ANO-RECTAL ANOMALIES: PHYSIOLOGICAL CONSIDERATIONS*

BY

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We have used a classification of ano-rectal anomalies following the concepts of Keith (1908), Stephens (1953a, b), Browne (1955) and Bill and Johnson (1958), as described elsewhere (Nixon, 1961).

The low anomalies in which the 'anus' or 'fistula' is below the puborectalis present few serious problems. The main groups are the covered and ectopic anus, and continence is achieved by adequately enlarging the opening where it lies by cutback and dilatation. Transplantation need only be considered later as a secondary procedure in the minority who need it for mainly aesthetic reasons. Puborectalis is intact around the bowel but the external sphincter is behind the anus in the common vulval and vestibular ectopic ani. The rectal sensation is not interfered with by the minor procedures used, but anal sensation must be deficient in these ectopics because the proctodaeum is itself deficient. Apparently it is not necessary under these conditions because the continence is good as shown by Partridge and Gough (1961) in their follow-up of the Great Ormond Street series.

In Swenson's procedure for Hirschsprung's disease it seems that the maintenance of anal sensation and the full sphincter complement usually allows continence in the absence of rectal sensation. (In Hirschsprung's disease the internal sphincter does not relax normally; but the same procedure has been used for severe megarectum without aganglionosis, and continence was still maintained.)

Thus it seems that there are two sensory and two motor elements normally involved in continence, but loss of one at least may not cause incontinence: rectal sensation (visceral); puborectalis (striated); anal sensation (somatic); external sphincter (striated); internal sphincter (unstriated). The internal sphincter seems to be more concerned with prevention of soiling than frank incontinence.

In the 'high' ano-rectal anomalies the bowel stops above the puborectalis sling, and ano-rectal agenesis

in the male is the common one. Our results with the abdomino-anal pull-through operation have been poor, only half of the survivors having adequate continence (Partridge and Gough, 1961). Certainly as they got older some poor results became fair and some fair results became good, and more will do so with further time and training. But this is a low standard to accept. It was observed that a good contracting puborectalis sling was no guarantee of continence even though the vulval ectopic anus can be completely continent with this muscle alone. In such cases anal sensation is clearly absent as there is no proctodaeal lining. It might be expected that rectal sensation would be destroyed by the mobilization of the bowel for the pull-through. If so an alternative procedure avoiding this mobilization might be expected to be more effective. On the other hand if the degree of agenesis is such that the more sensitive lower part of the rectum is undeveloped then this change could not be expected to help. Physiological studies were, therefore, carried out to investigate this point (R.P.C.) as described below. They showed that a sphincter reflex response to rectal distension was present in cases examined before operation but was lost in cases examined after operation.

Stephen's sacral approach (1953a, 1961) was also followed by interference with the reflexes as was my only case treated by Rehbein's (1959) operation. Hiatt and Santulli (1962) now lay great stress on the anal stage of the abdomino-anal operation so as to bring skin up into the anal canal as a sensory area. In the past I have found that the skin flaps of such a cruciate incision were prone to descend to the surface of the perineum and though they helped to avoid a troublesome ring stricture they did not stay within the canal to play a part in sensation.

An alternative attitude to correction, which might be more successful in avoiding damage to mechanisms and use all that are present, has therefore been considered: this is the concept of constructing a proctodaeum to reach up to the bowel rather than

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FIG.—Relation of sphincters in A normal and B ano-rectal agenesis.

mobilizing the bowel to bring it unnaturally far down. For much of the 'gap' is the result of an abnormal distance between the levator floor and the external sphincter, caused by the meeting of the ischio-rectal fat pads in the midline in the absence of the terminal bowel to keep them apart (Fig.). If the natal cleft can be deepened to its normal extent a 1-2 cm. canal extending through the pelvic floor is all that is needed to reach the end of the rectum. Such a procedure has recently been carried out by a modification of Stephen's operation on three occasions but a definitive technique has not yet been worked out. The purpose of this paper is rather to give an account of the pathophysiological findings.

Method

The apparatus was the same as that used to study megarectum (Callaghan and Nixon, 1964). The infants were too young to report sensation but the objective response to inflation of the balloon in the rectum could be recorded.

Results

Ano-rectal Agenesis. Ten cases were examined before and after operation (Table 1).

Abdomino-anal Pull-through. Post-operative response to 'rectal' distension was recorded in 12 cases aged $3\frac{1}{2}$ to 14 years. There was 'rectal' sensation in

TABLE 1

	No. Tested	Response to Rectal Distension	
		Induced Sphincteric Activity	Eventual Sphincteric Inhibition
Before operation	10	10	9
After operation			
Stephen's operation ..	5	2 + ? 1	0
Rehbein's operation ..	1	0	0
Modified pull-through ..	1	0	0

1, colonic (central abdominal pain) sensation in 6 and no sensation in 5 cases. Sphincter response was present in 6 and absent in 6 cases. Eventual sphincteric inhibition was absent in all 12 cases.

Post-operative studies were also made in cases with low anomalies. The findings showed that delay in treatment resulted quite frequently in the megarectum (rectal inertia) syndrome.

Low Anal Anomalies. These comprised covered, ectopic and stenotic anal anomalies (Table 2). Twenty-six cases were tested after operation at ages between 1 and 20 years. All showed co-ordinated sphincter responses to rectal distension; 8 had normal rectum; 18 had some degree of megarectum (rectal inertia).

This is of practical importance because of the common tendency of paediatricians to try to 'tide over' these babies until they are bigger before referring them to a surgeon. In fact the minor

TABLE 2

TYPE OF RECTUM IN TREATED LOW ANAL ANOMALIES

Type	Age at Treatment		
	< 3 months	3 months-1 year	1 year and over
Normal	4	2	1
Enlarged	1	1	0
Expanded	2*	4	0
Inert	1*	1*	9

* Inadequate treatment

procedure needed is better performed in the neonatal period to avoid the development of a megarectum (rectal inertia) syndrome secondary to the stenosis. Furthermore early operative treatment allows completion of the dilatations required in after-care before the baby reaches a psychologically more vulnerable age, and allows development of bowel control at the normal time.

Summary

Pathophysiological examinations in rectal agenesis show the presence of a sphincteric reflex response to rectal distension before operation and its loss after commonly used operations. Possible means of avoiding this are discussed.

Normal reflex responses were found in low anal anomalies (ectopic, covered, stenotic anus) after operation. The development of secondary megarectum was found when their treatment was delayed; early performance of the minor procedures is, therefore, advised.

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