Studies in Paediatric Urology

A Thesis submitted through the Department of Paediatrics, in the Faculty of Medicine, for the Degree of Master of Medical Science in the University of Adelaide

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Contents

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Brigid Dewan has given an enormous amount of time, effort and compassion, which has enabled me to complete this work; my children, Emily, Rachael and Simon have also made a significant contribution.

Declaration

This manuscript contains no material which has been accepted for any other degree in any University. To the best of my knowledge and belief, this manuscript contains no material previously published or written by any other person, except where due reference is given in the text. I give my consent for this copy of my thesis, when deposited in the University library, being available for loan and photocopying.

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Introduction

Paediatric urology is a developing subspecialty, the future of which is dependent on the furthering of an evidence based medicine approach to improving the standard of care for children with genitourinary tract disease. This document is a collection of laboratory and clinical studies in Paediatric Urology, including prospective and retrospective patient series, case reports of rare conditions; also, technical innovations are documented and reviewed in light of international experience.

The first of the studies presented has systematically looked at the *ex vivo*, radiological and histological anatomy of multicystic kidneys; the findings raise the prospect that the cystic changes are due to a combination of ischaemic and obstructive events.

The sheep laboratory study was an attempt to develop a neurogenic bladder model for use in experiments in bladder augmentation research: this study highlights the appropriate use of the laboratory to improve the interpretation of the clinical results of bladder transection as, from the results obtained, the previous enthusiasm for a trigonal division appeared to be based on poor evidence.

Bladder exstrophy is a rare condition of which relatively little has been published: understanding of the condition is enhanced by the provided summary of the developments over the centuries. As a result of previous research and lateral thinking about the management of bladder exstrophy, the transposition of the umbilicus during the neonatal bladder closure has become an appropriate improvement in the finesse of the surgery. Thus, the subsequent section of this volume is the presentation of the details of an omphaloplasty technique.

The next group of reports presents abnormalities of the penis, including phimosis. The discussion highlights the need for a clearer definition of phimosis and provides the results and commentary on a series of patients who were treated with steroid cream application: the mechanism of action of the non-operative therapy is also presented and discussed.

Priapism and diphallus are two rare conditions of the penis encountered in Paediatric Urology; both are appropriately discussed as case reports with a detailed review of the accumulated literature: six cases of priapism and two cases of diphallus are presented and their management discussed.

Pelviureteric junction obstruction is common, but preoperative identification of a lower pole vessel as the cause of the obstruction is only infrequently achieved. The usual finding of a kink and stenosis was thought more likely in the neonatal patient, but this had not been confirmed prior to this study. It is possible that the use of doppler ultrasound studies may help show which patients are likely to undergo further deterioration after a prenatal diagnosis of hydronephrosis, support for which comes from a finding that older patients are more likely to have a lower pole vessel as the cause of the PUJ obstruction than younger children.

A second study on pelviureteric junction obstruction is also presented: this study is of the different surgical approach required for the management of PUJ obstruction associated with a horseshoe kidney. There is little in the literature on how to appropriately manage these cases, and the indications for ureterocalycostomy appear to be poorly understood. Four patients, who underwent ureterocalycostomy were were studied following failed surgery. The need to consider anastomosis of the ureter to the lower pole calyx at the first operation is indicated by the findings in this group of patients.

Urolithiasis in children is known to be uncommon in developed countries, and uric acid calculi are a recognised feature of the Australian aboriginal children who presented for surgical management of urinary tract calculi in South Australia over a five year period: the lessons learnt are also discussed.

The disease of megacystis microcolon intestinal hypoperistalsis syndrome is presented as a case report which is augmented by a detailed analysis of the literature on this uncommon condition. The case presented also provides additional features of this syndrome.

The last chapter is of a technical innovation that uses renal vein cannulation for haemodialysis in renal failure patients, again highlighting an advance achieved by inventive thinking.

The compilation of these studies has allowed the author to review and formulate the arguments for what are hopefully a number of significant contributions to the improvement of paediatric urology standards.

Laboratory Studies in Paediatric Urology

A Study of the Radiological Anatomy of the Multicystic Kidney

Introduction

The typical "Bunch of Grapes" appearance of the multicystic kidney (MCK), first described by Cruveilhier in 1836 [1], was further studied by Schwartz [2] in 1936. Despite this long history, the pathogenesis of the MCK has still not been resolved and there is often confusion between cystic renal dysplasia and the MCK, the latter being differentiated by the presence of ureteric atresia or stenosis. The Potter classification adds to this confusion by having a case of urethral obstruction illustrated as a Type IIB kidney, the group that most closely resembles the MCK [3]. The coexistence of the ureteric and renal abnormalities suggests an obstructive aetiology [4,5], particularly as there is commonly a pelviureteric junction abnormality on the contralateral side [6]. However, the ureter is not always completely obstructed, and the upper ureter and pelvis can be dilated above a more distal obstruction [4,7].

A group of patients was studied to look at the prenatal changes, the preoperative function, the gross anatomy, and the radiological communications between the cysts and the ureter.

Materials and Methods

Eleven children were found to have an abnormal kidney by prenatal ultrasound (US) which was confirmed postnatally. All infants had a micturition cystourethrogram (MCU) and a MAG3 nuclear medicine scan, and each child had more than one postnatal US. The kidneys were removed for one or more of the following reasons: their large size (three), the presence of radionuclide activity on MAG3 scan (six), the need for other surgical intervention (two), or persistence of a significant amount of cystic tissue at 12 months of age (four). None of the patients had hypertension.

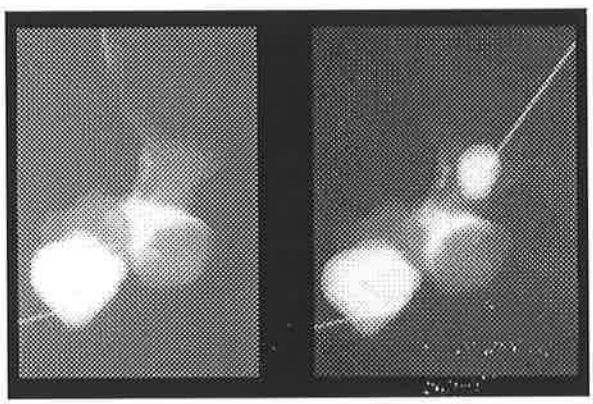
A nephrectomy was performed in each child and, if the ureter was not immediately evident, the retroperitoneal region at the pelvic brim was explored. After removal, the fresh specimen was immediately injected with water soluble contrast, firstly through the ureter if present, then into the region of the renal pelvis and, lastly, directly into the larger cysts. A number of radiographs of the injected kidneys were taken with increasing injection volumes. The volume of contrast and the number of injections varied with the size of the kidney and the degree of intercommunication of the cysts; the larger the kidney the greater the final number of radiographs and the larger the injection volume. In three cases a tubular structure connected the pelvis to a cyst and in two of these a 5 FG feeding tube was able to passed into the abnormal pelvis, through the connecting tubular structure into a lower polar cyst.

At the conclusion of the radiological studies the specimen was examined macroscopically for solid areas and note was taken of the position of these in relation to any function seen on the MAG3 study. The kidney was then placed in formalin to enable subsequent histological examination.

Results

Three kidneys were found to be hydronephrotic or normal at less than 20 weeks antenatal ultrasound, and the remainder were found to be abnormal on the first scan. Where sequential scans were performed the parenchyma was seen to become increasingly replaced with variable sized cysts; the cysts then increased in size and eventually the cystic renal mass decreased in volume. A postnatal nuclear medicine scan showed minimal but definite function in six kidneys. On MCU, one patient had vesicoureteric reflux (VUR), one had a contralateral pelviureteric junction (PUJ) obstruction and one had an ipsilateral ureterocele; the others were normal.

The 11 post-excision radiographic studies are shown in Figures 1-11. At the time of excision the ureter was absent in one patient (Fig. 10), the upper ureter and pelvis dilated in one (Fig. 7), two ureters were seen to be patent on the contrast study (Figs. 8+9) and one had a minute lumen seen histologically. One kidney had no communications between the cysts (Fig. 1) and the remainder had a variable degree of free flow as seen from Figures 7-16. Two had small tubular structures between some of the cysts (Figs. 2+3), and three had a large tubular structure coursing around the cysts, two of which allowed the insertion of a 5 FG catheter into the lumen of the cyst (Figs. 10+11). These interconnecting tubes had a urothelial-lining and a smooth muscle wall. The macroscopically solid areas correlated with function seen in the pre-operative nuclear medicine scans of the six affected kidneys that also showed concordant regions of dysplastic renal tissue, histologically. In the remaining kidneys, and other areas of the six with function on the nuclear medicine scan, no function, nor histologically renal elements, were recorded.



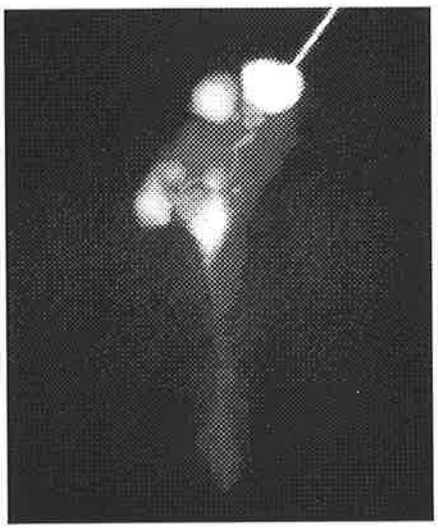
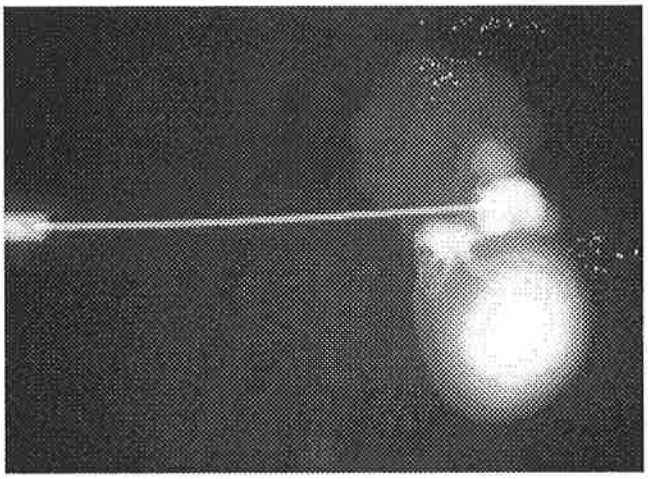


Figure 1 (left): Injection of the pelvic region failed to fill either the pelvis or any cysts. Filling of the upper and lower pole cysts was achieved by two separate injections. **Figure 2 (right):** Two separate sites of injection have resulted in the demonstration of small tubular connections between cysts.



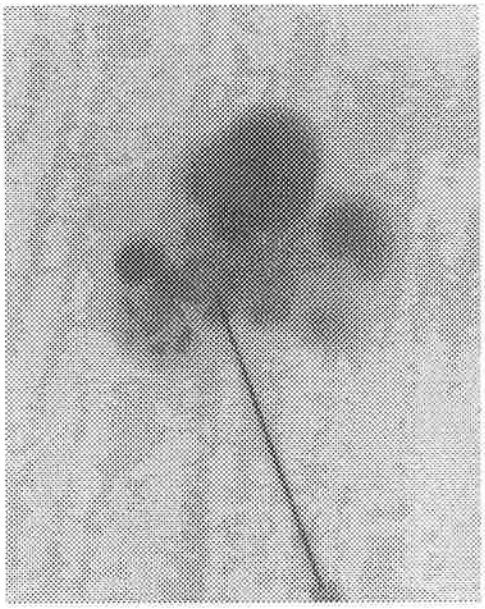
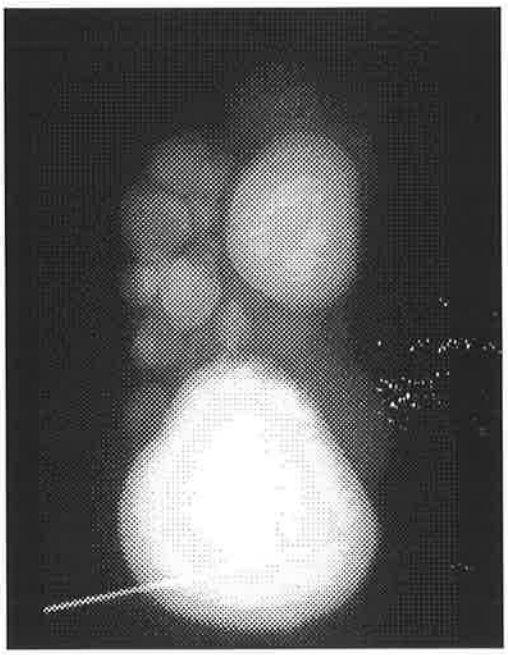


Figure 3 (left): Injection into the hilar region shows a narrow, short tubular connection with the larger lower pole cyst. **Figure 4 (right):** Free flow of contrast from an atretic pelvis into a large number of small cysts is seen.



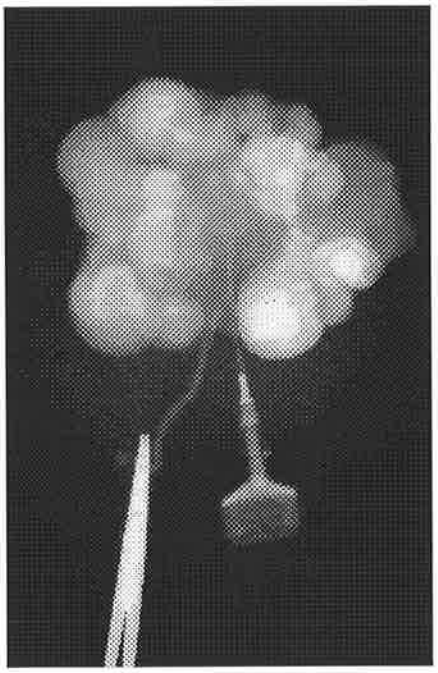
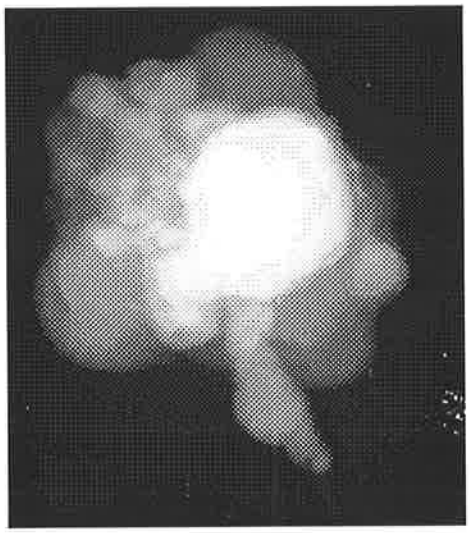


Figure 5 (left): A single injection into the lower pole filled one large and several small cysts throughout the kidney. **Figure 6 (right):** The abnormal renal pelvis was in free communication with all the cysts, but no tubular connections were seen.



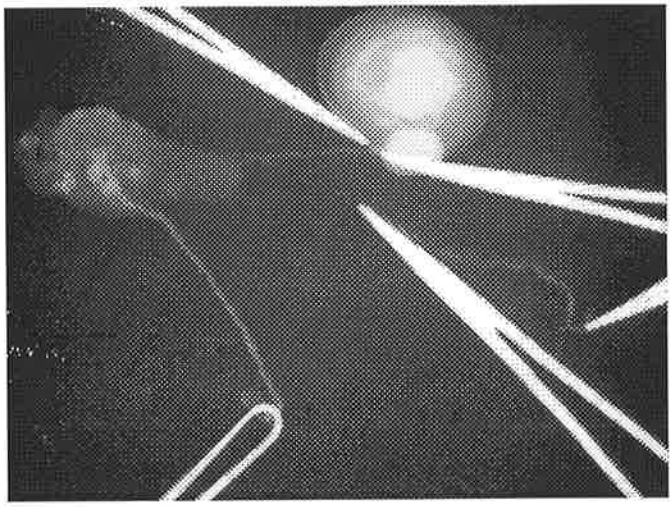
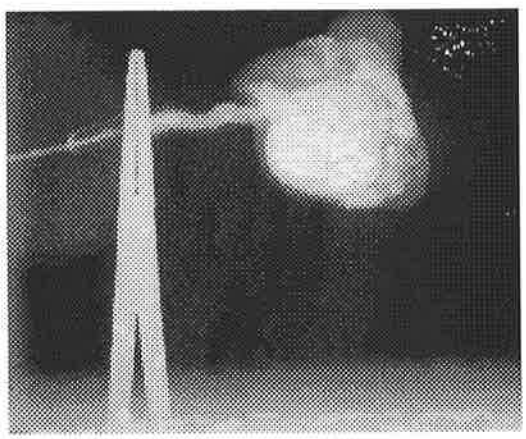


Figure 7 (left): The upper ureter and pelvis are both dilated, and communicate with all the cysts as demonstrated by a single injection into the renal pelvis. **Figure 8 (right):** Both segments of a duplex kidney are multicystic; the upper pole has ureteric continuity with the renal cysts, the lower pole cyst has a tubular communication to the pelvis.



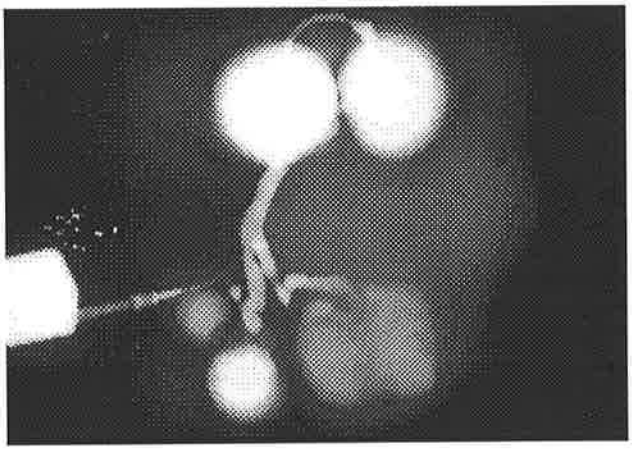


Figure 9 (left): Injection into an stenotic ureter, from the same side as a ureterocele, showed a patent ureter with filling of the kidney. **Figure 10 (right):** An extensive tubular network on the medial side of the cystic mass can be seen in spite of the ureteric atresia. The communication with the upper pole cyst could be intubated with a 5 FG feeding tube.

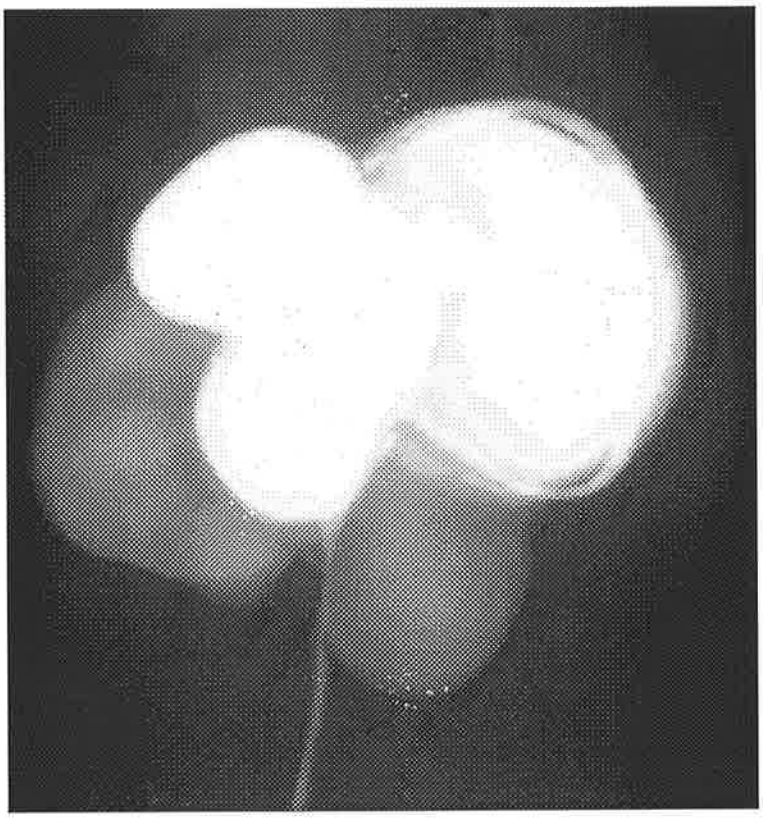


Figure 11: A long tubular structure is seen coursing around the lower pole cyst that is filled with contrast instilled into a rudimentary renal pelvis. The ureter is also seen to be minimally patent.

Discussion

Despite the MCK being the commonest cause of a renal mass in the newborn [8], and the first description occurring as early as 1836 [1], the embryology has not yet been elucidated. We have come to further understand the nature of this condition since prenatal US has been more widely used, enabling the observation that the MCK can develop from a minimally hydronephrotic kidney, which then may increase in size with an increase in cyst size and numbers, and eventually involute completely, even prior to birth [9,10]. Confusion occurs, however, because kidneys with multiple cysts can develop from different mechanisms. Nevertheless it would seem that there is a relatively homogeneous group that have the appearance of a "Bunch of Grapes" and ureteric atresia or some degree of stenosis. Previously it was felt that non-function [11,12] and non-communication of the cysts [7,13] were characteristics required for the diagnosis. This is now refuted by the above observations, and those of others [14], with both function in MCK's (six of 11) and communication between the cysts recorded (10 of 11) (Figs. 2-11) [15-18]. The *in vivo* radiographic studies of Saxton *et al.* [16] and Kullendorff *et al.* [17] and microdissections of Madewell *et al.* [19] also demonstrated the interconnecting tubules seen in a number of the above cases, but they did not mention the smooth muscle containing tubular structures coursing around large cysts which communicate with the lumen of one of the cysts (Figs. 9-11). However, these tubular structures were demonstrated in Potter's microdissections, but not associated with the MCK entity [3].

The communication between the cysts indicates that there has been a connection between the ureteric bud and the renal blastema, precluding a primary defect of the ureteric bud. A purely vascular accident mechanism has been proposed; this is supported by the presence of a rudimentary vascular pedicle in virtually all cases, [20,21] and the common association with crossed renal ectopia [22,23], duplex systems [24] and Horseshoe kidneys [6,25], where variations of the renal vessel anatomy are always present. However, a small kidney,

for whatever reason, usually has a small vascular pedicle, and ischaemia of the renal blastema alone does not explain all the features of the MCK.

The change from a minimally hydronephrotic kidney *in utero* to an MCK has been observed by Avni *et al.* [10]. Their finding suggests that a vascular accident, similar to that in small and large bowel atresia [26,27], may be the initiating event which leads to variable ischaemia of the ureter, pelvis and renal parenchyma, with the subsequent development of a cystic response of the kidney. Such a sequence would explain the morphological difference of the MCK from other cystic renal dysplasia. Recent animal studies provide some support to this theory; the angiotensin 2 receptor deficient mouse has an increased incidence of both the MCK and pelviureteric junction obstruction. Fetal studies of the model would suggest that the anomalies are due to a failure of apoptosis around the ureteric bud and renal blastema - the link may be the inability of blood vessels to grow through the 'rind' of cells at the pelviureteric junction; thus ischaemia plus obstruction occurs [28].

The degree of ureteric stenosis appears more variable than previously understood. Whereas a fibrous cord connecting an atretic pelvis to a normal lower ureter is the most frequent finding, the otherwise typical MCK can be seen with a dilated pelvis and upper ureter (Fig. 5) [4,15], a radiographically patent ureter [7,29] or complete absence of the ureter as shown in Figure 10 [7], findings that are not consistent with simple obstruction nor only a renal parenchymal vascular insult. If the aetiology is obstruction alone one would expect complete occlusion in all cases. Furthermore, complete ureteric occlusion in sheep produces a hydronephrotic kidney if performed late in gestation, but cystic dysplasia if the obstruction is early. However, the "Bunch of Grapes" of the MCK is not seen with obstruction alone [30].

A Study of the Effect of Supratrigonal Division of the Sheep Bladder

Introduction

The neuropathic bladder in children can usually be managed by conservative means, using anticholinergic medication and clean intermittent catheterisation (CIC). In a small number of cases, augmentation is indicated in the patient with a small capacity, high-pressure bladder in order to achieve continence and prevent renal damage. In recent years, there have been many studies aimed at developing new methods of bladder enlargement, particularly aiming to achieve a urothelial lined reservoir [31-40]. The literature reports bladder augmentation studies in a number of different species, including the rat, rabbit, dog, calf, pig and sheep, but only in Mau's study [41] was there an attempt to create a neuropathic bladder prior to the augmentation procedure. However, Mau's method produced a more widespread neurological insult. Sethia [42] produced unstable contractions in the bladder of the minipig, by a circumferential supratrigonal incision, whereas Gonzalez *et al.* produced an abnormal canine bladder by removal of its serosal surface [40]. Alternatively, Staskin *et al.* [43] found a decrease in the bladder capacity following bladder transection in the dog, and Choudhury and Mittra [44] divided the posterior portion of the supratrigonal bladder in dogs, producing a higher volume, lower pressure bladder, with essentially normal contractions.

As bladder augmentation procedures have been developed for the treatment of the functionally abnormal bladder, it was considered important to develop an animal model for the neuropathic bladder in which further study of bladder enlargement techniques could be undertaken. Thus, the current study was designed to assess the effect of a supratrigonal incision on the urodynamic function of the sheep bladder. A literature review of the clinical outcome for bladder transection was also undertaken.

Materials and Methods

After obtaining ethical approval, 11 male sheep aged five to seven months were anaesthetised using 20mg/kg thiopentone and maintained with a halothane, oxygen and nitrous oxide mixture via an endotracheal tube. One litre of 0.9% saline was administered via an intravenous catheter. A double lumen suprapubic catheter was inserted into the bladder, as described previously [45], and a urodynamic study performed with a fill rate of 30 ml/min of 0.9% saline (Fig. 12). The data were collected on specific purpose software, a hard copy produced and the information stored for later study. Following the urodynamic study, the bladder was exposed via a midline incision and a complete bladder transection, above the level of the ureteric orifices, was performed. Care was taken to ensure preservation of the vascularity by dividing only the muscle and mucosa of the bladder. A catheter was inserted through the dome of the bladder, which was closed in a single layer (Figs. 13-15).

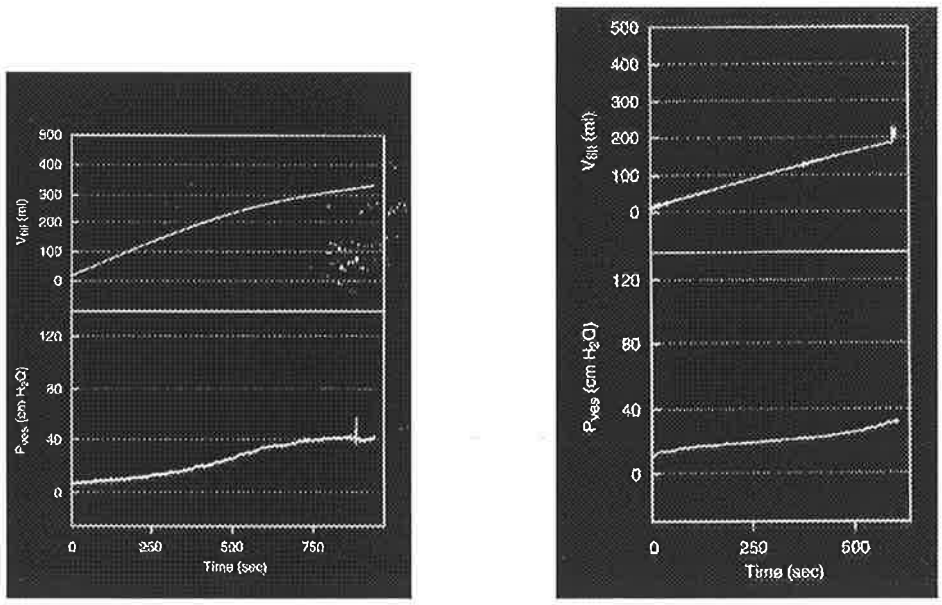
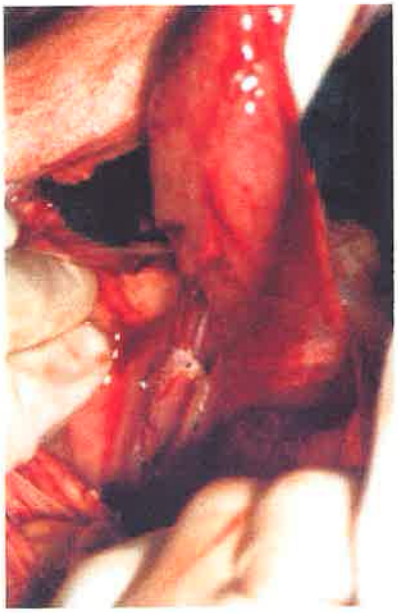


Figure 12: A urodynamic study (A) before and (B) six months after the transection of the bladder neck.



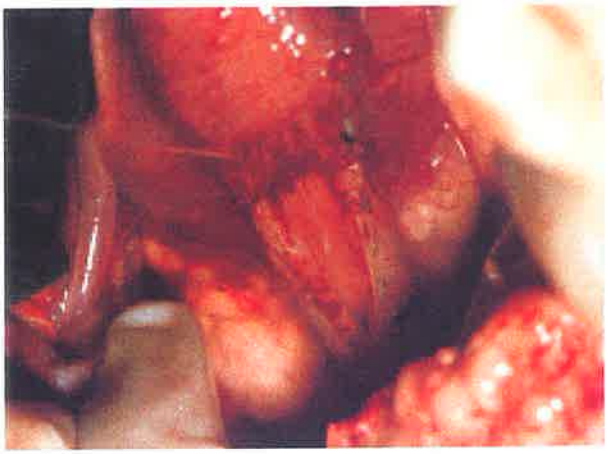
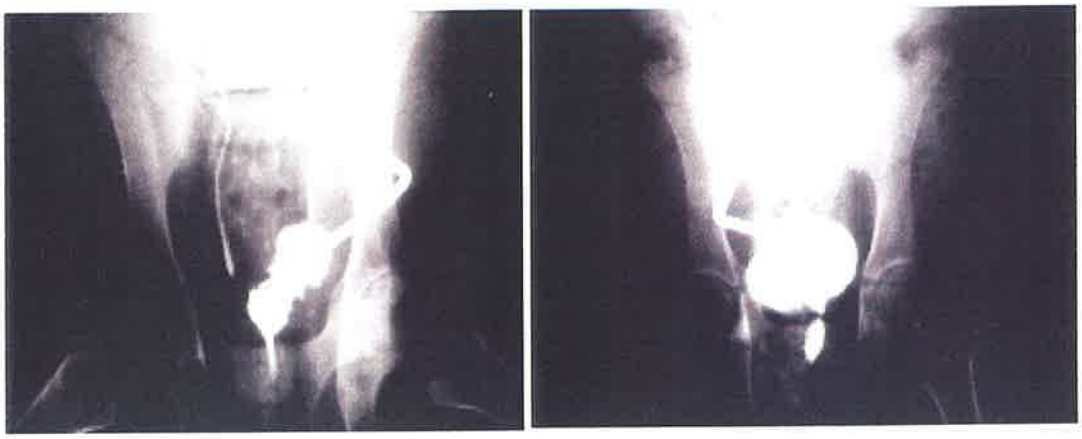


Figure 13 (left): The supratrigonal incision of the sheep bladder is commenced with the transverse incision of the anterior bladder neck, then completing transection, as shown. **Figure 14 (right):** The transection of the bladder has been completed and the closure is partly completed.



Figures 15 (left): A cystogram of a small contracted bladder with a poor urodynamic outcome; taken ten days after the transection. **Figures 16 (right):** A cystogram, taken 10 days after the bladder transection, of a bladder with a good urodynamic outcome. The difference between the two may have been due to ischaemia.

Ten days after the formation of the 'neurogenic bladder', a cystogram was performed and the catheter removed (Fig. 15 + 16). One of the sheep died at two weeks from an operative complication.

Six months after the supratrigonal transection, a further urodynamic assessment of the bladder was performed under a similar light general anaesthetic (Fig. 12). Following the urodynamic study, each animal was sacrificed with a lethal dose of barbiturate, and the bladder was removed for examination.

Urodynamic studies from six sheep aged 12 months old, under light general anaesthetic, were used as a control for comparison (Fig. 12).

The non-parametric Mann-Whitney μ test (also known as the Wilcoxon Rank Sum test) was used to analyse the urodynamic data. This test ranks the data, rather than making an assessment related to its size. Thus, it can account for a small sample size with an unknown distribution.

Results

The bladder volumes at the point of leaking during filling, for the study animals and the control group, are given in Tables 1 and 2. The median volume for the transection group increased significantly from 86 mls at six months to 245 mls at 12 months. At 12 months, the median volume for the control group was 165 mls. There was no significant difference between the transection and control groups at 12 months (p=0.18). One of the transected animals had a decreased bladder volume, which appeared to be due to ischaemia at the time of the transection. All other animals had an increase in bladder volume and no unstable contractions were seen in any of the urodynamic studies.

Sheep No.	*Volume at Leak Pressure (ml)*	
	Pre-Op.	*Post-Op.*
1	109	199
2	132	60
3	88	211
4	84	339
5	37	148
6	61	222
7	36	276
8	58	427
9	123	314
10	107	268
Median	**86**	**245**

Table 1: Bladder volumes of the study group animals, immediately before ***(Pre-op. group)***, and six months after the supratrigonal transection ***(Post-op. group)***.

Sheep No.	*Volume at leak pressure (ml)*
1	143
2	169
3	244
4	105
5	160
6	304
Median	**165**

Table 2: Bladder volumes of six control-group animals age 12-14 months.

Discussion

Animal Studies

The current study has shown that, in the sheep, supratrigonal transection does not produce either unstable bladder contractions, or a significant change in bladder volume. These data contrast with the findings of Sethia *et al.*, who used a supratrigonal transection of the Minipig bladder, finding bladder instability but no difference in compliance, when compared with the control animals [42]. Histologically, they did not find any increase in the density of the cholinergic nerve fibres, but recorded increased sensitivity to carbachol, potassium and acetylcholine in the muscular strips from the transected bladders versus the control bladders. Staskin *et al.* found supersensitivity to bethanocol following bladder transection with an increase in the density of cholinergic receptors in dogs and rabbits [43]. Unlike the above findings, they found a decrease in the bladder capacity. This may relate to the relatively early sacrifice of their animals at 120 days [43].

Choudhury and Mittra [44] also found altered bladder function in a dog model after posterior, supratrigonal transection-denervation of the bladder; their results, however, differ from those of Sethia *et al.* in that their technique resulted in an enlarged bladder at four weeks, but without unstable contractions [44]. Interestingly, Sibley [46] found changes in the obstructed pig bladder consistent with partial denervation, with increased sensitivity to acetylcholine and potassium, but decreased responsiveness to nerve stimulation of muscle strips. These changes are similar to those found by others after surgical denervation of the bladder of rats, cats and dogs.

Overall, the animal studies have had varied results which may relate to species differences and subtle differences in the operative techniques. One must therefore conclude that a highly predictable result is not possible. This would fit with the currently available literature on the clinical use of bladder transection.

Clinical use of Bladder Transection

While supratrigonal bladder transection has been used in the normal animal bladder, in an attempt to create the features of a neuropathic bladder, it has also been reported as a surgical method for denervation, to treat patients with unstable bladder contractions.

Essenhigh and Yeates [47] reported the use of open supratrigonal transection in 15 adults with bladder instability, but who had a satisfactory bladder volume and pressure under anaesthetic. Their cases were selected without detailed cystometric investigation and seven were followed for less than one year and 13 for less than two years. Improvement in day symptoms was reported as 'excellent' or 'very good' in 11 patients, 'good' in three and 'improved' in one. Essenhigh [48] and co-authors subsequently reported on 50 patients undergoing an endoscopic bladder transection, in which the procedure was deemed successful in only 16%. Bladder instability was demonstrated in all patients before operation and 93% post-operatively. Lucas and Thomas [49] had similarly bad results using an endoscopic approach, whereas Parsons *et al.* [50] had better results, with an excellent outcome in 57% of 30 patients and no change in 18%; despite these results they recommended further use of the technique.

In contrast to the animal study results, Turner-Warwick and Ashken (1967) have described 'supratrigonal denervation' for a hypersensitive bladder [51]. They used the term 'cystoplasty' for this procedure, which was performed on a 58-year-old woman with reduction of her bladder spasms. Gibbon *et al.* [52] performed a supratrigonal transection in seven patients with bladder instability but a normal bladder capacity, curing two completely and improving the other five. The bladder instability status of the group, however, was not discussed in detail and they did not divide the inferior vesical vessels as had Turner-Warwick and Asken [51].

Janknegt *et al.* [53] used the open technique of Essenhigh and Yeates in seven patients, with improvement in two and good results in five. Bladder instability was present in one with a good result and one with improvement. Six had night wetting only, all of whom had a stable bladder pre- and post-operatively. A further study published in 1995, suggested results that were encouraging for 12 patients who had an open operation which involved leaving a bridge of tissue above the left ureteric orifice. However, only three were cured, seven were improved and two had not changed.

Overall it would seem that supratrigonal transection has poorly predictable clinical and animal results; thus it probably has limited application in clinical management of bladder dysfunction in humans and the formation of a neurogenic bladder in the animal model.

Case Commentary

Despite the lack of validation of bladder transection in the management of the neurogenic bladder, the reported success in some cases prompted has lead to the use of the procedure in a difficult case of bladder spasm in a boy with cerebral palsy.

Operative Procedure

The patient's abdomen was opened via a previous suprapubic vesicostomy-formation incision, located caudal to the vesicostomy; the deep fascia was incised longitudinally and the bladder mobilised from the anterior abdominal wall. A transverse incision was commenced at the level of the anterior bladder neck, guided by the balloon of a urethral catheter. The transection of the bladder was then completed, passing above the ureteric orifices. Fortunately, it was possible to perform the bladder transection while leaving the vesicostomy in place. The bladder division was then closed with 3/0 Polyglycolic acid sutures and the bladder was drained via a urethral catheter, as well as the vesicostomy.

Clinical Outcome

The boy's recovery from the operation was uneventful, but the reduction in his bladder spasms lasted for only 12 weeks. He had commenced to have significant, painful deterioration of his spinal orthopaedic deformity and lower abdominal-wall spasms have lead to ongoing incontinence, which are now being managed with Botulinum toxin injections into his abdominal wall. The bladder pain appears to have been controlled with the transection operation.

Summary

This case report seems to further confirm the variability and unpredictability of the clinical application of bladder transection for bladder instability.

Clinical Studies in Paediatric Urology

Ureterocystoplasty

Introduction

Ureterocystoplasty, the newest technique for bladder augmentation, is rapidly becoming accepted as the ideal form of cystoplasty. The procedure involves incorporation of a strip of dilated ureter into the bladder, thus providing a urothelial lined augmented segment with a muscular support, adequate elasticity and good vascularity; thereby avoiding most of the complications inherent in enterocystoplasty [54-58].

Eckstein and Martin (1973) who reported a two incision, extraperitoneal removal of a poorly functioning kidney from a seven-month-old infant described the first ureterocystoplasty; they used a transverse bladder incision to incorporate the longitudinally incised ureter into the bladder. The authors also showed that the procedure can be performed following a ureteric reimplantation [59]: the patient, now in his third decade, remains well (Etker 1995 - Personal communication). Further experience with this procedure was not recorded until 1993. Wolf et al. (1993) [54], Bellinger (1993) [56], Churchill et al. [55] (1993) and Dewan et al. [60] (1994) then described the operation. Generally, ureterocystoplasty increases bladder capacity, improves continence and lessens vesicoureteric reflux. Unfortunately, the technique can only be used when the ureter is sufficiently dilated [58,61], however, attempts at dilatation of a normal-sized ureter have been studied experimentally. Using a rabbit model, progressive ureteral dilatation was developed over a 30-day period by instillation of saline through a subcutaneously implanted injection port. Ureters were dilated by at least 10-fold and augmentation cystoplasty then performed with these dilated ureteral segments, increasing the bladder capacity by an average of 260% [62]. Although this is interesting experimental work, there are a number of barriers to clinical application.

The way in which ureterocystoplasty is performed is still evolving; initially it was thought that it was essential to incorporate both the renal pelvis and dilated ureter to make the

augmenting patch adequate [54,55,60,63-65]. More recently, cases have been described whereby ipsilateral kidney function has been preserved, by performing a transureteroureterostomy and using the distal portion of the megaureter for augment the bladder [56,57,66,67]. Gosalbez, in 1996 [67] and Ahmed in 1998 [68], each described cases where they combined bilateral ureterocystoplasty, a transureteroureterostomy (TUU), and a Politano/Leadbetter ureteral reimplantation of one ureter, combined with a psoas hitch.

The 10 cases presented here have contributed to the further development of ureterocystoplasty. The first six cases provide insight into a new procedure, which combines the extraperitoneal, and renal preservation concepts; in five this was by approaching the contralateral kidney from the flank, on the side the ureter is to be used for the ureterocystoplasty. Several authors have already described the extraperitoneal approach to avoid the risk of contamination of ventriculoperitoneal shunts, facilitate subsequent peritoneal dialysis, minimise post-operative pain and reduce the period of hospitalisation [54-56,59,60,64,69-71]. The addition of TUU to the operation of ureterocystoplasty enables the patient to have a urothelial lined bladder augmentation, the advantages of staying outside the peritoneum, plus the benefits of renal preservation. Also, in the female, there is no pedicle to interfere with a Caesarean section if required in later years. ***Case 6*** developed both the "spare parts" concept of keeping a ureter, and the ability to use the one ureter for ureteric reimplantation and as an intubatable stoma.

The next four cases had a transperitoneal procedure, which was chosen because of the need for other intra-abdominal surgery and the desire to preserve both kidneys. ***Case 7*** is one of the few examples of ureterocytoplasty after ureteric reimplantation; ***Case 8*** had a concurrent pyelolithotomy; ***Case 9*** highlights the value of the procedure in developing countries, and the ability to remove a renal pelvic stone, and ***Case 10*** describes an adaptation of the Gosalbez/Ahmed concept of reimplantation and ureterocystoplasty of the same ureter.

Case Reports

Case 1: AM: (DOB - 21/3/92UR 874202) presented to the Royal Children's Hospital, Melbourne for management of her neurogenic bladder in October 1997. She had a lipo-meningocele and a dystrophic left lower limb, for which excision of the spinal lesion and amputation of her left foot had been performed. She had never been fully continent and had recurrent urinary tract infections for over 12 months. On examination she was noted to be febrile and unwell, therefore she was admitted for treatment of a subsequently confirmed urinary tract infection. An MCU identified a 50 ml, thick walled, high pressure, trabeculated bladder with grade V vesicoureteric reflux into the right kidney. Both kidneys were hydronephrotic, with significant reduction in size of the right kidney and US showed dilatation of both ureters, having been normal on US in 1996. A DMSA scan and an intravenous pyelogram (IVP) (Fig.17), indicated that the right kidney was virtually non-functioning and the upper pole of the left kidney was scarred; the left ureter was dilated on the IVP. She was started on anticholinergic medication and successfully commenced on clean intermittent catheterisation, following which cystoscopy and an extraperitoneal right nephrectomy and ureterocystoplasty were planned.

Operative Technique (Fig.18): The patient was placed in the right-side-up lateral position and a standard nephrectomy-length skin-crease incision was made between the 12th rib and the iliac spine; dissection was carried down through the muscle layers into the extraperitoneal plane and through Gerota's fascia. After inserting a Denis Browne ring retractor dissection was continued behind the peritoneum, anterior to the inferior vena cava and aorta. The left ureter was identified and stay sutures applied; a one centimetre longitudinal incision was made in the left ureter and a length of right ureter mobilised to form a Y-shaped ureteroureterostomy; the right to left ureteroureterostomy was performed with a 5/0 polyglycolic acid sutures. The right lower ureter was then mobilised down to the

mobilised down to the pelvic brim, preserving its blood supply. The wound was closed, after the insertion of a redivac drain: blood loss was minimal during the 70-minute procedure.

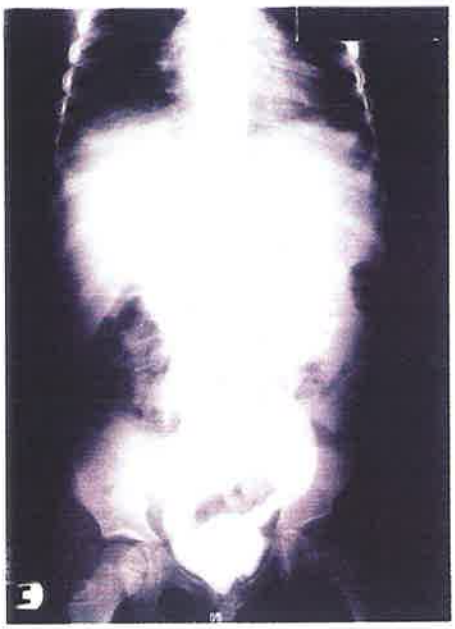


Figure 17: An IVP pre-operatively shows a virtually non-functioning right kidney and a dilated left ureter.

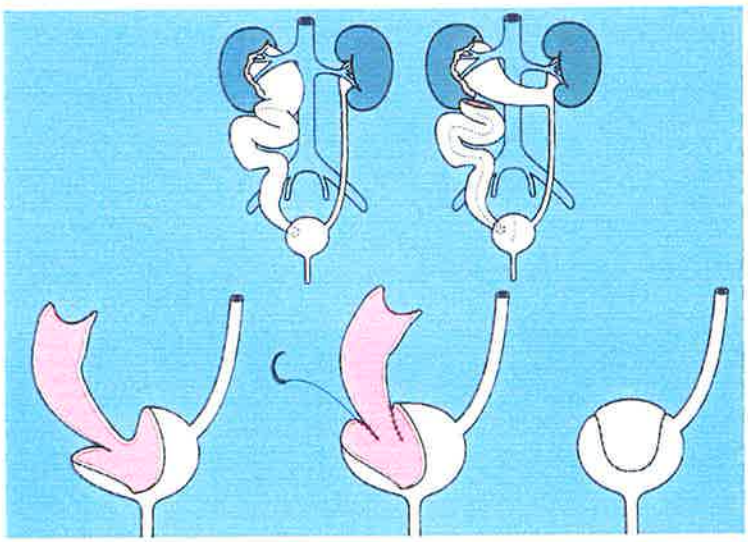


Figure 18: The illustration indicates the transureteroureterostomy and the ureterocystoplasty performed in ***Case 1-4***.

The patient was then placed in the supine position and a modified Pfannenstiel incision performed. The anterior wall of the bladder was mobilised and incised from the anterior bladder neck to the urachal remnant; dissection was then extended from to the orifice of the left ureter, then longitudinally to the level of the transection of the ureter (Fig.18). Two parallel suture lines were used to anastomose the right ureter to the opening in the bladder, thus completing the ureterocystoplasty. An 8 FG feeding tube was placed in the left ureteric orifice and secured with a 4/0 chromic catgut suture. The final position of the tip of the ureteric catheter was uncertain, but assumed to be one of the renal pelves. The bladder and abdominal incisions were closed around an intravesical, 10 FG Malecot catheter.

Post Operative Outcome: The patient recovered from the surgery uneventfully. One-month post surgery her catheterisation volumes were up to 280 ml and she was continent on three hourly CIC and infection free. Ultrasound showed that the hydronephrosis has resolved, and the right kidney contributed 9% of overall renal function on a nuclear renogram. No leak was seen on an MCU 10 days and in January 2000 she was dry both day and night, with a bladder capacity of 313 ml at 62 cmH_2O and a volume of 210 at 30 cmH_2O (Fig. 19).

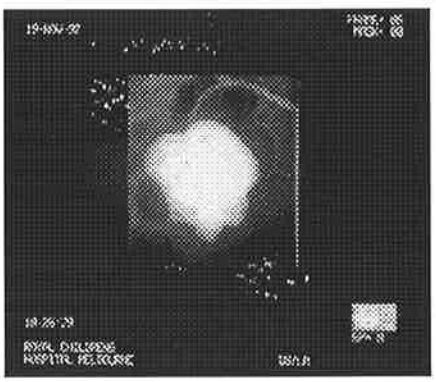


Figure 19: A post-operative cystogram shows a satisfactory bladder configuration 10 days after the ureterocystoplasty.

Case 2: This 7 year old girl had previously had a Mitrofanoff continent stoma formed for management of her neurogenic-bladder incontinence. Despite treatment with CIC and anticholinergic medication she developed right-sided VUR and renal parenchymal defects. She continued to be wet and was recorded to have a high-pressure bladder on repeat urodynamic study. Because she had recently developed VUR, and already had an appendicovesicostomy, it was decided to augment her bladder using an extraperitoneal left to right ureteroureterostomy and a ureterocystoplasty, utilizing the left ureter.

Operative Findings: The left kidney had good parenchyma, the ureters were approximally one centimetre in diameter, and the bladder wall was thick and trabeculated. The Mitrofanoff stoma was easily catheterised with a 10 FG catheter.

Operative Technique: The patient was placed in the left-side-up lateral position, the wound was opened via a muscle cutting incision, Gerota's fascia was entered and the kidney mobilised. The dissection was continued in the retroperitoneum, anterior to the inferior vena cava and aorta, until the right ureter was identified and stay sutures applied; a longitudinal incision was made on the medial aspect of the right ureter, to which a short length of the left upper ureter was anatomosed. The division of the left ureter was angled to facilitate a Y-shaped ureteroureterostomy, which was performed with 6/0 polyglycolic acid sutures. The left lower ureter was then mobilised down to the pelvic brim, preserving its blood supply. The lateral incision was closed, after the insertion of a wound drain.

The patient was then placed in the supine position and a modified Pfannenstiel incision performed. The anterior wall of the bladder was mobilised and incised from the anterior bladder neck to the urachal remnant; dissection was then extended to the orifice of the left ureter, then longitudinally to the proximal end of the ureter. Two parallel suture lines were used to anastomose the right ureter to the opening in the bladder, thus completing the ureterocystoplasty. An 8 FG feeding tube was placed into the proximal right ureter. The

bladder and abdominal wound were closed with the bladder drainage being via a 12 FG catheter in the appendicovesicostomy.

Post Operative Outcome: The girl has been well with a low pressure, good volume bladder. Unfortunately, little detail of the follow-up studies is available.

Case 3: Chandi, a seven-year-old girl had a cystoscopy at the Bangobandhu Sheik Mujib Rahman University Hospital in Bangladesh, on 22/06/98. She was confirmed to have a small volume, trabeculated bladder. On the 25/06/98 an extraperitoneal transureteroureterostomy and ureterocystoplasty were performed, after intermittent catheterisation had been instigated.

Operative Findings: As for the other patients undergoing this procedure, the bladder was thick walled and trabeculated, the ureters thick walled and dilated, with excellent blood supply to the ureter mobilised for the ureterocystoplasty. The transureteroureterostomy was easily performed through the extraperitoneal plane in front of the great vessels. An adequate length of ureter was available for the bladder augmentation. Upper tract drainage was achieved with a catheter inserted via the bladder during the ureterocystoplasty.

Operative Technique: The procedure was conducted in the same manner as for ***Case 1,*** using the extraperitoneal transureteroureterostomy (Fig.18).

Post Operative Outcome: The early postoperative course was uneventful apart from the child failing to comply with the intermittent catheterisation. Her urine cleared of blood in the first few days and her wound was satisfactory at the end of 10 days. She has been reported to be well, but no detailed studies have been conducted.

Case 4: LD was two years old at the time of presentation in Melbourne, with a history of recurrent urinary tract infections and urinary incontinence. A MAG3 scan and DMSA scan showed bilateral hydronephrosis with poor renal function and global parenchymal thinning. A cystogram showed a trabeculated Fir-tree bladder with bilateral grade V vesicoureteric reflux (Fig. 20). At cystoscopy a trabeculated bladder was identified, with a satisfactory ureteric tunnel on the right and a paraureteric diverticulum associated a slightly open ureteric orifice on the left. A urodynamic study, with a fill rate of 30 ml per minute, filled to a pressure of 30 cm H_2O at a total volume of 60 ml with reflux into the right ureter at a volume of 35 ml, into the right pelvicalyceal system at 45 ml and into the left ureter at 60 ml, following which the bladder pressure only rose after the pelvis and calyces appeared tense and distended: the study was conducted through a suprapubic catheter.

The child was commenced on CIC and anticholinergic and, because of the adverse changes on the urodynamic study combined with the already severe renal changes, was advised to have bladder augmentation performed.

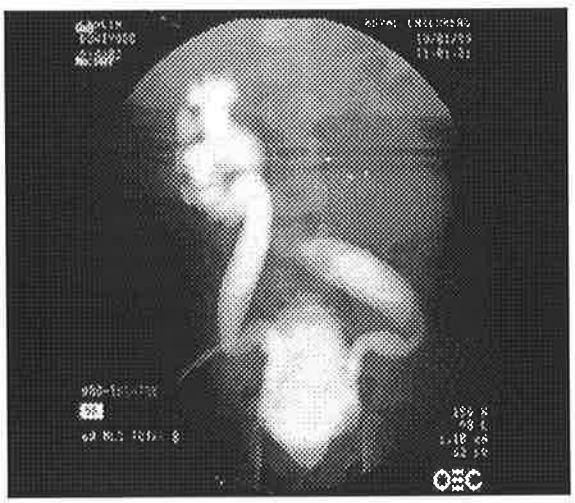


Figure 20: A preoperative cystogram of ***Case 4*** showing high grade bilateral VUR, plus a trabeculated bladder.

Operative Findings: The left kidney was hydronephrotic and adherent to the surrounding tissue, as were the left and right ureters. Through the suprapubic incision, the bladder was noted to be thick walled and trabeculated; the blood supply of the lower left ureter was satisfactory after mobilisation for the ureterocystoplasty.

Operative Technique: LD had a similar procedure to that described in ***Case 1***, for which she was first placed in the left-up lateral position and a skin crease, muscle-cutting incision made. Gerotia's fascia was opened and the adhesions around the kidney were divided. The left ureter and renal pelvis were mobilised, being careful to preserve the blood supply of the ureter. The right ureter was found retroperitoneally, in front of the aorta and the inferior vena cava. A longitudinal incision was made in the right ureter to which was anastomosed an obliquely incised left ureteropelvic junction, with 5/0 polyglycolic acid. The remainder of the upper left ureter was mobilised down to the level of the bony pelvic brim. A Cliney Mallecot nephrostomy catheter was inserted through the renal parenchyma, after closure of the posterior wall of the upper anastomosis; a redivac drainage tube was placed in the perinephric space. The left lateral wound was closed and the patient placed in the supine position. The bladder was approached through a standard suprapubic incision: the bladder was incised in the midline anteriorly, from above the bladder neck to the urachal remnant, then toward the ureteric orifice on the left. Care was taken to avoid the obliterated umbilical artery. A small opening was made in the peritoneum, which was closed with 6/0 Vicryl. The ureter was incised longitudinally over a 14 FG Nelaton catheter and the neo-bladder formed by two parallel sutures between the edges of the ureter and the open bladder. A 10 FG suprapubic catheter was left in the bladder and a redivac drain placed in the prevesical space.

Post Operative Outcome: The redivac drain was removed on Day 2. An antegrade study at one week showed flow into the bladder, with no hold-up at the anastomosis. However, there was relatively high pressure drainage into the bladder, therefore the dose of oxybutinin was increased to 3.75 mg per day; also local anaesthetic (bupivicaine) was

instilled into the bladder every four hours to reduce detrusor spasm. The bladder capacity was 150 ml, 10 days after the operation. Follow-up studies of the upper tracts with nuclear medicine and ultrasound identified that the dilatation had markedly improved, as has the functional components of the parenchyma (Fig. 21). Lencia was being managed successful on 3 hourly CIC, with no urinary infection and dry between catheterisation.

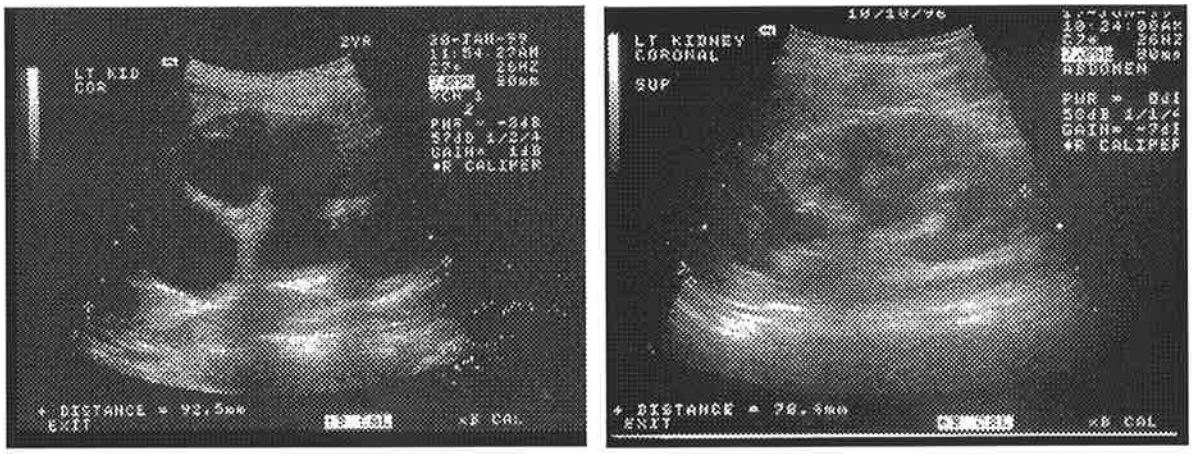


Figure 21: A preoperative ultrasound compared to the post-operative study, showing marked resolution of the hydronephrosis of ***Case 4***.

A urodynamic study performed on 1/12/99 gave a bladder volume of 180 ml at 20 cm H_2O and a total volume of 280 ml at 45 cm H_2O. The shape of the bladder was typical for a ureterocystoplasty and the endoscopy showed a smooth transition between the native bladder and the ureter.

Case 5: MU was noted at 23 weeks gestation to have moderate bilateral pelvicalyceal dilatation, and the bladder was enlarged. No follow-up studies were conducted, until he presented with sepsis at the age of five days. He was found on investigation to have bilateral hydroureteronephrosis, a duplex left collecting system, a thick walled bladder and urethral obstruction. Initial treatment with antibiotics and urethral catheter drainage of his bladder were supplemented with the insertion of bilateral nephrostomy tubes, oral

anticholinergic medication and intravesical Bupivicaine. Fulguration of the posterior urethral obstructing membrane was undertaken when he was biochemically stable and his sepsis controlled. Over a number of days, he was able to be rendered catheter free and was voiding normally. Over the next few months he failed to have satisfactory resolution of his hydronephrosis and his creatinine did not settle adequately. Therefore, having identified that his urethra had been adequately treated he was reinvestigated with a urodynamic study which identified that he had a bladder of 75 ml at 35 cm H_2O. In particular it was felt that he was not emptying his bladder sufficiently to maintain an adequately low, average bladder pressure; this conclusion was reached by identifying that his hydronephrosis improved with an indwelling catheter *in situ*. His bladder status on the latter investigation fitted with the trabeculated appearance seen on the original cystogram (Fig. 22).

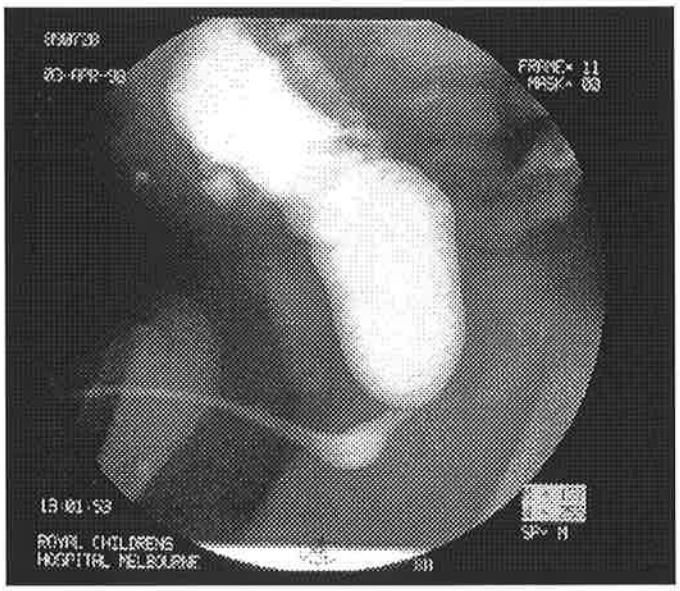


Figure 22: The cystogram of ***Case 5*** taken soon after the original presentation at 5 days of age. The marked trabeculation and thick walled nature of the bladder are evident.

Operative Findings: Adequate parenchyma was found on the left side and the pelvis-to-pelvis, and left ureter to right ureter anastomoses were performed without difficulty; in particular the retroperitoneal anastomosis from left to right was not difficult through the left lateral approach. The blood supply of the left ureters draining the left upper and lower poles was satisfactory after they had been fully mobilized (Fig. 23). The entry of the duplex ureters was though a 2 mm common wall (Fig. 24).

Figure 23: The blood supply of the upper and lower, left ureters for the duplex system for ***Case 5*** was satisfactory after they had been fully mobilized.

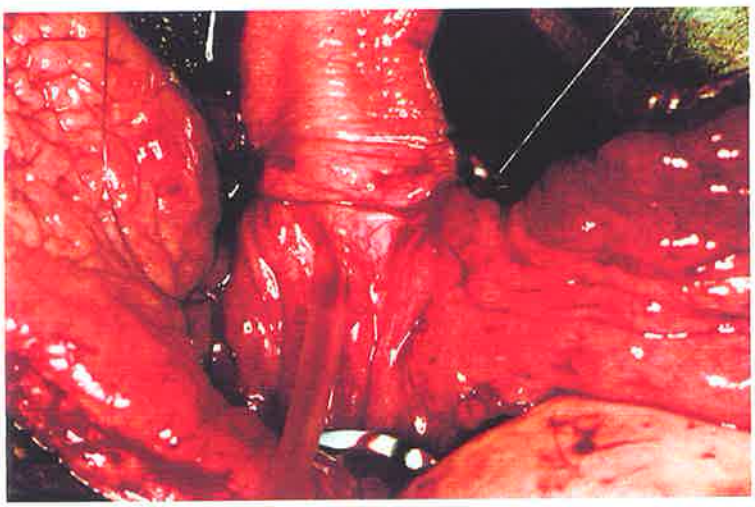


Figure 24: The above picture is of the entry of the duplex ureters into the bladder though a 2mm common wall in ***Case 5***.

Operative Technique: MU was placed in the left-side-up lateral position and a left transverse incision extended through the abdominal muscle, facilitating a retroperitoneal approach to the left kidney. Gerota's fascia was opened, the left kidney mobilized, and the upper and lower pole ureters of the left duplex system identified. Insertion of the lower pole ureter into the bladder was confirmed by placing methylene blue into the ureter and awaiting drainage via the indwelling catheter. The upper pole ureter was then divided transversely and a ureteropyelostomy of the upper pole ureter to the lower pole pelvis, via a single layer anastomosis using continuous 6-0 Monocryl, was performed. A 10 FG Malecot catheter was inserted as a nephrostomy. Retroperitoneal dissection enabled identification of the right ureter, to which the divided lower left ureter was anastomosed, forming a left-to-right extraperitoneal transureteroureterostomy. The lower portion of the two ureters on the left was then dissected to the level of the true pelvis. The lateral wound was closure in layers with 3-0 Vicryl and subcutaneous with 6-0 Monocryl, and steri-strips were applied.

Mitchell was then placed in the supine position, a Pfannenstiel incision was made, and the linea alba and bladder were opened in the midline anteriorly. The incision then extended to the orifice of the lower pole ureter (the most cranial on the bladder), and along the length of that ureter. The proximal half of the two ureters were separated from each other and the caudal (distal) ends were left attached to each other, to help augment the blood supply of each of the ureters. The ureterocystoplasty was completed and the upper pole ureter brought up to the umbilicus as a CIC stoma. As the bladder was closed a 10 FG suprapubic catheter was left *in situ,* and a 10 FG feeding tube was inserted, placed through the catheterisable stoma.

Post Operative Outcome: Mitchell made a rapid recovery and has been shown to have a decrease in the degree of upper tract dilatation, no leak through the umbilical stoma and is

able to empty his bladder without the need for CIC through the stoma. A urodynamic study at four months showed him to have a bladder of 235 ml at 35 cmH_2O, and a volume of 225 ml at 20 cmH_2O. At that stage he remained well without UTI's and with a good stream and large volume voiding episodes, but with inadequate emptying, thus he was commenced on intermittent catheterisation through the umbilical stoma.

Case 6: JU was noted prenatally to have marked renal tract dilatation. He was transferred to the Royal Children's Hospital when initial postnatal therapy had failed to achieve an adequate creatinine level and the dilatation of his upper renal tract remained severe. On arrival he was found to have debris in his renal tract, consistent with urinary tract infection. A MAG3 nuclear medicine study indicated that there was no function in the 1.0 cm left kidney and a cystogram showed the urethral changes consistent with a COPUM. Cystoscopy confirmed the presence of the COPUM, which was disrupted with a Fogarty balloon. Because of the infection, and marked dilatation of the ureters and pelvicalyceal systems, bilateral nephrostomy tubes were inserted. The creatinine level stabilized, as did the infection, but drainage into the bladder was poor. In view of the lack of function on the left kidney, poor emptying of the bladder and marked dilatation of the right side it was decided, rather than perform a vesicostomy or ureterostomies, to proceed with left nephrectomy and to allow bladder drainage by way of reflux through the left ureteric stoma. Subsequently, the bladder emptying remained inadequate; therefore the right system was tapered and reimplanted and the left ureteric tunnel was incised, producing a freely refluxing left system and a non-refluxing right lower ureter. In the next few days, the right nephrostomy tube was removed, after the insertion of a double J stent, which was subsequently removed at the time of demonstrating endoscopically that the right ureteric tunnel was widely patent, at which time his urethral obstruction was formally fulgurated.

JU remained well, but continued to have intermittent urinary tract infections and had passed a variable amount of urine from his stoma and penis. His bladder continued to be thick walled and his right kidney, although markedly improved, remained hydronephrotic. In view of the recurrent infections it was decided to proceed to reduce the redundancy of his renal tract, improve his bladder dynamics and give him stomal access for CIC. His bladder on preoperative investigation had a leak volume of 27 ml with decompression into the left ureter of 12cm H_2O and a total pressure of 30cm H_2O at 45 ml on 24/9/99. A left ureter ureterocystoplasty intubatable Mitrofanoff right lower ureter stoma and ureteric reimplantation of the upper two-thirds of the right ureter was planned.

Operative Findings: On 25 September 1999 the left ureter had a satisfactory blood supply at the end of mobilisation and longitudinal incision. The bladder was markedly thick-walled and fibrotic in appearance. The right ureter was tortuous and large in diameter with a thick wall. The blood supply appeared to be adequate to both the stoma-forming distal third, and the upper two-thirds, at the end of mobilisation and division.

Operative Technique: The patient was placed in the left lateral position and an elliptical incision performed around the left upper ureteric stoma. The dissection was carried down through the subcutaneous fat and muscle layers, whilst preserving the blood supply to the ureter. The retroperitoneal mobilisation was carried down to the pelvic brim, the ureter was oversewn and the wound closed in layers with 3/0 and 5/0 Vicryl. The patient was then placed in the supine position and a modified Pfannenstiel incision was performed, and the bladder dissected free from the anterior abdominal wall. An anterior midline incision was made in the bladder and across to the left ureteric orifice, over the dome. Extravesical dissection was performed on the left, being careful to preserve the blood supply of the left distal ureter. The ureter was incised longitudinally to its proximal end, and the skin attachment to the proximal end

was removed. The ureter was noted to be attached laterally and caudally producing a segmental out pouching of the bladder in that region, which was left undisturbed to preserve blood supply to the distal left ureter.

Attention was then turned to the right side where extravesical dissection identified the right ureter, the blood supply of which was preserved despite the dissection being carried up to the level of the renal pelvis. The length of the ureter necessary for reimplantation and for establishment of a right Mitrofanoff stoma was measured, and the ureter divided. A strip of skin in the right iliac fossa was then configured as an interposition flap to produce an intubatable stoma, centered at McBurney's point. The distal third of the ureter was bought out through the muscle layers to the site of the intubatable stoma orifice and anastomosed to the skin flap with 5/0 Monocryl. The proximal two thirds of the ureter was then passed behind the Mitrofanoff ureter and passed through a transverse tunnel in the upper part of the trigone. The intravesical ureter was sutured in place with 5/0 catgut with a relatively short tunnel after the ureter had been imbricated with interrupted 5/0 Monocryl sutures to decrease its huge lumen. An 8 FG ureteric catheter was placed into the right kidney through the ureteric orifice and secured with a bladder suture. A 10 FG Cliney Mallecot catheter was easily passed through the intubatable stoma and this was sutured into the bladder, also with 5/0 chromic catgut.

The ureterocystoplasty was then performed using continuous sutures of 5/0 Monocryl on either side of the ureter to anastomose the left ureter, the two halves of the bladder to the midline anterior portion of the bladder on either side. A further 10FG Cliney Mallecot catheter was left in the bladder through the vesicoureteric anastomosis. The wound was then closed with 3/0 and 5/0 Vicryl and the skin wound of the intubatable stoma was closed with 5/0 Monocryl. Catheters were sutured in place with 3/0 nylon, and the patency of the three catheters was assessed with flushing of saline.

Gentamicin and Ampicillin were given at the start of the procedure and Cefotaxime was continued post-operatively.

Post Operative Outcome: The patient was noted to have obstructive diuresis following the procedure and drained poorly through the vesicoureteric junction initially, therefore he had an indwelling stent inserted. After four weeks the stent was removed and the hydronephrosis was subsequently minimal, with the child on intermittent catheterisation. An ultrasound in January 2000 showed virtual complete resolution of the hydronephrosis and a cystoscopy and urodynamic study showed the bladder to have a volume of 130 at 20 cmH_2O, with an end fill volume of 160 at 35 cmH_2O. He had remained well, free of urinary infection and was voiding occasionally.

Case 7: This eight year old girl was incontinent, had high grade reflux into the right kidney and reduced function in both kidneys, particularly on the right. She presented for treatment during a Paediatric Urology teaching visit to the National Pediatric Institute in Hanoi, Vietnam, in 1998. The girl had previously had bilateral transtrigonal ureteric reimplants, but was found to have redeveloped VUR on the right. An ultrasound showed bilateral hydroureteronephrosis and *Pseudomonas Aeroginosa* grew from a urine specimen. The operation consisted of a transperitoneal left to right, transureteroureterostomy, a ureterocystoplasty and an appendicovesicostomy. This combination was chosen to facilitate renal preservation and provide an abdominal stoma for CIC. The latter because of the cultural perception of urethral CIC is inappropriate. The preoperative investigations showed a markedly trabeculated bladder, with only minimally dilated ureters (Fig. 25).

Operative Findings: Both ureters were dilated, but not as widely as for the ureters in the other cases, but with a good blood supply to the proximal left ureter after it had been mobilised and longitudinally incised. The previous ureteric reimplant had resulted in the

left ureter being cranial to the right ureter in the bladder, with a wide, but adequate length of the right ureteric tunnel.

Operative Technique: The patient was placed in the supine position and the previous midline incision was opened and extended to the mid-upper abdomen. The bladder was mobilised and the right colon and small bowel mesentery were dissected from the posterior abdominal wall, sufficient to allow for retroperitoneal exposure of the left renal hilum and the upper right ureter. The left ureter was located via dissection anterior to the inferior mesenteric artery; this was facilitated by combining with dissection of the ureter cranially from the pelvic brim. A length of proximal left ureter that would allow for a tension free anastomosis to the side of the upper right ureter was determined; the left ureter was divided and a ureteroureterostomy was performed with 6/0 polyglycolic acid sutures.

The distal left ureter was mobilised down to the bladder leaving a significant amount of periureteral tissue posterior to the lower ureter, ensuring that the lateral aspect of the pelvic portion of the ureter was not disturbed. The bladder was then opened longitudinally along its anterior surface, over the dome, to the orifice of the reimplanted ureter, then along the ureteric tunnel, and into and along the proximal ureter, paying attention to the preservation of blood vessels. The appendix was mobilised and a routine appendicovesicostomy was performed (Mitrofanoff) [72]. The opened ureter was sutured to the bladder with two longitudinal suture lines, as for the other nine cases. A ureteric catheter was inserted into the right ureteric orifice, to allow upper tract drainage; the bladder was drained via both a urethral and Mitrofanoff catheter.

Post Operative Outcome: The patient made an uneventful recovery. A cystogram at 10 days showed no vesicoureteric reflux. Twelve months after the procedure she was reported to be continent and infection free, with a low-pressure bladder, with a large unspecified

volume. The author was unable to obtain any further detail from the treating surgeons in Vietnam. The patient, however, had made an uneventful recovery in the first 10 days.

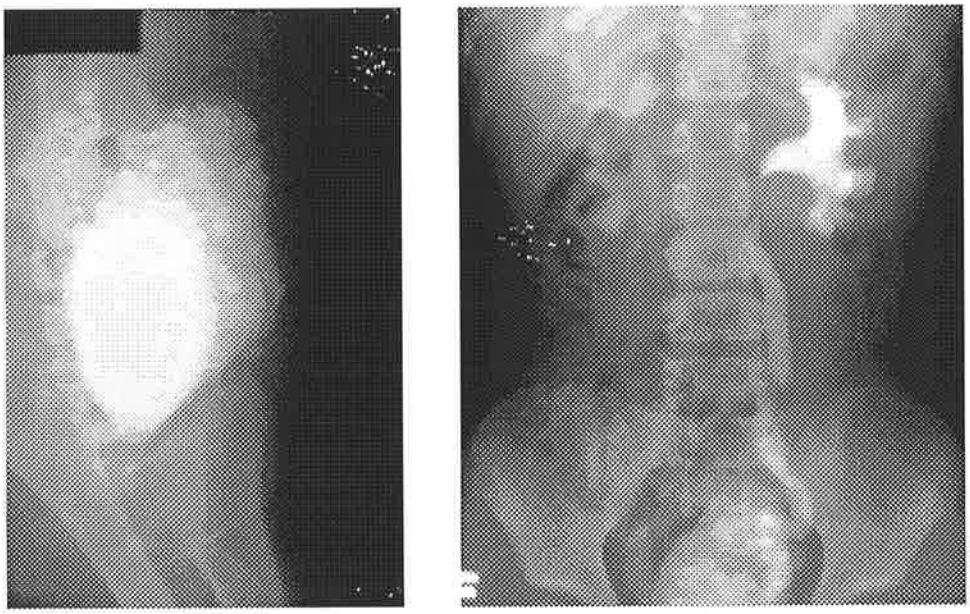


Figure 25: A cystogram and IVP of one of the Vietnamese patients - ***Case 7.*** The marked trabeculation and the relatively small ureters can both be seen.

Case 8: DA was born with an anorectal anomaly for which he initially had a colostomy and a subsequent anorectoplasty, in Papua New Guinea. Infection and infarction of the rectum complicated the procedure, with concurrent damage to his urethra. Consequently, he developed an iatrogenic rectourethral fistula, which was managed by diverting the faecal stream with a caecostomy. He had his fistula closed during a urethral repair, but continued to have urethral obstruction, therefore he had a vesicostomy formed, through which a more recent cystogram showed poor continuity of his urethra and a small bladder. He was also investigated for recurrent urinary tract infections which identified small kidneys on an IVP and a one centimetre stone in his left lower pole calyx: a "redo" anorectoplasty, repair of his urethra and closure of the vesicostomy were planned at the age of 10 years.

Operative Findings: The anal region was markedly stenotic and the prostatic urethra was deficient posteriorly, as well as being fibrotic. The pelvic musculature and the sacrum appeared to be adequate and the blood supply of the mobilised rectum was excellent.

After the vesicostomy was closed, the bladder was noted to be small volume (maximal capacity of 20 ml) and high pressure (>35 cm H_2O at capacity). The bladder was thick walled and both ureters were dilated. The kidneys were small and scarred, and a stone was situated in the left mid calyx.

Operative Technique: The patient was placed in the jack-knife position to facilitate a midline incision that allowed for mobilisation of the anus and lower rectum, and the performance of a redo Pena anorectoplasty. Once the rectum had been freed the defect of the posterior urethra was identified, and defined by excision of the overlying fibrous tissue. A graft of foreskin was then harvested and sutured in place with 5/0 polyglycolic acid sutures over an 8 FG urethral catheter. The rectum was then placed within the muscle complex and the posterior midline incision closed.

The patient was positioned supine and the bladder assessed with a large urethral sound, through the vesicostomy. The possibly adequate size of the bladder was not supported by a urodynamic study performed once the bladder was closed. Rather than then convert to a lateral approach to the left kidney a decision was taken to proceed to a transperitoneal ureterocystoplasty and concurrent transureteroureterostomy, with removal of the left renal stone. The abdomen was opened via a midline incision and multiple adhesions were divided, to allow the small bowel mesentery to be freed from the posterior abdominal wall, thus allowing access to the upper ureter and kidney bilaterally: the left renal pelvis was incised longitudinally and the renal calculus removed. The mid portion of the right ureter and kidney were dissected from the surrounding tissue, preserving their blood supply; the ureter was divided at a point that gave sufficient length of the upper ureter to join as an end

to side anastomosis to the left ureter. The anterior wall of the bladder was opened and the left ureteric orifice intubated with an 8 FG catheter that was passed into the right renal pelvis, through the transureteroureterostomy.

The lower right ureter was mobilised with a generous amount of surrounding tissue, following which the incision in the bladder extended to the right ureteric orifice and along the length of the ureter. The right ureter was then anastomosed to the bladder with two parallel sutures of 4/0 polyglycolic acid, around a suprapubic catheter and the abdominal incision was closed in layers.

Post Operative Outcome: The patient did not have any early post-operative complications and was reported to have been well up until reviewed during a further visit by the author to PNG six months later. He had remained continent of urine by day and night, and free of urine infections, but had occasional urgency and needed to void frequently.

The patient was reassessed on 13/3/99, under general anaesthetic. An 8 FG catheter could easily be inserted into the bladder, which contained 200 ml of clean urine. During a rapid fill urodynamic study the volume at 20 cm H_20 to be 220 ml, with an end fill volume of 260 ml, while the patient was not on anticholinergic medication.

Pressure (cmH_20)	Volume (ml)
8	130
9	160
14	200
20	220
38	240
40	250
50	260

Table 3: The pressures and volumes at the time of assessment of the bladder of this boy six months after his ureterocystoplasty.

Cystoscopy showed a large segment of the bladder wall to be made up of the ureter and the posterior urethra was irregular, but there was no stenosis and no adverse changes in the inlay graft tissue. Suprapubic pressure produced an adequate stream.

During the reassessment of this boy's bladder he had an examination of his anus, which showed that he had a minor degree of narrowing at the level of the skin, which was easily dilated to 12 Hagar and the caecostomy was therefore closed. His post-operative course was complicated by an episode of adhesive bowel obstruction, requiring reestablishment of his stoma. In October 1999, the bowel continuity was successful established. He was able to void adequately, with a large bladder volume; his occasional incontinence appeared to be due to lack of sensation, which was overcome by frequent timed voiding. His upper tract dilatation was resolved by the intervention.

Case 9: JFG, a five year old spina bifida boy suffered from recurrent urinary tract infections; he was found to have reflux nephropathy on a nuclear medicine study (Fig.

26), and a markedly trabeculated bladder with multiple diverticula on MCU (Fig. 27). Ultrasound identified bilateral hydroureteronephrosis and a thick walled bladder. An IVP and a nuclear medicine study showed marked thinning of the parenchyma, with obvious cortical defects. His initial investigations were in Australia and his operation was performed in Bandung, Indonesia.

Operative Findings: Both kidneys were enlarged with thinned parenchyma. The ureters were dilated and tortuous, with thickened walls and good blood supply at the completion of mobilization. The bladder was small volume, thick walled, with the multiple diverticula suggested by the MCU.

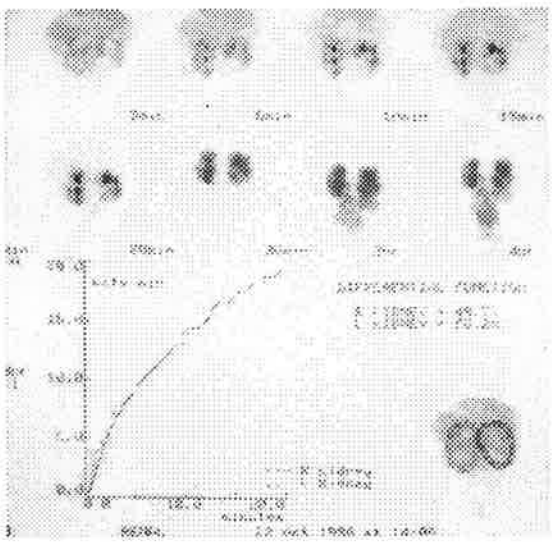


Figure 26: A nuclear medicine study of ***Case 9*** which indicates that the renal parenchyma is thinned, and the pelvicalyceal systems are hydronephrotic.

Operative Technique: The abdomen was opened through a midline incision, the bladder mobilised, as was the posterior abdominal wall attachment of the small bowel mesentry and

the right colon. The right ureter was mobilised to the level of the renal pelvis, carefully preserving its blood supply. A length of right ureter was selected which would facilitate an end to side anastomosis to the left upper ureter. The right ureter was anastomosed to the left with 5/0 polyglycolic acid suture. The bladder was then opened in the usual manner, from the anterior midline to the right ureteric orifice, then along the right lower ureter. The ureterocystoplasty was completed with two parallel sutures of 3/0 polyglycolic acid. An 8FG catheter was placed into the upper tracts through the left ureteric orifice and the bladder closed around a suprapubic catheter. The wound was closed in the usual manner.

Post Operative Outcome: JFG's early recovery was unremarkable and he was discharged well after 14 days. The catheters were be removed on day 10 and he was established on CIC prior to discharge. In the first few months he was infection free and continent, with a marked increase in his bladder volumes. Unfortunately, no further urodynamics have been able to be obtained.

Case 10*:* A seven-year-old boy with lumbosacral spina bifida, presented with urinary incontinence and hydronephrosis. His Indonesian parents had recently been instructed in CIC for management of his wetting and urinary tract infections, which, despite the addition of anticholinergic medication, did not control his incontinence. His preoperative imaging studies showed a hydronephrotic enlarged left kidney with a hydroureter. The right kidney was difficult to see on US but was thought to be present and mildly hydronephrotic. A nuclear medicine study showed good function of a hydronephrotic left system with no function on the right. A cystogram identified a trabeculated bladder with high-grade reflux into a dilated left pelvicalyceal system (Fig. 27). The subsequent operation was planned to consist of cystoscopy, cystometrography and circumcision; a ureterocystoplasty was to be undertaken if the bladder findings indicated the need for augmentation or, alternatively, a left ureteric reimplant would have been performed if the bladder proved to be satisfactory.

Cystoscopic and Operative Findings: The bladder was thick walled, trabeculated, poorly compliant, with a volume of 100 ml at 60 cm H_2O. No ureteric orifice was seen on the right side of the bladder, and subsequently the right renal bed was found to contain only an odd-shaped right lobe of the liver, confirming renal agenesis. The left ureter was hugely dilated and tortuous, with a wide ureteric orifice in a normal position, through which the cystoscope could be easily introduced.

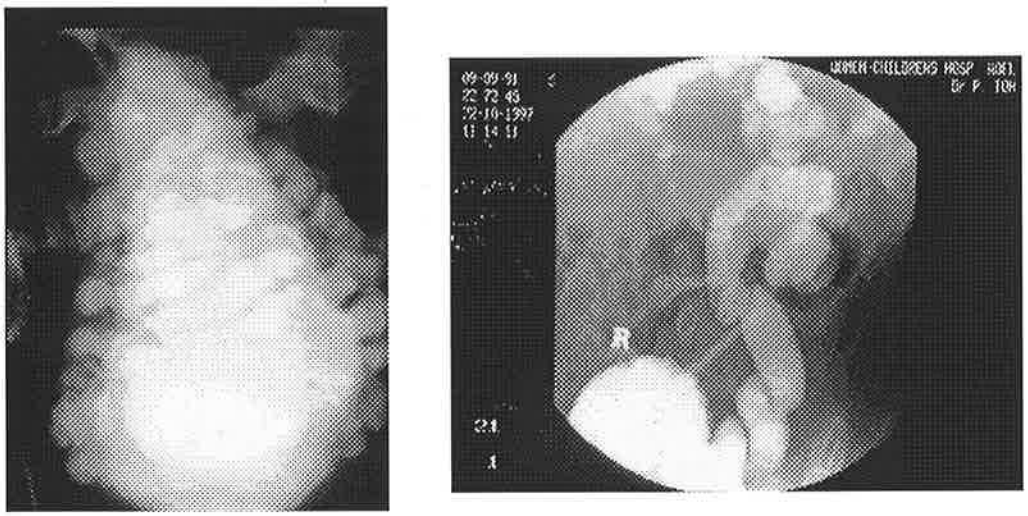


Figure 27: A cystogram of ***Case 9*** (left) showing the marked diverticula formation, compared a cystogram of ***Case 10*** (right) with a less trabeculated bladder with high-grade reflux into a dilated left pelvicalyceal system.

Operative Technique: Through a long midline incision the abdomen was opened, the ventriculoperitoneal shunt identified and isolated to the right side of the abdomen. The bladder was partly mobilised and the right paracolic gutter explored. The left kidney was hydronephrotic and a hugely dilated left ureter was seen in the left retroperitoneal space. The peritoneum was dissected from the mid-portion of the pelvis of the kidney, to the region of the vesicoureteric junction; great care was taken not to dissect behind the pelvic end of the ureter, thus preserving its blood supply. A decision was taken to perform a ureterocystoplasty and reimplantation of the upper ureter into the bladder as described by Gozalbez *et al.* [67]. The bladder was incised from the midline anteriorly to the right of the midline at the apex of the bladder, down to the ureteric orifice and longitudinally in the

lower two thirds of the ureter, having disconnected the lower ureter from the upper one third. The upper end of the ureter had an excellent blood supply and was reimplanted into the left half of the bladder, combined with a Psoas hitch of the bladder toward the lower pole of the kidney: papaverine was applied to the upper ureter to improve its blood flow. The neo-bladder was closed with a running suture to anastomose either side of the opened ureter to each half of the bladder. The front wall of the bladder ultimately consisted of the right half of the bladder and the ureterocystoplasty. A 10 FG Mallecot, suprapubic catheter, a transvesical ureteric catheter and a perinephric drain were left *in situ*.

Post Operative Outcome: The patient recovered uneventfully and a cystogram at 10 days after the surgery did not show any leak, or VUR (Fig. 28). Two weeks after removal of the ureteric catheter the upper tract dilatation had decreased from that seen pre-operatively. At three months he was continent with CIC with a bladder capacity of up to 400 ml and, on US, his upper tract dilatation had resolved completely. He has subsequently remained continent, his hydronephrosis has been satisfactory and he has been infection free.

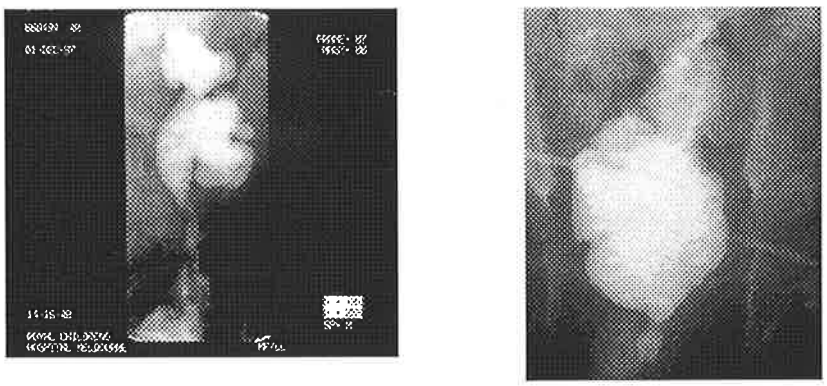


Figure 28: The combined antegrade pyelogram and cystogram of ***Case 10***, performed 10 days after his ureterocystoplasty for a neurogenic bladder and a single kidney.

Discussion

For almost a century, surgeons dealing with bladder dysfunction have recognised the need to enlarge the urinary bladder in certain highly selected instances. The indications for these procedures include a variety of conditions that have, as a common theme, low bladder capacity, poor compliance, and raised intravesical pressure. The effects of the bladder dysfunction include urinary incontinence, abdominal pain, recurrent urinary tract infections, vesicoureteral reflux and, in the worst cases, renal parenchyma injury [55]: unfortunately, a perfect and consistent solution for the dysfunctional bladder is not yet available. Conservative medical therapies include the administration of parasympatholitic drugs (administered both orally and intravesically), CIC, antibiotic prophylaxis and appropriate bowel management. When these fail to produce continence, or lead to kidney damage, surgery is usually recommended.

The underlying diagnoses of the dysfunctional bladder include posterior urethral obstruction [58,60,63,64,67], bladder exstrophy [57], a neuropathic bladder [57,58,64,67], ureteric duplication with reflux [58,66] and end stage renal disease [73], all of which are not infrequently seen with poorly functioning kidneys, with associated massive dilated, refluxing ureters. Often only one kidney is adversely affected: this has been described as a "pop-off" or VURD syndrome (unilateral vesicoureteric reflux and renal dysplasia) [74], in which unilateral vesicoureteric reflux appears to allow the ipsilateral ureter and its pelvicalyceal system to act as an expansive reservoir, thus preventing contralateral vesicoureteric reflux and renal damage by lowering the bladder pressure [75]. Occasionally the dilated poorly functioning system leads to persistent urinary tract infection, suggesting the need for a nephroureterectomy. However, the subsequent removal of the pressure-reducing reflux unit may lead to deterioration in function of the remaining kidney, or the need for a transplant kidney if both native kidneys have already failed.

Until recently the mainstay of surgical treatment was the removal of the refluxing, non-functioning system, and subsequent bladder augmentation, if increased bladder volume was required. Early experience with augmentation most commonly involved the use of ileum, colon and stomach. However, the complications of the use of gut have been increasingly recognised, including excessive mucus formation, bladder rupture, calculi, dysplasia and malignancy, metabolic acidosis and abnormalities of calcium metabolism [54-57,59,76-78]. More recently, the complications of the haematuria-dysuria syndrome, metabolic alkalosis and hypergastrinaemia have been found to be associated with the use of the stomach [79-81]. The paradox of removing renal tract tisssue during a nephroureterectomy and then performing a bladder augmentation with bowel has become more apparent as the complications of enterocystoplasty have been more widely recognised. Concurrently, techniques other than ureterocystoplasty have been developed for the formation of a urothelial-lined neo-bladder, highlighting the appropriateness of avoiding the incorporation of gut mucosa into the urinary tract. Autoaugmentation [82-85], the most widely used urothelial lined bladder augmentation, lacks a support to the urothelium, diverticulocystoplasty is limited to very few patients [86] and use of urothelial lined bowel and stomach segments is yet to be conclusively validated [32,33,38,87]. The ureter has all the appropriate layers and, not surprisingly, has become favoured, particularly when there is high grade vesicoureteric reflux [54-60,64]; in addition to improving the bladder, the procedure also eliminates the predisposition to urinary infection in the poorly draining ureter, without the need for ureteric reimplantation. Of the large number of ureterocystoplasty cases thus-far described, several have been two years of age or less, indicating the applicability to younger children [55-60,64], whereas ileocystoplasty and gastrocystoplasty are rarely considered appropriate to the very young, although they have been used [88,89].

Since Eckstein's paper the technique has been modified, and more refined forms of reconfiguring the ureters have been developed. The combination of ureterocystoplasty via an extraperitoneal approach with renal preservation is the main technical innovation

reported in this work, however, the use of ureterocystoplasty with a single kidney had also not previously been described. The use of a duplex system for ureterocystoplasty and intubatable stoma, facilitated by an ipsilateral pelvis-to-pelvis anastomosis and contralateral ureteroureterostomy, has also not previously been described ***(Case 5)***, nor has ureterocystoplasty after an earlier nephrectomy with ureter preservation ***(Case 6)***.

Originally the renal pelvis as well as the megaureter was thought necessary to ensure an adequate increase in bladder volume [54,55,60,63-65]. More recent studies have shown that use of the lower two thirds of a dilated ureter provides a considerable increase in bladder capacity which has allowed for ureterocystoplasty and preservation of the ipsilateral kidney [56,57,61,66,67,71]. This innovation includes forming a transureteroureterostomy and using the lower 2/3 of the divided ureter for bladder augmentation, as in eight of the 10 cases presented here, or the transected ureter is reimplanted into the bladder and the lower ureter is used for the augmentation, as in ***Case 10***. Thus, both ureters can be used to enlarge the bladder: Gosalbez, in 1996 [67] and Ahmed in 1998 [68], each described cases where they combined bilateral ureterocystoplasty, a transureteroureterostomy, and ureteral reimplantation of the one ureter into the bladder, using the Politano/Leadbetter procedure and a psoas hitch. ***Case 10,*** described herein, is a modification of Gosalbez/Ahmed's technique in a boy who had only a single dilated ureter available for both the augmentation and ureteric reimplant. The transient ureterovesical junction obstruction seen in Gosalbez's case was not seen in this boy, and continence and improved bladder dynamics were achieved. Other less common applications of ureterocystoplasty may include the incorporation of the ureter associated with massive reflux into a non-functioning lower pole of a duplex system. The duplex system use, as in ***Case 5***, has opened further options for rehabilitation of a high-pressure bladder attached to a duplex upper renal tract. Also, if bladder dysfunction occurs secondary to outflow obstruction from a ureterocele, the upper pole ureter of a duplex system can be used for the ureterocystoplasty [67,71].

Several authors have described the extraperitoneal approach first used by Wolf *et al.*, [54]. This procedure, using the double incision suggested by Eckstein in 1973, was used in six of the described cases, five of which involved both lateral and Pfannenstiel incisions to mobilise the ureter of a ureter-donating kidney. This approach, which has not been previously described, avoids the complications of the transperitoneal approach, minimizing post-operative pain and reducing the period of hospitalisation. Earlier series used a midline incision and transperitoneal approach when renal preservation and transureteroureterostomy were planned [54-61,63,64,67-71,73], as in cases 7, 8 and 9; in two of these additional intraperitoneal surgery was required.

Other variations of the operations have been described, including preservation of the ureteric orifice [54] that involves using only the extravesical portion of the ureter for the ureterocystoplasty, which results in a relatively short length of ureter for the cystoplasty. This technique is particularly valid if there has been a previous ureteric reimplant; however, I have incised the ureteric tunnel in all cases of ureterocystoplasty without additional complication. If a reimplanted ureter is used, greater care should be taken not to disturb the lateral blood supply of the distal ureter. Only one of the current group (***Case 7***) had a previous reimplant of the ureter used for the ureterocystoplasty.

Transureteropyeloplasty, a further modification, facilitates the concurrent improvement of delayed pelviureteric drainage, by anastomosing the upper end of the transected ureter to the pelviureteric junction of the contralateral side [61]. The pelvis of either kidney can be accessed during a transperitoneal procedure if stone formation has occurred, as was the case in the boy in Papua New Guinea ***(Case8)***.

Preoperative Management

Several steps need to be incorporated into the preoperative management of patients undergoing this procedure. It is important to ensure that the urethra is intubatable prior to performing a ureterocystoplasty, for two reasons; the first because the need for CIC should be anticipated after any bladder augmentation, and the second because a suitable ureter will often be found in a boy who has previously had urethral obstruction. Never the less, many of these patients will be able to void spontaneously after the procedure, as they do not have a neurogenic sphincter.

Also, in anticipation of the unusual event of the ureter or its blood supply not being suitable, the patient is prepared for enterocystoplasty, as was the case in all 10 cases in this study.

Pre-operative investigation includes assessment of the anatomy of the renal tract, which should be fully reviewed prior to the procedure. It would be expected that one would always use a MCU and either an IVP, antegrade pyelogram or retrograde pyelogram, depending on the findings of the initial investigations. Also, it is appropriate to assess the function of both kidneys with a nuclear medicine scan.

Postoperative Management

A suprapubic tube can provide urine drainage in the postoperative period either alone, or in conjunction with a urethral catheter. I have employed a retroperitoneal drain following the procedure, and continued prophylactic antibiotics. In this period of wound healing and volume accommodation, the advantages of the choice of ureteral tissue became immediately apparent with the visible absence of mucus. The ureteral mucosa also has an inherently better resistance to adherent bacteria than the bowel mucosa alternative.

Subsequent reports have documented a further 38 cases of UCP [57,58,60,61,63,64,66,67,69,71,73], probably far fewer than have been performed.

Follow-up of these cases varies from 3 to 40 months, with no deterioration of renal function and an improvement in bladder volume and capacity, voiding ability, bladder stability and continence usually recorded.

Outcome for Ureterocystoplasty Series

In the 16 cases of ureterocystoplasty I have been involved with, 10 are recorded in detail as part of this study. Overall, good bladder augmentation results, with respect to increasing bladder capacity and maintaining low bladder pressures have been obtained in all children who have had follow-up by the author; no significant adverse results were recorded in any of the patients, although calculi formed in the early post-operative phase of ***Case 1.*** Mucus formation has not been a problem, nor has bladder rupture.

The details for the follow-up results for the 10 patients are given in Table 4. All those patients followed for more that three months have had a substantial improvement in their bladder capacity. The five Australian and one PNG case have all had bladder volume increases in excess of four fold, and those on CIC have remained infection-free with marked improvement in their upper tract dilatation. The patients operated on in other countries have had less rigorous follow-up, but have all had technically successful operations, a smooth post-operative course and reports of good clinical outcome for their bladder enlargement, without other urological complications.

Patient	Age(y)	Sex	Initial	Country	Procedure	Pre-Op Vol	Post-Op Vol
1	5	F	AM	Australia	EPTUU	50	313
2	7	F	VF1	Vietnam	EPTUU	-	-
3	7	F	BDF	Bangladesh	EPTUU	-	-
4	2	F	LD	Austral/Nauru	EPTUU	60	280
5	1	M	MU	Australia	EPTUU + Duplex	75	235
6	1	M	JU	Australia	UCP + Mitrof	45	160
7	6	F	VF2	Vietnam	TUU	-	-
8	10	M	DA	PNG	TUU + stone	20	260
9	5	M	JG	Indonesia	TUU	-	-
10	7	M	JC	Austral/Indon	Single Kidney	100	400

Table 4: The pre and post operative bladder volumes for the 10 children who have undergone a ureterocystoplasty (UCP). EPTUU – extraperitoneal transuretero-ureterostomy: TUU – transureteroureterostomy: Mitrof – Mitrofanoff stoma.

Summary

Although ureterocystoplasty may not provide the same increase in bladder volume as can be expected from enterocystoplasty, intra-abdominal, bowel mucosal and nutritional consequences of routine cystoplasty are avoided. The procedure should be considered in the management of a high-pressure bladder with a refluxing megaureter.

History of Bladder Exstrophy Management

Introduction

Until recent decades, the life-style of bladder exstrophy patients would have been intolerable and the risk of malignancy of the bladder plate not insignificant [90,91] (Figs. 29-30).

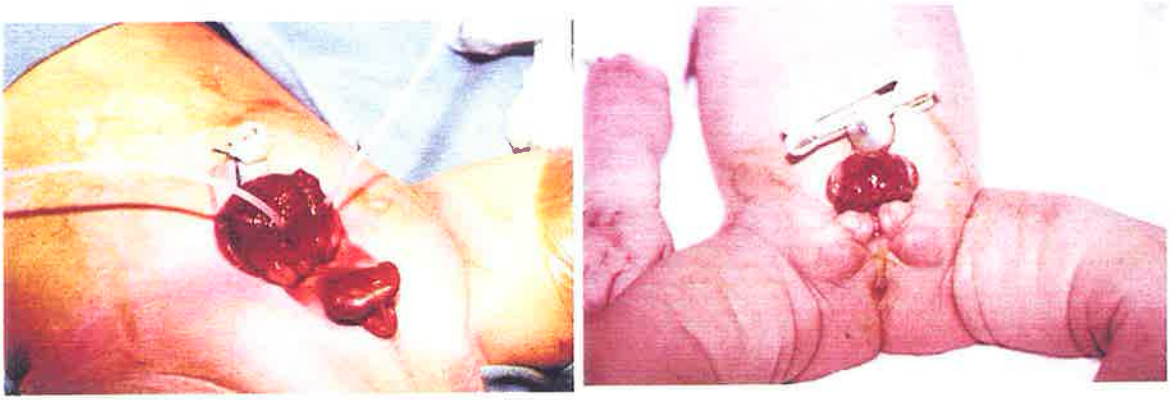


Figure 29+30: A male and female neonatal bladder exstrophy as they usually present in developed countries.

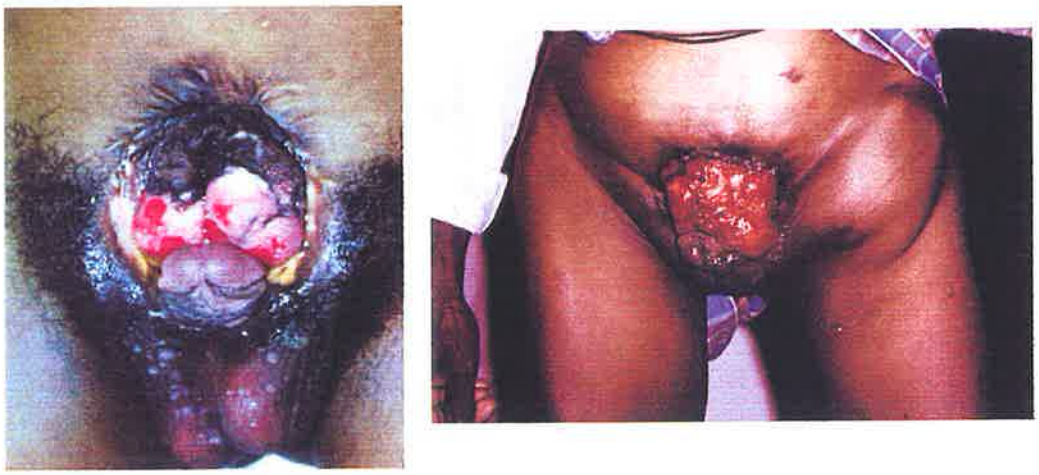


Figure 31+32: Chronic exposure, as shown in this 16 year-old (Fig. 31), can result in development of malignancy, as in this 26 year old man with squamous cell carcinoma (Fig. 32).

Urinary diversion, by ureterocolostomy, was used by Simon [92,93] on 5th July 1851, but it was not until 1942 that Young reported the first continent female patient after a primary repair [94]; the first continent male was recorded by Michon in 1948 [95]. Despite this early success Marshall and Muecke stated in 1970, "Surgical restoration of typical, classical exstrophy of the bladder to obtain normal or nearly normal activity (functional closure) is still an investigative program", based on a summary of 329 published cases from 1906 to 1966 in which there had only been 19% with a fair to good result [96]. And, in 1974, D.I. Williams wrote that "although functional reconstruction seems the logical treatment, and is certainly desired by most parents, many urologists are opposed to it on the grounds that the success rate is small" [97]. This pessimistic view is supported by a study of 12 Paediatric Urology units in Britain and Ireland by de la Hunt and O'Donnell in 1989. They found that 37 of the 81 born between 1975 and 1985 had been diverted [98], whereas no diversion had been undertaken at The Hospital for Sick Children, Great Ormond Street between 1978-1991 with 80% of the patients becoming continent [99]. Lepor and Jeffs also reported an 80% success rate with 28 cases [100] and the Mayo Clinic [90] had a 74% successful outcome with 18 children who had primary closure over similar years. Thus, most surgeons would now anticipate success from primary closure, combined with bladder neck reconstruction and bladder augmentation, although the view is not uniform [101] (Table 5).

The greater proportion of patients require further surgery to be able to be continent [99,101-104], therefore efforts to achieve dryness consume most of the resources in the management of the patient with bladder exstrophy. Intermittent catheterisation [105,106] and continent stomas [72,107-109] have significantly improved the prospect of these patients being dry with good kidneys, and new, urothelial lined bladder augmentation methods may remove some of the risks of current enterocystoplasty techniques [33,38,60].

Authors	Years covered	Bladder Number of patients	Bladder Augmentation rate (%)	Urinary Diversion rate (%)
Mesrobian *et al.* [90]	1918-83	103	0	69
Connor *et al.* [110]	1945-85	207	9	34
Husmann *et al.* [111]	1961-86	156	?	15
Mollard *et al.* [104]	1966-90	73	16	10
De castro *et al.* [112]	1970-91	85	14	0
Csontai *et al.* [101]	1972-89	42	0	83
Oesterling & Jeffs [113]	1975-85	144	1	10
Hollowell & Ransley [99]	1978-90	86	60	0
Perovic *et al.* [114]	1988-91	36	33	0

Table 5: The diversion and augmentation rates for bladder exstrophy from nine reports.

Discussion

With improved management of the major problems for the patient with bladder exstrophy, other aspects of their care have assumed greater importance. The evolution and current status of bladder exstrophy treatment is presented in the following discussion.

1. *Urinary Diversion*

Bladder closure, bladder augmentation or continent diversion have largely superseded diversion, either into the colon or via a non-continent conduit. However, occasionally the appearance of the bladder plate warrants considering the patient for urinary diversion, a technique that is still the preferred option by those who claim incontinence is not achievable with the bladder *in situ* [101].

Ureterosigmoidostomy and other diversion techniques were commonly used until primary bladder closure began to be popular, which coincided with the recognition that diversion of urine into the colon is associated with a 100 fold increase in the risk of colon adenocarcinoma, particularly at the ureterocolic anastomosis [115-118], including in those who had a rectal pouch [115,119]. It is recommended that these patients should have regular endoscopic, histological and biochemical follow-up.

Despite good continence rates resulting from ureterosigmoidostomy or a rectal pouch, the incidence of urinary tract infection and renal damage is high [90,110]; Segura and Kelalis identified 17 of 25 late deaths to be due to renal failure [120]. Thus most major centres have changed from urinary diversion to primary closure [90,110]. The infection and metabolic complication risks of both the ileal and colonic conduits are also high [90], the malignancy risk is still present [121], and ureteral and stomal complications are not uncommon [122].

2. *Bladder Closure and Concurrent Surgery*

a. Early Closure

It was not until neonatal anaesthesia and post-operative care were sufficiently developed that early closure of bladder exstrophy could be entertained. The main advantage of early surgery is the pliability of the neonatal pelvis, which enables an easier closure without pelvic osteotomy. This approach was advocated by Swenson in 1957 [123], initially used by Ansell [103] and is now the preferred option for most [99,104,110,124,125]. Also, delay in closure appears to increase the fibrosis and pseudopolyp formation, making the bladder more difficult to invert [123].

In those infants with a contracted, fibrotic bladder, which cannot be inverted, Büyükünal, after a series of animal experiments, advocated a combined peritoneum, muscle and skin flap, which he has used in six older children (without osteotomy) [126,127]. Alternative, but similar approaches, are to use either an anterior abdominal wall muscle or peritoneum flap or a lyophilized dural patch [128]. In 1869 Thiersch had used skin flaps to cover the everted bladder [129], and Young used a skin and fascial covering over the closed bladder in his 1942 case [95].

Following bladder closure the patient should be maintained on prophylactic antibiotics to prevent renal scarring from the usually present VUR. And, if the residual urine volume is greater than 50 ml, CIC should be instigated as both infection and residual volumes greater than 50ml impact significantly on the incidence of renal scarring in this group [111].

b. Paraexstrophy Flaps

Duckett first described paraexstrophy flaps in 1977 as a method of elongating the urethra [130]; the extra length may improve the prospect of subsequent continence and separation of the bladder from the prostate does make it easier to place the bladder in the pelvis. Successful use of this paravesical, shiny skin has been reported since Duckett's original description [103,104,114]. However, Gearhart *et al.* [131] reported a 40% complication

rate in 78 patients, suggesting ischaemia as the cause of the frequent complication of urethral stricture. Duckett, in an editorial comment, suggested strictures they saw were due to failure to extend the flaps alongside the proximal urethral plate [131]. Care should be taken to prevent ischaemia when mobilising these flaps, which should be reinforced by passing the mobilised interpubic band anterior to the paraexstrophy flap urethra [124,132].

c. Pelvic Osteotomy

Closure of the pelvis in the first 48 hours can be achieved without an orthopaedic procedure [103], and beyond two weeks an osteotomy is usually used. However, Marshall and Muecke [96] and more recently Csontai *et al.* [101] have not used osteotomy, even in the older patients.

Various approaches have been advocated by those who feel osteotomy is required; in 1906, Trendelenberg described disarticulation of the sacroiliac joints to facilitate the closure of the pelvic ring [133]; in 1958 Shultz used iliac osteotomies [134], which some use with the addition of external fixation [104], many use plaster immobilisation only, to which Scherz *et al.* added placement of a band of fascia lata across the symphysis [135]. Anterior innominate [136-138] and superior pubic ramus osteotomies also have their supporters [114,139-141], although the former can lead to the injury of the femoral nerves [136,138], and the latter to injury of the obturator neurovascular bundle.

A number of authors now favour osteotomy in the newborn period, thus converting a relatively tight closure to one without tension [104,138]. Mollard favours the posterior osteotomy [104], however the superior pubic ramus osteotomy is a simple procedure that does not require repositioning of the patient and which can easily be achieved in the neonate. The more aggressive use of a simple procedure is indicated by the findings of Jeffs *et al.* who have shown that the success of the initial closure impacts significantly on the prospect of continence [113]. During the osteotomy it is necessary to protect the vessels and nerves as they pass through the obturator foramen, which can be done by

visualising the neurovascular bundle in the neonate, added by inserting a retractor in older children. The principal advantage of the anterior pubic osteotomy appears to be the independent movement of the adductors to a more medial position [140]. Perovic *et al.* added a combination of wires and screws in older children, and highlight the need to fracture the inferior pubic ramus [114]; as was mentioned in the original description by Cook *et al.* in 1962 [139], but which is less important in the newborn. Schmidt *et al.* changed from the posterior iliac osteotomy to the superior pubic ramus osteotomy in 1977, thenused the technique in 15 patients, concluding that there were fewer incisions, less operating time (five vs three hours) and less blood loss with no difference in the orthopaedic outcome [141].

The most recent bladder exstrophy closure technique has been described by Kelly [142]. Initially his operation involved separating the inner and outer aspects of the pubic rami and thus dissociating the thigh adductors from the muscles of the pelvic floor. The procedure is reputed to improve continence by re-establishing the external sphincter to its normal position around the urethra. However, the external sphincter may well be an intrinsic part of the urethra and therefore not abie to be restored in such a manner [143]. Kelly now mobilises the inner layer of periosteum and muscle only, but also uses an anterior rectus flap to cover the bladder [142], as was reported by Marshall and Muecke who used the rectus fascia flaps instead of osteotomies [96].

Closing the pelvis is not only beneficial for the successful closure of the abdomen, but may also directly influence male sexual function as there seems to be a direct correlation between the interpubic distance and the length of the male phallus [144].

d. Herniotomy

Inguinal herniae occur more commonly in the bladder exstrophy group because of the lack of obliquity of the inguinal canal. The approach to the management of inguinal herniae is variable. Jeffs *et al.* suggested they should be treated when the hernia presents [145], whereas others routinely explore bilaterally for a hernial sac at the time of the primary procedure, particularly in males [103]. Support for the latter approach comes from a recent study by Stringer *et al.*, in which they reviewed 70 cases and found herniae in 86% of boys and 15% of girls, of which the majority of the herniae were bilateral. Incarceration and recurrence were both common [146]. The data reported by Husmann *et al.* were similar, but with a lower male incidence of 56% [147]. They also found that patients who underwent bladder preservation had a higher incidence of inguinal hernia than those who had urinary diversion. The much lower incidence reported by de la Hunt and O'Donnell (nine of 81) may have been due to under-reporting in a multi-centre study [98].

One aspect of the management of inguinal herniae which has not yet been assessed is the impact of the additional, lateral (although minimal) dissection on the success of bladder closure. It would seem reasonable to inspect the cord structures from within the pelvis at the time of the bladder closure and, if a hernia sac is present, perform a herniotomy, thus occasionally avoiding additional lateral subcutaneous dissection. If no sac is present the parents should be informed of the potential of developing a hernia, and if a symptomatic hernia subsequently develops both inguinal canals should be explored.

3. *Surgery and Other Intervention to Produce Continence*

a. Bladder Neck Reconstruction

Continence can be achieved in a small proportion of patients without further surgery to increase outlet resistance [90,99,110,123] (Fig. 29); however the majority now undergo a modification of the Young-Dees-Leadbetter bladder neck reconstruction in which the lower

modification of the Young-Dees-Leadbetter bladder neck reconstruction in which the lower bladder is tubularised and reinforced with detrusor muscle to form a longer urethra [148-150]. Young proposed the tubularisation [148], Dees added a proximal extension [149] and Leadbetter facilitated further proximal extension, by reimplanting the ureters higher on the bladder [150] (Fig.30). Young [95] used his tubularisation of the caudal portion of the bladder in the first successful case in 1942 and Marshall and Muecke [96] achieved continence in 10 of 12 in which bladder neck reconstruction was performed as part of the initial operation. Only one of eight without the extra urethral length became continent.

Figure 33: An endoscopic view of the bladder neck region three months after neonatal bladder exstrophy repair, without bladder neck reconstruction.

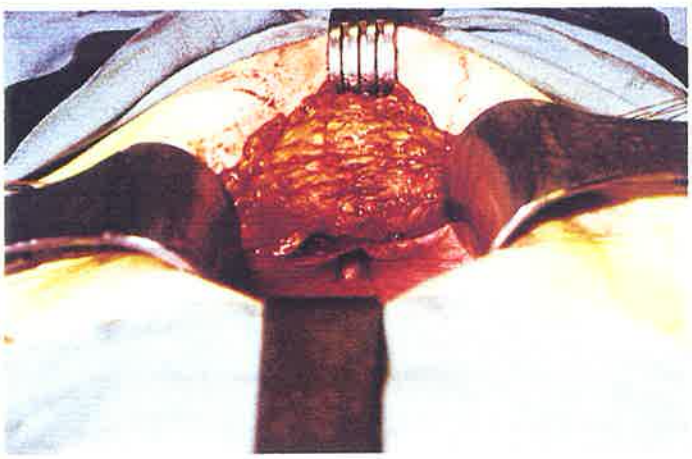


Figure 34: An intraoperative view of the bladder neck prior to a Young-Dees bladder neck reconstruction.

Meguerian *et al.* found urethral length to be the most important determinant of success in achieving a dry patient, and a bladder neck reconstruction length of 3.5 cm is considered necessary [100,150].

Modifications of the Young-Dees-Leadbetter technique have included the addition of an anterior suspension of the bladder neck [100], the use of the transtrigonal ureteric reimplant, and more recently the cephalotrigonal reimplant, reported by Canning *et al.*, which provides for a slightly longer urethral reconstruction [151]. Diamond and Ransley initially suggested surrounding the Young-Dees tube with silicone sheeting, later modifying the operation to include the addition of an omental wrap, because of the 25% urethral erosion rate. This manoeuvre facilitates the later insertion of an artificial sphincter, if required [152]. Wrapping a longitudinal detrusor flap across the bladder [153] and rotating a long flap circumferentially around the neo-urethra [154] have both been reported with success, but have not been widely reported in subsequent studies. Alternatively, Kropp developed a technique of tubularising the bladder neck and performing an antireflux style of implantation into the bladder; however when continence is achieved there is loss of the bladder neck safety valve mechanism which allows leakage at high pressures [155].

Bladder neck repair is now commonly used as part of the anticipated staged reconstruction, with wide variation in the proportion of patients requiring bladder augmentation to achieve the three to four hour dry interval considered necessary to be deemed continent (Table 6).

b. Bladder Augmentation

The success of primary reconstruction has improved with time, partly due to the use of bladder augmentation as a component of the staged procedure in those with a persistently small bladder. Table 5 indicates the change away from diversion and the percentage of patients who have required bladder augmentation and Table 6 shows the use of bladder augmentation as an adjunct to bladder neck reconstruction.

Authors	Years covered	Number of patients	Continence rate (%)	Bladder Augmentation rate (%)
Lepor & Jeffs [100]	1975-82	22	80	0
Canning *et al.* [151]	1986-92	75	75	0
Perlmutter *et al.* [125]	1971-89	22	77	36
Mollard *et al.* [104]	1966-90	55	89	0
Connor *et al.* [110]	1945-85	25	82	17
Hollowell & Ransley [99]	1978-90	79	80	41

Table 6: Continence rates after bladder neck reconstruction, including the proportion requiring bladder augmentation to achieve that success.

It can be seen from the tables 5 and 6 that the reliance on bladder augmentation differs between centres; however the general principle, i.e. augmentation when the bladder capacity is less than 60 ml, is widely held [110,125,138]. None-the-less, Gearhart found improvement in bladder capacity in 25 of 28 patients after epispadias repair and used bladder augmentation in only one of 23 previous failed repairs [138]. This relatively conservative approach to the use of bladder augmentation has merit, as incorporating intestinal mucosa into the urinary tract is not without complication.

All gut segments have been used in the exstrophy population, including the stomach in a small number of cases [112,156], from which complications of stone formation [104,112] and spontaneous bladder rupture [99,104] have all been documented. Metabolic complications have not been seen, but are an inherent risk from augmentation, and malignancy can occur because of the nature of the exstrophic bladder [90,91,157] as well as from the bladder augmentation [121].

c. Intermittent Catheterisation

One of the greatest contributions to the management of bladder exstrophy has been the advent of CIC as first published in 1972 [105,106]. The concept of regular emptying of the bladder by catheterisation has facilitated continence in those with a good volume, poorly emptying bladder, and provided a mechanism to safely empty small bladders that have been augmented. Many recent studies report the addition of CIC to the primary closure, bladder neck reconstruction and augmentation regimes [99,104,112]. To ensure that adequate emptying of the bladder is possible it is important that a urethra that is readily catheterised is formed during the urethroplasty phase of the repair. Further to the development of CIC emptying of the bladder came the use of catheterisable stomas created by using the appendix, ureter or other mucosal lined continent conduit, which has been attached to the anterior abdominal wall and bladder.

d. Continent Diversion

When reconstruction of the bladder is not possible, has failed to achieve continence because the outlet does has not provide satisfactory resistance, or the urethra is not adequate for CIC of a poorly emptying bladder, a continent stoma is indicated. This approach was popularised by the work of Mitrofanoff who described the attachment of the appendix to the bladder in an antireflux manner in 16 cases and the use of a continent ureteric stoma in two [72]. Since then, continent stomas have been used in large numbers of patients with a variety of different tubular structures forming the catheterisable stoma. Attachment to the skin can be either in the perineum [107], to lower abdomen or umbilicus [108,158], and the ureter [72,99,107,108,159], fallopian tube [159], vas, ileum [108,160,160], skin, stomach and colon tubes, and bladder flaps [161,162] have all been used as alternatives to the appendix. The same stomas have been used with either augmented bladders or continent intestinal urinary reservoirs [159,163].

To be successful the stoma needs to be cosmetically acceptable, non-pouting, continent, easily intubated and easily reached by the patient. The appendix was initially the stoma of choice [107,108], but the ureter is becoming increasingly used, and was the preferred option of Woodhouse *et al.* [159]. The results are usually satisfactory; however, late stricture, stomal stenosis and incontinence can occur [107,108]. Closure of the bladder neck appears to increase the risk of stone formation, particularly when bladder augmentation has included the incorporation of gut mucosa [108]; this may be overcome if bladder augmentation is performed with one of the urothelial lined techniques [33,38,60].

e. New Continence Technology

i. Artificial Sphincters

Artificial urinary sphincters have not been extensively used in the management of bladder exstrophy, mainly because it is felt that erosion through the scar tissue is more likely than through a normal urethra of a neuropathic bladder [164]. Small numbers of cases have been reported by several authors, with acceptable continence rates [99,110,125,156,165,166]. Urethral erosion was seen [99,104], but may be overcome by preparing the urethra with omentum and silicone as suggested by Diamond and Ransley [152]. However, they caution against the use of thick silicone sheeting which may in itself cause erosion, a complication which has resulted in occasional loss of the urethra [152]. It is this risk of urethral loss that motivates most Urologists to redo the bladder neck reconstruction and resort to a continent diversion if continence is not achieved, rather than use an artificial sphincter.

ii. Bladder Neck Injections

Polytetrafluoroethylene (Polytef) has been widely used for the management of incontinence in adults, with generally good results, but its efficacy in bladder exstrophy has not been established. Alternative injectables, such as silicone particles, have been used with success in a male patient, by Mollard [104]. Two studies have reported the use of glutaraldehyde cross-linked collagen; Caione *et al.* used collagen to treat 16 patients with either bladder exstrophy or epispadias, most of who required additional injections. They claimed to have achieved an increased urethral resistance, thus improving bladder capacity prior to bladder neck reconstruction [167]. Ben-Chaim and Gearhart injected collagen into 19 patients who required 33 injections but only achieved continence in 58%. They felt that the periurethral fibrosis might prevent the lifting of the mucosa from the underlying tissue necessary for success of the endoscopic technique [168].

4. *Genital Reconstruction*

a. Epispadias Repair

Repair of the male urethra is now considered by many to be an important part of establishing continence, thus urethroplasty is usually performed to encourage bladder enlargement prior to bladder neck reconstruction [113,169]. Surprisingly, this sequence was not used by Merguerian *et al.*, even though they found that urethral length was the most significant factor in developing continence [169].

Epispadias repair usually involves the formation of a Duplay tube that was initially described in 1880 [170]. In 1895, Cantwell provided a method of transposing the urethra to the ventral aspect of the penis [171], a technique revived and improved by Ransley's addition of the principle of chordee reversal by corporal rotation, as developed by Koff and Eakins [132,172]. The technique was initially not favoured because of the risk of urethral devascularisation. With magnification loops, antibiotics, better instruments and modern suture material this does not seem to be a problem, especially if a urethral "mesentery" is created; the ventral urethra is quite amenable to the often necessary CIC. When the urethra is significantly shortened, a pedicle island graft [124,132,173,174], a double island pedicle [173], or a free graft [116,175,176] can be interpositioned at the base of the penis, or one can use the H.H. Young tumble tube technique [116,132]. If the prepuce is not used for the urethroplasty it can be used as a vascularised preputial flap to cover the dorsum of the penis [132].

The preferred technique is currently the Cantwell-Ransley, as reflected by the results from Jeffs' group, in Baltimore, who have recently changed from using Young's technique to the Cantwell-Ransley procedure, with improvement in the fistula rate from 38% to 8% [177].

Kelly and Eraklis [178] reported a technique of more extensive mobilisation of the neurovascular bundle than usually suggested [176] and has not been subsequently reported. Many would consider the length defect to be intrinsic to the corpora and the operation

potentially unsafe [179,180]. Kelly also advocates forming a hypospadiac urethra at the time of bladder closure and performing a lateral flap urethroplasty performed at three years of age [142]. The addition of this stage to the urethroplasty seems unnecessary when satisfactory results have been obtained with the Cantwell-Ransley approach [102,177,180], although Kelly's procedure may have merit because of it is aimed at placing the proximal urethra within the sphincter mechanism.

It was believed that mobilisation of the corpora from the inferior pubic ramus would improve penile length, however it has recently been identified that the proximity of the pubi has a direct correlation on the length of the male phallus, and that closing the pelvis may improve the outcome [144,177], without the risks of extensive mobilisation. It is also appreciated that injudicious dissection to facilitate penile lengthening can damage the corpora and/or neurovascular bundle.

Excessive chordee can be repaired by either a reverse Nesbit procedure [181,182] or corporal rotation at the time of the initial epispadias repair [172], or by the use of an inlay graft to the corpora in patients having follow-up procedures [182].

b. Female Genital Reconstruction

At the time of the original closure, the introitus can be opened [124] and the clitoral hood constituted. In the neonate it is difficult to combine the two halves of the clitoris, which can be achieved by denuding the medial surface at a later stage [124]. The urethroplasty is often carried out as part of the bladder neck reconstruction, with extra length gained by raising a flap from the mons, as used by Gearhart *et al.* for female epispadias repair [183], or from vaginal wall and perineal flaps [184]. As for the epispadiac female, and the bladder exstrophy male, there may be an advantage in pre-empting the bladder neck repair with urethroplasty to encourage an increase in bladder capacity [183].

The mons often requires further reconstitution in later life because of the progressive widening of the lower end of the initial wound. A good cosmetic result can easily be obtained by lateral mobilisation, bringing together hair bearing skin [124]; at this stage the size of the introitus can be reassessed.

As the Mullerian duct structures are usually normal, pregnancy is obviously possible and, in fact, have been recorded in over 130 women [116]. However, during pregnancy both rectal and uterine prolapse are common, except where the vaginal orifice is stenotic [185]. The delivery should be via caesarean section for continent patients, with due precaution to avoid the pedicle of any bladder augmentation segment [91]. If urinary diversion has been performed, vaginal delivery should only be used if uterine prolapse occurs before delivery [186].

Two further considerations for women with a history of exstrophy are the state of their renal function during the pregnancy and their concerns about the transmission of the anomaly. Renal function should be monitored in accordance with the pre-pregnancy status, and both the men and women can be reassured that recurrence of the abnormality in offspring is very unlikely [186].

5. *Prenatal Ultrasound Diagnosis*

Prenatal diagnosis of bladder exstrophy has been recorded by a number of authors since first described, in 1986 [187-189]. The ability to make a diagnosis is based on the fetal bladder normally being identifiable at 15 weeks gestation. A poorly filled bladder can be differentiated from an absent bladder by locating the umbilical arteries as they run parallel to the lateral aspect of the bladder when it is in the normal position [189]. The fetal bladder normally empties every 50 to 155 minutes [190]; therefore if not seen in the first brief scan a further study, after an appropriate delay, should allow identification of the normal bladder. Failure to visualise the bladder, in the presence of normal kidneys and amniotic fluid,

should suggest bladder or cloacal exstrophy. Low insertions of the umbilical cord, separation of the pubic bones, a short penis or a bifid clitoris are also supportive of the diagnosis [188,189]. The bladder may appear as a mass on the lower part of the abdomen [188], or the lower abdomen may be irregular as can be seen in Figure 35. Careful US assessment for other congenital anomalies and chromosome analysis are recommended, as associated anomalies can be identified [90,98,101,191].

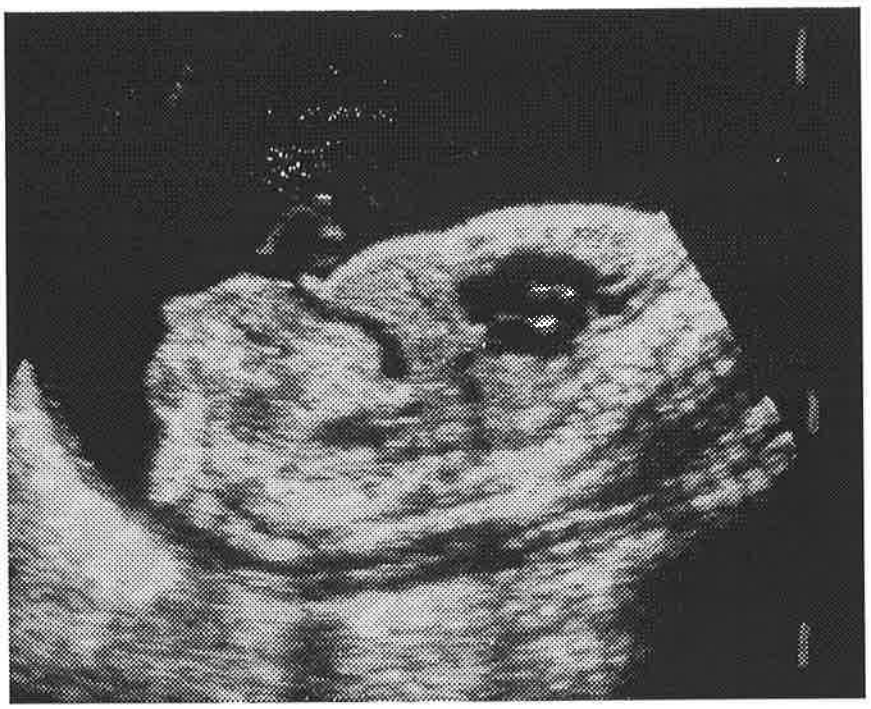


Figure 35: In this prenatal ultrasound study, the absence of filling of the fetal bladder, in the presence of normal kidneys and amniotic fluid, and the bulge seen on the lower abdomen (caudal to the umbilical vessels) suggests the diagnosis of bladder exstrophy.

Prenatal diagnosis facilitates arrangements for delivery in a tertiary institution and early surgical intervention, with potentially improved outcome. Unfortunately, despite the expectation of a satisfactory cosmetic and functional outcome, termination of pregnancy is often performed after the diagnosis of bladder exstrophy has been made.

Summary

Terry Allen's recent words, "I believe the final chapter on treatment of bladder exstrophy has yet to be written", will probably remain current for some time at an editorial comment to work by Jeff's group [110]. However, many new concepts have been added to the understanding of bladder exstrophy but until continence treatment has further improved, new ideas should continue to be fostered and scientifically applied. However, we have come a long way and increased attempts to address the relatively less important facets of management are timely.

Umbilical Transposition in Neonates with Bladder Exstrophy

Introduction

Bladder exstrophy is now managed with a staged surgical approach, giving a good prognosis for continence and normal renal function [99,104,169]. Early closure has contributed to the improved results, a factor that allows for the preservation of the umbilical cord for use for formation of an umbilical cicatrix. In order for the vessels to be used for the omphaloplasty, they need to be transposed from immediately above the vesical plate to the correct position on the abdomen (Fig. 36). The appearance resulting from this procedure is more normal than for a subsequent skin flap omphaloplasty, provided the umbilical stump is positioned correctly on the abdomen and sepsis is avoided at the time of the procedure.

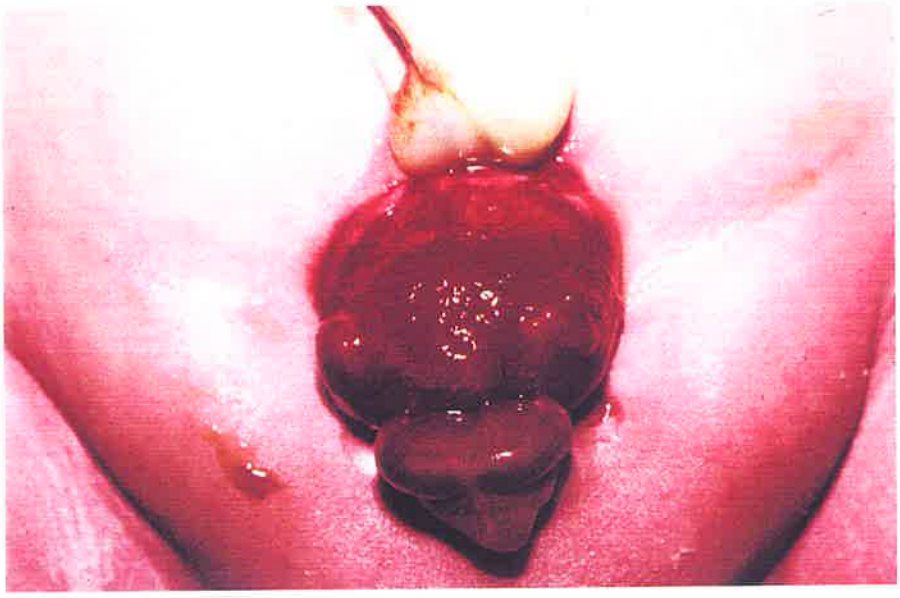


Figure 36: The relationship of the umbilical stump to the bladder prior to bladder closure. The boy also has bilateral inguinal herniae.

Patients and Method

Between October 1990 and July 1998, 12 patients with bladder exstrophy had a primary bladder closure, three of which were referred for surgery in the newborn period.

Early in the dissection the umbilicus is removed from the apex of the bladder plate with a small cuff of fascial tissue. During mobilisation of the bladder, the umbilical stump is preserved and the umbilical arteries are freed in the extraperitoneal plane until the base of the umbilical stump is able to reach the mid abdomen (a point above a line between the anterior superior iliac spines, equal to the distance the neo-symphysis is below that line). The linea alba is then sutured to the cuff of fascia adjacent to the umbilical vessels and the remainder of the wound closed in the usual fashion. Adding superior pubic ramus osteotomies in all these children reduced tension on the wound.

The umbilical stump was managed post-operatively by application of antibiotic cream on a regular basis, for five to seven days. Peri-operative intravenous antibiotics were administered in accord with the management of the bladder exstrophy closure.

Results and Comparison with Other Methods

Each patient has been followed for at least to 12 months; all have an umbilicus in the normal position with a satisfactory appearance (Figs. 37+38). None of the children had sepsis related to the umbilical stump, and none has developed an umbilical hernia needing further surgery. Primary closure of the bladder and anterior abdominal wall was achieved in each case, and there were no episodes of wound sepsis or breakdown.

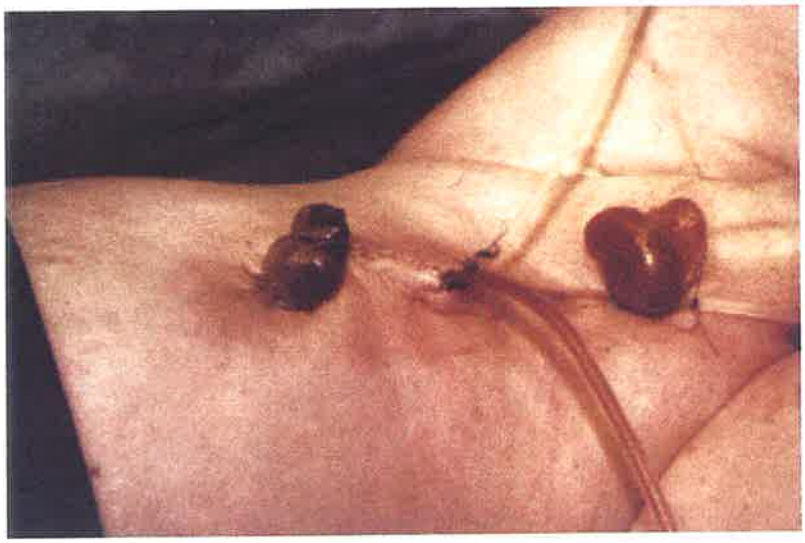


Figure 37: The umbilical stump in position at the end of the procedure that has closed the bladder exstrophy and repositioned the umbilical cord.

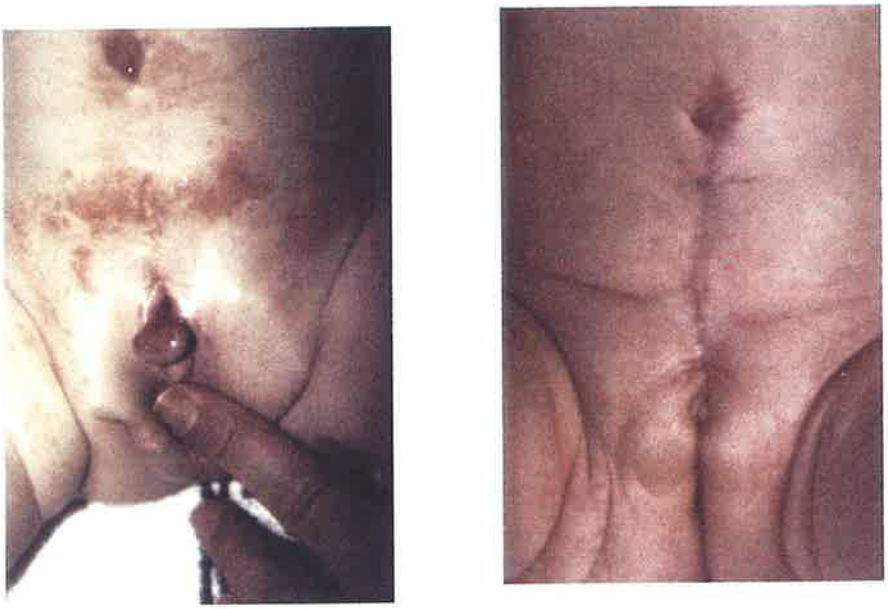


Figure 38: The healed umbilicus a male (A) and a female patient (B).

Ansell [103] advocated the early closure of bladder exstrophy, but did not transfer the umbilical cord to its normal location, which was first reported by Hanna in 1986, with good results [192]. However, Duckett and Caldamone, commenting on the 14 cases in which they used paraexstrophy skin flaps as part of the bladder reconstruction, mentioned one in which they had preserved the umbilicus. They blamed the loss of the paraexstrophy flaps on umbilical preservation and subsequent sepsis. They, therefore, abandoned preservation of the umbilical cord at the time of the primary bladder repair [124].

Sumfest and Mitchell [158] used a tubularised skin flap as part of a continent diversion procedure to create an umbilicus in 12 older patients; Gearhart in an editorial commenting on their operation supported the Hanna neonatal technique. Hanna and Ansong also provided a method of omphaloplasty in older patients having urinary reconstruction using a Y-V plasty [193]. In these older patients, either having a delayed closure or the latter stages of urinary tract reconstruction, an alternative approach is the use of 10mm wide laterally based advancement flaps which are attached to the deep fascia, a manoeuvre which is facilitated by excising cranial and caudal triangle of skin. This technique avoids the disadvantage of the Sumfest and Mitchell procedure [158], which results in a scar which is not in the midline, and gives a more uniform shape to the umbilical defect than the Y-V plasty of Hanna and Ansong [193].

Advantages and Disadvantages

Umbilical transposition produces a more normal appearing umbilicus than other techniques, but has a little increased risk of sepsis and the possible development of an umbilical hernia.

Comment

The prospect of a favourable outcome for the more complex problems encountered by the bladder exstrophy patients has necessitated refining the management of the less important aspects of their care. Obviously, careful attention to the technique of closure of the bladder is of paramount importance in the treatment of the neonate undergoing an early bladder closure, but provided the umbilical vein and arteries can be safely mobilised, preservation of this natural focus for development of the umbilical cicatrix can also be achieved.

Phimosis: is Circumcision Necessary?

Introduction

The prepuce is a vestigial structure that, at birth, is nearly always adherent to the glans and non-retractable [194]; the adherence is because of a common layer of squamous epithelium between the glans and the inner surface of the prepuce [195]. Thus, the foreskin completely clothes the glans during the years when the child is incontinent, protecting the glans against injury by minimising contact with sodden clothes or nappies [194].

World-wide, only one in seven males is circumcised [195], a rate which varies from country to country. In Australia and Canada, 40% of boys are circumcised, 80% in the United States of America and only 6% in the United Kingdom [196]. Non-surgical indications are common and include religious, cultural and social reasons. Thus, it is not surprising that circumcision is the oldest and most performed surgical procedure in the world [197]. However, the circumcision rate in Australia has declined over the last decade, partly due to the statements by the Australian College of Paediatrics and the American Academy of Paediatrics that neonatal circumcision is not medically indicated [196]. This is supported by Metcalf *et al.* who argued that good penile hygiene offers all the advantages of routine circumcision without the risks associated with surgery and anaesthesia [198]. As a result, circumcision for non-medical, other than religious reasons, is not encouraged [196]; exceptions include recurrent balanitis and paraphimosis, which are considered relative indications, and balanitis xerotica obliterans (BXO) which is usually, but not always, regarded as an absolute indication [196,199]. The most common stated medical indication for circumcision is phimosis; however, the definition of this condition is obscure in most publications.

What is Phimosis?

The normal prepuce gradually becomes detached from the glans, and retractable, as keratinisation of the epithelial layers occurs, aided by smegma which helps dissect the space between the glans and foreskin, preventing re-adherence [195]. Gairdner [194] and Oester [200] found that the prepuce is retractable in only 4% of newborn males, 20% at six months, 50% at one year, 90% at three years and 99% at 17 years of age. Therefore, a certain proportion of any age group has a non-retractable foreskin, especially before two years of age [201]. Little is known or written about the aetiology of true phimosis, despite the supposed frequency of phimosis varying from 4% to 10% [202].

In 1948, Winsbery-White defined phimosis as the congenital or acquired narrowing of the preputial opening, characterised by a non-retractable foreskin without adherence which can lead to retention of secretions under the foreskin with consequent irritation and balanitis. Interference with micturition can, on rare occasions, lead to subsequent backpressure effects on the bladder, ureters and kidneys [203].

Winsbery-White's definition of phimosis does not seem to be sufficiently exacting: a more precise, practical guide to the difference between a non-retractable and a phimotic foreskin is as follows; when the normal, but non-retractable infant foreskin is examined, attempted gentle retraction results in the distal part of the foreskin pouting, with the narrow portion of the foreskin being proximal to the tip of the prepuce (Fig. 39). Forced retraction of such a foreskin can result in longitudinal splitting, as is well demonstrated in the Figures presented by Stenram *et al.* [204]. However, true phimosis produces a cone shaped foreskin during the same gentle retraction manoeuvre, with a fibrotic, circular band which forms the most distal and narrowest part of the prepuce (Fig.41). Minor narrowing of a retractable foreskin, which does not split during attempted complete retraction, probably does not require intervention (Fig.40).

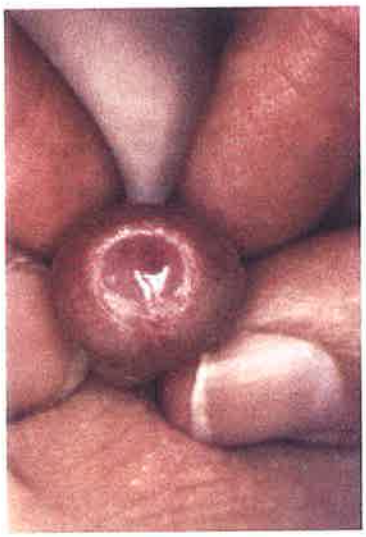


Figure 39: A ***normal, non-retractable infant foreskin,*** which has adherence of the foreskin to the glans.

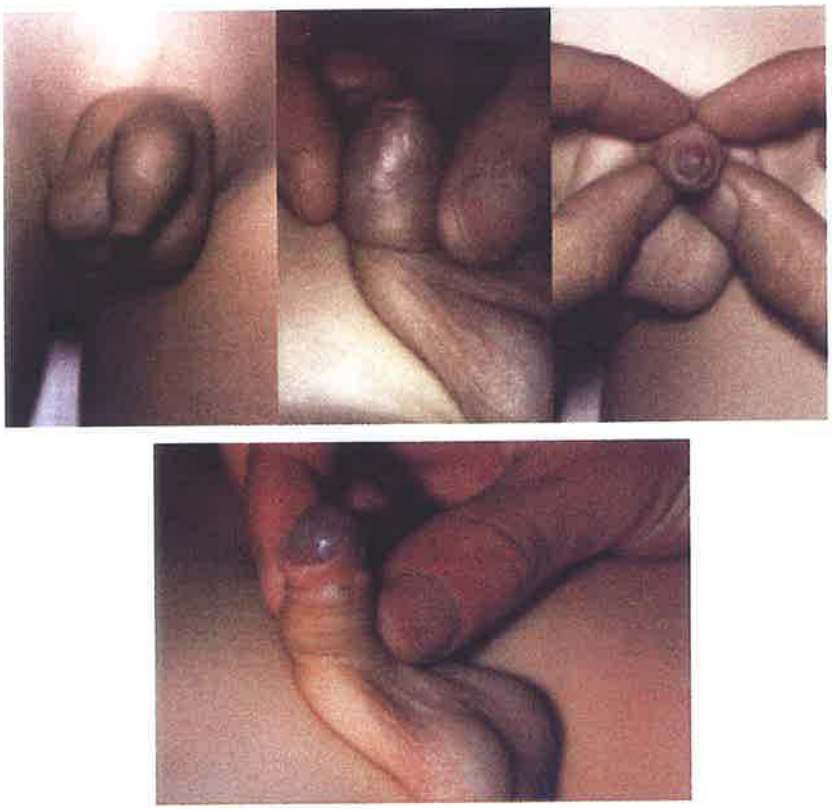


Figure 40: A foreskin with a minor phimosis. No narrowing is seen without attempted retraction of the foreskin (A). The narrowing (B) easily stretches (C) to allow full, gentle retraction (D).

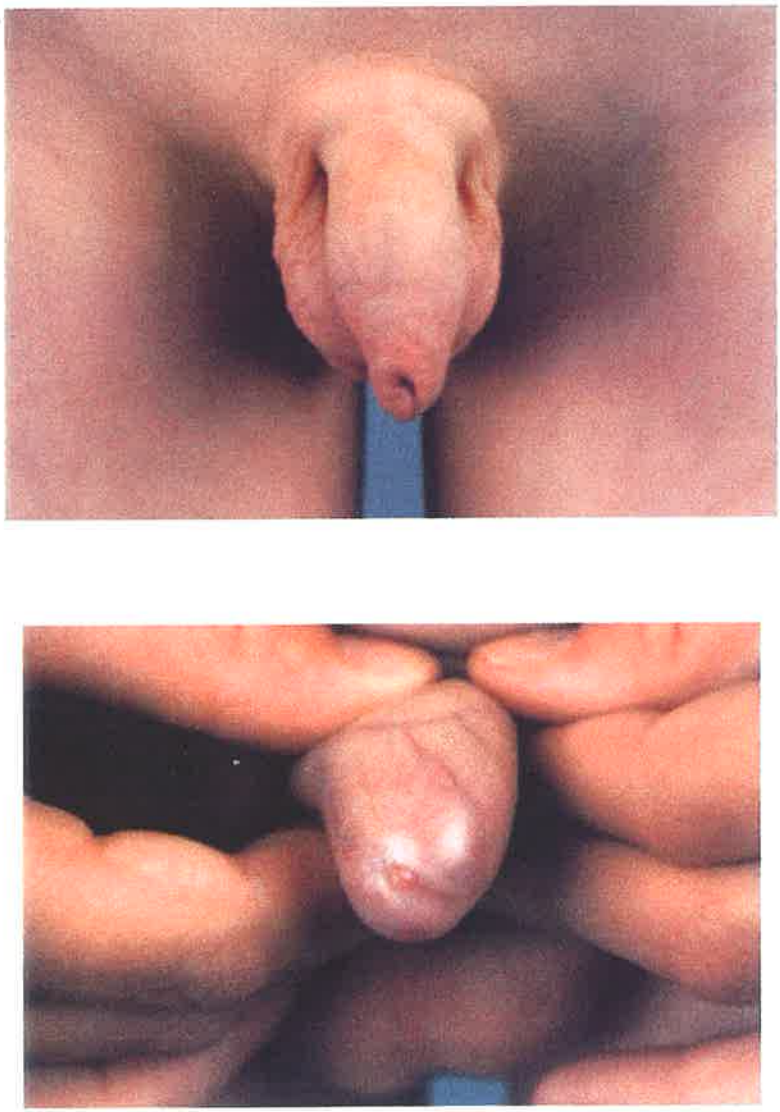


Figure 41: A case of true phimosis. The non-retracted foreskin can look normal (A). The partly retracted foreskin reveals a conical shape with a pinhole meatus (B).

Confusion about the definition of phimosis is highlighted by the study of Griffiths and Frank, who found that only 30 of 128 boys with a medical reason for referral to a Paediatric Urologist had true phimosis (although they did not give an exact definition). They suggested that ballooning and non-retractability is often over interpreted [205]. Obviously, the referring practitioner may have used the label of phimosis to facilitate referral, thus further confusing the debate on the appropriate management of phimosis.

Consequences of Phimosis

In 100 cases of balanitis, Escala and Rickwood recorded recurrent attacks in only 36%, leading them to suggest that circumcision was only indicated in recurrent cases. They reported that, while 4% of boys will develop balanoprosthitis, only 1% have true phimosis [206].

Phimosis, in the short-term, can cause ballooning during micturition, discomfort with voiding, infection and pain on erection. In the long-term, BXO and even obstructive renal damage disease can occur; also carcinoma of the penis is thought to be more common when phimosis is present. Poynter stated that BXO can develop at any age and appears to have little relationship to circumcision status [207]; it appears, however, to be rare below the age of five and cure is often affected by circumcision of a phimotic foreskin [208,209]. In examining multiple circumcision and post mortem specimens, Reddy *et al.* suggested that a non-phimotic foreskin with regular hygiene had no increased risk of penile carcinoma [210]. Further argument against circumcision for the prevention of carcinoma of the penis is given in two letters in the Canadian literature, based on the rarity of penile cancer [211,212]. Circumcision is considered by many to be the least problematic alternative way of managing the foreskin. This view is not supported by a New Zealand study of up to eight years follow-up in over 500 boys: penile problems occurred in 11.1% of uncircumcised boys and 18.8% of the circumcised children [213].

Techniques of Phimosis Management

Operative

A number of studies have looked at different ways of surgically managing the foreskin, including routine circumcision, sutureless circumcision [214]; modifications of preputial-plasty [215-217], dilatation with a balloon [218] or artery forceps [219] and forced retraction with local, general or no anaesthetic [220-222] have all been advocated. Unfortunately, none these studies have been prospective randomised trials.

The majority of circumcisions in America are carried out as a neonatal procedure, prior to discharge from the maternity ward. In a 10-year review of circumcision on 5,521 of 5,882 male births from 1963 to 1972, Ansell recorded the treatment outcome for a group circumcised with either a plastibell or the Gomco device. Complications included a 1% haemorrhage rate and less than 1% other complications [223]. In contrast, Persad found an 8% meatal stenosis rate with operation before one month of age [224] and overall complication rates ranged from 0.2% to 0.6%, reaching 35% if minor complications were also considered [195]. Some of these include ulceration of the meatus, infection (local and systemic), haemorrhage, removal of excessive or inadequate amount of skin, wound dehiscence, electrocautery damage to the penis, partial amputation and complications from anaesthesia [195,196,204,225,226].

Non-Operative

Steroid cream is a painless, less complicated and more economical alternative to circumcision in the treatment of phimosis. Wright had success of 80% (89 of 111 boys) with 0.05% Betamethasone [227] and Kikiros *et al.* showed improvement in 33 of 42 boys (78%) with 0.05% Betamethasone, 18 of 21 boys (86%) who used hydrocortisone, but they felt that Betamethasone worked more quickly [196]. Another steroid cream, 0.05% clobetasol propionate, has also been used successfully (70% - 54 boys) by Jorgersen and Svensson [228]. It is widely held that BXO does not respond to topical steroid treatment

[196,227], and circumcision is indicated. However, recent cases, presented in the series below would suggest that the combination of steroid cream and four quadrant incisions could produce a normal foreskin, despite the presence of BXO.

Prospective Study of Steroid Treatment

Materials and Methods

A total of 65 boys were treated with one of two different steroid creams from November 1992 until April 98. The boys presented with a range of conditions as shown in Figure 42: the age distribution, severity and treatment are shown in Figures 43-45. Those presenting with VUR were found to have phimosis at the time of examination after an MCU; the indication for investigation was either UTI or prenatal hydronephrosis: balanitis was not part of the initial illness in this group. Phimosis symptoms occurred in 30 patients and consisted of ballooning of the foreskin on micturition, non-retractability of the foreskin and/or balanitis. Those with an incidental diagnosis were usually identified to have phimosis at the time of consultation for an undescended testis, or requested examination of a sibling.

Fifty-eight boys have had adequate follow-up, some of whom had a second course of treatment and in some of whom therapy was changed from Hydrocortisone to Betnovate for their second course of treatment.

The age distribution is presented in Figure 43: the largest group were those who were less than six months of age, the next most common age group were those between four and five years of age: the severity of phimosis, according to the age groups, is also shown in Figure 43. Nine had a severely stenosed meatus, 23 moderately closed foreskin orifice and 33 had only a minor degree of narrowing of the distal end of the foreskin. The size of the hole and the degree of fibrosis were documented as individual data-points, such that some patients with BXO, i.e., a severe degree of inflammation

inflammation and scarring, had only a minor degree of narrowing of the preputial orifice: eight boys had BXO (Fig.44).

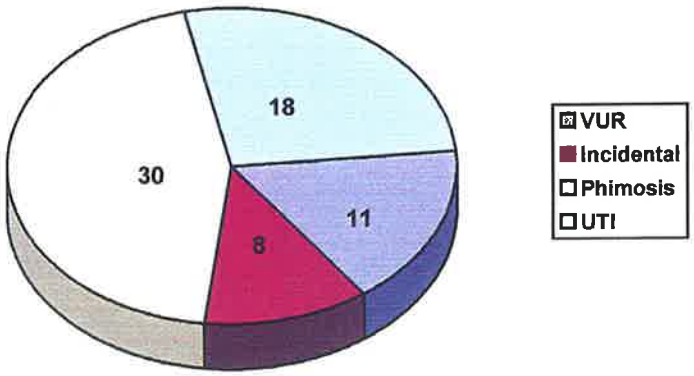


Figure 42: The nature of the presentation of boys with phimosis. The presentation of three cases was not recorded.

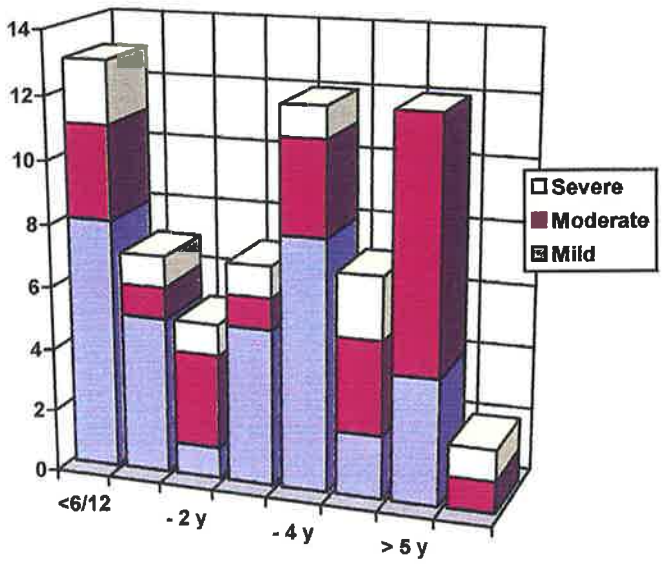


Figure 43: Age distribution and degree of severity of the narrowing of the preputial orifice.

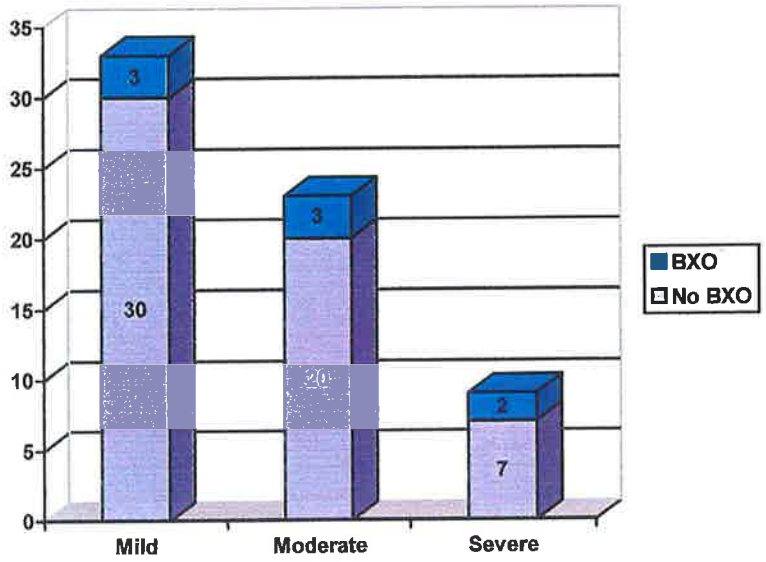


Figure 44: The graph shows the degree of preputial narrowing and the presence of Balanitis Xerotica Obliterans (BXO).

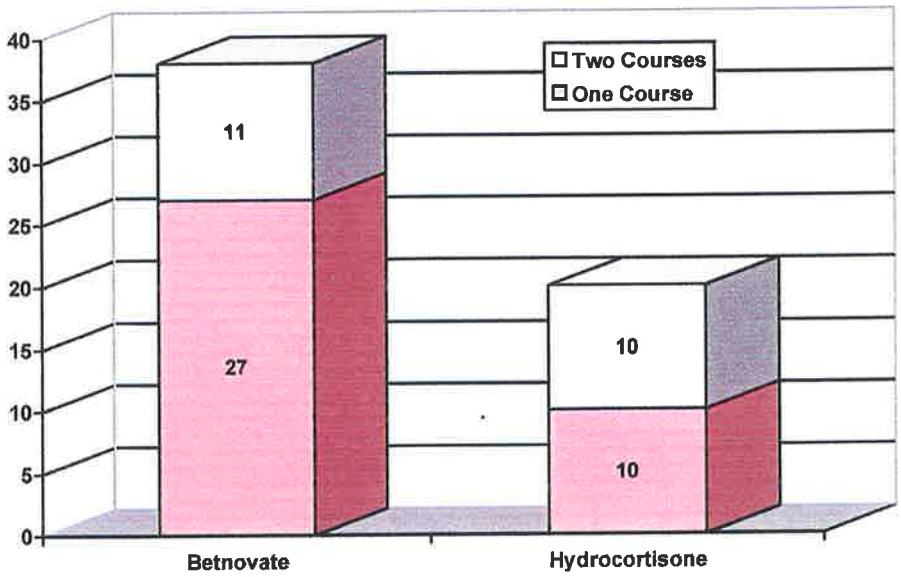


Figure 45: The number of patients who had a second course of steroid treatment for each of the drug treatment groups.

The parents were given either 1% Hydrocortisone or 0.05% Betamethosone (Betnovate) to apply to the partly unrolled foreskin. This they were to achieve by a gentle attempt to retract the foreskin, thus reducing the amount of redundant skin distally, to ensure the application of the cream to the narrow portion of the prepuce. Once the prepuce had become retractable, the parents were encouraged to continue to retract the foreskin when the boy was bathing and when he passed his urine or had his nappy changed (depending on the age of the patient). The parents were shown how to apply the steroid cream to the foreskin at the time of the initial consultation; they were asked to demonstrate their ability to place the foreskin in the optimal position for maximum therapeutic benefit of the steroid cream application. A six-week course was prescribed, after which the child was reviewed. If the foreskin was not easily retractable beyond the glans, the treatment was continued for a further six weeks. At the second consultation the parents were again instructed on the correct method of application of the cream, if necessary. If the foreskin had become retractable the parents were again reminded of the need to retract the foreskin when the child was in the bath and at the time of micturition, for a further three months. Figure 45 indicates that number of patients treated with each drug and the number that had a second course of that drug.

Those boys with BXO, who had persistence of the narrowing of the foreskin, underwent an operation that consisted of an incision of the foreskin orifice in four quadrants of the narrow portion of the prepuce.

Results

Most of the boys who had Betnovate required only one course of six weeks, but in many of those who used 1% hydrocortisone phimosis failed to resolve (Fig.46). Although, some required a second course and one required a course of Betamethasone when the standard first hydrocortisone course failed. All boys were reviewed at least two weeks after the cessation of the steroid cream. Failure may have occurred in some because the parents had not understood that the foreskin should be "unrolled" to reveal the narrow portion before the steroid cream was applied. In those who failed, or who recurred, the foreskin was treated with 0.05% Betamethasone, which seemed more effective if used as the initial management. Three of those treated with Betnovate subsequently went on to have a circumcision, whereas three of 20 with 1% hydrocortisone required operative treatment – resorting to further non-surgical treatment was a significant factor to these parents.

Boys with BXO were more likely to require a subsequent or longer course of the steroid cream application.

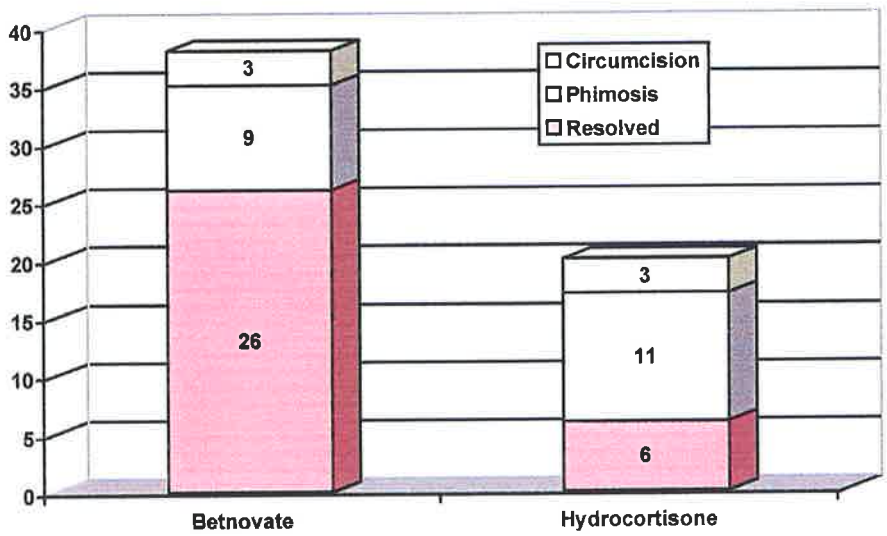


Figure 46: The outcome data, including recurrence, resolution or a subsequent circumcision, divided into the two different primary treatment groups.

Discussion

This study has indicated the high rate of success for the use of steroid cream in the management of boys with true phimosis, particularly if the parents are specifically instructed to unroll the foreskin before the cream is applied to the fibrotic part of the foreskin. If the parents are not instructed and specifically shown the technique of application, not only is the medication applied to the incorrect part of the foreskin, but also the additional therapeutic step of gentle retraction, thus stretching of the narrowing portion of the prepuse, does not occur. The risks of paraphimosis should be explained and the need to return the foreskin to the neutral position if retraction beyond the coronal groove is achieved. Importantly, the patient should be reviewed to highlight the need to maintain the diameter of the orifice of the foreskin with continued foreskin "physiotherapy": a second course of steroid cream was often necessary because of the failure of the parents to comply with the ongoing need for retraction of the foreskin.

What then is the action of steroids on the phimotic foreskin? There may be two possible mechanisms by which the steroid cream resolves the phimosis. Firstly, the anti-inflammatory and immunosuppressive effect (Figs. 47+48): according to Kragballe, corticosteroids stimulate the production of lipocortin [229]. The lipocortin formed inhibits the activity of phospholipase A2, which releases arachidonic acid, the precursor of prostanoids and leukotrienes (mediators of skin inflammation), from phospholipid [229]. Corticosteroids also inhibit mRNA responsible for interleukin-1 formation and several types of T-lymphocyte activities, including interleukin-2 production and mitogen-induced T-cell proliferation [229]. These actions of corticosteroids on arachidonic acid metabolism and interleukin-1 formation produce the anti-inflammatory and immunosuppressive effects. Corticosteroids not only inhibit early phenomena of inflammatory process (oedema, fibrin deposition, capillary dilatation, migration of leukocytes into the inflamed area and phagocytic activity) but also its later manifestations (proliferation of capillaries and fibroblasts, depletion of collagen and cicatrisation) [230]. Secondly, the skin thinning effect

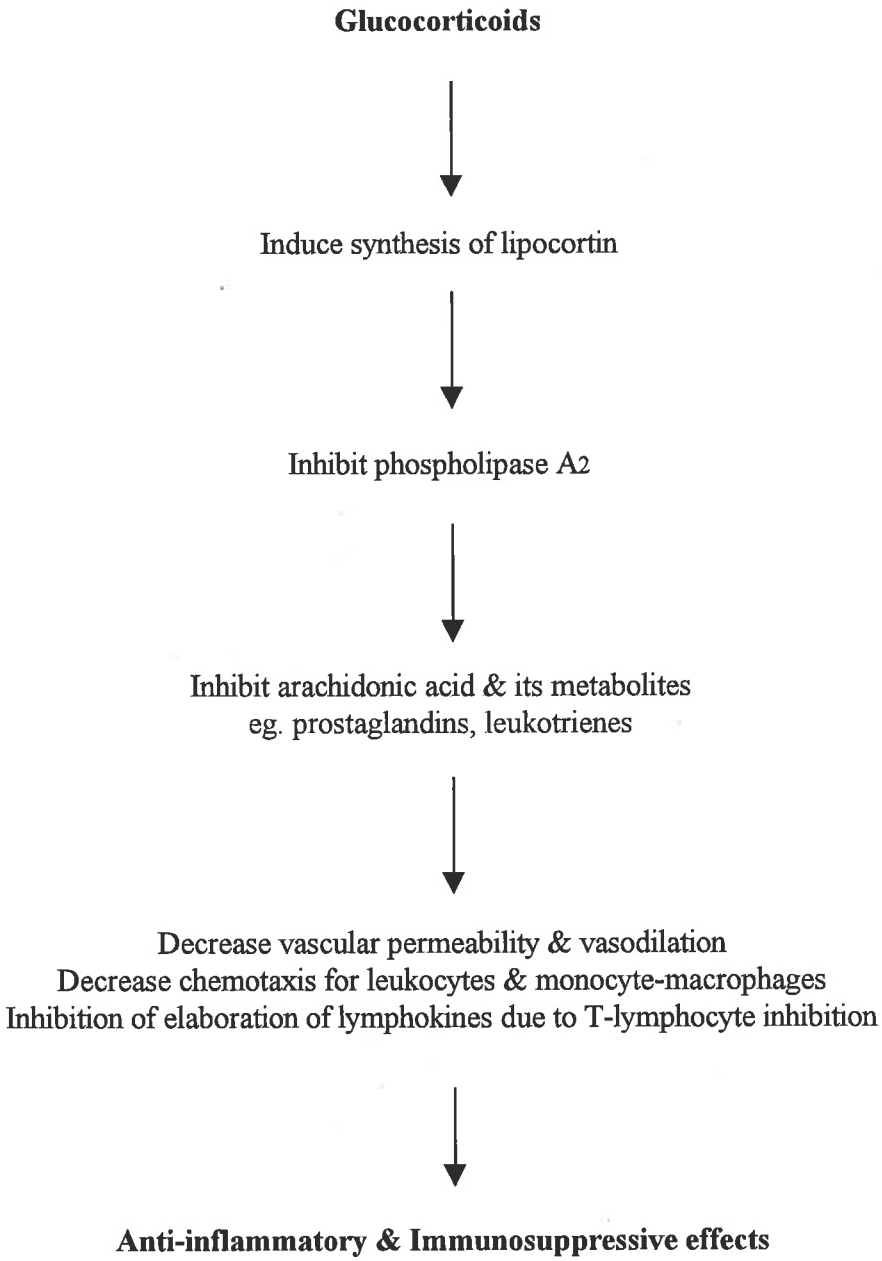


Figure 48: Anti-inflammatory and immunosuppressive effects of steroid on the skin.

Summary

Effective steroid treatment of phimosis is becoming increasingly important. Firstly, the majority of boys in Australia are now uncircumcised; thus the uncircumcised state is becoming the accepted norm. Secondly, in the current economic climate of finite health dollars, the cost savings from steroid treatment should be considered. Thirdly, the above steroid treatment does not appear to have significant complications and is painless. From the published results, as for the outcome of this study, 0.05% Betamethasone appears to be more effective than 1% hydrocortisone and more effective than Vaseline [234]. It would appear that daily retraction of the foreskin is necessary after initial success has been achieved and circumcision seems indicated when topical steroid treatment fails. However, a double-blind randomised controlled study of different steroids and placebo, with a research protocol which includes a clear definition of phimosis as part of the research protocol, is needed; a view which is supported by others [235]. Further research to elucidate the mechanism of corticosteroid action on the foreskin would also be appropriate.

Priapism in boys

Introduction

Priapism is a rare condition in childhood, which occurs most frequently in association with sickle cell disease, where conservative management is often appropriate. The following five cases, with differing aetiologies, and a review of the literature, highlight the need for aggressive therapy when conservative measures are not immediately curative.

Priapism is an abnormal penile erection, which is persistent, painful and generally unassociated with sexual arousal. The erection may be intermittent and can be distinguished from normal erection by turgidity of the intercommunicating corpora cavernosa, in combination with a flaccid corpus spongiosum and glans penis.

The term comes from the name of the Greek God, Priapus, the son of Aphrodite, the Goddess of sexual love, beauty and feminine fertility, agricultural production and good hunting, and was generally held to be responsible for producing beneficial results whatever the occupation being practised.

Several workers have expressed differing views as to the possible mechanism of erection and opinions on the pathophysiology of priapism have varied: Bochdalek (1854) [236] and Waldeyer (1899) [237] considered venous occlusion to be the prime factor, whereas Conti (1952) and others this century, have argued that tumescence is due to increased arterial inflow [238]. Recent experiments in animals have found corpora cavernosa tumescence to be the result of active relaxation of the sinusoidal spaces, active arteriolar dilatation and active venous outflow constriction acting in combination, rather than a single factor [239-241].

The understanding of the management of priapism has improved significantly over recent years from these animal studies of the physiology and from the evidence accumulated from the increasing use of intracorporeal agents to produce an artificial erection. For example, it is well recognised that papaverine, a smooth muscle relaxant, and phentolamine, an alpha adrenoreceptor antagonist, are both able to induce both erection and priapism when injected into the corpora cavernosa. Agents such as the alpha adrenoreceptor agonist, Metaraminol have, therefore, been used in treating persistent erection [242]. All of these agents, when used experimentally, have led to the recognition of the existence of two subgroups of priapism, one due to a low flow state and the other due to increased blood flow through the penis [243]. Thus, when the turgidity of the penis is less than maximal, conservative measures should be instigated, but when there is a persistently fully turgid penis, or when conservative measures fail to produce detumescence, aspiration, intracorporeal alpha adrenergic injections [242], embolisation of an arteriocavernosal fistula [244] or a caverno-saphenous shunt [245] may be necessary.

Fabry's disease (alpha-galactosidase A deficiency) is a lysosomal storage disorder inherited in an X-linked recessive manner and is characterised by specific cutaneous lesions, painful crises, and, in later life, progressive renal failure; ***Case 4*** was referred with an unusual presentation in a hemizygous boy. ***Case 5*** was seen following an episode of perineal trauma, which has only been rarely described in boys [246,247], and only in one previous case has an arteriocavernosal fistula been embolised in such a paediatric patient [246]. All six cases managed by the author are presented in detail and the unusual features are discussed.

Case Reports

Case 1: A 10-year-old boy awoke on the morning of admission with a painful erection. He had a three-day history of rash over his toes and buttocks, as well as mouth ulcers. On examination, he was found to have a small number of petechial lesions over his buttocks and toes, palatal ulcers and mild cervical lymphadenopathy, and his penis was turgid, except for the lax corpus spongiosum and glans. He also showed signs consistent with thoraco-lumbar junction transverse myelitis, which lasted only a few hours. Apart from isolating an Echovirus 14 from his urine, all other investigations were normal, including a full blood examination, lumbar puncture and myelogram. Local cold application and analgesia were the only treatments administered, with a resultant gradual resolution of the boy's priapism over 48 hours. At follow-up, his parents reported that he was experiencing normal erections, and he was neurologically normal when examined.

Case 2: This previously well, eight-year-old boy presented with a seven-day history of incomplete priapism. He had only mild penile discomfort and no difficulty in passing urine. On examination (including rectal examination) he was normal, apart from the finding of priapism. Unlike the other four cases, his penis was only slightly tender. Red cell morphology, white cell count, pelvic Xray, coagulation profile, mid-stream urine and a nuclear magnetic resonance scan of his spine, were all normal, except that his Coxsackie B_2 antibody titre was positive, then lower in the convalescent serum. Therapeutic penile aspiration produced a near-flaccid penis, which then continued to resolve over the following seven days; his erections have since been normal.

Case 3: Five days after an uneventful appendicectomy, this two-year-old boy presented with signs of pelvic sepsis. He had difficulty in passing urine and had a continuous painful erection with fluctuating levels of turgidity. He was treated with intravenous antibiotics, phenergan

sedation and cold application. The degree of penile turgidity declined over the following two days, after which he was discharged. On review, his parents reported him as having normal erections.

Case 4: An 11-year-old boy had been diagnosed as having Fabry's disease two years before presenting with a painful, persistent erection of 12 hours' duration. He was not on any medication. This boy, who is recorded as IV.16 in Figure 49, which represents his extended Caucasian family in which several members have had Fabry's disease. The diagnosis was biochemically established in the above patient at the age of two years when the family was investigated, after the diagnosis had been made in his two maternal uncles. All individuals shown in the pedigree have been enzymatically assessed by leucocyte studies, except for the individuals I.1, I.2, III.4, IV.8, IV.9, IV.10 and IV.17. The relative, I.1, is said to have died of Bright's disease, but further details are not available. Prior to this presentation, his only clinical problem had been occasional mild, short-lived leg aches.

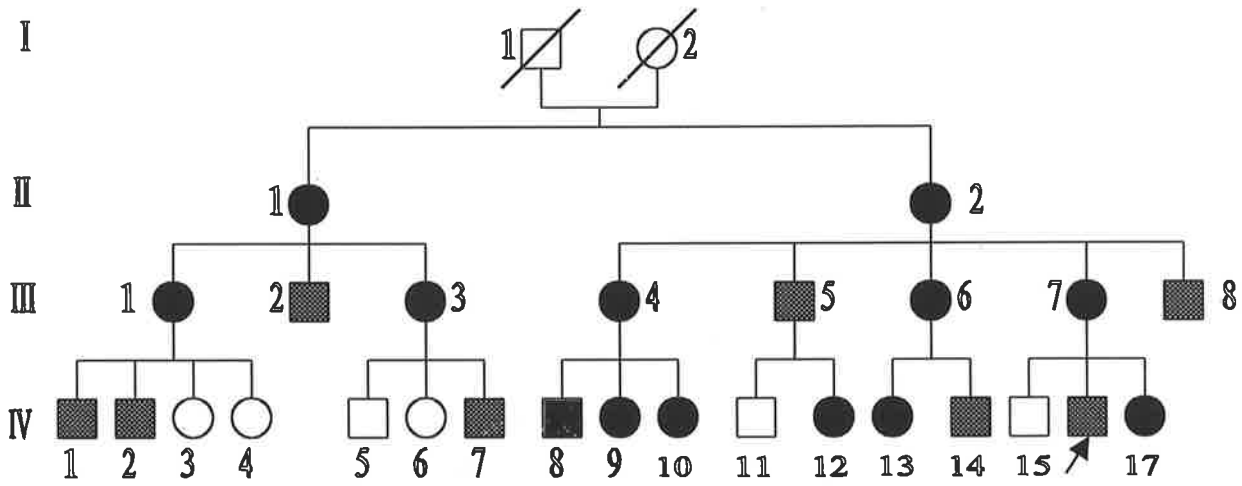
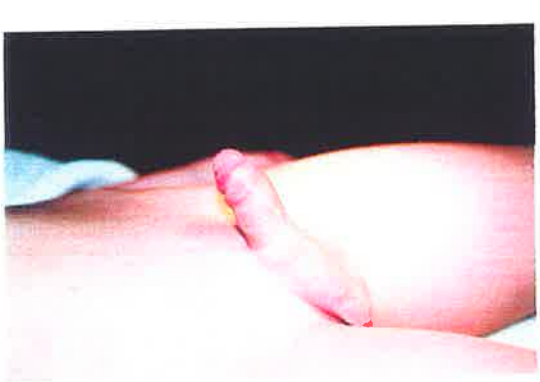


Figure 49: Pedigree data of family with Fabry's disease. Hemizygote. Obligate carrier on pedigree. Borderline heteroxygote enzymatically/obligate carrier of pedigree. Probable heterozygote. Clinically and enzymatically normal. Clinical/biochemical status unknown.

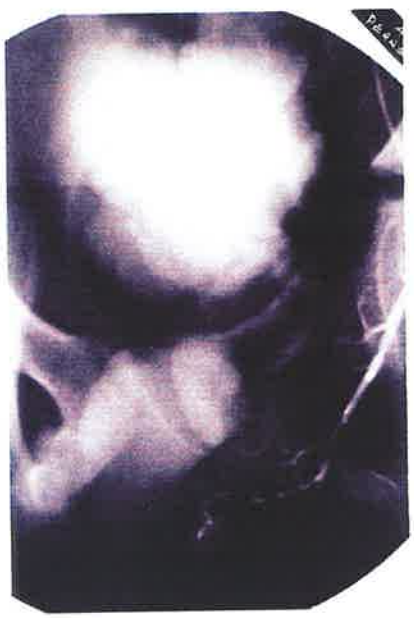
On examination, he was distressed, with turgid corpora cavernosa; he was a small boy (height and weight below the 3rd percentile for age), in the early stages of prepubertal sexual development, and was in obvious severe penile discomfort. The remainder of the examination was normal; in particular he had no clinical features of Fabry's disease (ie, corneal opacity, angiokeratomas, sweating abnormalities, hypertension, and haematuria). Extensive investigation revealed no aetiological factor for his priapism apart from the enzyme deficiency previously noted. An unsuccessful attempt at cavernosal aspiration was carried out in the casualty department; 24 hours after the onset he was given a general anaesthetic, the corpora cavernosa were aspirated and an attempt was made to instill one ml of Metaraminol, but rapid refilling of the corpora prevented adequate instillation and only partial, temporary detumescence was achieved. Repeat aspiration and urinary catheter insertion were performed 16 hours later, with a greater degree of detumescence. At each aspiration, initially a small volume of dark blood was obtained, after which bright blood and a high rate of inflow into the penis were noted; 48 hours after onset, further incomplete detumescence was produced by a third series of aspirations. An Ebbehøj cavernosa shunt was therefore performed (a narrow blade incision being made in the dorsum of the glans and extended into the corpus cavernosum on each side; the glans was then sutured). Over the following 12 hours, his erection returned to its initial turgid state. After a total of 72 hours, a second attempt at an Ebbehøj shunt failed to produce complete detumescence and under the same anaesthetic, a right caverno-saphenous shunt was carried out. Satisfactory but incomplete detumescence resulted, which resolved slowly and he was discharged 14 days after admission. A penile doppler study and US examination four months after discharge showed flow through the saphenous vein shunt and no fibrosis in the corpora. However, the cavernosal arteries could not be visualized and he had not experienced any subsequent erections. Surgical closure of the saphenous vein shunt was to be considered subsequently.

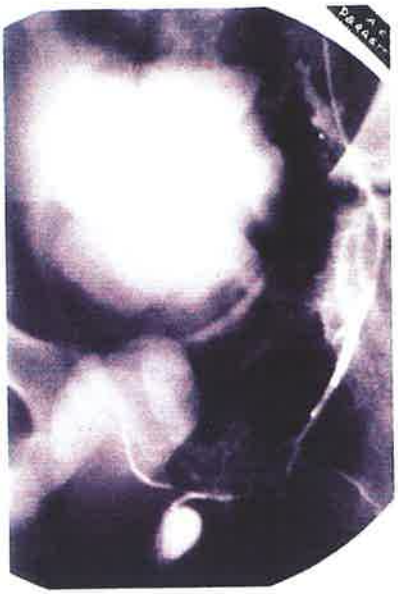
Case 5: A seven-year-old boy presented five days after a fall-astride injury. Bruising was noted immediately after the injury, but his priapism was not evident until the following morning. At the time of presentation his glans was flaccid, his corpora cavernosa turgid and his scrotum slightly bruised (Fig. 50); however he was not in pain and he was voiding normally. He was sedated with diazepam and pethidine, and an ice pack was applied, with no response. A general anaesthetic was administered and a caudal injection given, again with no effect on penile turgidity. The intracorporeal pressure was 39 mmHg and the penile blood gases were arterial. The aspiration and irrigation with heparinised saline failed to produce detumescence; therefore 0.25mg Metaraminol was injected through the glans, into the corpora cavernosa. An excellent, but fleeting response was seen, so two further injections were given, with a lesser response with each. Therefore, an attempt was made to create a communication between the glans and the corpora cavernosa by lacerating with a 19 FG needle via the glans; this was not successful but, as the penis was easily detumesced manually, no further treatment was given at this stage. Post-operatively he was commenced on the antiplatelet agent, aspirin, and oral antibiotics. Over the next 24 hours he developed increasing turgidity, bruising and oedema of the distal penile shaft, therefore an arteriogram was performed via the right femoral artery, with the intention of embolising an arteriovenous fistula. The left common penile artery (Fig. 52) fed an arteriocavernosal fistula, and detumescence occurred while the arterial catheter was in place. The erection partially recurred despite successful placement of autologous clot initially, then gelfoam (Fig. 53). Over the following eight hours the penile turgidity returned to the pre-arteriography state; therefore a further general and caudal anaesthetic were administered. The penis again contained blood with arterial gases levels at a pressure of 39 mmHg. Transient detumescence was obtained by two further injections of 0.25mg Metaraminol, but with time the penis returned to the same state after each treatment, therefore a left sided Grayhack, cavernosum-saphenous vein shunt was established [245]. An adequate length vein with a patent anastomosis was achieved. The corpora were still distended, but softer and easily emptied of blood following this procedure. His penis detumesced to a

emptied of blood following this procedure. His penis detumesced to a stuttering priapism over the next two days (Fig. 51), and a doppler study two weeks later showed poor flow in the venous shunt, but excellent flow in the corporal arteries on each side. He was noted by his parents to have normal nocturnal erections.



Figures 50 + 51: Priapism in the boy with an arteriovenous fistula, with detumescence after the embolization and the shunting procedure.





Figures 52 (left): The penis is partly detumesced with the angiography catheter in situ. An area of extravasation was shown confirming the presence of an arteriocavernosus fistula. **Figures 53 (right).** Following autologous clot and gelfoam embolisation, a stagnant column of contrast is seen in the left common penile artery and pooling at the site of the leak.

Case 6: A mildly mentally retarded boy presented to the emergency department of the Sunshine campus of the Royal Children's Hospital Melbourne for the second occasion in 18 hours, complaining of penile pain. The blood film was normal, there was no family history, rectal exam was normal, and he was not on any drugs; there had not been any history of perineal trauma.

On this occasion the Paediatric Urologist was contacted, the application of cold compresses was commenced and transfer to the operating room arranged. He was noted to have markedly turgid corpora cavernosa and a lax glans. He also was seen to have a candida rash on that half of his glans not covered by the partly phimotic foreskin. A general anaesthetic was administered, as was a caudal injection of bupivicaine. The glans on each side was aspirated of old blood, with a virtually unrecordable pO_2. The corpora were lavaged with a dilute heparin solution, until high-pressure arterial blood could be aspirated. Metaraminol 0.1 mg was then injected on two occasions, with the penis emptied of as much blood as possible prior to each of the injections, which proved difficult because of the high-pressure inflow. The improvement in the degree of tumescence was minimal, and fleeting. Several passages of a 19G needle through the spongiosus and cavernosus on each side, each through a single stab hole on either side of the glands, produced some improvement after each manoeuvre. The success of each attempt lasted only several minutes, although the penis could be at least minimally emptied by firm manual compression. Further heparin was used intermittently over the four hours taken to treat and wait after each new episode of treatment, because initially a minimal, variable response was achieved. The turgidity of the penis ultimately returned to the same as at the time of presentation; therefore a right Grayhack, corporosaphenous shunt was performed. As the clamps were removed the detumescence was near complete; the penis was easily emptied, but the penis regained a worrying degree of turgidity over the next 15 minutes. He subsequently had an epidural infusion whith no early response but achieved gradual detumescence over five days. Six weeks later he had not had any erections and the cavernosal vessels were recorded as

having no flow on doppler US. Six months later he was again reporting nocturnal erections and the flow in the shunt appeared to have ceased.

Discussion

Priapism is associated with a wide range of causes, with an increasing incidence in adults due to the use of intracorporeal agents to facilitate artificial erection. However the disease remains rare in children (Table 7), except where sickle cell anaemia is common [248,249]; however, despite its rarity, the condition has even been recognised in the neonate [250].

Idiopathic
- Haematological
  - Sickle cell disease
  - Glucose phosphate isomerase deficiency
  - Abnormal platlet aggregation
  - Hyperfibrinogenaemia

Local Malignancy
- Infection
  - Pelvic cellulitis
  - Mumps
  - Other virsus

Perineal Trauma
- Iatrogenic
  - Antihypertensives
  - Haemodialysis
  - Anticoagulants
  - Intravenous fat emulsions
- Neurological
  - Spinal injury
  - Transverse myelitis
- Other
  - Diabetes
  - Amyloidosis
  - Fabry's disease

Table 7: Aetiology of Paediatric Priapism.

Because of the wide range of causes of priapism (Table 7), it is essential that patients have a thorough history recorded and complete physical examination; also appropriate investigations should be performed before a diagnosis of idiopathic priapism is made. The experience with the six boys suggests the need to add evidence of a recent viral infection to the list of potential causes.

It has hitherto been considered that a paediatric patient usually has spontaneous resolution of his priapism, and intervention is not necessary. However, a review of the literature shows an impotence rate of 14% in a group of 70 patients taken from various published reports [250-256]. Two percent (1/50) were impotent after conservative management alone, while almost 50% (9/19) were impotent after surgical intervention. A study of 207 patients treated from 1973-1990 in Finnish hospitals showed that only 9/79 (11%) of patients treated conservatively gained relief of the priapism, whereas 59% of the surgically treated group did. The youngest patient was 8 yo. Early treatment was successful with injection therapy, and small shunts were successful if utilized early [255].

It is reasonable to contend that the high impotence rate after surgery is due to the delay in carrying out surgery in severe cases, which stems from the view that all paediatric priapism resolves spontaneously. This contention is upheld by the not-insignificant impotence rate in sickle cell boys with priapism (14%; 8/59). Two of six patients treated by the author resolved spontaneously, and the third settled following corpora cavernosa aspiration. In the fourth patient with long-standing underlying pathology and the fifth and sixth boy, detumescence was only achieved by surgical intervention.

Fabry's disease, the underlying disease in ***Case 4***, has been shown to be due to a marked reduction in alpha-galactosidase-A activity [257] which results in the accumulation of crystalline glucosphingolipids in the lysosomes of all body tissues and, in particular, in the

endothelial and perithelial cells of blood vessels, epithelial cells of the cornea, lens, renal glomeruli and tubules, autonomic nervous system ganglion cells, and cardiac muscle fibres. Neutral glycosphingolipids are important structural components of plasma membranes, possibly subcellular organelle membranes and transport complexes such as low-density lipoproteins. Their degradation is a result of the sequential action of specific exoglycosidases, most of which are lysosomal in location [258]. The patient with Fabry's disease and priapism had no angiokeratomata, which although unusual is not exceptional, as there have been reports of patients with Fabry's disease with none of the typical skin lesions [259,260]. In addition, the skin lesions often evolve with age and thus were less likely to be seen in this boy.

During the current illness he developed a painful crisis for the first time. Typically, these crises can be precipitated by excessive exercise, fatigue, rapid climatic changes, and emotional stress and are characterised by agonizing, burning pain in the palms and soles that may last for days. Relief is usually achieved by narcotic analgesia alone.

Priapism has been reported in two adults as a consequence of Fabry's disease [261] and in a 12-year-old boy with Fabry's disease following treatment with phenoxybenzamine for supposed associated autonomic dysfunction [262]. It would appear that ***Case 4*** is the youngest individual with Fabry's disease recorded to have developed priapism. The priapism was not precipitated by any exogenous factors and could have been due to either vascular occlusion or autonomic dysfunction producing the high-flow priapism. The prognosis for normal sexual function in this boy remains uncertain.

It was decided to perform a cavernosum-saphenous vein shunt, rather than perform more radical vascular surgery in ***Cases 5 + 6*** [245]. Detumescence was achieved in both, and the shunt was noted to have closed spontaneously two weeks after surgery; good flow was seen in the corporal vessels and normal erections eventually returned in both.

Although the corpora of ***Case 4*** were not fibrosed on US, he did not have an erection during an 18-month follow-up period. The presence of flow through the shunt suggests that ligation of the fistula would restore potency.

Evaluation of the management of priapism indicates the previous lack of understanding of the pathophysiology of erection. However, some of the earlier techniques are still effective, eg cold applications [275], treatment of the aetiology (leukaemia, sickle cell anaemia), sedation and analgesia. On the other hand, systemic anticoagulants, fibrinolytics, hypotensive agents, regional and general anaesthesia, are now recognised as being relatively ineffective.

Direct corpora cavernosa aspiration, followed by the installation of heparin, streptokinase [276] or alpha-receptor antagonists, is now widely advocated, especially the use of Metaraminol [242,277].

Several surgical techniques have been used, including incision and drainage, and rectal diathermy [275], both of which are now not encouraged. Caverno-spongiosum shunts, advocated by Quackels (1964) [278] and Howe (1969) [279], have been largely replaced by glans cavernosal shunts created by a narrow blade (Ebbehøj, 1975) [280], Tru-cut needle (Winter, 1971, 1976 [281,282]), or either formal anastomosis described by Al-Ghorab, in 1978 [283], or by using a Kerrison rongeur as described by Winter in 1981 [284]. The most popular, more extensive procedure has been the caverno-saphenous shunt developed by

Grayhack in 1964 [245]. Other alternatives include the dorsal vein-cavernosa shunt (Barry, 1976 [285]) and embolisation of the internal pudendal artery with autologous clot [244,267]. The latter two methods have been widely reported. Ulman *et al.* described modification of the Winter technique which involved the insertion of an "angiocath" via the glans on each side for up to 48 hours. This produced a bridge between the glans and the cavernosa in a 14 yo thalassaemia boy with a 4-day history. Normal erections were observed 2 months after presentation [254].

Occasionally, a more radical surgical approach is taken, such as that used by Upadhyay *et al.*, who, in a 19 year old black male, following 28 episodes of priapism, with 7 episodes in the 5 days prior to surgery, inserted inflatable prosthesis [251]. Injection therapy, a Winter shunt and an El-Ghorab shunt were tried by Sundaram *et al*, before they inserted prostheses in a patients after 5 days for recurrent priapism [253]. As a first step, despite the apparent lack of success, I would favour the more conservative approach of the Ebbehøj glans cavernosal shunt or the Grayhack caverno-saphenous shunt for resistant cases, and would not resort to this more aggressive approach initially. It should be remembered that if a shunt is created closure may be necessary, as demonstrated in a study of 26 patients who had either a Grayhack, Quackles, Al-Ghorab or Ebbehøj shunt; a permanent open shunt was found in 6 of 26 patients treated for priapism. Potency was restored in five of the six following closure of the shunt [256].

Recent work has demonstrated that both high and low flow states can produce prolonged erection, and each produces a different pattern of blood gases within the penis [239,243,286]. Given that hypoxia of the corpora eventually occurs in all cases, it is important to regard all cases of priapism as a surgical emergency [287-289]. Lue *et al. (1986)* also agree, but maintain that the degree of urgency is greater if the priapism is due to a low-flow rather than a high-flow state [241].

The anorectoplasty had placed the anus in the midline. A further short cleft located to the left of the midline was associated with an anal dimple and subcutaneous muscle. A ridge of fatty tissue was present between the orthotopic and ectopic cleft.

Radiological Findings

Plain radiographs showed the sacrum appeared to be bifid, and cystography performed through the posterior, left side penis showed the catheter to enter the left side of the bladder, with vesicoureteric reflux into the left lower ureter on filling of the bladder (Fig. 55). The position of the orthotopic urethra was suggested by an indentation in the lower central portion of the bladder (Fig.56). A voiding study was inadvertently not obtained. An intravenous pyelogram (IVP) showed bilateral renal tissue with malrotation of the left kidney.

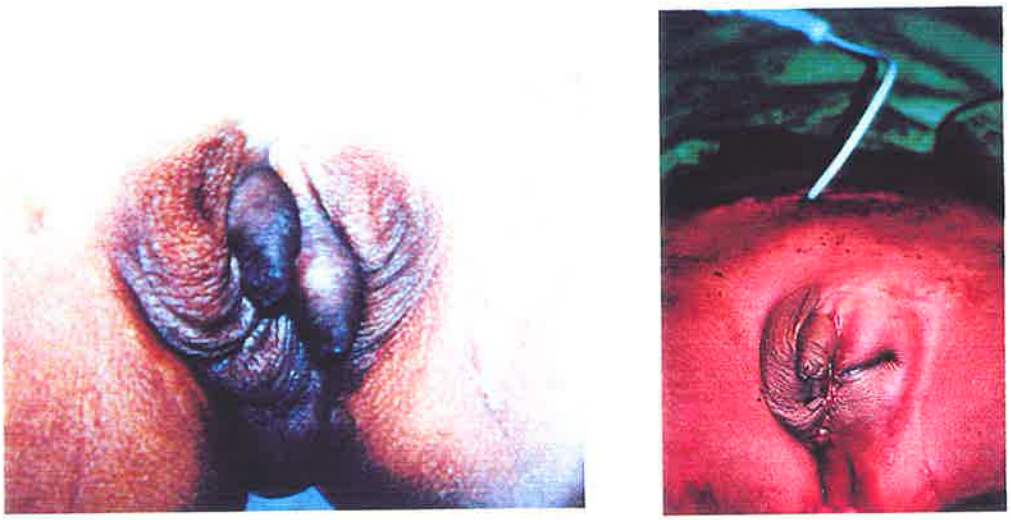


Figure 55: The two penises of this boy with diphallus are virtually equivalent pre-operatively. The post-operative appearance shows a single midline phallus.

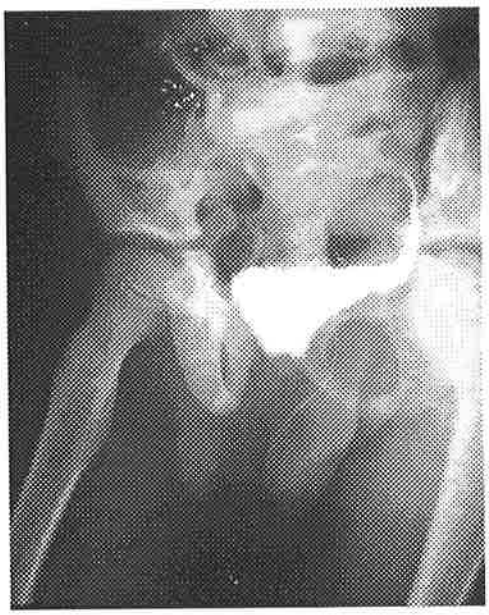


Figure 56: A cystogram shows reflux into the left ureter and the catheter can be seen entering the bladder via the orthotopic urethra.

Operative Findings

Cystsocopy could not be performed, due to the lack of facilities for a procedure being conducted in Papua New Guinea; therefore all findings were at the time of the open procedure. On palpation, each phallus had a normal pair of corpora cavernosa and a normal spongiosum. Both urethras opened into the bladder; the anterior urethra in the normal position at the apex of the trigone, the posterior penile urethra opened to the left of the midline. The left urethral orifice had a 'golf-hole' configuration. Only two ureteric orifices were noted in the bladder. A catheter passed into the bladder through either penis, confirming the connection of each urethra to the bladder.

Discussion

This case of diphallus illustrates the complex cluster of associated anomalies that occur, to a greater or lesser degree, with almost all of the few reported cases. It also highlights the importance of thorough investigation of the genitourinary system to delineate the full extent of duplication and other anomalies. Many similarities exist between the above case and that described by Kaufman *et al.* [293]; which, in addition, had a third continent perineal urethra, prostatic agenesis, vertebral anomalies and widely separated pubic bones. The kidneys and ureters were normal in their case, without VUR. The repair included total left phallectomy with plastic reconstruction of the external genitalia. Azmy *et al.* [294] report a male infant of unspecified karyotype with a complex group of anomalies including diphallus. The phalli each contained complete corpora cavernosa; both passed urine through glanular urethral orifices. Duplication of the bladder with VUR into the left renal moiety of a horseshoe kidney was noted. Multiple additional anomalies included: complex duplication of the lower gastro-intestinal tract with two patent anal dimples, duplication of ileum and colon, Meckel's diverticulum and omphalocele; skeletal anomalies of wide separation of pubis, levoconvex scoliosis, malsegmentation of right ribs, short sacrum and multiple vertebral body anomalies; diastasis of the rectus abdomini; in addition an intra-abdominal left testis was identified and the absence of the right testis was noted. The child was surgically managed by joining the two bladders side-to-side, performing a right orchidopexy, and removing the left phallus. A case of complete diphallus with many of the cluster of anomalies seen in the other cases was reported by Huang *et al.* [296]. A 28-year-old Chinese 46XY male with divarication of the recti, duplication of the symphysis pubis, pelvic asymmetry and a history of imperforate anus with a double gluteal cleft. He had two small penises with normal erectile function, the right having a normal glanular urethra with poor urine flow and the left a perineal hypospadiac urethra, which was the dominant voiding route. The scrotum was bifid and the left compartment contained the small solitary testis. The urinary tract was otherwise normal, aside from a papilloma in the

age and involved excision of the ectopic right penis including a complete urethral excision, via a combined intra and extra vesical approach. A right orchidopexy was also performed and the VUR was managed conservatively. Sharma *et al.* [299] report a case of diphallus in an infant of unspecified karyotype with a number of the common associated defects, but with the first reported association of ileal atresia and tubular duplication of the colon; the infant died of respiratory complications prior to definitive surgery.

These cases, and the Papua New Guinea boy, demonstrate the wide variety of anomalies associated with diphallus. Imperforate anus and other hindgut anomalies including duplication, vertebral anomalies including spina bifida, renal and urinary tract anomalies as well as the obvious external genital anomalies are all common in this rare condition, unified by the recognition that they represent duplication of the caudal body elements of mesodermal origin [292]. Between three to five weeks gestation mesoderm migrates caudally separating the urogenital sinus from the rectum. The genital tubercle, ultimately the phallus, results from merging of the paired columns of mesoderm around the lateral margins of the cloacal plate [292]. Hollowell *et al.* [292] suggested that diphallia may be the result of a defect in migration of mesoderm through the cloacal membrane in early embryogenesis. This hypothesis is supported by the associated defects arising predominantly in tissue of mesodermal origin. Thus, diphallia may be considered part of the more encompassing caudal duplication syndrome. With the exception of the case described by Karna and Kapur [297] (balanced translocation) all other reported karyotypes are normal. It has been suggested that defects in homeobox genes, which are thought to be master controller genes of differentiation, may be involved in caudal duplication, including diphallus [297]. The evidence for this is speculative and involves extrapolation from work on Drosophila species [300]. It appears from testicular biopsy [293] and seminal analysis [296] that fertility is compromised in some cases of complete diphallia. However, long-term follow-up suggests physical growth and development are normal [293,296]. Diphallia, although an exceedingly rare abnormality, demonstrates the

Pelviureteric Junction Obstruction: Age Presentation versus Pathology

Introduction

Pelviureteric junction obstruction is the most common cause of hydronephrosis in the neonatal kidney. In 1969, Johnston classified the pathogenesis of PUJ obstruction into two functional groups: 1) intrinsic disturbances, such as a narrowing or dysfunctional adynamic segment of the ureter and 2) extrinsic mechanical factors, such as an aberrant vessel to the lower-pole, which compresses the pelviureteric junction or upper ureter [301]. Although much has since been written about PUJ obstruction, the clinical significance of these anatomical variants needs to be further pursued.

In an attempt to identify patients that would benefit from the use of doppler US, a study was performed to define the population with a lower pole vessel as the cause of their PUJ obstruction.

Materials and Methods

This study is a retrospective review of 165 kidneys in 158 children between the ages of two days and 15 years who underwent pyeloplasty at the Department of Surgery of the Women's and Children's Hospital, Adelaide, between January 1990 and March 1996. All patients underwent an Anderson-Hynes pyeloplasty.

The operative records were reviewed with particular focus on the underlying pathology and the age at which the patients underwent surgery. Thirty-one patients (33 kidneys) were excluded from the study; fifteen patients had other abnormalities of the urinary tract that complicated the aetiology of the obstruction. These included seven duplications, five horseshoe kidneys, two

pelvic kidneys and one with crossed fused renal ectopia. Six operations were cases of "redo" pyeloplasty and in 10 cases, accurate records were not available.

The remaining 127 patients (132 renal units) were separated into two groups according to the aetiology of the obstruction and the Mann-Whitney rank sum test was used to compare the median age at operation for both groups. P-values of <0.05 were regarded as statistically significant.

Results

In 127 patients in this study, PUJ obstruction was more common in males (94) than in females (33) and occurred more often on the left (76) side than the right (46). There were five patients with bilateral obstruction. The hydronephrosis, leading to the diagnosis of obstruction was detected antenatally in 57% of patients. The remaining cases presented mainly with urinary tract infections (18%), pain (16%) or haematuria (2%); 7% had a variety of other presentations.

The most common finding at operation was a ureteral narrowing which occurred in 51 per cent of the renal units (65 of 132 kidneys). A kink at the PUJ was found in 23 units and a further 18 had an obstruction caused by a combination of narrowing and kinking. An obstructing, aberrant lower polar vessel was identified as the cause of the PUJ obstruction for 24 kidneys; in all cases the vessels crossed the ureter anteriorly; in three of these cases a concurrent intrinsic narrowing was also found.

The age at operation ranged from two days to 12 years for the intrinsic group, while the patients with lower-pole vessels were aged five days to 15 years. Only five of 24 patients (21%) with lower pole vessels underwent operation in the first year of life compared to 70 of 103 (68%) for the non-vessel group. The median and interquartile ranges for both groups are

extrinsic compression by an aberrant lower-pole vessel tends to present at a later age compared to intrinsic pathologies. Others have alluded to this difference [317,321,322] although it has never been confirmed. In a study reviewing 31 cases of PUJ obstruction in adults, Lowe and Marshall found associated lower-pole vessels in 52%, twice the rate reported in children [323]. Harman *et al.* described a bimodal presentation of the disease, the first peak being from two weeks to six months and the second from seven to eight years of age. Freedman *et al.* proposed that the underlying pathology in the neonate might differ inherently from that in older children who present clinically [324]. This is supported by the failure of late ultrasonography to detect significant hydronephrosis in children who subsequently presented with symptoms of obstruction [325] and Stephens proposed a gradual evolution of the distension ultimately leading to obstruction and pain [322]. His anatomical description was shown experimentally in canines by Koff, whose findings indicate that the precise anatomy of the pelviureteric junction determines the pattern of flow across the obstruction [326].

The findings for the study group reported here confirm the late presentation of vascular PUJ obstruction. With improvement in doppler US, this additional test may be useful in selecting out those cases where there is some inconclusive evidence of obstruction, either for closer follow-up or earlier surgery. The interpretation of the doppler findings should be modified by the clinical picture, US appearance and results of diuretic renography, remembering that the underlying pathology and vessel prevalence in equivocal cases that resolved has not been determined in the above study. A prospective study, involving follow-up of these ambiguous cases, after determination of the presence of lower-pole vessels through doppler US, is warranted.

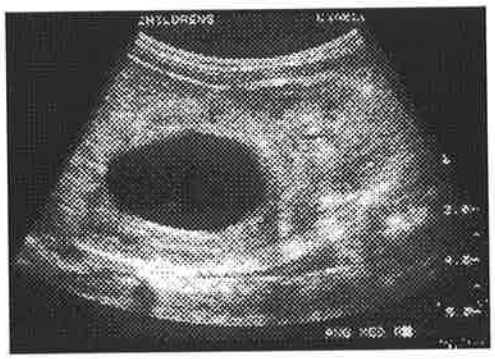
Urine in the obstructed pelvis was noted to be cloudy and microscopy and culture revealed *S. Aureus* infection. The patient was commenced on intravenous Flucloxacillin and a nephrostomy tube was left *in situ* to temporarily by-pass the constructed pyelo-pyelostomy.

Post-operative recovery was uncomplicated and the nephrostomy was removed at six days, after a nephrostogram revealed good drainage to the bladder at low pressure (Fig. 60). The patient was discharged on the eighth post-operative day and, when seen four months later, had good parenchymal thickness on the upper pole, with minimal hydronephrosis.

Discussion

This case represents an unusual complication of a duplex collecting system and challenges notions of conservatism with regard to timing of surgery in antenatally diagnosed hydronephrosis.

Duplex collecting systems arise embryonically from two buds from the same mesonephric duct. One or both buds may be displaced such that an ectopic ureteric orifice results. The more cranial of the buds drains the upper pole of the kidney, but becomes more caudal and ectopic to the lower pole ureteric orifice, as the distal mesonephric duct becomes absorbed into the urogenital sinus (Weigert-Meyer rule) [327].



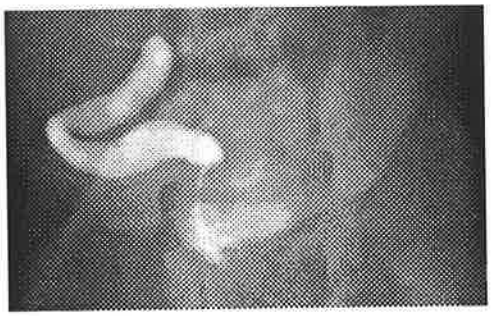


Figure 58 (left): An ultrasound showing dilatation of the upper pole collecting system, out of proportion to the degree of ureteric distension **Figure 59 (right):** The pre-operative cystogram with reflux into the dilated upper pole ureter which inserts ectopically into the urethra.

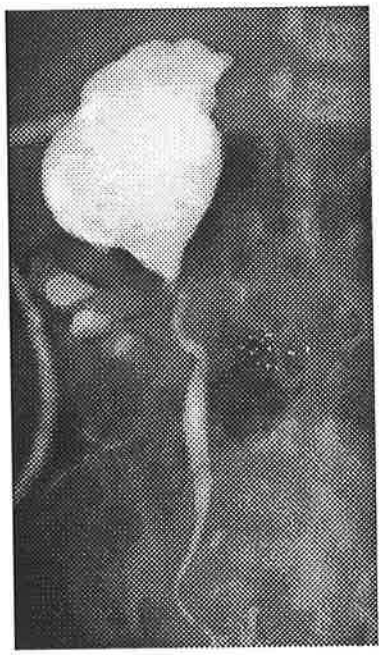


Figure 60: Contrast has been injected via the upper pole collecting system nephrostomy tube and drains through the pyelo-pyelostomy, into the orthotopic ureter, then into the bladder. The arrow indicates the junction of the upper pole collecting system with the lower pole pelvis.

A study of the impact of prenatal US on the morbidity and outcome of patients with duplex systems found that early recognition and treatment of duplication anomalies associated with hydronephrosis prevented considerable postnatal morbidity, with eleven times more patients with postnatally diagnosed duplication anomalies presenting with significant symptoms [328]. Furthermore, it was found that three times more upper pole moieties were functional and salvaged with minimal morbidity in those with antenatally diagnosed duplex anomalies compared with those that presented postnatally.

Renal segments removed from patients with duplex anomalies have been found to show changes related to obstruction and infection, rather than dysplasia [330]. It is possible, therefore, that functional upper pole moieties can be saved, using kidney-sparing procedures (uretero-ureterostomy/uretero-pyelostomy/pyelo-pyelostomy), where surgery is performed prior to the development of further infective/obstructive complications. In this case, infection was evident at five days of age, despite prophylaxis. Pyelo-pyelostomy was chosen for this child because the function study and the macroscopic appearance of the renal parenchyma supported preservation of the upper half of the kidney and the alternative approach would have required both a pyeloplasty and a megaureter reimplant in a newborn. Those with a poorly or non-functioning upper pole, more likely to be seen in postnatally diagnosed cases, usually require heminephrectomies or partial ureterectomies.

Had this patient not undergone surgery early in the neonatal period, it is likely that the combination of infection and obstruction in the upper pole moiety would have caused significant loss of renal function, if not major morbidity from pyelonephritis. Through early intervention, she was able to have a kidney-sparing procedure with minimal morbidity. The finding of pelvi-ureteric junction obstruction of the upper pole pelvis is rare and occurred in this case due to dilatation of the upper pole, with subsequent twisting and kinking of its ureter on the lower pole ureter. Prior to the report of this case it would appear that the entity described had not previously been reported.

Ureterocalycostomy for PUJ obstruction in the Horseshoe Kidney

Introduction

A horseshoe kidney is a common congenital renal abnormality resulting from fusion of the lower poles of each metanephros. The anomaly was first documented in early autopsy studies by DeCapri (1522) (quoted by reference [331]). The presence of a horseshoe kidney predisposes to a number of clinical problems, in particular, poor drainage of the renal pelvis, producing hydronephrosis that may be complicated by pain, infection and stone formation [332]. Factors predisposing to obstruction in Horseshoe kidneys include: high takeoff ureter with an acute pelviureteric junction angle; an isthmus over which the ureter must pass; aberrant blood vessels in the region of the PUJ; a relatively intrarenal pelvis with a medially placed inferior pole and congenital PUJ narrowing [333,334]. The condition may also be associated with abnormalities of other organ systems [335]. Historically, various approaches have been employed to manage PUJ obstruction in horseshoe kidney with various aspects of the above pathology in mind. These include division of the isthmus alone and subsequently in conjunction with nephropexy [333]; rotation flap of the pelvis; heminephrectomy [334] and more recently, endopyelo-plasty [65,336-338] with and without the use of laparoscopy [339]. Appropriately, the Anderson-Hynes pyeloplasty is still the more common primary procedure to manage PUJ obstruction in the horseshoe kidney [340].

Ureterocalycostomy has been previously described to relieve PUJ obstruction, particularly for the huge kidney with a dependent lower pole or where the pelvis is intrarenal [341-344]. Applied to the horseshoe kidney, ureterocalycostomy has the theoretical advantage of providing dependant drainage, while avoiding the need for ureteric passage over the isthmus. Here is described a series of four cases of horseshoe kidney with PUJ obstruction, in whom ureterocalycostomy was employed to establish adequate drainage of the renal pelvis. Each of these cases had a failed primary drainage procedure. With the lessons learnt from these cases

it would appear that ureterocalycostomy is a useful procedure in selected cases of PUJ obstruction in a horseshoe kidney.

Case Reports

Case 1: M J initially presented at three years of age with renal pain and a urinary tract infection, associated with poor control of his diabetes mellitus. Investigations confirmed the presence of a PUJ obstruction to the left side of a horseshoe kidney. Exploration of the kidney was undertaken at which the surgeon found the PUJ to be crossed by a lower pole vessel. An Anderson-Hynes dismembered pyeloplasty was commenced but, because of the impression that the anastomosis was under tension when anterior to the lower pole vessels, the PUJ was placed posterior to the offending vasculature. Subsequently the kidney remained dilated and poorly draining; plus the boy had further urinary tract infections and on-going poor diabetic control. An antegrade nephrostomy study, which incorporated balloon dilatation of the PUJ, was performed, without subsequent improvement in drainage. After referral at eight years of age, a diuretic renogram was followed by a second exploration, at which the ureter was again disconnected and lengthened by a rotational flap of the pelvis. The kidney was also rotated to facilitate the caudal aspect of the renal pelvis coming to lie in a dependent position. The patient was symptom free for two years, then developed further episodes of colicky pain and deterioration in the drainage of the kidney. At a third operation, when the boy was 11 years old, the ureter was found to be bound to the renal isthmus by fibrous tissue; the obstruction was resolved by performing a ureterocalycostomy. Following removal of the double J stent there have been no further symptoms, and the US and nuclear medicine studies indicate that the drainage is satisfactory 18 months later.

Case 2: CK presented with right-sided hydronephrosis and horseshoe kidney. Her initial surgical procedure involved resection of a portion of renal pelvis lying behind aberrant renal

hilar blood vessels when she was nine years old. A nuclear medicine scan five and eight years later showed obstruction and she underwent a redo pyeloplasty at aged 16 years, during which a nephrostomy was left *in situ*. Continued drainage occurred from the nephrostomy, with pain when the nephrostomy was clamped. Therefore, the patient was returned to the operating room on the 10th post operative day at which time a ureterocalycostomy was performed and a double J stent inserted. Follow-up studies showed minor persistent hydronephrosis, which had resolved on an US two years after her last operation.

Case 3: JA presented with a urinary tract infection for which he underwent renal exploration at the age of three years, after a nuclear medicine study had identified an obstructed left pelviureteric junction. A renal US showed hydronephrosis, parenchymal thinning and a horseshoe kidney; the lasix half clearance time was greater than 100min on the diuretic nuclear renogram. The operative procedure, as described in the original operation record, was a routine Anderson-Hynes pyeloplasty. The boy continued to have intermittent renal discomfort and poor drainage on a repeat nuclear medicine study, this led to a Whitaker test one year after his pyeloplasty; with an infusion rate of 13 ml/min; no pressure rise was seen in the kidney. The boy was subsequently transferred to my care, after further episodes of renal discomfort. Repeat radiological studies reiterated the marked calyceal and pelvic distension, with poor drainage during the lasix washout phase of the nuclear medicine study, leading to a further exploration of the kidney at six years of age. The ureter was found to exit the pelvis just cranial to a lower pole vessel under which it passed posteriorly, prior to passing anteriorly over the thick isthmus (Fig.61). The ureter was not sufficiently long to anastomose anterior to the lower pole vessel without tension; therefore the ureter was joined to the anterior calyx of the lower pole of the kidney. The postoperative course was complicated by a urinary tract infection, leading to early removal of the double J stent. Subsequently the boy has been free of renal pain and urinary infections; also, an US and nuclear medicine study performed 12 months after the

Subsequently the boy has been free of renal pain and urinary infections; also, an US and nuclear medicine study performed 12 months after the ureterocalycostomy showed significant improvement in the drainage of the renal pelvis and degree of hydronephrosis.

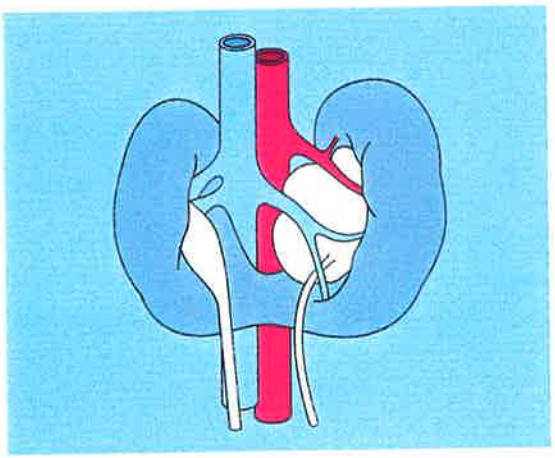


Figure 61: ***Case 3***. The ureter passes under a lower pole vessel, prior to passing over the renal isthmus.

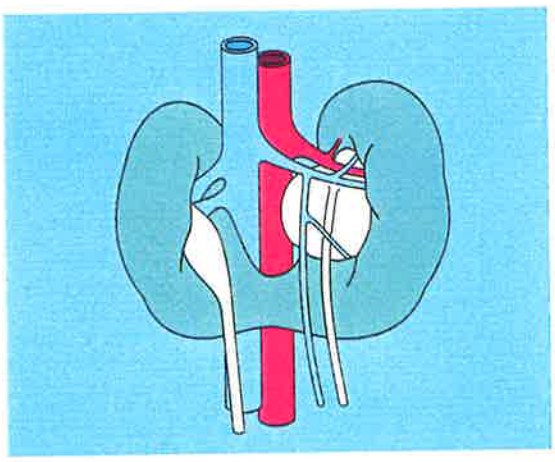


Figure 62: ***Case 4.*** The ureter arose from the cranial aspect of the left renal pelvis, followed the gonadal vein and was crossed anteriorly by a branch from the gonadal vein, just cranial to the renal isthmus.

given in Figure 57. The median age at operation was 3.1 months for the non-vessel group and 67.3 months for the vessel group; the difference between the two groups was statistically significant (Mann-Whitney, P<0.0001).

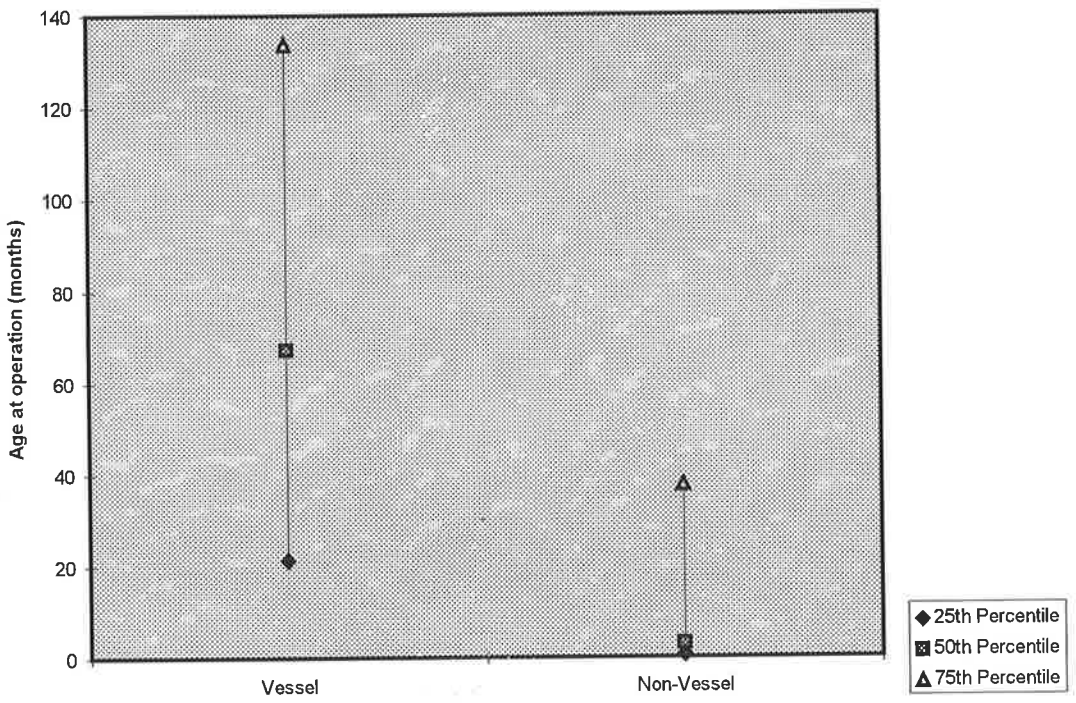


Figure 57: Median and interquartile range (25th, 50th, 75th percentile) for age at operation for vessel and non-vessel groups.

Discussion

Pelviureteric junction obstruction is the most common cause of obstruction of the fetal kidney and is more common in males than females. The exact cause is unknown, but several theories have been proposed based on embryological, anatomical, histological, and functional hypotheses. The lesion is bilateral in 10-40% of cases, usually with a different degree of severity for each side. When unilateral, the obstruction is more commonly left-sided and is often associated with anomalies of the contralateral kidney, including multicystic dysplasia, renal agenesis, duplication, horseshoe deformity, and renal ectopia. These common associations were again demonstrated in the above study. Anomalies of other organ systems are infrequent, and no consistent pattern of association has been found [302].

The widespread use of prenatal ultrasonography has resulted in a marked increase in the early detection of fetal hydronephrosis, the natural history of which is still being defined. The fate of antenatally diagnosed hydronephrosis remains uncertain. Studies have shown that the hydronephrosis in as many as 80% with persistent dilatation after birth either subsides or markedly improves within 12 months [303,304]. A study conducted by Koff and Campbell illustrated that many newborn kidneys with severe hydronephrosis and initial poor function were not obstructed and showed spontaneous improvement [305]. Flake noted that hydronephrosis appeared less severe on postnatal US, attributing the change to secondary effects of maternal hormones on smooth muscle of fetal renal pelvis [306]. In contrast, those with significant obstructive renal tract pathology can have a normal scan before 20 weeks gestation [307,308], an initially normal postnatal US [309] or even a non-obstructive diuretic renogram in the early newborn period [305].

If obstruction is confirmed, it is widely held that early surgery should be undertaken to relieve obstruction, to allow for maximum development of renal function. This is supported by Mayor

et al. who followed children with severe obstruction and found when obstruction was relieved in early infancy, creatinine clearance improved compared to older children who demonstrated deterioration in renal function [310]. King *et al.* reported a tenfold increase in renal function after pyeloplasty in infancy, compared to older patients [311] and Perez *et al.* this difference between the outcome for the different ages is partly due to contralateral hypertrophy, with a decreases in the ability of the affected kidney to recover relative function [312].

Currently, postnatal diagnosis of PUJ obstruction is achieved with a combination of US, nuclear medicine studies, voiding cystourethrography [313] and the Whitaker pressure-perfusion test [314]. Nuclear medicine function and drainage studies have assumed a major role, but the test has many limitations and should be interpreted in the light of the prenatal US history and the post-natal finding, including the clinical examination and history. Homsy *et al.* followed 17 kidneys with a non-obstructed or partially obstructed system, demonstrated on diuretic renogram, over a period of three to six months. Of the 17 renal units, eight (47%) showed improvement of pelvic washout, seven (41%) developed delay in drainage and two (12%) remained stable [304]. Dejter *et al.* observed five renal units with equivocal drainage half-time (10 to 20 minutes) for an average period of seven months, noting that two units exhibited significant improvement in drainage half-times to a non-obstructed pattern (<10 minutes), one deteriorated into obstruction (>20 minutes) while two units remained equivocal [315]. A more refined approach to the interpretation of the study may improve the predictability of the investigation [316].

A lower pole vessel as the cause of the pelviureteric obstruction, was found in 18% of renal units (24 of 132) undergoing operation in the above study. The prevalence of lower-pole vessels associated with PUJ obstruction in children exhibit a wide variance, with an incidence of 11%, 27% and 39% quoted [317-320]. Highlighting this anatomical variation may improve precision of follow-up. The results of the above study indicate that PUJ obstruction due to the

extrinsic compression by an aberrant lower-pole vessel tends to present at a later age compared to intrinsic pathologies. Others have alluded to this difference [317,321,322] although it has never been confirmed. In a study reviewing 31 cases of PUJ obstruction in adults, Lowe and Marshall found associated lower-pole vessels in 52%, twice the rate reported in children [323]. Harman *et al.* described a bimodal presentation of the disease, the first peak being from two weeks to six months and the second from seven to eight years of age. Freedman *et al.* proposed that the underlying pathology in the neonate might differ inherently from that in older children who present clinically [324]. This is supported by the failure of late ultrasonography to detect significant hydronephrosis in children who subsequently presented with symptoms of obstruction [325] and Stephens proposed a gradual evolution of the distension ultimately leading to obstruction and pain [322]. His anatomical description was shown experimentally in canines by Koff, whose findings indicate that the precise anatomy of the pelviureteric junction determines the pattern of flow across the obstruction [326].

The findings for the study group reported here confirm the late presentation of vascular PUJ obstruction. With improvement in doppler US, this additional test may be useful in selecting out those cases where there is some inconclusive evidence of obstruction, either for closer follow-up or earlier surgery. The interpretation of the doppler findings should be modified by the clinical picture, US appearance and results of diuretic renography, remembering that the underlying pathology and vessel prevalence in equivocal cases that resolved has not been determined in the above study. A prospective study, involving follow-up of these ambiguous cases, after determination of the presence of lower-pole vessels through doppler US, is warranted.

Upper Pole Pelviureteric Junction Obstruction

Introduction

Upper pole PUJ obstruction has never been described in isolation. This case presents the features of a girl with obstruction to the outflow of urine from the upper pole collecting system, due to a secondary rotation of the kidney. The additional significant finding of infection in the obstructed collecting system, despite perinatal antibiotics is also discussed.

Case Study:

RP was referred at three days of age, after an antenatal diagnosis of severe right-sided hydronephrosis. She was commenced on prophylactic antibiotics shortly after birth.

Renal US at three days of age revealed a right duplex kidney with dilatation of upper pole collecting system, out of proportion with the degree of ureteric distension, suggesting an element of pelvi-ureteric junction obstruction (Fig.58). The upper pole ureter passed beyond the bladder neck and appeared to insert ectopically into the urethra. A micturation cystourethrogram showed reflux into a markedly dilated right ureter, which inserted in the region of the external urethral sphincter (Fig.59). A MAG3 radionuclide renal scan revealed a right duplex kidney. The kidney contributed 58% of overall renal function, with an obstructed upper pole that functioned well.

When five days old the patient underwent cystoscopy, during which two ureteric orifices were identified, one on either side of the trigone. No definite ectopic ureteral opening could be identified in the urethra. A right pyelo-pyelostomy was performed, revealing the right kidney to be oedematous, inflamed and rotated postero-laterally by the massively dilated upper renal pelvis; consequently the upper ureter was kinked as it was crossed by the lower pole ureter, creating a secondary upper pole moiety pelvi-ureteric junction obstruction.

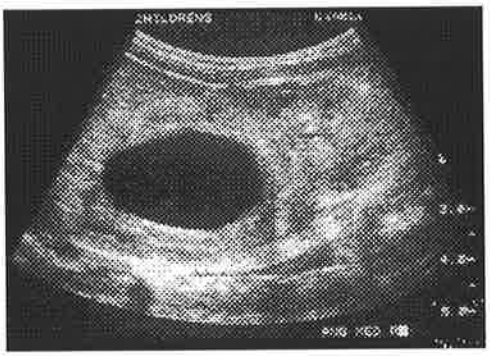
Urine in the obstructed pelvis was noted to be cloudy and microscopy and culture revealed *S. Aureus* infection. The patient was commenced on intravenous Flucloxacillin and a nephrostomy tube was left *in situ* to temporarily by-pass the constructed pyelo-pyelostomy.

Post-operative recovery was uncomplicated and the nephrostomy was removed at six days, after a nephrostogram revealed good drainage to the bladder at low pressure (Fig. 60). The patient was discharged on the eighth post-operative day and, when seen four months later, had good parenchymal thickness on the upper pole, with minimal hydronephrosis.

Discussion

This case represents an unusual complication of a duplex collecting system and challenges notions of conservatism with regard to timing of surgery in antenatally diagnosed hydronephrosis.

Duplex collecting systems arise embryonically from two buds from the same mesonephric duct. One or both buds may be displaced such that an ectopic ureteric orifice results. The more cranial of the buds drains the upper pole of the kidney, but becomes more caudal and ectopic to the lower pole ureteric orifice, as the distal mesonephric duct becomes absorbed into the urogenital sinus (Weigert-Meyer rule) [327].



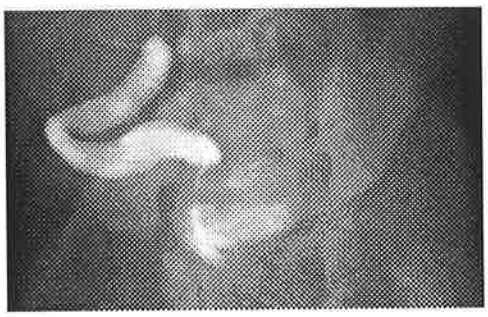


Figure 58 (left): An ultrasound showing dilatation of the upper pole collecting system, out of proportion to the degree of ureteric distension **Figure 59 (right):** The pre-operative cystogram with reflux into the dilated upper pole ureter which inserts ectopically into the urethra.

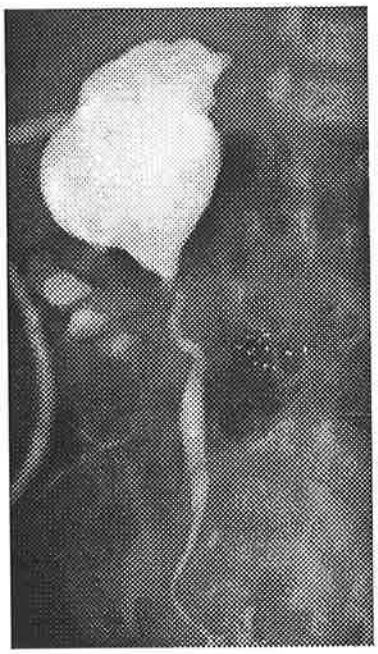


Figure 60: Contrast has been injected via the upper pole collecting system nephrostomy tube and drains through the pyelo-pyelostomy, into the orthotopic ureter, then into the bladder. The arrow indicates the junction of the upper pole collecting system with the lower pole pelvis.

In general, ectopic ureters are associated with dysplastic non-functioning renal parenchyma and are therefore more likely to require heminephroureterectomy. In this case, the upper pole function was relatively normal despite the presence of an ectopic upper pole ureter inserting below the bladder neck [328].

Pelvi-ureteric junction obstruction can arise from a variety of anatomical abnormalities, both intrinsic and extrinsic to the junction. The most common cause of extrinsic pelvi-ureteric junction obstruction is that caused by an aberrant or accessory renal artery or an early branching vessel to the lower pole of the kidney, seen in up to 40% of patients with pelvi-ureteric junction obstruction [327,329]. This case presents an unusual variation, where the obstruction does not involve the lower pole moiety, but rather the upper pole moiety as a result of the longitudinal rotation of the kidney and subsequent crossing of the upper pole pelvi-ureteric junction by the lower pole ureter. Primary upper moiety pelvi-ureteric junction obstruction has not, to my knowledge, been previously reported.

Antenatal US has allowed detection of abnormalities before renal function becomes impaired and thus non-operative and more conservative (kidney-sparing) surgical approaches to pelvic-ureteric junction obstruction/duplex systems have been suggested. The timing and indications for surgical intervention in infants with hydronephrosis detected on antenatal US is still controversial.

A number of studies have looked at the need for early postnatal intervention in cases of antenatally diagnosed pelvic-ureteric junction obstruction. In general, it has been shown that renal function in these neonates is often well preserved and rarely deteriorates in the short term. It has therefore been postulated that non-operative management, rather than surgical, can be utilized in those with preserved renal function (>40%), once a micturating cysto-urethrogram has excluded bladder outlet obstruction in boys and severe VUR. The value of antibiotic prophylaxis has also been questioned in these patients [324].

A study of the impact of prenatal US on the morbidity and outcome of patients with duplex systems found that early recognition and treatment of duplication anomalies associated with hydronephrosis prevented considerable postnatal morbidity, with eleven times more patients with postnatally diagnosed duplication anomalies presenting with significant symptoms [328]. Furthermore, it was found that three times more upper pole moieties were functional and salvaged with minimal morbidity in those with antenatally diagnosed duplex anomalies compared with those that presented postnatally.

Renal segments removed from patients with duplex anomalies have been found to show changes related to obstruction and infection, rather than dysplasia [330]. It is possible, therefore, that functional upper pole moieties can be saved, using kidney-sparing procedures (uretero-ureterostomy/uretero-pyelostomy/pyelo-pyelostomy), where surgery is performed prior to the development of further infective/obstructive complications. In this case, infection was evident at five days of age, despite prophylaxis. Pyelo-pyelostomy was chosen for this child because the function study and the macroscopic appearance of the renal parenchyma supported preservation of the upper half of the kidney and the alternative approach would have required both a pyeloplasty and a megaureter reimplant in a newborn. Those with a poorly or non-functioning upper pole, more likely to be seen in postnatally diagnosed cases, usually require heminephrectomies or partial ureterectomies.

Had this patient not undergone surgery early in the neonatal period, it is likely that the combination of infection and obstruction in the upper pole moiety would have caused significant loss of renal function, if not major morbidity from pyelonephritis. Through early intervention, she was able to have a kidney-sparing procedure with minimal morbidity. The finding of pelvi-ureteric junction obstruction of the upper pole pelvis is rare and occurred in this case due to dilatation of the upper pole, with subsequent twisting and kinking of its ureter on the lower pole ureter. Prior to the report of this case it would appear that the entity described had not previously been reported.

Ureterocalycostomy for PUJ obstruction in the Horseshoe Kidney

Introduction

A horseshoe kidney is a common congenital renal abnormality resulting from fusion of the lower poles of each metanephros. The anomaly was first documented in early autopsy studies by DeCapri (1522) (quoted by reference [331]). The presence of a horseshoe kidney predisposes to a number of clinical problems, in particular, poor drainage of the renal pelvis, producing hydronephrosis that may be complicated by pain, infection and stone formation [332]. Factors predisposing to obstruction in Horseshoe kidneys include: high takeoff ureter with an acute pelviureteric junction angle; an isthmus over which the ureter must pass; aberrant blood vessels in the region of the PUJ; a relatively intrarenal pelvis with a medially placed inferior pole and congenital PUJ narrowing [333,334]. The condition may also be associated with abnormalities of other organ systems [335]. Historically, various approaches have been employed to manage PUJ obstruction in horseshoe kidney with various aspects of the above pathology in mind. These include division of the isthmus alone and subsequently in conjunction with nephropexy [333]; rotation flap of the pelvis; heminephrectomy [334] and more recently, endopyelo-plasty [65,336-338] with and without the use of laparoscopy [339]. Appropriately, the Anderson-Hynes pyeloplasty is still the more common primary procedure to manage PUJ obstruction in the horseshoe kidney [340].

Ureterocalycostomy has been previously described to relieve PUJ obstruction, particularly for the huge kidney with a dependent lower pole or where the pelvis is intrarenal [341-344]. Applied to the horseshoe kidney, ureterocalycostomy has the theoretical advantage of providing dependant drainage, while avoiding the need for ureteric passage over the isthmus. Here is described a series of four cases of horseshoe kidney with PUJ obstruction, in whom ureterocalycostomy was employed to establish adequate drainage of the renal pelvis. Each of these cases had a failed primary drainage procedure. With the lessons learnt from these cases

it would appear that ureterocalycostomy is a useful procedure in selected cases of PUJ obstruction in a horseshoe kidney.

Case Reports

Case 1: M J initially presented at three years of age with renal pain and a urinary tract infection, associated with poor control of his diabetes mellitus. Investigations confirmed the presence of a PUJ obstruction to the left side of a horseshoe kidney. Exploration of the kidney was undertaken at which the surgeon found the PUJ to be crossed by a lower pole vessel. An Anderson-Hynes dismembered pyeloplasty was commenced but, because of the impression that the anastomosis was under tension when anterior to the lower pole vessels, the PUJ was placed posterior to the offending vasculature. Subsequently the kidney remained dilated and poorly draining; plus the boy had further urinary tract infections and on-going poor diabetic control. An antegrade nephrostomy study, which incorporated balloon dilatation of the PUJ, was performed, without subsequent improvement in drainage. After referral at eight years of age, a diuretic renogram was followed by a second exploration, at which the ureter was again disconnected and lengthened by a rotational flap of the pelvis. The kidney was also rotated to facilitate the caudal aspect of the renal pelvis coming to lie in a dependent position. The patient was symptom free for two years, then developed further episodes of colicky pain and deterioration in the drainage of the kidney. At a third operation, when the boy was 11 years old, the ureter was found to be bound to the renal isthmus by fibrous tissue; the obstruction was resolved by performing a ureterocalycostomy. Following removal of the double J stent there have been no further symptoms, and the US and nuclear medicine studies indicate that the drainage is satisfactory 18 months later.

Case 2: CK presented with right-sided hydronephrosis and horseshoe kidney. Her initial surgical procedure involved resection of a portion of renal pelvis lying behind aberrant renal

hilar blood vessels when she was nine years old. A nuclear medicine scan five and eight years later showed obstruction and she underwent a redo pyeloplasty at aged 16 years, during which a nephrostomy was left *in situ*. Continued drainage occurred from the nephrostomy, with pain when the nephrostomy was clamped. Therefore, the patient was returned to the operating room on the 10th post operative day at which time a ureterocalycostomy was performed and a double J stent inserted. Follow-up studies showed minor persistent hydronephrosis, which had resolved on an US two years after her last operation.

Case 3: JA presented with a urinary tract infection for which he underwent renal exploration at the age of three years, after a nuclear medicine study had identified an obstructed left pelviureteric junction. A renal US showed hydronephrosis, parenchymal thinning and a horseshoe kidney; the lasix half clearance time was greater than 100min on the diuretic nuclear renogram. The operative procedure, as described in the original operation record, was a routine Anderson-Hynes pyeloplasty. The boy continued to have intermittent renal discomfort and poor drainage on a repeat nuclear medicine study, this led to a Whitaker test one year after his pyeloplasty; with an infusion rate of 13 ml/min; no pressure rise was seen in the kidney. The boy was subsequently transferred to my care, after further episodes of renal discomfort. Repeat radiological studies reiterated the marked calyceal and pelvic distension, with poor drainage during the lasix washout phase of the nuclear medicine study, leading to a further exploration of the kidney at six years of age. The ureter was found to exit the pelvis just cranial to a lower pole vessel under which it passed posteriorly, prior to passing anteriorly over the thick isthmus (Fig.61). The ureter was not sufficiently long to anastomose anterior to the lower pole vessel without tension; therefore the ureter was joined to the anterior calyx of the lower pole of the kidney. The postoperative course was complicated by a urinary tract infection, leading to early removal of the double J stent. Subsequently the boy has been free of renal pain and urinary infections; also, an US and nuclear medicine study performed 12 months after the

Subsequently the boy has been free of renal pain and urinary infections; also, an US and nuclear medicine study performed 12 months after the ureterocalycostomy showed significant improvement in the drainage of the renal pelvis and degree of hydronephrosis.

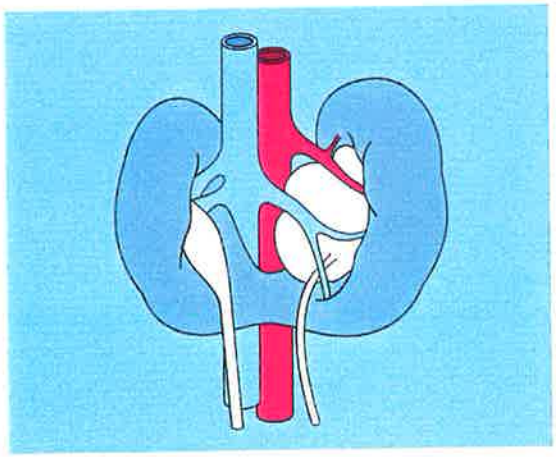


Figure 61: ***Case 3***. The ureter passes under a lower pole vessel, prior to passing over the renal isthmus.

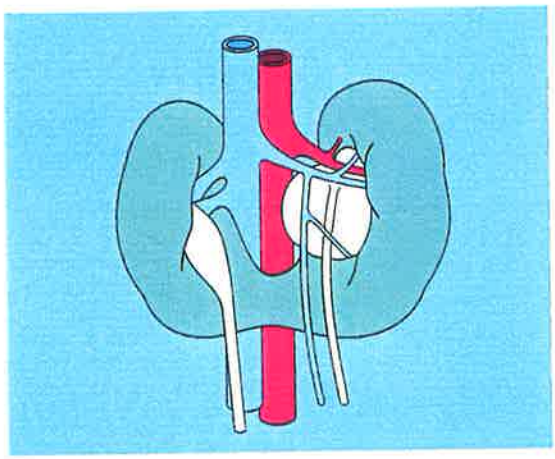


Figure 62: ***Case 4.*** The ureter arose from the cranial aspect of the left renal pelvis, followed the gonadal vein and was crossed anteriorly by a branch from the gonadal vein, just cranial to the renal isthmus.

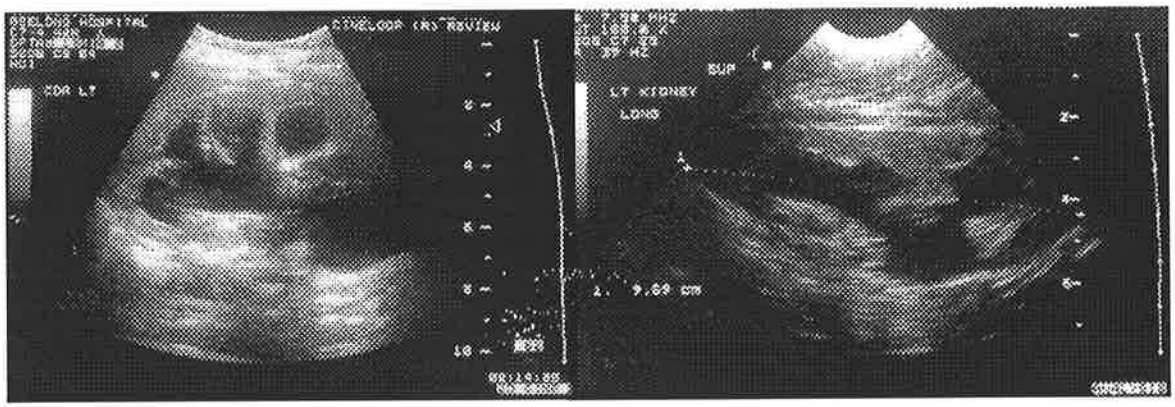


Figure 63: Ultrasound before and after final operation for ***Case 4***.

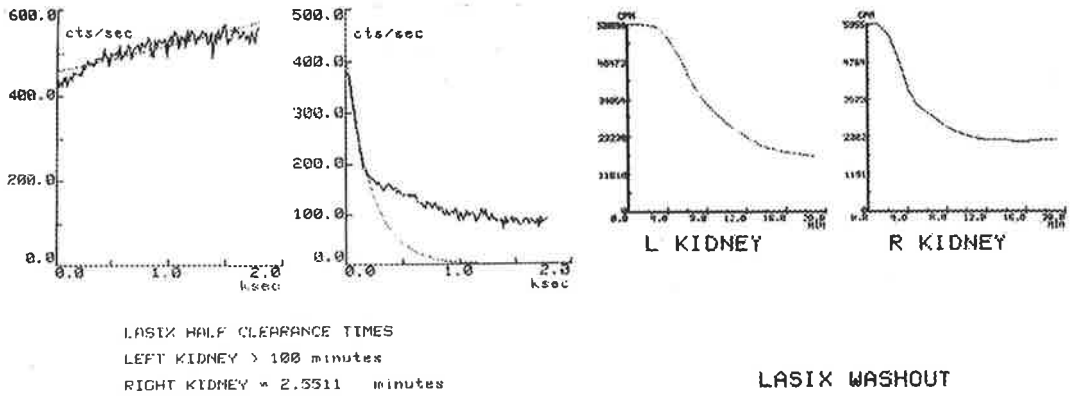


Figure 64: Nuclear medicine study before and after the final operation on ***Case 4***.

Case 4: (Fig.62-64) LW developed a urinary tract infection and haematuria which was found to be due to a pelviureteric junction obstruction on the left side of a Horseshoe kidney. The obstruction, which was suspected on an US, was confirmed on an IVP and subsequent retrograde study when five years old. At exploration, a high take-off of the ureter was noted and adhesions were divided: no pyeloplasty was attempted as the surgeon felt the division of the adhesions had resolved the obstruction. The girl presented again two years later, with intermittent episodes of abdominal pain which were initially thought to be due to constipation. Investigation with an US and a MAG3, lasix renogram showed marked hydronephrosis, delayed drainage and reduced function. A further operation was undertaken at which the ureter was seen to attach to the cranial extent of a long, left-sided pelvis. The ureter followed the left gonadal vein on the anterior surface of the renal pelvis, toward the isthmus of the horseshoe kidney. The ureter was crossed, anteriorly by a vein that was a branch of the gonadal vein. The ureter was mobilised, divided and anastomosed to the caudal extent of the pelvis and the anterior aspect of the lower pole calyx, giving the pelviureteric junction a dependent position without tension on the ureter. A diuretic renogram and an US three months after the procedure showed marked improvement, with further improvement 12 months after the surgery.

Discussion

Rovsing, in 1911, first described the division of the isthmus of horseshoe kidney in a patient presenting with long-standing nausea, vomiting and abdominal pain worsened by hyperextension of the trunk (Rovsing's sign). The horseshoe kidney was diagnosed by palpation under general anaesthetic, alone: division of the isthmus was performed under the assumption that abnormal position and passage of the ureters over the isthmus was the cause of the symptoms of obstruction. Symptomatic improvement was reported following the operation [345].

Donahue (1932) described the use of nephropexy, in addition to isthmic division, to successfully relieve hydronephrosis on the right, then subqsequently on the left in a case of horseshoe kidney. The 32 year old woman presented with a long history of right upper quadrant pain; bilateral hydronephrosis being demonstrated preoperatively by pyelography. At operation he describes an anterior dilated pelvis with an acute angle high take off ureter. The ureter passed forward, over the isthmus, and aberrant vessels were seen compressing the region of the left pelviureteric junction. The patient was free of hydronephrosis and symptom free at three months postoperatively. Donahue emphasized the importance of dependant drainage of the renal pelvis being established at the time of operation [333].

In 1937 Foley described a series of 20 patients treated by Y-V pyeloplasty to relieve PUJ obstruction, principally in non-fused renal units; advocating the principle of a dependant, funneled PUJ with minimal redundant pelvis [346]. This principles was reiterated by Lowsley in 1952 [347]. Successful use of reconstructive pyeloplasty in managing PUJ obstruction in horseshoe kidneys was supported by results of a series published by Culp and Winterringer in 1955 [348].

Pitts and Muecke (1975) [331] retrospectively reviewed a series of 170 patients with a Horseshoe kidney seen at the New York Hospital over a 40 year period. The most common surgical problem was hydronephrosis, secondary to either PUJ obstruction or calculi. Twenty-five patients underwent pyeloplasty for obstruction, 80% of which were cured by a single procedure. Six underwent unsuccessful division of the isthmus alone, all of which failed to ameliorate the blockage. Pitts and Muecke suggested that previously recorded successful treatment with symphysectomy was probably due to dissection of the PUJ rather then division of the isthmus. Given that the procedure may be complicated by haemorrhage or fistula formation with no prospective data suggesting proven benefit, the indications for the procedure are unclear, although it has been recently advocated in conjunction with pyeloplasty by a large Russian retrospective review [334] and described alone in a Chinese review [349].

More recently, endopyelotomy has been described as a technique for management of PUJ obstruction in horseshoe kidneys [336-338]. Although only employed in a small group of patients, persisting radiographic improvement of obstruction is documented at up to 52 months follow-up [336]. Computerised Tomography with angioplasty [65] and laparoscopic division of crossing vessels at the PUJ [339] are two techniques that have been recently described as treatment adjuncts. However, a vascular solution to a urological problem seems inappropriate, because of the potential loss of renal parenchyma. Currently, Anderson-Hynes pyeloplasty is the most common initial procedure in correcting PUJ obstruction in horseshoe kidney [340]. A success rate of greater than 90% in relieving obstruction with a single operation can be expected with pyeloplasty in normally rotated kidneys [340,350]. However, very little has been published on the outcome for pyeloplasty in horseshoe kidneys.

Ureterocalycostomy

Ureterocalycostomy has been previously described in the management of PUJ obstruction, but mainly for single systems. Jameson described the successful application of ureterocalycostomy in an eight-month-old boy with PUJ obstruction in a solitary kidney, with a nearly complete intrarenal pelvis [343]. Hawthorne modified the technique, advocating sufficient lower renal pole amputation to prevent the possible complication of ureterocalyceal stenosis [344]. More recently, a series of 35 cases has been described in which ureterocalycostomy has been employed as the initial procedure in selected cases of PUJ obstruction in adolescents and children, including 15 cases of Horseshoe kidney; no late deterioration was noted [342].

The four cases with a horseshoe kidney described above were treated by ureterocalycostomy following a failed initial surgical procedure. The series includes three cases where initial surgical intervention was an Anderson-Hynes pyeloplasty, two of which were followed by unsuccessful redo procedures. The third case was not suitable for a redo pyeloplasty owing to insufficient remaining ureteric length and the fourth had only division of pelvic adhesions as the initial procedure. In each case the position of aberrant blood vessels appeared to compress the PUJ or impede the formation of a tension free Anderson-Hynes pyeloplasty.

Ureterocalycostomy has the advantage of overcoming the configuration which contributes to obstruction, viz., passage of the ureter over the isthmus, compression from lower pole vessels and a relatively non dependant rotated pelvis, but has the obvious disadvantage that it is necessary to dissect renal parenchyma, increasing the risk of stenosis at the anastomotic site; the technique seems to have been effective, as the procedure was successful in the four cases in this series.

Given the usual success of the Anderson-Hynes pyeloplasty, ureterocalycostomy would not necessarily be the first line procedure in PUJ obstruction with a horseshoe kidney. It does however have a useful role in cases of PUJ obstruction in which dependent drainage does not seem possible without connection of the ureter to the lower pole calyx, either at its caudal or anterior aspect.

Urolithiasis in Australian Aboriginal Children

Introduction

Urolithiasis in adults has been extensively reported in the medical literature, while comparatively little has been written on paediatric urolithiasis [351]. These few reports reveal considerable variations in the incidence, aetiology and composition of childhood stones in different parts of the world [352-355]. The Aboriginal children of Australia have a high incidence of urolithiasis and this updated, retrospective review highlights the peculiarities of urolithiasis in this population and the success with alkaline dissolution therapy.

Patients and Methods

Thirty-six Aboriginal children, with urolithiasis were seen in the Urology Unit of Adelaide Children's Hospital from 1985 to 1991. The hospital records and radiographic results of all patients were reviewed with regard to age at diagnosis, sex, past medical and surgical history, presenting symptoms, results of biochemical, microbiological and radiological investigations, site, number and composition of stones, management and outcome.

Results

The sex and age distribution of 36 children is shown in Table 8. More than 70% of the children were below the age of two years. Twenty-six patients presented with fever (72.2%), nine with abdominal pain (25%), two with dysuria (5.6%), four with haematuria (11.1%) and two patients with acute renal failure. Thirty two patients had been treated for urinary tract infection (UTI) previously, 20 patients had a history of recurrent gastroenteritis, seven had been treated for recurrent respiratory infections and six had evidence of past or present otitis media. All 36 patients were below the 50th percentile in weight (22 were below the 10th percentile, eight were between 10-20th percentile and six

were between 25-50th percentile). All were below the 10th percentile for height. However, the weight and height measurements were based on percentile charts of Caucasians. A UTI (colony counts $>10^5$) was present in 27 patients at the time of presentation. A single organism was isolated from 19 patients and significant mixed growth was seen in eight. *Escherichia coli* was isolated from 19, *Proteus* in six, *Klebsiella* from six, *Streptococci* in three and *Haemophilus influenzae* in one.

Sex	Age Distribution (years)		
	<2	<5	<12
Male	15	5	3
Female	10	3	0

Table 8: The sex and age distribution of 36 Aboriginal children with urolithiasis.

Biochemical estimation of blood urea and creatinine were normal in 34 and two presented with acute renal failure; nine showed compensated metabolic acidosis. Serum calcium, phosphate and uric acid were normal in all patients. Estimation of urinary calcium, phosphate and uric acid were normal in the 18 patients tested.

The presence of stones was initially investigated with US and was positive for stones in all except one, in whom the presence of a stone in the renal pelvis was shown by IVP. Pelvicalyceal dilatation was shown on US in 15 patients.

Eighteen patients had an IVP prior to referral, all of which were abnormal; two with a non-functioning kidney and the other 16 showed at least one calculus. A DTPA scan was performed in 21 patients out of which 14 were abnormal, showing either delayed drainage

(12 patients) and/or reduced function (five patients). The presence of a horseshoe kidney in one patient was seen in both the IVP and DTPA scan. An MCU was done in nine patients and showed VUR in one. Pelviureteric junction obstruction was noted in the IVP in an eight-year-old male, but no malformations were detected in the other patients.

The anatomic location of the calculi in the urinary tract is shown in Table 9. Stones were unilateral in 26 patients and bilateral in ten; two patients had a stag-horn calculus. The distribution of bilateral stones was as follows: kidney in six, ureteric stones in two patients, and kidney on one side with a ureteric stone on the opposite side in two.

Location	No. of Patients
Kidney & Pelvis	34
Ureter	4
Bladder	1

Table 9: The anatomic location of the 39 calculi in the 36 patients.

Chemical dissolution was attempted for radiolucent stones which were not causing a significant obstruction: the therapy consisted of high fluid intake, oral antibiotics and either potassium citrate or sodium bicarbonate to achieve a urinary Ph>6.5. Fourteen patients were put on alkaline therapy, eight without surgical intervention and, in the remainder, following surgery. Five of the eight non-operated patients had their stones dissolved with alkaline therapy, while in one patient with bilateral pelvic stones, the stone disappeared on one side, and the other was removed surgically. In two other patients, the stones continued to increase in size while on alkaline therapy and hence were removed at operation. Post-operative alkaline therapy was also used for patients with minor residual concretions.

Calculi were removed primarily whenever there was significant obstruction, reduced function or when the stones were large. A total of 27 patients underwent stone removal; this included pyelolithotomy with pyeloplasty in one patient with a PUJ obstruction; a `kidney split' (longitudinal nephrotomy) in three patients, two of whom had stag-horn calculi and one a large pelvic stone with an intrarenal pelvis: One patient had Dormia basket extraction of bilateral ureteric stones.

Stone material was analysed in 29 patients. Twelve patients had urate as the predominant constituent and 16 had a mixture of urate and oxalate. In one patient, calcium oxalate and calcium carbonate were the major constituents. The period of follow-up ranged from five months to six years with a mean of 2.5 years. Apart from routine clinical examination and urinalysis, a follow-up US was performed in all patients. The first post-therapy US was within three weeks after initiation of alkaline therapy and three months following surgery; subsequent scans were at intervals depending on clinical indications. Fourteen patients with abnormal preoperative isotope scans had a post-operative nuclear medicine scan, with improvement in function in three, deterioration in one and no change in 10. A residual stone detected in one patient three months following surgery dissolved with alkaline therapy.

Discussion

The incidence of paediatric urolithiasis has progressively declined in industrialized societies, with near disappearance of vesical stones [353]. Upper urinary tract stones constitute nearly 80-90% of childhood urolithiasis in reports from North America and Europe [353]. Uric acid stones accounted for only 7.6% and 1.3% of all childhood stones in the North American and European series respectively. North American studies also found an average age higher than in this study; 11.2 years [355] and 15.7 years [356] compared to most being below two years of age in this group, and an average of 2.5 years in the Darwin area [357]. On the other hand, studies from Thailand, India and Indonesia show a high incidence of childhood stones, predominantly in the bladder. Vesical stones noted in these countries are mainly composed of ammonium acid urate and calcium oxalate [358] as are the renal and ureteric stones in Aboriginal children [359]. High cereal and low protein intake, recurrent gastroenteritis and dehydration are suggested to be responsible for childhood stones in the Australian Aborigines and those from developing countries [359-364].

Epidemiological studies on dietary habits of Aboriginal children in Central Australia show that they are often breast fed till the age of two years, supplemented by only cereals and bread; eggs and meat are only included in the diets of older children [360]. The urine concentration of ammonium ion and uric acid of children fed cereals is high, whereas the excretion of phosphates and pyrophosphates is low and as pyrophosphate is an effective inhibitor of calcium oxalate crystallization, a high cereal diet favours urolithiasis [360]. This dietary pattern may be a major factor in the higher incidence of urolithiasis in Australian Aboriginal infants and toddlers.

Nearly 70% of the children included in the above study were less than two years old, in contrast to the children with vesical stones from Thailand, India and Indonesia who were between three to seven years of age [303]. This may be due to the earlier screening

facilities available in Australia or more probably to the dietary patterns of Australian Aborigines noted above.

The normal values of serum and urinary uric acid in the above patients may be due to the temporal difference between the initiation of stone formation and clinical presentation, by which time the dietary pattern might have been altered. It is difficult to explain the predominance of upper tract stones in this series compared to the vesical stones in children from developing countries. In these children the vesical stone formation is explained by the crystallization occurring in the bladder where the urine remains stagnant for a period of time [358,360]. The Aboriginal children have a smaller renal pelvis compared to Caucasians, which may favour upper tract stone formation (Fig. 65). It is also possible that the vesical stones seen in children from developing countries may in fact originate in the upper tract and the increased incidence of upper tract stones seen in the recent studies in Thailand supports this [364].

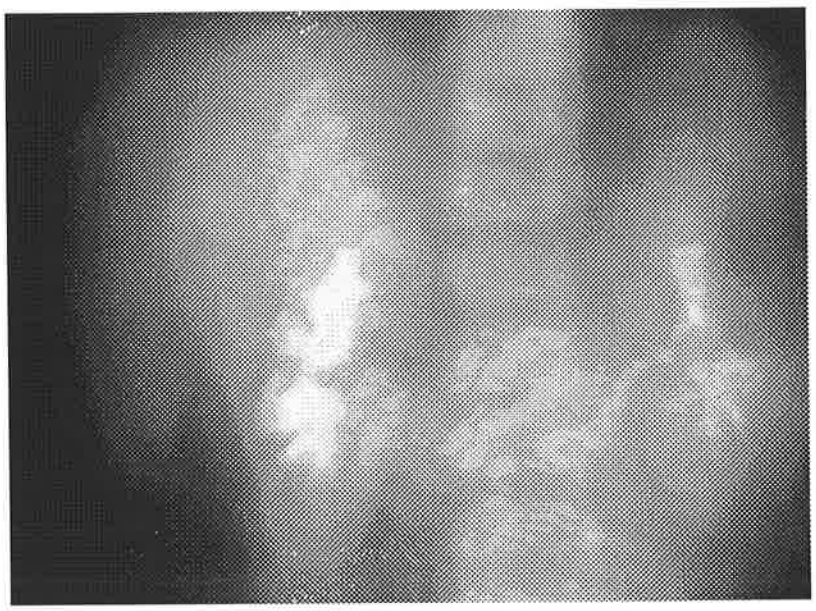


Figure 65: Intravenous urogram showing the small, intrarenal pelvis, with a calculus on the left side.

Escherichia coli, the major cause of urinary tract infection in the above cases, is probably secondary to the calculus rather than the initiating factor as even in the six patients with *Proteus* infection, the stones consisted of urate crystals, further suggesting that infection is secondary. The management of paediatric urolithiasis has been traditionally based on the use of drugs and surgical treatment. With the increasing use of endoscopic techniques and extracorporeal shock wave lithotripsy (ESWL) in adults, these techniques are being evaluated in some centres for childhood urolithiasis [353,355]. Dissolution of uric acid stones by alkalinisation of urine has been used in adult practice for many years but reported use in the paediatric age group is limited [357]. For successful alkaline dissolution certain criteria have to be met:

i) The stone must be non obstructing or only partially obstructing,

ii) Significant hydronephrosis must not be present.

iii) The patient must be under close follow up by physicians familiar with the problem.

Urinary pH should be frequently checked to verify adequate alkalinisation and parents should be warned that as the stone becomes smaller, it may move down the ureter from the kidney or pelvis and produce ureteric obstruction. In my experience, alkaline therapy was found to be safe and effective. Many Aboriginal children are treated successfully with alkalinisation of their urine without referral to a urology unit. Use of alkaline irrigation solutions in the urinary tract is an alternative form of chemical dissolution and may be indicated in patients with multiple or large stones or with stones at multiple sites in the urinary tract [365].

Views vary on surgical removal of staghorn calculi, especially where the choice between longitudinal nephrotomy ("Kidney-split") and extended pyelolithotomy is concerned. The advantage of the former method is that it gives good access for the removal of staghorn and multiple renal calculi [363]. The extended pyelotomy has a lesser traumatic effect on the kidney parenchyma but is technically more difficult in Aboriginal children due to the small size of the renal pelvis alluded to above. Percutaneous endoscopic lithotomy and ESWL do have a role in the management of these patients that will probably expand as technology improves.

Conclusions

In conclusion, this review of paediatric urolithiasis in the Australian Aborigines highlights the high incidence of upper urinary tract calculi, the predominance of urate and oxalate stones, the importance of dietary factors in the aetiology, the effectiveness of alkaline therapy and the rarity of recurrent stones. Further studies are required to elucidate the anatomical peculiarities of the renal pelvis and variations in the metabolism of urate that may be important in aetiology of the stones in this select group.

Megacystis-Microcolon-Intestinal Hypoperistalsis Syndrome

Introduction

Megacystis-Microcolon-Intestinal Hypoperistalsis syndrome (MMIHS) is a rare, autosomal recessive condition, characterised by those features suggested by its name, plus malrotation and hydroureteronephrosis. Far from being a simple case report, this section of this thesis is a compilation of 33 data points from each of the contributions to the literature, namely the 39 publications which describe 35 female and 23 male patients [366-404]. The 58 patients have all had intestinal hypoperistalsis and megacystis, with bladder volumes often in excess of 400 ml at birth [378,384,389,395,396,400,402]. There are, however, some children with otherwise typical MMIHS who have not had one or more of either malrotation [374,400], microcolon [373,381,393,403], microileum [366,372,382,403] or hydroureteronephrosis [378,380,382,395,398,404]. Infrequent findings in the infants are shown in Table 10. With regard to the pregnancy, polyhydramnios has been recorded in several cases [380,394,395,397,398,400,403,405].

The prognosis is poor, with only 17 of the 58 patients living beyond six months, and only one beyond five years [403], despite the presence of an apparently normal compliment of colonic ganglion cells recorded in most patients.

This case report describes a child with MMIHS and the additional features of a dysphonic cry, brachycephaly, low set ears, over-riding second toes, hydrometrocolpos and segmental colonic dilatation.

Prune belly [367,369,382,387,394,398,402]

Undescended testes [378,382]

Vesicoureteric reflux [379,387]

Exomphalos/umbilical hernia [366,405]

Renal dysplasia [367,369,404]

Urachal diverticulum [380,385]

Talipes, Meckel's diverticulum, patent ductus arteriosus and mandibular hypoplasia [401]

Cardiac rhabdomyoma [374]

Table 10: Uncommon isolated features recorded in cases with MMIHS.

Case Report

The index case for this study was a female infant, born at 35 weeks gestation, who weighed 2.1 Kg at birth. Her parents were a healthy, non-consanguineous Melanesian couple, in Papua New Guinea. The pregnancy and delivery were normal for a multiparous woman. However, the outcome for previous progeny is not known.

On examination the baby was hypotonic with a feeble dysphonic cry with brachycephaly, low set ears, and over-riding of the second toes. Her abdomen was grossly distended by two discrete masses, the anterior of which arose from the pelvis and extended to the level of the umbilicus; the posterior lesion occupied the left lower quadrant. Other clinical features included the absence of bowel sounds and the ability to express urine by lower abdomen pressure. The external genitalia were unequivocally female and other systems were

clinically normal. Clinical assessment of autonomic function (tear production, heart rate, blood pressure, respiratory rate and pupil responses) was normal.

A plain abdominal radiograph illustrated a dilated stomach and duodenum with absence of gas in the distal bowel (Fig. 66). Abdominal US showed a cystic mass occupying the pelvis, which appeared to be the vagina, and dilation of the left pelvicalyceal system; intravenous pyelography confirmed left sided hydronephrosis with lateral displacement of both ureters by a homogeneous cystic mass (Fig. 66). The vagina was catheterised and 85 ml of serosanguinous fluid drained, following which the cavity delineated radiographically. Renal function studies and chromosome analysis were not available in Papua New Guinea.

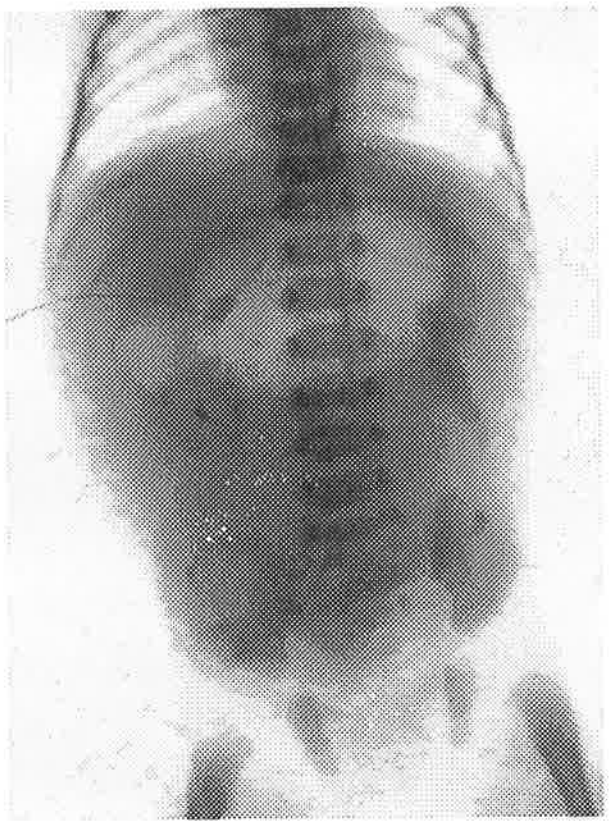


Figure 66: An IVP demonstrating left hydronephrosis and wide separation of the ureters because of an enlarged bladder and vagina with no gas beyond the duodenum.

The baby did not pass meconium and had persistent large nasogastric aspirates. She was therefore commenced on total parenteral nutrition. A laparotomy was performed because of the failure to pass meconium, persistence of the lower abdominal mass after drainage of the vagina, the dilated stomach and failure of gastric emptying.

The vagina, uterus and bladder were found to be dilated, though the urethra and the vaginal orifice were both patent. The distal half of the Sigmoid colon was also distended, but the more proximal colon was collapsed and there was no evidence of ano-rectal stenosis. The stomach and proximal duodenum were dilated, though not because of the midgut malrotation, which was present but not obstructive. The small bowel was collapsed and aperistaltic. Therefore, after correcting the rotational deformity, a gastroenterostomy was fashioned, and a low descending colon stoma and a mucous fistula were created.

Post-operatively the baby developed signs of necrotizing enterocolitis and was found to have a *Klebsiella aeroginosa* urine infection. She developed oliguria and coagulopathy, and died on the sixth post-operative day.

Autopsy confirmed the grossly distended lower colon, bladder, vagina and uterus. The separation of the bladder and vagina, with normal outlets from the bladder, vagina and rectum, was also confirmed. The gastroenterostomy was intact and there were no duodenal or jejunal atresias. Segments of intestine showed evidence of necrosis consistent with the clinical diagnosis of necrotizing enterocolitis. The vagus and laryngeal nerves were macroscopically normal, as were the larynx and the vocal cords. Histological study of the specimens from several sites along the gastrointestinal tract, including the dilated distal Sigmoid colon, showed normal ganglion cells.

Discussion

This child is the 59th to be described with features of the MMIHS, a condition first identified in five girls, by Berdon *et al.* in 1976 [370]. All of the 59 babies were of average weight (3.1± 0.8 kg) and 75% were born after 35 weeks gestation, as was the above index patient.

The principle components of the syndrome are an enlarged bladder, intestinal hypoperistalsis and a collapsed colon. However, four cases in the literature did not have a collapsed colon; these were patients who died at 11 months, five and 14 years [393,403,403] with a uniformly dilated large bowel, raising the possibility that this feature might accompany longer term survival. However, the above case and another, in which the child died at four days [366], suggest that segmental dilatation can be present at birth as an integral part of the phenotype. One other patient had a normal colon [384].

Most reports indicate the condition to be fatal in the newborn period from renal failure and sepsis, as in this case.

There have been a number of uncommon features associated with individual cases of MMIHS (Table 10). Previously unrecognized abnormalities in this case were dysphonia, brachycephaly, low-set ears, over-riding toes, and a cavernous vaginal vault and dilated uterus.

The antenatal diagnosis of MMIHS is a recent achievement that has already been reported in 24 cases. Two were identified as having megacystis as early as 16 weeks gestation [373,388] and, of the 20 cases with known gestation at diagnosis, the average timing of detection was 28±7.7 wks, usually with both megacystis and hydroureteronephrosis. A number of studies showed progression of the changes during pregnancy [373,377,394], from a normal US at 19 weeks to megacystis at 29 weeks [373], and from minor hydronephrosis at 21 weeks to megacystis at 25 weeks [377]. Intrauterine death [376] has

been recorded and one pregnancy was terminated because of the prenatal changes seen following a history of MMIHS in a sibling [388].

It is generally accepted that the condition is autosomal recessive [388,401] as there have now been seven cases with consanguineous parents [369,381,388,401] and seven pairs of affected siblings [369,370,376,377,388,394,401]. One infant had a brother with a Prune Belly, but not the other features of MMIHS. No chromosome abnormalities were found in those cases whose chromosomes were examined [388,394,404]. A report records Clomiphene use during the pregnancy of one case [375], but there have been no other reports linking the use of Clomiphene, and no other aetiological agent has been suggested.

In MMIHS there is functional obstruction of the large bowel and bladder, but histological features have been somewhat inconsistent, although most authors have found normal numbers of gut ganglion cells, as found in the Papua New Guinea case. However, Krook found immature ganglion cells in the rectum [382], Young *et al.* suggested that there was an increased number of ganglion cells in one specimen and an increase in the number of nerve fibres in the rectal biopsies of three other cases [403] and Kirtane *et al.* suggested there was a degree of dysganglionosis with fewer and shrunken neurons in some part of the affected bowel [381]. Taguchi *et al.* studied the distribution of several neurotransmitters in the small bowel of their patient, finding that there was an increase in Substance P and Leucine-Enkephalin, but a reduction of the levels of Vasoactive Intestinal Peptide and Peptide Histidine Methionin [395]. Bindyl *et al.* also used histochemical analysis and found abnormal neurons [371]. Alternatively, Puri *et al.* proposed that the disorder might result from a myopathy, based on the presence of vacuolar degeneration in electron micrographs of smooth muscle cells of the ileum and bladder [389]. It is difficult to bring these diverse findings together into a unifying hypothesis for the aetiology. Obviously, further detailed histologic and histochemical studies are required.

There is little evidence to suggest that intervention improves prognosis. In MMIHS, most reports are of single cases, many of which have had a number of operations; of the 58 reported cases, seven had a laparotomy only, 18 had a vesicostomy and 16 had a small bowel stoma of one form or other. Up to three operations per patient were performed and 25 cases had their first procedure in the first two days of life. Only 12 of the 58 cases in the literature did not have a surgical procedure and, despite their surgery, most of the 46 with intervention died. It seems that almost all children with MMIHS will die in spite of early aggressive medical and surgical treatment and, while it is usually possible to establish urinary drainage and to use antibiotics to minimize the risk of urinary tract infection, it is not possible to overcome the absence of gut motility.

Venous Access via the Renal Vein

Introduction

Long-term central venous access for intravenous nutrition and renal dialysis is fraught with complications. Thrombosis of large central veins is a particularly difficult problem that has become more common as more children are continued on long-term home parenteral nutrition. The patient in this report required venous access, not only for long-term nutrition but also for haemodialysis, prior to and following renal transplantation.

Case Report

This infant was born at term after a normal pregnancy and delivery. On his second day of life he developed necrotizing enterocolitis secondary to *Clostridium perfringens* infection and developed overwhelming sepsis. Post-operatively he developed renal failure and was shown to have renal cortical necrosis on biopsy and US scanning. In addition, he developed idiopathic protein losing enteropathy and required on-going support with parenteral nutrition. He required venous access for both dialysis and parenteral nutrition. This was initially through the right internal jugular vein, then the left external jugular vein, followed by the right external jugular vein. An attempt to establish peritoneal dialysis failed, due to obliteration of the peritoneal cavity by adhesions. At 11 months of age, after several episodes of central line sepsis, a further attempt was made to place a central venous line via the superior vena cava, which was abandoned; and a right femoral vein catheter was therefore inserted. It was subsequently felt that the femoral vein line would limit the transplant options in the future and a right renal vein catheter was inserted. The concern about the use of the lower limb venous access was supported by the findings of a later study showing a syncytium across the inferior vena cava (Fig. 67). The renal vein catheter worked well, but was again further complicated by infections, which resolved after changing the catheter over a guide-wire on two occasions. Gut perforation and peritonitis, secondary to cardiovascular instability complicated a renal transplant, performed at 40

months of age. The right renal vein catheter was changed a third time over a guide-wire; however, the line sepsis did not resolve and the line was removed and temporarily replaced with a groin vein catheter, by the Intensive Care team. Despite good function of the renal transplant, the improvement in renal function did not bring about the anticipated improvement in gut function, necessitating on-going parenteral nutrition. With due consideration to the possible need for venous anastomoses during further, subsequent renal transplant episodes, the groin catheter was removed and substituted with a left renal vein line. This functioned well despite the known occlusion of the inferior vena cava at the level of the diaphragm (Fig.68).

Operative Technique

The patient was placed on the operating table in a supine, but slightly twisted position, so the abdomen was in a semi-lateral position and the chest virtually horizontal. The kidney was approached retroperitoneally through a muscle cutting incision below the 12th rib. The kidney was mobilised on its pedicle and a purse-string 5/0 prolene suture placed in the posterior aspect of the mid-portion of the renal vein. A Cook single lumen cuffed catheter was passed through a subcutaneous tunnel and inserted into the renal vein. The position was checked with the image intensifier leaving sufficient catheter in the inferior vena cava to allow for growth. The wound was closed in layers with polyglycolic acid sutures. The catheter positions are shown in Figure 68 + 69.

Changing the catheter over a guide-wire was performed in the operating theatre under general anaesthesia. Dissection was carried down to the catheter in the subcutaneous tissues, the catheter clamped and divided, and a wire passed through the catheter, into the central vein. A dilator and peel-away sheath was passed over the wire and the new catheter was fed into the vein, and the position checked.

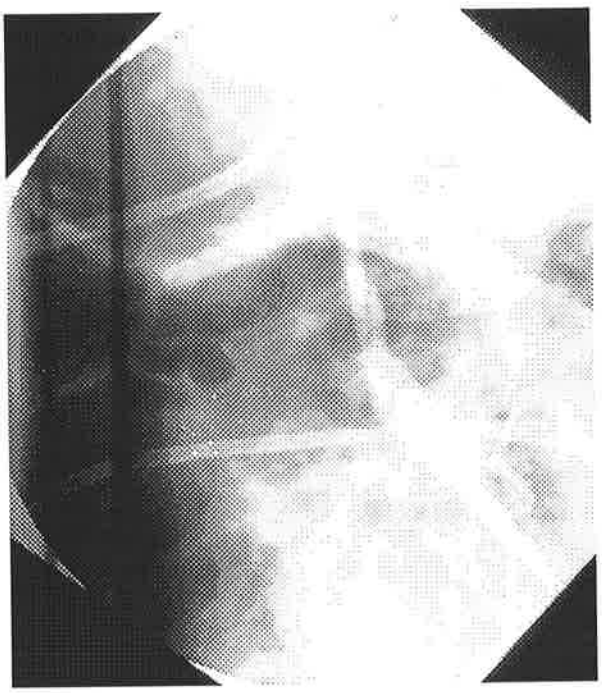


Figure 67: Venogram showing the synechium in the IVC and occlusion of the right common iliac vein.

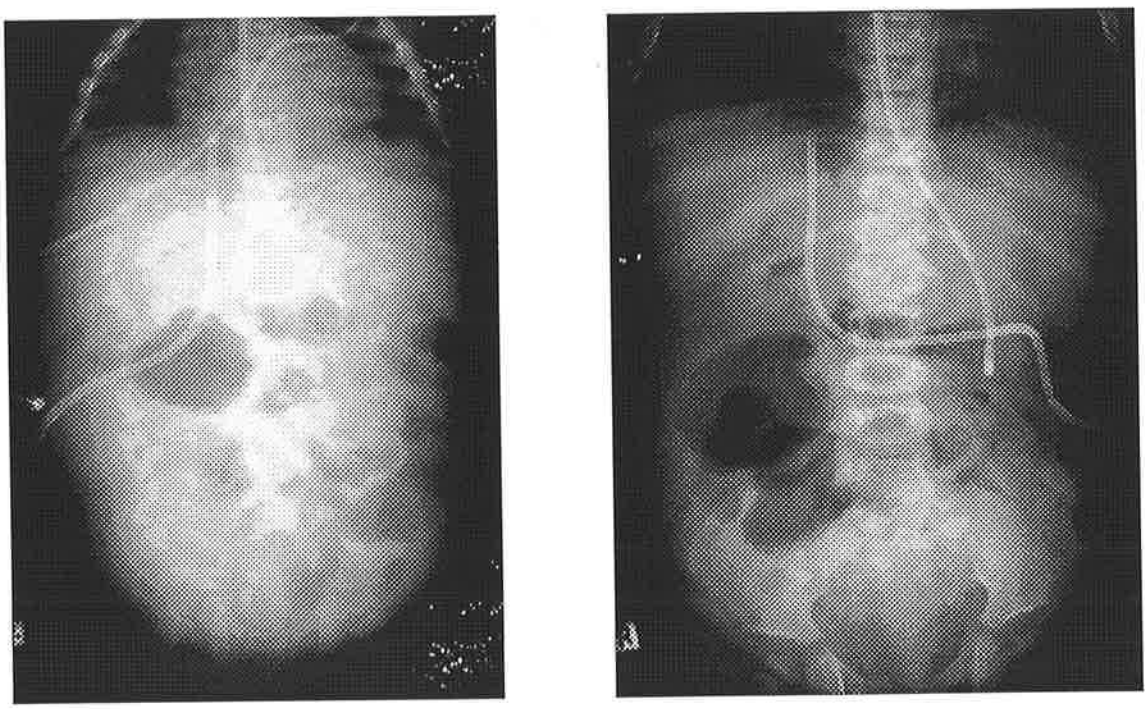


Figure 68 + 69: Radiograph of the catheter in situ in the right and left renal vein.

Discussion

In this patient, central venous access was required for both dialysis and parenteral nutrition. In addition it was felt important to try to preserve patent iliac veins for future renal transplantation. Thrombosis of large central veins is an uncommon but well recognised complication of central venous catheterisation with an incidence of 5-10% [406]. In children requiring long-term vascular access it is not unusual to experience increasing difficulty in replacing lines over the duration of the illness. A number of alternative sites of access have been described for use when the more conventional subclavian and jugular veins are not accessible. Inferior epigastric and saphenous veins have been used to gain access to the external iliac vein however this is associated with an increased risk of iliac vein thrombosis and thus jeopardise future renal transplantation. The gonadal vein has been used for long-term central venous access [407] but has not found universal favour [408] presumably as its smaller diameter makes it more prone to thrombosis and does not allow for the high flows required for dialysis. The internal mammary vein has been described for placement of a catheter in the superior vena cava [409] but would rely on good flow rates in the superior vena cava and patency in the distal innominate vein, a combination which may not be present when conventional access is failing. A novel approach described by Torosian *et al.* [410] involved passing a Dormia basket into the superior vena cava from below and maneuvering it to snare a catheter introduced into a thrombosed axillary vein under vision. The azygous vein is a well described route for central venous catheter placement but requires a right posterolateral thoracotomy for placement [411,412]. The catheter should be inserted below the junction between the azygous vein and the superior intercostal vein so that collateral flow in the latter vein is not interrupted. Direct cannulation of the right atrium has also been suggested, via an anterior right thoracotomy through the bed of the third costal cartilage but has not been widely reported in clinical practice [411].

In recent years long-term catheters have been placed into the inferior vena cava either directly at open operation [413] or using a percutaneous approach which may be either

direct or transhepatic [413-415] and has been reported for use in haemodialysis [408]. The theoretical risk of hepatic vein thrombosis secondary to such catheter placements has yet to be realized. The commonest new problem encountered in these series has been dislodgment of the catheters due to growth of children, or secondary to abdominal distension, where the catheter was tunneled to the anterior abdominal wall. A search of the literature and Medline did not find any other description of the use of renal veins for central venous access. The renal vein has the advantage of allowing the use of a catheter large enough for dialysis while giving a longer intravascular length to decrease the likelihood of dislodgment due to growth. The technique has the disadvantage of requiring an open approach for placement in a small child and an increased risk of renal vein thrombosis. Concerns regarding renal vein thrombosis are clearly less serious in a patient already requiring dialysis for renal failure. By using this approach, rather than the external iliac vein, it was possible to minimize the loss of pelvic veins needed for subsequent transplantation. Necessity has pushed clinicians into exploring new routes of access for long-term catheterisation of central veins; this paper presents another approach that may be used when options are limited, particularly where future transplantation demands preservation of iliac veins. When renal failure is already present, use of the renal veins for venous access has proven to be a useful alternative.

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Presentations

Bangladesh Society of Surgeons, Dhaka Paediatric Urology Symposium, June 1998 ***(Visiting Professor).***

1. Bladder exstrophy.

Royal Australasian College of Surgeons ASC, Sydney, May 1998.

2. Pyeloplasty in the horseshoe kidney.
3. Medical Management of Phimosis.

Royal Australasian College of Surgeons meeting, Adelaide, August 1996.

4. Phimosis - is circumcision necessary?

Royal Australasian College of Surgeons, Annual Scientific Congress, Melbourne, May 1996 ***(Invited).***

5. History of bladder exstrophy.

South Australian Urology meeting, October 1995.

6. Bladder-neck transection in a sheep model.

International Paediatric Surgery and Radiology meeting, Melbourne, March 1995.

7. Ureterocystoplasty in infants.

Asian Association of Surgeons, Bali, March 1995.

8. Ureterocystoplasty - the ideal bladder augmentation.

South Australian Urology meeting, October 1994.

9. Ureterocystoplasty in infants.

Royal Australasian College of Surgeons meeting, Adelaide, August 1994.

10. Extraperitoneal ureterocystoplasty.

Paediatric Urology Club, Adelaide, November 1993.

11. Extraperitoneal ureterocystoplasty.

Urology Society Australasian meeting, Hobart, March 1993.

12. Multicystic kidney: understanding the aetiology.

Paediatric Urology Club, Sydney, November 1992 ***(Invited).***

13. Multicystic kidney.

South Australian Urology Meeting, Adelaide, October 1992.

14. Multicystic kidney: the radiological anatomy.

Australian Urology Society Meeting, Adelaide, March 1992.

15. Compensatory hypertrophy with the unilateral multicystic kidney.

South Australian Urological Society, October 1990.

16. Preputial patch urethroplasty.

British Urological Association, July 1990.

17. Preputial patch urethroplasty.

Annual Scientific meeting, RACS, Dunedin, New Zealand, 1987.

18. Priapism in childhood.

Glossary

BXO	Balanitis Xerotica Obliterans
CIC	Clean Intermittent Catheterisation
COPUM	Congenital Obstructive Posterior Urethral Membrane
DMSA	Dimercapto-Succinic Acid nuclear Medicine scan
IVP	Intravenous Pyelogram
MAG3	Mercaptoacetyltriglycine; a test of renal function
MCU	Micturition Cystourethrogram
MMIHS	Megacystis Microcolon Intestinal Hypoperistalsis Syndrome
PUJ	Pelviureteric Junction
US	Ultrasound
UTI	Urinary Tract Infection
VUR	Vesicoureteric Reflux