

# PERMANENT URINARY DIVERSION IN CHILDHOOD

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by

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I AM GRATEFUL to the Council of the Royal College of Surgeons for allowing me to give this paper. It is a great privilege to deliver a lecture in Hunter's honour, and the collection of the material for this lecture has been of great value to me. To quote John Hunter: ". . . what more than all induced me to lecture, was the great advantage everyone finds by putting his thoughts into writing. A man can never tell how much he knows till he arranges his knowledge, and then he can tell how defective it is. . . ."

The subject of permanent urinary diversion in childhood has fascinated me for over 15 years for a variety of reasons. There is obviously no ideal solution to this problem. Surgeons both in this country and overseas hold widely different views on the advisability of any operation, the best operative method and the optimum time for operation. During recent years our views on the subject have become more accepted on both sides of the Atlantic, but they are still strenuously opposed in many quarters. The operation of transplanting the ureters into an isolated segment of ileum in children has been carried out by us for the last 10 years. Bricker first performed the operation in adults in 1950, and Bill *et al.* (1954) in Seattle, and Nash (1956) at St. Bartholomew's Hospital, both operated on children in 1954. We have by now considerable experience with this operation and are prompted to record this experience, remembering how long it took the medical profession to realize the bad long-term results of uretero-colic anastomosis. The history of uretero-colic anastomosis should be a warning to future generations of surgeons.

Uretero-colic anastomosis was first described by John Simon, of St. Thomas's Hospital, in 1852, for a child with ectopia vesicae. It testified, as he said, ". . . to the ardent wish of the surgeon to benefit his patient", but he added: "the risks are perhaps disproportionate with the annoyance of a malformation which improved apparatuses may render bearable". Simon's first attempt and many subsequent attempts by others were failures, and it was not until the beginning of this century, when Stiles (1911) and Coffey (1911) first devised the modern techniques for uretero-colic anastomosis, that the operation became a standard procedure.

It is a curious comment on medical thought that the disastrous long-term results of this operation were not widely recognized for four decades.

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There are two reasons which might explain this lack of vigilance in the profession, which is surprising, as complications of the operation were reported in the literature all through this period and more than 100 different techniques were described. Firstly, the majority of urinary diversion procedures were and are carried out on adults mainly for malignant growths of the bladder, and the expectation of life of these patients is short while complications take time to develop. Secondly, there was the enormous influence of Grey Turner, who in the 1929 Hunterian Oration painted a much too rosy picture of the fate of the 16 patients with ectopia vesicae for whom he had carried out uretero-colic anastomosis. He admitted that recurring pyelonephritis was a common occurrence in the 12 survivors, but added that these attacks were of no great consequence. By 1943, of the nine surviving patients nearly all had clinical evidence of some ascending infection, three had renal calculi, one had undergone a nephrectomy for gross pyonephrosis and one was lost to the follow-up. These are hardly reassuring results, but the operation remained nearly unchallenged until the early nineteen-fifties, when large-scale follow-up studies showed the frequency of such complications as recurrent pyelitis, hydronephrosis and hypercholaemic acidosis (Ferris and Odel, 1950; Harvard and Thompson, 1951; Jacobs and Stirling, 1952; Williams and Jolleys, 1952; Annis, 1953; Garrett and Mertz, 1954).

There is a lesson to be learned from the past and any new operation for urinary deviation must be subjected to a careful and critical long-term follow-up study. Such a study is only possible in children, as the expectation of life of most adults subjected to this procedure is too short. The operation of transplanting the ureters into an isolated ileal loop, which I have carried out on 70 children during the last 10 years, is a much more extensive procedure than uretero-colic anastomosis, and the end-result, necessitating the wearing of a urinary bag, is much less satisfactory. Only if the follow-up studies are much more satisfactory than those of other procedures and only if no better conservative or surgical method of treatment is available can there be any justification for the operation. "Operation", to quote John Hunter again, ". . . is a reflection of the healing art; it is a tacit acknowledgement of the insufficiency of surgery. It is like an armed savage who attempts to get that by force which a civilized man would get by strategem." And he concludes: "No surgeon should approach the victim of his operation without a sacred dread and reluctance, and should be superior to that popular *éclat* generally attending painful operations often only because they are so, or because they are expensive to the patient."

### **Indications for permanent urinary diversion in childhood**

Widely different views are held concerning the indication for permanent urinary diversion. So much has been written about this subject and so

different have been the opinions expressed that it is impossible to give here more than an outline of literature on the subject.

Time and further experience has not changed our views regarding the indication for diversion procedures since this matter was discussed at the 12th Annual Meeting of the British Association of Urological Surgeons eight years ago (Rickham, 1956). Broadly speaking there are four types of cases which might be considered for this type of operation: (1) the congenital neuropathic bladder, (2) ectopia vesicae and allied conditions, (3) other malformations of the lower urinary tract, and (4) certain cases with long-standing severe obstruction of the lower urinary tract.

*The congenital neuropathic bladder* is a common complication of myelomeningocele. It may also occasionally be associated with some types of rectal agenesis, especially those with absence of the lower three segments of the sacrum (Williams and Nixon, 1957).

Deficient nerve supply to the bladder was by far the commonest cause for which we carried out urinary diversion. Of the 96 children on whom a diversion procedure was performed during the last 10 years 58 had myelomeningoceles and 3 patients had rectal agenesis (Table I).

TABLE I  
CAUSES FOR URINARY DIVERSION

Spinal bladders .. .. .	59
Ectopia vesicae .. .. .	22
Cloaca .. .. .	5
Valves in posterior urethra .. .. .	5
Rectal atresia .. .. .	3
Malignant tumours .. .. .	2
Advanced tuberculosis .. .. .	1
Total .. .. .	97

There is nearly always associated involvement of the anal sphincters and the children suffer from faecal incontinence. This can usually be controlled by simple training, at first by the use of enemata and/or suppositories, later on by establishing a reflex action of the bowels every morning and frequently also every night. We have had very little trouble with faecal incontinence once the children reached school age, provided that they were conscientiously trained by parents or nurses during the toddler stage. In congenital neuropathic bladder secondary to myelomeningocele the bladder is either flaccid and there is no residual urine and minimal, if any, urethral resistance or there is overflow incontinence with a hypertonic, trabeculated bladder, often studded with multiple diverticula, massive reflux up the ureters, hydro-ureters and hydro-nephroses. In our series both types of bladders were about equally common, but in others cases with complete incontinence have predominated (Nash, 1957).

When discussing the treatment of congenital neuropathic bladder, one must realize that the problem is totally different from the acquired

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neuropathic bladder following spinal cord injuries at higher levels and that there is usually little hope of establishing an automatic emptying cord bladder in the patients with myelomeningoceles. As the involvement of the anal sphincters makes it impossible to achieve urinary continence by operation, it is understandable that many surgeons have been reluctant to perform operations for urinary diversion in this type of case. This explains why such procedures as expression of the bladder, catheter drainage and exploration of the spinal cord have been carried out so frequently in the past. We have practised all of these procedures without much success.

In the overflow incontinence type of bladder Cr  d   expression carried out by the mother should always be given a conscientious trial (Prince and Scardino, 1953). If the sphincter spasm is too great, treatment with antispasmodic drugs and, if this is not successful, weakening of the bladder sphincter by Y-V or transurethral resection (Emmett and Helmholtz, 1948) should be carried out. However, in these cases ureteric reflux and dilatation of the upper urinary tract nearly always develop in time. It has been stated that reflux and upper urinary tract damage is always the result of urinary infection (Mitchie, 1963), but this is not correct as it can be observed in newborn infants with neurogenic bladders. Urinary infection does, however, play a major additional role in further decreasing renal and vesical function, and determined attempts must be made to control it by chemotherapy.

Continuous vesical catheter drainage (Kennedy and Hodges, 1957; Conger and Taub, 1960; Rose and Smith, 1963) and the insertion of a suprapubic catheter after closing the urethra (Gross, 1953) have been practised widely in the United States. In our experience continuous catheter drainage of the bladders of small infants, whether suprapubic or transurethral, is an extremely dangerous procedure especially in the presence of upper urinary tract damage (Brandesky and Rickham, 1962).

Exploration of the spinal cord, division of adhesions and the tight filum terminale appears to have improved the urinary incontinence in a very few cases of highly selected patients (Thompson *et al.*, 1957; Thompson and Jackson, 1958). We have carried out this procedure in nearly 20 patients without ever having seen any benefit as far as bladder function was concerned.

While in girls no satisfactory appliance can be fitted, penile clamps and bags have been frequently recommended. Penile clamps (Campbell, 1951) are only mentioned in order to condemn them. The discomfort they cause in boys with unimpaired sensation and the damage they can cause in boys with anaesthesia of the external genitalia (Nash, 1963) are definite contra-indications. Penile bags can be of use provided the penis is large enough, the skin not anaesthetic and the bag does not interfere

with orthopaedic appliances, and we have a small number of boys who manage their bags very satisfactorily.

*Ectopia vesicae.* To-day the standard treatment of this severe malformation is plastic closure of the bladder (Sweetser *et al.*, 1956; Sweetser, 1954; Chisholm, 1962; Swenson, 1958). Unfortunately the majority of plastic closures of the bladder are unsatisfactory because of incontinence, residual urine, recurrent infection, stone formation and hydronephrosis (Rickham, 1957, 1958, 1961) and will also finally need urinary diversion.

The reasons for the unsatisfactory results of uretero-colic anastomosis carried out directly or by using Maydl's (1894) procedure, recently resurrected by Wallace (1961), have already been described. Any operation where the inefficiently contracting bladder is incorporated in the drainage system (Powell, 1956; Lowsley and Johnson, 1955; Cordonnier, 1957) and any operation where the anal sphincter is used to control urinary incontinence (Gersuny, 1898; Duhamel, 1957; Boeminghaus, 1956; Singer, 1959) must in the long run give rise to the same complications as uretero-colic anastomosis. Residual stagnating urine will accumulate, leading to reabsorption and infection and, in addition, the pressure necessary to overcome the tone of the anal sphincter will by necessity lead to back-pressure affecting the upper urinary tract. The large number of such operative procedures described in the literature show that none of these methods is satisfactory. They have certainly been disastrous in our hands. Urinary diversion operations will nearly always be necessary in the most extensive variety of ectopia vesicae, vesico-intestinal fissure (Rickham, 1960).

*Other malformations of the lower urinary tract*, such as cloaca, will also need urinary diversion operation as these patients have usually faecal as well as urinary incontinence.

*Lower urinary tract obstruction due to congenital malformations of the bladder neck or the posterior urethra* is a common condition in childhood; it is treated by removal of the obstruction. In the past, grossly neglected cases with enormously distended and sacculated bladders, massive hydro-ureters and hydronephroses were frequently observed. Now that paediatricians have become aware of the condition such cases are rare. On the other hand the improvement in the management of newborn infants with surgical conditions (Rickham, 1962*b*) has allowed some infants with enormous bladders and nearly completely destroyed kidneys to survive the immediate neonatal period. In these children the bladder is grossly distended and the vesical musculature stretched and often nearly completely replaced by fibrous tissue. Removal of the obstruction to the urinary flow will frequently not cure the urinary retention as the bladder muscle is unable to contract. Here urinary diversion may be a life-saving procedure. In the less severe cases the operation is only of a temporary nature, in some of the very severe ones diversion has to be

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permanent (Rickham, 1962a). We have carried out urinary diversion in some patients over eight years ago and have repeatedly attempted to reconstruct the normal urinary tract, but were prevented from doing so by the total inability of the bladder to contract even after prolonged periods of training, tidal drainage, drug treatment, etc.

### **The optimum time for urinary diversion**

Here again there is no unanimity of opinion. It has been argued that operation should be postponed until the child reaches the age of 6 to 8 or even 12 to 13 years, on the grounds that bladder control may improve in time (Swenson *et al.*, 1956; Pyrah, 1956; Williams, 1958, 1962; Spellman and Kickham, 1962). In our experience this has only rarely been the case; more commonly repeated attacks of urinary infection have caused deterioration of urinary control and function. In cases with retention overflow there is the added danger of further deterioration of renal function because of back pressure effects. Those who recommend delayed urinary diversion procedures seem to be curiously unaware of the psychological aspect of urinary incontinence. It seems cruel to allow a child dribbling urine to grow up as an outcast. There can be little doubt that the psychological impact of an operation which leaves the child with a urinary bag is a severe one. We have found that this effect is greatly minimized by operating early upon these children. Early operation has its own special difficulties both in operative technique and in bag management, but this is more than outweighed by the fact that by the time the child reaches school age he is accustomed to wearing the bag and in the majority of cases manages it without difficulties. He obviously realizes that he is different from his fellows, but provided home conditions are adequate he has come to accept this fact. The only example of marked psychological upset following the operation which we have seen occurred in a girl operated upon when she was eight years old and we have been again and again amazed at the cheerful acceptance with which children treat this disability. In cases with overflow incontinence it has been our custom to carry out a urinary diversion procedure whenever we were unable to halt the back-pressure effect by training or operation and we are of the opinion that procrastination in these cases is unwise. In children with flaccid bladder and complete incontinence the optimum time for urinary diversion is in our opinion two years of age. Prior to this age there is little to be gained, as even the normal child would live in nappies. If urinary diversion is carried out at this age both mother and child have ample time to accustom themselves to the bag management before school attendance, and when he reaches the age of five the child has nearly forgotten how it is to be without a urinary bag.

### **Operative procedures for urinary deviation**

The contra-indications for uretero-colic anastomosis have already

been outlined and the inadvisability of operations necessitating the use of indwelling catheters has also been mentioned. Operations allowing the urine to flow through the paralysed bladder and procedures where the anal sphincter controls the drainage system are also undesirable. The so-called tubeless cystostomy aimed to overcome the disadvantages of catheter drainage (Blackson, 1957; Rinker *et al.*, 1959; Carlson, 1960; Lapidès *et al.*, 1960). Our experience with this operation has not been a happy one. We have found that the paralysed bladder failed to contract after cystostomy and the urine stagnated unless the child was constantly nursed lying prone on a frame.

Finally, a urinary conduit can be constructed allowing the urine to flow out of an opening in the abdominal wall, where it can be collected in a bag. The caecum with the appendix as spout (Verhoog and Gräinne, 1909; Makkas, 1910), a loop of jejunum (Marion, 1912), the ileo-caecum with the appendix (Lengemann, 1912) or ileum (Gilchrist *et al.*, 1950) as spouts or the colon (Uebelhoer, 1952) have all been employed, but these procedures have not been generally accepted and only the isolated ileum has been widely used. The ileum is a more satisfactory conduit for urine than is the colon as it peristalses more rapidly, but it must not be forgotten that ileal peristalsis exists primarily for the churning of food, not for the propelling of urine (Boyarsky *et al.*, 1962). There is therefore a possibility of urinary constituents being absorbed through the ileal wall although not to the same degree as happens in the colon (Hopewell, 1959). The ileal loop must therefore be short and in adults should not exceed 6 to 8 inches. In infants and small children it must be still shorter, and we have found that an intra-abdominal length of maximum 4 inches, preferably 2½–3 inches, is all that is permissible (Rickham, 1961). In children the original Bricker (1950) procedure must therefore be slightly altered to allow the ileal loop to pass straight through the abdominal cavity from the promontory of the sacrum posteriorly to the skin, opening beside the umbilicus anteriorly. Provided the left free margin of the triangular mesentery of the isolated ileal segment is securely tucked down to the posterior abdominal wall, there is no danger of intestinal obstruction (Rickham, 1957).

The operation should not be carried out in the presence of advanced renal failure and gross pyelonephritis, and whenever we have disregarded this proviso we have been in serious trouble. The only two deaths secondary to operation which occurred in this series happened in children with advanced renal failure, who died of uraemia. Both these deaths occurred over eight years ago and since that time we have avoided performing the operation in the presence of advanced renal damage. Unfortunately a considerable proportion of the infants and children who come under observation have grossly damaged kidneys with a persistent blood urea of 100 mgm. per cent and more marked changes in the blood chemistry and advanced and uncontrollable pyelonephritis.

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For these children cutaneous ureterostomy is a life-saving procedure. Although this operation has been known to surgeons for a very long time (Simon, 1875), and although good results in a small number of cases have been repeatedly recorded in the past (Cabot and Scherer, 1935; Trabucco and Marquez, 1950; Keyes, 1940; Folsom and O'Brien, 1941), the operation has not been popular mainly for three reasons. Firstly, because clumsy bilateral collecting mechanisms had to be applied (Band, 1942; Carleton and Nagamatsu, 1950; Fish and Stevenson, 1949). Secondly, because nearly all patients with indwelling catheters will sooner or later suffer from gross pyuria (Humphreys, 1956); and thirdly, because there is a tendency to stricture formation in cutaneous ureterostomies.

Fortunately in the children under discussion all these disadvantages can be overcome. In these patients with advanced renal damage due to back-pressure the ureters are not only enormously dilated but also very tortuous and elongated. In 1958, my colleague, J. H. Johnston, and I described a method by which the openings of the two ureters were placed side by side in the midline of the abdomen below the umbilicus, necessitating the use of only one urinary bag. Subsequently a similar operation was described by Swenson and Smyth (1959) and Chute and Sallade (1961).

During the last eight years I performed cutaneous ureterostomies in 26 children with hydronephrosis, hydro-ureters and often advanced renal damage. Considering the precarious condition of many of these patients the results have been surprisingly good. The various bags which were designed (Fig. 1*a* and *b*) make indwelling catheters unnecessary, and the procedure is thus safer (Wosnitzer and Latimer, 1960). In order to prevent leakage the formation of a ureteric spout was introduced by Papin in 1927 and has recently been resurrected by Eckstein (1963). In some of our earlier cases we too constructed such a spout, but we found after a few years that stenosis invariably occurred. Since we have sutured the ureteric openings flush to the skin we have had no more trouble with stricture formation, and there has been practically no leakage from the bags.

Urinary deviation is obviously only a poor second best. Whether it will be possible to control bladder contraction by electric stimuli from implanted electrodes (Bradley *et al.*, 1963; Ellis, 1964) only the future will show.

### Analysis

During the last ten years I have performed 70 ileocutaneous ureterostomies; the ages at which operation was carried out are shown in Figure 2. Six children have died and one child was lost for the follow-up. It was thought that only patients operated upon more than a year ago should be considered for this study (Table II), and as nine children were treated less than a year ago there remained a total of 54 patients (Fig. 3).



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All these patients have had periodic clinical examinations, catheterization of the ileal loop, cultures of their urine, intravenous pyelograms and blood chemistry studies. At present all children are perfectly well and in all but three with advanced renal damage growth and weight gain has been satisfactory (Table III). In view of the fact that many of these patients had myelomeningoceles and hydrocephalus the high percentage of mentally normal patients is very gratifying; only one child had an intelligence quotient which made him not eligible for education.

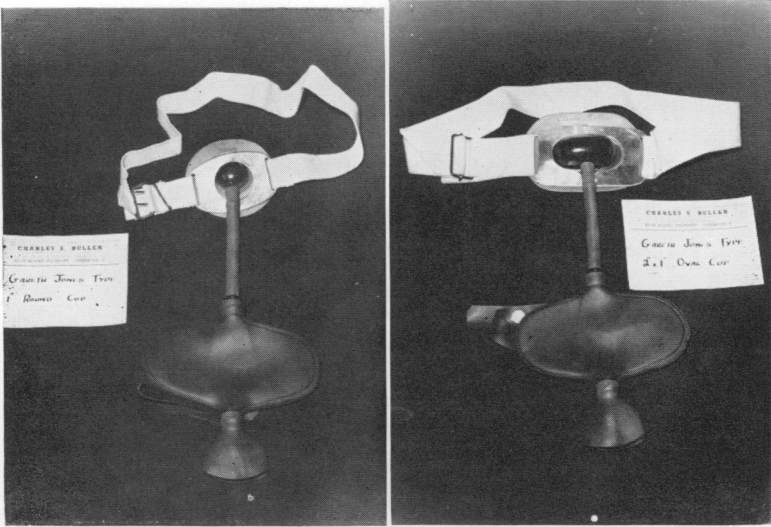


Fig. 1. (a) Bag for central cutaneous ureterostomy openings. (b) Bag for cutaneous ureterostomy openings lying side by side.

Amongst the 27 patients with cutaneous ureterostomies performed during the last eight years (Fig. 4) the death rate was much higher. This is not surprising as in this group there were many patients with advanced

TABLE II  
TRANSPLANTATION OF URETERS INTO ISOLATED ILEAL LOOP

Follow-up for more than 1 year ..	54
Operation less than 1 year ago ..	9
Not traced .. .. .	1
Deaths .. .. .	6
Total ..	<u>70</u>

renal disease and urinary infection. Only 14 patients qualify for the purpose of our study (Table IV). The length of follow-up can be seen from Figure 5. The proportion of children of average height and weight is

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lower than in the previous group, but most of them are of normal intelligence (Table V).

Before considering the present state of health of our patients in more detail, it may be advisable to discuss the complications which we have encountered in the past.

Amongst the ileal loop patients the only common complication was

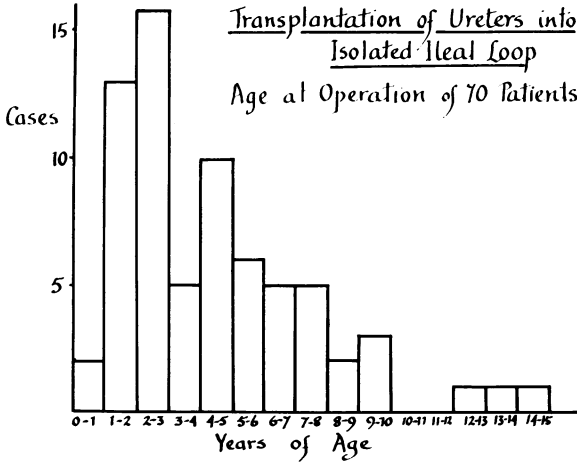


Fig. 2.

stenosis of the ileal stoma (Table VI). This complication is less common in adults and older children, although it may also occur in the older age groups (Creevy, 1960), but is very frequent in children operated upon at

TABLE III  
FIFTY-FOUR CASES FOLLOWED-UP FOR MORE THAN ONE YEAR

Sex ..	21 males	33 females
Physical development ..	51 normal	3 below normal
Mental development ..	47 normal	6 below normal but educable 1 ineducable

an early age. It is the price one has to pay for the advantages of early operation. The stricture may occur at any level between the peritoneum and the skin. This complication has become less common since we

TABLE IV  
CUTANEOUS URETEROSTOMIES

Follow-up for more than 1 year ..	14
Operation less than 1 year ..	7
Not traced .. ..	1
Deaths .. ..	5
Total .. ..	<u>27</u>

(21 both ureters transplanted; 6 single ureter transplanted)

routinely core out a disc of abdominal wall with the aid of a special hollow knife (Fig. 6*a* and *b*). Provided the child is periodically examined and the loop catheterized, and provided prompt action is taken if the stoma becomes stenosed or the volume of residual urine increases above one ounce, it is not a serious complication. If, however, the stricture is missed, the ileal loop will quickly distend with urine and its length and

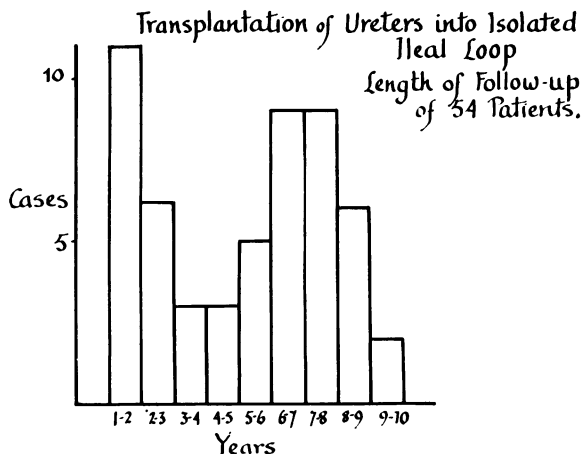


Fig. 3.

width will increase to enormous dimensions. As there is a mucosa-to-mucosa anastomosis between ureter and ileum (Fig. 6*c*), similar to Nesbit's (1949) uretero-sigmoid anastomosis, it is not surprising that hydro-ureters and hydronephroses rapidly develop. Hyperchloraemic acidosis with very low plasma potassium levels result and may cause severe clinical symptoms. Luckily all these changes are quickly reversible

TABLE V

FOURTEEN CASES FOLLOWED-UP FOR MORE THAN ONE YEAR

Sex . . .	6 males	8 females
Physical development . .	9 normal	5 below normal
Mental development . .	11 normal	2 below normal but educable 1 ineducable

once the condition has been recognized, the stricture excised and the ileal loop shortened. Nevertheless the possibility of this complication makes it imperative to examine children under five years at monthly or bi-monthly intervals, and even older children must be carefully watched.

Recurrent pyelonephritis, which was such a common complication, or uretero-colic anastomosis was not observed in this group of children, which is surprising as the urine in the ileal loop is only sterile in less than half of the cases. It appears that, in spite of the wide uretero-intestinal

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anastomosis, ureteric peristalsis can in the absence of back-pressure effectively prevent ascending urinary infection. We have had no case of post-operative intraperitoneal leak of urine or intestinal contents and peritonitis, which appear not infrequent in adult series (Wells, 1956).

Three of the four other complications observed were due to a technical error. Kinking of the loop due to intra-abdominal adhesions occurred twice and necessitated division of the adhesions (Fig. 7a). Stricture of the

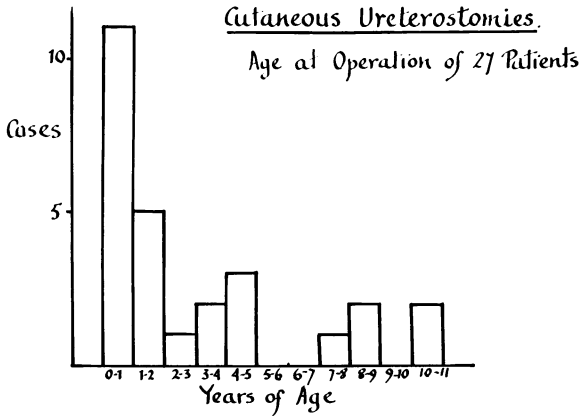


Fig. 4.

anastomosed ureter occurred only once in 138 anastomoses. One child developed a left hydro-ureter and hydronephrosis in the absence of strictures of the ileal stoma or the ureteric anastomosis. We were unable to find a reason for this phenomenon and wonder whether partial devascularization of the lower ureter and consequent defective peristalsis may be the cause.

TABLE VI  
COMPLICATIONS (ILEAL LOOP)

Stricture of ileal stoma . . . . .	28
Obstruction of loop due to kinking . . . . .	2
Stenosis of one ureteric anastomosis . . . . .	1
Dilatation upper urinary tract, cause unknown . . . . .	1
Deaths connected with operation . . . . .	2
Deaths not connected with operation . . . . .	4
(1 strangulated inguinal hernia; 2 hydrocephali; 1 malignant tumour)	

There were two operative deaths due to an error of judgement, an operative mortality of under 3 per cent comparing favourably with other published series (Annis, 1956; Hodges *et al.*, 1961). There have been no late deaths due to renal failure (Cordonnier and Nicolai, 1960).

Four of the six deaths were not related to the operation, the cause of death ranging from unrecognized strangulated inguinal hernia to

rhabdomyosarcoma. Three of the earlier cases with cutaneous ureterostomies (Table VII), where we attempted to construct a cutaneous nipple, developed strictures which had to be excised. Of the five deaths three were associated with the patients' gross renal damage. Considering the advanced renal damage of many of the patients it is surprising that there have not been more fatalities.

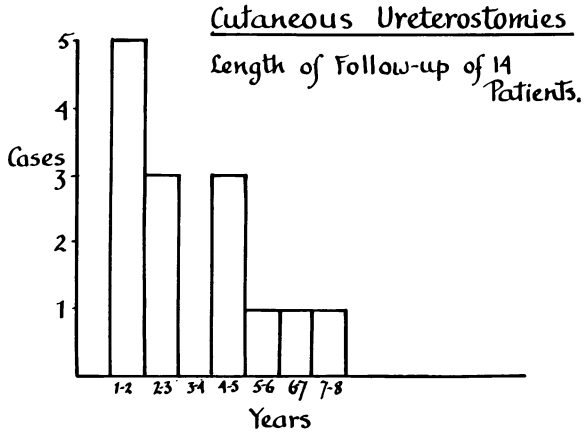


Fig. 5.

All the 54 patients with ileo-cutaneous ureterostomies carried out more than one year ago are in good health, have only a few millilitres of residual urine and no evidence of infection of the upper urinary tract. Pre-operatively 11 patients suffered from moderate and 10 from severe

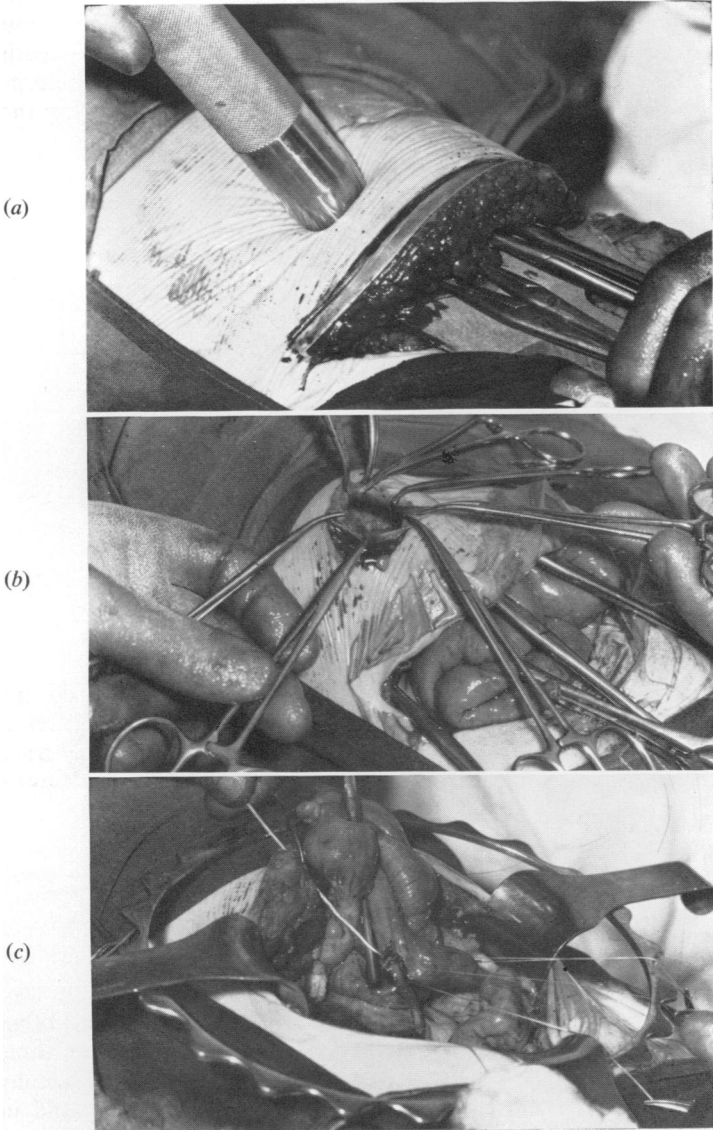
TABLE VII  
COMPLICATIONS (URETEROSTOMIES)

Stricture of the stoma	..	..	..	3
Deaths	..	..	..	5
Renal failure	..	..	..	1
Renal failure + infection	..	..	..	2
Meningitis	..	..	..	1
Malignant tumour	..	..	..	1

hydronephroses and hydro-ureters, the upper urinary tract of the other patients was normal. Apart from the four cases mentioned above, three of whom have only recently been corrected, intravenous pyelography showed improvement of the upper urinary tract in 16 children. and in 34 children there was no change (Figs. 7b-10).

The more important data of the blood chemistry were as follows: The blood urea was within normal limits in all but three patients and these patients had an even higher blood urea before operation. The serum chloride concentration of all patients was within normal limits and in no single instance was there a suggestion of hyperchloraemia,

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**Fig. 6.** (a) The hollow knife coring out a disc of abdominal wall for the ileostomy opening. Note the forceps steadying the peritoneum. (b) The abdominal wall opening for the ileostomy after a disc has been cored out by the hollow knife. The mosquito forceps have been used to pick up the peritoneum. (c) The mucosa-to-mucosa anastomosis between the left ureter and the isolated ileal loop is half completed. A probe is inserted through the opening into the ileum.

although this is reputed to occur in cases where the kidneys are severely infected pre-operatively (Parkhurst and Leadbetter, 1960). The plasma potassium level was normal in all but nine cases, where it was between 3 and 3.5 m.Eq/l. The plasma alkali reserve was also normal in all but nine patients, where it was slightly below normal at levels ranging between 20 and 22 m.Eq/l. Surprisingly the low plasma potassium levels and relatively mild acidosis did not always occur together, but low plasma potassium levels were found in the presence of a relatively high alkali

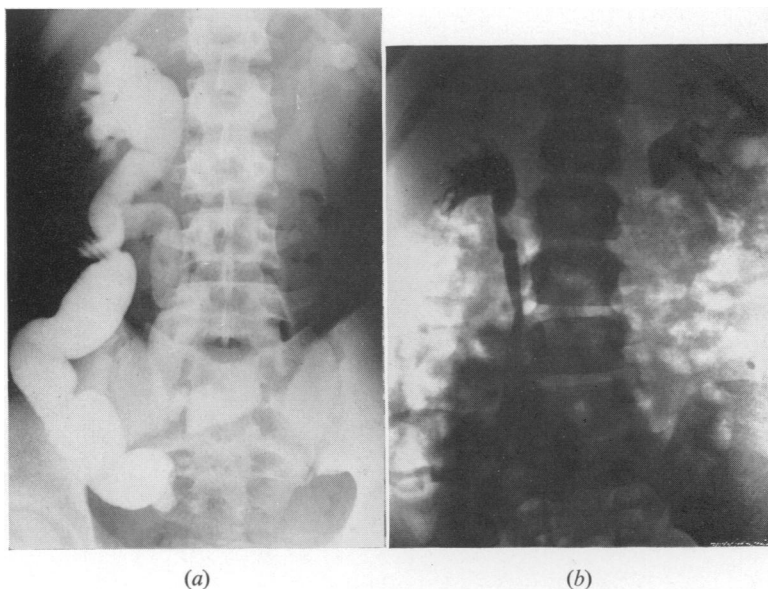


Fig. 7. (a) I.V.P. of patient with ileo-cutaneous ureterostomies. There was an intraperitoneal adhesion crossing the proximal part of the isolated ileal loop, causing back pressure and bilateral hydro-ureters and hydronephroses. Division of the adhesion was followed by complete recovery. (b) I.V.P. 10 years after transplantation of ureters into isolated ileal loop (ectopia vesicae).

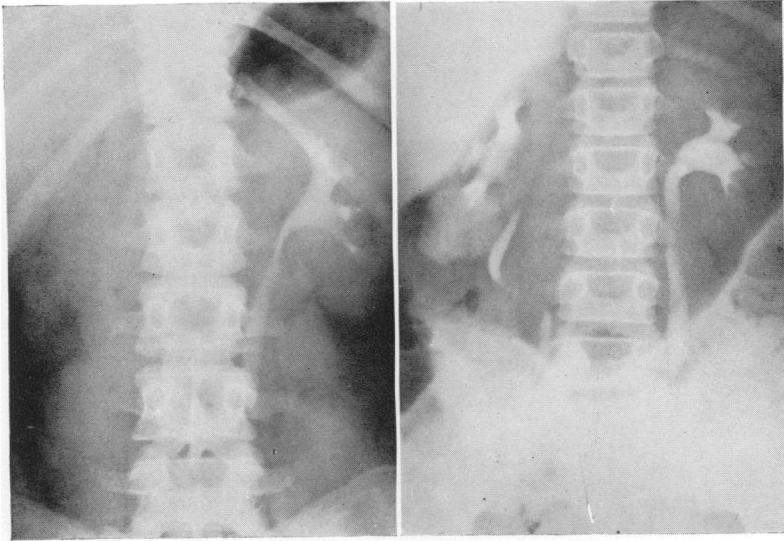
reserve and vice versa. These occasional changes in the potassium level and alkali reserve, which were not associated with any ill health or depression of renal function, may be explained by the fact that many of these children had been on prolonged administration of acid sodium phosphate solution in order to prevent ammoniacal dermatitis round the ileal stoma, which in some children can be somewhat troublesome. Although we still use this medicine in order to clear up the dermatitis we now refrain from prescribing it for too long a period.

When considering the 14 patients with cutaneous ureterostomies the outlook is less pleasant, because some of them have hardly any kidney substance at all and are perpetually at the brink of renal failure.

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Pre-operatively one patient had a moderate degree of hydronephrosis and in 13 the hydronephroses were severe. Post-operatively the upper renal tract has improved in seven and remained the same in five patients. In two the renal function was so bad that the intravenous pyelogram never showed any renal function at all.

In 10 children the blood urea was within normal limits, in one it was between 40 and 50 mgm. per cent and in two well above this level. In all cases it had improved since operation. The serum chloride level was



(a)

(b)

Fig. 8. (a) I.V.P. of patient with ectopia vesicae. Uretero-colonic anastomosis 14 years ago. Recurrent pyelonephritis necessitating right nephrectomy and severe hyperchloraemic acidosis. Transplantation of left ureter into isolated ileal loop 9 years ago. (b) I.V.P. of patient with cloaca who had her ureters transplanted into an isolated ileal loop 6 years ago.

normal in 10 patients; one had hypo- and two hyperchloraemia. Again there has been an improvement since operation in every case. The serum potassium was normal in all but one child, where it was slightly below the lower limit of normality. Six patients had a persistent and two of them a severe acidosis; these latter two are renal dwarfs. Both these patients could only be kept alive by the daily intravenous and intra-gastric administration of vast quantities of water for over a year and excreted urine of the specific gravity of between 1,001 and 1,002.

The satisfactory management of the urinary bag is of the greatest importance (Table VIII). Forty-three of the 54 patients with ureteric transplant into isolated ileal loops are dry day and night, seven had



occasional "accidents" during the day when they forgot to empty the urinary bag, and four had occasional wet beds at night. It is the younger child who will have accidents, but provided he has a conscientious mother he soon learns to eliminate these mishaps. These satisfactory results could not have been achieved without the help of Mr. Charles Bullen, who designed a variety of urinary bags to fit the special requirements of the various patients. The bags for cutaneous ureterostomies have already been described. Apart from the standard bags for the ileostomy

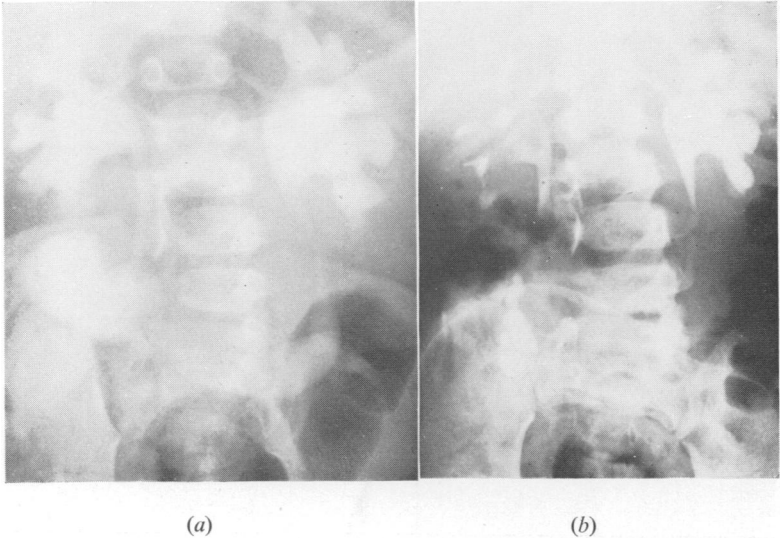


Fig. 9. (a) I.V.P. of patient with congenital neuropathic bladder and bilateral hydronephroses 6 months after transplanting ureters into an isolated ileal loop. (b) Same patient as in (a), 5 years later.

stoma we have found two types of bags especially useful: the infant bag with the lateral opening for night drainage (Fig. 11a), which is used

TABLE VIII  
BAG MANAGEMENT

1. <i>Ileal Loops</i>				
Dry day and night	..	..	..	43
Still occasionally wet during day	..		..	7
Still occasionally wet during night	..		..	4
2. <i>Ureterostomies</i>				
Dry day and night	..	..	..	8
Occasionally wet	..	..	..	4
Not yet wearing bag	..	..	..	4

for most children below school age and often for several years longer, and the bagpipe bag (Fig. 11b), incorporating a non-return valve, which is especially useful for children wearing callipers or sitting in wheelchairs.

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### Long-term management and social integration

During the last 20 years the surgery of congenital malformations has rapidly developed and many conditions which were formerly regarded as incurable can now be remedied. Unfortunately there is a misconception in most lay and even some medical minds, that surgery can restore these unfortunate children to complete normality. This is certainly not true for many of the severe malformations of the cardiovascular, central nervous, urinary and locomotor systems. The best that can at present

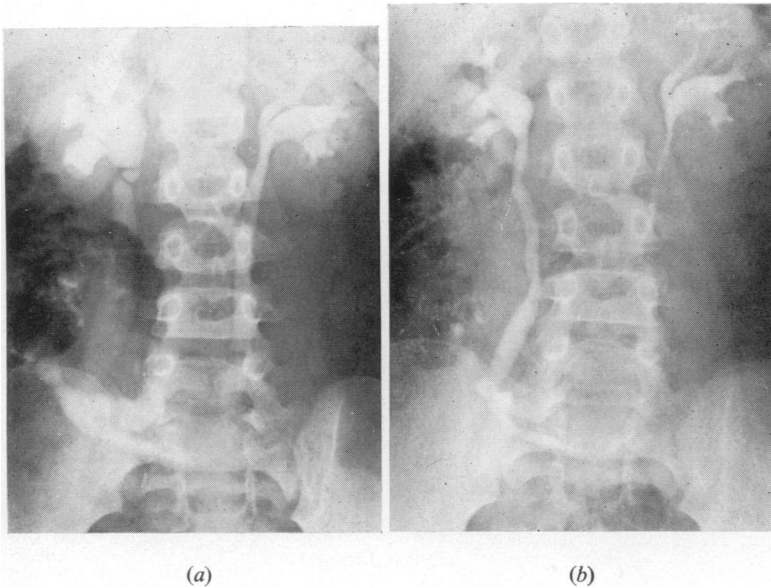


Fig. 10. (a) I.V.P. of patient with congenital neuropathic bladder 6 months after transplanting ureters into isolated ileal loop. (b) Same patient as in (a), 7 years later.

be hoped for in many of these children is to make them socially acceptable and allow them to live a useful and not too unpleasant existence. I think most doctors will agree that we as medical men cannot refuse treatment, but children like the ones under discussion need medical supervision for the rest of their lives. In order to do the best for these children there must be close co-operation between the various medical and surgical specialists treating the different malformations and, in addition, psychiatrists, physiotherapists, nurses, teachers and, last but not least, the parents have to be integrated into a team. Doctors are shirking their responsibility if they confine their efforts to the treatment of the malformations but regard education and training for employment of these children as a "social" question best left to the State or voluntary organizations.

At present many people do not seem to realize the size of the problem. There has been a public outcry about a couple of hundred thalidomide babies born in this country and money and medical and scientific resources have been mobilized in order to help them. Fortunately the thalidomide tragedy will not recur, but the thousands of children born every year with severe malformations of the central nervous and urinary systems escape public attention. There are only 97 cases of permanent urinary deviation in this series, and my colleagues at the two Liverpool Children's Hospitals operated on 25 others during the same ten-year period.

This is, however, not the whole story. We operate upon an ever increasing number of myelomeningoceles, at present about 150 a year; a considerable percentage of them will need urinary diversion in the future. As yet insufficient provision has been made for the rehabilitation

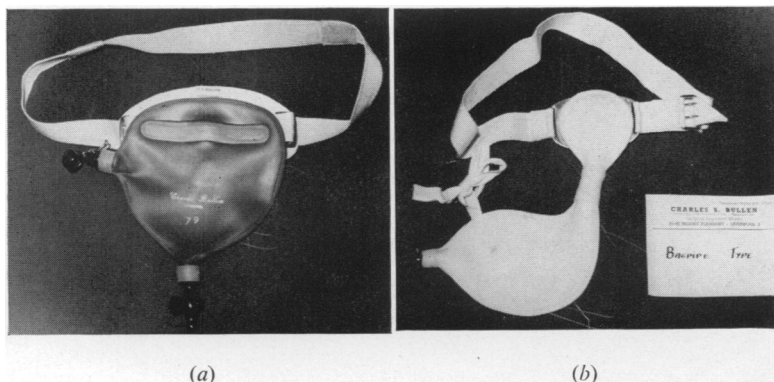


Fig. 11. (a) Infant ileostomy bag with side opening for attaching night tube. (b) The bagpipe ileostomy bag.

of this ever increasing number of children under the National Health Service. Centres for efficient primary and secondary treatment are urgently needed. The idea that these children are all mental defectives has been disproved by the figures presented here. Investigation into the fate of the 68 children on whom permanent urinary diversion operations have been carried out over a year ago shows the following (Table IX): 30 or nearly half of the children live at home or go to ordinary schools ranging from elementary to grammar and public schools. A number of these children wear orthopaedic appliances, quite a few have complete paralysis of the lower limbs and can only move in wheelchairs and full-length callipers. Seventeen children go to schools for physically handicapped children—excellent schools, but not primarily designed for this type of case as they have to admit a considerable proportion of mentally dull children. Quite a number of children in this series were reasonably bright, but because of poor home conditions could not be kept with their

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parents. Two children have home teaching, i.e. are visited by a teacher for a few hours each week, a thoroughly unsatisfactory arrangement. One child has been waiting for a home teacher for three years. Thirteen are too young for school and five have left the school and are studying or in employment, which was obtained after great difficulties, as most large firms demand medical examination of their employees and will not accept physically handicapped applicants.

TABLE IX  
FOLLOW-UP SCHOOLING AND EMPLOYMENT  
(68 cases operated over 1 year ago)

Ordinary school	..	..	..	..	30
School for physically handicapped	..	..	..	..	17
Home teacher	..	..	..	..	2
Home teacher not available	..	..	..	..	1
Too young for school	..	..	..	..	13
Left school	..	..	..	..	5
Student Architect					
Accountancy Clerk					
Glazier					
Mechanic					
Seamstress					

Provided the orthopaedic malformations are not severe the children tend to do very well at school and often none of their classmates realize that they wear a urinary bag. Some of the boys have excelled in games and have swum, played football, or competed in athletics for their school, but there is little doubt that the more handicapped patients would greatly benefit by more efficient physiotherapy, teaching and rehabilitation.

ACKNOWLEDGEMENTS

I should like to express my thanks to my registrars, house surgeons and ward sisters, without whose help this work would not have been possible. I am most grateful to Dr. Carol Martin for helping me to collect the material for the lecture.

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## SIR HUGH LETT, Bt.

IT IS WITH deep regret that we have to announce, at the time of going to press, the death of Sir Hugh Lett, Bt., K.C.V.O., C.B.E., a past President of the College. A full tribute to Sir Hugh will be published in the September issue of the *Annals*.

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## THE BRITISH CLUB FOR SURGERY OF THE HAND

AN INSTRUCTIONAL COURSE on the Treatment of Injuries of the Hand will be held at the Royal College of Surgeons of England on Friday, 9th October 1964, from 2 to 6 p.m., under the chairmanship of John Barron, F.R.C.S.Ed. The programme will be as follows:

Introduction . . . . .	.. ..	T. J. S. PATTERSON, M.D., F.R.C.S.
Treatment of Tendon Injuries .. ..	.. ..	R. G. PULVERTAFT, F.R.C.S.
Treatment of Nerve Injuries .. ..	.. ..	D. M. BROOKS, F.R.C.S.I.
Treatment of Fractures and Joint Injuries .. ..	.. ..	G. R. FISK, F.R.C.S. Ed., F.R.C.S.
Treatment of Skin Loss .. ..	.. ..	I. F. K. MUIR, M.B.E., M.S., F.R.C.S.
Amputation .. ..	.. ..	H. G. STACK, F.R.C.S.
Rehabilitation of the Injured Hand .. ..	.. ..	C. B. WYNN PARRY, D.M., M.R.C.P., D.Phys.Med., W/C., R.A.F.

Application to attend this meeting (registration fee, including tea, £1) should be addressed to the Honorary Secretary of the Club, H. Graham Stack, Westhay, Mount Avenue, Hutton, Essex.