INTRODUCTION

Ureterocele is a congenital cystic dilatation of the terminal portion of the ureter. Ureteroceles are a relatively common cause of urological problems observed. The defects may be unilateral or bilateral, and can be associated with normal, ectopic or duplicated ureters ***(Madeb et al., 2001).***

Ureteroceles arise from abnormal embryogenesis, with anomalous development of the intravesical ureter, the kidney and the collecting system. Because of the chronic obstructive nature of ureteroceles the activity of the affected renal unit also ranges from a normal well functioning kidney to a nonfunctioning dysplastic renal unit ***(Minevich et al., 2005).***

Few entities in paediatric urology present as clinical challenges, one of them is the ureterocele. Ureteroceles have varied effects as regards obstruction, reflux, continence, recurrent urinary tract infection and renal function; hence, each ureterocele must be managed on an individual basis as a unique entity and not by a simple algorithm. It's imperative for the treating physician to be acquainted with the multiple presentations, radiological appearances and treatment options of ureteroceles, as well as the complications to avoid. Such knowledge yields the best clinical results **(*Schlussel and Retik, 2002*).**

Ureteral anomalies are some of the most significant anomalies in all of pediatric urology because they directly affect overall renal function. These congenital problems may manifest acutely or insidiously. Similarly, if they are incorrectly treated, the adverse outcome may not be appreciated for years. Appropriate management is predicated on knowledge of the relevant embryology, anatomy, and physiology, as well as all the variants thereof. Finally, the urologist entrusted with the care of these children must be familiar with the many reconstructive techniques available so that an optimal outcome can be achieved ***(Schlussel and Retik, 2002).***

A large range of treatment options have been proposed in view of the extreme variety of urological lesions observed. Total or partial resections have been replaced by minimally invasive surgery, and more conservative endoscopic procedures, often modulated according to the type of lesion, the child's age and general state at the time of diagnosis and current practices ***(Sauvage et al., 2002).***

Epidemiology:

Campbell in 1951 reported the incidence of ureteroceles to be 1 in 4,000 autopsies of children. Malek and colleagues reported in 1972 an incidence between 1 in 5,000 and 1 in 12,000 in general pediatric admissions. Both of these estimates are probably low, suggesting that small ureteroceles were missed. Uson and colleagues reported a much higher incidence of 1 in 500 autopsies ***(Uson et al., 1961).***

Ureteroceles occur most commonly in whites and are unusual in blacks. Although females are affected four to seven times more often than males, the malformation is often more complex in male ***(Eklof et al, 1987).***

Some series have demonstrated a slight left-sided predominance; approximately 10 % are bilateral ***(Royle et al, 1971)***.

Of ureteroceles, 60% to 80% are ectopic ***(Brookes and Zeitman, 1998; Ericsson, 1954; Mandell et al, 1980)***, and approximately 80% are associated with upper pole ureter of a duplex kidney ***(Brock and Kaplan, 1978; Ericsson, 1954; Mandell et al, 1980; Stephens, 1963).***

Intravesical ureteroceles associated with single ureter are seen more in adults than in children and may be an acquired lesion ***(Thompson and Kelalis, 1964)***.

Although the severity of obstruction and hydroureteronephrosis is greater in children than in adults, single system ureteroceles usually are associated with better function and less hydronephrosis than are duplex renal units and their more commonly associated ectopic ureteroceles. ***(Rabinowitz et al, 1978)***

Single system ectopic ureteroceles are rare, usually occur in men, and may be associated with cardiac and genital anomalies. Associated urologic anomalies, especially renal

anomalies of fusion and ectopia, often occur with ureteroceles ***(Johnson and Perlmutter, 1980).*** When the ureterocele arises from the upper moiety of a duplex kidney, this upper moiety frequently displays renal dysplasia ***(Perrin et al, 1974; Snyder et al, 1983).***

AIM OF THE WORK

In this essay we aim to discuss the different types of ureteroceles as regards their clinical, diagnostic and therapeutic alternatives.

EMBRYOLOGY OF THE LOWER URINARY TRACT

Development of the three embryonic kidneys:

The embryonic kidneys are in order of their appearance, the pronephros, the mesonephros, and the metanephros. The first two kidneys regress in utero and the third becomes the permanent kidney.

- **Pronephros:**

In humans, the first evidence of pronephros is seen late in the 3rd week, and it completely degenerates by the start of the 5th week. The pronephroi develop as five to seven paired segments in the region of the future neck and thorax. Development of the pronephric tubules starts at the cranial end of the nephrogenic cord and progresses caudally. As each tubule matures, it immediately begins to degenerate along with the segment of the nephric duct to which the tubules are attached ***(Davies and Bard, 1998).***

- **Mesonephros:**

In humans, it serves as an excretory organ for the embryo while the definitive kidney, the metanephros, begins its development. Development of the mesonephric ducts (also called wolffian ducts) precedes the development of the mesonephric tubules. The mesonephric ducts can be seen at

about the 24th day as a pair of solid longitudinal tissue condensations developing parallel to the nephrogenic cords in the dorsolateral aspect of the embryo. Their blind distal ends grow toward the primitive cloaca, and soon fuse with it at about the 28th day. This fused region later becomes a part of the posterior wall of the bladder. As the ducts fuse with the cloaca, they begin to form a lumen at the caudal end. This process of canalization then progresses cranially, transforming the solid condensations into the definitive mesonephric ducts.

By the 4th month, the human mesonephroi have almost completely disappeared, except for a few elements that persist into maturity. Certain elements of the mesonephroi are retained in the mature urogenital system as part of the reproductive tract. In males, some of the cranially located mesonephric tubules become the efferent ductules of the testis. The epididymis and vas deferens are formed from the mesonephric (wolffian) ducts. In females, remnants of cranial and caudal mesonephric tubules form small, nonfunctional mesosalpingeal structures called the epoöphoron and the paroöphoron ***(Davies and Bard, 1998).***

- **Metanephros:**

The definitive kidney, forms in the sacral region as a pair of new structures, called the ureteric buds, sprouts from the distal portion of the mesonephric duct and comes in contact with the blastema of metanephric mesenchyme at about the 28th day ***(Davies and Bard, 1998).***

Development of the Ureter

At the 4 weeks' gestation, an out pouching arises from the distal mesonephric duct. This out pouching is the ureteric bud, and it interacts with a mass of mesenchyme that is the metanephric blastema. This interaction results in the ureteric bud's branching and developing into calyces, renal pelvis, and ureter ***(Moore, 1988).*** **(Fig. 1).**

The segment of mesonephric duct distal to the ureteric bud is the common excretory duct. This duct eventually is absorbed into the developing bladder and becomes part of the trigone. The point of origin of the ureteric bud is the ureteral orifice. When the common excretory duct is absorbed into the bladder, the ureteral orifice begins to migrate in the bladder in a cranial and lateral direction ***(Moore, 1988).***

By day 33 of gestation, the common excretory ducts (the portion of the mesonephric ducts distal to the origin of the ureteric buds) dilate and become absorbed into the urogenital sinus. The right and left common excretory ducts fuse in the midline as a triangular area, forming the primitive trigone. The ureteric orifice exstrophies and evaginates into the bladder by day 37 and begins to migrate in a cranial and lateral direction within the floor of the bladder. During this process, the mesonephric (wolffian) duct orifice diverges away from the ureteric orifice and migrates caudally ***(Merlini et al, 2004).*** **(Fig. 2).**

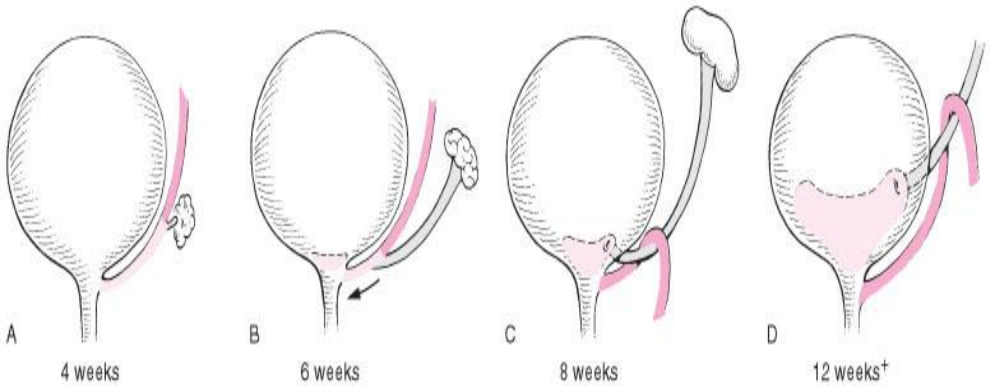

Fig. (1): The ureteral bud further develops into the ureter and induces the metanephric blastema to differentiate and become the kidney. The common excretory duct is progressively absorbed into the bladder and becomes the trigone. The mesonephric duct will become the vas deferens in the male ***(Schlussel and Retik, Campbell's urology, 2002).***

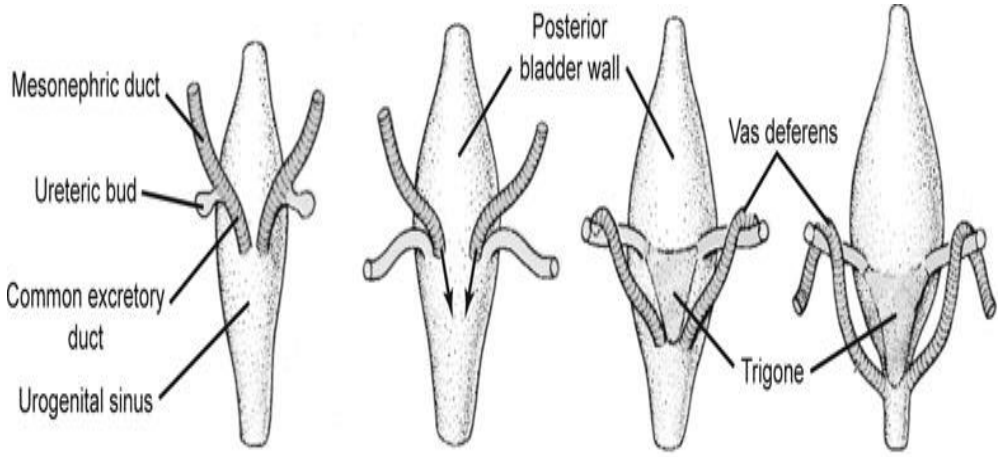

Fig. (2): Incorporation of the mesonephric ducts and ureteric buds into the bladder wall. Between the 4th and 6th weeks, common excretory ducts, the terminal portion of the mesonephric ducts caudal to the ureteric bud formation, exstrophy into the posterior wall of the developing bladder. The triangular region of exstrophied mesonephric ducts forms the trigone of the bladder. This process brings the ureteric bud openings into the bladder wall, while the mesonephric duct openings are carried inferiorly to the level of pelvic urethra. ***(Modified from Sadler TW: Langman's Medical Embryology. Baltimore, Williams & Wilkins, 1985)***

If two separate ureteric buds originate from the mesonephric duct, two complete and separate interactions will develop between the ureter and the metanephric blastema. The result is two separate renal units and collecting systems, ureters, and ureteral orifices. This complete duplication is synonymous with a duplex system drained by double ureters. The final position of the ureteral orifices has important clinical implications. Both ***Weigert (1877)*** and ***Meyer (1946)*** noted that there is a constant trigonal relationship between the upper and lower pole orifices. When performing a cystoscopic examination, it is important to remember this counterintuitive concept: the so-called lower or distally placed orifice is in fact the orifice of the upper pole, and the so-called higher or cranial orifice is the lower pole orifice. The lower pole orifice is more cranial and lateral to the caudad, medial upper pole orifice. To achieve these positions, the two ureters and orifices rotate 180 degrees clockwise on their longitudinal axes ***(Weigert 1877 and Meyer, 1946)*** **(Fig. 3).**

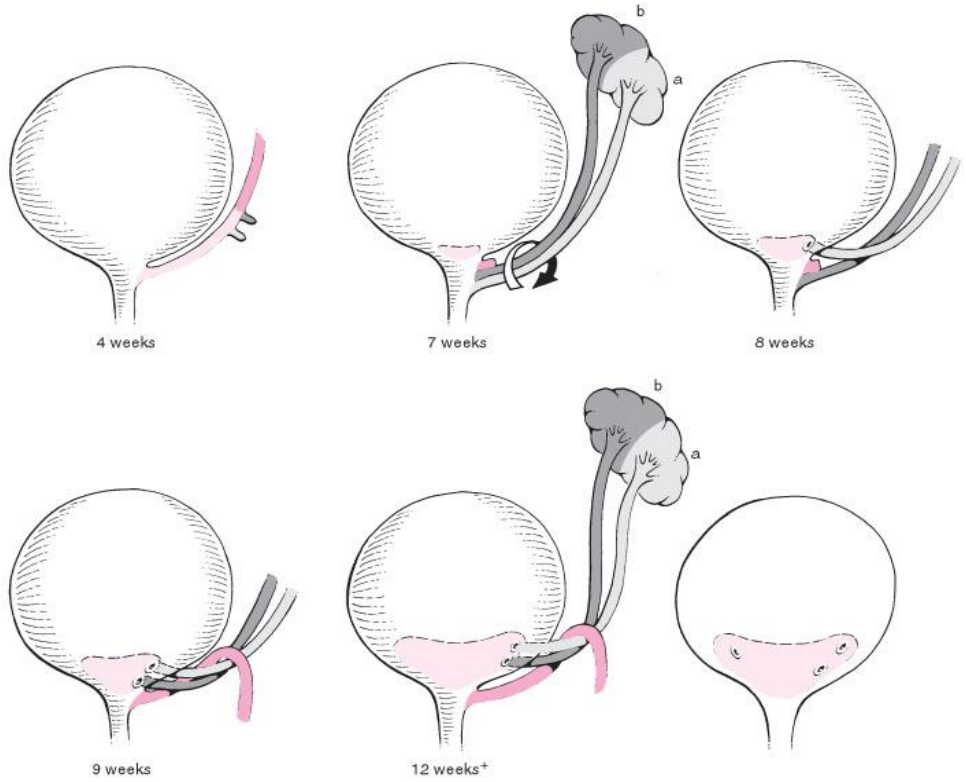

Fig. (3): The Weigert-Meyer rule is depicted. The upper pole ureter and the lower pole ureter rotate on their long axes to yield an upper pole orifice (b) that is medial and caudal to the lower pole orifice (a) ***(Schlussel and Retik, Campbell's Urology, 2002)***

The mechanism of ureteric orifice incorporation into the developing bladder is inferred primarily from clinical observations of duplex kidneys. The upper pole ureteric orifice rotates posteriorly relative to the lower pole orifice and assumes a more caudal and medial position. Weigert and Meyer recognized the regularity of this relationship between upper and lower pole ureteric orifices, which has come to be known as the *"Weigert-Meyer rule*". According to this concept, an abnormally lateral lower pole ureteric orifice may result from a ureteric bud's arising too low on the mesonephric duct, which causes premature incorporation and migration within the developing bladder.

In such a ureteric orifice, vesicoureteral reflux is more likely to occur because of an inadequate intramural tunnel. In contrast, an abnormally caudal upper pole ureteric orifice may result from a ureteric bud's arising too high on the mesonephric duct. It may drain at the bladder neck and verumontanum, or it may remain connected to the mesonephric duct derivatives (e.g., vas deferens) ***(Mackie and Stephens, 1977; Schwarz and Stephens, 1978).*** In females, the ectopic upper pole ureter may insert into the remnants of the mesonephric (wolffian) ducts or into the vaginal vestibule.

The ureter begins as a simple cuboidal epithelial tube, surrounded by loose mesenchymal cells, which acquires a complete lumen at 28 days of gestation in human. It was suggested that the developing ureter undergoes a transient luminal obstruction between 37 and 40 days and subsequently recanalizes ***(Alcaraz et al., 1991).*** It appears that this recanalization process begins in the midureter and extends in a bidirectional manner both cranially and caudally. In addition, another source of physiologic ureteral obstruction may exist as Chwalla's membrane, a two-cell-thick layer over the ureteric orifice that is seen between 37 and 39 days of gestation.

In humans, urine production is followed by proliferative changes in the ureteral epithelium (bilaminar by 10 weeks of gestation). The epithelium attains a transitional configuration by 14 weeks. The first signs of ureteral muscularization and development of elastic fibers are seen at 12 weeks of gestation.

In humans, the ureteral smooth muscle phenotype appears later than that of the bladder. Smooth muscle differentiation is first detected in the subserosal region of the bladder dome and extends toward the bladder neck and urethra, whereas smooth muscle differentiation of the ureter occurs later within the subepithelial region in the ureterovesical junction, ascending toward the intrarenal collecting system ***(Baker and Gomez, 1998).***

In the embryonic ureter and bladder, it is likely that epithelial-mesenchymal interactions are important in the development of urothelium, lamina propria, and muscular compartments, but the exact nature of this induction process is unknown. Before 10 weeks, elastic fibers are few in number, poorly developed and randomly arranged. After 12 weeks, these fibers become more numerous throughout the ureter and are seen with specific orientation ***(Escala et al., 1989).***

Embryology of ureteroceles:

At 37 days' gestation, Chwalle's membrane, a two-layered cell structure, transiently divides the early ureteric bud from the urogenital sinus ***(Chwalle, 1927).*** The stenotic orifice commonly seen in the ureterocele has led several researchers to postulate that this dilatation results from incomplete dissolution of Chwalle's membrane. Others have theorized that the affected intravesical ureter suffers from abnormal muscular development; without the appropriate muscular backing, the

distal ureter assumes balloon morphology ***(Tokunaka et al, 1981).***

A third theory implicates a developmental stimulus responsible for bladder expansion acting simultaneously on the intravesical ureter ***(Stephens, 1971).*** Alternatively, ***(Tanagho, 1976)*** hypothesized that the distal ureteral segment, which is incorporated later into the developing urogenital sinus may be acted upon by the same factors that causes the expansion of the urogenital sinus to form the bladder, together with a delay in establishing the lumen of the ureteral bud. Unquestionable evidence does not exist to support any of these theories, which, in fact, have very little effect on clinical practice.

ANATOMY

General Description

Each ureter represents the tubular extension of the renal collecting system, which courses downward and medially to connect the kidney to the urinary bladder. In the adult, the ureter is generally 22 to 30 cm in total length, varying with body size and habitus. As noted earlier, its origin at the ureteropelvic junction is often vaguely defined in the normal state. The ureter and collecting system extending to the renal papillae are lined by a transitional cell epithelium, identical to and contiguous with that of the bladder. Beneath this epithelium is a layer of connective tissue, the lamina propria, which together with the epithelium forms the mucosa ***(Kabalin, 2002)***.

When not distended by urine, the ureteral mucosa lies in longitudinal folds. Smooth muscle covers the renal calyces, renal pelvis, and ureter. In the ureter, this muscle usually can be divided into an inner layer of longitudinally coursing muscle bundles and an outer layer of circular and oblique muscle. In the normal state, the urinary effluent does not passively drain but is actively propelled from renal pelvis to bladder by the peristaltic action of the ureteral muscle. A thin layer of adventitia immediately surrounds the ureter and contains an extensive plexus of ureteral blood vessels and lymphatics that course longitudinally with the ureter ***(Kabalin, 2002)*** **(Fig. 4).**