Section of Proctology

President R W Nevin TD FRCS

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Discussion on Megacolon and Megarectum with the Emphasis on Conditions other than Hirschsprung's Disease

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Megarectum and megacolon have not been discussed since Gardiner's paper on megacolon in 1953. He adopted a classification which seemed generally acceptable, namely: (1) Obstructive or symptomatic (including C.N.S. lesions). (2) Aganglionic or Hirschsprung's disease. (3) Simple (also known as functional) – idiopathic, pseudomegacolon, dolichocolon, colon inertia. A better name for the second type should be a more-embracing term, rather than 'aganglionic' or 'achalasic', to include all abnormalities of the ganglion population: for in Hirschsprung's disease no ganglia are found in a segment and in Chagas's disease, due to the *Trypanosoma cruzi*, there may well be a quantitative diminution of ganglia. Gardiner discussed mainly the simple or functional group; much of this paper is devoted to this group, though the findings are not altogether in keeping with his. It is the presence of the megarectum as well as the megacolon which makes treatment in the adult difficult.

What is megarectum and megacolon? The condition may be defined as a state in which the bowel is persistently much larger in diameter than normal and intractably constipated. Spurious diarrhœa may sometimes be a symptom of this.

Obstructive megacolon and megarectum may occur at any age, from lesions pressing on the bowel wall from outside, such as pelvic tumours, and from lesions in the bowel wall or within its lumen, such as stenosis, strictures and tumours. These can be dealt with by removal of the obstruction; they are not true megacolon. Gardiner included central nervous system lesions such as spina bifida in the obstructive group. Many extensive neurological lesions cause megacolon; these are separate and difficult problems. A fourth type of megacolon should be included as 'disorders of the central nervous system'.

Neurological lesions and the obstructive type have been excluded from this survey which is of 53 cases, 28 cases being documented. Over half of the cases presented before the age of 30, and over a third between the ages of 11 and 30. Excluding those occurring in the first year, 24 were males and 23 were females. In the first year of life all were males: 5 presented during the first weeks of life, all due to Hirschsprung's disease; 1 started at five weeks and was thought to be functional.

There were 8 cases of Hirschsprung's disease in the adult proven by adequate rectal biopsy and 1 possible case where no biopsy was done. All the definite Hirschsprung's disease cases were male; the possible case is the only female. All were aged between 21 to 40 except 1, whose operation was carried out at the age of 13. Hirschsprung's disease in the adult is not uncommon. The aganglionic segment appears to be confined to the rectum and has not, as yet, extended far into the sigmoid. I have not seen any case of a colonic aganglionic segment without rectal involvement and most of such cases recorded are inadequately documented. The Swenson-type pull-through procedure in the adult is formidable, as the pelvis is much deeper than that of a child and the colon has reached gigantic proportions, perhaps up to 25 cm in diameter, and the bowel wall may be 4–6 mm thick. If a pull-through such as a Maunsell-Weir abdomino-anal procedure can be accomplished, the colon is apt to remain adynamic, as sometimes occurs in the infant and perhaps related in ætiology to that seen in neonatal intestinal obstruction (Nixon 1960).

The results of surgery for Hirschsprung's disease in the adult, at St Mark's and St Bartholomew's Hospitals, are not very good. Three of the 4 cases treated by anterior resection have poor results as they require frequent enemata; the fourth has a fair result but the bowel above the anastomosis remains loaded. These cases may

recur, as only a small part of the aganglionic segment was removed. One patient was treated by a Marden Black pull-through operation but though his bowels move he is not truly or adequately continent. Three cases have good results: 2 had abdomino-anal operations and the bowels move normally but one is impotent; the third is the questionable case of Hirschsprung's disease treated by colectomy and ileorectal anastomosis (a boy operated on at 13 years, who had a good result from a modified Swenson-type pull-through procedure, is excluded).

I wish to stress the fact that though most cases of Hirschsprung's disease present in the neonatal period, not all do so; if possible cases occur later an adequate rectal biopsy should be taken and if no ganglia are found an operation advised. It is doubtful if medical management should be advised permanently once a diagnosis of Hirschsprung's disease has been established, for sooner or later operation will become mandatory.

The simple or functional group of megacolon and megarectum when they reach adult life are difficult. In the child corrective training may achieve temporary improvement in a majority. Lee *et al.* (1950), Ravitch (1958*a*,*b*) and Hallenbeck & Waugh (1952) all agree that medical management is correct, except in cases with a long sigmoid, the dolichocolon which intermittently twists. This is in keeping with Gardiner's observations and sigmoid resection is advised for these cases. This medical management or sigmoid resection is not the answer for all and attention to clinical findings and the macroscopic appearance at laparotomy is rewarding. There is an inconsistency in the literature in regard to the condition of the anus in the functional case. Many times the anus is said to be relaxed and even gaping with soiling of the clothes, yet at other times a remark appears such as 'if the anus is spastic it must be dilated and if need be a sphincterotomy carried out'. Surely these two conditions are not one and the same disease.

Three different and distinct macroscopic appearances of the distal colon and rectum may be found: (1) Dilated and relatively thin-walled sigmoid, frequently associated with a rectum which appears normal or a little dilated. There is almost no true muscular hypertrophy of the bowel wall. (2) A dilated and grossly hypertrophic sigmoid and a dilated *atonic* or *hypertonic* rectum. It may be argued that one cannot appreciate bowel tonus, but this is, to some extent, possible clinically. Some cases do not seem to fit into these categories. The dilated thin-walled sigmoid is typical of dolichocolon, as Hallenbeck & Waugh (1952) pointed out. The volvulus which occurs in these people, and may be confirmed by scarring of the elongated mesentery, is intermittent; hypertrophy does not occur to any degree. The rectum is often a little dilated and sigmoidoscopy is easy, but this may be a reflex atonicity resulting from the mesenteric twist. These cases respond to sigmoid resection. Their history is often suggestive, with periods of intermittent obstruction. The anus is normal in appearance and to digital examination. Rectal sensation and the response to rectal distension are normal also.

In the dilated hypertrophic sigmoid and rectum there is frequently a history of bowel actions perhaps once a month. This type is not common but suggests distal obstruction and the anus is usually competent and unrelaxing. Strangely, rectal discomfort is not marked and sensation may be diminished in spite of a high rectal tone but the sphincters are not easily inhibited. In these cases with sigmoid and rectal hypertrophy there is failure of sphincteric inhibition, the cause of which is unknown. It is difficult to see what treatment can help until the condition is better understood. There is, however, a failure of physiological function, that is of sphincteric inhibition with rectal distension by fæces; the X-ray appearance often shows a high degree of tone with good segmentation despite the dilatation.

The dilated hypertrophic sigmoid with the dilated atonic rectum is perhaps more common than the previous group. Again the anus is closed. These people have little or no rectal sensation and a failure of sphincteric inhibition, despite a huge degree of rectal distension. Such is a patient who walked about with four litres of fluid in the rectum and large bowel without sensation or leakage. The balloon with which an attempt was made to register rectal sensation burst in her rectum without any response and she was not mentally abnormal. The rectal wall sensory mechanism apparently failed though no bladder upset was apparent. The treatment of these cases is a problem too.

Before resecting a megacolon it is advisable to make sure that (1) ganglia are present in a rectal biopsy, (2) rectal sensation is adequate, (3) rectal volume is not excessive in order to produce a sensory and reflex response, and (4) that sphincteric inhibition occurs with rectal distension. These investigations are essential if sphincteric tone is normal. The only cases, if obstruction is excluded, where the condition may be due to bad habit, psychological or mental disturbance are those in which soiling of the clothes is present. These cases show some diminution of rectal sensation requiring an increased volume of fæces to initiate it, but the sphincters are inhibited normally.

Senile rectal atony, with its consequent fæcal impactions in which propulsion fails, exhibits inhibition of the sphincters. Thus it, too, should be automatically excluded from those cases where surgery is to be considered. A trial of Prostigmin

is often helpful and may give a promising response in functional megacolon. The results, however, have not been lasting.

Abnormalities of rectal physiology which are associated with megacolon and megarectum have been mentioned – the hypertrophic megarectum, the atonic megarectum and failures of sphincteric inhibition. Clearly these are abnormalities of rectal function and colonic resections do little to improve them; it may make them worse, as a relatively more hypertonic colon is anastomosed to a relatively inert rectum. Colonic resections for these abnormalities are likely to meet with the same result as they did in Hirschsprung's disease when surgeons failed to appreciate the presence of a rectal obstructing lesion. In a series of 14 cases of ileorectal, ileosigmoid and cæcorectal anastomoses, sigmoid and anterior resections, 9 still have a loaded rectum requiring enemata to clear them and 2 have diarrhœa. Six ganglionectomies have been carried out, together with other procedures, and no beneficial results have been noted. Abdomino-anal procedures seem to offer better results. Chronic volvulus with a relatively thin-walled colon responded to sigmoid resections.

Two cases bear out Ravitch's observations, one with latent cretinism and one with myxœdema. The bowel habit was improved and the megacolon became less marked with thyroid therapy.

Thus a complete classification of megacolon is reached (Table 1).

Table 1
Classification of megacolon and megarectum

(1) Obstructive: Tumours, strictures, volvulus, &c.
(2) Faulty habits, including psychiatric problems
(3) Endocrine: Cretinism, myxœdema
(4) Central nervous system: Spina bifida, paraplegia, cauda equina, &c.
(5) Peripheral nervous system: Absent ganglia, reduced ganglia, abnormal end-organs or reflex response (insensitive rectum, failure of sphincteric inhibition)

Finally, megacolon, when the bowel is opened perhaps only once a month, suggests a disorder of rectal, not colonic physiology. Hypertrophy, atony and failure of sphincteric inhibition are the most reliable signposts and colonic resections will not cure them.

I should like to thank my colleagues at St Bartholomew's and St Mark's Hospitals for allowing me to review their cases.

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Mr Harold H Nixon (*London*)
The classification of megacolon and megarectum that has been found useful in management, the treatment used and the results achieved, will be reviewed. Much is owed to my past chiefs and especially to Sir Denis Browne.

I regard all megacolon as secondary, as there seems to be no good evidence of a congenital primary overgrowth of the colon. The work of Swenson & Bill (1948) and of Bodian *et al.* (1949) was an important step in distinguishing the type which is now named Hirschsprung's disease. This is the megacolon secondary to congenital aganglionosis of the distal bowel. Its distinguishing feature is the hypertrophy of overactive bowel proximal to the obstruction caused by the absence of peristalsis in the abnormally innervated distal bowel. The other types of megacolon are largely characterized by a passive loading of the bowel. Table 1 summarizes the clinical contrast between Hirschsprung's disease and the other types.

Table 1
Clinical differentiation of Typical Hirschsprung's disease and rectal inertia

	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

The anus in Hirschsprung's disease feels normal. In rectal inertia it is often soiled, but the sphincter usually grips normally. Occasionally one sees a patulous anus. Indeed one child was referred as having a congenitally defective anus described as being 'like an ill-fashioned colostomy'. The rocks of fæces were manually evacuated and within a week or two the sphincter tone became normal, the mucosal prolapse from ineffective straining had settled, and the anus then looked and felt normal.

The Swenson type of resection has proved sound in Hirschsprung's disease although there are still problems of management. Duhamel's (1960) operation has not yet proved its worth. Rehbein's recent follow up of cases treated by State's operation was not very encouraging (Rehbein & von Zimmermann 1960). Table 2 shows the results I have obtained during the past five years.

The group of passive megacolon is a composite one with differing ætiologies even though the radiological appearances are similar. The terminal reservoir or megarectum is the usual picture. It

Table 2
Hirschsprung's disease: personal cases 1956–1960 (Total 68)

Operation	*No.*	*Cure*	*Controlled*	*Failure*	*Death*
Swenson type	51	39	6	2	3 (+1 later death)
Duhamel	10	8	1	–	1
Nixon	2	2	–	–	–
Died before definitive treatment possible	–	–	–	–	5

Follow up over two years in 41
14 had some operation elsewhere before referral

corresponds with the clinical picture I have called rectal inertia. The colon drives the fæces into the rectum but the rectum relaxes to accommodate more and more instead of being stimulated to evacuate. A mass fills the pelvis and in severe cases reaches up to the ribs. Even in such cases observation at operation has shown that the massively distended bowel is almost all rectum and that the colon above is remarkably normal. Because flatus is passed easily, distension is not a feature, the masses are easily palpable or even visible and the general health is reasonable except in a few cases after years of neglect.

Less commonly a tubular dilatation of the distal or whole colon is seen. Its significance is still not clear. Clinically the cases seem similar. Dolichocolon is a radiological picture but I am not convinced that it has any significant anatomical basis in a congenital lengthening of the colon. The use of tannic acid in the barium enema can eliminate redundant loops or demonstrate their ability to empty effectively.

Several conditions may produce rectal inertia. The largest group appears to be that following trouble with bowel training. If training is too strict, too lax or too inconsistent the child learns to hold back the stool until the urge to defæcate passes off. If the mother fusses the child and makes an emotional and social occasion of potting then the child learns to hold back to attract attention. The painful passage of hard stools which follows the delay, frightens the child into further holding back and a vicious circle may persist until the rectum is enlarged and unable to evacuate completely even if the child wishes. Thus constipation and progressive loading of the rectum may continue long after the exciting cause has passed. Overflow incontinence may supervene and fluid motions pass around the impassable rock.

Similar events may follow some painful condition such as an anal fissure or difficult conditions such as prolonged recumbency in the treatment of orthopædic defects; or congenital deficiencies, in particular anal stenosis or neurological lesions of spina bifida or sacral agenesis. Some cases have a deeper psychological basis but in most of those of functional origin it seems to be a training problem which may have resolved years before the patient is seen – the constipation persisting because of the secondary enlargement of the rectum.

Use of the term encopresis to cover two different conditions causes confusion. That in which there is incontinence in the absence of a neurological lesion – analogous to enuresis – is probably entirely a psychiatric problem though it may be associated with some constipation and rectal loading. But overflow incontinence in severe constipation with rectal inertia (megarectum) is different. It is often due to less serious functional causes or sometimes to organic ones; though a secondary functional overlay is common in a condition which causes such social embarrassment.

Treatment of the inertia group of megacolon and megarectum is based on two principles. First that the bowel must be emptied completely and kept so, long enough to recover its tone and shrink to enable its contractions to become effective. Secondly the child must be trained to regular habitual defæcation and not allowed to wait for the call to stool. For this call is likely to be deficient, and if the child is allowed to put off defæcation then relapse is sure. One case of rectal inertia was unconscious of the inflation of an intrarectal balloon with 10 oz of fluid. After three months' treatment 2 or 3 oz caused marked discomfort. Balloon tests have not given evidence of primary abnormality of sensation except in overt neurological lesions.

The following regime has been used: (1) Manual evacuation of hard masses under general anæsthesia. (2) Daily rectal washouts – up to 20 pints of saline, a few ounces at a time. *Not* reduced when spontaneous actions begin. Minimum of two weeks, then at increasing intervals over the next four weeks or more. Hypertonic phosphate enemas or Dulcolax suppositories may be substituted after the bowel has been emptied in some cases. (3) Aperients as soon as the masses are evacuated: Senokot, Dulcolax or Mist. Neostigmin. (Follow up over many years has not revealed that regular use of aperients leads to habituation and increased need.) Aperients are withdrawn gradually. (4) *Training in regular attempts at defæcation.* This can rarely be managed at home and it is unfair to expect the district nurse to do the washouts unaided. Enemas are often useless, distressing, and sometimes dangerous or fatal. Besides water intoxication there is the risk of enema collapse – which occurs in any type of constipation with masses of impacted fæces.

Because parental ineffectuality is so often the basic cause these patients and their parents are followed up for a period of years.

Table 3 shows the results achieved – about one-third relapsing and needing further treatment but few being persistently refractory. The only death

Table 3
Megacolon and megarectum: total 46 cases (1956–1960)

Apparent ætiology	*Controlled by* *Regime*	*Regime repeated*	*Operation*	*Failure*
Management ('training')	9	2	2	1
Psychological	3	–	–	1
Anal stenosis, &c. ('organic')	8	5	3	–
Precocious onset	8	–	1	1 (Died)
	28	7	6	3

Follow up over two years in 21 of the cases
2 cases not traced

was in the uncommon but interesting group of precocious, often neonatal onset.

Table 3 includes only severe cases requiring in-patient treatment. During the same period at Great Ormond Street I saw 48 cases of constipation which were less severe and were treated as out-patients. Barium enema would have shown a degree of megarectum in many of these but did not seem necessary.

A longer term follow-up is more convincing in such a condition. Eight years ago I carried out rectal sensation tests on another group of patients including 16 cases of rectal inertia. Mr J Cohen and Mr B Gampel have been able to contact 13 of these. Table 4 shows the results after eight years.

Table 4
Rectal inertia – 8-year follow up: 16 cases (13 traced)

Ætiology	*No.*	*Cured*	*Much improved*●
'Management'	7	6	1
'Psychological'	1	–	1
'Organic'	1	1	–
'Precocious'	4	3	1
	13	10	3

● Slight symptoms, insufficient to need further treatment

The results of the sensation tests are shown in Table 5. The perineal (rectal) sensation was often obtained only by a very large bolus up to 10 oz in the balloon instead of the usual 2 or 3 oz. The failure to produce more than abdominal colonic sensation in 8 is, I think, secondary to the enlargement and not due to primary neurological defect. For in the 3 which were retested after treatment rectal sensation had been restored.

Table 5
Sensation tests
(Intrarectal balloon distended with 1 oz increments of water)

5 cases: Rectal sensation normal though some to larger bolus than usual
8 cases: Colonic sensation only; of which 3 were retested after treatment and then had normal rectal sensation

Occasionally the anatomical enlargement of the rectum is such that it cannot revert to normal and relapse is inevitable. These are perhaps the ones met in adults. I have found segmental colectomy useless and have resorted to abdomino-anal resection of the rectum as for Hirschsprung's disease. There was a definite hypertrophy of the rectal wall. The operation is only a substitute for the first part of treatment. Training in regular defæcation habits is also essential – the more so as a low resection will reduce the specific rectal sensation. I think it is useless to operate until the child is old enough to co-operate in this after-care.

It is not satisfactory to resect so as to leave the 7 cm necessary for full rectal sensation because the bowel is ballooned right down to the anal canal and the retained rectal segment is sufficient to form the beginning of another megarectum. It did so in the one case in which I left some bowel. I did this because the child had a treated imperforate anus with an incomplete sphincter and I feared incontinence if I went too low. In another case I have narrowed the rectum by excising a section anteriorly instead of excising it all, because previous segmental colectomies had been carried out elsewhere. Relapse had occurred and there was no mobilizable loop of colon to bring down. It is too early to assess the result.

Less Common Types of Non-Hirschsprung Megacolon of Precocious Onset

First a rare type of which I have had two examples – one fatal. They imitated Hirschsprung's disease in infancy in many of their features, clinically and radiologically. Both babies failed to thrive. They had marked gaseous abdominal distension and constipation but visible colonic peristalsis was not prominent. The barium enema showed marked distension of the colon proximally with an undilated distal segment. Some improvement was obtained with rectal washouts. In both my cases a colostomy was performed but it was immediately evident that there was none of the colonic hypertrophy so typical of Hirschsprung's disease. The colostomies did not help. The proximal colon still did not evacuate. The first infant died with fæcal masses still in the right colon. In the second case the colostomy acted and then discharged excessive fluid. After treatment of the electrolyte disturbance the colostomy was closed and the child recovered and now progresses well. It seems that there had been a functional paralysis of the colon. Histological examination of the wall revealed no abnormality of the intramural plexuses, colitis or other abnormalities.

Dr M Bodian pointed out that the head circumference of the first case was small for her age and the brain showed cerebral atrophy. He suggested the possibility of a central origin for the abnormal

motility. The head of the second case is well below the 10th percentile also, being 15 cm at the age of 6 months.

Others of precocious onset have presented the picture of the terminal reservoir. But the history of constipation has reached back even to the neonatal period and clearly training had nothing to do with their onset. Gairdner (1960) has called this congenital constipation. Harris *et al.* (1954) has suggested that it is the result of slight anal stenosis relieved spontaneously by the passage of stools or the examining finger. Perhaps a central nervous factor should be considered for 2 of our cases had difficult births and the more florid cases of acute functional obstruction in cerebral birth trauma are well recognized.

It is tempting to consider that this syndrome might result from a very short Hirschsprung segment, so short that perhaps only the internal sphincter was involved and unable to relax in coordination with the defæcation wave of peristalsis. Biopsies have failed to show any abnormality and certainly when aganglionic segments as short as 2 cm have occurred they have presented the typical Hirschsprung symptomatology, A few internal sphincterotomies were performed in reservoir cases but the results were not impressive.

In these conditions I find it very difficult to decide what is cause and what is consequence, what is inborn and what is learned.

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Colonic Motility in Megacolon

The term megacolon indicates no more than the radiological or operative finding of a grossly enlarged colon. The diameter of the colon at barium enema, however, will depend not only on the state of the organ at the time, but also on such factors as the volume of fluid and the pressure head used together with the patient's toleration of the procedure. For this reason, it is necessary to restrict the definition to patients who have, in addition, a history of severe and prolonged constipation with or without overflow incontinence. In this study, the motility of the pelvic colon of 16 patients is described. The sigmoid colon was dilated in all 16 and all but 1 had a megarectum. Two patients had biopsy evidence of aganglionosis (of the rectal segment) and in another the clinical diagnosis was congenital megacolon. The diagnosis was confirmed at operation in 5 cases and at post-mortem in 1.

Methods and Normal Patterns

The motility of the pelvic colon was studied by three miniature balloons (7 mm diameter × 10 mm) arranged in series and recording on a metal capsule optical manometer (Rowlands *et al.* 1953). The tube was passed into the colon with the balloons in position at 25, 20 and 15 cm from the anus so that simultaneous records could be obtained from three positions. Details of the method have been published elsewhere (Connell 1961*a*).

In the normal subject the principal wave seen represents a slow contraction lasting between 20 and 30 seconds and of varying amplitude up to 100 cm of water pressure. The contractions are segmental and areas of the colon separated by 5 cm nearly always show independent activity (Fig 1). Peristalsis, recognized as a wave of contraction sweeping caudally in sequence over the three balloons, is rarely seen.

Results

Patients with megacolon demonstrate relative colonic hypomotility. It is doubtful if this group of patients is in any respect homogeneous, and motility patterns tend to support this. The motility records fall into four groups:

(1) *Generalized hypomotility:* Records in the resting state in this group show generalized hypomotility. There are occasional spontaneous waves, never of great amplitude (Fig 2). Some show low-grade activity almost continuously. This activity has no apparent propulsive purpose.

These patients were studied late in their disease and it is difficult to say whether this hypomotility represents colonic inertia or a phase of exhaustion secondary to prolonged overactivity. Two of the 7 patients had been using laxatives excessively for many years, one taking ½ lb of senna leaves a day. It seems possible that the hypomotility was secondary to excessive purgation. Another patient aged 21 had a short aganglionic segment and the colonic inertia may have been subsequent to prolonged attempts to overcome the resistance of this segment. No ætiological factors could be detected in the other patients who had a history typical of idiopathic acquired megacolon.

(2) *Colonic inco-ordination:* Three patients presented with persistent colonic activity in all three channels, but differed from normal in that the motility pattern of each segment was almost identical (Fig 3). This implied that the colon, while not inert, had been unable to develop its normal segmental activity, thus permitting the accumulation of a mass of fæces. Recording devices placed in this continuous column of fæcal material necessarily recorded identical patterns.

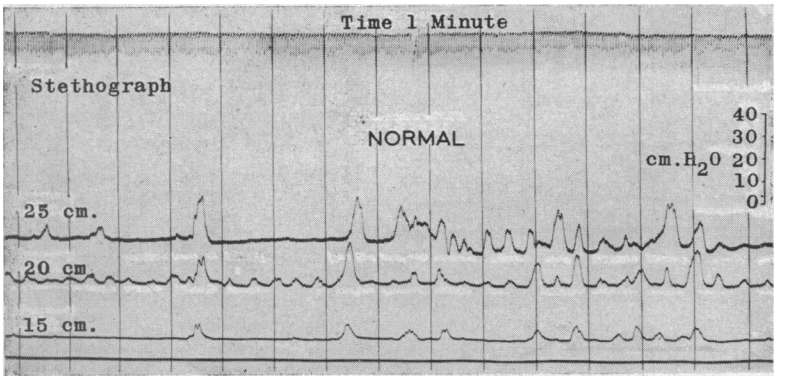

Fig 1 *Normal colonic motility showing segmental activity (reproduced from Connell 1959). In this and in all subsequent figures the lowest trace is from between 10 and 15 cm from the anus; the second from the bottom, between 15 and 20 cm from the anus; and the third from the bottom, between 20 and 25 cm from the anus. The upper trace is a respiratory record. Calibration in 10 cm steps, verticals = 1 minute in all figures*

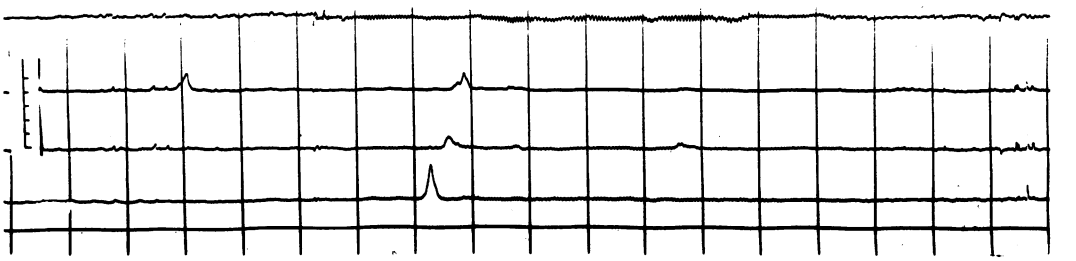

Fig 2 *Generalized hypomotility in megacolon*

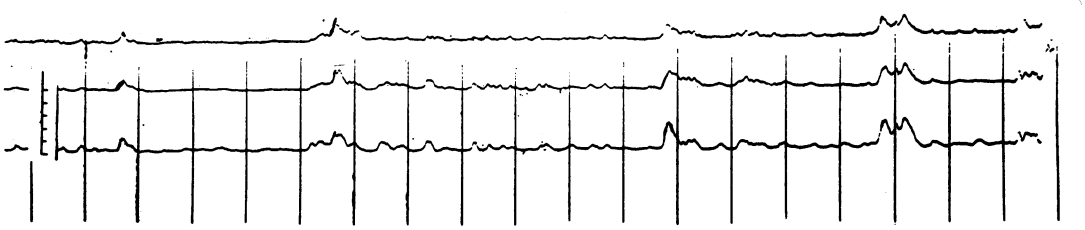

Fig 3 *Colonic inco-ordination in megacolon. The activity of each channel is similar*

Of these 3 patients one had a history typical of acquired megacolon, another, a boy of 16, had been diagnosed in childhood as Hirschsprung's disease on clinical and radiological grounds but had never had a full thickness biopsy. The third is of particular interest: she was aged 51 (a patient of Mr H Thompson's) and, following left hemiplegia, developed gross abdominal distension seen radiologically to be due to distension of the distal colon. The patient died of a myocardial infarct. A post-mortem showed that 25 cm of the descending colon was narrowed and hypertrophied but with normal mucosa; distally there was gross post-stenotic dilatation and muscular thinning of the sigmoid and rectum. Ganglion cells were seen in normal numbers in both areas of gut.

(3) *Segmental hypomotility:* A second group of 3 patients demonstrated inactivity in one segment only. Fig 4 is from a girl of 15 who was subsequently shown to have an aganglionic segment from the anus to 17 cm, although a barium enema did not reveal a constricted rectum. The lower trace is from the aganglionic segment. It is not amotile but shows low amplitude and apparently purposeless activity. The other 2 patients had different clinical histories: (*a*) A girl with acquired megacolon who had a subsequent resection of the colon which contained normal numbers of ganglion cells. (*b*) A middle-aged woman who was developing a megacolon at the same time as a progressive pyramidal lesion of all four limbs.

(4) *Normal records:* Three of the subjects had records which were normal in all respects. Each had an acquired megacolon but on crude testing 2 of the patients had grossly diminished rectal sensation without any abnormality of sigmoid sensation.

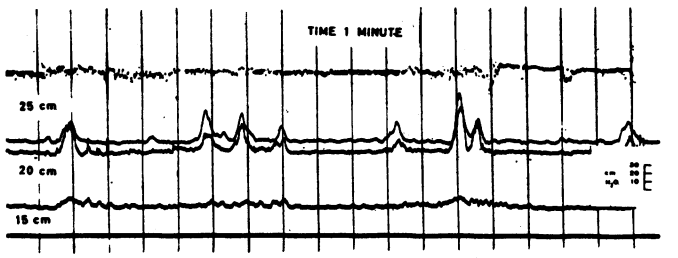

Fig 4 *Segmental hypomotility in megacolon. Only low-amplitude activity is seen in the lowest channel*

Effects of Drugs

(1) *Prostigmin*, in a dose of 0·5 mg S.C., has been studied in 7 patients. Five had depressed or inco-ordinated motility in all three channels. One had segmental hypomotility and one had an essentially normal record. With one exception all had acquired megacolon. In all but one the Prostigmin increased both the segmental and peristaltic activity. The difference is sometimes striking (Fig 5). The patient known to have an aganglionic rectal segment demonstrated the most marked increase in peristaltic waves (Fig 6) which coincided with the passage of soft fæces. Here it is probable that the Prostigmin augmented a response which was developing spontaneously. Almost certainly the recording points were above the aganglionic segment. One patient who had a normal basal record had no alteration of activity following injection of Prostigmin.

Of the 6 patients who had a good manometric response to Prostigmin, 4 are having a trial of neostigmine in a dose of 15 mg t.d.s. In a follow-up ranging from three months to over one year all are having satisfactory bowel actions on this regime. The other two did not have neostigmine but resection of the dilated bowel. Six months after surgery both patients again needed either laxatives or suppositories to achieve a satisfactory bowel action. The remaining patient who had a poor response to Prostigmin had a resection and was satisfactory four months afterwards.

(2) *Mecholyl:* Two patients, 1 a proved Hirschsprung's disease and the other suffering from an acquired megacolon, were given sufficient Mecholyl to produce systemic effects. In neither case was there alteration in motility.

Discussion

The intraluminal pressures of the colon have been determined. These represent one aspect of intestinal motility which is a term used to describe a number of different but related parameters, in-

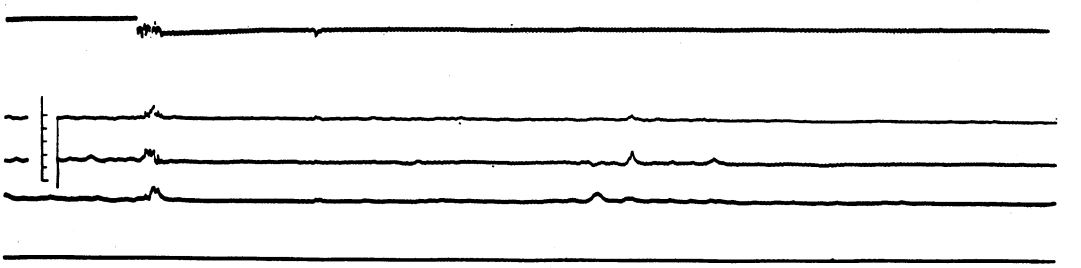

Fig 5A *Colonic activity before Prostigmin*

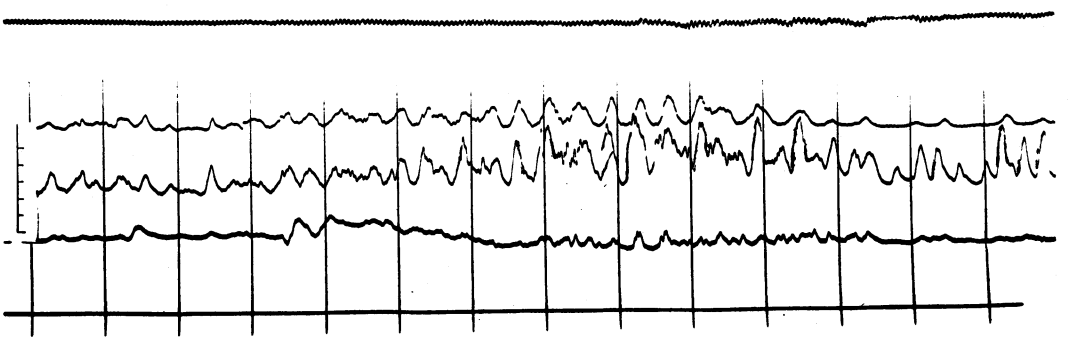

Fig 5B *Activity in same patient following 0·5 mg Prostigmin S.C.*

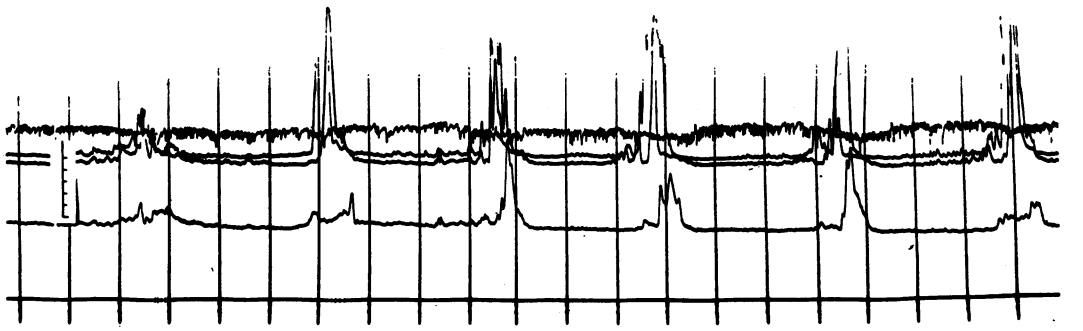

Fig 6 *Powerful peristaltic waves in patient with Hirschsprung's disease following 0·5 mg Prostigmin S.C.*

cluding transit through the bowel, the radiological or surgical appearances of bowel movements, intraluminal pressure changes, and bowel tone. In megacolon the tone of the colon is probably abnormal and information about this would be helpful; it is not possible to record the tone of smooth muscle directly in man. The other approach is to use miniature recording devices to study intraluminal pressures which reflect changes in tone although no absolute measures of tone can be made. It is often thought that the base line pressure of a motility trace is a measure of tone but this is not so as the most important component of the basal pressure is the hydrostatic pressure of the body tissue, and bowel tone appears to contribute very little to it. Understanding of the motor defect in megacolon will probably only be achieved when, in addition to measurements of intraluminal pressures, measurements of tone are also made.

These intraluminal pressures indicate that there is no defect of motor function common to all cases of megacolon. The whole group shows generalized hypomotility but there are important differences in different subjects which in this presentation have been grouped into four categories. This finding is consistent with the hypothesis that the anatomical abnormality of a megacolon may be common to a number of distinct physio-pathological processes. Generalized hypomotility and segmental inactivity have been seen in a minority of severely constipated subjects who do not have a megacolon (Connell 1961*b*). These disorders of motility may be better understood if considered functionally rather than anatomically. The anatomical abnormality may be the common end-result of a number of distinct pathological conditions such as colonic inertia, which may be either primary or secondary to excessive purgation, inco-ordinate colonic motor activity, colonic aganglionosis, and other obstructive lesions or diminished bowel sensation.

It is not possible to distinguish between colonic aganglionosis and megacolon resulting from other causes on the basis of intraluminal pressure measurements, but it has been suggested that some separation might be achieved on the basis of the colonic response to Mecholyl. Davidson *et al.* (1955) showed that in 4 of 6 patients with Hirschsprung's disease there was relaxation of the normally innervated bowel with no alteration in the motility of the denervated segments. In normal persons this differential relaxation was not seen. In the 2 patients tested with Mecholyl the results were not helpful but Davidson's observations warrant further assessment.

The disappointing results of surgery in acquired megacolon suggest medical treatment whenever possible. Although the number of patients in this series is small there was a good clinical response to Prostigmin where there had been a good manometric result. It is suggested that this Prostigmin response test is a useful guide to therapy in these patients.

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Megacolon: A Physiological Study

An experimental technique has been developed to investigate rectal physiology and provide measurable information about rectal activity, in contrast to the purely anatomical information obtained from barium enema studies.

Continuous electrical activity in the external sphincter ani has been demonstrated by electromyography (Floyd & Walls 1953). This is in marked contrast to striped muscle elsewhere (Fig 1) and is due to a continuous postural contraction which is unique in the muscles of the pelvic floor (Porter 1960).

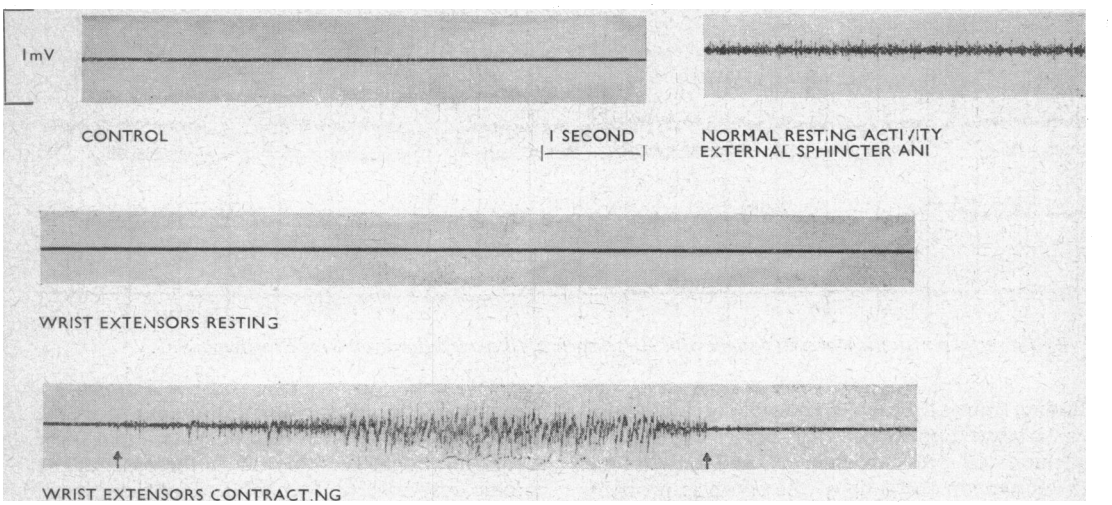


Fig 1 *The external sphincter shows continuous activity at rest. The wrist extensors are inactive unless a volitional contraction is made*

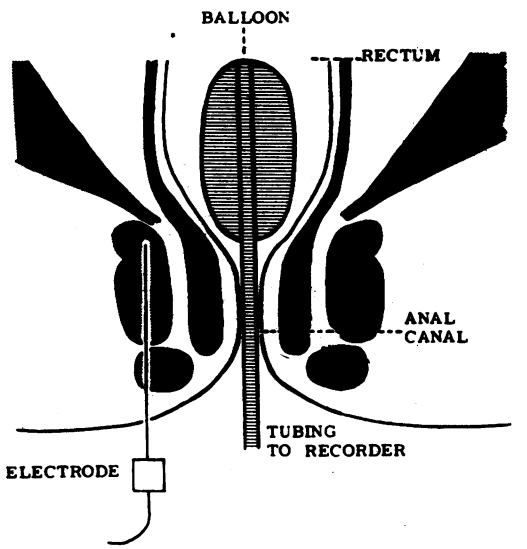


Fig 2 *A standard Miller-Abbott balloon lies in the rectum connected* via *a fine tube to a pressure recorder. A concentric needle electrode is inserted deeply into the external sphincter ani to record electrical activity*

This continuous electrical activity can be inhibited by rectal distension with a balloon (Fig 2), and the point at which it disappears recorded by means of needle electromyography. Inflation is carried out in 50 ml stages; initially the electromyogram (Fig 3) shows that the external sphincter is stimulated to increased activity; this is succeeded by progressive inhibition, which finally becomes complete.

Pressure changes within the balloon record the response of the rectal wall to distension (Fig 4A), which initially resists filling and then relaxes. These intrarectal pressure changes are conveniently expressed as a graph (Fig 4B).

In normal subjects rectal sensation, transient at first, always precedes inhibition of the external sphincter, which occurs with a volume of 150–200 ml, and a pressure of 45–55 mm Hg. The pattern of the rectal pressure graph (Fig 4B) is therefore constant.

In abnormalities of rectal function changes may be found in: (1) The threshold for reflex spincter inhibition. (2) The threshold for subjective rectal sensation. (3) The pattern of the rectal pressure graph.

It is worthy of note that in Hirschsprung's disease the responses to testing are normal. The sensory pathways for both conscious rectal sensation and reflex sphincter inhibition are therefore intact.

Idiopathic Megacolon

For convenience the cases of idiopathic megacolon have been classified by their radiological appearances: (1) Megarectum with a normal colon. (2) Megacolon with megarectum.

Megarectum with a normal colon: This group of patients presents early in life with a life-long history of constipation. Fæcal soiling and incontinence are common. Examination reveals a grossly loaded rectum and often a lax anal canal. The barium enema shows rectal distension with a normal colon above. Rectal biopsy is normal.

In two patients examined by the method described above the findings were grossly abnormal.

Case 1 Female, aged 17

The rectal pressure graph (Fig 5A) shows a slowly rising curve. This indicates a lax atonic rectal wall.

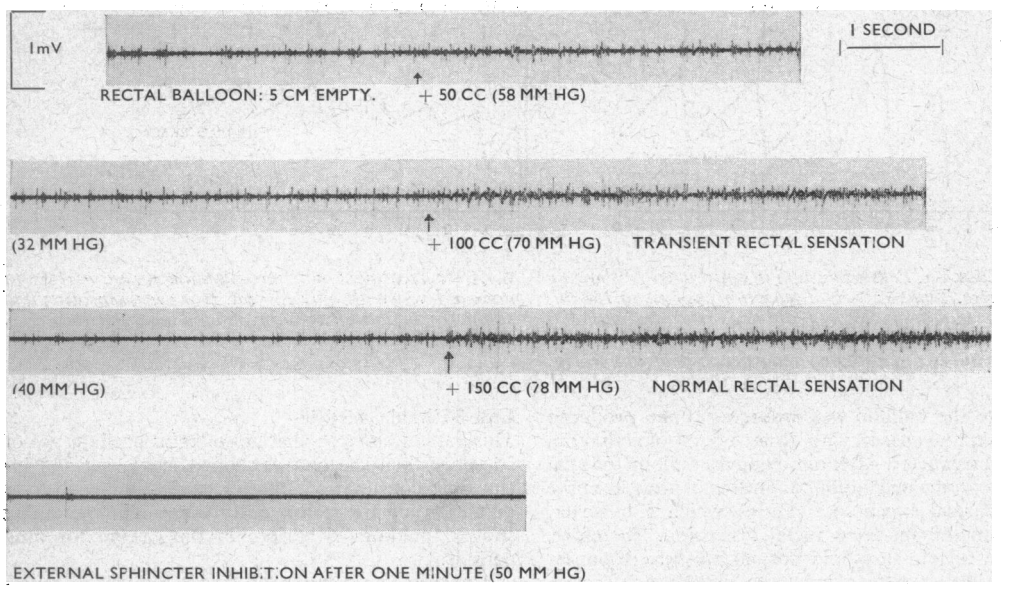
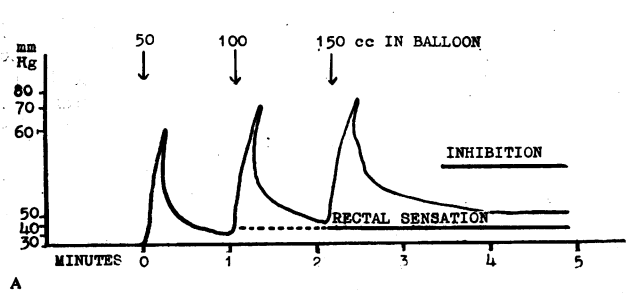


Fig 3 *Electromyogram of the external sphincter ani showing normal resting activity with an empty balloon in the rectum 5 cm from the anal verge. At each arrow 50 ml of air is injected into the balloon. This produces an increase in activity each time, which is succeeded by an increasing degree of inhibition. Inhibition is complete one minute after 150 ml has been injected. Rectal sensation precedes inhibition and increases in proportion to the degree of distension*



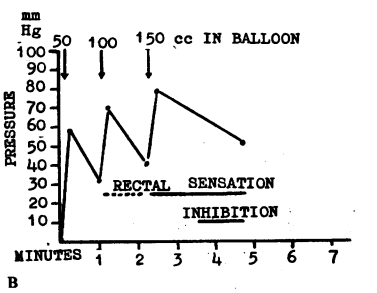


Fig 4A, *Pressure tracing recording intrarectal pressure changes in response to balloon distension in a male aged 34. At each arrow 50 ml of air is injected into the balloon. Initially the rectal wall resists distension and a peak is obtained; as the rectal wall relaxes the pressure falls to a new resting level over the course of a minute. Transient rectal sensation is represented by the broken horizontal line, the normal urge to defæcate by a solid line. Inhibition of the external anal sphincter is marked by the solid line labelled 'Inhibition'.* B, *Pressure graph expressing the pressure tracing plotted against time in minutes. It shows that the normal intrarectal pressure curve rises relatively steeply to 50 mm Hg with 150 ml in the balloon*

Rectal sensation and inhibition of the external sphincter are present, but the threshold for both is raised. The anal canal is lax.

After treatment with neostigmine for three weeks this patient had a regular bowel habit. Re-examination (Fig 5B) showed that rectal tone had improved, but the threshold for sphincter inhibition remains at a high level. Sensation is still impaired.

Case 2 Female, aged 21

This patient obtained a bowel action four times a day by means of oral Dulcolax. The signal for defæcation was abdominal colic and not rectal or perineal sensation. The rectum contained a large fæcal mass of which she was unaware and which she was unable to pass, in spite of marked anal laxity.

The electromyogram showed complete inhibition of resting activity in the external sphincter and puborectalis, undoubtedly due to fæcal distension of the rectum, which thus simulated the balloon in our experiments.

On inserting a balloon into the rectum a little continuous activity was induced in the external sphincter. This showed a slight increase with each 50 ml addition of air until 150 ml was reached, when external sphincter inhibition reappeared. In spite of this, an attempt to

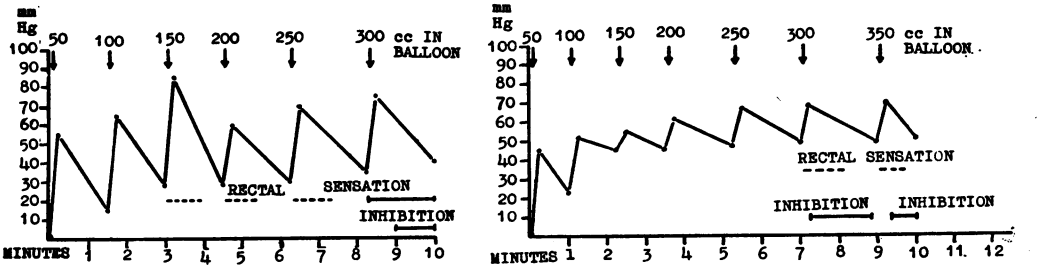


Fig 5 **Case 1** A, *Pressure graph in a girl aged 17 showing a slow rise. Transient rectal sensation appears at 150 ml, but a normal desire to defæcate does not arise until 300 ml have been added. Inhibition of the external sphincter appears at the same volume and a pressure of 40 mm Hg.* B, *After treatment with neostigmine pressure rises to normal levels with only 150 ml. However, sensation and sphincter inhibition require 300 ml and the former is only transient*

defæcate the balloon was unsuccessful and produced anal pain and spasm, with a sharp burst of activity in the anal sphincter. After removing the balloon the anal sphincter remained inhibited. The rectal pressure curve has a normal appearance (Fig 6), which is, however, superimposed on gross rectal distension. Therefore, above a certain threshold normal pressure responses and inhibition appear, but rectal sensation is grossly impaired.

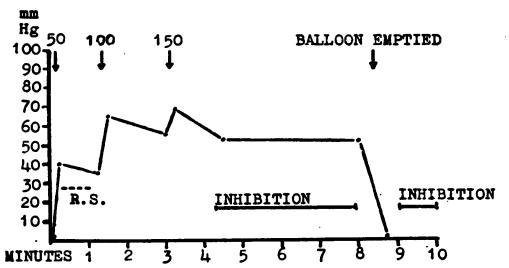


Fig 6 **Case 2** *The pressure curve rises to normal levels at 150 ml; inhibition of the external sphincter appears at this volume and a pressure of 50 mm Hg. On emptying the balloon sphincter activity returns temporarily, but is succeeded by inhibition again. Rectal sensation is transient with the first 50 ml only*

The experimental findings in these two patients are complementary. Rectal filling is hardly noticed at conscious level and sphincter inhibition requires a grossly abnormal fæcal mass. This is usually too large and hard to pass, in spite of the anal relaxation it produces. The attempt to defæcate stretches the anal canal abnormally, producing pain and spasm, and the attempt is abandoned. However, further filling above this level can initiate partial defæcation provided the fæces are soft. If, on the other hand, they are liquid the patient is incontinent as the external sphincter is already inhibited and rectal sensation is impaired.

Megacolon and megarectum: Two patients examined in this group provide a striking contrast physiologically, in spite of a similarity in their clinical histories and barium enema studies.

Case 3 Female, aged 36

This patient had a normal bowel habit until the age of 25, since then she had developed increasing and intractable constipation. Barium studies showed gross dilatation of the colon and rectum and screening showed pooling of barium in the cæcum for long periods.

The rectal pressure graph (Fig 7A) shows an extraordinary degree of rectal atony. The inhibitory threshold is enormously raised and rectal sensation is absent. Neostigmine produced neither significant rectal contraction nor clinical improvement.

Case 4 Male, aged 39

This man had a normal bowel habit until the age of 33. He then became increasingly constipated. The barium enema showed a similar degree of dilatation to that in the previous case, but no pooling in the cæcum.

The rectal pressure graph (Fig 7B) provides a striking contrast to that in Case 3. The rectum is hypertonic and hyperactive. The rectal sensory threshold is low and that for sphincter inhibition is normal.

Neostigmine restored this patient's bowel habit to normal. It is difficult to reconcile the experimental findings in this case with the radiological picture of megacolon. They are unlike our findings in any other patients in this series.

Discussion

The rectal pressure graph provides an objective index of rectal muscle function provided it is related to inhibition of the external sphincter, rather than subjective rectal sensation which is variable and sometimes absent. The response to treatment can be gauged from a series of graphs. Neostigmine may convert the rectal pressure curve to a normal pattern.

Eight of the patients investigated were treated with neostigmine; in one of them with extreme atony it failed to produce clinical improvement. It appears to act by improving peristalsis and increasing rectal tone. There is no evidence that it reduces the threshold for inhibition of the external sphincter when this is high.

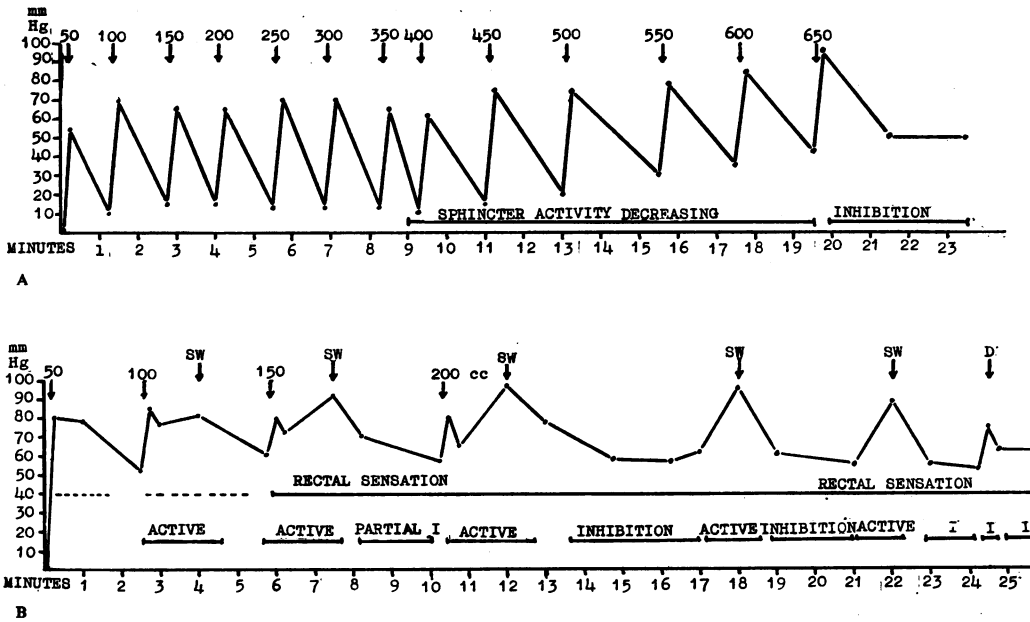


Fig 7A, **Case 3** *Intrarectal pressure fails to rise until 400 ml has been injected. Thereafter it rises slowly with increasing sphincter inhibition. This becomes complete at 650 ml and 50 mm Hg pressure. Rectal sensation is absent.* B, **Case 4** *Pressure rises steeply with only 50 ml; transient sensation appears at this volume. Above 100 ml rectal filling evokes rectal contractions: 'SW'. A normal urge to defæcate is experienced at 150 ml and inhibition is complete at 200 ml, though this is interrupted by each rectal contraction, but returns as the rectal wall relaxes once more. At 'D' the patient attempted to defæcate the balloon but failed, in spite of inhibiting the external sphincter on bearing down*

It is apparent that no constant pattern of behaviour in patients with idiopathic megacolon has been forthcoming from this study. This must be a stimulus to further research on a tantalizing problem.

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Megacolon and Megarectum in Chagas' Disease[1]

The object of this paper is to present the results of the surgical treatment of megacolon and megarectum in Brazil and a summary of recent contributions by Brazilian authors which have brought about a better ætiological and physiopathological knowledge of the disease. This presentation is an account of work carried out in the Medical School of Ribeirão Prêto.

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Nomenclature

Numerous eponyms have been proposed for the condition. Obviously 'idiopathic dilatation' had to be abandoned as soon as the pathogenesis of the disease was more clearly understood. 'Achalasia of the colonic sphincters' is not sound, since the existence of such sphincters, both from the anatomical and from the functional standpoint, is questionable. According to our clinical and surgical experience, no achalasic zone or ring has been found distal to the dilatation of the sigmoid colon or rectum. The terms 'megacolon' and 'megarectum' are acceptable on a roughly descriptive basis and because of tradition inasmuch as they refer to the colonic and rectal dilatation which constitutes an advanced stage of the disease. In early stages of this disease patients have a functional abnormality without dilatation of the intestine, and for this reason the terms megacolon and megarectum are not accurate.

According to our present knowledge of the disease, the better name is 'aperistalsis' of the colon and/or rectum. This term was suggested by Brasil (1955) for a similar condition occurring in the œsophagus and was extrapolated for the disease of the large bowel. It was officially recommended by the Brazilian Federation of Gastro-

enterology in 1959. 'Aperistalsis' is adequate because of its good physiopathological connotation: it means motor inco-ordination, defective motility, and disturbed or absent peristalsis without propulsive efficiency. This is what happens in the so-called acquired form of megacolon: the fæcal stagnation is a consequence not of distal mechanical obstruction but of a functional inco-ordination of the muscular activity in a long intestinal segment including the dilated bowel.

Incidence

A partial survey of the autopsy records in the Medical School at Ribeirão Prêto showed 69 cases of megacolon out of a series of 556 autopsies not including casualty deaths nor infants less than 1 year of age. In the Department of Surgery, 58 patients with rectocolonic aperistalsis were operated upon between March 1955 and April 1961. In this Department 4 beds in a ward of 30 are permanently occupied by such patients. If a selective policy for admissions were not adopted, all beds could be occupied by them and by those with aperistalsis of other hollow viscera, especially the œsophagus. This is a reflection of the high prevalence of the disease in Brazil, especially in the central areas.

In Brazil the geographical areas in which cases of aperistalsis are observed closely coincide with those where Chagas' disease (*Trypanosomiasis americana*) is endemic, and the majority of patients with megacolon and megarectum present the immunological characteristics and the electrocardiographic patterns of chronic Chagas' disease. Similar observations have been made recently in Chile (Atias *et al.* 1959) and Argentina (Rosenbaum & Cerisola 1958). There are reasons for believing that a methodical investigation of œsophageal and bowel dynamics in other countries in which Chagas' disease is known or has been observed (South and Central America and the Southern part of North America) would disclose a similar relationship.

Mega-œsophagus and megacolon, as segmental dilatations of the alimentary canal, are advanced pathological phases of a systemic disease and frequently occur together. In the above-mentioned autopsy series, there were 62 cases with mega-œsophagus of which 31 (50%) also had megacolon; out of 69 cases with megacolon 19 (27·5%) also had mega-œsophagus. In our series of 58 surgical patients with aperistalsis of the colon and rectum, 48 have had radiological examination of the œsophagus: 23 (47·9%) showed œsophageal aperistalsis and most of them had degrees of dilatation (mega-œsophagus). Recent investigations with myographic recordings (Vieira & Godoy 1961, personal communication; Vieira, Godoy & Carril 1961, personal communication) revealed a typical positive response by the œsophageal musculature in most cases of megacolon, even when there was no complaint of dysphagia. The reverse was observed, namely a positive response by the colon in cases of mega-œsophagus, either with or without obvious constipation.

Besides the œsophagus and the distal large bowel, other segments of the digestive tract and other hollow viscera present aperistalsis and may eventually become dilated. In this autopsy series, there were 5 instances of megagastrum, 2 of megaduodenum, 1 of megajejunum and 2 of megavesicula (dilatation of the gall-bladder). We have also seen megaureter. Similar findings have been reported by Etzel (1942) and Raia (1943).

Aperistalsis of the colon and rectum occurs at any age and in either sex. In our surgical series, the oldest and the youngest patients were 64 years and 10 months old respectively. The average age was 34·7 years. In our experience megacolon in infancy and childhood is not always of the Hirschsprung type. We have observed two children with 'megacolon': one of them had microrectum, diaphragmatic occlusion of the sigmoidorectal junction and dilatation of the left colon; the other, 10 months old, had megacolon and megarectum. Pathological studies have confirmed that the first had Hirschsprung's disease and the latter Chagasic aperistalsis.

No special study was made of race or occupation, but the majority of the patients come from rural districts. Similar observations were published by Freitas Jr. (1950) and Rezende (1956).

Pathology

Since the third decade of this century, when degenerative and inflammatory lesions in the myenteric plexuses were first described (Hurst 1925, 1934, Etzel 1937), there have been many contradictory ætiological theories, but there is unanimity in ascribing an important role to the destruction of the ganglion cells as the causative mechanism of œsophageal or colonic dysfunction and dilatation. Some authors, quoted by Cutait (1953) and Raia (1955), describe a distal aganglionic segment in all cases of megacolon. According to them, there is a functionally strictured distal segment, the rectum, in which the ganglion cells are absent, in contrast to the proximal dilated sigmoidal loop, in which they are present. This is true for Hirschsprung's disease, but in our experience at Ribeirão Prêto this is not true of aperistalsis or acquired megacolon and megarectum. In these there is a reduction in the number of ganglion cells throughout both the rectum and the colon. Moreover the quantitative reduction of the ganglion cells is observed in all segments of the alimentary tract, with the eventual appearance of other enteromegalic segments, especially in the

œsophagus (Amorim & Corrêa Netto 1932, Köberle & Penha 1959).

The conflicting findings and interpretations of the histopathological features of 'aperistalsis' are understandable and have a possible explanation in the following facts: (1) Most pathologists concentrate on the qualitative changes in the myenteric plexuses and do not pay attention to quantitative analysis. (2) The samples submitted to microscopic examination are not representative. As a rule, the specimen is either a small fragment taken arbitrarily from any part of the surgical or autopsy specimen, or a thin strip of the muscular layer of the sigmoidorectal junction or the œsophageal cardia obtained during a non-excisional operation, as, for example, the pelvirectal sphincterotomy (Corrêa Netto's procedure) and the gastro-œsophageal extramucosal myotomy of Heller. With such sampling, normal and abnormal nerves may be found or none at all. The result is inconsistent and inconclusive.

Köberle (1956, 1959*a*, *b*) and his assistants at Ribeirão Prêto have performed a quantitative analysis of the ganglion cells in serial sections of many blocks of various representative tissue fragments taken from each surgical or autopsy specimen. Ring segments 1 mm high of the circumference of the intestine and œsophagus, with serial sections from each ring, showed the following average numbers of ganglion cells.

For the colon and rectum, at various levels:

	Cæcum	*Transverse*	*Sigmoid*	*Rectum*
Normal	4,163	4,947	5,785	4,036
Megacolon	189	253	381	163
Percentage of normal	4	5	6	4

For the œsophagus, at three different levels:

	Upper	*Mid*	*Lower*
Normal	445	778	1,003
Chagas' disease with apparently normal œsophagus	122	261	391
Mega-œsophagus	0·1	1·5	2·7

This striking reduction of the ganglion cells has been consistently found in our specimens of the colon and rectum (Köberle 1956, 1957) as well as for the œsophagus (Köberle & Penha 1959). It is also observed in the heart (Alcantara 1959) and, but less markedly, in the remaining segments of the digestive canal and in the wall of other hollow viscera, e.g. the biliary ducts, the ureters, and the bronchi (Köberle 1959*b*).

The partial or complete aganglionosis of the œsophagus and of the colon was formerly ascribed to a nutritional deficiency of vitamin B_1 (Etzel 1935). But Chagas (Chagas 1916, Chagas & Villela 1922), in his original descriptions of the disease, pointed out the frequent finding of enteromegalies and suggested that the *Trypanosoma cruzi* could be responsible for them. Other Brazilian authors have emphasized the similar geographical distribution of Chagas' disease[1] and the enteromegalies as well as the high incidence of positive complement-fixation tests for *T. cruzi* in patients with mega-disease. Mayer & Rocha-Lima (1914) reported the finding of the parasite in the stomach wall of monkeys and Köberle & Nador (1956) presented further morphological support for this ætiological relationship by showing that the muscular fibres of the hollow organs harboured pseudocysts of the parasite in the leishmania phase: the pseudocysts rupture, causing an inflammatory reaction in the tissues between the two muscular layers with partial or complete destruction of the ganglion cells. Köberle and his co-workers also demonstrated, in human Chagas' disease and in rats inoculated with *Trypanosoma cruzi*, that the quantitative and qualitative changes of the nerve cells occur not only in the intestine but also in the heart (Alcantara 1959), in the bronchi (Köberle 1959*b*) and in the spinal cord (Schwartzburd & Köberle 1959). Köberle (1956) suggests that a neurotoxin liberated after the rupture of the pseudocysts and the disintegration of the leishmania forms of the *Trypanosoma cruzi* is the cause of the degenerative changes in the ganglion cells.

Other toxic factors may be responsible for such lesions and for aperistalsis under certain special conditions and in the countries where Chagas' disease does not occur. Worms & Leroux-Robert (1934) ascribed the ætiology of some cases observed in France after World War I to gas poisoning (Yperite). And Erwenich (1940) suspected that the ganglion cell destruction was caused by diphtheria toxin.

In South America, however, aperistalsis and enteromegaly are a manifestation of Chagas' disease. In Ribeirão Prêto we believe that this will be soon confirmed in other areas. The papers

[1]The *Trypanosoma cruzi* is the causative agent of Chagas' disease and is transmitted to man by blood-sucking insects belonging to the subfamily triatominæ. When the infected insects bite an individual there may be contamination of the wound by the insect's fæces, which contain numerous parasites. The vector chiefly bites the face and this is the reason for the nickname 'barbeiros' (barbers). There is multiplication of the trypanosome at the point of the initial infection. A few days later, the parasites disappear from the blood stream and invade smooth muscles and the myocardium. They multiply in these tissues in the form of leishmania and cause destruction of the adjoining tissue with the formation of cyst-like cavities. When these pseudocysts rupture, most of the leishmania are destroyed by the natural defences of the body but some of them reappear in the circulation. This cycle is repeated many times during the first weeks of this acute phase of the disease, during which diagnosis can be confirmed by direct examination of blood smears or by means of the 'xenodiagnosis', that is, allowing a laboratory-bred insect to bite the patient with examination of fæces ten days after. The symptoms are those of an acute infection, with fever, generalized adenopathy and a slight enlargement of the liver and spleen. Moreover, there is cardiomegaly with arrhythmia. About 10% of the patients die. The patients who survive go into the chronic phase of Chagas' disease which is mainly characterized by heart block, also dysfunction and dilatation of many hollow viscera. During this phase diagnosis can be confirmed by a complement-fixation test (the Machado-Guerreiro's test). Up to the present time no specific treatment for acute or chronic Chagas disease has been discovered.

by Verbrycke (1920), Vinson (1947), Templeton (1948), Emert (1952) and Bates (1952) on mega-œsophagus and megacolon, and those by Packchanian (1939) and Woody & Woody (1955) on trypanosomiasis seem to foretell this, since they come from or refer to the same geographical areas – the southern part of the United States. A few months ago Ingelfinger reported to Freitas (1960, personal communication) 4 patients with œsophageal aperistalsis observed in Boston, Mass. All had lived in the Southern States and one presented the same electrocardiographic pattern described in South American Chagasic patients. It seems desirable that every person with a positive complement-fixation (Machado-Guerreiro) test be submitted to a thorough radiological study of the œsophagus and of the large bowel. Moreover, every patient with rectocolonic or œsophageal aperistalsis should be investigated by ecologic anamnesi and a Machado-Guerreiro test (Freitas 1947).

The results of the latter in the Ribeirão Prêto series are quite impressive. The complement fixation was positive in 96·7% of the autopsy cases with enteromegaly and in 90% and 94·4% of the surgically treated cases of œsophageal and rectocolonic aperistalsis.

Another fact which favours the Chagasic ætiology of mega-œsophagus, megacolon and megarectum in Brazil is the observation made by Rezende & Rassi (1958) that, three to twelve months after the acute phase of Chagas' disease, the œsophagus, previously normal at fluoroscopy, shows radiological features of motor inco-ordination and dilatation, with the corresponding complaint of dysphagia. In 1950, similar findings were made by Nunan *et al.* (1952) in 13 children with megacolon and a previous diagnosis of acute Chagas' disease.

In 1959 Guimarães & Miranda described mega-œsophagus with leishmania in the organ wall in a rhesus monkey inoculated ten years before with *Trypanosoma cruzi*. Okumura (1960, personal communication), in São Paulo, has claimed the production of megacolon and mega-œsophagus in animals (dogs and mice) with experimental Chagas' infection.

The *Trypanosoma cruzi* is a parasite of muscular tissue but damages the ganglion cells, possibly through a neurotropic toxin. During the acute phase of Chagas' disease parasitic metastatic implants take place in variable degrees of intensity and frequency and with a variable topographical distribution. There is a local inflammatory reaction, formation of granulomas and the rupture of the pseudocysts with interstitial inflammation and toxic destruction of ganglion cells, the numbers of those destroyed depending on the intensity of the infestation. This accounts for the variability of the qualitative and quantitative morphologic features in a casual single small piece of tissue obtained in the chronic phase of the disease: whether there is further ganglion cell damage during this period is doubtful and merits further investigation.

The motor dysfunction is a consequence of ganglion cell destruction and denervation. Denervated muscular tissue, including the muscular tissue of the œsophagus and colon, when partially or totally deprived of its nervous control, becomes hypersensitive. Kramer & Ingelfinger (1951), with Mecholyl stimulation of the œsophageal musculature, demonstrated this hypersensitive response in cases of so-called 'cardiospasm'. With kimographic recordings of intraluminal pressure variations at various levels, our group in Ribeirão Prêto showed hypertonus and increased contractility after Mecholyl stimulation in Chagasic patients either with or without dysphagia as well as actual dilatation of the œsophagus (Vieira & Godoy 1961, personal communication). Our group has obtained a similar pattern in the large bowel, either with or without megacolon and megarectum (Vieira, Godoy & Carril 1961, personal communication). The colon reacts less intensely than the œsophagus to similar doses of Mecholyl and we do not know yet whether such a difference can be attributed either to the relatively less extensive neuronal destruction in the wall of the large bowel compared with the œsophagus or to a greater local production of cholinesterase in the former, as suggested by Fink & Friedman (1960). According to these findings, the hyperreactive Mecholyl test can be considered as an objective demonstration of the enteric motor dysfunction in Chagas' disease. The fact that the response has been obtained at various levels and even in the early and in the mild forms of the disease suggests that the neuronal degeneration and the corresponding motor inco-ordination are diffuse from the beginning of the disease.

The first dysfunctional stage is, therefore, hypertonus and hypercontractility, with or without clinical manifestations, depending on the degree of neuronal destruction and of the consequent motor inco-ordination (aperistalsis). This inco-ordination can be witnessed by fluoroscopy or during surgical exploration, and appears as scattered, arrhythmic, localized rings of contraction without any order or peristaltic efficiency. It is easier to see this in the œsophagus than in the colon or rectum.

As a result of motor inco-ordination and exaggerated reactivity, muscular hypertrophy soon follows. The muscular wall, although hypertrophied, has no propulsive effectiveness and there is stoppage of the intestinal content with

progressive dilatation and elongation of the loop (dolichomegacolon). This means stretching and mechanical stimulation of the muscular fibres, with the consequent additional hypertrophy, especially of the circular layer. The stagnant fæces are progressively desiccated and hardened (fæcaloma), leading to mucosal inflammation, ulcers and perforation.[1] In our surgical specimens of the dilated rectosigmoid we found no other lesions than inflammatory reaction and stasis ulceration. One patient with advanced aperistalsis of the colon and œsophagus died from peritonitis due to perforation of an ulcer in the sigmoid colon. Another complication of megacolon is acute volvulus, with or without interruption of the blood supply to the twisted sigmoidal loop. The tendency to volvulus is caused by tremendous elongation of the sigmoid colon with a characteristic thickening and retraction of the mesenteric tissues, probably a result of lymphangitis.

Two points merit discussion:

(1) Why is enteromegaly more often seen in the rectosigmoid and œsophagus, when the destruction of the ganglion cells is observed throughout the digestive tract? A possible explanation is that these two segments are more exposed to mechanical stress, since they receive and contain respectively the desiccated fæcal bolus and undigested food. The content of the remaining segments of the alimentary tube is more fluid. Mechanical burden is important. After proximal colostomy is performed, the resting 'megaloop' undergoes a striking regression. Similarly development of Chagasic cardiomegaly depends not only on cardiac denervation, but also on the work imposed upon the heart.

(2) Is there an achalasic or a strictured distal segment? Our experience gives no support to the theory of sphincteric achalasia of the rectum and of the colon. We have never found a real organic or functional distal stricture. In Hirschsprung's disease, it seems to be true that the aganglionic distal segment is functionally strictured. In Chagasic rectocolonic aperistalsis this is not found. We believe that the apparently narrow distal segment seen radiologically corresponds to the terminal part of the rectum that cannot dilate because dilatation is hindered by the neighbouring structures, particularly the pelvic musculature. For the distal œsophagus, a similar role is played by the ring and crura of the diaphragmatic hiatus.

Clinical Features

The outstanding symptom of aperistalsis of the colon and rectum is chronic constipation. Bowel movements occur with an interval of eight days to five months and must be induced by laxatives and enemata. The patient's tolerance to such prolonged constipation is astonishing. The lodged fæcaloma sometimes requires manual evacuation under anæsthesia (twenty-eight times in our series). It is often easily palpable through the abdominal wall. Rectal palpation detects dilatation and the impacted bolus. Malnutrition is a consequence either of anorexia and bad habits or of concomitant œsophageal involvement. In the latter case, there is a dysphagic complaint.

The chronic intestinal constipation is well tolerated. It becomes acute if a volvulus of the sigmoid colon complicates the picture. The diagnosis of the latter is straightforward. On six occasions (10·3%) acute volvulus caused admission to hospital, and in 2 patients there was a history of this complication.

X-ray studies are performed by barium enema, after removal of the fæcaloma. Fluoroscopic evaluation is made and films are taken before and after spontaneous evacuation. Contractility, tonus, motor co-ordination and emptying efficiency are evaluated. There is no indication for rectoscopic examination for either diagnosis or treatment, except in the special case of partial volvulus. For decompression, a tube is passed beyond the twisted rectosigmoidal junction.

It is worth emphasizing that the patients with Chagasic aperistalsis of the colon and rectum may have not only œsophageal involvement but also (1) dilatation of other segments of the digestive and urinary tract and the corresponding symptoms, (2) a cardiac condition (Ferreira-Santos *et al.* 1959), and (3) hypertrophy of the salivary glands (Vieira 1958).

Treatment

There is no curative treatment for 'aperistalsis' because the fundamental lesion, namely the destruction of ganglion cells, is permanent. Treatment is palliative, and recurrence after surgical treatment depends on the amount of damage to ganglion cells in the remaining intestine.

Lumbar sympathectomy, splanchnicectomy and pelvirectal sphincterotomy were used in the past for treating aperistalsis of the colon and rectum, but almost invariably failed. The present knowledge of the pathogenesis of the disease does not offer a sound basis for operation. Resection of the presumptive neurovegetative supply to the dilated loop increases the pre-existing intramural denervation. The external incision of the pelvirectal (Corrêa Netto 1934) and internal anal sphincters is as incomplete as Heller's operation for advanced mega-œsophagus. For many years Brazilian surgeons, including the

[1] In the œsophagus, the stagnation of food causes stasis œsophagitis, ulcers, bleeding, leucoplasia, diverticula, fistulæ and, eventually, carcinoma.

author, performed this sphincterotomy. Recurrence was the rule.

The best therapeutic approach to the problem is rectosigmoidectomy (Cutait 1953, Raia 1955, Ferreira-Santos 1959). The resection includes the dilated sigmoid and rectum with the exception of the distal 4–5 cm of the latter. Sigmoidectomy is unsatisfactory and leads to recurrence with dilatation of the left colon now anastomosed to the untouched aperistaltic rectum (Finochietto 1927, Riveros 1940). It is important to remove the entire denervated segments showing motor dysfunction, muscular hypertrophy, dystonia and dilatation, i.e. the part of the denervated intestine which has continuously endured the traumatic action of desiccated solid fæces. Even so recurrence sometimes occurs. At operation the surgeon cannot know the intensity and extension of the quantitative neuronal reduction in the remaining colonic segments.

The method used to complete the anastomosis is a matter of personal taste. Our experience includes:

Internal anal sphincterotomy		1
Right hemicolectomy		1
Left hemicolectomy		1
Total colectomy		1
Abdomino-perineal rectosigmoidectomy:		5
With Swenson's perineal anastomosis	3	
Intra-sphincteric pull-through operation	1	
Endo-anal pull-through operation	1	
Anterior suprapubic rectosigmoidectomy:		49
As the single operation	45	
The same plus anal sphincterotomy	4	
Total		58

Rectosigmoidectomy has been used more frequently for obvious reasons. Other colonic segments are resected when dilated, but this is exceptional. There are cases with total megacolon. Whether it is advisable or not to perform a total colectomy remains a controversial matter. In one case a total colectomy with ileoproctostomy was carried out, leaving behind almost the entire rectum, so that the latter would act as a reservoir and avoid the disadvantages of perineal ileostomy. The patient is doing well after more than two years, with two or three bowel movements a day. The prognosis is not good, however, as the patient had mega-ileum at the time of his operation. The second case is more recent, was submitted to a conventional anterior rectosigmoidal resection and is under observation.

When rectosigmoidectomy is contemplated, the anterior suprapubic procedure is preferred for all cases except young children. In these the Swenson technique is easier.

To avoid post-operative complications, an important point is to perform a three-staged operation, with primary colostomy of the transverse colon as the first step. Before the adoption of this policy dehiscence was frequent. Nine out of 18 anastomoses (50%) were complicated by fistula formation on the fourth post-operative day: all but one were treated by temporary colostomy, 2 had to be drained through the perineum and 4 which developed stenosis were submitted to a plastic re-operation. After a decision to perform an external stoma in the transverse colon as the first operative stage, the incidence of fistula after the second stage (resection and anastomosis) became lower: in a series of 34 consecutive resections, there were 4 cases of dehiscence, only 2 with complete fistula. None developed stenosis and healing was spontaneous. As a rule, the patient is discharged after eight or nine days, but a third stage, to close the colostomy, is required after four weeks.

When treating rectocolonic aperistalsis, the conventional measures of pre-operative bowel sterilization are not enough, whether using sulfa drugs or broad spectrum antibiotics or both. These are effective in cases of cancer or other diseases, but do not sterilize most cases of megacolon because of aperistalsis and stagnation. Of the utmost importance, therefore, is the mechanical cleansing of the colon by means of daily water enemas carrying the drugs for intestinal antisepsis through the rectum and through the distal colostomy. Seven to twelve days must elapse between the first and the second stage, and this will not be possible unless the distal gut is cleared of fæces.

In acute volvulus, there are two procedures. If no vascular damage is observed during laparotomy, operation will consist of untwisting the loop and transverse colostomy. Resection is delayed until bowel preparation is complete. In cases with necrosis of the twisted sigmoidal loop, resection is performed with double lumen sigmoidostomy. Resection of the rectum is done as a second stage, after sterilization.

Two technical details concerning rectosigmoidectomy should be mentioned. First, when a good end-to-end anastomosis is not possible because of a great difference between the diameter of the rectal stump and the descending colon, it is advisable to close the latter and to perform a side-to-end anastomosis. In 49 anterior resections of the rectosigmoid, this reconstruction was done on 8 occasions. The second point is the failure of the stumps to come together without tension, even after ligature and section of the inferior mesenteric vessels. In such a case an isolated ileal loop has been successfully interposed between the descending colon and the rectum.

In 58 operation cases there were no deaths. Follow-up studies were obtained in a few up to five years; 3 have constipation again with radiological demonstration of recurrent megaformation. The majority of the patients were operated upon in the last two years. Follow-up was

obtained in 70%: these are doing well with daily bowel movements.

Acknowledgments: I wish to record my thanks to all my colleagues in the Medical School of Ribeirão Prêto whose team-work made this investigation possible, but especially Professor Fritz Köberle, Professor J L Pedreira de Freitas and Dr Clovis B Vieira; also to Mr Ian Todd and Dr Basil Morson for their help with this lecture.

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Professor J C Goligher (*Leeds*)

I propose to confine my remarks to the two common forms of megacolon, namely Hirschsprung's disease and rectal inertia—especially the latter. For, since the brilliant pioneer work of Swenson & Bill (1948) and Bodian, Stephens & Ward (1949), there has been a not unnatural tendency in writings on the subject of megacolon to dwell on the rationale, technique, and for the most part satisfactory results of the Swenson type of operation for Hirschsprung's disease, and to dismiss rectal inertia in brief paragraphs, glossing over the lack of real insight into the condition and the unsatisfactory nature of its treatment. This is regrettable, more particularly as rectal inertia is, in my experience, commoner than Hirschsprung's. Thus of my 24 cases with megacolon only 4 suffered from Hirschsprung's disease, but 16 were examples of rectal inertia.

The first issue to be settled when a surgeon is faced with a case of megarectum and megacolon is to decide which of these two conditions he is dealing with. In some instances the diagnosis may be clinically and radiologically evident, as for example when the barium enema plate shows an appearance such as that revealed in Fig 1A, with a relatively contracted lower segment typical of Hirschsprung's disease, or alternatively as in Fig 1B where an enormous dilatation of the rectal ampulla extends down to the ano-rectal ring, suggesting that the condition is one of so-called rectal inertia. However, we have been assured by many authorities that the latter radiological appearance may not be incompatible with Hirschsprung's disease, when the aganglionic segment is exceptionally short, and for that reason we have usually done rectal biopsies on these cases, as recommended by Swenson (1958). But I must admit that so far we have not succeeded in uncovering any examples of Hirschsprung's disease, and I wonder how often Mr Nixon in his considerable experience has salvaged by rectal biopsy patients with aganglionic megacolon who had been relegated by the radiologist to the diagnostic 'dump heap' of idiopathic megacolon. Also I should be grateful if he would enlighten us on his technique of rectal biopsy. In theory it would seem essential to remove a portion of the rectal wall comprising both layers of the muscle coat in order to secure a specimen of the area in which Auerbach's plexus resides, but I understand that Mr Nixon normally contents himself with a mucosal biopsy. Is this really adequate, and does it give the pathologist a chance to express an opinion on the presence or absence of the essential ganglion cells?

In the management of cases of rectal inertia we have been much influenced by the teaching of the Hospital for Sick Children in Great Ormond

Street, and have usually employed the sort of conservative regime outlined by Mr Nixon. It is, however, a tedious business which does not seem to give much joy to anyone, least of all to the nursing staff, but in many cases it does enable the patients to avoid the extremes of fæcal impaction and incontinence that have usually been the symptoms bringing them to hospital in the first instance. What has been disappointing to me about this method has been the rarity of any substantial return of the rectal calibre to normal on objective examination. Even after several years the rectum is often found on palpation to contain an abnormally large accumulation of fæces, and the barium enema plates frequently show little or no diminution in the width of the rectal ampulla. However, this may be an indictment of our inadequacies in carrying out the regime. I should be interested to learn more of the objective results in Mr Nixon's cases. He did say that a considerable proportion of his patients were cured and many others much improved, but does this mean subjective or objective improvement?

In my mind, this failure to achieve a restoration of the bowel calibre to normal throws doubt on the whole idea underlying this conservative treatment – namely that the condition is due simply to habitual overdistension of the rectum from faulty bowel habits – and inclines me to the view that there is some intrinsic rectal abnormality in these cases. This is suggested particularly in those with a really enormous distension of the rectum extending into the upper abdomen. It is hard to believe that this can become normal again. A further fact pointing to an intrinsic abnormality is the finding – seldom emphasized in standard texts – that in many of these cases of idiopathic megarectum (actually 50% of my cases) there is an associated deformity of the anal region, usually an ectopic vaginal positioning of the anus. It may be objected that this is associated with anal stenosis and that the megarectum is secondary to this state of affairs. But this is not my experience; usually the ectopic anus is patulous, even more so in 2 of our 8 cases with combined megarectum and ectopia in which a 'back-cut' operation had been previously performed soon after birth.

The external sphincter in these cases with ectopic vaginal anus remains in the perineum, so that the anal orifice has no surround of striped muscle. This would seem to me to invalidate the hypothesis advanced by Mr Todd, and to some extent suggested by Mr Porter's work, that idiopathic megarectum may perhaps be due to some defect of the inhibition of the external sphincter ani that normally occurs on continued distension of the rectum – unless of course the puborectalis, which partly surrounds the ano-rectal junction, can be considered to act as a complete

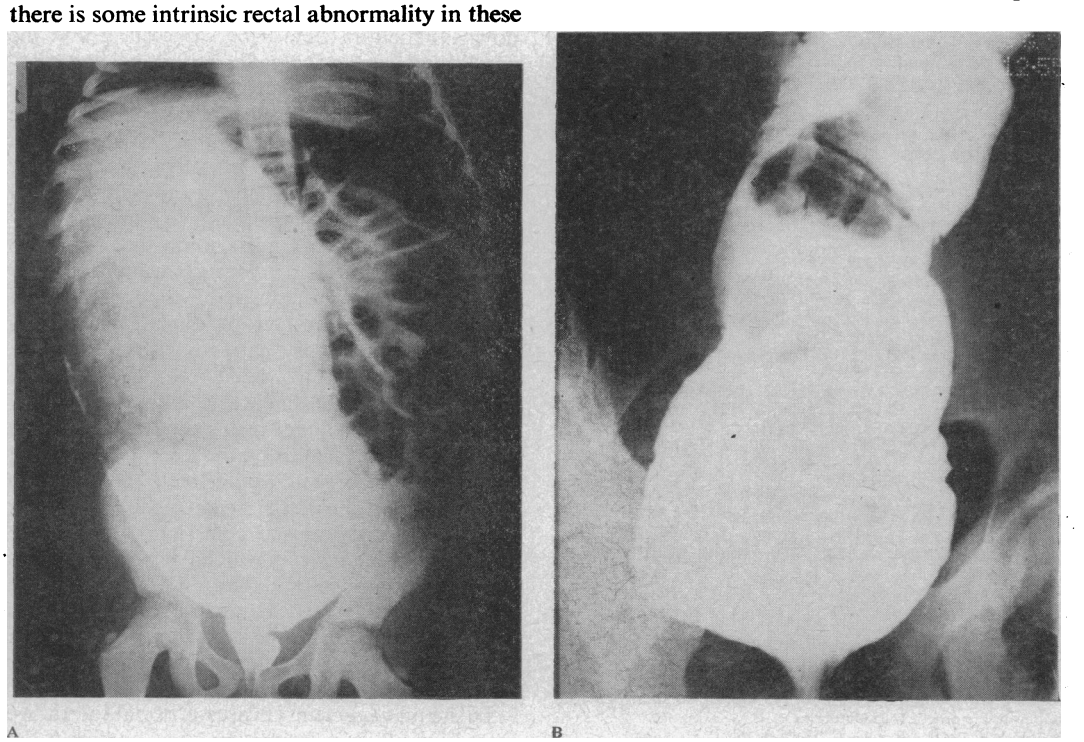

Fig 1 *Barium enema plates in cases of megarectum and megacolon.* A, *Hirschsprung's disease.* B, *Rectal inertia*

substitute for the external sphincter and to display this disturbance of proper relaxation. But I would emphasize that the anus seems patulous in most of the cases with ectopic vaginal anus that have come our way.

Another possible intrinsic abnormality is diminution of the contractile power of the rectal wall. But rectal biopsies have failed to show any quantitative diminution in the ganglion cell population of the rectal wall in these cases, comparable to that found in Chagas' disease, which Professor Santos has described. Also, though Dr Connell's motility studies did reveal a hypomotile condition of the rectal wall in some of his cases of megarectum, in others the pressure recordings were within the limits of normality. Also, as he mentioned, certain people who do not suffer from megarectum have similar hypomotile tracings. It would seem that lack of contractile power in the rectal wall cannot be held to be a consistent cause of megarectum.

Last to be considered as an ætiological factor is the possibility of some blunting of the normal rectal sensation, on account of which the patient allows fæces to accumulate without feeling any urge to defæcate. It seemed to me that there was some discordance between the sensory findings recorded by the opening speakers and our own findings in this matter. We have found that these patients with rectal inertia are quite sensitive to balloon distension of the rectum if a big enough balloon is used, but that, as in Mr Porter's cases, the sensation is experienced not in the sacral region but in the lower abdomen. Also, Mr Nixon claims that rectal sensation is perhaps initially much diminished but returns to normal as rectal distension is reduced by wash-outs, suggesting that there is no inherent sensory defect.

I feel compelled to say that, despite the interesting papers contributed to this Discussion, the precise nature of the essential abnormality present in many cases of idiopathic megarectum or rectal inertia still eludes us.

Surgical treatment of rectal inertia: It has generally been advised that these cases should not be operated on, because to remove the affected part of the rectum entirely would involve a low resection likely to interfere with continence. But there are times when the surgical instinct to excise a large mass rises superior to reason, particularly when faced with patients with a truly enormous megarectum! It is tempting in these cases to remove the dilated bowel in the hope, as Mr Nixon has put it, of giving the patient 'a fresh start in life'. I have twice succumbed to this desire and in neither case was the result encouraging. One young man had a low abdomino-anal resection and unfortunately developed leakage, sepsis and a host of complications for which I had eventually to remove the rectal remnant and give him a permanent colostomy. The other patient, a young boy, had a similar operation with a smooth postoperative course, but he has had indifferent rectal function since and still has to have enemata or wash-outs. I was therefore interested to learn that in Mr Nixon's cases submitted to resection the wash-out regime has also had to be continued. I noted Mr Todd's decidedly gloomy report on the results of different surgical procedures in his cases of megarectum submitted to radical surgery. As a corrective to his depressing record I should like to describe one operation that can be relied upon to give a good result in these cases – though at a price. That is a proximal iliac colostomy, preferably with a divided colon, placing the proximal end in the abdominal wall in the left iliac fossa and the distal end nearer the mid-line in the lower epigastric region. I have now performed this operation on 3 cases, 2 on girls of 8 or 9 years and 1 on a man of 21. Before operation the idea of an artificial anus was naturally repugnant to them and their parents, but now all three patients sing the praises of their colostomies, which seem to them like charms compared with the bother they used to have with their megarectum! I do not suggest that such a colostomy could often be indicated in the management of rectal inertia, but, when medical treatment has apparently failed to keep the patients reasonably comfortable, the surgeon should consider this simple and effective operation as an alternative to any form of resection, which is so frequently followed by continued symptoms and serious complications.

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Sir Denis Browne (*London*)

In the treatment of that variety of 'megacolon' which I consider is better called 'colonic inertia' there is one fundamental principle – that if the rectum is emptied the rest of the colon will look after itself. Purgatives only send more fæces down till the rectum is distended even beyond its already abnormal size; the wash-out with tube and funnel needs skilled administrators and is ineffective on masses of hard fæces; and the ordinary enema may vanish into the dilated bowel with unfortunate results.

What is needed is a method that acts on the rectum and lower colon, that is safe, clean and can be self-administered in privacy by older children or adults. The well-established though little-used technique shown in Fig 1 fulfils all these requirements. An ordinary rubber hot-water bottle is filled with a suitable amount of warm, approximately normal, saline and a tube with shut-off clip is screwed into its neck. The bottle is then

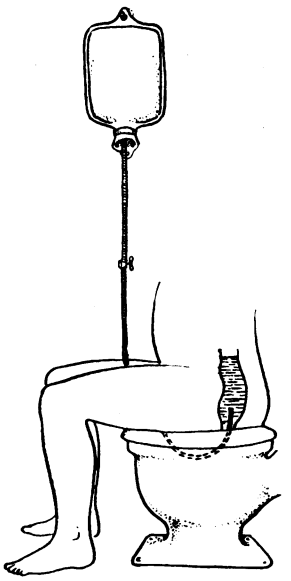

Fig 1

hung up at a height of three feet above the anus when the patient is sitting on a lavatory seat. The fluid is allowed to flow through a plastic tube till mild distension or desire to defæcate is felt, the patient having full control meanwhile by the shut-off clip. Then the tube is withdrawn and the fluid and everything that comes with it is allowed to drop through the relaxed anus: the vertical position of the trunk is a great factor in both safety and efficiency. This routine, taking a very few minutes and attracting no notice to its performance, can be carried out daily till the rectum becomes used to a new state of emptiness.

Small children can be seated on a wooden armchair with a suitable hole cut out, and the necessary control gained by a padded band over the thighs. The tube is inserted from behind, where they can neither see nor grab at it. The whole procedure is usually accepted quite easily, which is by no means the case when the child has to be held down in a horizontal position.

Mr Clive Butler (*London*)

I wish to emphasize the importance of Prostigmin in the treatment of both Hirschsprung's disease and so-called idiopathic megarectum. Since 1937 it has been my custom to have these patients screened in the X-ray department; after an intramuscular injection of Prostigmin it can be clearly seen if the dilated bowel responds or not. In favourable cases patients have been put on a course of oral Prostigmin combined with a suitable purgative; the results in a small number of cases have been encouraging.

[Mr Butler showed two slides illustrating the waves of contraction which occurred in the dilated rectum following intramuscular Prostigmin.]

Mr Ian P Todd, in reply, said that Professor Goligher's remark that a colostomy might give a satisfactory result in these cases bore out his own thesis that there was a disorder of rectal physiology, not colonic. The vaginal ectopic anus was patulous, as the external sphincter ani was not surrounding the orifice. There the puborectalis might not be inhibited normally by rectal distension or the rectum might be atonic and hypo-excitable, or it might be hyposensitive, a frequent finding in these cases. It was, however, frequently inadequate and obstructive.

Mr H H Nixon, also in reply to Professor Goligher, said: (1) He did not recall any case with 'enormous dilatation . . . down to the ano-rectal ring' in which biopsy had revealed Hirschsprung's disease. (2) The value of 'mucosal' biopsy depended on the submucosal and intermuscular plexuses both being absent throughout a Hirschsprung segment. (3) Tables 4 and 5 interpolated in the text gave a longer follow up of rectal inertia treatment. There was objective resolution of the megarectum. (4) The training regime had been used after operative removal of the rectal reservoir, but treatment had not been continued indefinitely.

Mr N H Porter, in reply to Professor Goligher, said that in 12 cases of idiopathic megacolon the position of the anus had been normal. The integrity of the voluntary sphincters had been verified clinically and by electromyography. Investigation of normal and paraplegic patients showed that reflex inhibition of the external sphincter ani and puborectalis was vitally important in normal defæcation. Without inhibition defæcation was difficult and might be incomplete so that fæcal accumulation occurred. Fæcal accumulation might be associated with a high threshold for reflex sphincter inhibition. The extent to which the anal canal would dilate was governed by its size and by pain, so that there was a limit to the dimensions of the fæcal mass which would pass. If the mass needed to establish reflex sphincter inhibition and relax the anal canal was abnormally large it would not pass, no matter how slack the anal canal might become. In this respect, therefore, a high inhibitory threshold would act as an inherent barrier to defæcation. This hypothesis could only be evaluated by further study and a long follow up of cases. Mr Porter questioned the validity of comparing this physiological problem with the host of anomalies, anatomical and functional, which arose in congenital maldevelopments of the pelvic floor.

In the experiments described rectal sensation referred to some kind of desire to defæcate, which was experienced in the perineal or sacral regions and not the abdomen. Finally, he emphasized that sphincter inhibition depended on a reflex arc. The afferent impulses arose from sensory endings in the wall of the rectum. If defects of the inhibitory mechanism existed they might affect any part of this arc. It was not, therefore, possible to exclude sensory defects from investigations at present.