Section of Proctology

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Megacolon and Megarectum in Older Children and Young Adults

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Classification and Terminology

Prior to the discovery of aganglionosis as the cause of Hirschsprung's disease, terminology was simple and adequate with regard to the facts known at that time. A clear distinction was made between idiopathic megacolon, also called Hirschsprung's disease, where no obvious cause of the dilatation and hypertrophy was found, and symptomatic (or secondary) megacolon, developing in patients with a demonstrable causative disorder such as stricture of the anus or rectum, lesions of the central nervous system or endocrine disorders.

With later concepts of Hirschsprung's disease, classification became a little more complicated. Instead of two main groups of megacolon, there were now three. Hirschsprung's disease formed the new group, characterized by congenital absence of ganglion cells from a distal segment of bowel. Idiopathic megacolon remained as a group of unknown origin, also called functional or psychogenic megacolon or megarectum, colonic or rectal inertia, or simply chronic constipation. Symptomatic megacolon remained unchanged.

This subdivision of megacolon has been more or less adequate for nearly twenty years but is now beginning to be outgrown. A number of new disease entities have been described, which closely resemble but are not identical with Hirschsprung's disease, and do not fit in under the headings of idiopathic or symptomatic megacolon. These new syndromes are not common enough to be important from a practical point of view. Theoretically, they are of great interest; we have to face them and to place them as adequately as we can in our poor knowledge of them. An extended basis for the subdivision and denomination of different types of megacolon is thus needed. Although many objections can be raised against terms currently used, it seems preferable to base an extended terminology on the old names. Everybody knows what they stand for, whereas a completely new terminology – even a logically perfect one – is apt to create confusion rather than clarification.

I have tried to group the different types of megacolon according to their underlying pathology as far as is known at the present time,

Table 1
Classification and terminology of megacolon

	Normal ganglion cells	
Abnormal ganglion cells	*Mechanism known*	*Mechanism unknown*
Absence		
HIRSCHSPRUNG'S DISEASE	SYMPTOMATIC MEGACOLON	IDIOPATHIC MEGACOLON
Congenital megacolon	Secondary megacolon	Functional megacolon
Congenital aganglionosis	(anorectal obstruction	Psychogenic megacolon
Hypoganglionosis	disorders of central	Megarectum
	nervous system, endocrine	Colonic inertia
	disorders)	Rectal inertia
		Chronic constipation
Degeneration		
ACQUIRED MEGACOLON		PSEUDO-HIRSCHSPRUNG'S DISEASE
(Chagas' disease)		(Segmental dilatation of the colon)
(Complication of anorectal surgery)		(Pseudo-Hirschsprung, Ehrenpreis)
		(Pseudo-Hirschsprung, Katz)
Immaturity		
of ganglion cells		

disorders with unknown ætiology and pathology forming a separate group (Table 1). Common synonyms for denomination are listed, the names given in capitals being those most used. The disease entities in parenthesis are subdivisions of a major group.

The first column comprises disorders caused by abnormality of ganglion cells: absence, total or subtotal (hypoganglionosis), degeneration and immaturity. If the term hypoganglionosis is judiciously used, denoting those cases only where isolated ganglion cells are present, this small group of patients should be included in Hirschsprung's disease, being a clinical entity. Degeneration and reduction in numbers of ganglion cells is the pathology found in Chagas' disease. This syndrome is caused by a trypanosome and is transmitted by an insect. The disease is endemic in certain areas of South America and is practically unknown elsewhere. A similar pathology has been found in a few patients developing a megacolon as a complication of anorectal surgery (Ehrenpreis 1965*a*, Nixon 1966). Presumably these changes have resulted from temporary impairment of the vascular supply to the segment of colon involved, much the same way as in experimentally produced megacolon (McElhannon 1960, Hukuhara *et al.* 1961). Immaturity of ganglion cells (Smith 1964, personal communication, Spencer 1966) is a physiological phenomenon in embryos and premature infants, which may be mistaken for absence of ganglion cells in evaluating rectal biopsy specimens.

Symptomatic megacolon is a well-defined group with a clear ætiology and needs no further comment in this connexion.

The third column deals with megacolon without histological evidence of any abnormality. So far we know practically nothing about the ætiology. Idiopathic megacolon and its various synonyms form the bulk of this group. These terms reflect some of the ideas on the origin of this condition. In recent years a trend towards a reduction or even elimination of the group of idiopathic megacolon in favour of other disease entities has been developing. In particular, Duhamel (1966) claims to have demonstrated distal aganglionosis or histological abnormalities of the structures of the internal sphincter muscle in a large proportion of these cases. Davidson & Bauer (1958) reported on a few patients showing a motility pattern in the distal rectum characteristic of Hirschsprung's disease, although there was no narrow segment of bowel and no abnormality of ganglion cells.

Personally I prefer the term chronic constipation, because it infers no more than we know and because to my mind constipation, not megacolon, is the essential feature of this disorder.

Until recently, megacolon developing in the absence of clear-cut causative factors has been an adequate definition of idiopathic megacolon only. Today we have reason to add a new group of disease entities to this definition. The term pseudo-Hirschspring's disease has been used to denote different things. Ravitch (1958) coined this name in presenting cases which are generally included under the heading of 'idiopathic megacolon', because he disliked the term 'idiopathic'. This term has, however, a strong position in medical terminology and should therefore be preserved. More recently two different disease entities have been described under the name of pseudo-Hirschsprung's disease, one by myself (Ehrenpreis 1965*b*), including cases described earlier under various names (Bill *et al.* 1957, Nixon 1961, Allegaert 1963) and one by Katz (1966). Finally, the same term was used as the title of a seminar, covering all the syndromes in this group (Bentley *et al.* 1966); this seems to be the most sensible way to use this term.

Pseudo-Hirschsprung's disease means a syndrome or a group of syndromes simulating Hirschsprung's disease. Besides the two entities reported by myself and by Katz, one further syndrome has been described, which belongs to this group: segmental dilatation of the colon (Swenson & Rathauser 1959). All these entities simulate Hirschsprung's disease clinically as well as radiologically, presenting with a narrow distal segment of bowel. No abnormality of the intrinsic plexuses has, however, been found in these cases. In this respect and with regard to our lack of knowledge of the ætiology, they resemble idiopathic megacolon. At the present time pseudo-Hirschsprung's disease should be considered as a group of megacolon, separate from true Hirschsprung's disease as well as from idiopathic megacolon.

I would like to end this short survey by quoting from a review by Bentley (1964): 'When simple histologic examination reveals no abnormality in the wall of the bowel, subtle intrinsic factors (possibly inborn) might still exist to impair the reflex of defecation. As knowledge of neurochemistry and smooth muscle activity increases, more sophisticated technics may develop to study the tissue excised from the rectum'.

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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