9. Renal duplication

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The complex embryogenesis of the kidney and drainage system accounts for the development and frequency of congenital abnormalities. A complete or incomplete double pyelocaliceal system is identified in 0.8% of postmortem examinations. As an incidental finding during intravenous urography, this percentage increases to 3%, of which in 40% of cases, the finding is bilateral.

The term “renal duplication” signifies the co-existence of two ureters separately draining the corresponding moieties of the kidney into the urinary bladder, which can present as an ectopic ureter or as an ureterocele. Duplication can be complete or incomplete. The function of both the upper and lower renal poles can be partially or totally affected, with the upper pole being the most commonly involved.

Embryologic development can best explain such abnormalities. The primordial ureter anlage of the drainage system emerges as a bud from the caudal part of the mesonephric duct and gradually ascends in a dorso-cranial direction.

A single ureteric bud that undergoes bifurcation will induce a variable degree of incomplete duplication of the upper renal segment. Complete duplication is the result of two ureteric buds, leading to the drainage of the kidney by two ureters.

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Typically, the second ureteric bud which corresponds to the upper pole ureter is longer and results in lower insertion within the bladder or is ectopic; the two ureters will naturally cross. Ureteral ectopia is commonly accompanied by varying degrees of dysplasia of the renal parenchyma.

The most frequent anomalies associated with renal duplication include ureterocele, incomplete or complete ureteral duplication and ureteral ectopia.

**Ureterocele**

Ureterocele is defined as a cystic dilatation of the terminal intravesicle ureter that is associated with the upper pole or is situated beyond the bladder neck. With an overall prevalence of 1 in 4000 births, it is more common among girls than boys.

Dysplasia of the renal parenchyma is usual and is often accompanied by impaired renal function.

Classification of ureteroceles is based on its location. The most common type is the ectopic ureterocele which is situated outside the bladder, as opposed to the intravesical or orthotopic ureterocele that is contained within the bladder. In 50% of ectopic ureteroceles, the lower pole ureter is raised causing vesicoureteric reflux, while an obstruction can lead to megaureter.

An association of contralateral renal duplication is recorded in 50% of cases, in some of which the ureterocele can induce reflux or obstruction in the upper moiety of the opposite kidney. Obstructive megaureter can prevent drainage of the bladder, causing contralateral renal impairment.

An ectopic ureterocele can cause secondary weakening of the muscular wall at the trigone and bladder neck resulting in disturbed urine continence. Albeit a rare presentation, a prolapsing ureterocele into the urethra has been recorded in girls.

Observed in females, orthotopic ureterocele is located strictly intravesically and is generally associated with a single kidney system. The ureter can be dilated and renal function slightly impaired.

Caecoureterocele is a rare entity associated with an ectopic ureter located in the urethra below the bladder neck. The corresponding upper pole of a double renal collecting system is non-functional.

**Clinical presentation**

Urinary stasis presents with heaviness and pain in the loin, frequently with urine infection and occasionally haematuria. The ureterocele can also cause lithiasis. Acute or chronic urine retention is the result of obstruction. Urethral
prolapse of the ureterocele through the external urinary meatus is sometimes observed in girls, while male infants can display urine retention mimicking posterior urethral valves.

The problem may be detected earlier on examination for pyelonephritis. Dysuria and cystitis are possible later symptoms.

**Diagnosis**

A distended ureter and ureterocele can be easily detected on an antenatal sonogram. Postnatal sonography will reveal the dilatation of the ureter extending to the upper pole and the ureterocele. Other lower pole problems may also be identified, including vesicoureteric reflux and pelviureteric junction obstruction.

DMSA scintigraphy is particularly effective in assessing renal and upper pole function; it may occasionally also prove helpful in detecting cryptic duplication.

Micturating cystourethrography (MCU) will disclose ipsilateral and bilateral vesicoureteral reflux in patients with duplex system ureterocele and assess the degree of an intraurethral prolapsed ureterocele.

Intravenous urography can disclose incomplete unilateral duplication and cryptic duplication in the contra lateral upper tract.

Magnetic resonance (MR) urography is the ideal imaging modality for displaying the anatomy of the upper and lower moiety in both kidneys but involves sedation of the child or anesthesia. Among this group of evolving techniques, is the development of fast three-dimensional scanners. Combined with MR urography, this method constitutes a major advance in the investigation of the duplex system.

Performing cystoscopy under anesthesia is helpful in locating the ureteral orifice; it can also facilitate differential diagnosis between ureterocele and ectopic megaureter.

**Treatment**

The timing, method and type of intervention is reliant on several factors, such as early diagnosis, renal function, location and presence of reflux.

Prophylactic antibiotic treatment is generally indicated if a child with a ureterocele is clinically asymptomatic and displays non- or hypo- renal function of the upper pole without evidence of significant lower pole or bladder outlet obstruction. A severe obstruction and urinary tract infection warrant an immediate endoscopic incision or puncture. In 25% of patients, decompression of the ureterocele is achieved without reflux. Should this fail,
an open operation may prove necessary. Secondary surgery is indicated in the
presence of significant vesicoureteric reflux or obstruction of the ipsilateral or
contralateral ureters and/or bladder neck. The type of surgery varies from
partial nephrectomy to complete unilateral reconstruction.

An endoscopic ureterocele incision is preferred in cases of poor upper pole
function and an ectopic ureter posterior to the bladder draining distally into the
vagina. It is also indicated if a prolapsed ureterocele prevents emptying of the
bladder. Given the risk of iatrogenic vesicoureteric reflux, the incision should
be made as close to the bladder wall as possible.

A virtually total loss of upper pole renal function is treated by
heminephrectomy and ureterocelic aspiration via the ureteral stump.

Pelvic anastomosis of the upper and lower poles, otherwise known as
pyelopyelostomy, can be performed providing renal function is satisfactory.

Ureterocele incision and reimplantation of the conjoined ureters is
indicated, preferably during the first year of life, if renal function is good and
dilatation of the lower pole ureter small. If the upper pole is non-functional,
upper pole heminephrectomy can be performed in conjunction with
ureterectomy and ureterocele incision, followed by reimplantation of the
ipsilateral lower pole ureter. This procedure represents major surgery and cases
of caecoureterocele present the risk of damage to the bladder neck and striated
sphincter mechanism resulting in a vesicovaginal fistula.

Double ureter

Incomplete ureteral duplication is ascribed to premature branching of a
single ureteric bud during antenatal development. The point at which these two
ureters join is dependent on when bifurcation took place; the most common
point is in the lower third of the ureter.

In complete ureteral duplication, the ureter draining the upper moiety is
predisposed to obstructive pathology while the ureter draining the lower
moiety has an inclination for vesicoureteric reflux.

Eighty per cent of ectopic ureters are associated with a duplicated
collecting system. At a male to female ratio 1:5, it is more prevalent among
females. The embryonic ectopic ureter is attributed to a disturbance in the
caudal section of the mesonephric duct. Hence, the ectopic ureteral orifice
should be sought in components deriving from the duct. In 60% of males, it is
found in the posterior urethra above the verumontanum and in 40% it is
located in the seminal tract - the epididymis, vas deferens and seminal vesicles.
In 35% of females, it is situated in the urethra, in 30% in the vaginal vestibule
and in 25% the lower section of the vagina. Rarely, it can be located in the
uterus or the fallopian tube.
**Clinical picture**

Symptoms for incomplete ureteral duplication are non-specific and include abdominal pain and urinary tract infection, while those for complete duplication will depend on the presence of obstruction or vesicoureteric reflux. Ureteral ectopia is associated with renal dysplasia and ureteral dilation, resulting from reflux, obstruction or dysmorphism.

In females, symptomatology resembles that for urinary tract infection: stenosed ureteral orifice with purulent vaginal discharge. Infrasphincteric ureteral ectopia in girls at nursery or school age causes urinary incontinence which can be nocturnal or daytime or both, albeit urine control is normal, while in boys it is not associated with incontinence since the ureter terminates within the sphincter mechanism. However, it is frequently accompanied by urinary tract infection and recurrent epididymo-orchitis.

**Investigations**

Imaging tests are the same as those previously mentioned for ureteroceles, i.e. sonography, nuclear studies, MCU, intravenous or MR urography and cystoscopy. The majority of ectopic megaureters are diagnosed early on ultrasonography. The “drooping lily” sign identified on urography represents inferolateral displacement of lower pole moiety and displacement of upper calyces of the lower pole collecting system. All these modalities constitute useful diagnostic tools for function assessment, reflux detection, and exclusion of ipsilateral compression of the lower pole and urethral obstruction. To investigate female incontinence, filling the bladder with methylene blue may be enlightening, although MRI has tended to replace this method.

Loss of uncoloured urine indicates ureteral ectopia.

**Management**

The timing and type of intervention is dependent on renal function of both moieties, the presence of ureteral dilatation, vesicoureteric reflux and the type of anatomic abnormality. Consequently, the decision of a heminephrectomy, anti-reflux surgery and ureteral reimplantation, pyelopyelostomy or nephrectomy will be made following assessment and evaluation of the above.

Incomplete duplication is conservatively managed initially. If surgery is necessary due to infection, anastomosis of the upper ureter with the lower pole pelvis is recommended. The co-existence of lower moiety pelvic-ureteric junction stenosis in the lower pole requires reconstruction surgery or heminephrectomy if the lower moiety is non-functional. Complete ureteral
duplication surgery depends on the renal function of the moiety corresponding to the abnormal ureter. Upper pole dysplasia is common and such cases are treated by heminephro-ureterectomy or double ureteral reimplantation without affecting the individual ureteral orifices. If upper pole function warrants preservation, urethral reconstruction is also a consideration. The surgical indications for suprasphincteric ectopic ureter are also relative. Heminephrectomy is appropriate if upper pole function is negligible. Vesicoureteric reflux can be managed by ureterectomy. If pole function is adequate, ureteral en-bloc reimplantation is recommended. Infrasphincteric ureteral ectopia is invariably managed by excision of the affected upper renal pole. Again, if upper pole function is good, reimplantation or pyelopyelostomy is a consideration. The possible presence of bilateral renal duplication should always be investigated.

References


