Completely Duplicated Ureters - Various Surgical Procedures

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Congenital complete duplication of the upper urinary tract with separate orifices of double ureters, ureterocele of one of these orifices, or abnormal ectopic position of the orifices beyond the trigonum or even beyond the bladder requires precise diagnosis and diverse conservative and operative management.

Ultrasonography (USG), excretory urography (IVP), voiding cystography (VCUG), cystoscopy, and in selected cases also radioisotope examinations allow for precise diagnosis of the abnormality. USG can reveal duplex kidney and assess its size, thickness of its parenchymal layer, ureteral dilatation and presence of ureterocele in the bladder. IVP helps in assessment of the secretory function of the upper and lower renal system, and the course of the ureters but does not inform one about the presence of separate orifices of a duplicated ureter. Cystoscopy can reliably confirm or rule out the presence of ureteral orificies of a duplicated ureter, and ectopic orifices in the cervix, urethra, vagina or even perineum.

In this anomaly there is frequently hypoplasia of one pole, in the form of a cyst or as a rudimentary fragment of a sclerotic kidney which may be nonfunctional and thus be overlooked in IVP tests. Asymmetry of the kidney or a downward change in the position of the lower pole as a result of pressure exerted by the upper pole cyst should, however, be considered [1].

Ureteral duplication is frequently associated with vesico-ureteral reflux, most often into the lower system, less often into both systems, and rarely into the upper one [11]. The orifice of the lower ureter is most often situated on the lateral side of the trigonum, the reflux is then a consequence of a very short or even absent submucosal tunnel [8,11]. In the case of a common orifice of both ureters or of the presence of a paraureteral diverticulum, the reflux can be involving both systems.

Ectopic ureters mostly drain the upper pole of the duplicated kidney and are usually accompanied by vesico-ureteral reflux, impairment of urine flow or infection [15]. Ureter with an ectopic orifice in the cervix or urethra (in the part proximal to the sphincter or in the sphincteric part) can be revealed by cystoscopy and also during cystography, owing to the reflux into this ureter [16]. The orifice in the urethra, distal to the external sphincter, occurs exlusively in girls and cannot be detected by cystography. Continuous urine outflow with normal periodic or paradoxic voiding are characteristic features of this anomaly [1]. Exceptionally the orifice may be situated in the vaginal vestibulum.

Ureterocele most frequently involves the orifice of the upper pole in the bladder. It may be so large that it fills up a major part of the bladder, and via urine outflow impairment may cause hydronephrosis involving the upper calycopelvic system. In the presence of a large ureterocele, reflux into the other ureter on the same side may also develop. This is most often due to the presence of the ureter in the wall of this cyst and to consequent lack of support, preventing close apposition of the anterior wall of the ureter to the posterior when the bladder is full [8].

Complete duplication of ureters with reflux not responding to conservative treatment requires surgical intervention. If the secretory function of the lower and upper renal system is normal, there is indication for their reimplantation [2,7,8] or for ureteroureterostomy [4,16].

If ureteral duplication is associated with ureterocele or if the orifice of one these ureters is ectopic in the urethra or in the perineum, then most frequently the system drained by these ureters is severely damaged and there is an indication for nephrectomy [2,3,6,15].

Patients and methods

In the years 1979-1993 we treated surgically 83 children, 13 boys (16 %) and 70 girls (84 %), for complete duplication of the upper urinary tract. In 13 cases the anomaly was bilateral. The youngest child treated surgically was 4 months old and the oldest was 15 years old. Mean age 4.5 years. Prior to the operation, for evaluation of the lower and upper urinary system, VCUG in 81 but IVP was carried out in every case. Each patient was assigned a grade of reflux based upon the international classification. No cystoscopy was done routinely; it was performed in 49 children for additional evaluation of the bladder and urethra.

In 18 children 99m Technetium DTPA renal scan was done. All patients were placed on continuous antibacterial prophylaxis after surgical treatment. In all children the control radiological examination comprised VCUG, whereas IVP was carried out in only 42 cases. In the other children who before the operation displayed slight renal damage, the state of the upper urinary system was controlled by USG.

Results

The following operations were done: in 24 cases submucosal reimplantation of the ureters by the classical methods in one block, in 48 cases the ureters were implanted separately after their separation by our procedure, and in 20 cases heminephroureterectomy was performed. Three children were, because of recurrent reflux, operated twice, and two children three times.

Vesico-ureteral reflux into duplicated ureter was found in 74 children, and it was bilateral in 6 cases. Reflux into the lower pole moiety was present in 54 cases (in 6 patients with bilateral duplication the reflux was into both lower pole moieties), in 14 cases the reflux was into both the upper and lower moieties, and in 6 (in 1 with bilateral duplication) only into the upper pole moiety.

In 9 children no reflux was found; in 6 with ureterocele and in 3 with ectopic orifice (5 of them had bilateral duplication). Additional anomalies detected in children with satisfactory function of both systems are recorded in Table 1. In 67 children ureteral reimplantation was performed usually by the Politano-Leadbetter method or by our own procedure. In the years 1979-1983 duplicated ureters were reimplanted in one block, in 24 cases, using the Politano-Leadbetter method in 22 and Cohen's method in 1 patient; in 1 case the submucosal tunnel was only elongated. Before the operation, reflux grade III was found in 18 children (17 duplicated ureters) and grade IV in 8 children (10 duplicated ureters). In 14 cases three ureters were bilaterally reimplanted, because of additional reflux into the single ureter of the contralateral kidney.

Table 1. Associated anomalies

Ectopic ureterocele	4
Ectopic ureter	6
Bladder diverticulum	10
Renal agenesis	1

During the reimplantation of duplicated ureters in one block, one bladder diverticulum and two paraureteral diverticula were removed, three ectopic ureters were reimplanted and three ureteroceles were excised (two from the second kidney system). In one case distal ureteral part was excised after previous heminephrectomy of the second, also duplex kidney.

Follow-up control examinations of 24 children after reimplantation of 27 duplicated ureters showed in 5 children (5 duplicated ureters: 18.5 %) reflux recurrence. Two of them were reoperated on, however, without success. No recurrence of reflux into the reimplanted single ureters of the second kidney was noted. This rather high proportion of reflux recurrence led us to try a new procedure involving separation of the duplicated ureter and reimplantation into separate submucosal tunnels [9].

Between 1983-1993 this procedure was used for reimplantation of duplicated ureters in 48 children. In 40 cases this was their first operation and in 8 cases the children have been previously treated without success by the classical methods. Five of them in our department. In 23 cases there was unilateral duplication of the right urinary tract and in 21 cases similar duplication on left side, while 4 children had bilateral duplication (in one of them reimplantation was done only on one side).

Grade III reflux was present in 17 cases (17 double ureters), grade IV in 16 (19 double ureters), and grade V in 6 children (6 double ureters). One

child without preoperative reflux had ureteral reimplantation done for ectopic orifice of one of the double ureters. Reoperations were performed in 6 cases for persistent grade III reflux, in 1 case for persistent grade IV reflux and in 1 case for persistent grade V reflux.

In these 8 children with reflux persisting after unsucessful operations by the classical methods, reimplantation was done by our own procedure. Three of them have been previously operated on elsewhere without success, and five by us. Two of them have had two operations before.

Reflux into the double ureters and into the single ureter of the second kidney was found in 16 children. However, reimplantation of three ureters was performed in 17 cases - in one additional case this was done for stenosis of the orifice of the single ureter. Bladder diverticulum was excised in 4 cases, paraureteral diverticulum in 3 cases, ureterocele in 3 and ectopic orifice in 1 child. In two children with massive elongation and dilatation of ureter the folding procedure was done [10]; in one of them with ureterocele the ureter was 12 mm in width, and in the second case with orifice stenosis it was 45 mm wide.

Control radiological examination of 40 children subjected to the first operation by our procedure showed reflux persistence in two cases (5 %). As concerns the above-mentioned 8 children treated by reoperation, in only one case reflux into the sclerotic lower part of the duplicated kidney persisted. The presence of duplication with ectopic ureter was the indication for operation in 11 children. Most of them [10] were girls. Table 2 shows the sites of the ectopic orifices.

The ectopic ureter was on the right and left side in 5 and 5 cases, respectively. In one child with bilateral duplication both ureters of the lower systems had orifices in the urethra. VCUG was done in 9 cases; it revealed reflux into the ectopic ureter in 7 of them into the upper system. In one girl with purulent oozing and in another with urine outflow from the orifices at the urethrovaginal septum, cystography was abandoned.

Table 2. Ectopic ureters

Ectopic orifice	Girl	Boys
Bladder neck	3	
Urethra proximal	2	1
Urethra distal	1	
Vestibule	3	
Not identified	1	

Cystoscopy performed in 8 children confirmed the presence of ectopic ureter and showed its orifice. In one child no orifice was found. Heminephroureterectomy was done in 5 cases. In view of the good secretory function of the renal system, in 6 children submucosal reimplantation of ureters was carried out: in 3 by the Politano-Leadbetter technique and in 3 by our own procedure.

Among 14 children with ureterocele heminephrectomy was done in 9, whereas in 5, after excision of the cyst, reimplantation of the double ureters was carried out (by the Politano-Leadbetter method in 2, and by our procedure in three cases).

Excision of the upper or lower pole with the ureter was done in 20 cases. In four of them with bilateral duplication, reimplantation of the contralateral double ureter was performed at the same time or later, because of vesico-ureteral reflux. Three children were subjected to reoperations. In one case the ureteral distal part was excised as reflux into that stump continued; in another case bladder diverticulum was excised. In one boy with sclerosis of the second single kidney damaged by vesico-ureteral reflux nephrectomy was done.

Discussion

Complete duplication of ureters, with good secretory function of both systems and presence of high reflux requires surgical intervention. The various methods proposed include uretero-ureterostomy, pyeloureterostomy; common sheath ureteral reimplantation usually by the methods of Politano-Leadbetter, Cohen or Lich-Gregoire, and our own procedure involving separation of the distal parts of the duplicated ureter and reimplantation in separate submucosal tunnels [9].

Until 1983 in our department we had reimplanted double ureters by the widely used classical methods. In the group operated on by these methods failures have occurred in 18.5 % with reflux recurrence. This rather high percentage of failures, though comparable with that reported from other centres [8], has induced us to seek another, more effective method of reimplantation of duplicated ureters.

The distal end of the duplicated ureter is along the terminal 2-3 cm surrounded by a common sheath (Waldeyer's sheath) that makes the ureters stiffer and less yielding to compression by urine present in the bladder under normal pressure. Moreover the blood supply to the terminal part of the ureter depends not only on the vessels of Waldeyer's sheath but also on those of the branches of the ureteral arteries derived from the inferior vesical artery and middle anal artery, as well as on the longitudinal arteries in the external membrane (adventitia) of the ureter [14].

On these grounds, in 1983 we decided to reimplant duplicated ureters after their separation. This procedure was used in 40 new patients and in 8 cases with reflux persistence after operations performed previously by the classical methods (five of them had been operated on unsuccessfully in our department and three in other centres). Two of these children have had two unsuccessful operations. One of them has been operated on by us by the Politano-Leadbetter method with distal uretero-ureterostomy [15] has been done again without success. After dissection of the ureters and separation by our new procedure, the adequate length of the ureters enabled their reimplantation in two separate submucosal tunnels. Reflux failed to recur after this operation. A similar result was obtained after use of this procedure in the other child that previously had been subjected to two operations by the Politano-Leadbetter method.

Early follow-up and long-term results (in some cases after 10 years) revealed in all children the absence of ischemia, necrosis of the reimplanted ureter and stenosis after operation, these being the most feared complications after separation of ureters [2]. Also the follow-up results support further use of this procedure (Fig. 1). Among children operated on for the first time, in only two cases (2 double ureters) control examinations pointed to vesico-ureteral re-

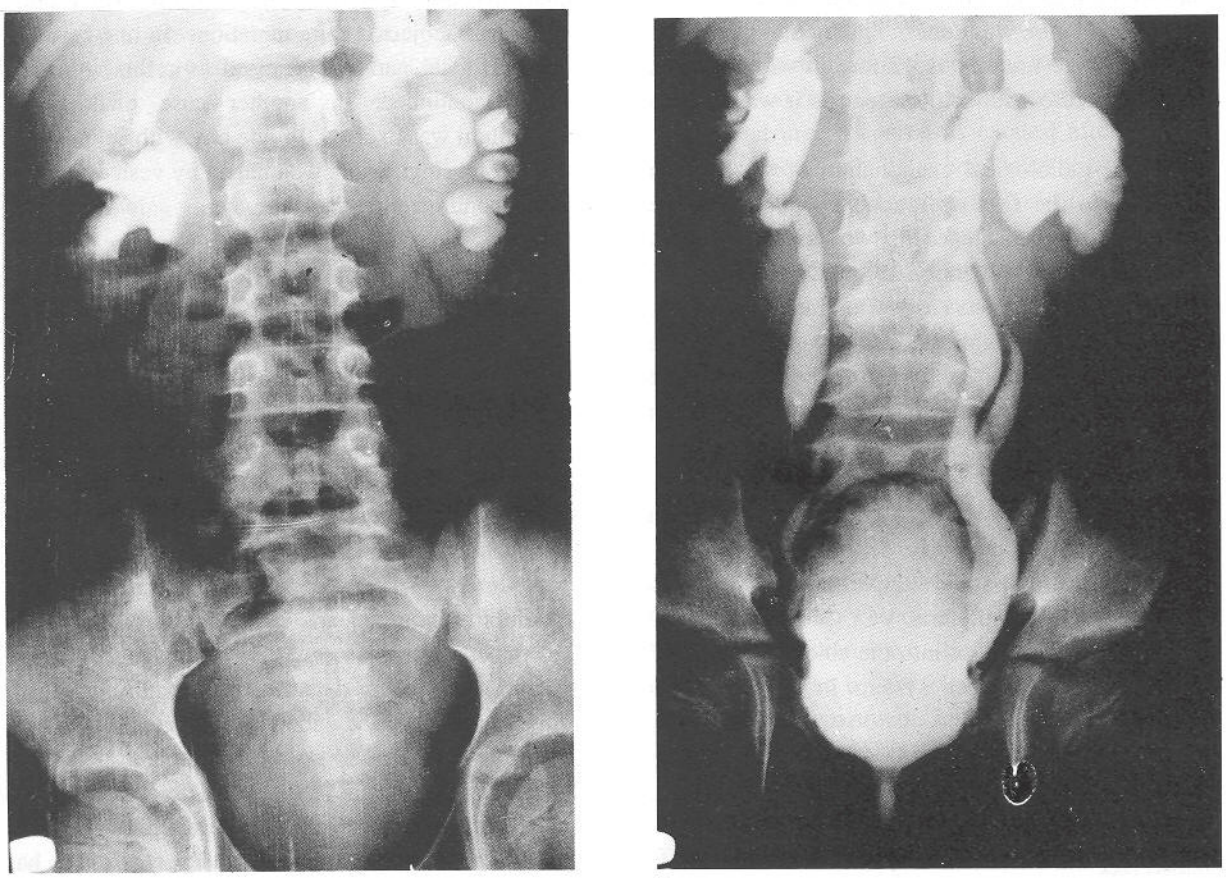

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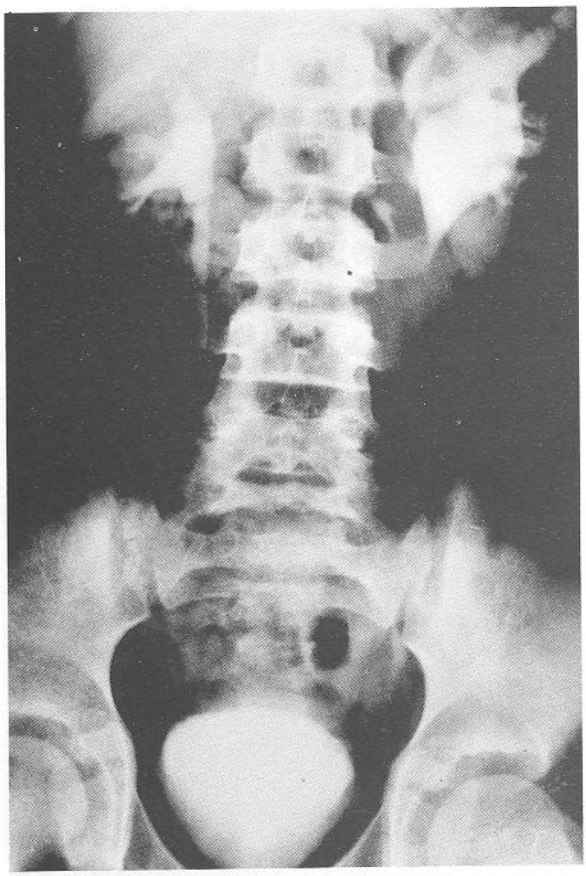
Figure 1. Results of bilateral ureteral reimplantation (on the left double ureters by our procedure). **A)** Preoperative IVP, **B)** Preoperative VCUG-vesicoureteral reflux involving both renal pole on the left and single kidney on the right), **C)** Postoperative IVP, **D)** Postoperative VCUG-small bladder diverticulum.



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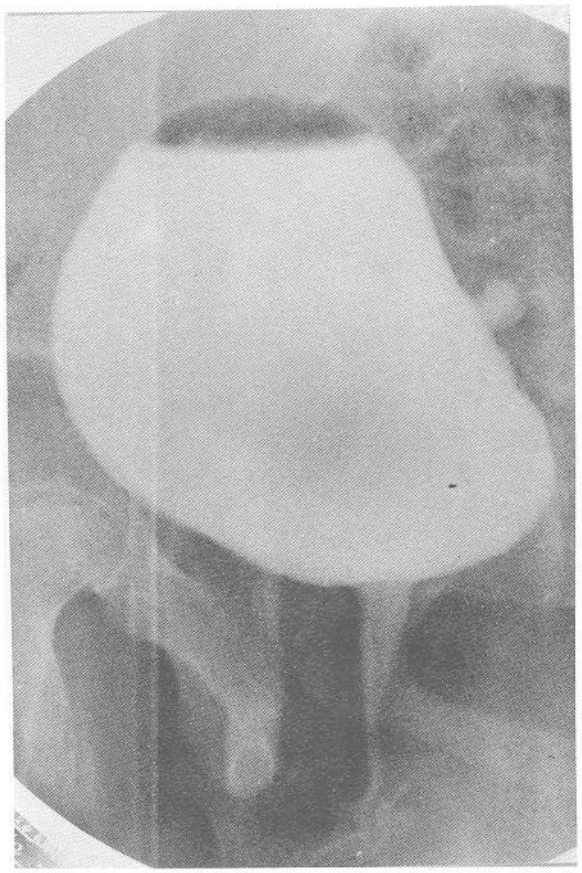

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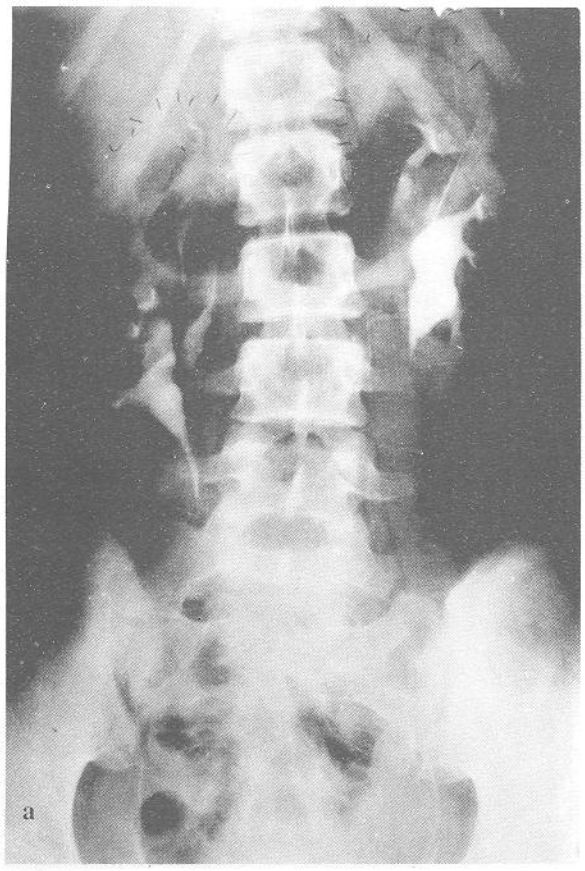
Table 3. A comparison between the results of reimplantations of the double ureters by Politano-Leadbetter (P-L), Cohen methods and Kalicinski procedure

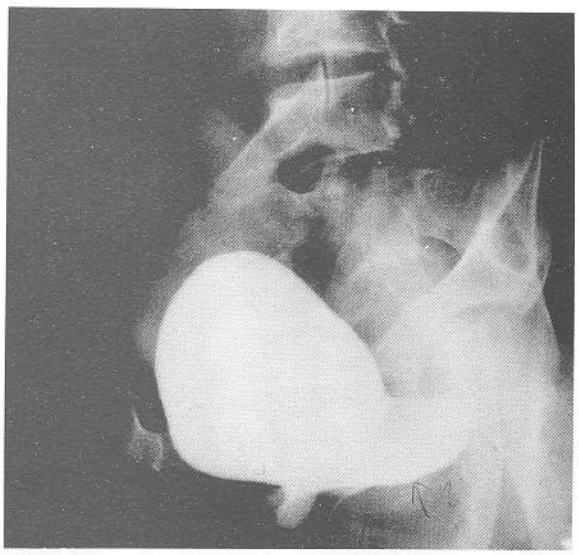
	Children	Double ureters	Success	%
P-L, Cohen methods	24	27	22	81.5
Kalicinski	38	40	39	97.5
Kalicinski+folding	2	2	1	
Kalicinski-reoperation	8	9	8	
Total	48	51	48	94.1

flux. This was observed in a boy in whom the submucosal tunnel was prolonged by suturing the mucosa over the ureter of the lower pole, and in a boy in whom folding of the ureter of the lower pole was done for enormous dilatation of the ureter (45 mm). Only one of the children with reoperations displayed reflux persistence. Because of pre-existing high-grade damage to the renal parenchyma of the lower pole and of persistence of reflux after the operation done by our procedure, lower heminephroureterectomy was performed. In the overall material (Table 3) treated by our procedure (48 children-51 double ureters), persistence of reflux occurred in 3 children (3 double ureters: 6.0 %).

We believe, that the good follow-up results of our procedure, evidently better in our material than those of common sheath ureteral reimplantation, encourage further use of this technique. In children in most cases separation of the ureters is not a problem and is relatively easy technically. Difficulties are encountered most frequently on separation of the distal end along the terminal 1-2 cm where the ureters are closely connected. Injury to the ureters in this part occurred in only one child. After excision of the damaged part, the ureters were reimplanted into separate submucosal tunnels.

Treatment of ureteral duplication with ectopic orifice of one of the ureters in the sphincter part of the urethra requires in most cases excision of the upper renal pole (Fig. 2) displaying nearly completely atrophic parenchyma and markedly dilated ureter. The operation can be done in one step using flank extraperitoneal approach that leaves a ureteral stump [1]. On opening of the bladder it is indicated to dis-





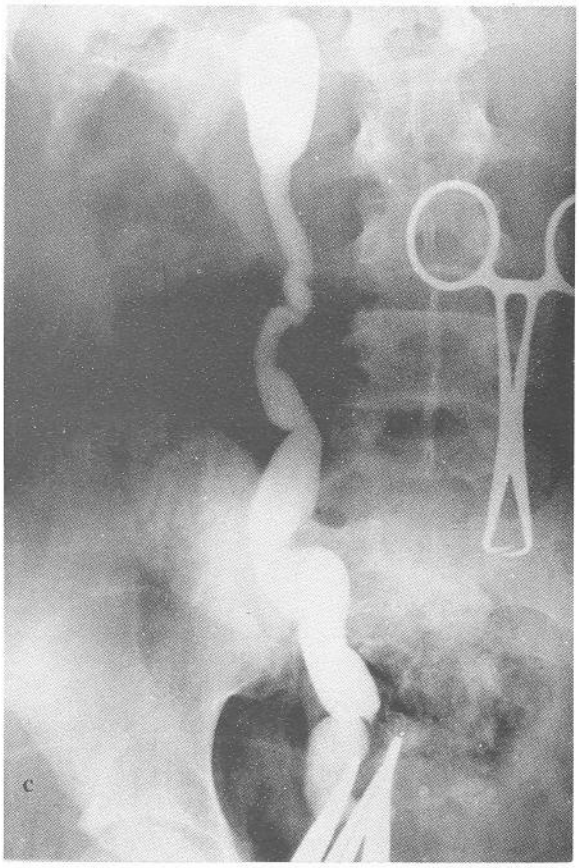
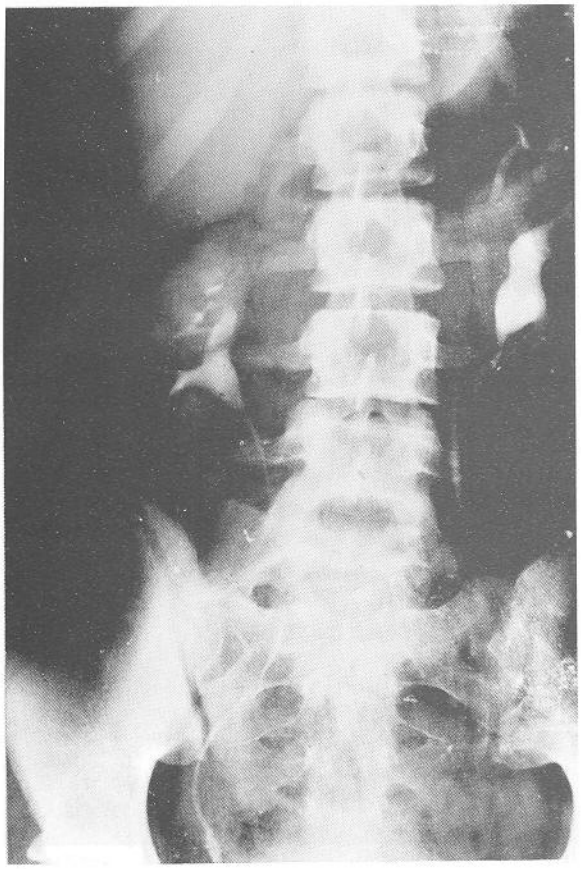


Figure 2. Results of right upper pole heminephroureterectomy. A) Preoperative IVP-bilateral duplex kidney, B) Preoperative VCUG-reflux into ectopic ureter, C) Retrograde pyelography, D) Postoperative IVP, E) Postoperative VCUG.

sect the ureter within the bladder but not from the urethral orifice of the ureter, since this could damage the sphincter. If the ureteral ectopic orifice is not situated in the sphincter area, the secretory function of the upper renal pole is usually good, as urine outflow is not impaired.

In most of these cases it is possible to reimplant the ectopic ureter into the bladder. The orifice of the ureter ending on the margin of the urethral opening or the urethrovaginal septum is usually stenosed. In these cases serious damage to the ureter and respective pole is an indication for heminephroureterectomy. This operation was performed in two girls with these symptoms.

Surgical treatment of ureterocele by transurethral incision is proposed by some authors [6,12]. In our material this was never done. Taking into account the possibility of vesico-ureteral reflux development with further damage to the corresponding renal pole, as well as the presence of additional abnormalities (reflux to the lower pole or to the second kidney), we prefer radical operation with excision of ureterocele, reconstruction of the bladder wall and reimplantation of the ureters (Fig. 3) if both parts of the kidney show good function [13]. If the upper renal pole is significantly damaged or not functioning, heminephroureterectomy is the method of



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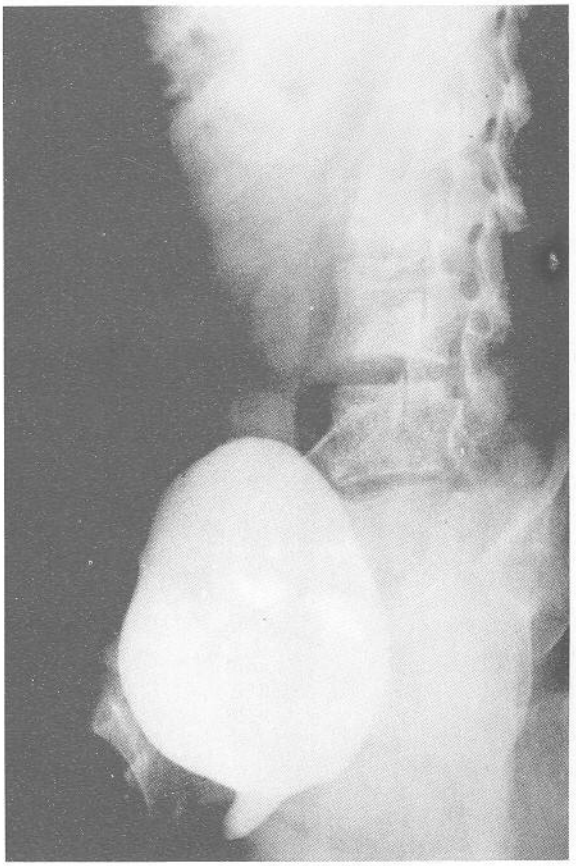

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choice. Radioisotopic examination is of help in taking the decision between partial nephrectomy and attempt at saving the damaged system. Some authors recommend primary heminephroureterectomy without excision of ureterocele [2,3,5].

The rationale for this approach is that postoperatively the ureterocele and the ureteral stump usually will collapse and there will be no subsequent problems. We believe that ureterocele can be the cause of persistent urinary tract infection and we remove it radically. In only one child (out of 9) heminephroureterectomy was performed without cystotomy and without excision of ureterocele. During 3 year period follow up of this case showed one recurrent infection.

The choice of the surgical treatment method (retroperitoneal or by way of cystotomy) may be influenced also by the presence of reflux into the second ureter on the same or on the contralateral side. There are two variants of management in the case of severe damage to one renal pole. One of them involves cystotomy, separation of the ureter from the damaged system and its reimplantation into the abdominal wall skin, after previous submucosal reimplantation of the second, usually normal ureter. The assessment of urine secretion by the decompressed system and control radioisotopic examination are decisive of the further surgical management: reimplantation of the ureter into bladder or its removal together with the corresponding renal pole.

In 7 cases ureterocutaneostomy was done. Unfortunately, follow-up (1-10 months, on the average 2.5 months) that revealed urine outflow of 70-150 ml (urine of low specific weight 1.004-1.005) and radioisotopic examination showed no improvement of the secretory function. In all 7 cases, decompression of the damaged system by ensuring free urine outflow failed to give significant improvement of the secretory function. In view of the young age of these children, and of the presence of damage to the second kidney in some of them, we attempted to save part of the kidney. In all cases we eventually carried out resection of the damaged pole with its ureter.

In the presence of a nonfunctioning pole showing

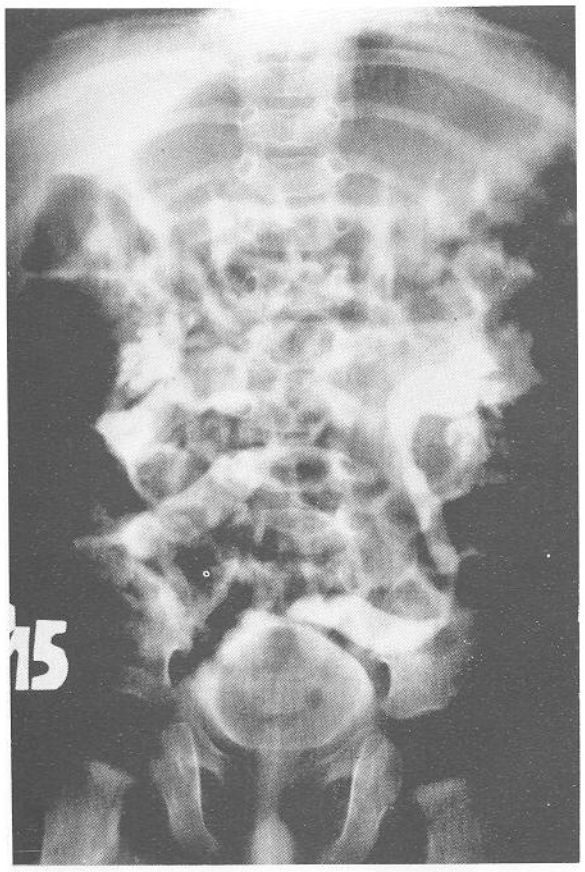

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Figure 3. Results of left upper pole heminephrectomy with reimplantation of the lower pole ureter reimplantation of the contralateral ureter after folding procedure. A) Preoperative IVP-ectopic ureterocele of the left upper pole, B) Preoperative VCUG-bilateral reflux, dilatation and tortuosity of right single ureter, C) Postoperative IVP, D) Postoperative VCUG.

no contrast filling on IVP, one-step resection of the damaged renal pole and nearly complete ureterectomy is preferred. The operation can be done through flank or transabdominal approach. In small children, the linea alba approach gives particularly good exposure to the kidney and enables excision of the dilated ureter down to the bladder and even removal of ectopic ureter from the retrovesical space. Separation of both ureters, the dilated and the normal one, requires a very skillful and precise surgical technique. One-step heminephroureterectomy was done in 13 children; in 4 of them at the same time, the second ureter of the duplicated system was reimplanted, in two also contralateral ureter. In one child with stenosis of the orifice of the single ureter, resulting in ureteral dilatation of 12 mm, reimplantation was done after the folding procedure.

In only one child, during ureterectomy of the dilated ureter of the nonfunctioning pole we damaged the other ureter of normal size. Application of several interrupted sutures and a drain in the retroperitoneal space sufficed for healing. Two children, after resection of the damaged pole by the retroperitoneal approach, had reoperations for recurrent urinary tract infections and persistent reflux into the ureteral stump. In spite of contradictory reports concerning leaving of ureteral stump [3,16] in these two children infections resolved after suprapubic excision of the distal ureter.

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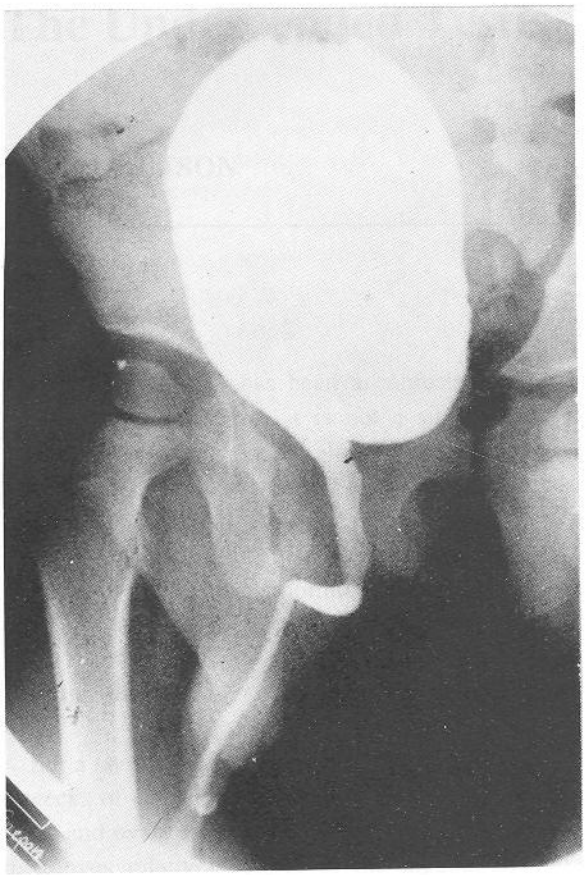

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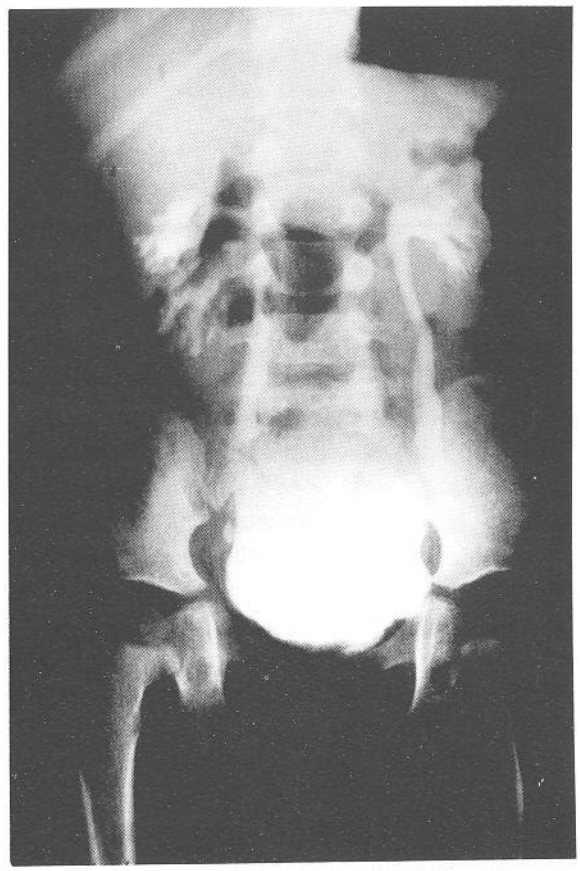

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