Section of Proctology

President Ian Todd MS

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President's Address

Some Aspects of Adult Megacolon

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Megacolon has not been the subject of a Presidential Address before, perhaps because little is known about it (though much has been written), or perhaps because it is a curiosity and of no great importance in this country. By definition it consists of large intestine with greatly increased diameter and hypertrophied wall. Unfortunately it is impossible to be sure whether the wall is thick or thin until the abdomen is opened. Thus from the diagnostic angle any large-calibre colon must be considered to be a megacolon.

My interest in this field was first aroused when I was resident surgical officer at the Hospital for Sick Children, Great Ormond Street, in the mid-1940s. At this time, before the work of Swenson *et al.* (1949) and Bodian *et al.* (1949), all cases were called Hirschsprung's disease or megacolon, the terms being used indiscriminately, as is unfortunately so even today. A local neurological abnormality to account for megacolon had been suggested, before Swenson's work, by at least three authors, though not by Hirschsprung himself.

At Great Ormond Street Dr Margaret Hawksley was giving spinal anæsthetics to these patients and some, but not all, emptied their bowel on the operating table. My chiefs, Barrington-Ward, Twistington-Higgins and Denis Browne, allowed me to try the effects of methacholine bromide and, though hypotension was caused in many, the results were generally not encouraging. It was not until much later, when I visited South America and met Cutait, Ferreira-Santos and Køberle, that my interest was again stimulated (*see* Ferreira-Santos 1961, Cutait 1965, Køberle 1960). The aganglionic segment had by then been described, and in Brazil megacolon of the Chagas type had been shown to be due to a quantitative myenteric ganglionic degeneration. Here was a quandary: two neurological lesions of ganglia producing, in Chagas' disease, a megacolon with some hypertrophy of muscle, and, in Hirschsprung's disease, a narrowed colon but with a greatly hypertrophied megasegment proximally in which the ganglia and plexus were apparently normal. These macroscopic differences were impressive and were carefully noted, together with the subjective sensation of tonus, in all cases of megacolon which we operated upon (Todd 1961).

Work by Dr Barbara Smith (1967) on the neuropathology of the plexus en face has partly borne out some of the earlier rather tentative hypotheses, but a reappraisal of cases and material available suggests that in megacolon there may be not only neurological changes but possibly muscle and other changes which could be the primary defect. It seems established that congenital absence or severe deficit of ganglia, accompanied by submucosal nerve changes, leads to a narrowed unrelaxing nonpropulsive segment, above which the more normal bowel becomes enlarged and thickened as a result of work hypertrophy; this is Hirschsprung's disease. In Chagas' disease there is a quantitative destruction of ganglia due to toxins from *Trypanosoma cruzi* which enter the patient through the bite of an infected Triatoma fly. This leads to increasingly unsatisfactory propulsive efforts by the diseased intestine where most work is required and where the contents are most solid until it becomes entirely decompensated; surgery is then required to relieve the obstruction. Other processes which destroy the neurological control of colonic propulsion might similarly lead to megabowel, and these have now been demonstrated: cathartic colon shows such changes, and may be recognized histologically (Smith 1968). Whether the damage is due to cathartics or to an inherent weakness or susceptibility of the nervous control is not known, for many people

who take large doses of laxatives over many years do not suffer ill effects. These people need to change the type of medicine from time to time when it becomes ineffective – perhaps this is transient damage. Ogilvie's syndrome is possibly a more central form of nerve interruption, due to neoplasia, which leads to megacolon.

Parks *et al.* (1962), Porter (1962) and others have taught us much about electromyography of the sphincters. Failure of sphincteric inhibition, that is relaxation of the anal canal musculature, when the rectum is filled, leads to a physiopathological obstruction and a hypertrophic megarectum proximally. The rectum becomes huge, sensation is dulled by the prolonged distension but no sphincteric inhibition can be demonstrated. These patients are never incontinent.

Provided there is no central or spinal lesion, incontinence of fæces usually signifies a neglected habit, with later dulling of sensation. The megarectum is secondary to this dyschezia and usually, though hypertrophic, becomes atonic. Rectal dyschezia may theoretically, and probably in fact, be due to a primary asensory rectum with normal sphincteric inhibition.

What else might upset the neurological propulsive mechanism? Perhaps other toxic substances or infecting agents can be responsible. There is some evidence that cytotoxic agents may have this effect, and we have all seen altered bowel habit with distension in patients receiving these drugs. It is probable, however, that the changes are transient and often reversible, so no megacolon results. We have seen a few cases which show inflammatory changes in the plexus, suggestive of an infective or protozoal agent.

When nerve damage is shown, it seems unlikely that the atony or hypertrophy previously mentioned is primary. However, there are a few cases of megacolon in the records at St Mark's Hospital where the bowel wall is paper-thin from overdistension, and obviously incapable of normal propulsion, but where no gross neurological changes have been shown. It would be reasonable to suggest that in these cases there is a primary muscular disorder or degeneration which causes 'pseudo-obstruction' and megacolon. Not all these patients are old, so that senile degeneration is not a satisfactory explanation; possibly this is in some way similar to thin-walled diffuse diverticulosis.

Connective tissue deficiencies such as scleroderma and Ehlers-Danlos syndrome cause constipation but seldom megacolon, although spontaneous rupture occurs in Ehlers-Danlos. Mr A S Till, of Oxford, has a case of scleroderma with gross megacolon, and Dr R A Kemp Harper has radiographs of this condition. Vascular deficiency seems to cause stenosis rather than enlargement of diameter. Volvulus is an intermittent closed-loop obstruction which usually remains relatively thin-walled, i.e. distended only, with no muscle hypertrophy. Other mechanical obstructions, apart from untreated congenital anorectal deformities, do not continue long enough to cause a megacolon.

Table 1
Theoretical etiological factors in megacolon and megarectum

1. Mechanical obstruction:	3. Muscle:
Congenital anorectal	Deficiency?
deformities	Degeneration?
Volvulus of colon	4. Connective tissue:
2. Neurological:	Ehlers-Danlos?
Congenital deficiencies	Scleroderma?
Degenerative effects	5. Vascular abnormalities??

Theoretically possible causes of megacolon and megarectum are summarized in Table 1.

How should a case of megacolon be investigated? The history may help. Has it been present since birth? Has it been apparently precipitated by pregnancy? Where has the patient lived? Have laxatives been used for many years, with lessening effect – and, if so, of what sort? The significance of pregnancy is not known, but in this series there are 2 cases (and possibly a third) of true Hirschsprung's disease which presented during the puerperium and were unremitting until treated. Chagas' disease is found only in a limited area of the Americas, though it is possible that similar diseases occur elsewhere. Megacolon occurs in both East and West Africa and various explanations have been put forward, but I have no personal knowledge of them. One patient who lived for many years in India shows changes in the plexus which are identical with those of Chagas' disease, though *Trypanosoma cruzi* is unknown in India. As for laxatives, senna and possibly the anthraquinones seem particularly dangerous in the development of cathartic colon.

X-ray examination is obviously helpful. A plain X-ray may show gross fæcal loading; though this is diagnostic, it is not often helpful in differentiating the cause of the megacolon. However, a colon distended by gas suggests either volvulus or the elongated thin-walled variety. A volvulus may collapse if a tube is inserted into it through a sigmoidoscope: and the thin-walled colon may if the anus is dilated, for the large intestinal muscle may not have the power, even with voluntary straining, to overcome normal sphincter tone.

With a barium enema the narrow segment of Hirschsprung's disease is best seen in the lateral view in the adult (Fig 1), as it may be short and obscured in other views by the huge megasegment. Demonstration, by barium enema, of dilatation down to the anorectal ring does not help in further differentiating the cause, and alteration in the segmentatory pattern (an unusual finding) gives no clue.

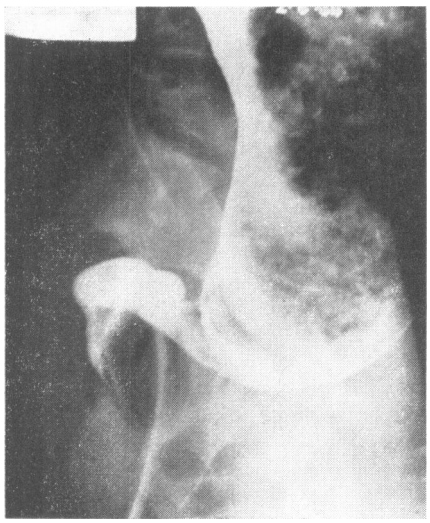

Fig 1 *Hirschsprung's disease, with narrowed segment seen in lateral view*

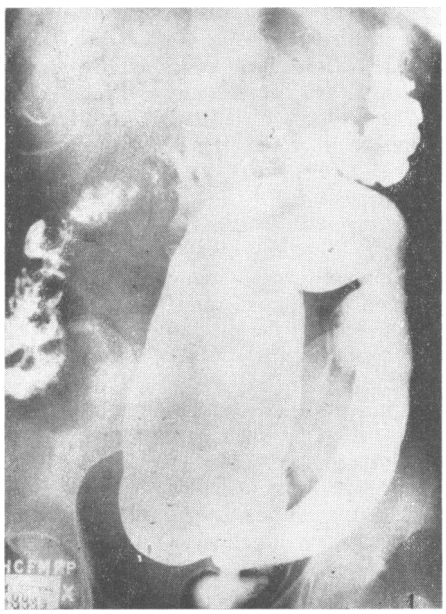

Fig 2 *Chagas' disease*

Transit time or failure of markers to progress beyond a certain point may occasionally be useful, but this is of more help in chronic constipation without megacolon. In some cases small intestinal transit may be slow, perhaps suggesting a generalized intestinal disease rather than disease of the large intestine alone.

Chagas' disease affects the whole intestine, though its effects are normally seen only in those parts which propel solids and thus have more work to do (Fig 2). There are, however, 3 cases

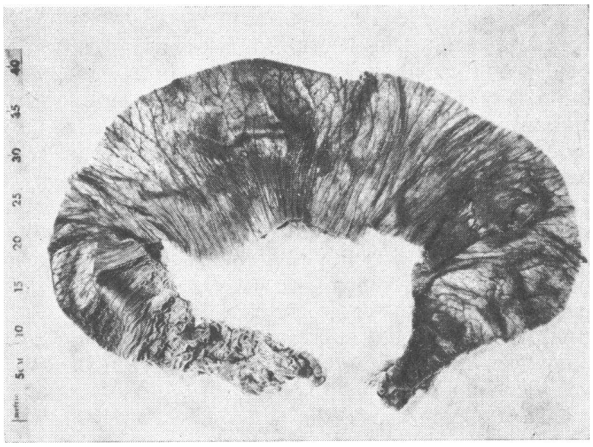

Fig 3 *Paper-thin transverse colon*

in the series, though only one is proven, that have myenteric damage which is neither of Chagas type nor aganglionic, with generalized failure of intestinal propulsion and dilatation.

Motility studies are not very helpful, though they have been carried out in many cases. Electromyography of the sphincteric mechanism combined with sensory responses has, however, been most useful. Normal sensation is provoked by stretch distension of the rectum by about 150–200 ml, but huge amounts may be required. Inhibition of sphincters occurs at about this level, though there may be gross variations. But sensation and inhibition do not necessarily go hand in hand, and it is not known where the primary defect lies. The rectum may be hyposensitive, with normal inhibition; or the sphincters may not be inhibited by huge volumes, in a rectum initially sensitive. The latter could well be secondary loss of perception due to prolonged overdistension, with a primary abnormality of sphincteric inhibition.

Sigmoidoscopy is seldom a very helpful investigation. A narrowed segment and a cavern above may be observed, but there are cases in which the instrument drops in to 25 cm; often these turn out to be cases of neglected habit.

Rectal biopsy should probably be done last, as the procedure could interfere with other investigations. A full-thickness strip should be taken under full anæsthesia from just above the anorectal ring, in order to rule out Hirschsprung's disease. The en face histological investigations cannot be carried out on so small a piece, and a 2·5 cm square of resected specimen is required. Thus a tentative diagnosis must be made in cases of megacolon, and treatment carried out before one's calculated guess is confirmed – a far from satisfactory sequence. Only at laparotomy can hypertrophy, atony or thinning be verified.

Treatment

I believe adult Hirschsprung's disease is best treated by the Duhamel operation. For this to be technically possible, removal of quite a large amount of the mega hypertrophied bowel is required. In the adult the coned segment may be foreshortened by fæces packed into it by the powerful colon above, but Smith (1967) has shown ganglionic and plexus changes in the megasegment proper; this is probably the cause of lack of propulsion post-operatively, which may in some cases lead to enterocolitis and death. This condition may not respond to sphincteric dilatation, sphincterotomy or washouts, which are often advised.

State's operation was done in 2 cases, and in both there was leakage for a time; in both the leak eventually closed spontaneously, in one case after a month. This bears out the contention that State's procedure, which is identical with a low anterior resection, leaves too much of the aganglionic segment, which remains partly obstructive. It must, however, be admitted that late results seem satisfactory, whereas the Swenson pull-through is technically difficult in the adult, may result in poor control, and is most liable to damage the sacral outflow.

With some trepidation I state that Chagas' disease in the adult should be treated by resection of the decompensated megasegment and a really low extraperitoneal rectal anastomosis. Cutait has given up pull-through procedures in favour of the Duhamel operation because of poor continence. This suggests that abdominal resection would be wiser, as sphincteric inhibition is normal in this condition and the sphincters may well be relatively inadequate by the time operation is undertaken, particularly in the adult.

Paper-thin large intestine seems to do well with resection of the thinned portion, whether sigmoid alone, as in volvulus, or a larger amount if the condition is diffuse. The ileocæcal valve should be retained whenever possible in mega disease, with a cæcorectal anastomosis if a long resection is desired, for the sphincters may well have been subjected to prolonged inhibitory stimuli from the distended rectum. Short sigmoid resections for anything except volvulus have given almost uniformly bad results.

The one remaining major problem of treatment is the megarectum which is hypertrophic down to and below the pelvic floor. Some cases are atonic, suggesting a propulsion failure, but this is not proven; there could be a primary sensory defect. Others must be due to sphincteric obstruction which has not been relieved by extensive sphincterectomy. A proximal colostomy has been satisfactory, when acceptable to the patient; in 2 cases the colostomy was later closed, on the presumption that the rectum had long been overdistended and had become secondarily atonic and unable to empty, but might have recovered; both worked normally for a time but then reverted to their old ways.

Perhaps the impression has been given that all cases come to operation, but obviously this is not so. One must, however, be more than ever wary of the prolonged use of cathartics. Almost all the totally decompensated cathartic colons show a severe degree of melanosis. Saline osmotic substances, such as Epsom salts, which carry water to the large intestine, should be tried first. They are also useful as a test of good sphincter control. Recently the use of large doses of pantothenic acid has been impressive, but it is not known whether this has a sound pharmacological basis or whether a deficiency could exist leading to failure of propulsion. Where the main problem is rectal inertia or true dyschezia it is absolutely essential that the rectum be kept empty; this can be achieved only by washouts or suppositories.

One reason for slow transit in constipation is lack of bulk. This is, however, rare in megacolon. Methyl cellulose and its derivatives are of no use in the mega conditions.

Prostigmine, often given as Rae's mixture in cases of constipation, occasionally contracts what may be a lazy colon which has become dilated. With true megacolon its effect is disappointing.

Material Analysed

This series is based on 90 cases of megacolon, 37 of which are entirely my own; I also saw, and sometimes operated upon, many of the others. The sex incidence is almost equal: 43 females and 47 males. The longest period without any bowel action was four months, in a case of Hirschsprung's disease.

Twenty patients had adult Hirschsprung's disease; one suffered from Chagas' disease, and this has been reported elsewhere (Todd *et al.* 1969); one probably had Ogilvie's syndrome, and 7 (3 males, 4 females) presented with megacolon secondary to anorectal stenosis of congenital origin. There was no case of iatrogenic stenosis causing megacolon.

Eighteen cases presented with a very thinned colon. Of these, 5 were probably straightforward sigmoid volvulus. In 9, however, the thin colon involved almost the whole of the large intestine, particularly the transverse colon (Fig 3), and in the remaining 4 rather more than the sigmoid alone; one wonders if these are not primary muscular defects or degeneration. The ganglia are normal, as is the plexus in those in which it has been studied.

Twenty-five have had no operation; biopsy has been done in 22; electromyographic and sensory studies have been carried out in many cases and in 6 have suggested neurological abnormalities

of either the sensory response or the sphincteric inhibitory mechanism.

Eighteen cases have been operated upon and, of these, 5 have shown plexus lesions which are not of Hirschsprung type. Smith (1968) has reported her findings in cathartic colon, and it is probable that many of the present cases, had they been adequately examined, would have shown neuropathological changes.

To summarize, adult megacolon needs to be adequately investigated. Biopsy, sensory and electromyographic studies are the most helpful. Resection of a so-called redundant sigmoid loop rarely cures, unless it is a true volvulus which shows a relatively thin wall. If the colon is paper-thin, then it is decompensated and propulsion is inadequate; resection of the whole thinned length gives good results. When possible the ileocæcal valve should be preserved, for in chronic constipation sphincter reflexes may be dulled and control poor. Beware the thick hypertrophic colon: unless this is secondary to the aganglionic segment of Hirschsprung's disease, surgery may alleviate but cannot cure because some distal obstructive process must be present.

What of extensive internal sphincterectomy, about which much is written in pædiatric circles? Theoretically it might be useful in the diffuse thin-walled condition in which the intestine inflates rapidly with gas. One must, however, be very wary of its use in adult megacolon and megarectum, as the sphincters have been subjected for years to excessive inhibitory stimuli and may well be none too competent.

In conclusion, it must be remembered that our knowledge of this condition is far from complete. As an example, one patient had had several colonic resections for a mega condition of the large intestine, the final anastomosis being an ileorectal one. In spite of this a mega-ileum developed, and yet there was no evidence of a neurological abnormality of the bowel and the ganglia were intact.

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Meeting May 27 1970

Papers

The Clinical Value and Limitations of Liver Scanning

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Radioisotope scanning procedures are increasingly used in medical diagnosis and in many cases, particularly in suspected brain and liver disease, may be regarded as routine investigations.

Scanning is probably the best available method of examining the liver, not only to confirm or exclude a clinical impression of liver enlargement but also to demonstrate the shape and position of the organ and to detect space-occupying lesions. Large series have shown that it is more reliable than liver function tests in the diagnosis of liver tumours, with a higher detection rate and fewer false positives (Czerniak 1964).

An ideal method of establishing the diagnosis is by tissue examination – open biopsy. However, Conn & Yesner (1963) have shown that blind biopsy detection of liver metastases can be as low as 50%. This detection rate can be improved by the use of liver scanning to select the proper biopsy site. Poulose *et al.* (1969) report 5 cases in which liver biopsy was initially negative but repeat biopsy after selecting the site from the scan image yielded positive results for deposits.

There have been many papers in the last ten years advocating liver scanning, reporting results and suggesting technical improvements. There have been few contributions dealing specifically with colonic and rectal carcinoma and probably one reason is that the patients concerned come to laparotomy in any event for relief of symptoms. Although this paper deals with scanning only in broad terms, it is interesting to note that, of all metastases, those from colonic cancer should be most amenable to detection by scanning as they are reported to be often particularly large (Edmondson & Anderson 1966).

Raven (1957) discussed the type of patient who would benefit from isotope scanning. He found that approximately 30% of patients with liver metastases from carcinoma of the colon would be suitable for hepatectomy, and 28% of those with carcinoma of the rectum. In these patients scanning would be helpful in assessing the operative