Prevalence of Congenital Duplication of Collecting System in Pakistan

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The urogenital system is more likely to have birth defects than any other system. Of all the urinary tract anomalies, Double ureter is the most common one. It may be incomplete (bifid ureter) or complete duplication. In order to collect data for this various medical colleges were approached & 120 cadaveric kidney pairs were examined for the presence or absence of double ureters. It was found that 02 out of all examined kidney pairs were showing this anomaly. One pair was having bilateral double ureters while other was showing right sided ureteric duplication.

Key words: Congenital Anomaly, Double ureter, Collecting System

Ureters are paired muscular tubes lying retroperitoneally, carry urine through peristaltic contractions from renal pelvis to urinary bladder. It is 25-30cm long & 3mm in diameter showing three areas of constriction & having a valve at its bottom end. At renal pelvis ureter is posterior most to renal vein & artery. Abdominal part of it is related to psoas major behind & gonadal, colic, ileocolic vessels, duodenum & sigmoid mesocolon in front. Pelvic part of ureter is related anterosuperiorly with ductus deferens in males & it lies behind the ovary in females then it obliquely enters the urinary bladder.

Embryologically ureter develops between 4th to 6th weeks of gestation by apperance of ureteral bud of mesonephric duct. Cranial part of bud joins metanephric blastema & induces the formation of renal parenchyma, major & minor calyces. Mesonephric duct caudally is incorporated into cloaca, which forms bladder trigone. When there is early splitting of ureteric bud splitting may be partial or complete resulting in partial or complete ureteric duplication. If two ureteric buds arise complete double ureters are formed. The upper pole ureter with long submucosal tunnel opens usually outside the bladder i.e medially & inferiorly where as lower pole ureter with short submucosal tunnel opens into the bladder i.e above & lateral to upper pole ureter resulting in vesicoureteral reflux afterwards. So in case of complete ureteral duplication the upper ureter is more likely to be associated with ectopic insertion, stenosis, ureteroceles, &/or obstruction & infection. Whereas the lower ureter which has a normal insertion is frequently associated with vesicoureteral reflux (VUR) due to incompetent valve mechanism.

A person with partial duplication may remain asymptomatic but with complete duplication of ureters a patient may come with recurrent urinary tract infection, ureteric stones or incontinence of urine especially in females.

This anomaly may be discovered in a patient of hypertension, proteinuria or renal insufficiency during evaluation.

Radiologically double ureter can be seen on USG (for ureteroceles), IVU (drooping liy sign), excretory urogram, voiding cystourethrogram (for VUR) antegrade pyelography (for 2nd ureter), cystoscopy (for ectopic orifice), CT scan, MRI.

Ureteral duplication alone requires no specific intervention. If it is associated with vesicoureteral reflux, obstruction or ureteroceles, an appropriate medical therapy & surgical correction is required which may includes ureteral reimplantation with ureteropelvicyostomy (joining Upper pole ureter with lower pole ureter) or ureteroureterostomy (joining upper pole ureter with lower pole ureter). Treatment of vesicoureteral reflux with dextranomer hyaluronic acid copolymer is an alternative to open surgery.

Materials and methods:
A study on cadavers was conducted from January 2001 to June 2004 to investigate the presence of double ureter or normal single ureter with each kidney pair. The study was designed after accidental finding of double ureter in right kidney during routine dissection in Anatomy department of FMH College of Medicine & Dentistry, LHR. In order to collect relevant data Anatomy Departments of various other medical colleges of Lahore like KEMC & FJMC were approached. All kidney pairs were observed for the presence of double ureters. The kidney pairs that were found to have normal single ureters were listed in-group A, while kidney pairs having double ureters were listed in-group B.

Results:
Group A: out of 120 pairs of dissected cadaveric specimens of kidneys 118 were showing bilateral single ureters lying posterior most in renal pelvis.
GROUP B: 02 kidney pairs out of 120 pairs of dissected cadaveric specimens of kidneys were having double ureters. One pair was showing bilateral double ureters & one pair was showing unilateral Right sided double ureters.
Table showing percentage of anatomical variation

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Anatomical Variations in Kidney pairs

Discussion:
Double ureter is the most common anomaly of urogenital system. 10-15% of population shows this anomaly. It is much more common in females than in males. 8% of children being evaluated for UTI were having ureteric duplication. The incidence of incomplete ureteral duplication is 1 in 25 individuals & that of complete duplication is 1 in 500 individuals. One-sided complete duplication of ureter results in 40% chances of complete duplication abnormality on other side. 10% siblings may be affected by this anomaly.

According to a study of 3600 intravenous pyelograms by Erdem Gmisburun the percentage of ureteral duplication was 2.61% while in our study it is 2%. According to his research it is more common in females, unilateral duplication is 6 times more commonly seen than bilateral one. Incomplete duplication was 72.9% i.e. incidence is more than complete variety.

The cause of double ureter is unknown. Double ureter occurs as a congenital anomaly in association with other abnormalities of different systems in certain syndromes like: Trisomy 13, Trisomy 18 (Edwards syndrome) which occur due to the extrachromosomal presence. In Caudal duplication syndrome there is duplication of caudally lying organs like vagina, cervix, distal colon & ureter.

Maternal Diabetes increases the chances of developmental anomalies of skeletal system heart & urogenital system (ureteral duplication, renal agenesis ureterocele & hydronephrosis) in newborns. The infants conceived by intracytoplasmic sperm injections & invitro fertilization have high risk of developing birth defects of CVS, musculoskeletal system & urogenital system (duplex ureter).

Many anomalies are sometimes associated with double ureters like ureterocele, ureteral ectopia, vesicoureteral reflux, ureteropelvic junction stasis, ureteric calculi causing urinary retention.

Jee LD, Rickwood AM, William MP & Anderson PA have done an ultrasonographic study in neonates for detection of anomalies associated with duplex system. Ureteral duplication has been reported along with. Primary adenocarcinoma & transitional cell carcinoma. S.Shimizu & H.Kojima has reported a case of pregnant female who developed an infection with Serratia marcescens & was having double ureter, which caused spread of infection to chorioamnion resulting in spontaneous abortion.

It was presented in Asian transplant congress in Bangkok that kidney with double ureter can be taken & transplanted safely. Geyky & Knight R have reported a successful transplant of kidneys with bilateral double ureters. The knowledge of Anatomic variation i.e. of double ureter is quite important in choosing the most appropriate type of operation for vesicoureteral reflux,
renal calculus & ureterocoele. Because if renal function is intact then surgery is the treatment of choice.  

Conclusion:
This knowledge of ureteric duplication is important for Anatomists, Paediatricians (clinicians) for diagnosis of certain syndromes & surgeons for planning surgery in complicated cases of double ureter & above all for successful transplantation of kidneys with double ureters.

References: