Functional defecation disorders in children

Novel insights into epidemiology, evaluation and management

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GENERAL INTRODUCTION AND OUTLINE OF THE THESIS

Parts of this introduction have been published as:

Childhood constipation: finally something is moving!
Ilan J.N. Koppen, Carlo Di Lorenzo, Miguel Saps, Phil G. Dinning, Desale Yacob, Marc A. Levitt, Marc A. Benninga

&

Chapter: Functional Constipation in Children
Ilan J.N. Koppen, Marc A. Benninga
PHYSIOLOGY OF DEFCATION

The dynamics of defecation are complex and rely on intricate processes involving the autonomic and somatic nervous system, pelvic floor muscles, and internal and external anal sphincters. After passing the stomach, duodenum, jejunum and ileum, chyme is transported from the terminal ileum into the caecum. Subsequently, luminal contents are moved distally through the colon while gradual desiccation and mixing occurs, resulting in progressively solid fecal material. The propulsion of feces is achieved by complex colonic motor patterns. Several different colonic motor patterns have been described, with the most well recognized being high-amplitude propagating contractions, which are associated with the mass movement of colonic contents and spontaneous defecation in healthy adults. Anterograde propagation of feces through the colon leads to filling of the rectum, which induces relaxation of the internal anal sphincter, allowing feces to travel further down the anal canal; this reflex is known as the recto-anal inhibitory reflex. Subsequently, sensory stimuli triggered by rectal distension and by the contact between fecal material and the mucosa of the proximal part of the anal canal result in an urge to defecate. At this point, voluntary contraction of the external anal sphincter and pelvic floor muscles can postpone defecation, by moving the fecal load higher up in the rectum, until place and time are appropriate for defecation. When defecation is initiated, voluntary relaxation of the external anal sphincter and the pelvic floor musculature (i.e. the puborectalis muscle and levator ani) allows for defecation to occur. During defecation, gentle increase of intra-abdominal pressure aids in expelling stools from the rectum.

FUNCTIONAL DEFCATION DISORDERS

Defecation disorders are common in children and account for a substantial amount of visits to general practitioners, pediatricians and pediatric gastroenterologists. In the absence of an underlying organic cause, these disorders are considered functional defecation disorders. Functional defecation disorders encompass two distinct entities: functional constipation (FC) and functional nonretentive fecal incontinence (FNRFI). Chapter 1 describes a systematic review on the worldwide prevalence of FC and FNRFI.

Functional Constipation (FC)

FC is characterized by infrequent evacuation of hard stools, which can be painful to evacuate. Moreover, FC is frequently accompanied by fecal incontinence and abdominal pain. These symptoms are known to have a significant impact on a child’s well-being and health-related quality of life.
**Pathophysiology**

The pathophysiology of FC remains incompletely understood. FC can occur at all ages, but the median age of onset of FC in children has been reported to be 2.3 years, around the time of toilet training.\(^\text{13}\) Aside from the high occurrence around the time of toilet training, FC may develop during infancy concomitant with changes in feeding, e.g. transitioning from breastfeeding to formula-feeding or at the introduction of solid foods.\(^\text{14}\) These findings suggest that both dietary and behavioral factors play an important role in the pathogenesis of FC. Furthermore, an association between functional defecation disorders and overweight has been suggested in the literature, but studies on this topic have reported conflicting results. In chapter 2, a cross-sectional population-based study on the association between overweight and FC is presented, while chapter 3 systematically reviews the literature on this theme.

One important etiologic factor of FC is stool withholding behavior. This often occurs after a negative experience such as a hard, painful or frightening bowel movement.\(^\text{15}\) Stool withholding behavior can lead to the accumulation of a large fecal mass in the rectum that is difficult to evacuate, this is known as fecal impaction. Fecal impaction may lead to overflow fecal incontinence which is the involuntary loss of soft stools that leak around the solid, obstructing, fecal mass. This bothersome symptom has been reported to be present in 75-90% of children presenting with FC.\(^\text{16}\)

**Functional Nonretentive Fecal Incontinence (FNRFI)**

Fecal incontinence is defined as the loss of stools in places inappropriate to the social context at least once per month in children with a developmental age of ≥ 4 years.\(^\text{17}\) In the vast majority of children with functional fecal incontinence, this symptom is associated with FC.\(^\text{18,19}\) However, in the remaining subset of children, no signs of fecal retention are present; these children suffer from a disorder classified as functional nonretentive fecal incontinence (FNRFI).\(^\text{17,19}\)

**Pathophysiology**

The pathophysiology of FNRFI is incompletely understood and its etiology is considered to be multifactorial.\(^\text{20-25}\) Interestingly, urinary incontinence is common in children with FNRFI.\(^\text{26-28}\) Prevalence rates of daytime and nighttime urinary incontinence in FNRFI patients range between 14-50% and 20-47% respectively.\(^\text{19,26,29}\) Vice versa, 11% of children with dysfunctional voiding, urge incontinence or bladder overactivity have FNRFI.\(^\text{27}\) It has been hypothesized that the co-occurrence of urinary and fecal incontinence in otherwise healthy children without signs of fecal retention may reflect a combined, possibly neurodevelopmental or behavioral elimination disorder.\(^\text{30,31}\)
Functional defecation disorders and all other functional gastrointestinal disorders are diagnosed according to the symptom-based Rome criteria, which have been developed by expert clinicians through literature review and a consensus process. When the Rome criteria were first established in 1990, these criteria only applied to adults. With the introduction of the Rome II criteria in 1999, specific criteria were also established for functional gastrointestinal disorders in children. The Rome II criteria were revised in 2006, culminating in the Rome III criteria which made a distinction between different pediatric age groups; infants/toddlers and children/adolescents. More recently, in 2016, the revised Rome IV criteria have been published, the Rome IV criteria for FC and FNRFI are shown in tables 1 & 2.

### TABLE 1. Rome IV criteria for functional constipation for infants/toddlers and children/adolescents.

<table>
<thead>
<tr>
<th>INFANTS/TODDLERS</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>G7. Diagnostic Criteria for Functional Constipation</strong></td>
</tr>
<tr>
<td>Must include 1 month of at least 2 of the following in infants up to 4 years of age:</td>
</tr>
<tr>
<td>1. 2 or fewer defecations per week</td>
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<tr>
<td>2. History of excessive stool retention</td>
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<tr>
<td>3. History of painful or hard bowel movements</td>
</tr>
<tr>
<td>4. History of large-diameter stools</td>
</tr>
<tr>
<td>5. Presence of a large fecal mass in the rectum</td>
</tr>
<tr>
<td>In toilet-trained children, the following additional criteria may be used:</td>
</tr>
<tr>
<td>6. At least 1 episode/week of incontinence after the acquisition of toileting skills</td>
</tr>
<tr>
<td>7. History of large-diameter stools that may obstruct the toilet</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>CHILDREN/ADOLESCENTS</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>H3a. Diagnostic Criteria for Functional Constipation</strong></td>
</tr>
<tr>
<td>Must include 2 or more of the following occurring at least once per week for a minimum of 1 month with insufficient criteria for a diagnosis of irritable bowel syndrome:</td>
</tr>
<tr>
<td>1. 2 or fewer defecations in the toilet per week in a child of a developmental age of at least 4 years</td>
</tr>
<tr>
<td>2. At least 1 episode of fecal incontinence per week</td>
</tr>
<tr>
<td>3. History of retentive posturing or excessive volitional stool retention</td>
</tr>
<tr>
<td>4. History of painful or hard bowel movements</td>
</tr>
<tr>
<td>5. Presence of a large fecal mass in the rectum</td>
</tr>
<tr>
<td>6. History of large diameter stools that can obstruct the toilet</td>
</tr>
<tr>
<td>After appropriate evaluation, the symptoms cannot be fully explained by another medical condition.</td>
</tr>
</tbody>
</table>

### TABLE 2. Rome IV criteria for functional nonretentive fecal incontinence for children/adolescents.

<table>
<thead>
<tr>
<th>H3b. Diagnostic Criteria for Nonretentive Fecal Incontinence</th>
</tr>
</thead>
<tbody>
<tr>
<td>At least a 1-month history of the following symptoms in a child with a developmental age older than 4 years:</td>
</tr>
<tr>
<td>1. Defecation into places inappropriate to the sociocultural context</td>
</tr>
<tr>
<td>2. No evidence of fecal retention</td>
</tr>
<tr>
<td>3. After appropriate medical evaluation, the fecal incontinence cannot be explained by another medical condition</td>
</tr>
</tbody>
</table>
CLINICAL EVALUATION

Because functional defecation disorders are clinical diagnoses, evaluation is primarily based on a detailed medical history and a thorough physical examination. Additional diagnostic tests are usually not necessary unless an organic cause is suspected.

History

Important information from the medical history includes details on defecation frequency, fecal incontinence episodes, stool form and consistency, withholding behavior, and pain during defecation. In order to help describe stool consistency, the Bristol Stool Form Scale (BSFS) can be used (Figure 1). Although the BSFS is commonly used in children, its reliability for the use in young children who are not toilet trained and wear diapers has been debated. The aim of chapter 5 was to evaluate the agreement between verbal report and the BSFS in the assessment of stool consistency among parents of infants and toddlers. In chapter 6, a new visual stool form scale for infants and toddlers wearing diapers is introduced; the Brussels Infant and Toddler Stool Scale.

When evaluating symptoms, alarm symptoms suggestive for an organic cause should be sought out. They include delayed passage of meconium, a history of bloody stools without the presence of a fissure, failure to thrive and severe abdominal distension. Furthermore, if parents report smearing of feces, this should raise the suspicion of sexual abuse.

Physical examination

Physical examination should be performed in all children with defecation disorders to establish the diagnosis and to detect potential alarm signs. Assessment of weight and height is of key importance since failure to thrive can be indicative of an organic cause of constipation. Physical examination primarily consists of examination of the abdomen, the perianal region and the lumbosacral region. Abdominal examination mainly focuses on the detection of a palpable fecal mass or scybala. Perianal inspection should be performed in all children; the physician should look for anatomic abnormalities, perianal feces, fissures, scars and erythema. Fissures can be a sign of hard or large stools, but can also be a sign of sexual abuse. Hematomas in the perianal region are also highly suspicious of abuse. Special attention should be paid to abnormal behavior during physical examination (e.g., sexual acting out, extreme fear). In children who already fulfil 2 or more of the Rome criteria, a digital rectal examination is not necessary for the diagnosis of FC. However, the procedure does provide valuable information, e.g. by enabling detection of a rectal fecal mass and assessment of anorectal sensation and sphincter tone. Moreover, in a child with functional fecal incontinence in whom the diagnosis is uncertain, a digital rectal examination may help in differentiating between FC and FNRFI.
Psychological evaluation

Between 30-50% of children with fecal incontinence have been reported to have a comorbid emotional or behavioral disorder. Because of this high psychologic comorbidity rate, clinicians are encouraged to observe, explore and enquire about emotional and behavioral problems and to refer children for psychological evaluation if necessary. Children with fecal incontinence have been reported to display significantly increased rates of separation anxiety, specific phobias, generalized anxiety, attention deficit hyperactivity disorder (ADHD) and oppositional defiant disorders. In chapter 4, the association between ADHD and functional defecation disorders is evaluated.
DIAGNOSTIC TESTS

The utilization of diagnostic tests is seldom indicated in the workup of children with functional defecation disorders. However, in selected cases diagnostic testing can be useful, e.g. if an organic cause is suspected or to help differentiate between FC and FNRFI if the diagnosis is uncertain.

Laboratory testing

Laboratory testing (e.g. for hypothyroidism, celiac disease or hypercalcemia) in children with defecation disorders is only indicated when there is a suspicion for an underlying organic disease, it does not belong in the routine workup of children with functional defecation disorders.

Radiology

Abdominal radiography

A plain abdominal X-ray is not an appropriate tool to diagnose constipation. The sensitivity and specificity rates are unsatisfactory, and low inter- and intra-observer reliability have been reported for the different scoring systems (Barr, Leech, Blethyn) that are used to evaluate fecal load based on abdominal X-rays.33,42,43

Colonic transit time

In the diagnostic workup of children with functional defecation disorders, measurement of the colonic transit time (CTT) is sometimes helpful, especially in children with severe and persisting symptoms or in cases where the diagnosis is unclear. Currently, the most widely used method to determine CTT in children is the radiopaque marker test, which is cost effective and simple to perform.44 Radiopaque markers are ingested orally and the amount of intra-abdominal markers is then determined using an abdominal X-ray.16,45,46 A CTT <62 hours is usually considered to be normal, while a CTT >62 hours suggests slow transit constipation.46 An extremely prolonged CTT of more than 100 hours indicates a severe form of constipation.46 Another method to determine CTT is radionuclide scintigraphy; after ingestion of radioactive isotopes, colonic transit is measured with a large-field-view gamma camera. Scintigraphy is a more novel technique than the radiopaque marker test and its use in children is less widespread.48-50 In children with functional fecal incontinence without signs of constipation in whom the diagnosis is unclear, a colonic transit study can be useful to discriminate between FC and FNRFI; in these children a normal CTT suggests FNRFI.33,39
Contrast enema
Contrast enemas are used to identify anatomic abnormalities of the anorectum. After infusion of contrast fluid into the rectum an abdominal X-ray is obtained, visualizing the distribution of contrast fluid in the distal gastrointestinal tract. Contrast enemas can be useful to detect mechanical causes of constipation such as anatomical abnormalities or complications after colorectal surgery.51

Ultrasonography
Transabdominal ultrasonography has been used to measure the transverse rectal diameter.52,53 An increased rectal diameter (>30 mm) is often considered to be suggestive for fecal impaction.54,55 Although transabdominal ultrasonography is a promising technique, there is currently insufficient evidence that the transverse diameter can be used as a reliable predictor of constipation and fecal impaction in children.33,56

Manometry
Manometry assesses intraluminal pressure and contact force in the gastrointestinal tract, thereby providing insights into contractile motor patterns of the gut.

Anorectal manometry
Anorectal manometry provides information about anorectal neuromuscular function. This test allows assessment of the rectoanal inhibitory reflex, anal sphincter pressure, rectal sensation, and defecation dynamics. The rectoanal inhibitory reflex is a physiological reflex that induces the internal anal sphincter to relax in response to rectal distension, thereby enabling defecation to take place. To assess the rectoanal inhibitory reflex during anorectal manometry, rectal distension is generated by inflating a balloon attached to the end of the manometry catheter. Since anorectal manometry can be used to assess the rectoanal inhibitory reflex, it is used as a tool to rule out Hirschsprung's disease.33 Hirschsprung's disease is the most common congenital gut motility disorder. It is characterized by the absence of enteric ganglions (aganglionosis) in the distal gut. Due to this aganglionosis the physiologic rectoanal inhibitory reflex is absent. The performance and analysis of anorectal manometry data requires considerable experience and therefore the procedure is primarily performed in specialized centers.

Colonic manometry
Colonic manometry is used as a tool to identify abnormalities in colonic contractile activity. Although protocols differ between centers, colonic manometry studies generally include recording periods in a fasted state, after ingestion of a meal and, particularly in children, after administration of a stimulant laxative.57
Most published pediatric colonic manometry studies have been performed with low-resolution colonic manometry catheters with 8 recording sites (with sensors being spaced 10-15 cm apart). These studies tended to focus on the presence or absence of high-amplitude propagating contractions.\textsuperscript{58–61} Since these motor patterns are associated with spontaneous defecation, their presence, occurring either spontaneous or in response to chemical stimuli, is considered to be an important marker for colonic neuromuscular integrity.\textsuperscript{4,5,62} However, with the development of high-resolution colonic manometry catheters, more detailed information on colonic motor patterns has become available and it has become clear that the colon exhibits more refined colonic motor patterns as well.\textsuperscript{3} In adult studies utilizing high-resolution fiber-optic manometry, several other motor patterns have been identified prior to and after a meal, the most prominent of which is a distal colonic cyclic motor pattern (2-6 cycles per minute), that increases in prevalence after a meal and propagates in a predominantly retrograde direction.\textsuperscript{3} The slow wave frequency of this motor pattern suggests that it is being generated by the pacemaker cells (interstitial cells of Cajal) within the submucosal plexus while its increase after a meal suggests that it can be influenced by extrinsic neural innervation. In adult patients with slow transit constipation an increase in this cyclic activity was not observed after a meal, potentially indicating an abnormality in the extrinsic neural innervation of the colon.\textsuperscript{63} Alternatively a complete absence of cyclic propagating activity may indicate abnormalities in the intrinsic neural pathways. \textbf{Chapters 7 and 8} describe findings from high-resolution colonic manometry studies in children with intractable FC. \textbf{In chapter 9}, results from contrast enemas and colonic manometry in children with FC are compared.

\section*{MEDICAL MANAGEMENT}

Conventional medical management of functional defecation disorders in children consists of nonpharmacological and pharmacological interventions. Nonpharmacological treatment recommendations for FC and FNRFI are similar, while the pharmacological treatments for both disorders are in contrast with one another. In 2014, the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition (ESPGHAN) and the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN) published a guideline for the evaluation and management of childhood FC.\textsuperscript{33} In \textbf{chapter 10}, we assess the awareness and implementation of this guideline among physicians from the United States and the Netherlands.
Nonpharmacological management

**Education**

Education is the first step in the non-pharmacological treatment of FC and FNRFI. This should include an explanation of physiological defecation dynamics, tailored to the developmental age of the child. In children with FC, the negative chain of events that may have been prompted by a painful defecation experience should be explained to parents and, if possible, children. The concept of overflow incontinence and the pivotal role that withholding behavior plays in the pathophysiology need to be explained.

**Toilet training program**

In order to prevent the occurrence of fecal impaction and to decrease the risk of fecal incontinence episodes, it is important to evacuate stools regularly. In children with a developmental age of ≥4 years, this can be established by introducing a toilet training program, with scheduled toilet sit moments throughout the day, usually after most meals and after coming home from school. The toilet sit moments are scheduled after a meal to benefit from the gastrocolic reflex which increases colonic peristalsis and tone in response to ingestion of a meal. To motivate children to maintain this toilet training program, a reward system can be introduced. By rewarding the child for completing toilet sit moments, the child is positively reinforced to comply with therapy. A non-accusatory approach of both physicians and parents is of key importance since children may feel guilty or embarrassed, especially about episodes of fecal incontinence. Only rewarding periods without fecal incontinence is therefore not recommended, as this may increase feelings of guilt and can be experienced as punishment for having fecal incontinence.

**Lifestyle interventions**

Insufficient fiber intake is associated with FC and a normal fiber intake should therefore always be recommended. However, there is insufficient evidence to support the use of supplementary dietary fiber in excess of the daily recommended intake. Epidemiological studies have suggested an association between a low fluid intake and FC, but there is insufficient evidence for a favorable role of fluid intake exceeding normal recommended amounts in the treatment of FC. Therefore, a normal fluid intake based on the age of the child is recommended in children with FC. Although higher levels of physical activity may be associated with a decreased risk of developing FC, no studies have been performed to assess the effect of increasing physical activity to treat FC in children.
Pre-, pro- and synbiotics
Prebiotics are nondigestible compounds that induce the growth or activity of microorganisms and probiotics are live bacteria or fungi that confer a health benefit for the host. Synbiotics are defined as products that contain both pre- and probiotics. Pre-, pro- and synbiotics have been suggested as potential treatment modalities for FC in children. In chapter 12, we systematically review the literature to investigate the efficacy and safety of pre-, pro- and synbiotics in the treatment of childhood FC.

Pharmacological treatment of functional constipation
The pharmacological treatment of FC consists of treatment with laxatives and involves three steps; disimpaction, maintenance treatment and weaning.

Disimpaction, maintenance treatment and weaning
Fecal impaction occurs in approximately 50% of children with FC. The fecal mass needs to be evacuated prior to initiating maintenance treatment in order to increase treatment success. Disimpaction can be achieved with enemas or oral intake of high-dose oral polyethylene glycol (PEG) (1-1.5g/kg/day) during 3-6 days. High-dose PEG and sodium docusate enemas have been found to be equally effective for disimpaction and although high-dose PEG is associated with a higher risk of fecal incontinence, PEG is recommended as first choice for disimpaction because it is administered orally.

After successful disimpaction, maintenance therapy with osmotic laxatives should be initiated to prevent the re-accumulation of feces. Osmotic laxatives are poorly absorbed by the intestinal wall, causing osmotic water retention in the intestinal lumen. This softens the stools and increases peristalsis through intestinal distension, facilitating easy and frequent bowel movements. PEG is the first choice osmotic laxative in children with FC based on its effectiveness and safety profile. Chapter 11 describes a cross-sectional survey study on therapy adherence in children with FC treated with PEG.

Maintenance treatment should be gradually weaned rather than abruptly discontinued in order to prevent a relapse. Weaning can be considered when symptoms are stable for at least 1 month under maintenance treatment, which means that children have a defecation frequency of ≥3 times per week and do not fulfill any other Rome criteria. It is recommended to evaluate symptoms again two months after cessation of treatment, to prevent or detect relapses.
Pharmacological treatment of functional nonretentive fecal incontinence

Unlike the pharmacological treatment of FC, the use of oral laxatives in children with FNRFI is contraindicated. Oral laxatives may even increase the fecal incontinence frequency by making the stools too soft to retain. Anecdotal evidence suggests that loperamide may have a beneficial role in the treatment of FNRFI. As an opiate receptor agonist, loperamide decreases peristalsis and increases the internal anal sphincter tone. Moreover, loperamide may improve sphincter function and thereby prevents involuntary loss of stools. When prescribing loperamide, it is important to carefully monitor patients to prevent the development of constipation. Based on scarce evidence, the antidepressant imipramine may also have a beneficial role in treating FNRFI. Imipramine has an anticholinergic action, its mechanism of action may be similar to that of loperamide decreasing motility and increasing sphincter tone. However, due to cardiovascular side effects and the risk of suicidal ideation, tricyclic antidepressants should be prescribed only under close clinical supervision.

Transanal irrigation

Transanal irrigation is an advanced medical treatment option designed to assist in evacuating stools from the bowel, it can be used in both FC and FNRFI if (non)pharmacological treatment options have failed. During transanal irrigation, water is infused into the colon through a rectally inserted irrigation device to mechanically wash out feces. If timed well and adequately performed, these washouts can induce regular emptying of the colon resulting in a continent state. Although study samples have been small, this form of colonic irrigation has been shown to be effective in the management of fecal incontinence due to constipation in children with neurogenic defecation disorders (e.g. spina bifida or Hirschsprung’s disease), anorectal malformations, FC and FNRFI. A study on treatment efficacy and parental satisfaction in children with intractable FC treated with transanal irrigation is presented in chapter 13.

SURGICAL MANAGEMENT

In severe cases of FC, when pharmacological treatments have failed, surgery is sometimes considered as a treatment of last resort. Currently, there are no evidence-based guidelines for the surgical management of FC and approaches vary between centers. In chapter 14, we assess the diagnostic and surgical approach of pediatric surgeons and pediatric gastroenterologists towards children with intractable FC.
Antegrade continence enemas
Antegrade continence enemas (ACE) can be achieved by surgically creating an external entrance into the intestinal lumen, usually the cecum, to enable flushing fluids into the colon. There are different ways to establish this. A common surgical procedure to achieve ACE is the Malone appendicostomy. This procedure involves connecting the appendix to the abdominal wall and creating a valve, allowing catheterization for fluid administration and at the same time preventing leakage of stools. Another technique to achieve ACE is via percutaneous cecostomy, a minimally invasive procedure that involves the percutaneous introduction of a cecostomy tube. Outcomes of ACE are estimated to be favorable in 82% of cases, although complete resolution of constipation and fecal incontinence with reversal of the ACE stoma is achieved only in 9.5% of cases.90

Sacral nerve stimulation (SNS)
Sacral nerve stimulation (SNS) aims at stimulating the anterior ramus of sacral spinal nerves S3 and S4 via surgically positioned electrodes connected to a pulse generator that is implanted subcutaneously in the lateral buttock and can be switched on and off at will. The implantation of the device is often preceded by a period of peripheral nerve evaluation, which involves the placement of a temporary lead in an attempt to predict the outcome of SNS. Although the exact mechanism of SNS is incompletely understood, promising results have been published in adults and children with FC.91–96 However, study designs, outcome measures and follow up durations vary widely between studies and sample sizes are generally small.91,97 Larger randomized-controlled studies with long-term follow-up are required to gain more insights into the efficacy of SNS in the treatment of childhood FC. In chapter 15, we evaluate long-term efficacy of SNS in children with constipation and describe patient benefit and parent satisfaction.

Ostomies and resections
In severe cases of intractable FC, surgery may prove to be beneficial.90 Besides operations to achieve ACE, which are discussed above, surgical management of FC may involve botox injections, bowel resection or formation of a temporary or permanent diverting stoma.98 In children with a megarectum or megasigmoid, which can be demonstrated by contrast enemas, a rectosigmoid resection may alleviate symptoms.98 Surgery is generally performed in a step-up approach, beginning with the least invasive procedure and proceeding to more invasive treatment modalities only after failure of the previous step. In chapter 16, outcomes of surgical management in children with intractable FC at a single tertiary children’s hospital are presented.
PROGNOSIS

Despite pharmacological interventions approximately 40% of children with FC referred to a pediatric gastroenterologist still has symptoms after 5 years and even after 10 years 20% of children suffers from FC symptoms. In these patients, symptoms may persist into adolescence or even adulthood despite laxative treatment. This underlines that there is still a need for better and more effective treatments of childhood FC.

FNRFI often is a long-lasting problem and treatment can prove to be quite challenging. After 2 years of intensive treatment in a tertiary center approximately 30% of FNRFI patients are cured. Most recover before they reach adulthood; nevertheless at 18 years of age 15% will still struggle with fecal incontinence. Regular follow-up visits are recommended to motivate children and their parents to adhere to treatment and to prevent relapses.
REFERENCES

General introduction and outline of the thesis


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