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Research Article

MANAGEMENT OF SINGLE SYSTEM ECTOPIC URETER AN OVERVIEW

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ABSTRACT
An ectopic ureter is characterized by an ectopic ureteric orifice outside the posterolateral extremity of bladder trigone mostly often with renal duplication. If it drain single kidney, it is called single system ectopic ureter (SSEU). In female child this entity leads with pseudo incontinence (Urinary incontinence with normal pattern voiding). The presumed embryologic mechanism is that the ureteric bud arises more cranial than usual from the mesonephric duct, so that it is taken in to the bladder wall at a lower level during the seventh week of gestation or does not join the bladder wall at all. Treatment can be nephroureterectomy or ureteric reimplantation.

Key Words: *Single System Ectopic Ureter, Pseudo Incontinence, Dysplastic Kidney*

INTRODUCTION
An ectopic ureter is characterized by an ectopic ureteric orifice outside the posterolateral extremity of bladder trigone & present predominantly in female children with duplicated kidney (Richard *et al.,* 2007). If ectopic ureter drain single kidney, it is called single system ectopic ureter (SSEU) (Choudhary *et al.,* 2001). Single system ureteral ectopia and associated congenital dysplastic kidney is surgically curable etiology of incontinence with other wise normal pattern of voiding in female child. .In contrary to western literature proportion of incidence of duplex system EU & SSEU is reversed in Asian literature (Joshi *et al.,* 2009, Gangopadyaya *et al.,* 2007)

MATERIALS AND METHODS
From April 2000 to May 2010, eight cases of single system ectopic ureter were managed in department of pediatric surgery PGIMS rohtak, Haryana (India). In a retrospective review, case notes, including presentations, findings on examination, imaging studies, operative details, pathological findings & postoperative outcome were studied. Purpose of this study was to inquire into the presentation, methods for diagnosis & therapy of single system ectopic ureter managed in our institute.

RESULTS
Out of eight cases seven were female & one was male, their age ranging from 3.5 to 13 years, with a mean of 7.4 years. All patients were case of unilateral ectopic & presented with complain of urinary incontinent (including male patient) with normal voiding pattern. Routine blood & urine examination was normal in all patients. Ultrasonography (USG) of abdomen & pelvis could localized the both kidneys in seven patients, in one 13 years old female child who was referred to us as right solitary kidney with urinary incontinent but with combine efforts of radiologist & surgeons left small dysplastic kidney could be localized with USG. Though in three patients kidney were very small but because of massive ureteric dilatation we could localized dysplastic renal tissue. All patients were subjected to examination under anaesthesia & genitoscopy prior to exploration. On examination under anaesthesia we could localized & succeed in intubations of the ureteric opening situated at vestibule in three patients ectopic opening in rest

patients could not be seen. On exploration all kidneys was found in lumber region & ureter was grossly dilated in three patients (Fig:-1) & all patients were underwent nephroureterectomy, urinary incontinence was disappear in all patients with in 24 hr of postoperative period. Histopathology was suggestive of dysplastic kidney in five patients & hypoplastic kidney in three patients.

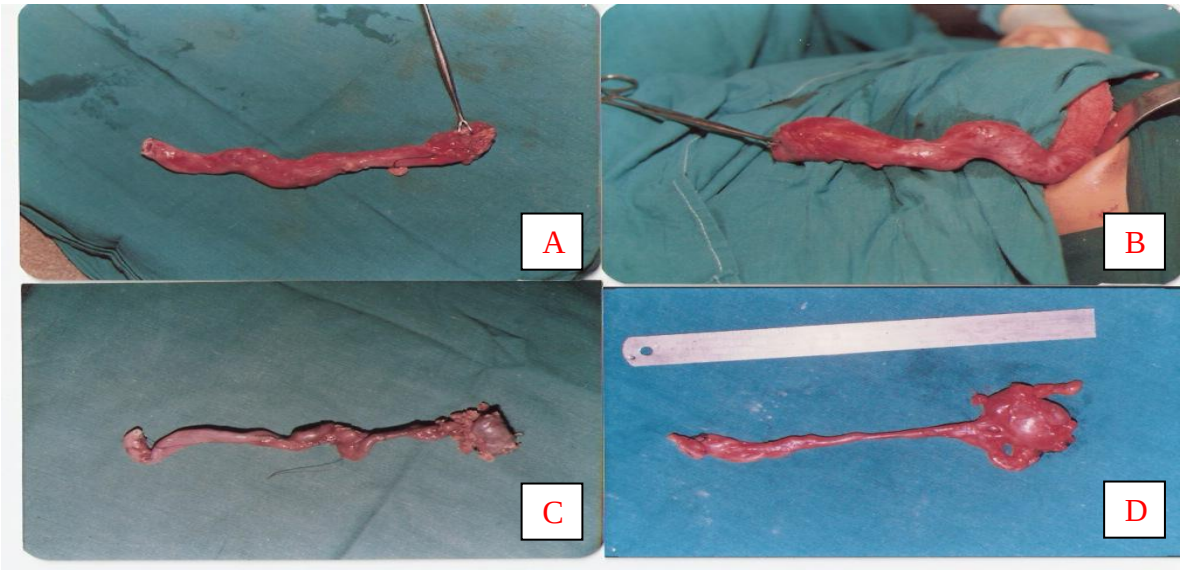

Figure 1: A is nephroureteric specimen B showing per operative photograph showing massively dilated & tourtuse ureter of C & D showing mild to moderate dilatation of ureter with dysplastic kidney in a pile of fate

DISCUSSION

The word ectopia is derived from the Greece word ex (out) & topos (place), any ureter whose orifice terminate any where other than the normal trigonal position is consider ectopic however, in general practice, term ectopic ureter is meant to imply a ureter whose orifice terminate even more caudally, such as in urethra or outside urinary tract(Richard et al.,2007). There are two distinct form of ectopic ureter (EU), one draining a duplex kidney & other connected to single kidney, the latter being termed SSEU (Choudhary et al.,2001).

The incidence of ureteral ectopia was approximately 1 in 2000 in one series of autopsies in children. Many patients remain asymptomatic & true incidence is difficult to determine (Richard *et al.,* 2007). In females ectopic ureter occurs two to three times more commonly than in males. More than 80% of females with ectopic ureters have duplex systems. The majority (75%) in males are singles (Gangopadhyaya *et al.,* 2007) .In contrary to western literature proportion of incidence of duplex system EU & SSEU is reversed in Asian literature (Joshi *et al.,* 2009, Gangopadhyaya *et al.,* 2007). In our series seven out of eight patients were female & one was male.

Associated anomalies with SSEU included renal dysplasia, anal stenosis, urethral-rectal fistula & bladder agenesis (Weight *et al.,* 2006). Decrease renal function in draining unit is frequently encountered which may result from primary dysplasia, obstruction, vesicoureteral reflux or recurrent infection (Wunsch *et al.,* 2000). In our patients we encountered hydroureter in four cases which may be because of stenosis at the ureteric orifice, renal dysplasia (in five patients) & renal hypoplasia (in two patients).

The presumed embryologic mechanism is that the ureteric bud arises more cranial than usual from the mesonephric duct, so that it is taken in to the bladder wall at a lower level during the seventh week of gestation or does not join the bladder wall at all. Maizels & Stephens postulated that abnormal growth of

spine preclude normal renal ascent & may even be responsible for renal agenesis & hypoplasia. Abnormal location of the ureteral bud probably leads to abnormal interaction with nephrogenic blastemia & renal dysplasia (Maizels and Stephens, 1979).

It is also postulated that failure of vascular supply during embryological development prevent kidney from developing normally & causes formation of a small primary organ that contain embryonal tissues. Some researchers believe that it is the obstruction of ureter during an early phase of the embryo that stops development of the kidney (Gangopadhyaya *et al.,* 2007). But all above theories can't explain the massive dilatation of ureter that was found in four patients of our series.

.A small, dysplastic, poorly functioning single kidney associated with an ectopic ureter may be difficult to localize, leading to delay in diagnosis or even to misdiagnosis as a solitary kidney (Choudhary *et al.,* 2001). The delay in reaching the correct diagnosis is greater in children who do not show evidence of functioning renal tissue on the side of the ectopic ureter. Borer *et al.,* 1998 reported a mean time of 5.7 years after presentation to reach the correct diagnosis. Kesavan *et al.,* 1977 reported a similar earlier series with delayed diagnosis. A high index of suspicion with advanced diagnostic tool which enable to detect very small poorly functioning kidney is necessary to change scenario.

High index of suspicious is key point of diagnosis. B mode ultrasound & nuclear scan or IVP recommended for localizing the kidney. Small dysplastic kidney may miss in USG & Nuclear scan. With nuclear scan we can detect relative function of kidney which would help in deciding the method of treatment (nephrouretectomy vs ureteric reimplant). MR urography considered most sensitive method of localizing kidney & site of ureteric insertion. MRU picked up the dysplastic moieties and their location as well as functional status and also depicted the course of the ectopic ureter (Joshi *et al.,* 2009).

Jeong *et al.,* 2007 reported series of 16 patients of SSEU underwent laparoscopic nephroureterectomy with incidence of small bowel injury in one case. Joshi et al (2009) reported the series of eight cases of SSEU underwent laproscopic nephroureterectomy with uneventful recovery. Park et al (2009) described the laproscopic nephroureterectomy with use of single port in patient of SSEU.

Treatment can be nephroureterectomy or ureteric reimplantation. Dysplastic kidney & those with function of less than 10% are normally removed (Gangopadhyaya *et al.,* 2007). For our patients excision was a simple & valid treatment .reimplantation indicated in case of bilateral (Wunsch *et al.,* 2000) or with good renal function. In case of bilateral single ectopic ureters, subsequent malformation of the bladder trigone and bladder neck may result in additional voiding dysfunction, and ureteral reimplantation alone may not solve the urologic problems (Heuser *et al.,* 2002). Similar to our experience Borer et al have reported complete dryness in seven girls of SSEU after nephroureterectomy. But Choudhary *et al.,* (2001) reported persistent wetting for few month in patients of SSEU after nephroureterectomy & purposed that it may be related to inherently weak urethral sphincter.

Conclusion

Single system ectopic ureter (SSEU) most often associated with dysplastic Kidney and presented with pseudo incontinence and can be manage by nephrouretectomy and it has very good prognosis in term of urinary continence.

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