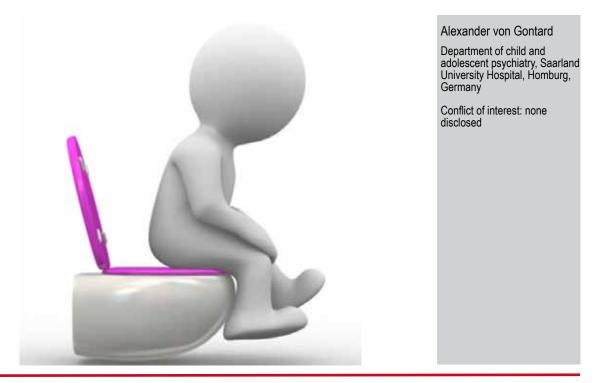
DEVELOPMENTAL DISORDERS

ENCOPRESIS

Chap

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Two major forms of encopresis can be differentiated: encopresis *with* and *without* constipation. The specific differentiation into these two subtypes is of utmost importance as they differ according to pathophysiology, clinical features and, especially, treatment (i.e., the former does not respond to laxatives, while in the latter they are essential in treatment). The aim of this chapter is to give an overview and practical approaches to assessment and treatment of these two subtypes of encopresis.

DEFINITION AND CLASSIFICATION

According to ICD-10 (World Health Organization, WHO, 2008) and DSM-IV-TR (American Psychiatric Association, APA, 2000), encopresis is defined as both voluntary and involuntary passage of feces in inappropriate places in a child aged four years or older, after organic causes have been ruled out. Despite major similarities between the two systems, they differ regarding essential points, especially the definition of subtypes. In ICD-10, one episode per month for at least three months is needed. ICD-10 does not define subtypes nor differentiate clearly between encopresis with and without constipation. The duration required for diagnosis in DSM-IV-TR (2000) is the same but it distinguishes between encopresis with and without constipation.

Table C.5.1Rome-III diagnostic criteria for functional constipationand non-retentive stool incontinence (Rasquin et al, 2006)

Functional constipation*

Must include two or more of the following in a child with a developmental age of at least four years with insufficient criteria for diagnosis of irritable bowel syndrome:

- Two or fewer defecations in the toilet per week
- At least one episode of fecal incontinence per week
- History of retentive posturing or excessive volitional stool retention
- · History of painful and hard bowel movements
- Presence of a large fecal mass in the rectum
- History of large diameter stools that may obstruct the toilet.
- * Criteria must be fulfilled at least once per week for at least two months

Non-retentive fecal incontinence*

Must include all of the following in a child with a developmental age at least four years:

- Defecation into places inappropriate to the social context at least once per month
- No evidence of an inflammatory, anatomic, metabolic, or neoplastic process that explains the subjects symptoms
- No evidence of fecal retention
- * Criteria must be fulfilled for at least two months

Based on current research and validation studies, the most precise taxonomy was proposed by pediatric gastroenterologists in their Rome-III classification for functional disorders of the gastrointestinal tract (Table C.5.1) (Rasquin et al, 2006). The first difference is that the neutral term of *fecal incontinence* was chosen instead of encopresis. This means, that fecal incontinence and encopresis can be used as synonyms (as in this chapter). The second change is that constipation was defined as the main disorder that can - but does not necessarily have to - be associated with soiling. Constipation is, in fact, more common than encopresis: many constipated children do not soil. The main symptoms of functional constipation can be seen in Table C.5.1. The definition of constipation cannot be based on a low defecation frequency on the toilet alone: some children pass stools every day but still retain fecal masses in the rectum and colon. Therefore, additional clinical signs and symptoms are needed and these are listed explicitly in the Rome-III criteria. The authors chose a shorter duration - two months - to encourage early treatment and to avoid chronification. Two symptoms must be present: a low defecation frequency; soiling; holding maneuvers; painful, hardened stool; and large fecal masses.

For encopresis without constipation, the neutral term of *non-retentive fecal incontinence* was suggested. The etiology of this subtype is not known and much less research has been generated for this disorder than for constipation (Bongers et al, 2007). Except for a shorter duration of two months, the criteria are similar to those of DSM-IV.

EPIDEMIOLOGY

Encopresis is a common disorder affecting 1% to 3% of children older than four years (the definitional age). Three different trajectories can be defined (Heron et al, 2008):

- Children with chronic encopresis over many years
- Those with relapses, and
- A group with a tendency to remit spontaneously.

In a Dutch study, 4.1 % of 5-6 year olds and 1.6 % of 11-12 year olds were affected (van der Wal et al., 2005). Prevalence depends on the definition used; for example, 5.4 % of 7-year old children soiled, but only 1.4 % once or more per week (Joinson et al, 2006). Encopresis can persist into adolescence and even young adulthood. Without constipation (i.e., non-retentive fecal incontinence), 49% of children soiled at the age of 12 years, and 15% at 18 years in a long-term follow up (Bongers et al, 2007). The prognosis of constipation is less favorable: only 80 % had a good outcome by the age of 16 years, and 75% to 80% at 16-27 years (Bongers et al, 2010).

Three to four times more boys are affected by encopresis than girls (Bellman, 1966). Encopresis occurs almost always during the day (Bellman, 1966). Nocturnal encopresis is more often associated with organic causes and requires a more detailed somatic assessment.

Most children from the age of four years onwards have one bowel movement a day – with great individual variability (Bloom et al, 1993). Chronic constipation is a more common disorder than encopresis. According to a large meta-analysis, the median prevalence was 9% world-wide with different definitions used (van den Berg et al, 2006). This meta-analysis included studies from countries such as Italy, US, Hong Kong, Japan, Finland, Turkey, Brazil, Saudi Arabia, among others.

CLINICAL SIGNS AND SYMPTOMS

As the differentiation of the two subtypes – constipation and non-retentive fecal incontinence – is decisive for treatment, it is important to know the basic signs and symptoms of each. Based on the research of Benninga et al (1994; 2004), as well as others, the main differences between the subtypes are shown in Table C.5.2.

Children with encopresis with constipation (or functional constipation) have a reduced number of bowel movements with large stools of altered consistency (too soft or too hard). They often experience pain during defecation. Abdominal pain and reduced appetite are typical. The colon transit time is increased, abdominal and rectal masses are palpable. In sonography, the rectal diameter is increased (> 25 mm). Often, daytime urinary incontinence and even enuresis co-exists. Additional emotional and behavioral disorders are found in 30%-50% of them and, finally, laxative therapy is helpful.

Children with encopresis without constipation (non-retentive fecal incontinence) do not have many of these symptoms. They have daily bowel movements of normal size and consistency. Pain is not frequent and appetite is

	Encopresis with constipation	Non-retentive fecal incontinence
Bowel movements	Seldom	Daily
Large amounts of stools	Yes	No
Normal stools (consistency)	One half	Nearly all
Pain during defecation	One half	Seldom
Abdominal pain	Often	Seldom
Appetite	Reduced	Good
Colon transit time	Long	Normal
Palpable abdominal mass	Often	None
Palpable rectal mass	Often	Never
Rectal diameter (sonography)	Increased	Normal
Daytime urinary incontinence	One tenth	Seldom
Nocturnal enuresis	One third	Tenth
Comorbidity with behavioral and emotional disorders	30%-50%	30%-50%
Laxative therapy	Helpful	Not helpful, even worsening

Table C.5.2 Differences between encopresis with constipation and non-retentive fecal incontinence

good. Colon transit time is normal and no stool masses can be palpated. Enuresis and urinary incontinence are less common, while comorbidity with psychological disturbances is similar (30%-50%). Finally, laxatives have no effect – can even worsen the soiling.

ETIOLOGY

Fecal continence is achieved following night-time and daytime urinary continence (Largo et al, 1978; 1996). Early toilet training can lead to early fecal continence in infancy and toddlerhood but has no influence on the development of encopresis at the age of 4 years. Encopresis can be preceded by delayed development and difficult temperament in the child, as well as maternal depression and anxiety (Joinson et al, 2008).

Functional constipation

Genetic factors do play a role in constipation but less so in encopresis. The concordance rate for constipation for monozygotic twins is 70%, for dizygotic twins 18%. The risk of occurrence is 26% if one parent is affected, 46% if both parents, 40% if father, and 19% if mother is affected (Bakwin & Davidson, 1971).

Functional constipation often develops after an acute constipation, which affects 16%-22% of young children. This can be triggered by a wide variety of psychological factors such as stressful life events, losses, family conflict, as well as somatic factors such as anal fissures, changes of diet, intensive toilet training and medication (Cox et al, 1998). Irrespective of the initial triggers, a sequence of chronic stool retention can follow. Acute constipation causes painful defecation and avoidance of defecation, inducing a paradoxical contraction of the external anal sphincter, which becomes habitual. Avoidance of bowel movements leads to accumulation of fecal material in the rectum and colon. Transit times are increased, peristalsis and sensation are decreased. Fluid withdrawal leads to large, hard fecal masses and a megacolon. Encopresis occurs because of interference of these masses with rectal function and by fresh stools bypassing the hard fecal masses.

Non-retentive fecal incontinence

In contrast to the clear model of functional constipation, the etiology of non-retentive fecal incontinence is not known (Bongers et al, 2007). It cannot be caused by psychological factors alone, as the comorbidity rate of both types of encopresis is the similar: 30%-50% of cases (Benninga et al, 1994; 2004; von Gontard et al, 2011).

COMORBIDITY

Overall, 30-50% of all children with encopresis have a comorbid emotional or behavioral disorder. This means, that three to five times more children with encopresis have additional disturbances in comparison to non-soiling children.

In a large population-based study of children aged 7½ years, children with encopresis had significantly increased rates of separation anxiety (4.3%), specific phobias (4.3%), generalized anxiety (3.4%), ADHD (9.2%) and oppositional defiant disorder (11.9%) (Joinson et al, 2006). In other words, children with fecal incontinence show a heterogeneous pattern of both internalizing and externalizing disorder comorbidity. Regarding subtype, encopretic children with constipation

Table	e C.5.3 Standard and extended assessment of encopresis
Star	ndard assessment (sufficient for most cases)
• •	History Questionnaires and scales Physical examination Sonography (not in all centers) Screening for behavioral disorders or full child psychiatric assessment
Exte	ended assessment (only if indicated)
•	Stool bacteriology Radiology: - Plain abdominal x-ray - Colon contrast X-ray - MRI of colon Manometry Endoscopy and biopsy

have similar rates of behavioral scores in the clinical range as children without constipation (Benninga et al, 1994; 2004). In other words, they cannot be differentiated according to the behavioral comorbidity.

DIAGNOSIS

The assessment of children with encopresis should be as non-invasive as possible, and should always include parents or other caregivers. For most children, a basic evaluation that can be conducted in many primary care settings is sufficient (Table C.5.3).

The history is the most important aspect of assessment. If conducted correctly and empathically, the most relevant information will be gathered through the history. It is worthwhile to take enough time during the initial consultation. Useful questions for a detailed history can be found in Appendix C.5.1.

Questionnaires can be a useful and time-saving procedure to gain information and to check if the information obtained through history is complete. A short questionnaire is shown in Appendix C.5.2.

A very useful chart is the Bristol Stool Chart. Seven types of stool forms are depicted ranging from "separate hard lumps, like nuts (hard to pass)" (type 1) to "watery, no solid pieces, entirely liquid" (type 7). The scale enables parents and children to identify the predominant type of stool easily and without lengthy descriptions. The course of treatment can also be monitored using this scale.

Each child should have a physical examination. Both a general pediatric and a neurological examination are recommended. The perianal and perigenital areas should be inspected. Spine, reflex differences and asymmetries of buttocks should be noted. A rectal examination should be performed at least once. If sonography is available, this can replace the rectal exam if no organic form or fecal incontinence is suspected. If available, sonography of abdomen, kidneys, bladder and retrovesical region is helpful. The most important finding is an enlarged rectal diameter of >25-30 mm in children with constipation (Joensson et al, 2008). In these cases, a rectal examination can be avoided. If sonography is not possible, a standard rectal exam should be performed and rectal masses can be palpated.

Due to the high comorbidity rate, a child psychiatric assessment is recommended in child psychiatric settings. In other settings, screening with validated questionnaires (such as the Child Behavior Checklist; Achenbach, 1991) is recommended. If scores are in the clinical range, further assessment or child psychiatric referral is recommended (von Gontard et al, 2011).

All other examinations are not routinely indicated – only if an organic type of fecal incontinence is suspected. It is important to avoid unnecessary and invasive investigations. Further details regarding assessment can be found in von Gontard and Neveus (2006).

Differential diagnosis

Somatic causes are present in 5% of children with chronic constipation and must be ruled out. These include anatomic causes such as anal fissures, abscesses, skin tags, dermatitis, anal stenosis and other ano-rectal malformations. Metabolic and endocrine causes include cystic fibrosis, celiac disease, cow milk intolerance/ allergy, diabetes mellitus and hypothyroidism. Neurological causes include cerebral palsy, spina bifida and myelomeningocele. Constipation can also be caused by various drugs (von Gontard and Neveus, 2006).

The most important differential diagnosis is Hirschsprung's disease, which affects 1:5000 infants and is an intrinsic neuromuscular disease of the gastrointestinal tract characterized by aganglionosis. Symptoms begin in the neonatal period with vomiting, abdominal distension, and food refusal. Older children can have small caliber stools, while soiling is a rare symptom. Most cases (80%) are diagnosed by the age of 4 years (Felt et al, 1999).

Organic causes in non-retentive fecal incontinence are much less frequent affecting a maximum of 1% of children. The most important are infectious diarrhea, neurological conditions such as *spina bifida occulta* and postoperative residual symptoms.

TREATMENT

Following assessment, children and parents are given detailed information on the subtype of encopresis (psychoeducation). As in the treatment of enuresis, unspecific factors such as enhancing motivation and building a good therapeutic relationship are helpful. Feelings of guilt, dysfunctional parental attributions ("my child is doing this on purpose") and frustration can be verbalized. Ineffective parental interventions such as punishment or non-indicated medication can be discussed with parents. If the child's food intake is restricted to low fiber foods, a change in the child's diet can be useful. Also, the amount of fluids should be increased, as many children do not drink enough during the day.

Toilet training

A baseline period with observation and recording is not necessary. Instead, toilet training is initiated right from the start. This training is indicated for both

types of encopresis. Children are asked to sit on the toilet three times a day, after mealtimes. This time is especially useful as the postprandial defecation reflexes are then most active. Children are asked to sit on the toilet five to ten minutes in a relaxed way, for this, it is important that their feet touch the floor. Otherwise a little foot-stool should be provided. These toilet sessions should be experienced in a positive way: children are allowed to read comics, books, play with computers or cell phones, draw pictures, etc. They do are not expected to pass urine or stools every time. These toilet sessions are documented in a chart as depicted in Appendix C.5.3. If necessary, the co-operation of the child can be enhanced positively by a simple token system with small rewards. All criticism or punishment should be avoided.

Laxatives

In *non-retentive fecal incontinence*, this toilet training is the main aspect of treatment. Laxatives are not indicated. In children with *constipation*, toilet training is combined with laxatives: first *disimpaction*, then maintenance treatment.

Disimpaction is necessary to evacuate fecal masses at the beginning of treatment. This can be performed rectally or orally. In rectal disimpaction, enemas are applied. The most widely used and recommended are enemas containing phosphates such as sodium hydrogen phosphate and sodium monohydrogen phosphate. Recommended doses are 30ml/10kg of bodyweight or half an enema for pre-school children, ¾ to one enema in school children. Often, these have to be repeated several times. An alternative is oral disimpaction with polyethylenglykol (PEG; macrogol). Large doses of up to 1.5g/kg body weight per day are given. Sufficient oral fluids are required for this osmotic laxative to be effective.

Once large stool masses have been passed, a lower maintenance dose should be given. Through these procedures success is achieved in 80% of children with rectal and 68% with oral disimpaction, both leading to a normalization of colon transit times (Bekkali et al, 2009).

Maintenance treatment

After successful disimpaction, a long-term maintenance treatment over a minimum of six months to two years should follow to avoid re-accumulation of stool masses (Felt et al, 1999). In addition to toilet training three times a day after mealtimes, oral laxatives are given. The standard and most effective laxative is polyethylglykol (PEG; macrogol), a long, linear polymer that binds water (Candy & Belsey, 2009; Pijpers et al, 2009). Side effects such as abdominal pain are rare. The initial dose is 0.4g/kg bodyweight per day in two doses. If stools are too hard, the dose is increased, if too soft, reduced. The therapeutic range varies from 0.2g/ kg to 1.4g/kg bodyweight per day (Nurko et al, 2008). Lactulose, a disaccharide, is less effective and has more side effects. The dosage of liquid lactulose ranges from 1ml/kg to 3ml/kg bodyweight per day in one to three doses.

If comorbid emotional and behavioral disorders are present, these need to be treated separately. Untreated comorbid disorders will reduce adherence and compliance and outcome of encopresis treatment will not be optimal.

COURSE AND OUTCOME

The untreated long-term course of both types of encopresis is not favorable. Constipation and non-retentive fecal incontinence can persist into adolescence and even young adulthood (see epidemiology). Therefore, encopresis needs to be treated actively and patients should be seen at regular intervals. In case of constipation, laxatives should be given for long enough; two years or longer in some children.

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Appendix C.5.1

TAKING A CLINICAL HISTORY OF ENCOPRESIS AND CONSTIPATION: EXAMPLES OF QUESTIONS*

Presentation

- Do you know why you are here?
- Is it because you soil your pants?
- Do you want to talk about it?

Frequency of encopresis

- Does your child soil during the day?
- How often does he** soil during the day (days per week; days per month)?
- How often does he soil each day (times per day)?
- At what time of day does he usually soil (morning, noon, in the afternoons, evenings)?
- Does your child soil during the night?
- How often does he soil during the night?

Symptoms

- If your child soils, how large are the stools (only smearing; smearing and stool masses; only stool masses)?
- What is the consistency of his stools (hard, soft, watery – show the Bristol Stool Chart)?
- In which situations does he soil?
- Does he soil in stressful situations?
- Can he postpone defecating if no toilet is available, e.g. while driving? If yes, for how long?

Relapses

- Has your child ever had a period in its life without soiling?
- If yes, what was the longest period?
- At what age did this occur?
- Was there a reason for the relapse (no, constipation, diarrhea, pain while

defecating, going to kindergarten, going to school, birth of a sibling, separation of parents, other life events)?

Toileting behavior

- Does he wear a diaper?
 - If yes, when (during the day, at night only, both day and night)?
- How many days per week does he pass stools into the toilet?
- How many times per day does he defecate?
- How large are the stools in the toilet?
- Do you have to send him to the toilet? If yes, is this effective?
- Does he enough time when going to the toilet? If yes, how long?
- Does he play or read while sitting on the toilet?

Associated symptoms

- Does your child go to the toilet regularly, at certain times of the day? If yes, when?
- Does he have difficulty passing stools?
- Does he have to strain?
- Is defecation painful?
- What is the consistency of the stools in the toilet (hard, soft, watery, with different consistencies, with blood – show Bristol Stool Chart)?
- Does he complain of stomach or abdominal pain? If yes, how often?
- How strong is this pain?
- When does he experience stomach pains? Before or after meals?
- Is pain relieved after going to the toilet?
- Does he pass wind?

Perceptions and reactions after soiling

- Does your child notice when he has soiled?
- Do you notice when he has soiled?
- How do you notice it?
- Does he tell you when he has soiled?
- If no, does he try to conceal it? Does he hide his underpants?
 - How does he react when he has soiled (indifferent, no reaction, sad, anxious, disappointed, ashamed, desperate, angry or other reactions)?
- Who removes the stools from the clothing (or the bed)?
- Is he upset about the soiling? If yes, how much?
- If yes, how does this distress show?
- Is he motivated for treatment?

Reaction of parents and others in the child's environment

- How do you react when your child has soiled?
- Are you distressed by his soiling? If yes, how much?
- Have you punished him because of the soiling?
- Do you think he soils on purpose?
- Who knows that he soils?
- Has he been rejected by peers because of the soiling? If yes, how?
- How often does this occur?
- Has he been unable to take part in activities because of the soiling? If yes, which activities (school outings, swimming, others)?
- Does he engage in sports? If yes, what sports?

Eating and drinking habits

- How much fluid does your child drink per day?
- Does he prefer to eat low fiber foods? If yes, which ones?
- Does your child prefer to eat other specific types of foods? If yes, which ones?

Previous treatment

- Has your child taken laxatives? If yes, which ones? For how long?
- Has he been examined because of the soiling? If yes, where and when?
- Has he been treated because of his soiling? If yes, where and when?
- What have you personally done to treat the soiling?

Family history

- Has anyone else in your family soiled?
- Have members of your family been affected by constipation? Nighttime or daytime wetting?
- Has anyone had illnesses of the kidney, stomach or bowels?

* von Gontard & Neveus, 2006; von Gontard, 2010.

**He or she as appropriate

Appendix C.5.2

ENCOPRESIS QUESTIONNAIRE – SCREENING VERSION*

Questions	Answers	
Frequency of encopresis		
Does your child soil during the day?		yes
		no
How often does your child soil during the day?		days per week
		days per month
How often does your child soil per day?		times per day
Does your child soil during the night?		yes
		no
How often does your child soil during the night?		nights per week
		nights per month
Encopresis symptoms		
If your child soils, how large are the stool masses?		only smearing
		smearing and stool masses
		only stool masses
What is the consistency of your child's stool?		hard
		soft
		watery
Relapses		
Has your child ever had a period in its life without soiling?		yes
		no
If yes, at what age did this occur? - from age of		years; months
- to age of		years; months
Toileting behavior		
On how many days per week does your child pass stools into the toilet?		days per week
How many times per day does your child defecate?		times per day
How large are the stool masses in the toilet?		small
		medium
		large

What is the consistency of your child's stool in the toilet?	hard
	soft
	watery
	with blood
Is defecation painful for your child?	yes
	no
Does your child have stomach or abdominal pains?	yes
	no
Perceptions and reactions after soiling	
Does your child suffer emotionally due to the soiling?	yes
	no
Is your child motivated for treatment?	yes
Have you punished your child because of its soiling?	yes
	no
Wetting	
How often does your child go to the toilet to urinate?	 Times per day
Does your child wet during the day?	yes
	no
If yes, how often?	 Days per week
Does your child wet at night?	yes
	no
If yes, how often?	 Days per week
How much fluid does your child drink per day?	 Litres per day

*von Gontard and Neveus, 2006 ; von Gontard, 2010.

	DO	CUMENT	ING TOIL	DOCUMENTING TOILET TRAINING (ENCOPRESIS CHART)*	G (ENCOPF	RESIS CH	ART)*	
Date		Monday	Tuesday	Wednesday	Thursday	Friday	Saturday	Sunday
Morning								
Sent to toilet (\rightarrow)	oilet (\rightarrow)							
Went alone (!)	one (!)							
Pants:	dry (D)							
	Wet (W)							
	Stools, small (S)							
	Stools, large (L)							
Toilet:	Urine (U) Stools (S)							
Midday								
Cont to toilet	nilot / J							
Went alone (!)								
Pants:	Drv (D)							
	Wet (W)							
	Stools, small (S)							
	Stools, large (L)							
Toilet:	Urine (U)							
	Stools (S)							
Evening								
Sent to toilet ($ ightarrow$	oilet (\rightarrow)							
Went alone (!)	one (!)							
Pants:	Dry (D)							
	Wet (W)							
	Stools, small (S)							
	Stools, large (L)							
Toilet:	Urine (U)							
	Stools (S)							
*von Gontar	*von Gontard and Neveus, 2006; von Gontard, 2010.	von Gontard, 20	10.					

Appendix C.5.3