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URINARY DIVERSION IN CONGENITAL PARAPLEGIA

By D. F. ELLISON NASH, F.R.C.S.

TWENTY years ago we were faced with the most appalling cases of advanced uropathology in children who had been born during the war, and whose apparently hopeless condition hardly warranted priority treatment when bombs and rockets were falling on our cities. Perhaps the most important weapon that came into use was the miniature resectoscope, and this transformed the picture of retention both in boys and girls. Better radiological study and constant additions to our antibiotic armamentarium certainly improved the situation, and the introduction of the light-weight Chailey urinal alleviated the incontinence problem in the boys.

The first major publications on urinary diversion in this country arose from the 12th Annual Meeting of the British Association of Urological Surgeons in 1956. The mortality from ileal loop urinary diversion had been considerable, but the applications of the procedure both in adults and children had been widespread and few surgeons had extensive personal experience. Of the 10 cases in children that I reported, all had severe previous urinary damage with recurrent infection, some with stones (Nash, 1956).

One girl was fully rehabilitated and six years after operation, while she was in perfect health, was murdered and must thus be excluded from this series. Another died from electrolyte imbalance following gastro-enteritis on holiday during which she received no hospital treatment—that was three years after operation. One girl who had been uraemic to the point of coma was operated on at the age of 9 and died at the age of 16 in a mental hospital undoubtedly from progressive renal infection aided by mismanagement. There was only one boy in this first group and he had been extremely toxic and anaemic, but improved greatly after operation and survived five years, dying eventually with acute rheumatic carditis unconnected with his original troubles. The only patient who underwent operation for a rather rapid deteriorating infection and obstruction died at 3 months after operation; with one's present knowledge and the antibiotic cover which is available 10 years later, this operation would I think have been postponed. Yet one other died about a year after operation at the age of 14, from peritonitis following an enema. Of the eight girls who underwent operation (that is excluding the murdered one), four are alive 11 years after operation. One has produced two normal children after normal pregnancies. Thus four out of eight girls operated on 10 years ago or longer are socially acceptable, wage earning, having progressed through normal puberty and education.

In the very first number of *Paraplegia* in 1963 are recorded these words, 'Although the pattern of disease in paraplegics is changing, urinary infection with all its sequelae remains the major underlying cause of mortality in chronic paraplegia' (Tribe, 1963). If this was the state of affairs in acquired paraplegia, what of the prospects in the congenital form with all the attendant complications and the further passage of years?

Very little has yet been published on long-term follow-up of these children. Most of the recorded work on ileal loop diversion has been in adults with malignant disease. This presentation is based on 20 years' personal experience in the management of children with varying degrees of paraplegia from birth or infancy. The study has involved some 473 patients in whom the causative lesion was spina bifida or dysplasia in 448 (95 per cent. approx.), sacral agenesis in 16 (3 per cent. approx.) (fig. 1), tumour and infant myelitis 8, and Pott's disease 1.

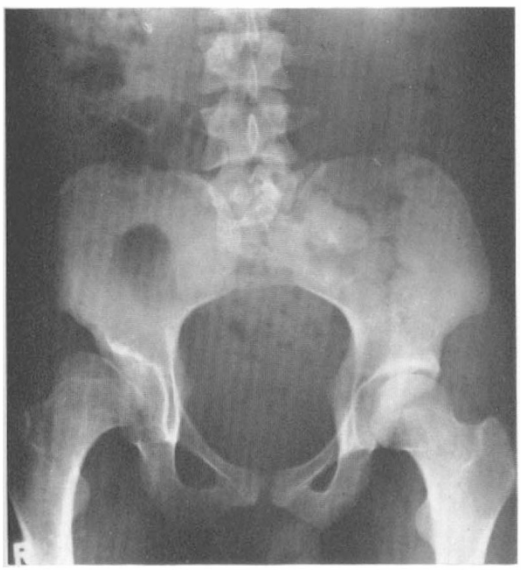

FIG. 1

Sacral agenesis. In this condition the peripheral nerves fail to develop and the pelvic floor is unsupported.

Special Problems of Congenital Paraplegia. There may be some who think that no effort should be made to preserve the life of a child born to suffer the limitations imposed by a myelocoele with or without hydrocephalus, and in order to dispel lack of interest in this particular topic I must place before you some very basic facts.

Some form of overt spina bifida with minor or major paralysis is found in approximately 1 in every 400 live births. With the present birthrate this means that approximately 2500 babies are born every year with spina bifida. It has been shown without doubt that delay in closing the myelocoele—an open wound of the spinal cord—decreases the likelihood of motor recovery where paralysis is already present, and very greatly increases the onset of paraplegia where none is seen at birth. The remarkable thing is that some of the worst cases will survive with little or no treatment, and on this account we must never assume that a particular baby is 'not worth doing'. I maintain that it is the surgeon, and not the obstetrician or paediatrician, who must make this decision, and this demands immediate access to informed surgical opinion. These babies cool very quickly and cannot respond to the normal drop in environmental temperature as does a normal baby. Immediate post-natal care, incubation and 'birthday' surgery gives a survival figure round 75 per cent. for the first year. The introduction of the Spitz-Holter valve, in spite of its complications, has made early surgery both necessary and safe.

The care of the urinary tract begins at birth. Not many babies are born with retention, but some already have damaged bladders and hydronephrosis.

The Distinctive Features. When we contrast this condition with that of acquired paraplegia we find ourselves faced with a crop of associated handicaps. Hydrocephalus is common, sometimes with mental defect—but by no means always; there is involvement of the cerebellum and brain-stem further affecting balance. With spina bifida there is frequently a primary abnormality in the urinary tract such as fused kidneys or unilateral agenesis, or duplication (figs. 2-4).

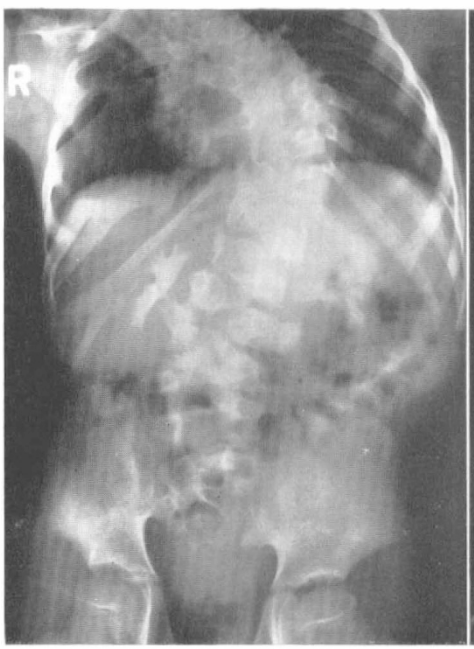

FIG. 2

Gross spinal and thoracic dysplasia makes the siting of a urinary stoma critical. This pyelogram shows a 'pancake' fused kidney to the right of the midline.

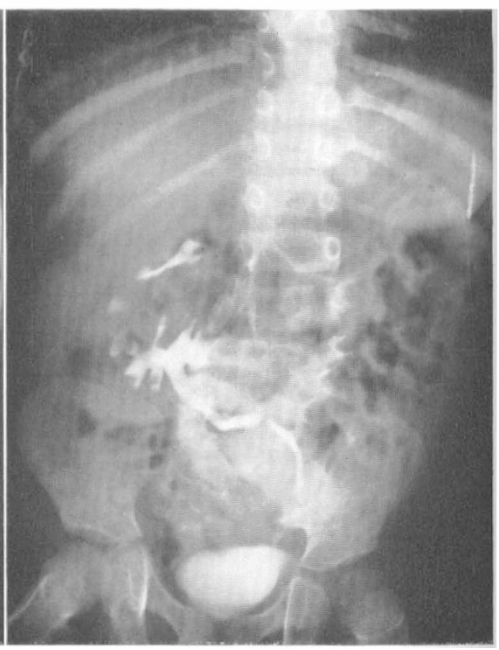

FIG. 3

Spinal dysplasia with crossed ectopia of kidney and fusion (pancake kidney).

(2) There is no norm of sensation in bladder or bowel, nor is there a straining mechanism. Since the child has no experience of these functions there is no language of communication in training the individual, or in explaining the situation.

(3) Because there is lifelong incontinence, there is no social experience related to the wet/dry antithesis. This does not matter much in the toddler stage but even at nursery-school age the child becomes conscious of her disability. The child herself cannot appreciate the import of the typical local authority report which reads, '. . . the school finds this child socially unacceptable', municipal politesse for '*she stinks*'.

(4) Because of pre-natal paralysis and perhaps post-natal neglect, deformity of varied degree is present and this affects mechanically the pelvic floor, the

problems of movement and accessibility to perineum or abdominal wall in surgery. The deformity may be so great as to exclude the possibility of constructing an ileal loop urinary diversion.

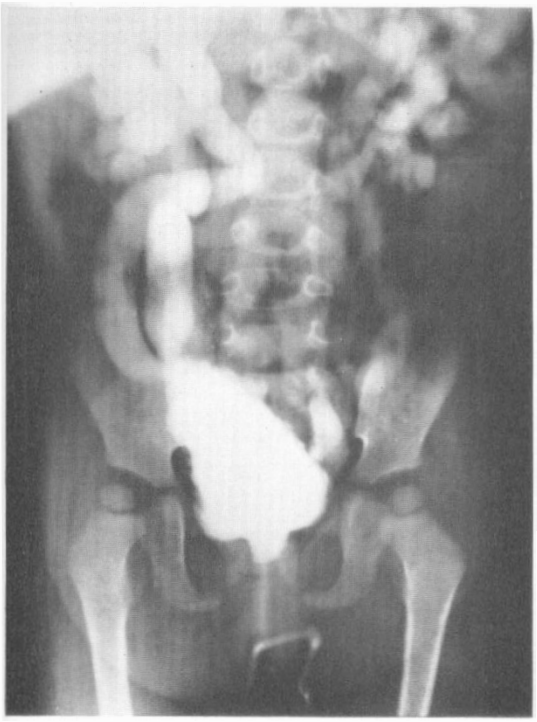

FIG. 4
Severe back pressure effects and primary renal abnormality—right-sided duplication.

(5) At present far too few children get adequate urological supervision from the day of birth. Some unfortunately still fall on the stony ground of the 'let them die' school of thought; some fall into the hands of skilful craftsmen, and while they are saved from immediate death, they succumb later when the unrecognised weeds of urinary infection spring up and choke them. Even in the best organised centres this programme of total management has not been going more than 10 years and therefore the really long-term results are unknown. Up to the moment when paraplegia strikes the adult the bladder muscle and innervation has been normal and the higher centres have been trained, and the urological problem is essentially one of adaptation and patient management.

(6) There is no physical perception related to sex, no arousal instinct, and consequently no physically based consciousness or anxiety related to incapacity. It is of course impossible for us who are normal to understand this situation. Puberty brings with it the problems of prolapse in the girl, and prostatic calcification in the boy.

Early Management of the Urinary Tract. It is vital to be constantly on the lookout for retention and to deal with it when it occurs. In my experience this can be done almost without exception by transurethral bladder-neck resection both in boys and girls. It very rarely has to be repeated in the girls and occasionally a fistula into the vault of the vagina develops: this is immaterial in view of the fact that incontinence was almost certainly going to occur anyway. It is perhaps worth mentioning in passing that in the group of children that are supervised adequately from the start of life, retention and back pressure seem far less common. Regular emptying by straining or manual expression in infancy undoubtedly diminishes the risk of major structural damage to the urinary tract.

Three methods of diversion are available.

(*a*) Ureterostomy.
(*b*) Suprapubic drainage of the bladder by catheter or vesicostomy.
(*c*) Ileal loop diversion—ileo-cutaneous ureterostomy.

There are occasional indications for ureterostomy, for instance in acute renal insufficiency with thick enlarged ureters (fig. 5).

There are no indications for suprapubic bladder drainage or vesicostomy as a definitive diversion. The stoma in vesicostomy is ill placed and the operation has no place whatever in dealing with the child's lesion (fig. 6).

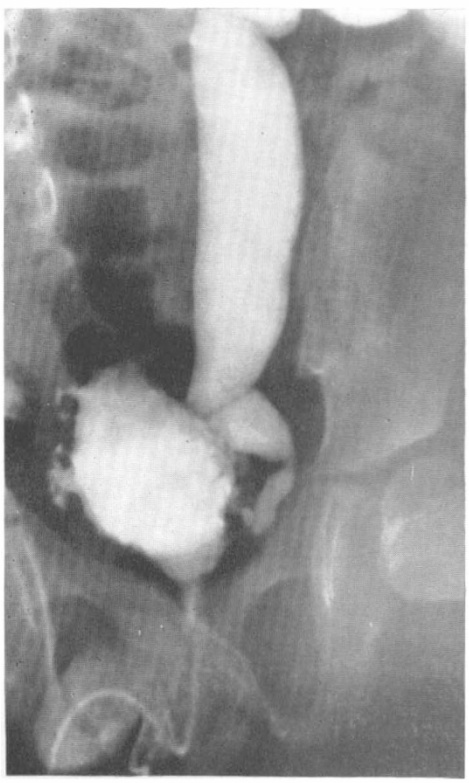

FIG. 5

Gross reflux from a diverticulated bladder. This is suitable for cutaneous ureterostomy drainage (bilateral) if infection cannot be controlled.

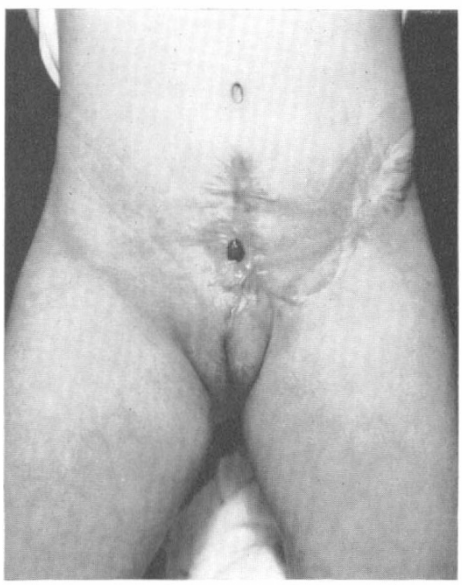

FIG. 6

The impossible situation produced by vesicostomy. The keloid irregular scarring prevents satisfactory attachment of a collecting appliance. This had to be excised and replaced by an ileocutaneous ureterostomy.

The principles of the ileal loop operation are well known. The loop is an active conduit and not in any sense a bladder. Electrolyte disturbances are unknown unless the spout is faulty or obstructed.

In considering an ideal method of diversion, these questions need to be answered:

1. Is it safe, (*a*) in short term? (*b*) in long term?
2. Does it meet the social, aesthetic and sexual requirements of the girl?
3. Does it enable the girl to be independent of hospital and nursing care?
4. Is it relatively cheap in the maintenance of appliances?

The answers to these four questions in relation to the ileo-cutaneous ureterostomy are:

1. *Is it safe?* The operative mortality is nil—although no one would pretend that it is a minor operation, or that there have been no near misses.

In long term, morbidity depends obviously on the extent of pre-operative renal damage. The revision rate is very low, and the long term renal hazard is probably unaffected by this particular method of diversion, but rather by pre-existing renal damage.

2. *Does it meet the social aesthetic and sexual requirements of the girl?* In relation to social needs, this method permits a full education curriculum limited only by the orthopaedic incapacity. It permits full engagement in social activity, dancing, camping and swimming, again limited of course by the orthopaedic incapacity.

It permits normal hygiene measures at puberty and removes urinary incontinence as a barrier to intercourse later.

It permits normal childbirth, though many of these girls have a small or contracted pelvis and Caesarian section might be necessary.

3. *Does it enable the girl to be independent of hospital and nursing care?* Provided the patient has two functioning arms and can reach her abdominal wall, she can achieve independence in relation to the voiding of urine.

4. *Is maintenance relatively inexpensive?* Apart from the provision of adhesive patches, and perhaps four belts and four bags in each year, there is virtually no maintenance.

Selection of Cases for Operation. *Age.* In some centres this operation has been undertaken in very young children but I can see absolutely no point in doing this for the paraplegic child. In my view the operation should not be undertaken as the *primary* treatment for lower urinary tract *obstruction* in the paraplegic. Such obstructions should be dealt with by transurethral resection in the girls in every case. I have not found it necessary to do open operation for Y-V plasty. From the point of view of managing the incontinence, a child under the age of 5 will not be able to cope with the apparatus herself, and such a child is perfectly acceptable in school in nappies. To those therefore having the care of the child there is no advantage in early operation. Over the age of 5 the situation is quite different. Even the backward child begins to be conscious of the disability of incontinence and with increasing weight, and in the ambulant with increasing activity, the constant wetness adds to skin infection and limits the quality of life. Any children can learn at the age of 6 or 7 to deal with the appliance completely by themselves and this seems to me the time to undertake the procedure. Although the operative mortality in my hands has been nil, I do not feel I can ever pretend to parents that this is anything less than a major operation. Only twice have I been faced with any post-operative revulsion against the spout, but I believe this

general acceptance is because I have waited until both the parents and the child were convinced that this operation was necessary. In many cases I have adopted the procedure of acquainting the child and the parents with the appearance of the spout and the problems of management by introducing them to another child. In our special school at Coney Hill the girls know all about it when the time comes for the operation to be undertaken, and they bring pressure to bear on me to speed the day when they will be dry. In contrast to the healthy adolescent or adult struck down by paraplegia, these children have never been able to worship their twisted body form, and therefore the acquisition of a 'stoma' does not produce the same aesthetic trauma in the congenital paraplegic, as it might with the young girl who had been normal in adolescence.

I think also that the complication rate, particularly due to recession of the spout, is likely to be higher in the younger children owing to the alteration of growth.

The timing of the operation must be decided in relation to the girl's total management and if any orthopaedic operations of a major nature are contemplated such as a psoas transplant, or hip osteotomies, the ileal loop diversion should be deferred as it is very much easier to deal with such children during their orthopaedic care by the use of an indwelling Foley catheter.

Operative Management. There are certain points which have direct bearing on the preparation and treatment of the child while under the care of whoever is supervising the total paraplegic management.

(1) Obstruction must be dealt with by adequate bladder neck resection, or by a long period of indwelling catheter drainage, following the same principles as the old-fashioned two-stage management of prostatic obstruction. Anaemia must be treated.

(2) The large bowel must be adeqately evacuated as there is very little doubt that chronic constipation of any degree adds very materially to the difficulties of the post-operative course. I insist on the use of intramuscular neostigmin and enemas for several days before operation as practically all these children are overloaded.

(3) In the post-operative management undoubtedly a large fluid throughput is the most important factor especially if there has been previous renal damage. I have only once seen electrolyte imbalance arising from the loop and this was when there was obstruction at the outlet. The ileal segment is in no sense a bladder. It is an active conduit from which there is no significant absorption of electrolytes and urea.

(4) Bacteriology of the urine emerging from the loop is apt to be misleading. If the general health of the child is good, one can be guided by the appearance of the urine rather than by laboratory reports. If there is any clinical suggestion of pyelitis then quantitative bacteriology is helpful and maintenance antibiotics are used.

(5) *Revision of Spout.* As with any form of ileostomy, stenosis in the abdominal wall, prolapse or elongation of the spout are occasional complications.

Although at the original construction the blood supply of the turned back mucosal cuff is coming up the mesenteric vessels, once the spout has become

established the outer layer gets sufficient blood supply from the skin to enable one to amputate a redundant segment by simple transection and over-sewing of the cut layers.

(6) Unfortunately some operators have left the children with minute spouts or skin flush buttons. These invariably contract and become stenosed. The spout should be at least 2 in. long, and the mucus which is produced by the outer layer acts as a protective for the skin, limiting urinary excoriation. There is far less trouble with leakage from the appliance if the spout is long enough to hang over the lip of the flange (fig. 7).

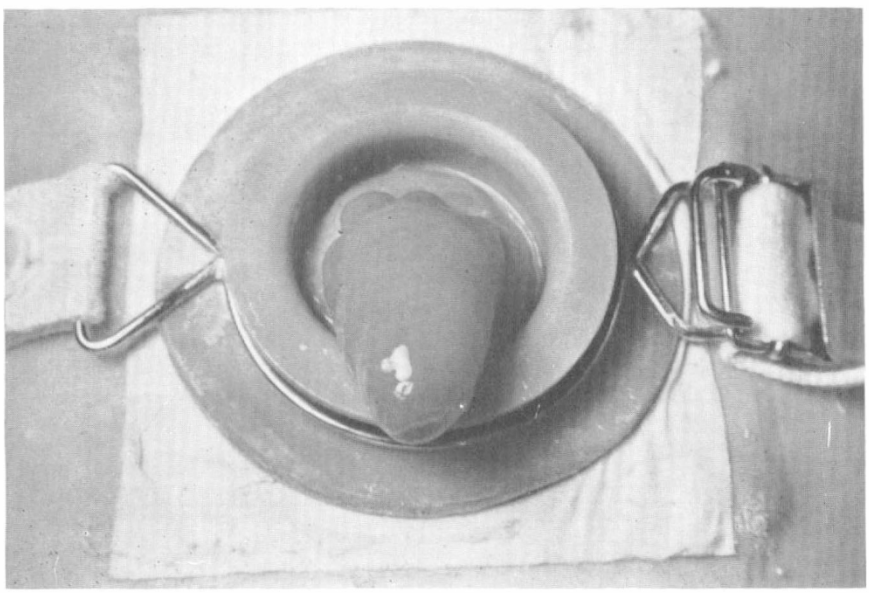

FIG. 7

Urinary ileostomy. The 'spout' should be long enough to hang over the flange.

Bleeding occasionally occurs from the spout following trauma, but the stoma must be placed at the original operation at a site which will not interfere with the wearing of orthopaedic appliances such as braces. A St. Mark's type flange with a 'canopy' is an additional protection.

Subsequent Investigations. Excretion pyelography is performed three months after operation and if dilatation is found where it was not present before, the pyelogram should be repeated at a further three-month interval. It is quite common to find post-operative upper-tract dilatation which persists for three or four months but settles down to normal.

During the first year blood urea and haemoglobin estimation are required every three months, or at more frequent intervals if abnormalities are found. A catheter should be passed into the natural bladder at the end of the first post-operative week, after one month, and at routine three-monthly follow-up visits

if it is found that there is any accumulation of mucus or pus. In spite of adequate pre-operative drainage (by transurethral resection) some girls have required post-operative transurethral resection to provide adequate drainage and prevent suppuration in the residual bladder. One of these was the patient who married, and the accumulation occurred after the first natural childbirth.

Personal Series. From a personal series of 473 cases of congenital paraplegia 70 have so far been subjected to ileal loop urinary diversion at various ages and stages of their pathology, some after years of neglect. The operative mortality has been nil. This relatively small proportion of the total is not an accurate picture of the numerical need for diversion. Many of the 473 have not yet reached the required age. Some have died of intercurrent disease, or of problems related to hydrocephalus.

TABLE I

Age at Operation

5-10 years	37
11-15 years	27
16 years and over	6
Total	70

Exactly 50 girls have been followed up, or could have been for four years or more. There were nine deaths, one by murder, one from peritonitis following enema. *There was thus a fallout rate of seven* (14 *per cent.*) *due to renal disease.*

TABLE II

The follow-up periods for survivors have been:

10 years	6
9 years	7
8 years	6
7 years	5
6 years	6
5 years	4
4 years	7
Total	41

TABLE III

The fall-out rates have been:

Deaths	1st year	3
Deaths	3rd year	1
Deaths	4th year	2
Deaths	10th year	1
		7 Renal
		2 Unconnected
Total		9

Nineteen (38 per cent.) had transurethral bladder neck resections before diversion (including five of the seven who died of renal disease, and this is an indication of the extent of pre-operative renal insufficiency).

Stoma revision was required in two (4 per cent.), one shortening, one stenosis (two others required dilatation).

Four patients developed stones (8 per cent.).

One kidney has had to be removed due to obstruction (1 per cent of kidneys at risk).

Summary

The distinctive problems of congenital paraplegia are compared with those of the acquired form, particularly with regard to associated handicaps, renal immaturity, lack of perceptual experience and social deprivation in early life.

Reference is made to the management of retention of urine and to the changing urological picture as age advances.

The satisfactory management of male incontinence is discussed.

Against this background are enumerated the requirements of the ideal urinary diversion. The isolated ileal loop operation, or ileocutaneous ureterostomy is discussed. A series of 70 patients is surveyed and the criteria for operation are presented in relation to pre-operative management, morbidity and mortality.

Long-term follow-up data are given concerning 50 consecutive girl patients operated on between 1955 and 1963 selected for ileal loop diversion from 473 paraplegic children under observation since 1946. The operative mortality was nil. The subsequent fallout rate due to death from the paraplegia or related conditions has been seven (14 per cent.), three in the first 4 months, one in the 3rd year, two in the 4th year and one in the 10th year.

Ileal loop urinary diversion is claimed to be the most satisfactory method of dealing with incontinence arising from congenital paraplegia in the female, and should not be undertaken before the age of five.

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URINARY DIVERSION IN PARAPLEGIA

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The prime objective in the urological management of spinal cord injuries is to preserve renal function by ensuring an unobstructed drainage of the entire urinary tract. Failure to do this constitutes an important cause of morbidity and mortality. Despite the tremendous progress made in the treatment of particularly traumatic lesions of the spinal cord, renal failure and uraemia still foreshortens many of these lives.