FROM THE GUEST EDITOR

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Ectopic ureter is a rather rare condition but frequent urinary leakage or very complicated urinary tract infection empirically bring to mind such a malformation. In Japan we commonly perform ultrasound examination by ourselves in daily clinic, analogous to the stethoscope of a pediatrician. In most patients an ectopic ureter can be suspected easily by ultrasonography. Further examinations such as a voiding cystourethrogram (VCUG), vaginography, and DMSA scan are helpful to develop a management plan. Definitive diagnosis is usually not perplexing in most of the patients. While preschool aged females with an ectopic ureter may be seen frequently by a physician, their parents are apt to be made to feel that urinary leakage is a developmental phenomenon appropriate for such age, even when the wetting is continuous. In this age group, dysfunctional voiding is rarely considered likely, and we should be careful before making a final decision.

Regarding management, many such patients have a dysplastic kidney or segment of kidney, and nephrectomy or heminephrectomy by open surgery or laparoscopic approach will be warranted, especially for single vaginal ectopic ureter. However, many parents are reluctant to agree to have the involved kidney removed. Consequently we often try to preserve the upper kidney in a duplicated system. We prefer an upper tract reconstruction, such as a pyelo-pyelostomy, through a flank approach, rather than en bloc ureteral reimplantation because the ureter from the upper kidney is always entirely anomalous. In bilateral ectopic ureter of single systems, a more rare condition, the management may be more complicated because the bladder is not well developed and staged surgery is an interesting possibility. Simple fenestration between the bladder and ureters is performed as an initial operation, and after the bladder is enlarged significantly, reimplantation is performed as a second step. In some cases, persisted urinary leakage after surgery is another unpleasant problem. Preoperative evaluation of bladder neck and urethra will be important.

In this issue, I chose three topics. Dr. Kakizaki presents a most interesting discussion of the embryology and the anatomy of the ectopic ureter. The subsequent two papers deal with management. Dr. Yoshino discusses the upper tract approach and Dr. Shimada the lower tract approach.

FROM THE EDITOR

Anthony A. Caldamone, M.D.

This month the 7th Annual Asian Pacific Association of Pediatric Urology (APAPU) will be meeting from September 29-30 in Kyoto, Japan. The meeting is being organized by Saburo Tanikaze and his pediatric urology team. The APAPU has been in existence for 7 years and has a membership of about 150 pediatric urologists. Dr. Tanikaze and his team have put together this issue of the Dialogues on the ectopic ureter. It is fascinating for me to learn about the differences in the anatomy, variability, and approaches based on cultural differences in the Japanese experience. The ectopic ureter is an entity with significant variability both in the upper and lower urinary tracts and may represent a significant challenge when the precise anatomy needs to be defined.

This issue covers the complete spectrum of the disorder. It is extensive in its discussion of surgical options as well. The embryological and anatomical discussion by Dr. Kakizaki is written clearly. The sections written by Drs. Yoshino and Shimada are contrasting and provide the reader with a valid rationale for the different surgical approaches.

This issue will serve as an excellent reference source for the ectopic ureter. Dr. Tanikaze and his contributors are to be commended.
Ectopic Ureter - Embryological and Anatomical Considerations

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In the English literature 70 to 80% of all ectopic ureters are associated with duplicated collecting systems, particularly in girls. Single ectopic ureter is far less common, occurring more frequently in male subjects with drainage to the genital tract. In female patients with unilateral single ectopic ureter, the ureter usually opens to the vestibule or urethra. A single ectopic ureter to the vagina is rare.

In contrast, about 70% of ectopic ureters in Japan are associated with a single collecting system and occur more frequently in female subjects with drainage to the vagina. Table 1 shows demographic data of 30 patients with ectopic ureter that were seen in our institute since 1983. The exact reason for this racial difference in the types of ectopic ureter is unknown.

| Table 1 - Characteristics of 30 cases with ectopic ureter in Hokkaido University Hospital |
|---------------------------------|------------------|------------------|------------------|
| Ectopic ureter opening          | No. of cases     | Female:Male      | Right:Left       |
| **Single system: 18 cases**    |                  |                  |                  |
| Gartner’s duct cyst            | 9                | 9:0              | 6:3              |
| Vagina                        | 5                | 5:0              | 1:4              |
| Urethra                       | 4                | 2:2              | 2:2              |
| **Duplex system: 12 units in 11 cases** |          |                  |                  |
| Vagina                        | 4                | 4:0              | 2:2              |
| Urethra                       | 7 (8 units)      | 3:4              | 4:4              |
| **Renal agenesis: 1 case**     |                  |                  |                  |
| Bladder neck*                 | 1                | 1:0              | 1:0              |
*blind-ending ectopic ureter associated with uterus didelphys and ipsilateral vaginal obstruction

Normal embryology

Early in the fourth week of gestation, the distal segment of the mesonephric duct (Wolfian duct) gives rise to the ureteral bud, which grows dorsally toward the metanephric blastema, the kidney progenitor. The ureteral bud gives rise to the urinary collecting system (intrarenal collecting ducts, calyces, pelvis and ureter) and also acts as an inducer of differentiation of the metanephric blastema to form nephron units. Between 6 and 8 weeks of gestation, the segment of the mesonephric duct distal to the origin of the ureteral bud (the common excretory duct) is incorporated into the anterior part of the cloaca, providing separate entry of mesonephric duct and ureter into the cloaca. With local growth, the ureteral openings migrate in a craniolateral direction to reach their respective destinations in the developing bladder trigone. Simultaneously, the mesonephric ducts descend with the urogenital sinus.

During week 6, the paramesonephric ducts (Mullerian ducts) arise lateral to the mesonephric ducts. The mesonephric ducts and paramesonephric ducts are intimately related anatomically and developmentally. Growth of the paramesonephric ducts proceeds in a caudomedial direction and eventually they cross the mesonephric ducts ventrally and fuse into a single midline channel that proceeds to empty in the urogenital sinus between the openings of the 2 mesonephric ducts. The fused segments of the paramesonephric ducts become the uterus and vagina, whereas the ununited segments form the fallopian tubes. In the male the mesonephric duct structure eventually forms the epididymis, vas deferens, seminal vesicles and ejaculatory ducts, while in the female the mesonephric duct is absorbed, resulting in the vestigial remnants along the fallopian tubes and proximal uterus and within the cervix and in the anterolateral wall of the vagina. The remnants of the mesonephric ducts within the cervix and vaginal wall are referred to as Gartner’s duct.

Based on these embryological steps, it is quite apparent that the mesonephric duct has a critical role in the formation of the ureter, kidney and paramesonephric derivatives (uterus and vagina).

Embryological abnormalities in ectopic ureter

If the ureteral bud arises more proximally (cranially) than normal on the mesonephric duct, the ureteral orifice has less time in the bladder to undergo its normal craniolateral migration and results in a ureteral orifice more medial and caudal than is usual. An even further proximal position of the ureteral bud on the mesonephric duct may result in the ureteral orifice remaining on the mesonephric duct, causing the ureteral orifice to terminate outside the bladder and urethra. In the female, an ectopic ureter draining into the Gartner’s duct can rupture into the adjoining fallopian tube, uterus, upper vagina, or vestibule. An ectopic ureter opening into a Gartner’s duct cyst is an uncommon variant of female ureteral ectopia.

Ectopic ureter is associated with varying degrees of renal anomalies. According to the “bud theory” proposed by Mackie and Stephens, the greater the distance of the origin of the ureteral bud from the normal take off section of the mesonephric duct, the more abnormal the renal segment, although they only studied duplex kidneys. Renal hypoplasia associated with ectopic ureter is the product of inadequate interaction between the ureteral bud and metanephric blastema.

Anatomical aspects

In the female, ureteral ectopia most commonly occurs in the urethra or vestibule, and less frequently in the vagina. In a female patient with an ectopic ureteral orifice distal to the bladder neck, urinary incontinence (ureteral incontinence) is the most prevalent symptom. However, ureteral ectopia opening into the vestibule without urinary incontinence has been reported in rare cases. Urinary continence in these cases is probably due to the incorporation of the ectopic ureteral end in the wall of the urethral sphincter. In the male, the posterior urethra is the most common site of termination of an ectopic ureter. Drainage into the mesonephric structures involves the seminal vesicle three times more often than the ejaculatory duct and vas deferens combined. In single ectopic ureter, the trigone on the affected side fails to develop. In a case with bilateral single ectopic ureter there is no stimulus for the development of the bladder trigone, bladder neck and a large part of the urethra. Thus, in bilateral single ectopic ureter, the structure vital for urinary continence is significantly defective, causing severe urinary incontinence.
Single vaginal ectopic ureter with or without a Gartner’s duct cyst is sometimes associated with abnormalities of the female reproductive tract. An abnormality of the mesonephric duct can explain the fusion anomaly of the paramesonephric ducts (bifid uterus or uterus didelphys), owing to the known influence of the mesonephric duct on the early stages of development of the paramesonephric duct as stated above. Approximately 200 cases have been reported with unilateral obstruction of duplicated paramesonephric ducts associated with ipsilateral renal dysgenesis. The combination of unilateral renal dysgenesis, ipsilateral Gartner’s duct cyst and ipsilateral paramesonephric duct obstruction is also reported, though extremely rare. Gartner’s duct cyst often appears posterior to the bladder. However, if it protrudes into the bladder, it may mimic a ureterocele. Similar conditions also may occur in a dilated ectopic ureter. Clinically, the formation of an acute angle of the wall of ureterocele with the wall of the bladder is an important feature in differentiating a ureterocele from a Gartner’s duct cyst or dilated ectopic ureter located posterior to the bladder. When cystic dilatation within the pelvis without ureteral dilatation is found in girls with ipsilateral renal dysgenesis, the possibility of a Gartner’s duct cyst should be considered. A vaginal mass associated with ipsilateral renal absence on imaging study is characteristic of the malformation complex featured by uterus didelphys and a septate vagina with unilateral vaginal obstruction. It may present with hydrocolpos at birth or with hematocolpos at puberty.

Renal anomalies

Ectopic ureter is associated with varying degrees of renal anomalies. According to the “bud theory” by Mackie and Stephens, the more remote the ureteral opening, the greater the degree of renal maldevelopment. However, in our experience renal dysplasia of vaginal ectopia is not as severe as expected from its ectopic orifice position or as compared to the male counterpart with a similar degree of genital ectopia. Because vaginal ectopic ureter can be explained embryologically by rupture of Gartner’s duct into the vaginal lumen, it may be that the true ureteral orifice is not as ectopic in some cases of vaginal ectopic ureter.

In contrast, single ectopic ureter opening into the Gartner’s duct cyst are associated with more dysplastic kidney than vaginal ectopic ureter without cyst formation. The corresponding kidney is sometimes absent in cases with single ectopic ureter opening into the Gartner’s duct cyst. This anomaly may be analogous to ectopic ureter opening into the seminal vesicle in the male counterpart. It is speculated that this anomaly results from a maldevelopment of the mesonephric duct in early fetal life.

From a clinical point of view, single ectopic ureter opening into the vagina or Gartner’s duct cyst is associated with a poorly functioning kidney that most often is not worth preserving, while the corresponding kidney is worth preserving in about 50% of cases with single ectopic ureter opening into the urethra. Regarding ectopic ureter associated with duplex system, about 25% and 30% of corresponding upper kidney segment has some function that is worth preserving in vaginal and urethral ectopic ureter, respectively.

References


Flank Approach in the Management of Ectopic Ureter

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The ectopic ureter drains into the bladder neck, urethra, wolffian or müllerian structures. It is either obstructive or refluxing and urological intervention is usually necessary. The surgical procedure is selected according to the renal function of the affected kidney and the associated abnormalities that need repair at the bladder level. The site of the opening of the ectopic ureter does not affect the choice of procedure directly. The surgical management of the ectopic ureter branches with two main approaches. One is the flank approach and the other is the lower tract approach. The feature of the flank approach is described.

Single system

In the single system, the selection of the approach depends on the renal function. When the affected kidney shows minimal or no function on the isotope renography, it is almost always dysplastic and nephrectomy with subtotal ureterectomy via the flank incision is justified. The kidney with ureter draining into the genital system is included in this category. The renal function with ectopic ureter into the urinary system varies and the kidney that has some function should be preserved with a lower approach.

Duplex system

In the duplex system, the flank approach is chosen when there is no major abnormality of the lower urinary tract. The advantage of this approach is that the decision whether to save or discard the involved kidney can be made with the direct visualization during the surgery. Compared to the single system, poor function of the affected moiety on the nuclear renography does not always mean dysplasia of the respective system. Hydroureteronephrosis is often a sign of preserved renal function as it indicates urine production to some extent and function may be recoverable after resolving the obstruction. Heminephrectomy is recommended when the moiety is clearly dysplastic. The removal of the odd looking parenchyma with many cysts relieves the future anxiety of malignancy and hypertension. Ureteropelviostomy is the option when the upper pole moiety shows the normal appearing parenchyma without signs of cystic dysplasia. Although the upper half of the duplex normal kidney contributes a maximum of 20% to the overall renal function, the postoperative function of the affected moiety is usually around 10% of the total function.
Renal function and (hemi)nephrectomy

When the function of the affected kidney is marginal, how much function is enough to justify the removal of the kidney? Dialysis is initiated when renal function falls between 10 and 15% in children. The commonly used cut-off comes from this idea. With dialysis, it becomes difficult to maintain effective exchange in the total absence of renal function. Survival outcome is significantly related to the residual renal function.\(^1\) Even function as low as 5% is meaningful in that situation. Although there is no absolute cut-off, it is important, therefore, to leave the salvageable kidney if it represents little risk, and to maintain function as much as possible considering the future long life in children. There is another issue to be considered in terms of function. When the affected kidney has no consistent function, the (hemi) nephrectomy cannot have any influence on the remaining kidney. However, when the affected kidney has some function, the sudden nephron reduction after nephrectomy might cause damage to the remaining parenchyma, through renal hypertrophy or hyperfiltration phenomenon. There is also a possibility of unknown mechanisms such as immunopathological reactivity.\(^2\)

Ureteral stump after incomplete ureterectomy

Complete ureterectomy of the ectopic ureter carries a risk of injury to the adjacent organs such as the ipsilateral healthy ureter and vagina, or to jeopardize the spincteric function around the bladder neck and proximal urethra. In either nephrectomy or heminephrectomy, the ureter is incompletely excised in order to avoid such risks. The distal ureteral stump after partial ureterectomy, however, can cause symptoms such as hematuria, bacteriuria, pyuria, and lower abdominal pain and the long refluxing stump and voiding dysfunction seem to be the risk factors. However, incomplete ureterectomy as distal as possible from the flank incision is sufficient because the morbidity of a ureteral stump is less than 10%, whether it is refluxing or obstructed.\(^3,4\) Cases with ectopic ureter have less of a chance of re-operation.\(^5\) The effective management of the symptomatic stump is excision although the less invasive endoscopic technique might be an alternative.

Laparoscopic surgery

Laparoscopic surgery has become popular and nephrectomy is performed in children as easily as in adults. Laparoscopic heminephrectomy in children is also done in experienced hands. In the single system, the position of the kidney with ectopic ureter varies. The location of the kidney needs to be precisely identified before surgery to determine the skin incision level in open surgery. On the other hand, the location of the kidney does not matter in the laparoscopic approach as the whole abdomen can be manageable. Another advantage of the laparoscopic surgery is the removal of the distal ureter. In laparoscopic surgery, the ureter can be removed safely under the direct vision deep into the pelvic cavity.\(^6\) The distal ureteral stump can be much shorter than that through a single flank incision. Still the risk may overweigh the benefits of the complete ureterectomy. In the duplex system, when upper pole salvage is desired, meticulous technique similar to the dismembered pyeloplasty is needed in performing a ureteropyelostomy. The current laparoscopic approach does not seem to reach the level of the open anastomosis as yet. Considering the possible conversion from heminephrectomy to ureteropyelostomy, open surgery is recommended in the duplex system. Robotic surgery has been shown to be beneficial as well.

Our series

There have been 48 cases of ectopic ureter in Kobe Children’s Hospital (1979 – September 2003) and Aichi Children’s Center (October 2003 - 2005). Forty two were female and 6 male. Age at surgery ranged from 2 weeks to 10 years. Younger children presented with urinary tract infection and older girls presented with incontinence. Five cases had the prenatal diagnosis of hydrenephrosis. Twenty six females had single and 16 had duplex systems. Four males had single and 2 had duplex systems. In total, 30 (62.5%) were single systems, similar to other Japanese reports. Nephrectomy through a flank approach was selected in 20 cases. The ureteral orifice was located in the urethra in 2 males and in the vagina in 13 female. The precise opening was unknown in 5. The majority showed no uptake on DMSA scan. Small dysplastic kidneys were found at surgery. Laparoscopic nephrectomy was done in the most recent three cases. Eighteen cases were associated with duplex system. All showed upper pole hydronephrovortex. Flank approach was selected in 14 cases. Heminephrectomy was done in 8 and ureteropyelostomy was done in 6. The upper pole showed little uptake on the DMSA scan in all heminephrectomy cases. Upper pole function was significant preoperatively in 3, and minimal in 3 in the ureteropyelostomy group. The postoperative course was uneventful in all cases but one. Temporary urinary leakage was seen in one case with ureteropyelostomy. The leakage stopped on the 7th day following surgery and no further complication was seen. After ureteropyelostomy, all hydrenephrosis improved markedly and when the upper pole function did improve, it never represented more than 14% of the overall kidney function. Cases that showed hydrourerter before surgery demonstrated the hydrourerter to some extent behind the bladder after the surgery. It gradually decreased in size or became undetectable on follow-up ultrasonography. Only one girl demonstrated one episode of pyuria for which the ureteral stump may have been responsible. Although no additional surgery was done in any of the cases.

Surgical procedure

A subcostal oblique incision is made beneath the 12th rib. Muscles are divided and the retroperitoneal space is exposed. After opening Gerota’s fascia, the upper pole is exposed. The final decision is made by the macroscopic finding of the upper pole. When heminephrectomy is chosen, the further dissection is made around the upper half of the kidney not only to expose the whole upper moiety but also to prepare for control of the unexpected intraoperative bleeding. The vessels to the upper moiety are divided during the dissection. The upper pole ureter is identified and divided. Isolation of the upper pole ureter gives better exposure at the junction between the upper and lower poles and minimizes the potential of leaving part of the upper collecting system. Care should be taken to protect the lower healthy ureter. The proximal ureter is freed and the upper pole parenchyma is excised. The demarcation of the upper and lower poles is clear when the upper moiety is apparently dysplastic (Figure A). Hemostasis of the renal edges is achieved by running sutures. The cut edges of renal parenchyma are approximated if possible. The distal ureter is subsequently mobilized as distal as possible and divided and closed with a transfixing suture after aspiration of urine in the distal dilated ureter. It is possible to remove the ureter down to the pelvic brim through the flank incision in younger children.

Once the decision to preserve the upper pole is made, there is no need for further aggressive dissection. The upper pole ureter is isolated distally and the upward dissection is made to the level of the lower pole pelvis right beneath the lower pole hilum. The upper pole ureter is cut (Figure B) and anastomosed to the adjacent lower pole renal pelvis or ureter using the 7-0 absorbable running sutures (Figure C,D). Discrepancy in size between the ectopic and recipient ureter are common but it
usually is not bothersome. A drain is placed after ureteropyelostomy but not after heminephrectomy. The operative time was not different in our series between the heminephrectomy and ureteropyelostomy group. There was a significant difference in blood loss between groups. The mean blood loss was 71g (15g to 153g) for heminephrectomy and 15g (4g to 30g) for the ureteropyelostomy. Unexpected bleeding was encountered during heminephrectomy only when demarcation of the moi¬eties was not clearly identifiable. Besides, even after the complete division of the upper pole vascular pedicles, there remains a vascular network between the upper and lower pole parenchyma and blood loss can be experienced at times. It seems that ureteropyelostomy is an easier, safer and more reliable procedure in cases with healthy upper pole parenchyma macroscopically.

**Figure A:** Arrows indicate the demarcation between the upper (up) and lower poles

**Figure B:** Arrows show the hilium lower pole with the (a:) proximal upper ureter, (b:) distal upper ureter, (c:) lower pole pelvis.

**Figure C:** Posterior anastomosis of ureteropyelostomy is completed

**Figure D:** Completed anastomosis

References

Ureteral Reimplantation in the Management of Ectopic Ureter

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The term “ectopic ureter” has generally been used to apply to a ureter whose orifice opens at the bladder neck or more distally into the mesonephric duct structures. An ectopic ureter is more common in females, and is often seen in association with complete ureteral duplication. Racial difference is seen in that between 70% to 80% of ectopic ureters are associated with duplicated system in English literature, while a single system ectopia accounts for about 70% to 80% of total in Japanese literature and most of them drain into the vagina. Surgical management of ectopic ureter depends on several factors including the function of affected kidney or renal moiety, duplicated or single system, position of the ectopic orifice, size of the ureter, association of vesicoureteral reflux or obstruction, and probably the age and gender of the patient. Because most of the ectopic ureters drain poorly functioning renal segments, upper pole heminephrectomy or total nephrectomy in a single system is often justified. If the function of the affected kidney or upper renal moiety is worth preserving, ureteral reimplantation is one of the recommended interventions. In addition, the scar after ureteral reimplantation is less conspicuous than that of upper tract open surgery. Herein I describe the anatomy of the lower portion of the ectopic ureter, and discuss the surgical procedure of ureteral reimplantation. Rare cases of this entity are also presented.

It is generally true that the more distal or remote from the normal position the ureter opens, the more abnormal or dysplastic is the affected kidney, although vaginal ectopic ureters sometimes drain small but sufficiently functioning kidneys which may justify renal preservation. However, there is no general rule to define what extent of renal function is worth salvaging. The usual feature of the excretory urogram is a nonvisualizing or poorly visualizing upper pole in a duplex system that is massively hydronephrotic. Isotope renal scanning using 99mTc-DMSA has proved to be the most appropriate method to evaluate renal function, although differentiation between the upper pole and the lower pole may be difficult. In single system ectopic ureters, split renal function on renal scintigraphy can help to choose the surgical option. In duplicated system, I usually select renal preserving surgery if ultrasonography and/or MRI shows hydronephrotic images of the upper moiety.

In single system ectopic ureters that open at the bladder neck, the hiatus in the bladder is also ectopic being more caudal than normal. Urethral ectopic ureters run a short submucosal course in the bladder base to reach the urethra. The intravesical part of the ureter is short and the hiatus lies close to the bladder neck. The meatus may be normal in size, stenotic or grossly patent. In vestibular ectopic ureter, the orifice lies to one side of the external urethral meatus or at the posterolateral lip on the urethrovaginal bridge. The ureter runs from its hiatus at the bladder neck in a submucosal course along the urethra. In the vaginal ectopic ureter, the orifice may open at the base of the hymen or in the lateral wall of the vagina or cervix. In the latter instance, the ureter bypasses the bladder and enters the vagina. In the former case, in which the ureter opens in the vaginal portion of urogenital sinus origin, it may run the same course as the vestibular ectopic ureter. Patients with this type of vaginal ectopia may not present with urinary incontinence because the ureter passes through the urethral sphincter mechanism. If the ectopic ureter joins the mesonephric duct remnant (Gartner’s duct), the urine is stored in the duct. A soft cystic mass composed in part of ureter and Gartner’s duct may project into the vaginal lumen. It may sometimes protrude from the vaginal orifice.

Management of the terminal ureteral stump is controversial. The terminal stump, if left long enough, may result in postmicturition urinary dribbling, and predispose to urinary infections and pyohydroureter. In cases of vesical neck ectopia, intravesical dissection and mobilization of the ureter, although difficult in some cases, may reserve enough ureteral length for successful reimplantation. The new hiatus is created cranially and laterally by dividing the detrusor. When the ectopic orifice is located distal to the vesical neck, an extravesical approach is utilized to initially identify the ureter outside the bladder.

The ureter should be mobilized as inferiorly as possible without sacrificing the inferior vesical pedicle where it is ligated and cut. Some authors advocate intramural dissection of the terminal stump. This method, total excision of the ureteral stump, includes opening of the terminal ureter into the ectopic hiatus by cutting the overlying detrusor, suturing the most distal end of the ureter from within the ureteral lumen, and re-approximating the hiatus and the detrusor muscles. The ureter is usually transected just proximal to its entrance in the hiatus. A psoas hitch may be necessary to reduce tension for reimplantation. I prefer a mini-hitch to the umbilical ligament. This conservative approach leaving a short ureteral stump behind rarely causes postoperative complications. In males, if the ureter joins the vas deferens or seminal vesicle, it takes a direct course bypassing the bladder. The stump of an ectopic ureter does not require routine excision. Ligation of the vas deferens can be postponed for several months in children who have had episodes of epididymitis to see whether it recurs after nephrectomy.

In cases of duplicated system, the extravesical course of the two ureters usually demonstrate a consistent relationship in the vicinity of the vesical hiatus with the upper pole ureter posterior in most of the cases. The intravesical segment of both ureters are closely bound in a common sheath, and cannot be separated without causing potential injury or ischemia of one or both ureters. If the upper pole ureter opens at the bladder neck or in the urethra, the two ureters run separately in their intravesical course and can be divided without causing ischemia to the lower pole ureter. During surgery one must remember that, despite the muscle layers being normally constituted in the extravesical ureters, the intravesical segment of the ectopic ureters in duplicated systems is only partially muscularized. Based on the histological features of the ectopic ureters, it is recommended to remove the ureteral stump including its submucosal segment. If upper pole has significant function, one of the options is to reimplant both ureters into the bladder. When reflux or obstruction is demonstrated in one or both ureters, a common sheath reimplantation is the preferable method of treatment. The lower pole ureter is dissected from within the bladder. The upper pole ureter, which is usually dilated, is transected just outside the bladder to avoid separation of the common sheath. The risk of injury to the mate ureter may outweigh the benefit of complete ureterectomy. In many instances tapering of the upper ureter is necessary to reduce its diameter. When excisional tapering is selected, tailoring is made along the ureteral length opposite the common wall. This procedure, however, can be technically challenging especially in small children.
Single ectopic ureter - bilateral or solitary kidney

Bilateral single ectopic ureter and a single ectopic ureter in a solitary kidney are rare anomalies. The bladder neck develops in the position of the urogenital sinus between the orifices of the mesonephric duct and the ureter. If ureter migration ceases in the position of mesonephric duct orifices, there is no formation of the trigone and bladder base. The bladder neck becomes very wide, poorly defined, and incompetent. In the male with this anomaly, the bladder grows well because the intact urethral sphincter mechanism provides a variable degree of urinary control. On the contrary, in rare instances in the female with this pathology draining into the distal urethra below the sphincter or into the vestibule, the bladder had no chance to store and distend with urine resulting in bladder hypoplasia. Renal dysmorphism is commonly observed as well as genital and anorectal anomalies.6

Ureteral reimplantation alone does not solve the urological problems due to insufficient development of trigone, bladder neck, and resultant bladder capacity.7 Ureteral reimplantation with a concomitant bladder neck reconstruction or sling procedure has been attempted expecting gradual increase in bladder capacity. However, the results have been poor, and eventually in most cases urinary diversion or augmentation cystoplasty to create an adequate reservoir was required.8 Drainage of the reservoir is usually accomplished with CIC.

We performed a staged procedure in a 5-month-old girl with single system ectopic ureter in a solitary kidney and hypoplastic bladder of about 5 ml in capacity.9 In the first procedure, side-to-side anastomosis was done between the dilated ectopic ureter and the lateral wall of the small bladder. The fenestration was small, about 5 Fr in caliber. Nine months later the bladder capacity increased to about 70 ml. Ureterocystoanastomosis was performed 11 months after the first procedure. She is now 17 years old, and is dry day and night without episodes of urinary infection.

Pseudoureterocele-type ectopic ureter

Pseudoureterocele-type ectopic ureter is complicated anatomically, and is easily misdiagnosed and incorrectly treated as an ectopic ureterocele. The embryology of this entity is not fully elucidated but the pathology suggests a single ectopic ureter draining into a mesonephric duct or Gartner’s cyst.10 The cyst may extend caudally behind the bladder and the urethra, and may rupture into the bladder or urethra, injuring the continence mechanism of the bladder outlet (Figure E). It may also rupture into the vagina resulting in a vesicovaginal fistula. Endoscopy reveals a wide-open bladder neck and a very thin bladder wall in a unilateral trigone on which side the dysplastic kidney drains. Endoscopic periurethral injection of bulking agents is ineffective because the material does not accumulate around the bladder neck to cause coaptation. Reconstruction of the bladder neck by Young-Dees-League or Kropp procedure seems difficult because the detrusor backing around the posterior bladder neck and the trigone is totally lacking. The Pippi-Salle procedure may be the better option after re-approximation and reinforcement of the trigone.11 It may be safer to perform a simultaneous augmentation cystoplasty and to create a Mitrofanoff continent stoma.

References

Figure E: Pseudoureterocele-type ectopic ureter

![Diagram](Image)
Ectopic Ureter

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