

HIRSCHSPRUNG'S DISEASE: A NEW SURGICAL TECHNIQUE*

BY

F. SOAVE

From the Department of Paediatric Surgery, Institute 'G. Gaslini', Genova, Italy

Method

The operation is by a combined abdomino-perineal approach.

The purpose of this communication is to describe a new surgical technique for Hirschsprung's disease. This consists in pulling down the colon, and passing it through the rectal canal whose structure is preserved intact. In this way mobilization of the narrowed recto-sigmoid segment is effected entirely by an extramucosal approach, without pelvic trauma. The colon is then resected from the perineal side.

The choice of the extramucosal route for mobilization of the recto-sigmoid segment was prompted by papers by Rehbein and Romualdi on the treatment of cases of high-level imperforate anus with rectourethral fistula or prostatic anus: after opening, cleaning and cutting across the upper rectum they removed the mucous membrane at the level of fistula.

Abdominal Stage. After laparotomy the narrowed recto-sigmoid segment is prepared above the intact pelvic floor with the tract of the colon to be resected (Fig. 1). If the colon is considerably distended by gas, it is emptied by introducing a needle at a tangent to the colon axis: no suture is needed where the needle is introduced. Novocain in 0.5% solution is infiltrated into the seromuscular coat of the recto-sigmoid segment above the pelvic floor, care being taken to avoid involvement of the mucous coat. In this way the entire circumference of the recto-sigmoid dilates at the desired point due to infiltration of the serous and muscular coats.

A longitudinal incision is made with a scalpel above the intact pelvic floor into the infiltrated region, and separation of the seromuscular layer from the mucous layer is carried out with dissecting scissors and with the

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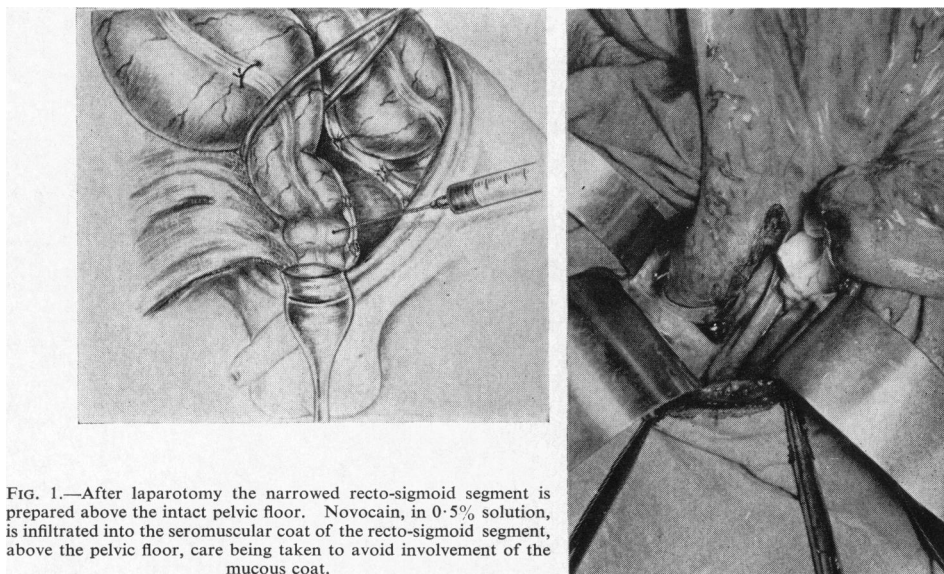


FIG. 1.—After laparotomy the narrowed recto-sigmoid segment is prepared above the intact pelvic floor. Novocain, in 0.5% solution, is infiltrated into the seromuscular coat of the recto-sigmoid segment, above the pelvic floor, care being taken to avoid involvement of the mucous coat.

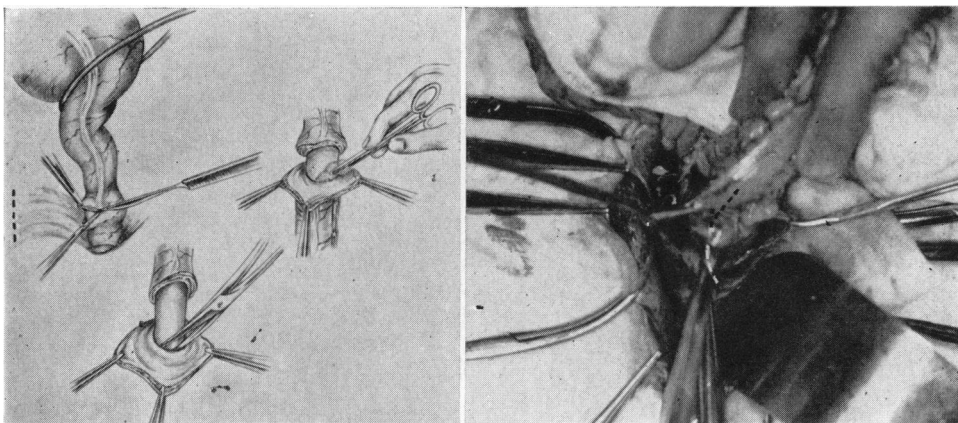


FIG. 2.—A longitudinal incision is made above the pelvic floor with a scalpel, and separation of the seromuscular layer from the mucous layer is begun and completed with dissecting scissors and with the aid of a small gauze pad held by a Klemmer forcep, until the proximal seromuscular coat is separated from the distal seromuscular coat and the underlying mucous coat is completely detached.

aid of a small gauze pad held by a Klemmer forcep, until, around the entire circumference of the bowel, the proximal seromuscular coat is separated from the distal seromuscular coat and the underlying mucous coat is completely detached (Fig. 2).

Dissection. Beginning at the edge of the distal seromuscular coat, with long curved dissecting scissors and with the aid of a small gauze pad soaked in adrenaline and held by a Klemmer forcep, the seromuscular coat is, progressively and carefully, detached from the underlying mucous coat (Fig. 3). This

manœuvre is carried out without trauma to the pelvic floor and with little difficulty or loss of blood, since the adrenaline acts as a haemostatic agent and only a few thin vessels are ligated.

Dissection is carried out, under visual control as far as possible and then with the right index finger, and should be extended distally as far as possible towards the anus (Fig. 4). The dissection of the mucous coat is easy in newborns and in older infants and children. At the point where the rectum joins the anus, at a level half-way up the levator ani muscles, tougher adhesions are found between the muscular and mucous coats. These are

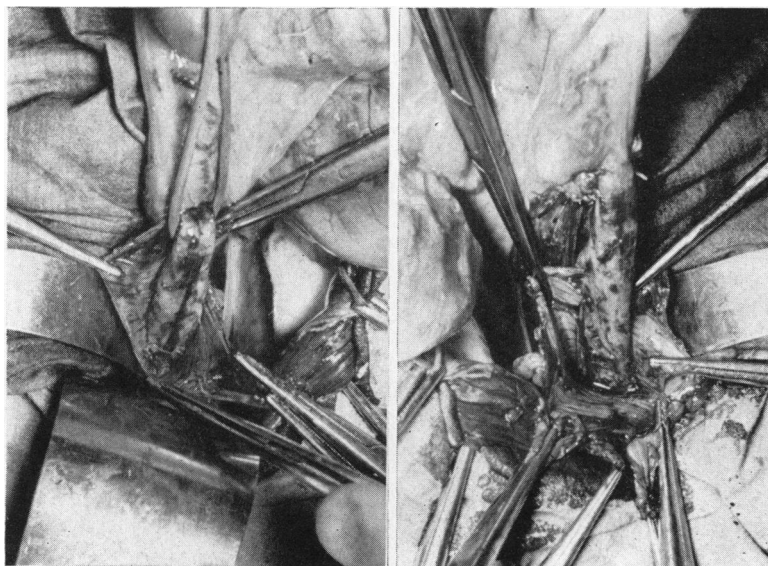


FIG. 3.—At the edge of the distal seromuscular coat, with long curved dissecting scissors and with the aid of a small gauze pad soaked in adrenaline and held by a Klemmer forcep, the seromuscular coat is detached from the underlying mucous coat.

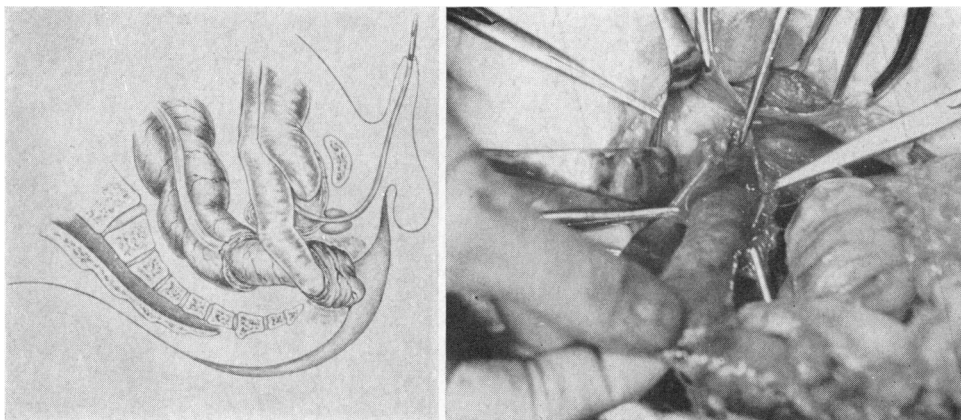


FIG. 4.—Dissection is carried out under visual control as far as possible and then with the right index finger and should be extended as distally as possible towards the anus.

carefully resected with the scissors (either by the abdominal or intra-anal route during the perineal stage) according to the anatomical situation.

By separating the coats of the recto-sigmoid segment by the extramucosal route as far as the ano-rectal line, pelvic innervation, together with the hypogastric, lumbosacral and sacral plexuses are spared (Fig. 5). This completes the intra-abdominal stage of the extramucosal detachment of the rectum. Before proceeding to the perineal stage a silk thread is stitched onto the colon to mark the point at which it is to be resected.

Perineal Stage. The anal canal is widely dilated and the mucosal cutaneous junction of the anus is stretched with four clamps in order to ensure as extensive a prolapse and exposure of the intra-anal mucosa as possible. A

scalpel (Fig. 6) or the scissors are used to make a slit 1 cm. above the muco-cutaneous junction in the intra-anal mucosa; the incision is then completed around the entire circumference.

In this way a tract of distal anal mucosa remains intact together with the perimucosal tissues.

The terminal mucous coat of the rectum is detached in its entire circumference, working upwards; a number of muscle fibres are resected near the level of the levator ani muscles which are thus spared. Preparation of the ano-rectal mucous coat is completed when the index finger of the abdominal operator, inserted between the muscular and mucous coats, encounters the finger of the perineal operator (Fig. 5).

The mucous coat of the recto-sigmoid is thus completely detached from its outer muscular and serous coats. Of

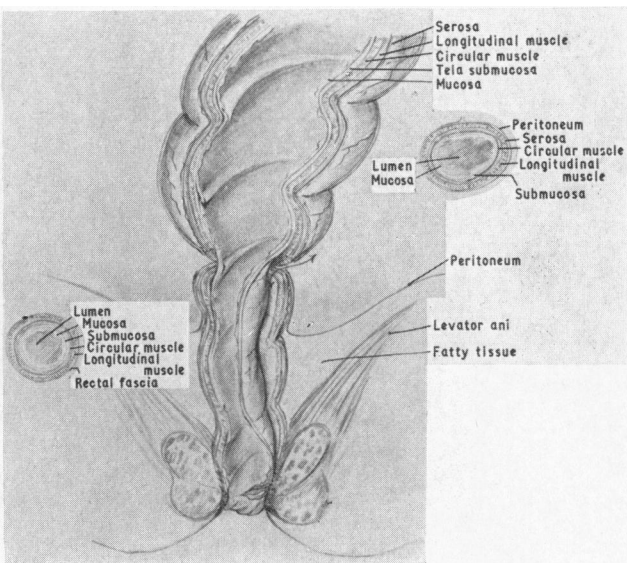
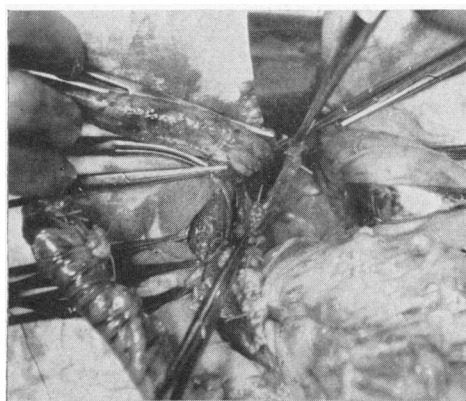


FIG. 5.—By separating the coats of the recto-sigmoid segment by the extramucosal route as far as the ano-rectal line, pelvic innervation, and the hypogastric, lumbosacral and sacral plexuses are spared.

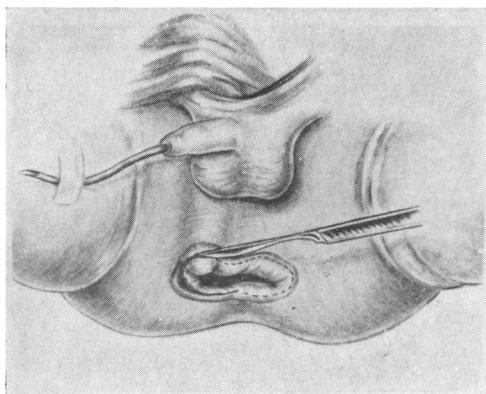


FIG. 6.—The anal canal is widely dilated and the mucosal-cutaneous junction of the anus is stretched with four clamps in order to ensure as extensive a prolapse and exposure of the intra-anal mucosa as possible. A scalpel or the scissors are used to make a slit 1 cm. above the mucous-cutaneous junction in the intra-anal mucosa; the incision is then completed around the entire circumference. In this way a small circular tract of distal anal mucosa remains intact together with the perimucosal tissue. The terminal mucous coat of the rectum is detached in its entire circumference working upwards: a number of muscle fibres are resected near the level of the levator ani, which are thus spared (Fig. 5).

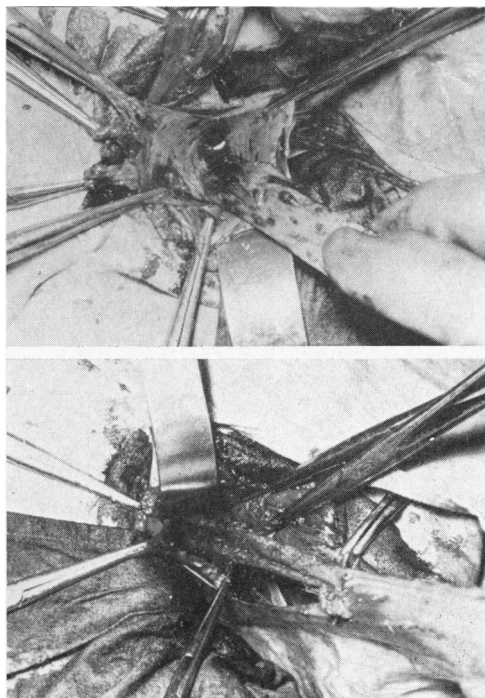


FIG. 7.—Of particular interest is the elasticity of the recto-sigmoid mucous coat. As the detachment manoeuvre proceeds, the mucous coat stretches, unlike the seromuscular canal which does not alter in length.

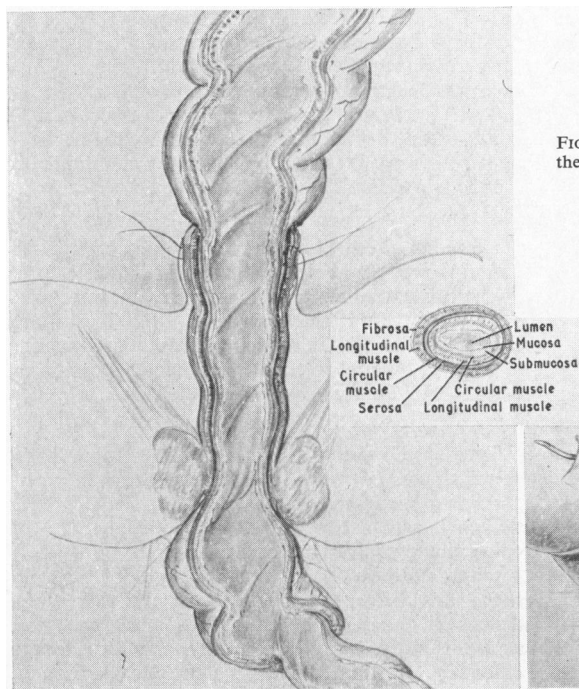


FIG. 8.—The mucous coat of the recto-sigmoid is withdrawn through the perineum or anus and the colon is pulled down beyond the anal orifice until the point of resection.

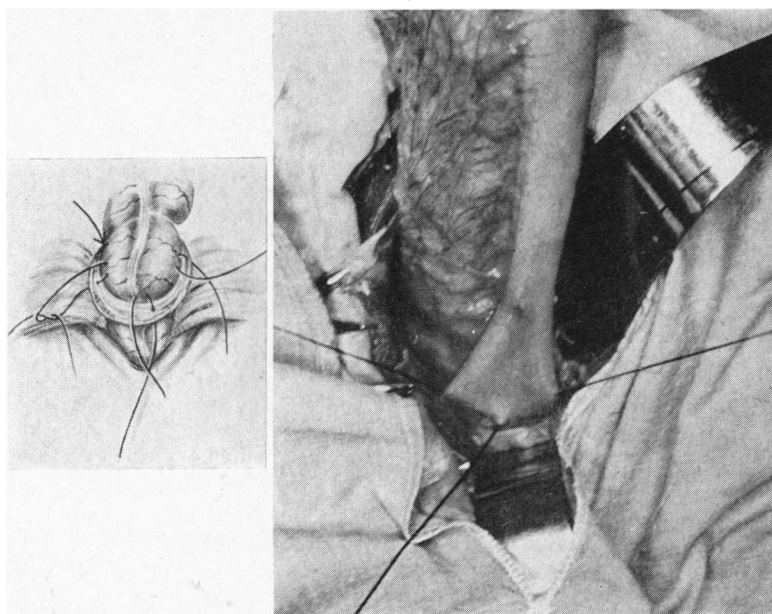


FIG. 9.—Drainage of the recto-anal canal. A Penrose drain is introduced between the lowered colon and the seromuscular coat to avoid accumulation of blood or serum above the pelvic floor, in the abdominal cavity and between the serosa of the colon and the rectal muscular canal.

particular interest is the *elasticity* of the recto-sigmoid mucous coat (Fig. 7). As the detachment proceeds, the mucous coat stretches, unlike the seromuscular canal obtained during the manoeuvre, which is resistant and does not alter in length. This resistance facilitates detachment of the recto-sigmoid mucous coat while the edge of the seromuscular layer is held with four clamps.

In this way the ano-rectal canal remains intact outside the recto-sigmoid mucous coat, and it is possible to spare the external and internal sphincters, levator ani muscles and the hypogastric, lumbosacral and sacral plexuses.

Pulling Down Recto-sigmoid (mucous coat) and Colon.

The mucous coat of the recto-sigmoid is withdrawn through the perineum or anus, and the colon is pulled down beyond the anal orifice until the point of resection marked by the silk thread. This 'pull-through' is carried out without danger of intra-abdominal infection, nor is there any risk to the innervation of the bladder, genital organs and sphincters (Fig. 8).

After preparing the colon, the pull-through may sometimes be a little difficult because of the presence of residual faecal masses in the colon, in spite of appropriate pre-operative preparation. In this case a big rubber tube is introduced into the recto-sigmoid segment and colon, and by means of a cleansing enema the faeces are softened and can then be easily expelled by gently squeezing the large bowel from the abdominal side.

Drainage of the Recto-anal Canal. With the aid of a Klemmer forceps a Penrose drain is introduced between

the lowered colon and seromuscular coat to avoid accumulation of blood or serum above the pelvic floor in the abdominal cavity (Fig. 9) and between the serosa of the colon and the rectal muscular canal.

After fixing the Penrose drain in position (indicated by the arrow) (Fig. 9), the round edge of the seromuscular layer protruding above the pelvic floor is stitched with four silk stitches to the outer surface of the colon. This achieves the twofold objective of closing the pelvic floor and attaching the colon to the seromuscular coat. Within two to four days the Penrose drain is removed.

Resection of the Colon. After the abdomen has been closed the lowered colon on the perineal side is resected at the predetermined point, leaving a stump protruding 5 to 10 cm. from the anus (Fig. 10). This stump is anchored with four silk stitches to the skin of the perineum (Fig. 10). A rubber tube is then introduced into the colon. The Penrose drain is visible.

Tube in Lowered Colon. A rubber tube or rectal sound 1 cm. or more in diameter (depending on the child's age) is inserted into the lumen of the colon and tied to it with a silk loop attached peripherally to the colon stump protruding from the anus.

Gases and intestinal contents are drained through the tube; this helps to avoid post-operative abdominal distension, and it may be left in place for several days, as a suture is not required: generally it is expelled spontaneously. Within 10-12 days a circular adhesion forms

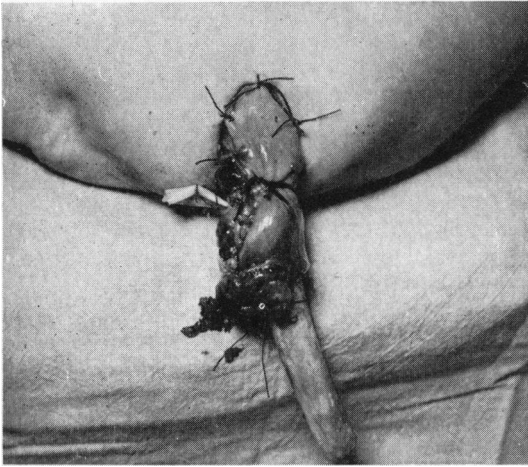


FIG. 10.—The lowered colon is resected at the predetermined point leaving a stump protruding 5-10 cm. from the anus. Penrose drain is visible.

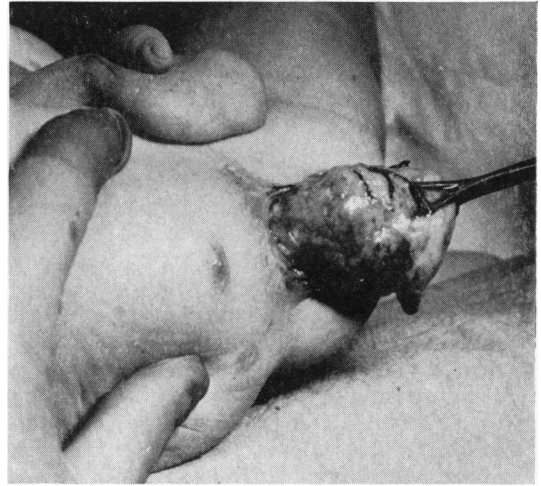


FIG. 11.—Generally within 15-20 days a circular adhesion forms between the serous coat of the colon, the mucous wall of the anus and the fibromuscular rectal canal.

between the serous coat of the colon, the mucous wall of the anus and the fibromuscular rectal canal (Fig. 11).

Resection of Colon Stump. After 15-20 days the colon

stump protruding from the anus is cut by cautery, and it is necessary to ligate the bleeding vessels that are found in the severed segment. The severed end withdraws into the anal canal after a few hours or sometimes immediately with slight finger pressure (Fig. 12).

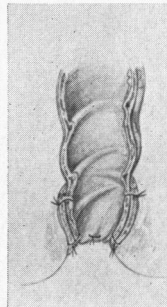
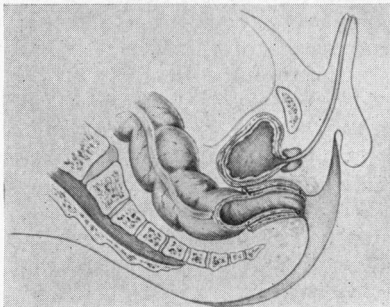
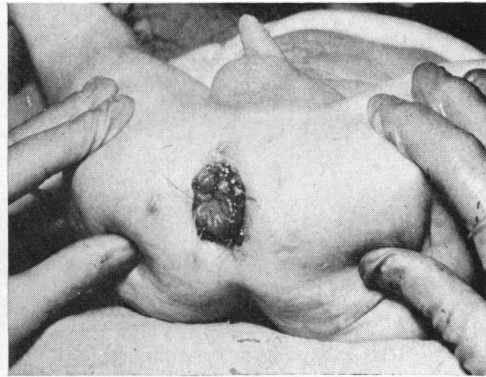
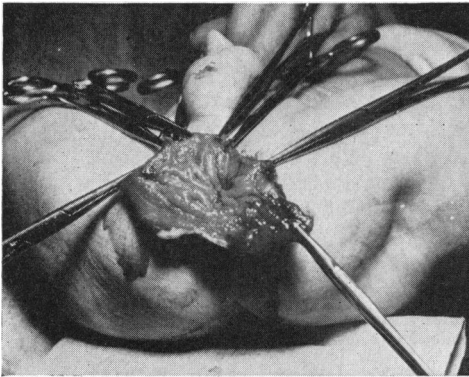


FIG. 12.—After 15-20 days the colon stump protruding from the anus is cut by cautery; the severed end withdraws into the anal canal after a few hours, sometimes immediately with slight pressure from the finger.

Discussion

By means of this technique which, although simple, requires considerable care, it is possible to achieve (without suture) an anastomosis between colon and anus, which is in good functioning order in several weeks, by which time the oedema of the colon stump (withdrawn into the anal canal) is resolved.

Digital examination of the rectum, following removal of the colon stump (after 15-20 days by cautery), enables one to follow the progressive resolution of oedema. At first this is felt as a protruding circular band within the intestinal lumen between the anus and the lowered colon (spontaneous anastomosis line), giving the impression of a rigid circular section. However, after daily dilatation, within six to twelve weeks it cannot be palpated, and the finger easily penetrates into the newly-formed anal canal.

Dilatation. Daily dilatation of the new anal canal is very important and is easily carried out during the period of oedema reabsorption, i.e. two to three months. Indeed no retraction remains at the level of

the ano-colon anastomosis, and there is only a circular ring between the anus and colon: although initially rigid this disappears within two to three months. The object of early dilatation is to avoid constriction of the colon during its post-operative period of readaptation. The oedema persists for several weeks and the walls of the colon require a little time to regain their normal peristaltic activity. By preserving the newly-formed ano-rectal canal with stimulation by dilatation the danger of faecal accumulation and irritation of the mucosa is avoided.

Before the patient is discharged, the mother is instructed in the dilatation procedure with a conic dilator with progressively increasing diameter, from to 1.2-5 cm. In this way only one dilator is required and need only be inserted once to obtain the desired degree of dilatation. Dilatation must be continued until the ano-colon anastomosis is calibrated.

Post-operative Physiology of Anal-colon Anastomosis. The segment of intact circular anal mucosa that remains after the operation is functionally normal and controls the defaecation reflex when the newly-formed rectum is full of faeces. Defaecation is dealt with by the colon and levator ani. These are still attached to the rectal muscular coat which has been left *in situ* and which, within a few weeks, adheres to the walls of the lowered colon.

The seromuscular rectal canal preserves its topographical anatomical relation with the levator ani and the nervous pelvic plexuses: these plexuses regulate functional stimuli for the colon and, together with the anal reflex and colonic peristalsis, ensure, better than any operation, the motility of the newly-formed rectum.

With this technique the colon is pulled down to the level of the anus, thus avoiding the risk of functional spasm and obstruction to faecal passage. This spasm often occurs after Swenson's operation, since the rectal segment remaining after the pull-through manoeuvre and end-to-end suture is always rather long and may also be aganglionic. This makes it susceptible to reflex stimuli which in turn lead to persistence of the functional obstruction and favour the accumulation of faeces and post-operative enterocolitis. Faecal accumulation and secondary post-operative enterocolitis are caused by functional (or organic) obstruction and must be avoided (so as not to revert to the pre-operative pathology) by removing the final direct cause of the aganglia, namely: *the continued presence of the non-functioning rectal segment.*

With the proposed technique this danger is avoided since the colon reaches the level of the anus.

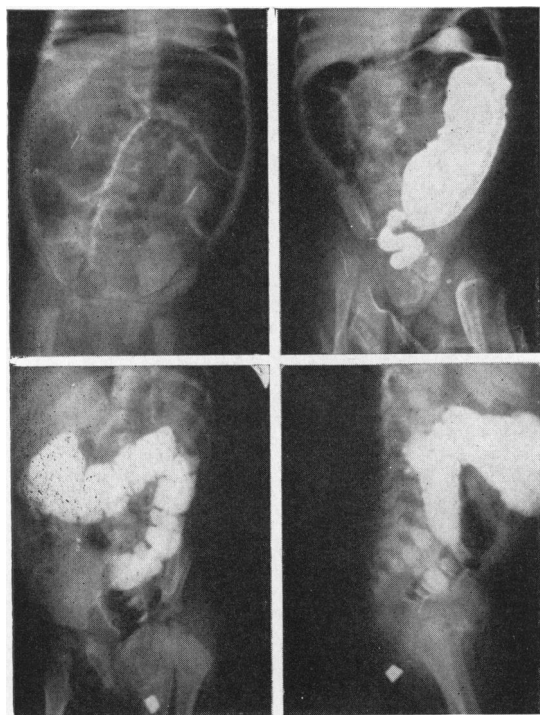


FIG. 13.—Pre-operative and post-operative radiographs of the 6-month-old infant. These show the morphological and dynamic features of the lowered colon and the peristaltic elasticity of the new rectum.

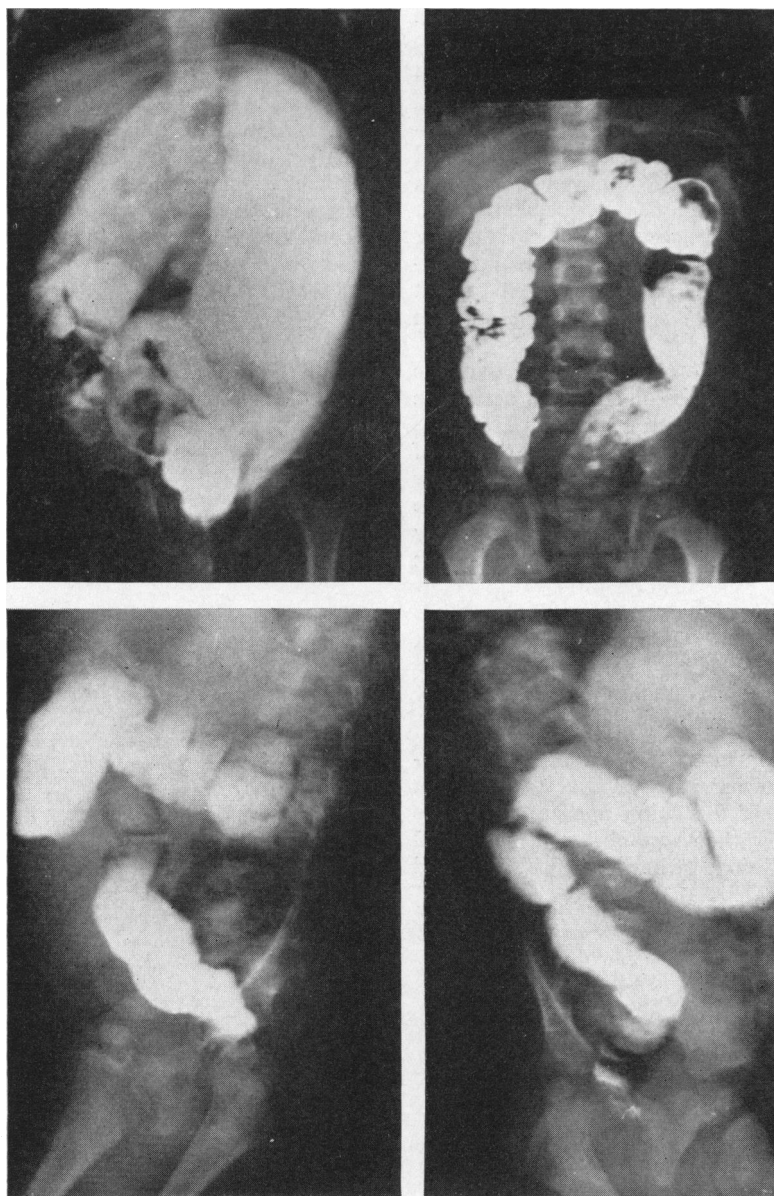


FIG. 14.—Pre-operative and post-operative radiographs of the 2-year-old infant. The final outcome of the operation is excellent as verified by radiography of the distal colon and new rectum.

Clinical Findings. The technique was used in 14 patients aged respectively 2 months (one patient), 5 months (three patients), 6 months (one patient), 2 years (three patients), 5 years (two patients), 6 years (two patients) and 7 years (two patients). The post-operative course was excellent in each case, and the tube ensured canalization of the bowel within one to two days. No bladder disorders

occurred; sphincter control was complete from the beginning. A number of patients were followed up for over 20 months and according to the parents the final outcome was excellent in each case.

Fig. 13 shows the morphological and dynamic features of the lowered pulled-through colon and the peristaltic elasticity of its most distal tract; this crosses the ano-rectal canal which has remained

intact after extra-mucosal detachment of the recto-sigmoid segment. This patient (Fig. 13) was a 6-month-old girl who had had enterocolitis and obstruction due to faecaloma of the narrowed segment and who at the age of 21 days had been submitted to operation for the formation of a sigmoid anus. At 6 months the child was operated on in a single stage with the technique described here. She did well and was in excellent health seven months after the operation. Anal dilatation was practised immediately and continued for four months. At present the newly-formed rectum is soft and of uniform structure.

Fig. 14 shows the morphological and dynamic features of the lowered colon and the peristaltic elasticity of the new rectum segment. This case concerns a 2-year-old boy with a classical picture of Hirschsprung's disease and a very long narrow recto-sigmoid segment. The final outcome of the operation was excellent, as verified by radiography of the distal colon (neo-rectum), and seven months later the child was in excellent health.

Dilatation was practised by us and the child's mother for five weeks. Intestinal function is at present normal.

Conclusions

The proposed non-suture technique of colon-anal-anastomosis, following extramucosal mobilization and pull-through of the colon appears to be the surest technique for the re-establishment of normal physiological conditions of intestinal peristalsis and

of function of the new rectum, since it spares pelvic innervation and does not cause disorders of the bladder, genital organs or internal and external sphincters.

The proposed technique is a radical and not a palliative treatment of Hirschsprung's disease and may be used in newborns, older infants and children. Post-operative dilatation is very important and must be continued daily for two to three months and more, if necessary.

BIBLIOGRAPHY

- Duhamel, B. (1959). Exclusion du rectum avec abaissement rétro-rectal et trans-anal du colon. Applications et premiers résultats d'une technique nouvelle. *Mém. Acad. Chir.*, 85, 192.
- Ehrenpreis, T. (1961). Long-term results of rectosigmoidectomy for Hirschsprung's disease, with a note on Duhamel's operation. *Surgery*, 49, 701.
- Hirschsprung (1888). Stuhlträgheit Neugeborener in Folge von Dilatation und Hypertrophie des Colons. *Jb. Kinderheilk.*, 27, 1.
- Pellerin, D. (1962). Le traitement chirurgical de la maladie de Hirschsprung par la résection-anastomose exteriorisée sans suture. *J. int. Coll. Surg.*, 37, 591.
- Rehbein, F. (1959). Operation der Anal- und Rectumatresie mit Recto-Urethral-fistel. *Chirurg*, 30, 417.
- Romualdi, P. (1960). Eine neue Operationstechnik für die Behandlung einiger Rectummissbildungen. *Langenbecks Arch. klin. Chir.*, 296, 371.
- (1961). Treatment of some particularly difficult cases of anus prostaticus. *Riv. Chir. pediat.*, 3, 27.
- Soave, F. (1963a). Die Nathlose colon-ano-stomie nach extramuköser Mobilisierung und Herabziehung des Rectosigmoids. Zur chirurgischen Behandlung des M. Hirschsprung. *Zbl. Chir.*, 88, 31.
- (1963b). Una nuova tecnica chirurgica per la terapia della malattia di Hirschsprung. *Osped. Ital.-Chir.*, 8, 3.
- (1963c). Une nouvelle technique chirurgicale pour le traitement de la maladie de Hirschsprung. *J. Chir. (Paris)*, 86, 5.
- , Bertolini, A. and Mantero, R. (1962). Il trattamento chirurgico del morbo di Hirschsprung secondo il principio di Duhamel. *Inform. med. (Geneva)*, 17, 21.
- Swenson, O. (1957). Follow up on 200 patients treated for Hirschsprung's disease during a ten-year period. *Ann. Surg.*, 146, 706.