. A fissure may become inflamed due to bacterial infection and chemical and mechanical irritation. A hypertrophied anal papilla proximally and a skin tag distally to a fissure are signs of chronic infection and should raise suspicion of Crohn’s disease.

**Diagnosis.** Typical symptoms are blood streaked stools, with or without painful defecation. The diagnosis is made by direct inspection by gently retracting the perianal skin. If a fissure is seen, further digital rectal examination and proctoscopy are unnecessary. The most common location is posterior midline, but in infants fissures may be seen in all locations, and in female infants a common site is the anterior midline. Atypical appearance of the fistulas, such as signs of chronic infection, multiple location, and irregularity should be investigated by biopsy and colonoscopy to rule out Crohn’s disease, immunodeficiency states, venereal infection, tuberculosis, and malignancy.

**Treatment.** In children, most idiopathic anal fissures heal spontaneously in a few months. Treatment of a symptomatic fissure consists of lubricant, e.g., Vaseline to ease passage of stools and management of constipation with dietary measures or laxatives. After the vicious circle of painful defecation and fecal retention is interrupted, healing of the fissure and cessation of hematochezia usually occurs between 4 weeks and 4 months. Topical glyceryl trinitrate ointment and calcium channel blocker diltiazem cure are only marginally better than placebo (Nelson et al. 2011). Fissures recalcitrant to conservative treatment may respond to intersphincteric botulinum toxin injections. Under general anesthesia, 15–25 IU of toxin is injected into four quadrants of the internal part of the sphincter complex. Healing occurs in several weeks. A recalcitrant fissure should be biopsied to rule out Crohn’s disease.
Hemorrhoids

True hemorrhoids are uncommon in children, excluding those with portal hypertension (Heaton et al. 1993). They may occur in conjunction with rectal vascular malformations. Clinically detectable internal hemorrhoids, with external extension, occur also in otherwise healthy children but are rare. They are usually asymptomatic, but the child or parents have noticed something protruding from the anus. A more common finding is a prominent venous plexus at the anal opening, which may be a source of rectal bleeding in constipated children. Hemorrhoidal venous plexus may rarely thrombose causing pain, as in adults with acute thrombosis of external hemorrhoids. Adult type hemorrhoids begin to occur in adolescents. In otherwise healthy children, asymptomatic hemorrhoids do not require surgical therapy. Major bleeding does not occur. Symptomatic patients with portal hypertension may require treatment. Banding or sclerotherapy controls the symptoms in the majority of cases.

Vascular Malformations

Vascular anomalies in the distal bowel are usually venous malformations, instead of hemangiomas as previously thought. Intestinal bleeding from vascular malformations is rare in children. The most common sites are the distal colon and rectum (de la Torre et al. 2002). In the minority of cases, the vascular lesion is a part of a systemic disease such as Klippel-Trenaunay (congenital varicose veins, cutaneous hemangiomas, and ectatic hypertrophy of the lower limbs) and Osler-Rendu-Weber (multiple telangiectasia) syndromes. A typical symptom of rectal vascular malformations is recurrent hematochezia that may sometimes be profuse. The patient may also present with hemorrhoids. The diagnosis of vascular anomalies may be difficult. Endoscopic examination may show unspecific local inflammation, and dilated or distorted vessels are rarely visible. Magnetic resonance imaging and angiography are the best imaging modalities to evaluate intestinal vascular lesions. Nonoperative methods may be used to control bleeding, but a permanent cure is best achieved by a complete resection of the lesion. Lesions that extend to the low rectum and anal canal are best treated by endorectal pull-through and coloanal anastomosis (Fishman et al. 2000). This sphincter-saving operation eradicates bleeding episodes for extended periods of times if not permanently.

Infantile Proctocolitis

Apart from anal fissures, the most common cause of hematochezia in infants younger than 3 months of age is eosinophilic proctocolitis. The infantile proctocolitis typically presents at the age of 3–4 weeks with fresh blood streaks mixed in mucous stools. Usually, there are no other symptoms, and the growth and development of the infant are normal. Colonoscopy shows colitis that is often patchy and rarely extends beyond left colon. Histology reveals marked eosinophilic infiltrate; some patients may have also elevated eosinophilic count in peripheral blood. An allergic etiology has been suggested because of these findings but is found in a minority of cases. The condition is self-limiting and symptoms usually subside within a few weeks. Dietary changes may be helpful in reversing the symptoms.

Perineal Groove, Skin Tags, and Polyps

The perineal groove is a mucosal-covered depression that runs in the midline from the anus to the vestibulum. It occurs exclusively in female infants and is often, but not necessarily, associated with an anterior location of the anus. Epithelialization occurs spontaneously during the first years of life. Perianal skin tags may be isolated or occur in conjunction with anal fissures or Crohn’s disease. As such, they are usually symptomless and should be left alone. Rectal polyps may protrude from the anal opening especially during defecation. Most of them are hamartomatous juvenile polyps and are easily excisable transanally or endoscopically. Colonoscopy is warranted to rule out polyposis.
### Anal Canal Duplication

Anal canal duplication is a rare, usually symptomless, congenital anomaly occurring almost exclusively in girls (Koga et al. 2010; Trecartin et al. 2019). They are one to several centimeters in length with an external opening just outside the sphincter muscles in the posterior midline. Communication with the anal canal is exceptional, and some may be associated with a presacral mass (teratoma, meningocele) or urinary tract malformations. Treatment involves complete excision using perineal or posterior sagittal approach depending on extension of duplication and associated presacral pathology.

### Conclusion and Future Directions

Besides anorectal malformations and Hirschsprung’s disease, pediatric anorectal disorders include a variety of pathologies. Although usually benign, their prompt recognition and adequate treatment is important, because they often cause significant discomfort and symptoms. Their pathophysiology and best treatment modalities continue to evolve with ongoing active research.

### Cross-References

- Anorectal Anomalies
- Crohn’s Disease
- Hirschsprung’s Disease
- Ulcerative Colitis

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