is invariably filled with air and is more peripherally situated. Caffey\(^3\) has produced evidence against these cysts being of congenital origin, and he believes that most, if not all, of the so-called 'congenital cysts' may be explained on the basis of check-valve bronchial obstruction. The bronchial elements in the cyst wall can be reasonably explained by the purely mechanical factors of bronchial dilatation beyond bronchial obstruction.

Perhaps the most reasonable view is that of Campbell,\(^4\) who holds that developmental bronchopulmonary malformations, through a variety of check-valve malformations, may give rise to lung cysts. Embryonic blockage of the lumina of the bronchial buds occurs, and single or multiple cystic cavities, lined with bronchial structures and filled with secretions, results. This hypothesis would also explain the finding of cysts in sequestration of the lung.

MALFORMATIONS OF THE ANUS AND RECTUM\(^*\)
A REPORT ON 85 CONSECUTIVE CASES
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'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.\(^3\) In this connection I should like to quote Willis Potts\(^4\) 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, 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\(^10,11\) and based on the work of Wood Jones\(^11\)

TABLE I. ANORECTAL MALFORMATIONS, 85 CASES

<table>
<thead>
<tr>
<th>Rectal agenesis, 32 cases</th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Recto-urethral fistula (M)</td>
<td></td>
<td>27</td>
</tr>
<tr>
<td>2. Recto-vesical fistula (M)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. Recto-vaginal fistula (F)</td>
<td></td>
<td>2</td>
</tr>
<tr>
<td>4. Without fistula (F, M)</td>
<td></td>
<td>2</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Anal anomalies, 47 cases</th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Ectopic anus (26)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>(a) Perineal (M)</td>
<td></td>
<td>1</td>
</tr>
<tr>
<td>(b) Perineal (F) 'Shot-gun anus'</td>
<td></td>
<td></td>
</tr>
<tr>
<td>(c) Vestibular (F)</td>
<td></td>
<td>16</td>
</tr>
<tr>
<td>(d) Vaginal (F)</td>
<td></td>
<td>7</td>
</tr>
<tr>
<td>2. Covered anus (18)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>(a) Ano-perineal 'fistula' (M)</td>
<td></td>
<td>15</td>
</tr>
<tr>
<td>(b) Ano-bulbar 'fistula' (M)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>(c) Ano-velar 'fistula' (F)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. Imperforate anus (M)</td>
<td></td>
<td>2</td>
</tr>
<tr>
<td>4. Imperforate anal membrane (M)</td>
<td></td>
<td>1</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Gross malformations, 6 cases</th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>M = Male F = Female</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

and Sir Arthur Keith.\(^12\) In this scheme 2 broad groups of abnormalities are recognized, viz. rectal and anal (Table I).

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
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. Even the arbitrary figure of 1.5 cm. is not a satisfactory criterion, and the wise surgeon will depend on anatomical landmarks rather than a ruler in coming to a decision.

3. The almost blind acceptance of the concept of 'fistula' between the rectum and urinary tract, vagina or skin. Such 'fistula' occur in more than 70% of cases 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' 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 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. 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.

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 extversion 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. 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 anterior occipital protuberance (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.

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. The levator ani itself may be poorly developed particularly in cases associated with absence of more than 2 sacral vertebrae. 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—'shOlgun 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 vestigial 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. 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'. A rare variant in boys (1 of our cases) is the so-called 'anoubular fistula'. 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
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. In
very rare, exaggerated examples of this condition the

venous return is also covered and the anus projected forwards,
together with the vagina to the base of the clitoris. 

(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, 
and the anal dimple is also absent or rudimentary. The external
sphincters are poorly developed or rudimentary and in-

cluded with the bulbocavernosus. 

(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. All the

muscles are intact except the superficial external sphincter
which is rudimentary or absent. 

B. Embryology

Broadly speaking, it may be said that the rectum is derived
from the entodermal cloaca and the anal canal from the
ectodermal cloaca (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 communica-
tion 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. Then the rectum has to
find a new 'vent' by migrating to the perineum and

establishing a communication with the proctodeum or anal
canal. The primary error is probably failure of fusion between the fused Rathke's folds below and the

urorectal septum above. Since this provides the bowel with a vent, migration is arrested and the rectum terminates
in this region, i.e. the prostatic urethra. Concomitant arrest
of development of the anal canal is common. The
development of the anal canal is associated with the

differentiation of the parablast alica of the urogenital sinus

(pee nile 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). 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'.

(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. 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 parablast alica 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. 

(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. The anterior ends of the anal tubercles,
however, do not fuse; this occurs at a time when the rectum

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. 'Cut-back' procedure for vestibular anus. The
anus has healed by granulation and tends to gape as indicated or

become stenosed.

Fig. 16. Vulvar anus. The anus is covered by skin only and a

track of meconium passes forwards along the midline raphe,

just deep to the skin to burst through on the scrotum, or

on the ventral surface of the penis.

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 reconstruction 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. Con-
inence is satisfactory and normal childbirth possible.

Fig. 2. Ladd and Gross' classification of anorectal anomalies:
Blind rectal pouch with normal anus. 'Fistulae' may occur in
association with any of these.

Fig. 10. Rectal agenesis. 'Blank' perineum with raphe crossing
the normal site of the anus. Note the small buttocks and ab-

sence 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

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.
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'.

(d) Imperforate Anal Membrane

This is due merely to failure of breakdown of the proctodeal membrane or anal plate.

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 rectovesical 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 on 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).

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', 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 'shotgun 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 anovulbar 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
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.4

(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

<table>
<thead>
<tr>
<th>Type of Anorectal Malformations</th>
<th>Anomalies of Sacrum and Coccyx</th>
<th>Anomalies of Genito-Urinary System</th>
<th>Other Malformations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anal (47)</td>
<td>3</td>
<td>5</td>
<td>7</td>
</tr>
<tr>
<td>Rectal (32)</td>
<td>16</td>
<td>21</td>
<td>6</td>
</tr>
<tr>
<td>Gross (6)</td>
<td>6</td>
<td>6</td>
<td>6</td>
</tr>
<tr>
<td>Total (85)</td>
<td>25</td>
<td>32</td>
<td>36</td>
</tr>
</tbody>
</table>

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 common10,14,17 and malformations of the heart, oesophagus, intestinal tract, and nervous system may also occur.6,28 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 malformations20 and deformities of the external genitalia do occur.

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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.

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 Rice29 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's10,17 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.20 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 pre-
mature 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.11 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.)

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 which is
affected 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
bulbocavernous 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 anomalies. 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 levator9 (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 im-
possible except in those rare cases that have a complete procto-
den. However, it is our impression that the earlier the definiti-
operation is done, the more satisfactory are the functional
results.

In anal anomalies the prognosis is excellent provided ill-
vised attempts at ‘perineal mobilization’ and ‘transplanta-
tion 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 enlarge-
ment 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 ‘imper-
forate 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’.
IMMUNIZATION OF THE RURAL BANTU AGAINST DIPHTHERIA IN THE NORTHERN TRANSVAAL

I. IMMUNITY PRODUCED AFTER ONE, AND AFTER TWO, INJECTIONS OF ALUM-PRECIPITATED TOXOID

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In the rural Bantu population of the Northern Transvaal diphtheria has occurred sporadically during the last 10 years, with no major outbreak and with no obvious seasonal

| TABLE I. RURAL DIPHTHERIA IN NORTHERN TRANSVAAL: AVERAGE ANNUAL INCIDENCE PER 100,000, 1951-57 |
|-----------------------------------------------|---------------|----------------|---------------|
| District | Rural European | Rural Bantu |                |
|------------------------------------------------|---------------|---------------|
| Burbernton | 68·8 | 7·5 |                |
| Groblersdal | 13·8 | 20·2 |                |
| Letaba | 58·9 | 4·0 |                |
| Lydenburg | 27·8 | 9·0 |                |
| Nelspruit | 34·8 | 9·5 |                |
| Pietersburg | 21·0 | 6·4 |                |
| Pilgrims Rest | 10·3 | 2·3 |                |
| Potgietersrust | 17·4 | 6·0 |                |
| Sibasa | 6·1 |                |                |
| Soutpansberg | 24·5 | 5·8 |                |
| Warmbaths | 12·5 | 3·9 |                |
| Waterberg | 15·8 | 14·1 |                |

* No Europeans

In Table I the incidence per 100,000 of the population (all rural Europeans and all rural Natives) is recorded. The difference in incidence between the races is striking.

The incidence in the Pietersburg district, where the experiment about to be recorded took place, is given in Table II. Although these incidences are unsatisfactory from a public-health standpoint, it is clear that no major outbreak took place. In 1950, when 38 cases, the highest incidence, were seen, the monthly distribution from January to December was as follows: 0, 10, 2, 9, 7, 5, 2, 0, 1, 0, 2, 0.

In Table II the age distribution of rural Bantu diphtheria is given, from 1951 to 1957 for all the districts of the area and from 1948 to 1957 for the Pietersburg district. The age distribution is similar in both areas, indicating that the same kind of conditions prevail generally and that the experimental results obtained in the Pietersburg district would be valid for the 11 other districts. The relatively low incidence in the 0–1 year group could have been predicted in view of the work of Mason et al.1 who showed that young Bantu babies, both rural and urban, have a high level of circulating antitoxin, maternally transmitted. When this decreases and eventually disappears after the age of about 6 months the incidence of diphtheria increases, to decline only after the age of 9 years, when silent and overt infections, spread over the years, have stimulated the production of a satisfactory active immunity.

In a large area like the Northern Transvaal, where the rural Bantu population is scattered, communications poor, immunization centres rather widely separated and the available trained medical personnel (medical officers, nurses and aides) already occupied with many other aspects of preventive

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* Reprints obtainable from J. H. Mason, South African Institute for Medical Research, Johannesburg.