Faecal soiling and anal achalasia

Overflow soiling from chronic accumulation of faeces in the rectal ampulla is a distressing problem in children. In as many as one-sixth, the underlying cause is anal achalasia, involving a failure of the anus to relax reflexly\(^1\) that can be shown by anorectal manometry.\(^2\) Most of the children in this group are of preschool age and many will present as toddlers. Some, but not all, will have a history of sluggish defecation in infancy. Anal achalasia usually proves to be due to ultrashort segment Hirschsprung’s disease, with typically abnormal intrinsic neurohistology evident in appropriate specimens from the wall of the rectal ampulla. A suitable biopsy can be obtained by anorectal myectomy,\(^3\) which has proved to be a benign procedure. The close correlation between the abnormal histological findings and the abnormal response to anorectal manometry has discouraged the widespread use of the latter for diagnosis, as, in addition to special skill, it requires time and highly specialised and costly equipment. Moreover, as myectomy appears to assist effective treatment, it has come to be practised relatively widely by paediatric surgeons for the diagnosis and management of ultrashort segment Hirschsprung’s disease.\(^4,6\)

The facilities needed for manometry and, to a lesser degree, myectomy are rather sophisticated and specialised, and only accessible to a few favoured paediatricians. Some of those less favoured may consequently fear that they are ill equipped to offer appropriate management to a proportion of the children that they see with faecal soiling. Happily, this fear may be groundless, for most patients with anal achalasia can be managed effectively without confirmatory tests to establish the diagnosis, and without myectomy, even for ultrashort segment Hirschsprung’s disease. This was demonstrated 38 years ago by Hurst,\(^6\) who showed that anal dilatation was helpful, at a time when it was not considered appropriate to examine the intrinsic neurohistology of the part. It is possible today, in centres with facilities to interpret the histology of small specimens taken from the short aganglionic zone in the rectal wall by suction mucosal biopsy,\(^7\) to diagnose ultrashort segment Hirschsprung’s disease without requiring myectomy specimens. It has also become clear that stretching the anus under general anaesthesia will help to relieve the symptoms of anal achalasia for a considerable time, in much the same way as the daily use of Hurst’s massive anal dilator. If required, it is not unduly hazardous or difficult to repeat the stretching of the anus at intervals.\(^8\)

Incidentally, an essential preliminary to myectomy is stretching of the anus, so that where myectomy has been performed in the management of ultrashort segment Hirschsprung’s disease it is impossible to decide the proportional benefit from the myectomy and the anal stretch. It follows that where there is a mass of faeces dilating the rectal ampulla with consequent faecal soiling, empirical stretching of the anus under general anaesthesia may prove helpful in treatment.

The general principles underlying the management of chronic constipation in children have been summarised by Bentley,\(^9\) but the following regimen can be used usefully to manage megarectum and rectal inertia, with or without anal achalasia. The presence of an abnormal accumulation of faeces in the rectal ampulla of a patient can be deduced from the history and confirmed by physical examination. Where there is a faecal mass in the rectum a plain abdominal radiograph will confirm the distension of the rectal ampulla and give a good indication of the amount and distribution of faecal accumulation in the large bowel, without the distress and cost of a barium enema examination. A large pelvic mass can impede emptying of the bladder and predispose to urinary tract infection, so that urine culture is appropriate, with the possible need for further investigation and treatment of urinary tract infection. Thereafter, examination under general anaesthesia allows a detailed examination of the anus for other lesions, general confirmation of the physical findings, and stretching of the anus.

Digital evacuation is undertaken for any massive accumulation of faeces in the rectum. Detention as an inpatient is then helpful, as a child who is removed from his home into the relatively ordered discipline of a children’s ward will often accept, without a distressing struggle, phosphate enemas*.

*Fletcher’s disposable phosphate enema (Pharmax Ltd.).
administered by a skilled children's nurse. Enemas are repeated twice weekly until the large bowel is emptied of faeces. Thereafter the reflex of defecation, as distinct from colonic peristalsis, will require continued stimulation, preferably without either excessive distension of an oversized rectal lumen, or the skilled administration of enemas or suppositories. Fortunately this can be achieved by bisacodyl (Dulcolax) given by mouth instead of by more direct application to the rectum as a suppository. It is also useful to prevent rock-hard stools and increase faecal bulk by giving 50% lactulose syrup in moderate doses. The medication is adjusted to achieve full evacuation of formed faeces (rather than an incomplete bowel motion) once every 24 to 48 hours. This will continue to stretch the anus. The preferred time for evacuation is late afternoon, after the hurry associated with the school day.

When the pattern of evacuation is stable and satisfactory, treatment can continue at home, but the follow-up should be prolonged, as interested supervision reduces the chance of relapse. Should this occur, the stabilisation regimen, including the anal stretch, may need to be repeated.

Happily, behaviour problems associated with faecal soiling commonly diminish with cessation of the symptom, but occasionally they persist to require further attention. Semantics aside, the management of true encopresis is a separate and distinct problem.

References
1 Hurst, A. F. (1934). Anal achalasia and megacolon (Hirschprung’s disease; idiopathic dilation of the colon). Guy’s Hospital Reports, 84, 317-350.

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