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THE DUHAMEL OPERATION FOR HIRSCHSPRUNG'S DISEASE

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The best operation is that which offers the maximum of benefit with the minimum of interference. It is bad surgery to employ a complicated destructive operation if some simple and conservative procedure will deal efficiently with the condition present. Charles F. M. Saint[5]

In 1957 Bernard Duhamel,[1] of Paris, described and illustrated in detail a technical modification of the Swenson's operation for Hirschsprung's disease. At the 6th annual meeting of the British Association of Paediatric Surgeons, held in Liverpool in June 1959, he reported on 11 cases of Hirschsprung's disease treated by this technique and claimed that he as well as some French, Spanish and Swiss surgeons found that the functional results were always perfect and that radiological studies showed complete regression of colonic dilatation to a normal diameter in 2-4 months.[2] At the annual meeting of the British Association of Paediatric Surgeons held at Great Ormond Street, London, in July 1960, Duhamel indicated that his results continued to be excellent,[3] and by that time several British paediatric surgeons had adopted the procedure for the treatment of Hirschsprung's disease.

In August 1960 we saw a patient suitable for treatment by Duhamel's operation. This was a Coloured boy of 4 years who had had a previous unsuccessful sigmoidectomy (as recommended by David State[7]). The operation proved to be simple despite the previous surgery, blood loss was minimal, there was a striking absence of operative shock, and his convalescence was remarkably smooth.

In a previous publication[4] it was pointed out that Hirschsprung's disease appeared to be comparatively common in Cape Town, and our experience since that time has confirmed this impression, particularly with regard to 'neonatal Hirschsprung's disease'. We decided, therefore, to conduct a trial of the Duhamel procedure, and from August 1960 until September 1961 have had the opportunity of performing the operation on 15 infants and children. In all fairness to the Swenson procedure,[8] which we used with very satisfactory results on 42 patients between 1953 and 1960, it must be emphasized that our objective in changing to the Duhamel operation was simply to undertake a clinical trial and not to decry a well-tried and established method of treatment. It is the purpose of this paper to describe briefly the technique of Duhamel's operation and to report our results to date. Comparison of our present results with our results of the Swenson's procedure must necessarily await further trial and a longer follow-up period.

MATERIAL

There were 15 patients. In 7 the aganglionic segment involved rectum and lower sigmoid only (short segment); in 4 it extended to the mid-descending colon; in 3 the whole of the left half of the colon was affected; and in 1 the disease involved the whole colon and 5 cm. of the terminal ileum (Fig. 1).

DIAGNOSIS

The diagnosis was made on the following criteria:

1. *Clinical presentation.* Nine infants presented with acute neonatal intestinal obstruction. Five of them required emergency surgery during the first month of life — 4 colostomies and 1 ileostomy — and in them the appearances of the bowel at operation were typical of Hirschsprung's disease. One of these patients had a subsequent Swenson's operation and one had a subsequent sigmoidectomy performed. Four of the infants were treated conservatively, and their subsequent course was typical of Hirschsprung's disease.

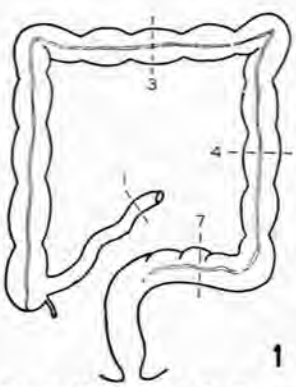

Fig. 1. Hirschsprung's disease — 15 patients (see text).

Four infants and children presented later in life, but from the history it was clear that their 'bowel-trouble' dated from birth. One of these had had a Swenson procedure performed elsewhere.

In 2 patients, aged 1½ years and 3 years respectively, the histories were indefinite and could not clearly be dated from birth.

Physical examination in all the older children showed the typical pot-belly with splayed-out ribs, poor nutrition, and an 'empty' rectum.

2. *Barium enema.* This was carried out in 10 patients, and in all of them the typical picture of Hirschsprung's disease was demonstrated (Figs. 2 and 3).

3. *Rectal biopsy.* This was performed on 14 patients by the technique developed at the Red Cross Hospital and described by Shandling[6] at the 2nd Congress of the South African Association of Surgeons in September 1960. In all of them there was complete absence of ganglion cells. (A rectal biopsy was not performed on the child previously operated upon elsewhere, because the pathologist's report on the excised rectosigmoid was available.)

4. *Biopsy of colon at laparotomy.* This was performed in all the patients, the purpose being to ensure that the proximal line of section was through normal bowel. In 8 of the patients the first biopsy taken from apparently uninvolved bowel (i.e. dilated and hypertrophied), revealed no ganglia, and a more proximal section had to be made

(this was necessary in both patients who had had previous Swenson procedures performed).

5. *Histology of excised colon.* In all but 1 of the patients the aganglionic segment extended proximal to the peritoneal reflection, and therefore the diagnosis could be confirmed by microscopic examination of the operative specimen even though the aganglionic rectum had been left *in situ.*

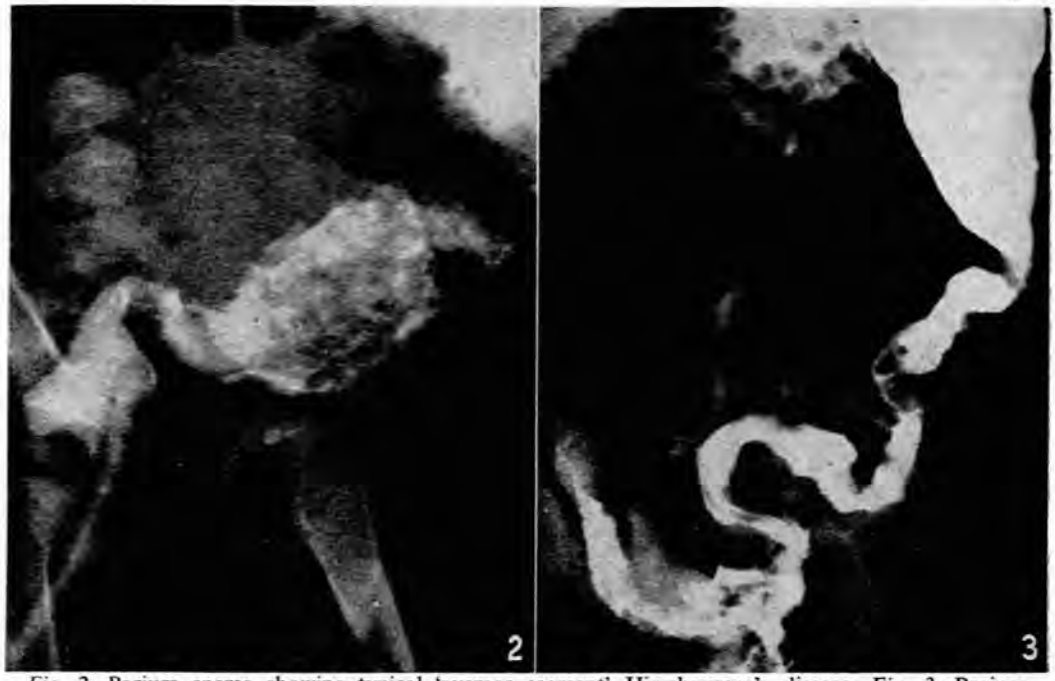

Fig. 2. Barium enema showing typical 'average segment' Hirschsprung's disease. *Fig. 3.* Barium enema showing 'long segment' Hirschsprung's disease.

TECHNIQUE OF THE OPERATION[1,2]

In 7 patients the operation was performed as a primary single-stage procedure at ages varying from 5 months to 9 years; in 3 patients it was performed as a single-stage corrective procedure after previous failed surgery, and in 4 patients as a two-stage procedure, the second stage consisting of excision of the colostomy (or ileostomy) and the Duhamel operation. In 1 baby colostomy was performed at 3 weeks, the Duhamel procedure at 5 months, and closure of the colostomy at 8 months (Table 1).

TABLE I. DUHAMEL OPERATIONS PERFORMED AT THE RED CROSS WAR MEMORIAL CHILDREN'S HOSPITAL, AUGUST 1960 TO SEPTEMBER 1961

Age at operation	*No. of patients*
Less than 3 months	2
3 - 6 months	4
6 months - 2 years	4
Over 2 years	5
Total	15

Single operation	7
Staged operation	5
Re-operation	3

Position

The lithotomy-Trendelenburg position, as used for synchronous combined excision of the rectum, was employed. In older children small Lloyd-Davies leg crutches were used, and in infants and small children improvised rests made of padded Kramer splints. A Foley's catheter was inserted into the bladder and strapped to the thigh.

The Abdominal Approach

Laparotomy was performed through a long, left, lower paramedian incision (in 3 patients this encircled the left transverse colostomy previously made), and the colon was inspected to verify the diagnosis. The lower sigmoid area was the common site of transition from normal to aganglionic colon. Proximal to the lesion, the colon was dilated and hypertrophied. It then coned down into the narrow segment, the cone of transition usually being about 3 - 5 cm. long.

After the diagnosis had been checked, the field was cleared by packing the intestines into the upper abdomen and holding the bladder forward by stay sutures steadied by haemostats.

In selecting the proximal level of section due consideration was given to the fact that ganglion cells are absent from the area of coning and often from the most distal 5 - 15 cm. of dilated bowel. The first biopsy of colonic wall was therefore taken from dilated bowel, approximately 10 cm. proximal to the transitional area. This biopsy was taken as a routine before proximal division of the bowel and sent for frozen section to ensure that the transection was indeed through bowel with normal ganglia.

After careful mobilization of the mesosigmoid by division of congenital bands which fix it to the lateral pelvic wall, the blood vessels in the mesentery were carefully inspected and a suitable vessel earmarked on which to mobilize the bowel.

An incision was then made on the lateral aspect of the base of the mesocolon as it leaves the common iliac vessels, and the left ureter was identified, exposed, and swept laterally away from the posterior attachment of the mesosigmoid. The right ureter was also identified and pushed aside through an incision to the right of the mesocolon. (In small babies the ureters lie very close to the midline and are easily dragged up in the mesentery of the hypertrophied bowel.)

It was usually necessary to mobilize the lower descending colon as well as the sigmoid, and in long-segment cases the mobilization included freeing of the splenic flexure by

the usual incision along its lateral border and division of the gastro-colic omentum.

The mesocolon was then divided. The line of division extended outwards from the point of division of the main blood supply to the margin of the colon selected for transection. Care was taken to skirt the lowermost intact arterial arcade. The colon was then transected between crushing clamps and the proximal end temporarily closed with a running suture, the ends of which were left long (Fig. 4).

The distal colon was then detached from its mesentery up to the rectosigmoid junction and the rectosigmoid angle lifted forward from the promontory of the sacrum. The cellular retrorectal space was opened up by cutting through the mesorectum and blunt dissection carried downwards in the midline using the finger and/or a swab on a holder until the pelvic floor was reached (Fig. 5). (When the dissection is low enough the finger passes over the tip of the coccyx.) A clear presacral plane of cleavage was easily found and the dissection was practically bloodless. By sweeping the finger from side to side this presacral space was enlarged until it was large enough to accommodate 2 or 3 fingers comfortably.

The rectum was then divided between clamps, 1 cm. proximal to the peritoneal reflection, and the specimen removed. The rectal stump was oversewn, invaginated, and covered by pelvic peritoneum as in Hartman's operation for carcinoma (Fig. 11).

The length of the proximal colon was now tested to see whether it could be brought down to the anus. Inadequate length was almost always due to tethering by the blood vessels and could usually be overcome by judicious division of restraining arcades. Sometimes it was necessary to divide a large arterial trunk, such as the left or even the middle colic, in which case more bowel had to be sacrificed. Although the general rule should be to conserve as much colon as possible, attempts at bringing down descending or transverse colon often put an undue strain on the vessels of supply, and in such cases it was deemed wiser to use the ascending colon hinged on the right colic or even the ileo-colic vessels. (In them the appendix was removed as a routine.) Also, in cases where the bowel above the cone was grossly enlarged, the subsequent steps of the operation were facilitated by removal of as much as possible of the dilated bowel along with the aganglionic segment.

At this stage the perineal part of the operation was begun.

While the perineal stage was being completed, the abdominal operator loosely approximated the margins of the pelvic peritoneum around the colon where it descended into the hollow of the sacrum. (The pelvic peritoneal flaps should not be too tightly sutured because of the risk of forming a dead space under the peritoneum.) Neither the sacral hollow nor the peritoneal cavity were drained.

The Perineal Approach

The anus was dilated and the rectal stump thoroughly cleansed by swabbing and irrigation. A special speculum was inserted into the anal canal which kept it wide open without encroaching on its posterior margin. Adrenaline, 1 in 300,000, was injected into the posterior submucous space to reduce oozing during the subsequent dissection. (Swabs soaked in this solution were also used.) (Fig. 6.)

An incision was made along the posterior circumference of the anus at the line of the ano-cutaneous junction (Fig. 6). Using scissors and the finger, the posterior wall of the anal canal was then dissected off the external sphincters up to the ano-rectal ring (Fig. 7). At the upper border of the ano-rectal ring the dissecting finger was thrust backwards through the remains of the longitudinal coat of the rectum into the previously opened-up retrorectal space (Fig. 5). This opening was then dilated up to about the size of a no. 11 - 13 Hegar's dilator.

The stage was now set for bringing the colon down to the anal verge. Before this was done, however, 4 or 5 sutures were passed through the distal lip of the ano-cutaneous incision. These were left long and attached to their needles for use in the ano-colic anastomosis to be performed later.

A long, curved haemostat was then introduced through the opening in the posterior wall of the rectum and advanced along the retrorectal space until the points appeared in the peritoneal cavity. The ligatures on the end of the colon were then grasped and by gentle traction on the forceps from below and guidance of the bowel from above, the colon was drawn down behind the rectum and through the gap in the anal wall to the level of the skin incision (Fig. 8). The lower end of the colon was reopened (Fig. 9) and its posterior margin sutured to the distal lip of the ano-cutaneous incision, using the sutures previously inserted into the latter.

Two Kocher forceps were then applied, one blade into the lower colon, the other into the rectum (Fig. 11). Their points met in a V high up in the bowel and their bases were held apart so that they came to lie at the lateral angles of colon and anus (Fig. 10). The forceps were tightened and tied together. These forceps served not only to crush the adjacent walls of colon and rectum, but also to prevent retraction of the colon and ensure haemostasis. After a few days when the forceps cut through, a longitudinal opening between colon and rectum was produced and a common chamber was formed (Fig. 12).

RESULTS

The postoperative course of all our patients, including the small babies, was remarkably smooth. Operative shock was minimal, and the children remained remarkably comfortable despite the forceps projecting from their anal orifices. There was no urinary retention (the catheter was usually removed at the completion of the operation) and spontaneous stools were passed within 2 - 4 days. The enterostomy forceps cut out in about a week (3 - 10 days), when the patients were allowed up and about. Excoriation of the buttocks presented a temporary problem in 2 small babies who had very long segments removed — one had an ileo-rectal anastomosis and the other an anastomosis of splenic flexure to rectum — but this is obviously something which cannot be avoided. There were no deaths or major complications, and none of the children developed postoperative entero-colitis.

It is still too early to assess the long-term results. Suffice it to say that all the children appear to be perfectly continent of faeces and that there have been no urinary disturbances. The older children have normal rectal sensation and are back at school, living perfectly normal lives, while the tiny babies have tolerated the operation very well and are thriving. Postoperative radiological studies

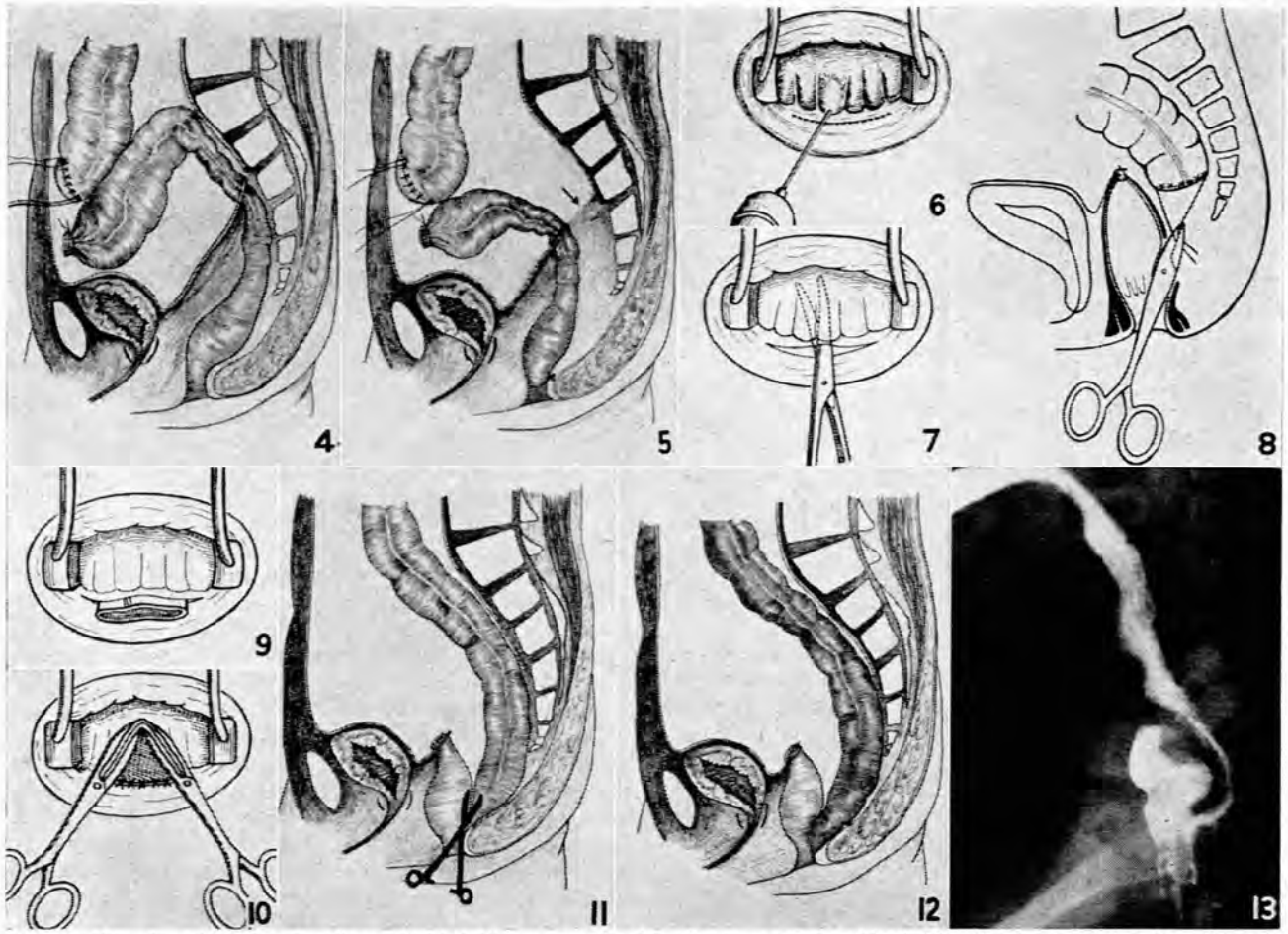

Fig. 4. The sigmoid colon has been divided 10 cm. proximal to the 'cone of transition'. The distal cut end has been ligated and the proximal cut end temporarily sutured. *Fig. 5.* The recto-sigmoid has been drawn forward away from the sacrum and the cellular retro-rectal space entered (upper arrow). The lower arrow indicates the site where the retrorectal space will be entered from below at a later stage. *Fig. 6.* The anus is held open by means of a speculum and the posterior wall infiltrated with 1 in 300,000 adrenaline to reduce bleeding. The dotted line indicates the line of incision along the posterior circumference at the ano-cutaneous junction. *Fig. 7.* The incision is deepened between the external and internal sphincters by scissors-dissection to separate the posterior wall of the anal canal from the external sphincter. *Fig. 8.* The mobilized colon is drawn down behind the rectum and through the gap in the posterior anal wall to the level of the skin incision. *Fig. 9.* The lower end of the colon projecting into the anal canal has been re-opened and redundant bowel removed. *Fig. 10.* The points of the crushing forceps meet in a V high up in the bowel and their bases lie at the lateral angles of colon and rectum. *Fig. 11.* The posterior wall of the 'pulled through' colon has been sutured to the distal lip of the ano-cutaneous incision, and Kocher forceps have been applied to crush the adjacent posterior rectal and anterior colonic walls. Note that the rectal stump has been oversewn and peritonealized. *Fig. 12.* The final result. The crushing forceps have cut through leaving a wide opening between rectum and colon. *Fig. 13.* Postoperative barium enema showing the colon entering the lower end of the rectal stump. Note that the colon is normal in calibre.

after intervals of 1 - 3 months have shown complete regression of colonic dilatation (Fig. 13). Even at this early stage, therefore, we feel that the results are most encouraging. We agree with Duhamel that the operation has the following special advantages:

1. Extensive pelvic dissection is avoided.
2. The nerve supply of the bladder is not disturbed.
3. Rectal sensation is preserved.
4. A wide anastomosis is performed.
5. Separation of the anastomosis is minimized.
6. It is suitable as a re-operation after previous failed surgery.
7. It is safe during the first few months of life.
8. Early ileo-rectal anastomosis is possible in patients with aganglionosis of the entire colon.

In view of the above we feel that this *simple and conservative procedure will deal efficiently* with Hirschsprung's disease, and we intend continuing with the clinical trial.

I wish to thank Dr. J. F. W. Mostert, Superintendent of the Red Cross War Memorial Children's Hospital, for permission to use the hospital records, and Messrs A. Katz and R. van der Riet who operated on 3 of the patients in this series. I am indebted to Mrs. P. Kottler and Mrs. H. L. F. Currey for the diagrams and to Mr. G. McManus for the photography.

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