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Megarectum in the Older Child

I would stress that these remarks apply only to the older child with an intractable condition causing symptoms which make him and his fellows inevitably bowel conscious.

Definition
This condition has been called 'terminal reservoir', 'megarectum' or 'rectal inertia'. The last term is preferred to 'colonic inertia', which has also been used, because the colon seems to work very well in packing fæces into the accommodating rectum. The colon appears remarkably normal even when the rectum reaches up to the ribs. Real tubular dilatation of the colon seems rare although dolichocolon is sometimes seen as a temporary radiological appearance which disappears when the colon is stimulated to contract.

Constipation is taken as meaning incomplete evacuation, not merely an infrequent bowel habit.

Symptomatology
These children may present to the surgeon only when spurious diarrhœa develops caused by chronic constipation with impaction of fæcal masses and overflow incontinence. The constipation commonly dates back to the toddler 'training' period. Fæcal masses are easily palpable per abdomen as well as per rectum. The rectum is dilated down to the anal canal. The typical patient is not seriously ill though often irritable, off his food and 'tetchy'. Rarely a neglected case becomes ill, with abdominal distension, wasting and even physical immaturity simulating Hirschsprung's disease. There is commonly a functional overlay from the embarrassing symptom but it seems that this may be an accompaniment rather than a cause of the condition in the majority.

Ætiology
The majority seem to arise as a training problem, the child taking against potting and acquiring a habit of holding back. Dr Oppé will discuss this stage. If mismanaged, physical changes may arise in the rectum which lead to a vicious circle, incomplete evacuation and further rectal enlargement.

There are several less common ætiological groups: other training problems (e.g. prolonged recumbency during other illness); persistent psychological upset; congenital constipation (Coekin and Gairdner); secondary to local factors: fissure-in-ano, anal stenosis, neurological, e.g. spina bifida.

Encopresis is a different problem. The word should be restricted by analogy with enuresis to incontinence without recognizable physical cause. The soiling is not an overflow incontinence and there is no marked constipation or megarectum.

Physiology
Control depends on two sensory and two motor factors, not all of which are essential. Rectal and anal canal sensation are distinct. The striated muscle sphincter controls incontinence and the internal sphincter prevents soiling between times.

Our earlier tests were by simple balloon inflation in the rectum to produce subjective sensory responses as used by Goligher & Hughes (1951). They showed that a very large bolus was necessary to cause the sensory response to distension and in some only colonic sensation was elicited. In 3 cases re-tested after treatment a normal response to a more normal sized bolus returned, suggesting a secondary loss of the call to stool rather than a primary sensory defect.

Callaghan & Nixon (1964) used a more elaborate instrument measuring objective pressure changes and electromyographic responses to rectal distension, like Porter (1961). They showed changes in these children of varying severity labelled as: (1) *Enlarged* – giving a normal response with a raised threshold. (2) *Expanded* – giving a response after a loading volume. (3) *Inert* – giving an abnormal response, so that reflex inhibition of the sphincter preceded any sensation of filling – a situation clearly likely to allow soiling.

Cases from Dr Berg's psychiatric clinic were also studied. They showed the same changes but, on average, of a lesser degree suggesting that the place of referral depended on differences of degree rather than of basic characteristics (Table 1).

Table 1
Rectal function in severe chronic constipation

Rectum	*Surgical clinic*	*Psychiatric clinic*
Inert	6	5
Expanded	4	9
Enlarged	1	3
Normal	1	5
Total	12	22

J Lawson (unpublished observations) has now developed a probe for more detailed study of the anal canal. Four chambers in line connected to Statham transducers and an Offner recorder measure the pressure responses to various stimuli simultaneously in different zones of the anal canal. This allows differentiation of internal and external sphincter activity. EMG records of the external sphincter are synchronously recorded. I believe the method to be original in allowing the pressure profile of the anal canal to be measured synchronously without moving the probe and hence introducing further stimuli.

Cases have been seen in which there is a failure of relaxation of the external sphincter in response to a rectal pressure wave, as described by Porter (1961). Two cases have had 'dissociated sensation' in that distension which produced a pressure wave down the rectum produced no sensation until the anal canal was reached. However, these seem exceptional.

Cine barium enemas have also been used. It has been observed that return to more normal size and contractions occurs in some after treatment. In others the rectum remains inert and is emptied passively by intra-abdominal pressure, like toothpaste being squeezed from a tube; such patients seemed unaware that they were using an abnormal method of evacuation.

Pathology

In gross cases there is some myohypertrophy. This might be explained entirely by Laplace's law, the tension in the wall at a given pressure being proportional to the radius of the cylinder.

Others have reported minor changes in the intramural nervous tissue, a 'forme anale de la maladie de Hirschsprung'. Such changes must be difficult to interpret and I have had the experience of the same slides being reported as normal at one hospital and abnormal at another. We have as yet been unable to demonstrate the physiological reaction typical of Hirschsprung's disease in the anal canal of any of these children and have observed that aganglionic segments at least as short as 2 cm can produce the typical clinical picture of Hirschsprung's disease rather than that of rectal inertia.

Table 2
Usual clinical picture

	Hirschsprung's disease	*Rectal inertia*
Onset	Neonatal	Training period
Constipation	+	+
Distension	+	−
Peristalsis	++	−
Rectum	Empty	Loaded
Soiling	−	+
General health	Poor	Unimpaired
Risk to life	High	Negligible

Diagnosis

Table 2 compares the typical findings in inertia and Hirschsprung's disease. The latter may be clinically mild or modified by management. However, it seems that overflow soiling does not occur in Hirschsprung's disease although uncontrollable diarrhœa may occur during attacks of enterocolitis. Radiology and/or biopsy are diagnostic.

Management

This must be prolonged because one has to redevelop a lost habit. With such prolonged care the outlook seems good. I was able to trace 14 of 17 cases eight years after they were studied: 13 were completely well and the fourteenth had such minor trouble he had not thought it worth consulting a doctor.

Conservative treatment is in the following stages: (1) Explanation and encouragement. (2) Empty bowel. (3) Keep empty to recover 'tone'. (4) *Teach regular efforts at defæcation without waiting for a 'call to stool'* – a 'by-the-clock' pattern.

Berg commented that medical treatment often consisted only of 'wash 'em out and chuck 'em out' and showed how unsatisfactory this was. Part of the syndrome is often a mother who does not cope well and continued aftercare is essential. Even then a third may relapse before finally responding.

Measures to Evacuate

(1) Fæcalomas may need manual evacuation under general anæsthesia.

(2) Daily rectal washouts of saline, not enemas, using up to 20 pints, a few ounces at a time. This is usually ineffective at home and upsetting in the emotional atmosphere engendered by the attempts.

(3) Hypertonic phosphate enemas may be adequate in milder cases and are much less laborious for child and nurse or mother. Some older children can learn to give their own.

(4) Aperients such as Senokot or Dulcolax are invaluable but must be used in a dose adjusted to the child's often enormous tolerance – perhaps 4 or more tablets in a single daily dose.

Berg showed that his 'senna and psyche' regime could remove large masses which I would have expected to require passive removal and the ability to use such a management at home is especially valuable in the younger child (Berg & Jones 1964).

A very few older children have developed a rectum so large that even after colostomy it could not contract down to a reasonable size. Occasionally I have had to resort to resection. Since the rectum is the involved organ this has been a

Swenson type procedure and is very tedious. I would stress that even then the removal of the rectum is only the first stage in treatment and must be followed by advice on habit training, perhaps with aperients and even enemas until a regular habit is achieved. It would seem that such extreme measures would rarely be needed if the earlier stages were adequately managed, but perhaps proctological surgeons see more of these long-neglected cases in adults (Todd 1961).

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The Myenteric Plexus in Hirschsprung's Disease

The normal myenteric plexus lies between the two muscle coats of the intestine and is a network of ganglion cells joined by small nerve trunks. If the gut is sectioned transversely little can be seen of this plexus and if the pathology is to be assessed it must be opened out and cut in the same plane as the plexus. If 100 μ frozen sections are cut in this plane and impregnated with silver, the anatomy of the plexus can be seen in the normal and the abnormal (Smith 1967).

Surgical specimens of colon from two cases of Hirschsprung's disease were studied, a female aged 26 and a male aged 28. They showed considerable dilatation with a short contracted segment at the lower end. Standard paraffin sections showed a short area of aganglionosis with bundles of unmyelinated nerve fibres. Silver impregnations from the lower end showed occasional random ganglion cells and a network of unmyelinated nerve fibres with a similar pattern to the normal network but without ganglion cells at the angles. The fibres in these nerves were fine and tightly packed, quite different from the thick straight fibres seen in the normal. Further up the gut these bundles contained numbers of fragmenting axons and ganglion cells became more numerous. Some of these ganglion cells were situated in the angles of the unmyelinated trunks. Much of the plexus when it did appear was disorganized, with axons going in all directions and often curling back again. There was no normal plexus on the specimens. The fact that a few ganglion cells were seen at the lower end in spite of their absence on paraffin sections is related to the thickness and orientation of the frozen material. One would need to cut approximately 150–200 serial 5 μ paraffin sections to see as much plexus as in one of these sections.

It has been shown by Hukuhara *et al.* (1961) that if the ganglion cells are destroyed by anoxia, the peristaltic wave does not pass but fæces get through because there is normal gut above. In these cases it is possible that the clinical symptoms are related to the combination of a partial physiological obstruction combined with poor or absent peristalsis for some distance proximally. Either condition alone might not be incapacitating.

The embryology of this condition may be related to arrest of the normal development. Jones (1942) and Kuntz (1952) have shown that the parasympathetic nerve supply develops originally as axons from the central nervous system either in the brain stem or the sacral cord. At a later date, ganglion cells migrate down these axons to form the adult plexuses. It is possible that the unmyelinated bundles in Hirschsprung's disease, which are known to be cholinergic, are the remains of the original sacral outflow. There is some suggestion in this material that they are breaking up and being replaced by a more adult type of plexus and that ganglion cells are taking up their positions in the angles of the original nerve fibres. For some reason this process has not gone to completion leaving the abnormalities we see in Hirschsprung's disease.

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Megacolon and Megarectum in Older Children

In the absence of organic obstruction to the outflow of fæces, massive distension of the rectum and anal canal must be due to failure of the propulsive mechanism, to failure of the anal sphincter to relax and permit egress of stool, or to active retention of fæces by contraction of the anal and perineal musculature. Once fæcal accumulation has occurred it is likely that secondary disturbances of both the sensory pathways and motor mechanisms take place so that the normal mechanisms governing both fæcal continence and defæcation become impaired.

The older child presents with a complex pattern of bowel dysfunction which I regard as the terminal stage of stool withholding. The chronically over-distended lower bowel is incapable of