VESICO-INTESTINAL FISSURE*

BY

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The great advances in paediatric surgery during the last two decades have brought about a change of attitude towards infants and children suffering from severe congenital malformations. A large number of these anomalies can now be satisfactorily treated and following the operation the child will be normal or nearly normal.

There are, however, still malformations or malformation complexes which are thought to be either untreatable or unjustifiable to treat as the child will remain more or less handicapped even after the most successful operation. These are mainly malformations of the central nervous or genito-urinary systems. It is my belief that in the next decade paediatric surgery will have to come to grips with these problems. We shall have to re-examine the contention that these children will never make useful members of society and are better left to die. We shall have to realize that with modern methods in physical education and rehabilitation much more can be done. This has been shown, for instance, in the management of spastic children. We shall also have to admit that with antibiotics and modern nursing care many of these children do not die or only die after a long time, a prolonged agony for them and their families alike.

I am prompted to this introduction because the condition which I am about to discuss is either not mentioned at all in the textbooks of paediatric surgery or urology or, if briefly described, it is dismissed as untreatable. There have been a few references in the literature to attempts at operation but as far as I can ascertain they have all ended in failure (Swan and Christensen, 1953). This prompts me to describe our experience in the treatment of this condition.

Definition and Description

The name vesico-intestinal fissure was introduced by Schwalbe in 1909, but the malformation has been

given a number of different names, such as ectopia vesicae with Meckel's diverticulum opening on the exposed bladder surface, extrophy of the bladder and colon, ectopic cloaca, etc. (Keith, 1908; Campbell, 1951; Williams, 1957).

In this condition there is an ectopia vesicae, which is often split into two lateral fields by an intervening zone of ectopic intestinal mucosa. If two kidneys are present, each ureter opens into the corresponding bladder field, but frequently one kidney is absent and malformation and displacement of the kidney is common. The intestinal mucosa belongs to the ectopic caecum. The opening of the appendix is often visible and the lower ileum tends to prolapse through the ileo-caecal valve forming a striking, bright red, sausage-shaped tumour. The colon is rudimentary and usually ends in front of the sacrum; the anus and rectum are always absent (Fig. 1). Duplication of some part of the lower intestine, such as the appendix or colon, is not infrequent. There is always an exomphalos situated above the ectopic intestinal field and a small myelo-meningocele or meningocele is frequently present; in one of our cases it only developed two months after birth.

The penis or clitoris is usually, although not invariably, split and the two halves widely separated. In one of our cases we were able to demonstrate, with the aid of serial sections of the autopsy specimen, that there were two complete penes each with a completely formed urethra, corpora cavernosa and spongiosa and prostate gland (Fig. 2).

Incidence

We saw our first case of vesico-intestinal fissure in 1953 and then considered it to be a great rarity. There are, however, a very large number of reports of this malformation in the literature since Meckel described the first case in 1812. In the last six years we have seen four cases as compared to 22 cases of ectopia vesicae and it appears quite possible that the condition is more common than is usually realized as many of the apparently hopeless children may not reach surgical centres.

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Embyrology

Vesico-intestinal fissure has been of great interest to embryologists because the malformation throws some light on the embryology of the cloaca, the lower abdominal wall and the causation of ectopia vesicae. In this context it is impossible to give a detailed description of the various opposing views regarding the embryological aetiology of the condition which we discussed in a previous article (Hall, McCandless and Rickham, 1953). It suffices to state that the hypothesis recently brought forward by Patten and Barry (1952), and especially their resurrection of Keibel’s (1896) view that the genital tubercles are primarily paired structures, has done much to explain most important features of this intriguing malformation (Fig. 3).

In the very early embryo the paired genital tubercles develop near the cephalic border of the cloacal membrane towards the end of the second week of intra-uterine life. They migrate towards each other and by the fifth week they fuse in the midline, reinforcing the cephalic border of the cloacal membrane (Fig. 3 (A1, A2, A3)). It is thought that in ectopia vesicae the genital tubercles develop somewhat caudally to their normal position, approximately opposite the point where the urorectal fold comes to the surface. If they now fuse in the midline the genital orifice will be cephalic and the anal orifice caudal to them (Fig. 3 (B1, B2, B3)). We developed this theory a stage further and assumed that if the paired genital tubercles develop still more caudally than with ectopia vesicae, they can now only fuse in the midline caudal to the cloacal membrane. There would then be a deficiency of secondary mesoderm over the whole of the hypogastic area and the extensive cloacal membrane would break down. If the membrane ruptured immediately after the formation of the tailfold, it would expose the dorsal and lateral walls of the hindgut. As Johnston (1913) has pointed out, the whole of the gut distal to Meckel’s diverticulum is derived from the hindgut. It will be seen that the above hypothesis explains the intestinal ectrophy and the frequent association with widely separated split penis or clitoris (Fig. 3 (C1, C2, C3)).

Treatment

Some details of the treatment of our four cases may be of interest because these show all the pitfalls in the treating of this condition and also how we were able to learn from our mistakes.

Case 1. The first case, a boy weighing 5 lb., was admitted moribund and any active treatment was out of the question. He died on the third day. A careful post-mortem dissection enabled us to form a detailed picture of the pathological anatomy of the condition.

Case 2. One year later we saw a second almost identical case when a few hours old. This infant was grossly premature, weighing 3½ lb. At operation the extrophied intestinal field was excised and on entering the abdomen it was found that the whole of the distal colon was duplicated (Fig. 4). This too was excised and the lower end of the ileum was pulled through to the perineum (Fig. 5). The two ectopic bladder fields were then united in the midline, the small exomphalos was repaired and the abdomen closed (Fig. 6). The infant stood the operation well, but it soon became apparent that the remaining length of the intestine was insufficient for adequate food absorption. In spite of numerous blood transfusions, plasma, protein hydrolysate and
Case 3. The third infant with vesico-intestinal fissure, another premature boy weighing 4½ lb., was admitted six months later (Fig. 7). Bearing in mind our unfortunate previous experience, we carefully preserved the ectopic intestinal field with its blood supply and closed the everted caecum and ascending colon. The distal colon, ending blindly in front of the sacral promontory, was freed and drawn down to the perineum. The two bladder fields were then closed in the midline as before. The exomphalos in this case was, however, a very large one and having already taken up most of the slack of the abdominal wall by suturing the bladder fields together, it now became absolutely impossible to cover with skin the abdominal defect produced by excision of the exomphalos. After several unsuccessful attempts at closure had been made, a piece of sterile nylon sheath was sutured to the mobilized abdominal skin across the defect. To our great surprise the child stood the operation quite well and when the nylon sheath finally separated two weeks after operation, there was granulation tissue underneath it, retaining the intestine inside the abdominal cavity (Fig. 8). Unlike Case 2, this infant thrived, the number of stools was not too excessive and he gained weight satisfactorily. The area of granulation tissue was covered by split skin grafts, and when 3 months old the child had doubled his birth weight.
Fig. 4.—Excised specimen from Case 2. The duplicated ascending colon, hepatic flexure and rudimentary double transverse colon are shown.

Fig. 5.—Case 2. The terminal ileum has been pulled through a hole made in the perineum.

Fig. 6.—Case 2. At end of operation.

Fig. 7.—Case 3. Note large exomphalos.

Fig. 8.—Case 3. After separation of nylon sheath.
VESICO-INTESTINAL FISSURE

Fig. 9.—Case 4.

Fig. 10. Case 4, aged 1 year. Note incisional hernia.

Fig. 11.—Case 4, aged 20 months, wearing urinary bag.
He then developed acute mechanical intestinal obstruction and, as conservative treatment was of no avail, his abdomen was opened. As was to be expected, the whole of the small intestine was glued together and closely adherent to the anterior abdominal wall. Only with the greatest difficulties was it possible to relieve the obstruction by dividing numerous adhesions. Although the child recovered from the operation, he had two further attacks of acute intestinal obstruction during the next three weeks, each of them necessitating a laparotomy. He finally died following the third laparotomy.

**Case 4.** We were somewhat disheartened by this experience and when a fourth case presented 18 months ago we devised a new plan of attack. Instead of opening the abdomen by incising the amnion covering the exomphalos, we now decided not to touch the exomphalos at all, but to enter the abdomen through the opening made by dissection of the intestinal field. We then wanted to proceed as in Case 3, repairing the caecum and pulling the terminal colon down to the perineum, and to close the abdomen by uniting the two ectopic bladder fields. If we found that there was sufficient abdominal wall and skin we would repair the exomphalos; if not, we would treat the exomphalos conservatively, employing Grob's (1957) method.

This child, a boy weighing 5½ lb., born on August 17, 1957, had a medium-sized exomphalos, a split penis and a small sacral myelo-meningocele (Fig. 9). His legs moved well, both testicles were descended. At laparotomy on August 18, he presented a picture identical with that of Case 3 with the exception that he had only one kidney, which was situated in the pelvis and had a very short ureter running to the left half of the ectopic bladder. After the bladder fields had been united it was found that there was enough slack in the abdominal wall to allow a complete repair of the exomphalos. The child stood the operation well. The post-operative period was complicated by an attack of mechanical obstruction when the child was 2½ months old. At laparotomy several adhesions had to be divided. The child made an uninterrupted recovery, gained weight slowly but steadily and started to stand up when 14 months old. He had a large incisional hernia (Fig. 10). There was no faecal continence, his perineal colostomy acting twice a day.

On February 20, 1959, it was decided to cut the left ureter off the ectopic bladder and, if possible, to perform a cutaneous ureterostomy in the left iliac fossa. At operation it was found that the length of the ureter was only about 1½ in. An end-to-end anastomosis between the ureter and a segment of isolated left ileum was therefore performed. In order to do this and to use as short an isoperistaltic piece of ileum as possible, it was necessary to rotate the ileal segment and mesentery through 180°, and to bring the loop out in the left iliac fossa. The dissection was made rather difficult by the numerous intra-peritoneal adhesions, but it was possible to rotate the loop without interference to its blood supply and to bring a 3½-in. long segment of ileum out in the left iliac fossa. Although the kidney is now draining urine practically straight upwards, there has been no stasis, and no hydronephrosis has developed. There is hardly any residual urine in the isolated ileal loop, possibly because of the vigorous peristalsis of the segment of intestine.

On April 23, 1959, the two ectopic bladder fields were excised and, after mobilization of the skin, it was possible to suture the wound under some tension. Most of the wound healed by first intention, but a small area in the centre sloughed. At the same time the two corpora cavernosa were divided from the widely separated pubic bones and the two penile halves were without difficulty brought together in the midline. No attempt was made to unite these two halves, but the posterior urethra and a small part of the vesical trigone were repaired.

The boy is now 22 months old and very well. He walks well, talks and appears of normal intelligence. He wears the infant's ileostomy urinary bag previously described (Rickham, 1956, 1958), and can be kept dry for two days at a stretch (Fig. 11). He is incontinent of faeces, but, in our experience, faecal incontinence is not a very great problem when the child gets older, and with the aid of a suitable diet, enemata and training it should be possible to keep him clean. He has intelligent parents and, with luck, his progress should be satisfactory.

In conclusion it should be said that vesico-intestinal fissure, the most severe malformation of the lower urinary tract, is perhaps not as rare as we previously supposed. After trial and error and committing (in retrospect) several stupid mistakes, we believe that the treatment outlined above may prove successful in the majority of cases and that the end result may well produce a useful member of the community.

Miss Isabella Forshall operated on Case 2 and kindly permitted its inclusion in this paper.

**References**


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Vesico-intestinal Fissure

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