MALFORMATIONS OF THE ANUS AND RECTUM*

A REPORT ON 85 CONSECUTIVE CASES

J. H. Louw, Ch.M.

Professor of Surgery, University of Cape Town, and Head of the Department of Surgery, Groote Schuur Hospital and Red Cross War Memorial Children's Hospital, Cape Town

'Imperforate anus' is a congenital anomaly which is commonly encountered in practice. Indeed, it is the commonest of the congenital malformations that are incompatible with life.[1] The incidence of this anomaly is about 1 in 5,000 live births[2] and we have had the privilege of treating no less than 85 cases during the past 6 years.

It should be clear that a large number of surgeons encounter the lesion during their career and that operations on babies with this condition are often performed by men who are unfamiliar with the technique and results of surgery in these cases.[1] In this connection I should like to quote Willis Potts[3] who recently stated: 'People are biased (most people are) but 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 rectovesical fistula when he never before has seen such a case nor witnessed the operative repair? . . . Such cases . . . are apt to be poorly handled. If the defects are not properly corrected at the first operation, these patients may be forever incontinent or doomed to a permanent colostomy.'

The following are examples of some of the mistakes that are still being made in the diagnosis and management of imperforate anus:

1. The popular idea that 'imperforate anus' *in its common form* is an anomaly where only a thin membrane prevents the escape of meconium (Fig. 1) 'which bulges the membrane *through the anal canal* as a plum-coloured mass'.[4] Although Gross[2] found the anomaly in 2·8% of his cases of anorectal malformations, it is generally agreed that this kind of membrane is excessively rare,[5] and Denis Browne,[6] in his wide experience, has never encountered a case. We have made the diagnosis on one occasion in our series of 85 cases, but the 'membrane' was no longer intact and we assumed that it had been perforated by the person who had originally attended the infant. It should therefore be clear that in the treatment of 'imperforate anus' the advice that 'all that is necessary is to incise the septum crucially'[7] is seldom applicable.

The commonly accepted classification of malformations of the anus and rectum is that of Ladd and Gross[8] in which 4 main visceral types, with and without 'fistulae' are recognized (Fig. 2). This classification, however, is based on an erroneous interpretation of the embryology (*vide infra*), and fails to act as a guide to treatment and prognosis. We have adopted a modification of Denis Browne's[9] classification elaborated by Douglas Stephens[10,17] and based on the work of Wood Jones[11] and Sir Arthur Keith.[12] In this scheme 2 broad groups of abnormalities are recognized, viz. rectal and anal (Table I).

TABLE I. ANORECTAL MALFORMATIONS, 85 CASES

Rectal agenesis, 32 cases	
1. Recto-urethral fistula (M)	27
2. Recto-vesical fistula (M)	1
3. Recto-vaginal fistula (F)	2
4. Without fistula (F, M)	2
Anal anomalies, 47 cases	
1. Ectopic anus (26)	
(*a*) Perineal (M)	1
(*b*) Perineal (F) 'Shot-gun anus'	2
(*c*) Vestibular (F)	16
(*d*) Vaginal (F)	7
2. Covered anus (18)	
(*a*) Ano-perineal 'fistula' (M)	15
(*b*) Ano-bulbar 'fistula' (M)	1
(*c*) Ano-vulvar 'fistula' (F)	2
3. Imperforate anus (M)	2
4. Imperforate anal membrane (M)	1
Gross malformations, 6 cases	

M = Male F = Female

2. The common and most dangerous practice of attempting to bridge the gap between anus and colon by a perineal approach.[1] This is only too frequently done in unsuitable cases at the expense of muscles, nerve supply and blood supply, and the child is left a rectal cripple. A popular textbook of surgery[7] advises perineal dissection when 'the distance

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between the end of the rectum and anal dimple is not more than 2 inches (5 cm.)', but most enlightened authorities on the subject agree that such dissection is highly dangerous when the distance exceeds 1·5 cm.[1,2,13-16] Even the arbitrary figure of 1·5 cm. is not a satisfactory criterion,[10,17] and the wise surgeon will depend on anatomical landmarks rather than a ruler in coming to a decision.

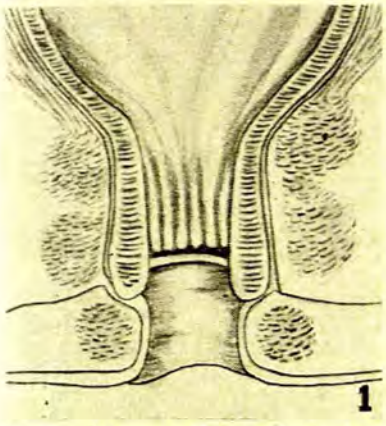

Fig. 1. Diagram of imperforate anal membrane often referred to as the 'common form' of 'imperforate anus'. In practice it is excessively rare.

3. The almost blind acceptance of the concept of 'fistulae' between the rectal pouch and urinary tract, vagina or skin.[2,15,16] Such 'fistulae' occur in more than 70% of cases[2] and while this idea of a 'fistula' prevails, treatment by 'transplantation of the fistula' will continue, and must remain unsatisfactory. This idea is based on an erroneous interpretation of the embryology which is still tenaciously accepted by many people despite Wood Jones'[11] observation more than 50 years ago that 'the generally accepted view of the development of the human hind end offers no explanation of some of the abnormalities which are commonly found clinically in this situation', and Sir Arthur Keith's[12] finding that 'In the series of malformations now described to you (114) there is not one single form that is satisfactorily accounted for thereby'. Most 'fistulae' should be treated by much simpler and less destructive procedures than 'transplantation'.

4. The mistake is made of advising inguinal colostomy in patients where a colostomy seems necessary.[7] In doing this, the redundant sigmoid loop, which should be left free for future reconstructive surgery, is sacrificed. A colostomy, if required, must be placed in the transverse colon to leave as much distal colon as possible for use at the definitive operation.[1,9]

ANALYSIS OF MATERIAL

Our cases of 'imperforate anus' have been divided into the 2 main groups, viz. rectal and anal (Table I). There were 32 cases of rectal agenesis and 47 with anal malformations. (A third group consisting of 6 cases and designated 'gross malformations' has been included for the sake of completeness.) This classification has enabled us to determine the necessary form of treatment and to estimate with reasonable accuracy the prognosis in each individual case. The 2 groups differ in many respects, viz. anatomically, embryologically, clinically, radiologically, as well as therapeutically and prognostically. These differences will be discussed briefly. (In the group with gross malformations the whole 'hind end' was deformed with multiple congenital anomalies and extroversion of the cloaca. These malformations are not amenable to surgery and will not be discussed.)

A. Anatomy

The most important difference between rectal and anal anomalies is to be found in their relationship to the levator ani muscle.[10,17] In rectal anomalies the bowel ends blindly above the levator whereas in anal anomalies it extends through the levator and is embraced by the puborectalis sling. It should be noted that in the newborn, the level of the levator is just below a line drawn from the symphysis pubis to the sacrococcygeal junction (P–C line, Fig. 3). In the normal infant this line marks the positions of verumontanum in males, the external os of the cervix in females, the peritoneal reflection, and the third fold of Houston.[10,17,18]

Rectal Agenesis, 32 *Cases*

Boys are much more frequently affected than girls (29 of our cases, Table I). In the common type in males the bowel ends on the pubococcygeal line and communicates *via* a minute 'fistula' into the prostatic urethra (Fig. 4a). The bowel rarely terminates in the bladder (1 of our cases). The anal canal is often absent and the external anal musculature in this region is usually entirely vestigial.[10,17,19] The levator ani itself may be poorly developed particularly in cases associated with absence of more than 2 sacral vertebrae.[10,14,17,20] In females the fistula opens high up into the posterior fornix of the vagina (Fig. 4b). A very rare variant in both sexes is agenesis without a 'fistula'.

Anal Anomalies, 47 *Cases*

Two common and 2 rare varieties can be recognized (Table I):

(*a*) *Ectopic anus*, 26 *cases*. Girls are much more commonly affected than boys (25 of our cases, Table I). The rectum passes through the levator which forms an adequate sling around it but, instead of opening on the surface at the proctodeum, it passes obliquely forward and in females opens through a somewhat stenosed orifice as follows:

(i) On the perineum—'shotgun anus' with near-normal external anal muscles (Fig. 5a, 2 cases). The normal strip of skin separating vaginal and anal orifices is absent.
(ii) Into the vestibule—'vestibular anus' with poorly developed external anal muscles (Fig. 5b, 15 cases).
(iii) Into the lower third of the vagina—'vaginal anus' with vestigal external anal muscles (Fig. 5c, 7 cases).

In males the anus is simply placed more anteriorly than normal (Fig. 5d, 1 case), thus close to the posterior border of the scrotum. The external anal musculature is practically normal.

The proctodeal pit is obliterated in all these cases and the external anal musculature deficient in most. At best, the deep portion of the external sphincter is well-developed.

(*b*) *Covered anus*, 18 *cases*. Boys are most commonly affected (Fig. 6a, 16 cases). In them the perineal raphe extends posteriorly over the anal site where it is usually thickened to form a ridge or tag. The rectum terminates just deep to this tag opposite the bulb of the urethra.[10,17] The anal orifice is covered by skin and is often projected forwards along the median raphe to the perineum, scrotum, and penis, or even as far as the frenulum. The deep external sphincter is well developed, but the superficial part is represented by a thin collection of fibres passing in a sagittal plane on either side of the 'anoperineal fistula'.[10,17] A rare variant in boys (1 of our cases) is the so-called 'anobulbar fistula'.[10,17] The perineum is absent, the lumen of the anus is projected forwards into the bulb of the urethra, and hypospadias usually coexists.

In girls (Fig. 6b, 2 cases) the anus is similarly covered by skin and projected anteriorly parallel to the perineal plane

LADD AND GROSS CLASSIFICATION2

to open at, or close to, the fourchette in the fossa navicularis—'anovulvar fistula'. The orifice is constricted and the patients usually have some male characteristics, e.g. enlarged clitoris.[21] In very rare, exaggerated examples of this condition the vestibule is also covered and the anus projected forwards, together with the vagina to the base of the clitoris.[6]

(*c*) *Imperforate anus* (*Fig.* 7, 2 *cases*). This condition occurs almost exclusively in males. The rectum terminates blindly at the *upper* border of the bulbocavernosus,[10,17] and the anal dimple is absent or rudimentary. The external sphincters are poorly developed or rudimentary and incorporated with the bulbocavernosus.[10,17]

(*d*) *Imperforate anal membrane* (*Fig.* 8, 1 *case*). A thin membrane stretches across the lumen of the anorectal canal at the site of the anal valves, i.e. at the level of the triangular ligament or lower level of the bulbocavernosus.[10,17] All the muscles are intact except the superficial external sphincter which is rudimentary or absent.[22]

B. Embryology

Broadly speaking, it may be said that the rectum is derived from the entodermal cloaca and the anal canal from the ectodermal claoca (Fig. 26). It must be understood, however, that there is coordination in the differentiation of these two primitive structures, and that errors in the development of one may be accompanied by errors in the development of the other. More details cannot be discussed here.

The development of the *rectum* is intimately associated with that of the urogenital sinus—bladder, urethra, and internal generative organs (Fig. 9). At first there is free communication between the two structures, but this soon closes off due to downgrowth of the urorectal septum and ingrowth of the lateral folds of Rathke—Fig. 9.[23-25] Then the rectum has to 'find' a new 'vent' by migrating to the perineum and establishing a communication with the proctodeum or anal canal.[5,12,26]

In 'rectal agenesis' the development of the rectum is usually arrested at an early stage before it has closed off from the urogenital sinus.[26] The primary error is probably failure of fusion between the fused Rathke's folds below and the urorectal septum above.[25] Since this provides the bowel with a vent, migration is arrested and the rectum terminates in this region, i.e. the prostatic urethra. Concommitant arrest of development of the anal canal is common.[10,17]

The development of the *anal canal* is associated with the development of the pars phallica of the urogenital sinus (penile urethra in males and lower third of vagina and vestibule in females) and with the formation of the genital folds and anal tubercles (Fig. 9).[9,21,22,27] The usual primary error is either imperfect development or premature fusion of the anal tubercles, or excessive posterior fusion of the genital folds with obliteration of the 'proctodeal pit'.[10,21,22]

(*a*) *Ectopic Anus*

The anal tubercles and inner genital folds do not fuse to form a perineum and the rectum then fails to migrate posteriorly.[5,22] The ectopic anus may open more anteriorly on the perineum, but more frequently there is 'compensatory' incomplete fusion of the lateral folds of Rathke and the establishment of a communication between the terminal rectum and pars phallica of the urogenital sinus (vestibule and lower third vagina in females). The condition is more common in females because posterior fusion of the genital folds is normally defective.[22]

(*b*) *Covered Anus*

There is excessive posterior fusion of the labioscrotal folds and also of the posterior portions of the superficial part of the anal tubercles.[9,21] The anterior ends of the anal tubercles, however, do not fuse;[22] this occurs at a time when the rectum

Fig. 2. Ladd and Gross[8] classification of anorectal anomalies: 1. Stenosis. 2. Persistent membrane. 3. Blind rectal pouch. 4. Blind rectal pouch with normal anus. 'Fistulae' may occur in association with any of these.
Fig. 3. Diagram of normal anatomy: (*a*) Male, and (*b*) female. Note the relationship of the levator ani muscle and pelvic viscera to the pubococcygeal line (P–C).
Fig. 4. Diagram of rectal agenesis. The rectum terminates on the pubococcygeal line proximal to the levator ani. (*a*) Male with recto-urethral 'fistula', and (*b*) female with rectovaginal 'fistula'.
Fig. 5. Diagram of ectopic anus: (*a*) 'Shot-gun perineum' in female, (*b*) vestibular anus in female, (*c*) vaginal anus in female, and (*d*) perineal anus in male.
Fig. 6. Diagram of covered anus: (*a*) Male, 'ano-perineal fistula', and (*b*) female, 'anovulvar fistula' or vulvar anus.
Fig. 7. Diagram of imperforate anus.
Fig. 8. Diagram of imperforate anal membrane.
Fig. 9. 'Ectopic vaginal anus' with chronic constipation and overflow incontinence resulting in excoriation of vulva and perineum.
Fig. 10. Rectal agenesis. 'Blank' perineum with raphe crossing the normal site of the anus. Note the small buttocks and absence of natal cleft.
Fig. 11. Rectal agenesis. Urine containing meconium has left a distinct stain on the diaper.
Fig. 12. 'Shot-gun perineum'. Anal and vaginal orifices open side by side without an intervening skin bridge.
Fig. 13. 'Vestibular anus'. A stenosed anal orifice is situated in the vestibule. A probe introduced into this orifice passed in a cranial direction.
Fig. 14. 'Covered anus'. The anus is covered by a triangular skin tag and immediately in front of this meconium can be seen shining through the perineal skin. Examination of this area through a lens revealed a minute 'microscopic anus' with escape of gas and a 'fly-speck' of meconium.
Fig. 15. Covered anus. The anus is covered by skin only and a track of meconium passes forwards along the midline raphe, (*a*) just deep to the skin to burst through on the scrotum, or (*b*) on the ventral surface of the penis.
Fig. 16. Vulvar anus. The anus is covered by skin and meconium tracks forwards just deep to the skin to open in the fossa navicularis at the fourchette. A probe introduced into the opening passed dorsally, parallel to the perineal skin.
Fig. 17. Imperforate anus. The anal dimple is absent and filled in by a ridge of tissue. No meconium can be seen shining through.
Fig. 18. Diagram illustrating the principle of the 'cut-back' operation in vulvar, vestibular and low vaginal anus.
Fig. 19. Photograph of method of 'uncovering' a covered anus (Fig. 15).
Fig. 20. Anal stricture in a case of rectal agenesis treated by abdomino-anal reconstructive surgery.
Fig. 21. Excoriation of buttocks due to incontinence following abdomino-anal reconstruction of rectal agenesis.
Fig. 22. Result of 'cut-back' procedure for vestibular anus. The anomaly has been converted into the 'shot-gun' variety. (*a*) After 3 weeks, and (*b*) after 3 months.
Fig. 23. Result of 'uncovering' a covered anus; (*a*) After 1 week, and (*b*) after 3 months.
Fig. 24. Result of perineal anoplasty for imperforate anus. The anus has healed by granulation and tends to gape as indicated or become stenosed.
Fig. 25. Result of 'cut-back' procedure in vulvar anus. The posterior extension of the labial folds can still be seen. Continence is satisfactory and normal childbirth possible.

has already migrated, hence meconium collects under tension just deep to the perineal raphe and in due course finds its way forwards in the loose connective tissue just deep to the raphe, bursting through to the surface at any point between the proctodeum and tip of the penis. (In the anovulvar variety there is also failure of fusion of the inner genital folds.)

(*c*) *Imperforate Anus*

There is imperfect development and premature fusion in the midline of the anal tubercles, but the migrating rectum finds no external 'vent'.[22]

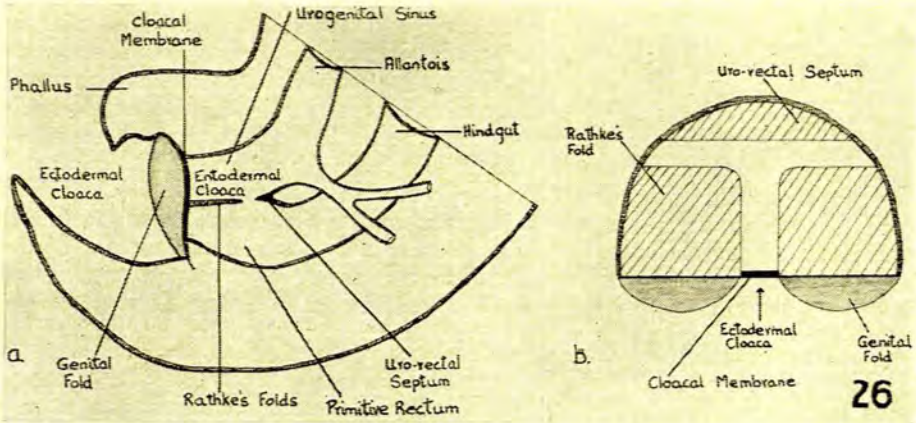

Fig. 26. Diagrammatic representation of human cloaca at 12-14 mm. stage. (*a*) Sagittal section, and (*b*) coronal section. Note the downgrowth of the urorectal septum and ingrowth of Rathke's folds which divide the cloaca into urogenital sinus ventrally and rectum dorsally. At the same time the genital folds and anal tubercles develop into the external genitalia and anal canal.

(*d*) *Imperforate Anal Membrane*

This is due merely to failure of breakdown of the proctodeal membrane or anal plate.[10,17]

C. Clinical Features

1. *Intestinal Obstruction*

The most striking and important clinical difference between rectal and anal anomalies concerns the development of intestinal obstruction.

In most cases of *rectal agenesis* symptoms and signs of acute intestinal obstruction become manifest within 48 hours. This happens in all cases without a fistula and also in boys with the common anomaly, viz. recto-urethral 'fistula', because the 'vent' is only minute. In the rare cases of recto-vesical fistula and rectovaginal fistula the opening may be large enough to deflate the bowel.

In most *anal anomalies* acute intestinal obstruction does *not* develop. In all varieties of ectopic anus the orifice is large enough to act as an efficient 'vent' during infancy. Later in life, however, when the stools become more solid, these children are prone to chronic constipation with faecal impaction, secondary megacolon, and overflow incontinence (Fig. 9). In patients with covered anus the thin skin which occludes the anal orifice usually ruptures spontaneously, and the same probably applies to that rare anomaly, imperforate anal membrane. Only in cases with imperforate anus and imperforate anal membrane does the patient develop acute intestinal obstruction, and differentiation from rectal agenesis must depend upon other features.

2. *Appearance of Perineum*

Rectal agenesis. In most of the cases the perineum is 'blank', often with a longitudinal median ridge running from the site of the normal anal position to join the median raphe of the scrotum (Fig. 10). Stimulation of this tissue may cause puckering due to contraction of the underlying vestigial muscle. The perineum may bulge when the baby cries, but meconium cannot be seen shining through the skin. There may be a dimple instead of a ridge and in rare cases a normal anal canal (Ladd and Gross, type 4) may be present. In many cases the buttocks are unusually small and the natal cleft obliterated (Fig. 10).[14]

In boys suffering from the common variant (recto-urethral fistula) there may be evidence of a communication with the urinary tract, e.g. 'the sign of green urine',[14] emergence of gas or a minute speck of meconium from the urethral meatus, flecks of meconium on the diaper (Fig. 11), or meconium in a centrifuged specimen of the urine. In females meconium may discharge from the vulva and, on passing a speculum, the fistula may be seen opening into the posterior fornix of the vagina.

Anal anomalies. The appearances depend on the type of anomaly:

(*a*) Ectopic anus. The anal orifice, which is somewhat stenosed but otherwise normal in appearance, is situated anterior to the normal position, e.g.:

(i) In the perineum both in males and females. In the latter case Denis Browne refers to the anomaly as 'shot-gun perineum' (Fig. 12).
(ii) In the vestibule (Fig. 13). A probe introduced into the orifice passes obliquely upwards, but can be felt through the perineum.
(iii) In the lower third of the vagina. A probe inserted into the orifice passes cranially, and only with forceful depression can it be felt through the perineum.

(*b*) Covered anus. In boys there is an epithelial fold suggestive of a hypertrophied perineal raphe extending over the usual anal site where it is often thickened to form a triangular posterior 'tag' (Fig. 14). Close scrutiny of what appears to be an intact perineum will reveal a bluish tinge shining through the skin and a small spot of meconium, no larger than a fly-speck, emerging from a microscopic opening adjacent to the 'tag'—the microscopic anus of Denis Browne (Fig. 14). In other cases a bluish track filled with meconium can be seen extending forwards (Figs. 15a and b). It usually bears little epithelial pearls and opens at the root of the scrotum or on the ventral surface of the penis. (In the anobulbar variety the anomaly is associated with a cleft scrotum and atypical hypospadias.)

In the vulvar anus of girls the anal dimple is also absent and filled by a central sagittal raphe. A track runs forwards and opens into the vulva close to the fourchette in the fossa navicularis (Fig. 16). A probe introduced into this opening passes directly backwards under the perineal skin. The

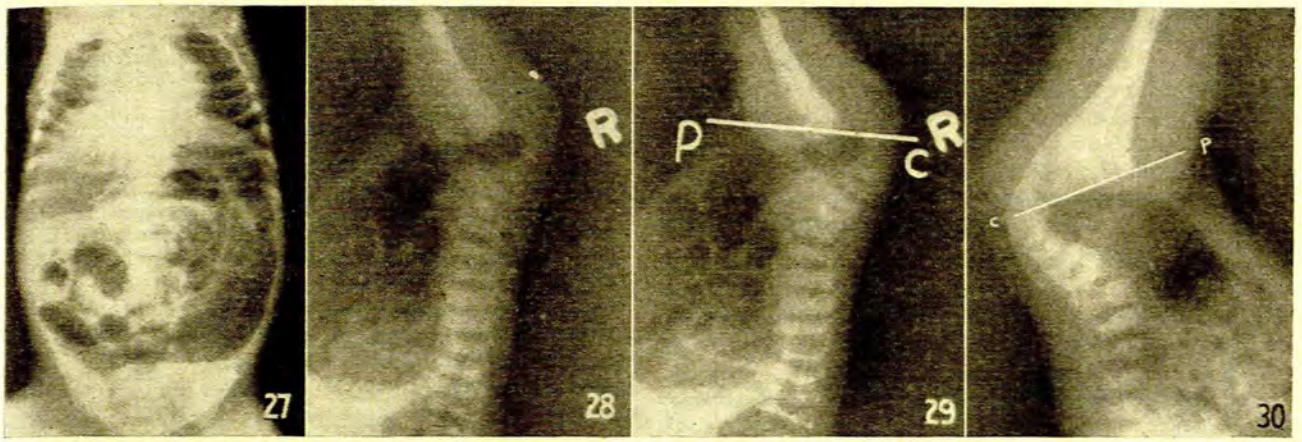

Fig. 27. Rectal agenesis. Flat A.P. plate (child erect) showing distended small and large bowel loops and fluid levels due to intestinal obstruction. (A similar picture is found in cases with imperforate anus.)

Fig. 28. Rectal agenesis. Flat, lateral plate (child inverted with marker at normal site of anus). The distance between the terminal gas shadow and anus is greater than 1·5 cm.

Fig. 29. Rectal agenesis. Flat, lateral plate (child inverted and pubococcygeal line inserted). The terminal gas shadow ends at the P–C line.

Fig. 30. Imperforate anus. Flat, lateral plate (child inverted and pubococcygeal line inserted). The terminal gas shadow extends caudal to the P–C line.

clitoris is often enlarged and resembles severe hypospadias so that in extreme cases of so-called 'covered vagina' the external genitalia resemble those of a boy.[6]

(*c*) Imperforate anus (Fig. 17). The appearance of the perineum is similar to that in rectal agenesis with a median perineal ridge, but there is *no* evidence of a recto-urinary fistula.

(*d*) Imperforate anal membrane. It is said that a normal anal canal is present and that the membrane bulges through it as a plum-coloured mass which becomes more obvious when the child cries.[4] When the membrane has ruptured the area may be stenotic with ragged epithelial tags at the level of the anal valves.

3. *Associated Anomalies*

Here, again, there is a striking difference between rectal and anal malformations (Table II).

TABLE II. SERIOUS ANOMALIES ASSOCIATED WITH ANORECTAL MALFORMATIONS

Type of anorectal malformations	*Anomalies of sacrum and coccyx*	*Anomalies of genito-urinary system*	*Other malformations*
Anal (47) ..	3	5	7
Rectal (32) ..	16	21	23
Gross (6) ..	6	6	6
Total (85) ..	25	32	36

In rectal agenesis there are frequently other abnormalities. Among these, vertebral anomalies are the commonest (50% of our cases), particularly the absence of sacral vertebrae.[10,14,17,20] These have an important bearing on the development of the levator muscles which are incompletely developed when more than two segments of the sacrum are missing. Urinary anomalies (70% of our cases) are very common[10,14,17] and malformations of the heart, oesophagus, intestinal tract, and nervous system may also occur.[2,28] In females associated malformations of the uterus and vagina are common.[10,17]

Anal anomalies are seldom associated with other serious anomalies (Table II), although vertebral malformations[20] and deformities of the external genitalia do occur.

D. Radiological Features

In patients who develop acute intestinal obstruction (mostly rectal agenesis; rarely imperforate anus) a straight X-ray of the abdomen will reveal distended small and large bowel with fluid levels (Fig. 27). This has to be differentiated from neonatal Hirschsprung's disease, the meconium-plug syndrome, and peritonitis with ileus. In cases with an adequate 'vent' the plates will be normal.

The position of the blind rectal pouch may be determined by holding the infant upside down and taking lateral films of the abdomen and pelvis. Gas in the bowel rises to the top, displacing meconium from the terminal end. By the Wangensteen and Rice[29] method a marker is placed on the proctodeal site and the distance between this and the terminal gas shadow measured (Fig. 28). In rectal agenesis the distance is greater than 1·5 cm. and in anal anomalies less than 1·5 cm. We prefer Stephen's[10,17] method whereby the relation of the gas shadow to the pubococcygeal line (upper border of symphysis pubis to last piece of sacrum) is determined. In rectal malformations the bowel terminates on or above this line (Fig. 29), while in anal anomalies it stops well below the line (Fig. 30) at, or caudal to, the lowest ossified segment of the ischium.[10,17]

E. Treatment

As a general principle early operative treatment is needed in every case.[14] This is particularly important in cases with rectal agenesis or imperforate anus with acute obstruction, but in all cases delay carries with it the risk of excessive stretching of the rectum and later 'rectal inertia' with chronic constipation and overflow incontinence.

Rectal Agenesis

The treatment is difficult because a high urinary fistula usually has to be dealt with and the bowel brought through the pelvic floor in front of the puborectalis sling before an orifice can be fashioned. A simple perineal approach is inadequate and destructive and is now universally condemned.[10,14,17,22] A combined abdominoperineal pull-through operation such as described by Rhoads *et al.*[30] must

be performed; this is best done soon after birth. Infants of 3 days and less tolerate this extensive operation very well.[1] After this age, however, the operation becomes extremely difficult because of distension, and then a transverse colostomy should be performed. Colostomy is also necessary in premature infants weighing less than 4½ lb. and in infants with other serious malformations. In such cases the definitive operation should not be unduly postponed because of the risk of urinary infection. We usually wait until the child weighs 10-12 lb. (age 3-4 months).

Anal Anomalies

These anomalies can all be rectified by a relatively simple perineal procedure.[6,9,10,14,17]

1. *Ectopic anus.* In the perineal variety operation is not justified, but regular dilatations are required. Vestibular anus is treated by the Denis Browne 'cut-back' technique.[9] One blade of a pair of scissors is inserted into the anal opening and guided posteriorly under the skin by external palpation until it approaches the natural anal site. Then the scissors are closed, thus performing a midline episiotomy (Figs. 18a, b, c, d). The wound edges are not sutured and post-operative dilatations are carried out daily for 3 months and then over longer intervals for the next 3-6 months. Vaginal ectopic anus may be similarly treated, but the orifice tends to remain tilted into the posterior wall of the vagina and secondary transplantation of the anus to the normal site is almost always necessary. (The best time for this procedure is probably the 4th year.[14] Alternatively, the orifice may be dilated regularly until the infant is about 6 months old, when primary transplantation of the misplaced anus may be performed.[31])

2. *Covered anus.* This condition is treated by a somewhat similar 'cut-back' technique. The 'track' is laid open from the front backwards, up to the point where it dips down to reach the anal site (Fig. 19). Epithelial excrescences are trimmed off to avoid tags. Regular post-operative dilatations are also necessary. (In girls with 'anovulvar fistula' the cut-back procedure is particularly satisfactory.)

3. *Imperforate anus.* This is the only anomaly that is treated by a formal perineal exploration through a midline incision extending from the scrotum or vagina in front to the coccyx behind. Preliminary 'needling' of the perineum to determine the exact depth of the blind end may be useful. The plane of dissection must be immediately posterior to the bulbocavernosus and during mobilization the levator 'sling' must *not* be divided but retracted. Prolonged post-operative dilatations are necessary.

4. *Imperforate anal membrane.* This anomaly is easily treated by cruciate incision of the membrane followed by dilatations.

F. Prognosis

There is a great difference in the prognosis of rectal and anal malformations. In *rectal agenesis* the prognosis is poor for the following reasons:

1. The infant soon develops acute intestinal obstruction, and delay in diagnosis is often responsible for pre-operative or operative fatalities.

2. Associated anomalies which may be incompatible with normal life are common.

3. A major abdominoperineal operation, which in itself is a serious risk, offers the only hope of a reasonable result.

4. The results of surgical treatment are poor because:

(*a*) Post-operative strictures are common (Fig. 20).

(*b*) Continence is defective on account of lack of normal rectal sensation, absence of external anal muscles, and often poor development of the levator[19] (Fig. 21). However, these children can usually be trained to social cleanliness when old enough to cooperate, and this is preferred to a permanent colostomy. Despite most extravagant claims by many American surgeons, we are convinced that normal continence is impossible except in those rare cases that have a complete proctodeum. However, it is our impression that the earlier the definitive operation is done, the more satisfactory are the functional results.

In *anal anomalies* the prognosis is excellent provided ill-advised attempts at 'perineal mobilization' and 'transplantation of fistulae' have been avoided. Acute obstruction is rare. Other serious malformations are uncommon. The defects can usually be corrected by a simple procedure and continence is very satisfactory in most cases. Stricture formation with the development of chronic constipation and rectal inertia is, however, an important sequela which must be prevented by regular post-operative dilatations. Providing the enlargement of the anus has been adequate and is kept adequate, (Figs. 22-24) bowel function is virtually normal, and in females normal childbirth can take place (Fig. 25). This is indeed fortunate, because in our experience these 'lesser anomalies' occur in more than half of the cases of 'imperforate anus'.

CONCLUSION

In conclusion, I can do no better than quote from a recent article by John Scott,[19] who has done a great deal of work on this subject both at the Hospital for Sick Children, Great Ormond Street, London, with Harold Nixon, and at the Boston Floating Hospital for Children, with Orvar Swenson:

'The surgical treatment of the imperforate anus is difficult and fraught with many disappointments. Only those surgeons who have special training and experience in operating on these small patients and who are prepared to supervise the long period of bowel training which is usually required should undertake their management. Parents must be encouraged to persevere with the unpleasant tasks that are often required of them, and the surgeon must place himself at their disposal at all times to give advice and hope. Only special pediatric surgical centres can provide the expert and efficient nursing care which is so valuable in these cases, and with modern communications babies can be transported rapidly and safely. There is no greater tragedy than the child condemned to a lifetime of misery and embarrassment by errors in the initial surgical treatment and lack of interest in the after care'.

ADDENDUM

Since the completion of this article another 14 patients suffering from malformations of the anus and rectum have been treated. The distribution of the various types of lesion was in keeping with our previous experience, but, strangely enough, there were 2 cases of imperforate anal membrane. Both these babies presented with acute intestinal obstruction. In the first a colostomy was performed elsewhere, and in the second the presence of a membrane situated approximately 1 cm. from the anal verge and *not* bulging through the anus, was diagnosed on clinical and radiological examination, and therefore treated by a cruciate incision and subsequent dilatations.

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