ANO-RECTAL MALFORMATIONS — THEIR MANAGEMENT AND SEQUELAE

MICHAEL KATZEN, M.B., B.CH. (RAND), F.R.C.S. (EDIN.), *Department of Surgery, University of the Witwatersrand, Johannesburg*

Considerable difference of opinion is expressed in the literature concerning the morbidity and mortality associated with congenital malformations of the ano-rectum. In order to clarify impressions previously gained in the management of these cases, a survey of 41 unselected patients treated for this condition at the Transvaal Memorial Hospital for Children, Johannesburg, between 1949 and 1960, has been undertaken. An attempt has been made to follow these patients up either by letter or by personal examination.

It must be appreciated that a large number of these patients were referred to the hospital after having received temporary or definitive treatment elsewhere and the management of many patients in no way reflects the routine or the methed used by the surgeons in attendance at the Transvaal Memorial Hospital. This series also serves to illustrate the marked difference in the standards of management of this condition outside and inside recognized paediatric institutions, and bears out the words of Willis Potts[1]: 'Is it quite fair that a surgeon who would not think of operating on a brain tumour or repairing a cleft palate should tackle a case of atresia of the rectum in a male infant with a recto-vesical fistula, when he never before has seen such a case nor witnessed the operative repair? If the defects are not properly corrected at the first operation, these patients may be forever incontinent or doomed to a permanent colostomy'.

An excellent review of this subject, covering most of its facets, was published by Prof. J. H. Louw in this *Journal* in 1959,[2] and it is not proposed here to describe the embryology of the condition or the detailed anatomy and clinical presentation of the various anomalies. Only the treatment, X-ray diagnosis, associated anomalies, morbidity and mortality will be discussed.

PRESENT SERIES

The classification of ano-rectal malformations in the present series is similar to that used by other recent authors[2,3] and is based on the original classifications of Browne[4] and of Stephens.[5,6] The classification of Ladd and Gross[7] has fallen out of favour, since most of these malformations appear to fall into their 'type 3' and their grouping provides no guide to the embryological aberration, treatment or prognosis.

The present series is depicted in Table I and is com-

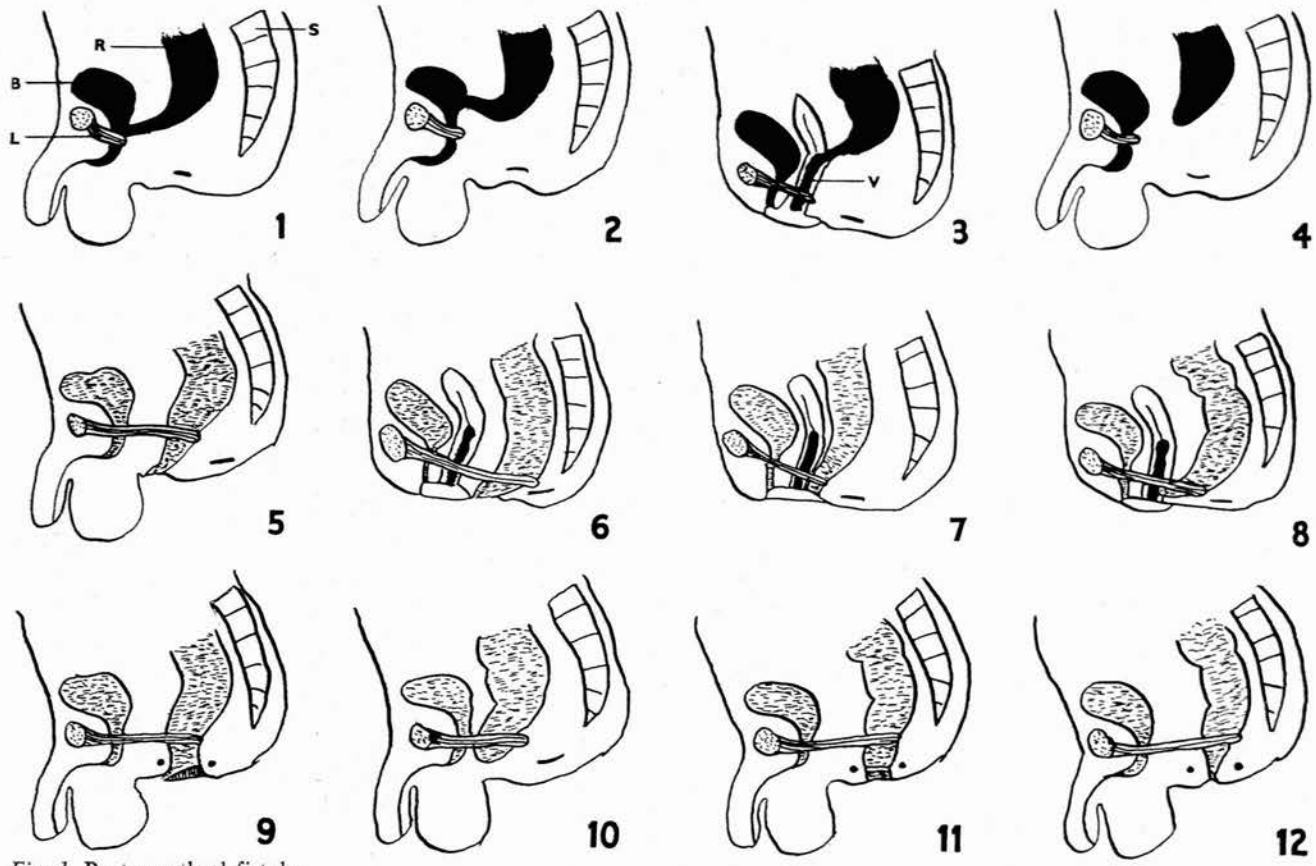

Fig. 1. Recto-urethral fistula.
Fig. 2. Recto-vesical fistula.
Fig. 3. Recto-vaginal fistula.
Fig. 4. Rectal agenesis without fistula.
Figs. 1 - 4 represent various types of rectal agenesis.
Fig. 5. Perineal ectopic anus (male).
Fig. 6. Perineal ectopic anus (female).
Fig. 7. Vestibular ectopic anus.
Fig. 8. Vaginal ectopic anus.
Fig. 9. Covered anus (ano-perineal 'fistula').
Fig. 10. Imperforate anus.
Fig. 11. Imperforate anal membrane.
Fig. 12. 'Microscopic' or stenosed anus.
Key: R=rectum, V=vagina, B=bladder, S=sacrum, and L =levator ani.

pared with the incidence of anomalies in the series of Louw[2] and of Partridge and Gough.[3] Partridge and Gough do not include the group 'imperforate anus' in their series, and there appears to be a disproportionately large number of patients in this group in the present series as compared with that of Louw. This matter is discussed more fully later.

The abnormalities encountered are depicted diagrammatically in Figs. 1 - 12. No examples of covered anus with ano-bulbar or ano-vulvar 'fistulae' were encountered in this series, nor were any gross abnormalities, e.g. persistent cloaca. There were no cases of rectal atresia.

RECTAL AGENESIS

The 18 cases in this group are enumerated in Table II, together with the primary treatment used, the age at which a 'pull-through' procedure was carried out, the complications, and the deaths.

TABLE I. COMPARISON OF THREE SERIES OF ANO-RECTAL MALFORMATIONS

Condition	*Transvaal Memorial Hospital*	*Louw*[2]	*Partridge and Gough*[3]
Rectal agenesis:			
Recto-urethral fistula (M) ..	13	27	
Recto-vesical fistula (M) ..	1	1	
Recto-vaginal fistula (F) ..	2	2	
Without fistula	2 (M, M)	2 (F, M)	
Total	18	32	114
Anal anomalies:			
Ectopic anus	11	26	98
Perineal (M)	2	1	
Perineal (F)	2	2	
Vestibular (F)	5	16	
Vaginal (F)	2	7	
Covered anus	4	18	53
Ano-perineal 'fistula' (M)	4	15	
Ano-bulbar 'fistula' (M) ..	0	1	
Ano-vulvar 'fistula' (F) ..	0	2	
Imperforate anus (M)	5	2	0
Imperforate anal membrane ..	2 (M, M)	1 (M)	3
'Microscopic' (or stenosed) anus	1 (M)	0	38
Total	23	47	192
Gross abnormalities	0	6	12

M=male, F=female.

1. *Recto-urethral Fistula (Fig. 1)*

Of the 13 patients with recto-urethral fistula, only 1 was treated at birth by the recommended primary 'pull-through' operation described by Rhoads *et al.*[8] This child died suddenly from an unknown cause the day following operation.

Eight patients were treated primarily by colostomy only, and 1 by colostomy combined with a perineal approach to divide the fistula and bring down the rectum. This latter operation, performed by a surgeon not on the staff, resulted in a persistent recto-urethral fistula, which was repaired and the colostomy closed when the child was 9 years old. One patient, in whom a duodenal atresia coexisted with the rectal malformation, was treated by colostomy and duodeno-jejunostomy, but he died shortly after operation.

One colostomy was performed because a tracheo-oesophageal fistula coexisted; the child died from a leak from the oesophago-oesophageal anastomosis. Two colostomies were performed because the birth weight was only 5 lb. Colostomy was performed in the other 4 patients as the method of choice, with a view to performing a 'pull-through' operation at a later date.

Four of the colostomies were followed by 'pull-through' procedures, 2 at 6 months of age and 2 at 1 year. A fifth patient did not return for a further operation. Follow-up of these patients showed that 3 had prolapse of the rectal mucosa, and 1 of these needed trimming. Two babies are still too young for follow-up, though the mother of one states that the child's diapers are always soiled (aged 9 months). One child is untraced, and the fourth was seen at 5½ years with no sphincteric control and a very constricted anus, which had to be dilated under general anaesthesia.

Of the 10 colostomies performed, 6 were transverse, 3 inguinal and 1 'near the splenic flexure'. One inguinal colostomy and the 'splenic flexure' colostomy caused considerable difficulty when 'pull-through' operations were later performed, necessitating resection of the distal recto-sigmoid and anastomosis of the colon to skin in one case.

2. *Recto-vesical Fistula (Fig. 2)*

This child underwent an abdomino-perineal 'pull-through' shortly after birth. Redundant mucosa had to be trimmed at 1 year, and at the age of 4 the child passes 4 stools a day, but the mother states that the child has no rectal sensation and has to be taken to the lavatory as a routine and forced to pass a stool.

3. *Recto-vaginal Fistula (Fig. 3)*

Of the 2 patients in this group, one had a transverse colostomy performed at birth and died from a cardiac abnormality 2 weeks after discharge.

The other child was operated on at an outside hospital at birth, when a colostomy and a 'pull-through' operation were performed. On admission at 3 months, it was found that a stricture had formed just above the recto-cutaneous junction. A further 'pull-through' procedure was done, which broke down, resulting in a recto-vaginal fistula. This was followed by a small-bowel obstruction necessitating laparotomy. The child died a few weeks after this last operation, performed elsewhere, of '?chest infection'.

4. *Rectal Agenesis without Fistula (Fig. 4)*

The first child in this group died of renal failure 4 months after a left inguinal colostomy was performed. Fused kidneys were found at postmortem examination.

The second child died aged 4 days, from haematemesis, following a 'pull-through' operation at 2 days. At postmortem examination a large acute ulcer of the pylorus was found.

ANAL ANOMALIES

1. *Ectopic Anus*

(*a*) *Perineal (male)*—*Fig. 5.* In the first patient in this group, the 'fistula' was situated very far anteriorly and was found to be stenosed up to the level of the levator ani. The 'fistula' was excised, and the rectum mobilized *via* the perineum and sutured to skin at birth. The child died of a cardiac anomaly at 4 months.

TABLE II. RECTAL AGENESIS

		Treatment	Age at 'pull-through'	Remarks	Deaths
Recto-urethral fistula	1	Colostomy at birth – later 'pull-through'	6 months	Premature. Prolapse of mucosa	
	2	Colostomy at birth – later 'pull-through'	6 months	Prolapse of mucosa. Later stricture	
	3	Colostomy at birth – later 'pull-through'	1 year	Prolapse of mucosa, trimming needed	
	4	Colostomy at birth – later 'pull-through'	1 year	Urinary infection ++. Herniation of colostomy	
	5	Colostomy at birth + perineal approach		Persistent recto-urethral fistula	
	6	Colostomy only		Did not return for 'pull-through'	
	7	Colostomy only			Associated tracheo-oesophageal fistula
	8	Colostomy only			Prematurity
	9	Colostomy only			'Cardiac' + micrognathia
	10	Colostomy + duodenojejunostomy		Associated duodenal atresia	'Postoperative', ? other anomalies
	11	Primary 'pull-through' operation	2 days		? 'shock' after operation
	12	Perineal approach only		Persistent recto-urethral fistula	
	13	4 unknown previous operations		Persistent recto-urethral fistula	
Recto-vesical fistula	14	Primary 'pull-through' operation	2 days	Prolapse of mucosa, trimming needed	
Recto-vaginal fistula	15	Colostomy only			'Cardiac'
	16	'Pull-through' + colostomy	2 days	Severe stricture. Further 'pull-through' at 3 months	Intestinal obstruction + '? chest infection'
Rectal agenesis without fistula	17	Colostomy only			Renal failure—fused kidneys
	18	Primary 'pull-through' operation	2 days		Haematemesis—acute pyloric ulcer

TABLE III. ECTOPIC ANUS—VESTIBULAR AND VAGINAL

		Age at presentation	*Operation performed*	*Age at operation*	*Follow-up*
Vestibular	1	3 years	Transplant of anus	3 years	Persistent faecal impaction after operation
	2	1 day	Transplant of anus	1 day	Mild colonic inertia at 8 months
	3	3 years	Transplant of anus	3 years	Incontinent on discharge. No further report
	4	3 months	Transplant of anus	3 months	Quite normal at 11 years
	5	6 weeks	Colostomy at 6 weeks. Transplant of anus	3 months	Fairly severe colonic inertia at 4 years
Vaginal	6	3 months	Transplant of anus	3 months	Severe colonic inertia at 2 years
	7	2 weeks	Abdomino-perineal 'pull-through'	3 weeks	Died—'inhalation of vomitus'

In the second patient, the anus was transplanted posteriorly at the age of 2 months. The reason for the preference of this procedure to a 'cut-back' procedure or simple dilatation is not known. This child, after 5 years, has a megacolon with excessive constipation unresponsive to everything except rectal washouts. There is no stricture present. The child is also a 'cardiac', is mentally retarded and has craniosynostosis, for which an operation has been performed.

(*b*) *Perineal (female)—Fig. 6.* The first patient in this group presented at 9 months with faecal impaction so severe that a left inguinal colostomy was performed. Considering the possibility of superimposed Hirschsprung's disease, a biopsy of distal colon was done which showed normal ganglion cells. Rectal biopsy was also done; this showed no ganglion cells. However, this latter biopsy was not thought to be a representative specimen. The colostomy was closed 4 months later and, when seen 1 month after this, the child was again found to have faecal impaction.

The second child in this group was admitted when 27 days old, with extreme constipation, and was treated with regular dilatations. When seen again after 3 years, she was found to have anal stenosis with severe constipation, and dilatations and rectal washouts were recommenced.

(*c*) *Vestibular and vaginal (Figs. 7 and 8).* These two groups are considered together, since their management is essentially similar. The 7 patients are enumerated in Table III, together with their age of presentation, treatment and follow-up. Two of these patients presented with severe faecal impaction, one at 3 years and one at 6 weeks. The latter had a colostomy performed and intense diarrhoea ensued from which the child almost died. Six patients had a posterior transplantation of the anus performed. On 1 it was performed at birth, on 3 at 3 months, and on 2 at 3 years. Of the 5 patients followed-up, 2 suffered recurrent bouts of faecal impaction, necessitating daily enemas, when seen at the age of 4 years. One child, aged 4, always soiled her underclothes, required dilatation every second day, and suffered occasional diarrhoea, but her mother is sure that rectal sensation is intact. One child, when seen 4 months after operation, aged 3½, appeared incontinent of faeces, and 1 child, aged 11, had completely normal bowel function. The seventh child of this group underwent an abdomino-perineal 'pull-through' procedure at 3 weeks. An erroneous diagnosis of high recto-vaginal fistula had presumably been made. This child died the day following operation from what appears to have been aspiration of vomitus.

2. *Covered Anus—Ano-perineal 'Fistula' (Fig. 9)*

All the children in this group were males. Two were treated by 'de-roofing' the sinus in the perineum and by anal dilatations. There is no follow-up of these 2.

The third patient was unaccountably treated by left iliac colostomy. This child had a concomitant gross sacral abnormality and died of cardiac failure on the 14th postoperative day.

The fourth patient had a $\wedge$-shaped raphe over the anus and was treated at an outside hospital with a 'descending' colostomy at 2 days of age. On admission, all that was necessary was to snip away the raphe and dilate the anus. The colostomy was subsequently closed.

3. *Imperforate Anus (Fig. 10)*

There appears to be a disproportionately large number of cases of imperforate anus in this series. However, it has been confirmed by the operative findings that the rectum in each case extended to within ½ - 1 cm. of the perineal skin and no 'fistula' to the perineum was present. All the patients were males and this is in accordance with the findings of Stephens.[6]

In 1 of the 5 patients a perineal dissection alone was performed, and the rectum was mobilized and sutured to skin. No follow-up of this patient has been possible.

In 1 patient a perineal approach failed to mobilize the rectum adequately and an abdomino-perineal approach had to be used. When seen 3 years later, this child was found to have an anal stricture with a massive megacolon. Despite treatment, the megacolon has persisted up to the age of 8, with overflow incontinence a prominent feature.

In 2 patients a perineal exploration was carried out at birth and the operation notes state that a recto-urethral fistula was found and divided during the dissection. A supplementary suprapubic cystostomy was performed in both. In the first child a follow-up at 3 years revealed 'satisfactory bowel action'. In the second, urine was passed through the anus post-operatively. At the age of 5 years a perineal repair of the recto-urethral fistula was performed. At the age of 8, a urethral stricture was diagnosed and a bladder calculus removed. The child, at this stage, did not appear to have complete rectal control.

The last child in this group was found, on admission, to have signs of peritonitis in addition to the rectal deformity. X-rays in the inverted position showed gas well below the pubo-coccygeal line[5] and erect plates demonstrated free gas below the diaphragm. Laparotomy disclosed free gas and bowel content in the peritoneal cavity. A perforation of the bowel could not be found and a caecostomy was performed with peritoneal drainage. The child died on the 15th postoperative day.

4. *Anal Membrane (Fig. 11)*

The first patient in this group was treated by incision of the membrane and dilatations. This patient was also operated on for tracheo-oesophageal fistula, and follow-up at 15 months revealed normal bowel action.

In the second patient, the membrane had ruptured before admission. The rectum was freed and sutured to skin. It is not known why this treatment was adopted instead of dilatation alone. At 4 months there was evidence of slight stricture formation, but this responded well to repeated dilatations.

5. *'Microscopic' anus (Fig. 12)*

This patient presented when 8 days old. All that was required was repeated dilatation. At 6 years this child was treated for chronic fissure-in-ano and now, at 8 years, has perfectly normal rectal function.

DISCUSSION

Rectal Agenesis

There are 5 possible methods which may be employed in the treatment of these patients:[3]

1. Division of the recto-urethral, -vesical or -vaginal fistula and abdomino-perineal 'pull-through' of the rectum in one stage.

2. Division of the fistula and rectal pull-down *via* the perineal route only.

3. Division of the fistula and rectal pull-down by a sacral approach.[9]

4. Division of the recto-urethral fistula and transverse colostomy, the definitive rectal 'pull-through' being performed later.

5. Simple colostomy with later rectal 'pull-through'.

Most authorities favour method 1,[2,3,9-11] since it has the advantage of restoring the child to normal as soon as possible. Though 2 of the 4 'pull-throughs' performed on neonatal patients in this series resulted in fatalities, it is not suggested that this is an indictment of the procedure, since there were other reasons for both these deaths. Most authorities agree that the mortality associated with this procedure in the neonatal period is not higher than if it is performed at a later stage.[3]

Method 2 is frowned upon. This operation is exceedingly difficult, causes considerable trauma to vital pelvic nerves and musculature and, in all 3 patients managed in this way in this series, resulted in persistent recto-urethral fistulae which required multiple further operations to effect their closure.

Method 3 was not used in any patient in this series and, though attractive anatomically, appears technically difficult.

Method 4 was not used in this series and appears to have the disadvantage of allowing the rectum to 'ride-up' after division of the fistula, thus making subsequent mobilization difficult and extensive.

Method 5 was used in 11 patients in this series. In 5 there was an associated abnormality from which the children ultimately succumbed, and 2 children were premature. In 4 it was used as the procedure of choice as a preliminary to later 'pull-through', though further indications might have included possible inexperience of the operator in the technique of the 'pull-through' procedure, or late referral of patients when excessive bowel distension might have made a 'pull-through' extremely difficult.

There are a number of points against the use of primary simple colostomy:

(*a*) Older infants resent dilatations and parents find them difficult to manage.

(*b*) The older infant tends to develop excessive excoriation of the buttocks with its attendant hazards.[3]

(*c*) Colostomy itself might be a hazardous undertaking in the neonatal period *(vide infra).*

(*d*) Urinary tract infection is avoided.

In defence of primary colostomy followed by later 'pull-through', it must be stated that, although this series is small, there appeared to be no substantial difference in the functional result obtained from either primary or secondary 'pull-through'. It must be emphasized, however, that the 'pull-through' should follow the colostomy with as little delay as possible, i.e. when the child weighs 10 - 12 lb. The hazard of waiting longer is illustrated in one patient in this series in whom the secondary procedure was performed 1 year after the colostomy, during which time he suffered recurrent urinary infections which inflicted considerable renal damage. Hyperchloraemic acidosis has also been reported in these patients as a result of absorption of urine from the distal colon.

It is interesting to note that in all cases of abdomino-perineal 'pull-through', done for recto-urethral fistula, a concomitant suprapubic cystostomy was performed. Though this is not usually believed to confer any added benefit, it is noted that it produced no untoward complication in this series.

Anal Anomalies

Analysis of this group of cases raises several important points:

1. Most of the patients with ectopic anus presented with severe constipation and it is evident from the follow-up that treatment had been delayed for so long in many cases that 'colonic inertia' with its attendant sequelae had been allowed to develop. It is stressed by recent authors[3,12] that, if definitive treatment is not carried out early, it must at least be assured that the ectopic anus is kept well dilated and functioning adequately until the operation is performed.

Considerable controversy exists about the best operative treatment for vestibular or vaginal ectopic anus. The 'cut-back' procedure of Denis Browne[4] appears to have lost favour and most authorities now recommend formal transplantation of the anus to its normal position.[3,11,12] The 'cut-back' may, however, be used as a preliminary procedure in the neonatal period if the anus is stenosed, but the new anus so formed usually retracts to its former position and a formal transplantation becomes necessary later.

Transplantation should not be long delayed, though opinions differ about the optimum age for its performance. Varied ages, e.g. birth, 6 months,[11] and 4 years[12] have been offered as optimum times for operation, and the present series does not indicate any advantage in performing the operation at any particular age, provided the ectopic anus functions normally up to the time of surgery.

In almost all cases in this series, the transplantation was performed through a vertical incision joining the ectopic anus and the proposed new anal site. Though decried by authors[11,13] who favour a transverse incision, the longitudinal incision used here does not appear to have caused excessive scarring and retraction of the anus to its former site.

2. Two cases of imperforate anus in this series appear to have been associated with recto-urethral fistula. This association is exceptionally uncommon, although Stephens[6] has reported recto-urethral fistulae below the level of the verumontanum. However, Stephens,[6] in defining the entity of imperforate anus, stated that the anterior wall of the rectum is closely attached to the urethra. It is possible that this close attachment of urethra caused tenting of this structure when traction was put on the rectum in these cases, and this was mistaken for a fistula and divided. A recto-urethral fistula and eventual urethral stricture developed in 1 of the patients postoperatively, so that this explanation seems feasible in at least 1 case. However, it must be admitted that, since this is a retrospective analysis, there might be a discrepancy in the hospital notes and these 2 cases might have been examples of rectal agenesis with recto-urethral fistulae.

3. Most authors claim that operations performed on children with anal anomalies give generally good results.[2,3,5,14] This is not borne out by this series *(vide infra).*

X-ray Diagnosis

All the patients in this series admitted to hospital shortly after birth were X-rayed in the inverted position with a marker over the anus, according to the method of Wangensteen and Rice.[15] Twenty such X-rays were available for study.

It was found that all the patients with rectal agenesis showed the rectal gas bubble, as anticipated, above the pubo-coccygeal line.[5] In only 2 patients with anal anomalies did the gas bubble fail to appear below the pubo-coccygeal line, presumably because of inspissated meconium in the rectal stump or because air had not had sufficient time to reach the anus before the X-rays were taken. However, these 2 cases were readily diagnosable on clinical grounds alone.

It is suggested that this discrepancy does not detract from the value of X-rays in the diagnosis of anal anomalies, and that they are of great value when used in conjunction with the physical signs.

Associated Abnormalities

The associated congenital abnormalities found in this series are enumerated in Table IV and are those mentioned in the patients' hospital notes or found at postmortem examination. The incidence is probably much higher than stated, since many abnormalities may only be discovered in later life and many, e.g. in the urinary tract, require specific investigation for their diagnosis.

TABLE IV. ASSOCIATED ANOMALIES

Anomaly	*Rectal agenesis (18 cases)*	*Anal anomalies (23 cases)*
Cardiac	2	2
Urinary tract:		
Crossed ectopic kidney	1	1
Fused kidneys	1	—
Hypospadias	1	—
Tracheo-oesophageal fistula	1	1
Duodenal atresia	1	—
Micrognathia	1	—
Coloboma of the iris	—	1
Craniosynostosis	—	1
Sacral abnormality	3	2

It is generally reported that associated abnormalities are more commonly found with rectal agenesis than with anal anomalies,[2,3,5] and Forshall,[16] in her series of 53 patients with rectal agenesis, found only 29 who did not suffer from other serious abnormalities. In the present series no considerable difference was found in the incidence of concomitant anomalies between the 2 groups. The high incidence of urinary tract abnormalities, as reported by others,[2,3,5,16] is also noted.

Of the 27 cases where X-rays were suitable for examination, sacral abnormalities were found in 5 — 3 in association with rectal agenesis and 2 with anal anomalies. Among these 5 patients there were 2 with hemivertebrae and 3 in whom there were only 4 sacral vertebrae. The incidence of sacral abnormalities is reported by some authors[2,6,17] to be much higher than that found in this series. The abnormalities are of great significance, particularly with regard to future rectal function, and they should be carefully looked for when the child is first X-rayed. Though sacral abnormalities are reported to be more frequent in association with rectal agenesis than with anal anomalies,[17] this fact is not borne out in this series and does not appear to be a useful diagnostic point in distinguishing between the 2 conditions radiologically.

Mortality

The causes of death in this series are enumerated in Table V. This list includes children dying up to 9 months of age, and the mortality rate of 31·7% is higher than in most series

TABLE V. CAUSES OF DEATH *

Cardiac anomaly	4
Renal anomaly	1
Prematurity	1
Perforation of bowel (? site)	1
Haematemesis (acute pyloric ulcer)	1
Small-bowel obstruction + chest infection	1
Tracheo-oesophageal fistula	1
Associated duodenal atresia	1
Postoperative:	
? Shock	1
Aspiration of vomitus	1
Total	13

*Mortality rate = 31·7%.

TABLE VI. RECTAL FUNCTION

Condition	*Age at FU*	*Stricture*	*Incontinence*	*Colonic inertia*	*Normal*
Rectal agenesis	4 years	—	—	+++	—
	5½ years	+++	+++	—	—
	1 year	—	—	+++	—
	1 year	—	+++	—	—
	3 months	+++++	—	—	—
Ectopic anus:					
Perineal Male	5 years	—	—	+++++	—
	1½ years	—	—	+++++	—
Perineal Female	3 years	+	—	+++++	—
Vaginal and vestibular 'fistula'	3 years	—	—	+++	—
	4 years	—	+++	—	—
	9 months	—	—	—	+
	11 years	—	—	—	+
	4 months	+	—	+++++	—
Imperforate anus	8 years	+++++	—	—	—
	3 years	—	—	—	+
	8 years	—	—	+++	—
Imperforate anal membrane	4 months	—	—	+++	—
Stenosed anus	8 years	—	—	—	+

+=mild, +++=moderate, +++++=severe, FU=follow-up.

(varying from 18%[3] to 36%[6]), which usually include only deaths occurring during the first admission to hospital.

It will be noted that of the 13 patients who died, death was due to an associated congenital abnormality in 7 and to prematurity in 1. In only 2 was operation (an abdomino-perineal 'pull-through') the direct cause of death.

RECTAL FUNCTION

The rectal function achieved in 18 of the surviving 28 patients is presented in Table VI and includes those who developed rectal stricture. Some patients were not yet old enough for follow-up to be of value and these were excluded from the Table. Children operated on only a few months previously were included, however, if there were definite signs of complete incontinence, stricture, or gross constipation with faecal impaction necessitating manual evacuation. It is possible that some children did not present for follow-up examination because their bowel function was quite normal, and this might mean that the results are perhaps a little better than those depicted in the Table.

Constipation

It will be noted that the commonest problem is that of *constipation*. This may progress to faecal impaction, necessitating manual evacuation or (as in one patient) colostomy, or ultimately to megacolon. There are a number of causes of constipation in these cases:

1. *'Colonic inertia'*. This term was originally coined by Denis Browne[13] and the condition may result from anal stenosis, pain on defaecation from fissure-in-ano (often caused by over-zealous dilatations), or defective bowel training. Loss of rectal sensation is also an important factor, resulting in persistent postponement of the act of defaecation. The basic mechanism of the condition is a failure to empty the rectum adequately with resultant distension followed by atony of the rectal musculature, which in turn further inhibits rectal emptying. It has been shown by various authors[3,14,16,18] that the condition is more severe in rectal agenesis than in the various 'low' abnormalities. However, 4 of the most severe cases of colonic inertia in this series occurred postoperatively in children with ectopic anus with prior stenosis, who presented for the first time with severe faecal impaction. It would appear that anal stenosis must be avoided at all costs. Babies with ectopic anus who are not operated on at birth should have regular dilatations even though bowel action is apparently normal at the time, since colonic inertia develops insidiously and its early symptoms often go unnoticed by the child's mother.

Anal dilatations are the sheet-anchor of treatment after all operations performed for ano-rectal malformations, and it must be ensured that the mother is performing these adequately at home. These dilatations should be continued until the surgeon is completely satisfied that there is no further chance of stenosis occurring — a difficult decision, but one which can usually be taken about 3 - 6 months after operation. Partridge and Gough[3] suggested that a No. 14 Hegar dilator should pass with ease by the third week after operation.

Laxatives of some kind or other are invariably necessary in the management of colonic inertia. If possible, these should be mild, e.g. milk of magnesia, 'California syrup of figs', or 'colace', but occasionally these are ineffective and purgatives containing senna ('senokot') or phenolphthalein might have to be used. Liquid paraffin should be avoided since it causes the stool to pass too easily and predisposes to stenosis.

Suppositories, e.g. 'dulcolax' or glycerin, may be used and are found to be very effective by some authors. However, many children object to them *(vide infra)* and many mothers find their insertion 'a nuisance'. Carbachol has occasionally proved of value in severe cases.

Enemas are best avoided, though the very occasional patient will respond well to them. Most children find them extremely distasteful and they may be hazardous in the hands of unskilled mothers. If faecal impaction occurs, it is best treated by manual evacuation under general anaesthesia. This is far kinder than persistent bowel-washouts and insertion of suppositories, which usually succeed in instilling terror into the child and fear of future procedures.

Bowel training is essential in children with colonic inertia and requires great tact and persistence on the part of the mother. Over-zealous 'potting' with rigid discipline may be just as harmful as a *laissez-faire* attitude to this problem, and these mothers require guidance and encouragement from the surgeon.

2. *Stricture*. There are 3 cases of established ano-rectal stricture in this series and 2 of mild stricture which responded to dilatations. All these strictures resulted from severe scarring following sepsis at the mucocutaneous anastomosis. Minimal postoperative sepsis at this site is regarded by most authorities as inevitable and this serves to underline the importance of postoperative dilatations. Once a stricture is fully established, further treatment is most unrewarding and there is frequently little to do except provide the child with a permanent colostomy. Prevention is unquestionably better than cure.

3. *Hirschsprung's disease*. Hirschsprung's disease coexisting with ano-rectal malformation has been reported only twice in the literature.[15,19] It has been sought for as a cause of persistent constipation in these cases by other authors, but never found. In 2 patients in the present series, biopsies of colon were taken at the time of operation, but ganglion cells were found to be present. Scott,[20] in dissections of cases of rectal agenesis, found the nerve supply to the rectal stump to be intact in all those examined.

4. *Psychogenic*. Though this is never a primary cause of constipation in these patients, it may be a secondary one. Children with constipation caused by colonic inertia frequently revolt against the use of dilatations, suppositories, enemata, and over-enthusiastic 'potting'. They may develop severe negativism and refuse to cooperate, becoming a trial to both mother and doctor. Psychiatric advice may be necessary in some of them.[21]

Incontinence

This is the other major sequela which may supervene in patients operated on for ano-rectal malformations. It may be of two types:

(*a*) Caused by faecal impaction with overflow. This has been discussed above.

(*b*) True incontinence. This may be caused by congenital absence of pelvic structures, e.g. muscles and nerves, subserving rectal continence, or damage to these structures at operation. Congenital absence of the structures is nearly always associated with sacral agenesis.[17] There is little one can do to assist continence, and the

reconstructed ano-rectum becomes virtually a perineal colostomy.

Attention to detail while operating on patients with ano-rectal malformation will assist in preventing incontinence from damage to vital structures. Points of importance are:

1. During the 'pull-through' procedure, the rectum should be passed through the anterior part of the levatores ani in order to make full use of the sphincteric action of the pubo-rectalis sling.[6]

2. Extreme care must be taken to prevent damage to pelvic nerves. Denis Browne[13] reported using graduated Hegar dilators instead of forceps or scissors to fashion a tunnel through the pelvic floor.

3. The rectum must be handled very gently, and not stripped, in order to retain its nerve supply, which is nearly always intact.[20]

Perhaps the best summary of the problem of rectal function in children with rectal agenesis is provided by Isabella Forshall:[16]

'... The prognosis for faecal continence is, however, much better than is generally supposed, provided the mothers and children are managed with patience and understanding. Few, if any, of these children with agenesis of the terminal rectum are continent of faeces in the usual sense of the term. They will, however, if carefully managed, in time be able to lead normal lives. The age at which they may be expected to become clean is much later than for a normal child, usually somewhere between 5 and 7 ...

'The only hopeless cases are those with a stricture. The worst mistake in management is to allow the child to become constipated. Some children are clean all day if a colon washout is given well before the time of starting for school; others remain clean if they take a dose of Epsom salts before breakfast to empty the colon. It is sometimes worth arranging that the child goes to school in the morning only for the first year, until he gains confidence and to prevent him from being left all day in soiled pants if he should have an accident. The fortitude of some of these children is remarkable; they refuse foods they like but which they have learned give them loose stools which they cannot control. As Potts has said — "for these children diarrhoea is a nemesis". Both the mother and child need to know that their medical adviser is behind them, interested, hopeful, and ready with encouragement and advice. Under no circumstances should the child ever be spoken of as being dirty.'

It will be seen from Table VI that, of the 18 children followed-up, only 4 had normal or acceptable rectal function for their age. However, 7 of those with mild stricture, incontinence or colonic inertia are still under the age of 5, and further improvement in their condition can still reasonably be expected. Though the results are better in the group with 'low' abnormalities than in those with rectal agenesis, they may nevertheless be regarded as relatively unsatisfactory.

COMPLICATIONS

Other than the complications noted previously and those associated with any abdominal surgery, particularly in the neonatal period, there are two large groups for consideration:

1. *Urinary Tract Complications*

(*a*) *Persistent recto-urethral fistula* after operation. This occurred in 4 children in this series, all following a perineal approach. All 4 required multiple attempts at repair before closure was ultimately effected, confirming the experience of others.[3]

(*b*) *Urethral stricture.* This occurred in 1 child, following ligation and division of a recto-urethral fistula. In order to display a recto-urethral fistula, it is customary to put slight traction on the rectum. This causes tenting of the urethra analogous to the tenting of the common bile duct when traction is put on the cystic duct during cholecystectomy. For this reason it is advisable to close off the fistula a small distance away from the urethra, to avoid narrowing or mucosal damage. It is interesting to note that Gross[10] advised that it is not necessary to close the urethral side of the fistula at all, Potts[11] advised that it be oversewn, while Partridge and Gough[3] stated that it should be transfixed and ligated.

(*c*) *Urinary infection.* Severe urinary infection may be caused by delay in performing the 'pull-through' operation following colostomy in a patient with a recto-urethral fistula. This was demonstrated in 1 child in this series. Urinary tract infections are also commonly found together with ano-rectal abnormalities, owing to associated renal-tract anomalies.

(*d*) *Urinary incontinence.* This may occur in association with absence of sacral nerves, commonly found with sacral agenesis, or may follow damage to these nerves during reconstructive operations on the ano-rectum. No child with urinary incontinence was encountered in this series.

2. *Complications of Colostomy*

Colostomy was performed on 15 of the 41 patients in this series. One patient almost died from intractable diarrhoea following colostomy, and one suffered a herniation of bowel and stomach around the colostomy which necessitated repair and nearly caused the death of the infant. A few children suffered from minor prolapse of colonic mucosa. Nine children undergoing colostomy died, all from causes unrelated to the colostomy.

Of the 15 colostomies, 8 were transverse, 5 were sigmoid, 1 was placed at the splenic flexure, and there was 1 caecostomy. The caecostomy was performed for perforation of the colon, the site of which was undetermined. It must be emphasized that in cases of ano-rectal malformation any colostomy sited on the left side of the colon is likely to interfere with further procedures and should be studiously avoided.

SUMMARY

1. The various methods of treatment, X-ray diagnosis and complications in a series of 41 unselected patients with ano-rectal anomalies are reviewed.

2. There is a considerable disturbance of rectal function following correction of ano-rectal anomalies. This series illustrates that function in cases of rectal agenesis is uniformly poor, and that contrary to accepted teaching the results in 'low' abnormalities leave much to be desired, especially in the entity of ectopic anus.

3. The high mortality rate associated with ano-rectal abnormalities (31·7% in this series) is due mainly to the high incidence of coexisting congenital anomalies.

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