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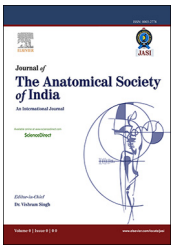

Case Report

# Multicystic dysplastic kidney with twice duplicated distal ureter having ectopic blind endings

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ABSTRACT

Congenital abnormalities of the kidneys, urinary collecting system, and bladder come at an incidence of roughly 1% of total live births. These urinary abnormalities form a major problem in urological studies both of interest to the surgeons and anatomists, of which inverted "Y" shaped ureter, ectopic ureter, blind ending ureter, and multicystic kidney are a very rare spectrum of anomalies. Coexistence of such a complex spectrum of anomalies in the same individual is of immense surgical importance.

During dissection, retro peritoneum of intra uterine dead (IUD) male fetus of 28 weeks gestation a combination of multicystic dysplastic kidney with a rare ureteral anomaly that is not seen frequently was observed. The normal left renal anatomy was completely distorted. Ureter divided twice in an inverted "Y" branching pattern, the two left limbs entered the wall of rectum and urinary bladder of same side respectively and right limb opened ectopically into urinary bladder. The gross and microscopic features were in favor of Multicystic Dysplastic Kidney and Ureter. Congenital renal dysplasia may be explained by an abnormal induction of metanephric blastema by migrating ureteric bud. The embryological basis of inverted "Y" shaped ureter is not clearly understood, we suggest that it is due to longitudinal splitting of ureteric bud. This type of anomaly of the ureter may represent real traps in the interpretation of abdominal imaging, particularly in CT scanning.



## 1. Introduction

Congenital malformations represent defects in morphogenesis during early fetal life. Congenital abnormalities of the kidneys, urinary collecting system, and bladder come at an incidence of roughly 1% of total live births.[1] Multicystic dysplastic kidney (MCDK) is a congenital maldevelopment, in which the renal cortex is replaced by numerous cysts of multiple sizes, resembling a bunch of grapes, occurring in 1 in 1000 to 4300 live births, most often on the left side.[2] MCDK shall be predominant in boys than girls. Most common age of presentation was within first month of life,[3] and MCDK can be diagnosed by high resolution ultrasonography in the neonatal period. Multicystic kidney is usually unilateral, segmental or focal associated with abnormalities of collecting system.[4] It

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may remain clinically silent throughout life or may involute with time. In children who survive, there is an increased risk of Wilm's tumor and renal cell carcinoma.[5] 50% of MCDK are associated with other urological defects such as ureteral atresia, reduplication, meckel syndrome, neural tube defects, Kallman syndrome, agenesis or dysplasia of the kidney, and agenesis or cysts of the seminal vesicles.

Of the ureteric anomalies, bifid ureters and double ureters are more frequently seen, but triple ureters are reported by Boopathi in 1 out of 11 patients.[6] According to the Committee on Terminology, Nomenclature, and Classification of the Section on Urology of the American Academy of Pediatrics, ureteral ectopia is defined as a ureter that terminates into an abnormal location.[7] Several authors have described the possible ectopic endings of ureter in male and females. The common sites in decreasing order in males are the posterior urethra, prostatic utricle, seminal vesicle, ejaculatory duct, vas deferens and in females urethra, vestibule, vagina, cervix and uterus, but the list remains without rectum.[8] Ureteral ectopia is 2–12 times more common in females.[9] We report, to the best of our knowledge, a rare occurrence of the coexistence of all these anomalies in a single individual.

## 2. Observation

Autopsy done in a male intra uterine dead fetus of 28 weeks, obtained from Obstetrics and Gynecology Department of Government Medical College, Thrissur, a combination of multi cystic dysplastic kidney with ureteral anomaly was observed

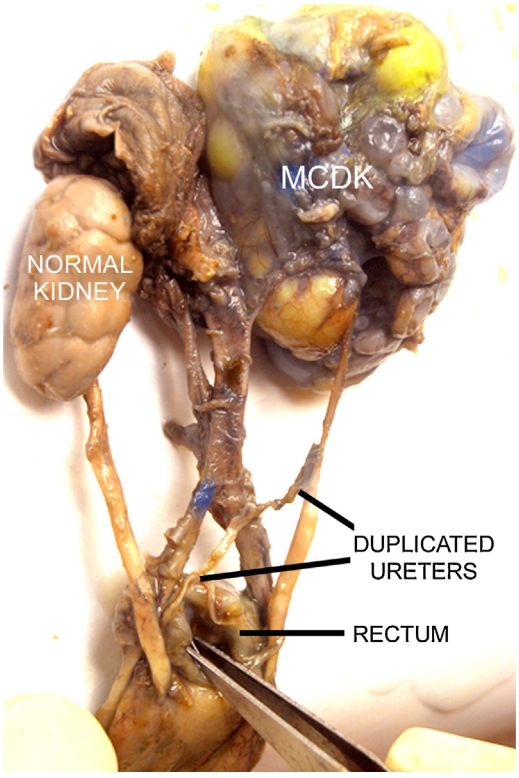


**Fig. 1 – Showing gross appearance of kidney with multiple cysts of varying sizes resembling bunch of grapes and duplicated distal part of ureter.**

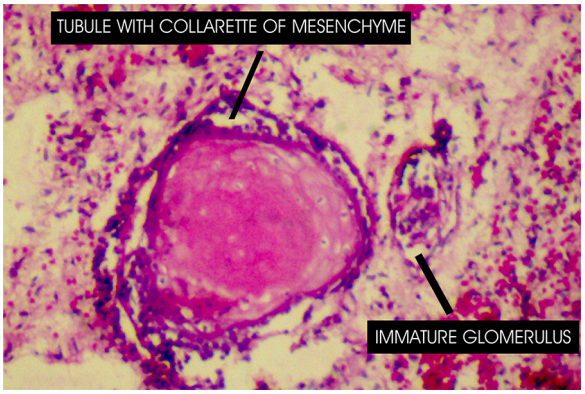


**Fig. 2 – Microsopic appearance of left kidney stained with Hematoxylin and Eosin showing immature glomerulus and multiple tubules with dysplasia.**

(Fig. 1). The gross and microscopic appearance of the left kidney and ureters was observed in detail. Normal renal anatomy was completely distorted. Left kidney measured $10 \times 7 \times 4$ cm, dilated and cystic, extending from T10 to L5 vertebra. Cysts were of varying sizes from 2 to 12 mm, walls thin and contained clear white fluid. The opposite kidney was normal in shape, size, and structure.

Three cms distal to its formation left ureter divided into two in an inverted "Y" manner. The left division had a normal termination into urinary bladder. The right division after entering pelvis again had an inverted "Y" branching. Both limbs ended blindly as well as ectopically, right one into urinary bladder and left into rectum. Such a variant ureter presented here was not seen in any of the available literature. Small diameter of ureter hindered introduction of a probe. Malformations of other organs were not observed. Photographs of kidney were taken after injecting two different color solutions into the cyst. All the five limbs of ureter, bladder and kidney sections were taken for histo-pathological studies for confirmation.

Microscopically kidney showed multiple dilated disorganized tubules and ductules lined by single layer of low cuboidal epithelial cells with collarrette of mesenchyme; cartilage confirming dysplasia (Fig. 2). Section of the duplicated parts of ureter showed lumen lined by transitional epithelium and smooth muscle. Smooth muscle in the walls of ureter confirmed using mallory's trichrome stain (Fig. 3).

## 3. Discussion

Anatomical variations and congenital anomalies of kidney and ureter were well described by Edmund Papin and Daniel Eisendrath. Usually ureteral reduplication is associated with a hypoplastic kidney that develops around second ureter which may end blindly below. In general, the more ectopic the uereteral orifice, the more the involved moiety of kidney.[10] In this case the multi cystic dysplastic kidney is associated with partial reduplication of distal ureter which ends blindly below