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Chapter 1

Childhood Encopresis — Pathophysiology, Evaluation and Treatment

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1. Introduction

Encopresis is defined as persistent faecal incontinence without associated anatomic abnormality. It is a common, disabling condition of children often associated with functional defaecation disorders potentially open to nonsurgical treatments. It is considered to be primarily a disorder associated with chronic constipation, with stool retention in 96% of children over the age of four years presenting with faecal incontinence. [1]

The symptom of constipation is defined broadly as difficulty or delay in defaecation [2], often associated with large calibre stools and the presence of stool in the rectal ampulla. [3-5] Anecdotal evidence from parents often shows a transition from early simple constipation to chronic constipation, faecal retention and soiling. [6]

Constipation may not be easy to identify on history. Barr et al. noted that 45 percent of children when asked the most commonly used screening question for constipation relating to frequency of bowel motions gave an answer within the normal range. [5] These children were obviously constipated based on other criteria. Some children with encopresis have daily bowel motions but apparently incomplete evacuation as evidenced by periodic passage of very large amounts of stool. [7] Conversely, infrequent defaecation alone may not indicate constipation as this may simply represent the lower limit of normality. [8] In the literature the terms ‘constipation’ and ‘faecal retention’ are used interchangeably.

A period of continence is often defined as a period of at least one month without faecal incontinence. However, the frequency of soiling episodes does not necessarily correlate with severity of constipation. [9] Soiling in childhood encopresis is variable in quantity and frequency and may occur in the range of monthly to many times a day. It can occur at night but rarely exclusively so. [10] Children may have always soiled (primary) or may have
commenced soiling after a period of continence (secondary) with a third to half of children presenting with primary encopresis. [11, 12]

The underlying pathologies resulting in childhood faecal retention remain relatively poorly understood. Colonic motility, large gut innervation, cyclic anal activity, bowel sensation and evacuation release, as well as behavioural factors may all contribute in varying degrees to the condition known generically as constipation with secondary encopresis. Encopresis involving soiling without evidence of stool accumulation and in the absence of any obvious anatomic abnormality does occur but there are no studies which have specifically and usefully examined functional disorders in this group.

Some authors attribute soiling and the generally associated faecal retention solely or predominantly to psychogenic causes. Halpern [13] reviewed the child rearing practices and personality types said to precipitate encopresis, but pointed to the lack of firm evidence associating inadequate parenting with soiling. Children with encopresis have been found to have a higher incidence of maladjustment than the normal population, but this is far from universal and more importantly has been observed to improve with symptomatic remission. [11, 14-16] It is likely therefore that the symptom of encopresis is at least contributing to the behaviour problems rather than the reverse. The stress on the child, his/her family, friends and teachers resulting from prolonged faecal incontinence is obvious but it should not distract from the need to identify and correct any functional abnormality that may exist. [16]

2. Epidemiology and natural history

Boys are far more prone to encopresis than girls, with the prevalence of encopresis amongst 7 to 8 year old boys 2.3 percent and amongst girls of 0.7 percent in the classic study from Stockholm. [17] The Isle of Wight survey found that 1.3 percent of 11 year old boys and 0.3 percent of girls were incontinent of stool [18], and similar data has recently been reported from the Netherlands. [19]

Approximately 70 to 80% of children presenting with encopresis are boys. [10, 11, 20, 21] The proportion of boys in studies of children with chronic constipation is approximately 60 percent. [3, 22-24]

In adults the picture is reversed with women being more inclined to suffer from constipation, [25] and incontinence. [26, 27] However, in the clinical group of 276 patients described by Speakman and Henry, if men and women who had had previous surgery or trauma and the women who had a history of difficult vaginal delivery were discounted then the remaining patients consisted of 6 women and 4 men. Obviously when comparing childhood and adult prevalence of any disorder it is necessary to discount adult conditions for which there is no paediatric equivalent.

Faecal incontinence tends to be underreported in medical histories, [28] likely leading to systematic underestimation of its incidence and prevalence in adults. In one study, only 5% of patients with self-reported faecal incontinence had this recorded in their medical history. [28]
Of the 46% controls who responded to the questionnaire, 5% indicated they experienced faecal soiling. It was suggested that either the doctor might be reluctant to treat the problem, or alternatively that they regarded it as a minor symptom. Therefore assessment of the recovery rate of children with encopresis by comparison of reported prevalences is open to gross inaccuracies. It seems likely that the same bias occurs in paediatric reporting. In their study of 176 consecutively referred children with constipation, Arhan et al. [29] reported a referral diagnosis of encopresis in 8% but in fact 68% of the 176 children suffered from this symptom.

There are functional differences in continence mechanisms between normal adult men and women. These include greater activity of both sphincters in men and a lower rectal volume to reach the threshold for desire to defaecate in women. [30-32] There are dangers inherent in extrapolation from the physiology of adults to children but it has suggested that gender differences also exist in paediatric anorectal function which might explain the greater proportion of boys with anorectal dysfunction. To date most studies which have included a comparison group of control children have not found evidence to support this suggestion. Corazziari did study 78 healthy children as a comparison group for 63 chronically constipated children and found no gender difference in stool frequency or total gastrointestinal transit time. [24] Only 25 (13 boys) children had manometric studies and there were no gender differences identified. Similarly, Meunier et al. found no significant gender differences in two control groups of normal children (n= 32 and 31). [23, 33]

3. Physiology of normal continence and defaecation

In order to investigate continence and defaecation disorders it is first necessary to understand these processes in a normal person. Unfortunately the literature only provides detailed studies of normal adults and so we are in the position of having to extrapolate these findings to children.

Stool frequency in Western communities decreases in the first years of life and then appears to plateau, but there is some evidence that this is not the case in developing communities for which there are no significant age-related differences. [34] Corazziari [24] observed that bowel frequency was significantly higher in children younger than three years than those of 3 to 12 years but found no difference in total gastrointestinal transit time. Normal frequency in the older age group was considered to be between 4 and 9 bowel actions per week. In young adults 5 to 12 bowel actions per week can be considered to be normal with males defaecating significantly more frequently than females. [35]

Continence is maintained by the physical resistance to the passage of faeces moving from the rectosigmoid into the rectum and thence through the anal canal. [36] Stool transfer into the rectum usually occurs as a result of colonic high-amplitude propagated contractions, which are more likely to occur after wakening and meals. The rectum is generally collapsed before the arrival of faeces, which then result in distension, rectal contraction, a sensation of urgency, reflex relaxation of the internal anal sphincter and semi-voluntary relaxations of pelvic floor muscles. If defaecation does not occur, rectal contractions and the sense of urgency slowly
subside with the rectum accommodating to continuing distension. Resistance to the movement of stool into the rectum allows its accumulation in the distal colon.

The movement of faeces into the distal rectum from the sigmoid colon is impeded by its two lateral angulations and its spiral folds. [37] Resistance to movement through the anorectum is provided by the sharp anteroposterior angulation and the anal sphincters. The anorectal angle is maintained by the striated pelvic muscles, mainly the puborectalis. The anal sphincters form a high pressure zone consisting of two overlapping muscles: the internal anal sphincter (IAS) composed of smooth muscle, and the external anal sphincter (EAS) composed of striated muscle. Tonic change in the IAS is entirely reflex whilst that in the EAS is under voluntary control. Contraction of the puborectalis sling in conjunction with contraction of the EAS is thought to assist the role of this sphincter. It does not appear to play as important a role in the maintenance of continence as the EAS.

The two sphincters can function independently of each other, depending on the need to accommodate faecal matter, ascertain the nature of the rectal contents, preserve continence or to defaecate. At rest the sphincters maintain a high pressure zone which has an asymmetric profile with the highest pressures in the outermost sphincter area. [37] The asymmetry is largely maintained by contraction of the EAS which predominantly surrounds the distal anal canal and is submaximally tonically active under resting conditions. However, approximately 80 percent of the total sphincter tone is due to the activity of the IAS. Cyclical variations in resting pressure within the anal canal including spontaneous relaxations of the sphincter have been observed in adults [32, 38] and in children. [39] There is a reduction in IAS tone, resting EAS activity and colonic motor activity during sleep in adults. [37] There is some suggestion that the EAS undergoes periodic change in tonic activity. [40]

With the arrival of sufficient faeces in the rectal canal to cause it to distend to a threshold volume there is a reflex relaxation of the IAS accompanied by contraction of the EAS. This rectoanal inhibitory reflex (RAIR) is associated with an increase in rectal pressure due to rectal contraction and within one second a transient sensation. [41] The triggering of the RAIR appears to be dependent on the rate of rectal distension: slow continuous filling allows a greater volume to collect before the IAS relaxes. Further increases in rectal contents beyond this threshold produce a gradation of sensation from that of wind, to an urge to defaecate, to the experience of pain. [42] Following each increase in rectal contents and volume, the EAS recovers resting tone after the brief increase in activity. There is however a rebound increase in resting pressure of the IAS and the baseline rectal pressure increases for a period accompanied by an increase in rectal contractions. [30]

The rectal contractions reduce earlier with slower rates of filling but accommodation of the rectal contents can occur longitudinally without the necessity of relaxation of the rectal wall. [43] The increases in rectal contractions and axial pressures possibly tamp the stool into the proximal anal canal thereby increasing the defaecatory urge. [44] As the volume increases the relaxation of the IAS increases in strength and duration until recovery no longer occurs.
and there is a sustained relaxation. Parallel to this the contraction of the EAS increases in strength and duration, maintaining continence. Prior to sustained relaxation of the IAS during the resting phases, Frenckner [45] determined that the IAS is responsible for just over half the anal tone and is therefore still important in maintaining continence.

The progression from mild to acute urgency generally occurs with the attainment of sustained increase in rectal tone and dilatation of the IAS. The sense of urgency is likely due to activation of stretch receptors in the proximal rectum or sigmoid colon. [43] There is disagreement about whether acute urgency and sustained relaxation of the IAS always occur together in normal children [4, 16, 46] while in adults Sun et al. [43] found no evidence for this.

There are significant differences between age groups in normal children in both the maximal tolerable rectal volume, and the threshold volume required to elicit rectal contractions (rectorectal reflex). [33] The thresholds for these increased and decreased respectively with increasing age.

Although some component of the EAS response to the inhibitory reflex is spinal (as it is observed to an extent in paraplegic patients) [47] depending on the level of the lesion it is susceptible to conscious control and therefore must be modulated by CNS involvement. [48] During sleep there is no diminution of the IAS response but there is a significant reduction in the EAS component.

Very high levels of rectal distension can be associated with reflex abolition of both EAS and IAS activity causing a profound reduction in anal pressure [41] resulting in automatic defaecation. [49] This has been noted in normal children [4, 50] and adults. [51] This reflex is present in paraplegic patients with intact peripheral nerves and distal spinal reflexes so it is probably autonomous. [47]

The sensory receptors are complex in that not only the presence but also the nature of rectal contents are perceived, and the sensation due to IAS relaxation is felt differently from that due to rectal distension. [52] Receptors exist in the anal canal and may exist in the rectum and the muscles of the pelvic floor. There is disagreement about the origins of rectal sensation, whether mediated by receptors in the pelvic floor and not in the rectum, or whether as Sun [43] and Loening-Baucke [53] have postulated there are at least two types of rectal receptors: rapidly adapting mucosal receptors and slowly adapting mechanoreceptors in or on the rectal wall, as well as the possibility of some in the sigmoid colon.

Sun [41] found that the duration of IAS relaxation and sensation in adults were not correlated although the former was always shorter than the latter. However, a strong association was found between the durations of EAS contraction and sensation. Transient sensation was not generally perceived if rectal contractions were not elicited and the EAS did not contract unless perception occurred. [30] Buser et al. [52] found that some adults with faecal incontinence did experience sensation at a time when EAS contraction was absent, so postulated that the EAS contracts as the result of rather than the cause of sensation. Read and Read [54] have suggested that the role of anal sensation receptors, as opposed to the rectal complex, may not be to
preserve continence but to identify the rectal contents or signal the end of defaecation. [49] If this is the case then the RAIR allows testing of the rectal contents by these receptors, providing conscious information on which suitable actions may be taken. It has been suggested the spontaneous cyclical IAS relaxations fulfil the same purpose. [38]

An alternative or supplementary mechanism for the identification of the physical nature of faeces may be associated with the different rates of distension of the rectal wall accompanying the propulsion of material from the distal colon. As well as differences in RAIR thresholds, rapid rectal distension has been found to produce a different sensation from gradual distension to the same volume so that distinction between these may provide the discriminatory information. [43] However, whatever the order and origins of stimuli, it is obvious that once the rectal contents have reached the threshold for reflex relaxation of the IAS then at least subconscious awareness of stool in the rectum and immediate contraction of the EAS are essential for the preservation of continence. The ability to experience a sense of urgency before profound reflex anal dilatation occurs is likewise essential.

With an increase in intra-abdominal pressure there is a reflex compensatory increase in EAS activity to a level which provides an anal pressure in excess of the rectal pressure. [30] This allows continence to be maintained when coughing, sneezing, blowing up balloons, laughing or any other activity which poses a threat by its effect on abdominal pressure. Voluntary squeezing or tightening up of the EAS to maintain continence involves no increase in intra-abdominal pressure. [6]

Voluntary defaecation takes place in three phases. Initially there is an increase in abdominal pressure and rectal pressure brought about by closure of the glottis, fixation of the diaphragm and contraction of abdominal, perianal and hamstring muscles combined with contraction of the puborectalis sling and both sphincters. [37] Then the pelvic muscles relax allowing straightening of the rectoanal angle and of both sphincters. The normal anorectal angle at rest is approximately 90° and increases to 125° during straining. At the same time strong colorectal contractions assist expulsion of the stool and the anal sphincters relax. Electrical activity in the EAS is greatly reduced at this stage. Schuster [49] suggested that this relaxation takes place when the threshold for automatic defaecation is reached. As defaecation proceeds the rectal pressure gradually falls. The third stage involves the return to the original state after a rebound contraction of the anal sphincters.

It can be seen from the complex nature of continence and defaecation that there are many opportunities for problems to occur both through physiological deficits and disordered processes. [36] Insufficient IAS or EAS resting tone, inadequate or delayed EAS response to the rectoanal inhibitory reflex, elevated or absent threshold of sensation from rectal distension and a blunted feeling of urgency have all been proposed as possible causes or at least contributors to faecal incontinence. Inadequate colonic propulsion, failure of the IAS to relax, inappropriate contraction of the EAS and puborectalis, failure of the levators to lift the pelvic floor, luminal obstruction or an impairment in the central control of defaecation may singly or in combination result in obstructed defaecation. Failure to relax the striated musculature of the pelvic floor during straining has been termed anismus [55] and probably results in incomplete evacuation, faecal retention, chronic distension of the rectum, and possibly
concomitant reduction in sensation leading to soiling or, at least, to its continuation. Attempts to identify pathophysiology which may be present in children with faecal incontinence have largely concentrated on studies of resting anorectal pressure and motility characteristics, sensation, the RAIR and the investigation of anismus.

4. Treatment

The aim of treatment is to for the patient to achieve the ability to be in charge of his/her own continence and defaecation. To this end any significant faecal impaction needs to be relieved and a regular output established. Treatment for encopresis falls into three stages with the first being initial disimpaction with commencement of maintenance laxatives or prokinetic agents. The second stage is the establishment of a good bowel habit by the use of behaviour modification; and thirdly, the correction, if necessary, of abnormal defaecation dynamics. The first two modes of treatment are frequently adequate to resolve the problem but if the encopresis is refractory. Many children respond well but there is undoubtedly a group who continue to have long term problems past puberty.

4.1. Laxatives

Laxative treatment regimens vary in detail but generally aim to produce one to two bowel actions per day. The extent of the faecal retention determines the type of medication. Polyethylene glycol (“macrogol”) based regimens are increasingly accepted as a first line, but there is still an occasional place for stimulant laxatives such as senna derivatives or bisacodyl. Enemas and suppositories are now only infrequently used for disimpaction. Increased fibre is of use only if the current intake is inadequate.

4.2. Behaviour modification

Concurrently with laxative medication, a star chart with a reward system both for successful defaecation in the toilet and for soil-free days can be used as positive reinforcement aimed towards achieving an improvement in toileting habit. Regular sits three times a day for 5-10 minutes with a minimum of distraction is an effective regimen In addition, clarification of the physiology of encopresis to parents and children to alleviate guilt is very important, as is attentive follow-up to maintain compliance and monitor progress. In a referred population of children presenting with encopresis this regimen can be expected to result in complete remission from soiling in approximately half and in addition to be independent of laxatives in the same or less.

4.3. Biofeedback for treatment of anismus

The rationale for the development of biofeedback had been to provide a correction of disturbed anorectal dynamics, and especially for paradoxical sphincteric contraction or anismus. The method recommended for biofeedback generally is the same as, or an adaption of,
anorectal manometry with some sort of visual or auditory feedback of sphincteric contraction. Unfortunately critical evaluation in controlled studies has failed to provide evidence of superior efficacy to standard treatments. [62, 63]

5. Discussion

Encopresis in childhood is an important cause of soiling, with socially disabling consequences. It is usually associated with constipation and is thought to be secondary to periodic relaxation of the anal sphincters in the presence of a loaded rectum with secondary seepage. The pathophysiology of disturbed anorectal function is relatively poorly studied in children and results often interpreted with data obtained from adult studies. Most children do have some type of manometric abnormality and many have a degree of rectal enlargement. Dynamic abnormalities also exist and the best studied is paradoxical sphincteric contraction or ‘anis-mus’.

Treatment regimens which include a combination laxatives for disimpaction and maintenance, together with behavioural interventions centred around encouraging toileting are generally effective but there is a group of children who go on to have significant long term problems.

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References


