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Original Research Article

A HUMAN CADAVERIC STUDY ON INCIDENCE AND MORPHOLOGY OF ANATOMICAL VARIATIONS OF KIDNEY AND URETER WITH EMPHASIS ON ITS EMBRYOLOGICAL, GENETIC AND CLINICAL SIGNIFICANCE

Shweta B Shambharkar *[1], Shabana Borate [2], Suresh Gangane [3].

*[1] Assistant Professor, Dept of Anatomy, Govt. Medical College, Gondia, Maharashtra, India.

[2] Associate Professor, Dept of Anatomy, Grant Govt. Medical College, Mumbai, Maharashtra,India.

[3] Ex Professor and Head, Dept of Anatomy, Grant Govt. Medical College, Mumbai, Maharashtra, India.

ABSTRACT

Background: Kidneys are amongst the common sites of congenital abnormalities and anatomical variations. Congenital anomalies of kidney and urinary tract (CAKUT) constitute approximately 20 to 30% of all anomalies identified in the prenatal period and a leading cause of renal failure in children. The common variations pertaining to kidney are polycystic kidney, unascended kidney, horseshoe kidney with fused upper or lower pole, atrophic kidney, lobulated kidney, malrotated kidney, bifid pelvis or ureter, most common being bifid pelvis and pancake kidney is a very rare variant.

Materials and Methods: Fifty human adult cadavers were included in our study; observed and studied over a period of three years in the Department of Anatomy, Grant Govt. Medical College, Mumbai, during routine dissection.

Results: In our study we found, 01.01% of renal agenesis, 01.01% of fused pelvic or pancake kidney, 01.01% of malrotated kidney, 02.02% of unascended kidneys, 05.05% hypoplastic or atrophic kidneys, 07.07% lobulated kidneys, 05.05% polycystic kidneys, 02.02% of bifid pelvis and 03.03% of triplicate pelvis.

Embryological basis: The development of kidney begins at the fourth week of gestation; the failure of proper inductive interaction between the ureteric bud and the metanephric blastemal can lead to various congenital anomalies. Anomalies can result due to abnormal development, ascent, rotation and migration.

Genetic basis: CAKUT are either sporadic, familial, syndromic or non-syndromic. Transcription factor 'WT1' produced by mesenchyme of the metanephric blastemal helps in epithelialization of ureteric bud. Congenital abnormality occurs when there is mutation of genes that regulates the expression of WT1.

Conclusion: Renal anomalies are one of the commonest anomalies which may remain unnoticed till adulthood. The knowledge of anatomical variations of kidney and ureter is of utmost importance for surgical and uroradiological interventions. Hence an early detection and proper follow-up may be helpful in better management and increased survival rates.

KEY WORDS: Variations, Polycystic, Atrophic, Lobulated, Unascended, Pancake, Triplicate.

Address for Correspondence: Dr.Shweta B Shambharkar, Plot No.43/B, Dhadiwal Layout, Suyog Nagar Square, Post: Parwati Nagar, Nagpur, Maharashtra-440027, India.
Phone: 9960810368 **E-Mail:** shambharkar.shweta4@gmail.com

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INTRODUCTION

Congenital anomalies of kidney and urinary tract (CAKUT) are common findings on fetal ultrasound [1]. It is common in children and represent approximately 30% of all prenatally diagnosed malformations [2]. CAKUT play a major role in morbidity and mortality. It accounts for approximately 3.3 to 11.1% incidence in general population and about 50% of all congenital abnormalities [3]. It occurs at a frequency of 1 in 500 live births and are a common cause of renal insufficiency in childhood [4]. Many of the anomalies range from mild asymptomatic malformations such as double ureter or minimum renal pelvic obstructions to severe pathologies such as renal agenesis, renal dysplasia, horseshoe kidney etc. which are many times fatal [5].

Male preponderance is seen in renal agenesis. Unilateral renal agenesis (URA) commonly affects approximately 1 in 1000 births while bilateral is reported to be 1 or 2 in every 10000 births [6]. In URA, left kidney is more commonly absent compared to right. URA may be asymptomatic and often incidentally diagnosed by USG or CT secondary to another condition with the contralateral kidney demonstrating compensatory hypertrophy. Bilateral renal agenesis is invariably fatal [7].

Renal fusion anomalies are reported in 0.1-0.2% of all live births. The most extreme form of renal fusion is the pancake kidney, thought to make less than 10% of all renal fusion anomalies [8]. Congenital renal fusion anomalies characterized by either partial or complete fusion of the two kidneys are represented by horseshoe kidney, crossed renal ectopia with or without fusion and fused pelvic cake kidney. Though these anomalies may remain asymptomatic in certain cases they may be associated with pathological conditions like nephrolithiasis, hydronephrosis, vesicoureteral reflux and renal neoplasms [9]. These renal fusion anomalies exhibit abnormalities of position, migration, rotation and vascular supply. They occur frequently in males [10]. Pancake or cake kidney is one of the rarest types of renal ectopia. The two kidneys are fused at the medial borders of each pole to produce doughnut or ring-shaped mass. When there is more extensive fusion along the entire medial aspect of each kidney, a disc or shield shape is created. The pelvis is anteriorly placed and ureters remain uncrossed. Each collecting system drains its respective half of the kidney and does not communicate with the opposite side [11].

Rotational anomalies of kidney are a rare entity [12]. Though rare this type of anomaly has wide implications in the context of advanced surgical procedures and diagnostic evaluation of kidney donors. Anomalies of renal rotation are associated with renal ectopia and the fusion anomalies but may be exhibited by the kidneys which are otherwise normally placed [13].

Failure of the kidneys to ascend into the renal fossa in utero results in ectopic or unascended kidney. Such kidney is often found in pelvis however it may be placed higher up in lower lumbar region. Pelvic kidneys often become hydronephrotic due to obstruction of the anteriorly placed ureter and an anomalous arterial supply [14].

Children with renal dysplasia may have abnormal renal tubules and tend to lose essential water and sodium in urine. Hypertension and proteinurea may develop in children with renal dysplasia and further aggravate renal function [15]. Hypoplasia usually occurs due to inadequate ureteral bud branching and results in small kidney with histologically normal nephrons though few in number [16]. Renal atrophy is characterized by shrinkage of kidney due to loss of nephrons. Several primary renal diseases and acute or chronic pyelonephritis may cause renal atrophy. Obstructive uropathy may cause a higher urinary pressure within the kidneys causing damage to nephrons [17]. The diminutive kidney is one weighing less than 100 grams. There are several causes: Hypoplasia in which the kidney is miniature or rudimentary at birth due to arrested development; Aplasia in which there is no true kidney, only remnants of parenchyma and vascular pedicle; Pyelonephritic atrophy resulting from infection and obstruction, in which atrophy due to nephrofibrosis usually takes place in a kidney of normal size at birth, although it may occur in a hypoplastic

kidney [18].

Multiple lobulations of kidney are witnessed throughout the fetal life [19]. Most of them disappear during the first year of birth but differing degrees of lobulations may persist in adult life. It is caused due to incomplete fusion of developing renal lobules [20].

Cystic diseases of kidney are heterogenous, comprising of hereditary, developmental and acquired disorders. They account for 6-8% on dialysis. Adult polycystic kidney disease (ADPKD) is a major cause of chronic kidney disease. ADPKD is an autosomal dominant disease with high penetrance and occurs in 1 out of 400 to 1000 persons and accounts for 5 to 10% of chronic renal failure [21].

Each kidney has only one ureter but there are cases where ureteral duplication or triplication can be seen that can be grouped under congenital anomalies of the kidney. The renal pelvis can also be found as duplicate or triplicate pelvis as an anatomical variant as that of ureter. Ureteral duplication may be incomplete or complete. Incomplete duplication of ureter is known as bifid ureter, said to be present if there are two separate ureters at the proximal aspect and they join at any point below ureteropelvic junction but before entering the bladder; whereas complete duplication is when there are two separate ureters that are continuous and enter the urinary bladder separately [20].

Most cases of CAKUT are sporadic and are limited to urinary tract but some of them are syndromic or associated with positive family history. To understand the basis of human renal anomalies, knowledge of kidney and urinary tract development is necessary [4].

Family histories revealed atleast one member with a known kidney or urinary tract disease in 50% and CAKUT in 22.9% of the families in asymptomatic first degree relatives of patients with CAKUT [22].

Considering the importance and rising incidence of renal malformations, we undertook a cadaveric study on 50 cadavers during routine dissectionin Grant Govt. Medical College, Mumbai, over a period of three years. Clinical studies on patients and uroradiological studies are available in literature but cadaveric studies are scarce. So our study could be a great help to clinicians, surgeons, radiologists, nephrologists to understand the possible variations that could be found during various procedures.

AIMS AND OBJECTIVES

1. To know the incidences of various types of congenital anomalies of kidney and ureter.
2. To study the anatomy of variations of kidney and ureter.
3. To understand the embryological and genetic basis of the variations of kidney and ureter.
4. To study the clinical significance of the variations found.
5. To correlate the findings of the present study with the findings of the previous workers.

MATERIALS AND METHODS

The present study was undertaken on 50 embalmed human cadavers allotted to MBBS students for routine dissection in Grant Govt. Medical College, Mumbai over a period of three years. Both male and female cadavers were included in the study. The cadavers were donated by relatives with consent and with death certificate. None of them had any pathological lesions, traumatic lesions or surgical procedures in the abdominal regions. These were the incidental findings during routine dissection of cadavers.

In the present study, amongst 50 cadavers 31 were male and 19 were female, --50 were right and 49 were left kidneys (one kidney was absent in a cadaver). As per the Cunningham's Manual of Practical Anatomy Volume-2 (Thorax and Abdomen)[23] anterior abdominal wall and abdominal cavity was opened. All the abdominal organs were removed for exposure of the posterior abdominal wall. Details of the position and external appearance of kidney in situ were noted. The right and left kidneys and the surrounding tissues were removed en bloc and the organ examined and studied grossly and as horizontal sections. The normal anatomy and the variations observed were studied in detail. The results were presented as percentage. The variations were analysed between male and female, right and left kidneys.

OBSERVATIONS AND RESULTS

The study of renal anomalies of kidney was an observational study in which 50 human cadavers (31 male and 19 female) were dissected in Grant Govt. Medical College, Mumbai. Observation was made on dissected specimens for the presence of any variations in the morphology of kidney and ureter. We exclusively studied the variations of kidney-proper and ureter. Following variations were studied:-

a) Morphological variations of kidney like renal agenesis, lobulated kidney, small or atrophic kidney, cystic kidney, fused kidney.

b) Variations in location like ectopic kidney or unascended kidney.

c) Variations in rotation like rotated kidney.

d) Variations in morphology of ureter like bifid pelvis and triplicate pelvis.

e) All the above anomalies were said to be present since birth, but the attempt was made to differentiate it from acquired deformities like congenital atrophic kidneys differentiated from acquired pyelonephritic atrophy.

f) A thorough study was done for the presence of any other associated anomalies.

Table 1: Variations found in Kidney and Ureter in Present Study.

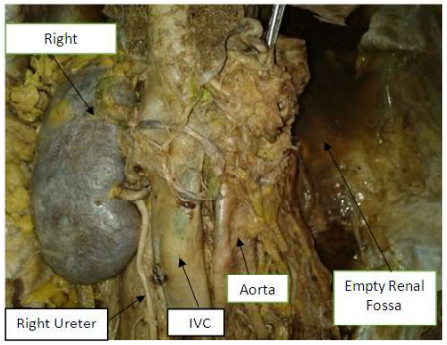
Total Variation Found		%
27/99		27.27%
Variation Found		**%**
Male	15/31	48.38%
Female	09/19	47.36%
Variation Found		**%**
Right	15/50	30%
Left	10/49	%

Variation	No	M	F	R	L	B/L	%
1.Renal agenesis	1	1	-	-	1	-	1.01
2.Fused kidney	1	-	1	-	-	-	1.01
3.Malrotated kidney	1	1	-	1	-	-	1.01
4.Unascended kidney	2	1	1	1	-	-	2.02
5.Atrophic kidney	5	3	1	2	3	1	5.05
6.Lobulated kidney	7	4	2	5	2	2	7.07
7.Cystic kidney	5	2	2	4	1	1	5.05
8.Bifid pelvis	2	2	-	1	1	-	2.02
9.Triplicate pelvis	3	1	2	1	2	-	3.03
Total	27	15	9	15	10	3	27.27

Abbreviations: M-Male; F-Female; R-Right; L-Left; B/L-Bilateral

Renal Agenesis: Unilateral absence of kidney was found in one cadaver, a male cadaver on left side.

Fig.1: Left Renal Agenesis.



Fused pelvic kidney: A fused unascended pelvic kidney was found, the ascent being limited by the aortic bifurcation. It was supplied by a single renal artey, coming from right common iliac artery; drained by a single renal vein, draining into left common iliac vein; and had a single ureter draining into urinary bladder at right vesico-ureteric junction. The hilum is anteriorly placed with arrangement of structures from above downwards as renal artery, renal vein and ureter.

Fig. 2: a. Unascended Fused Pelvic Kidney.

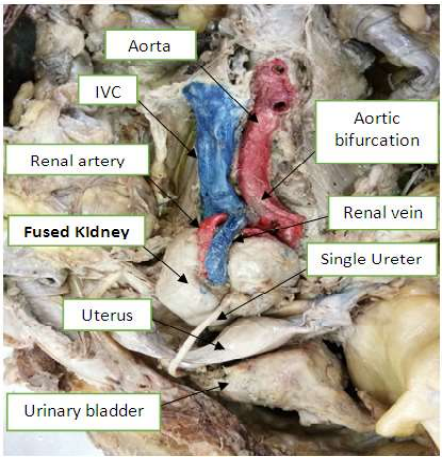
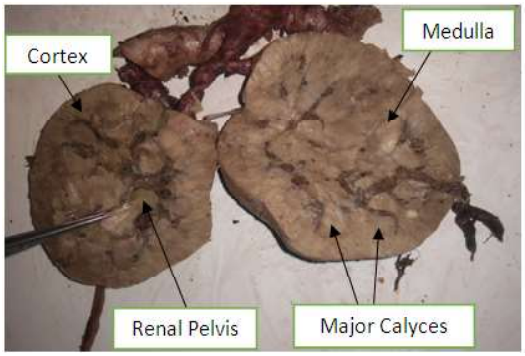
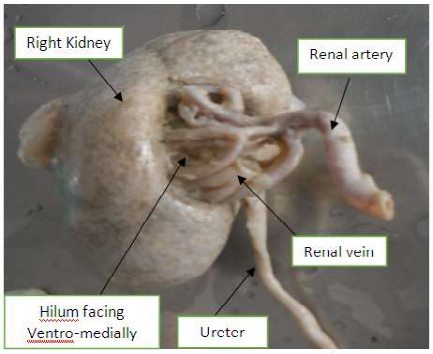


Fig. 2: b. Fused Pelvic Kidney cut section.



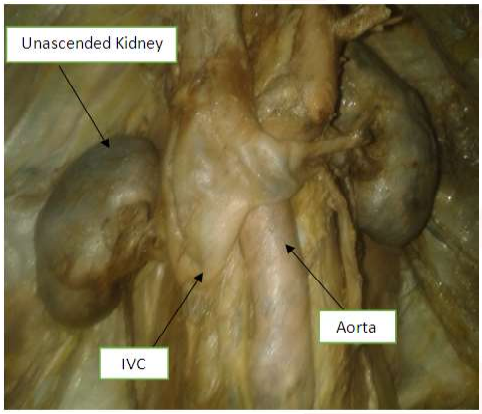
Malrotated kidney: An incomplete malrotation of kidney was found in one male cadaver on right side with hilum facing ventromedially.

Fig.3: Malrotation of kidney- Ventromedial.



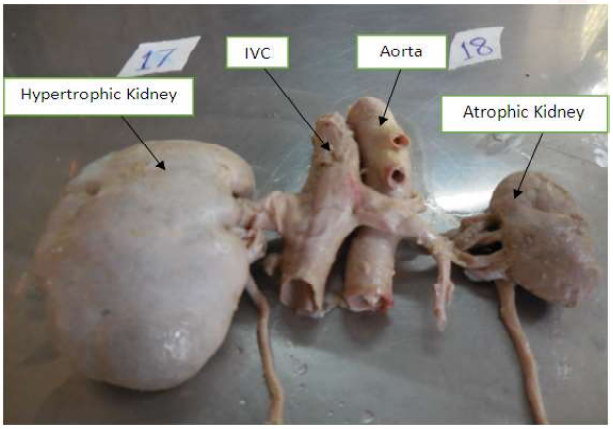
Unascended kidney: In our study we found 2 unascended kidneys, 1 pelvic fused kidney shown in figure 2, and another shown below. In this case, right kidney was placed from the lower border of L_2 to upper border of S_1 in a male cadaver.

Fig. 4: Unascended Kidney.



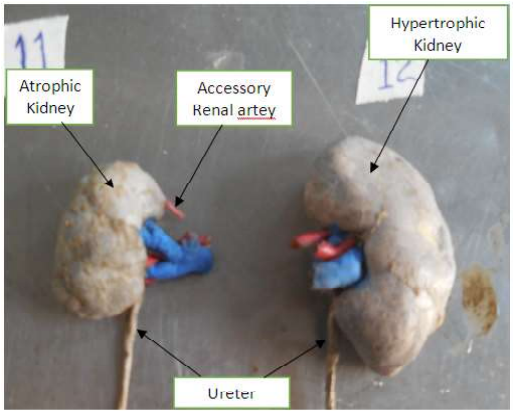
A) Atrophic kidney: In our study, we found 5 atrophic kidneys,4 in male cadavers and 1 in female cadaver, amongst which one male cadaver had bilateral atrophic kidneys. Atrophic kidneys were found 2 on right side and 3 on left side. In this case, left kidney is very small or atrophic weighing 16gms, as a result of which there is compensatory hypertrophy of fellow kidney.

Fig. 5: Left Atrophic Kidney with compensatory Right hypertrophy.



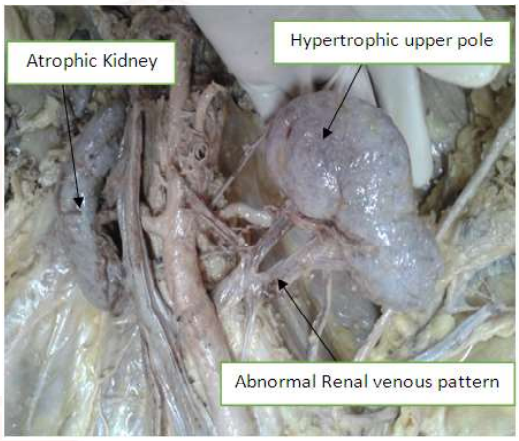
In this case, right kidney is atrophied weighing 35gms, with hypertrophic left kidney.

Fig. 6: Right Atrophic Kidney.



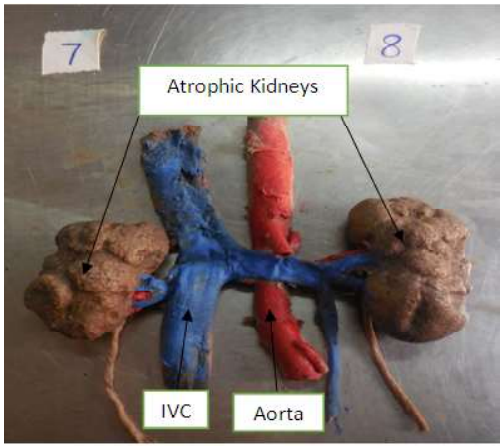
In this case, right kidney is present as a thin slender mass of parenchyma and left kidney had uneven hypertrophy of upper pole.

Fig. 7: Right Atrophic Kidney.



In this case, both kidneys are atrophied also has persistent fetal lobulations.

Fig. 8: Bilateral Atrophic Kidneys (with bilateral lobulations).



Lobulated kidneys: Total 7 kidneys were lobulated in our study, in 3 male cadavers, bilaterally in 2 male cadavers, unilaterally in 2 female cadavers, 5 on right side and 2 on left side. In this case, both kidneys had persistent

fetal lobulations with normal microscopic and functional characteristics.

Fig. 9: Bilateral Lobulated Kidneys.

Cystic kidneys: We found 5 cystic kidneys, 3 in males with one cadaver having bilateral cysts and 2 in females,4 right and 1 left. In this case, a huge cyst was present at the upper pole and a small cyst on the anterior aspect.

Fig. 10: a. Polycystic Kidney with a large cyst.

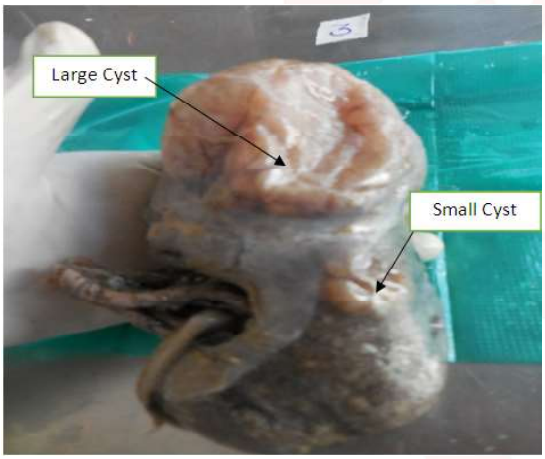
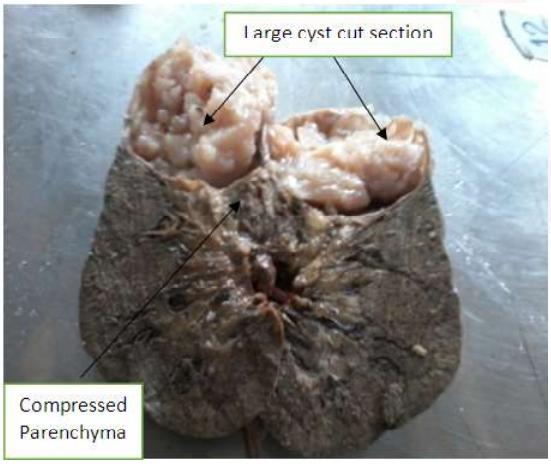


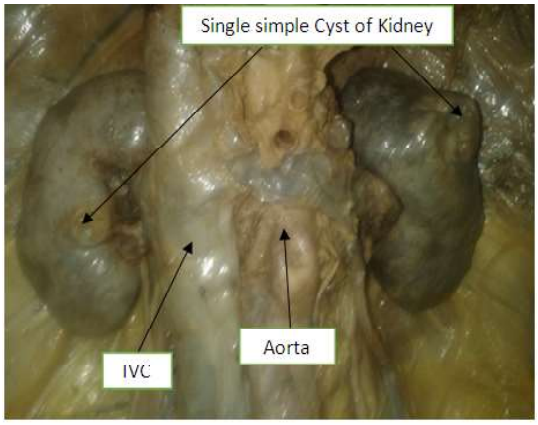
Fig.10: b. Large Cyst cut-section showing compression.



Of Parenchyma

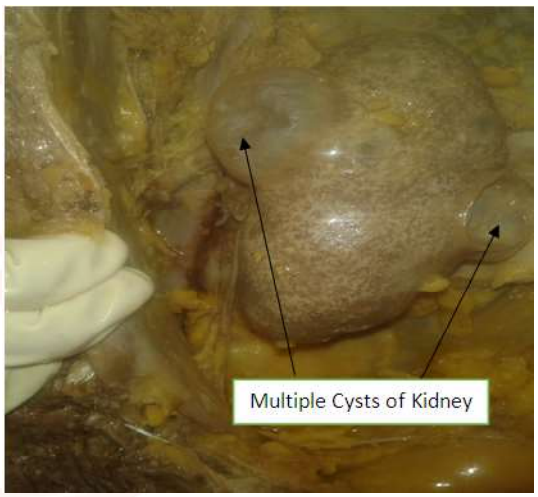
In this case, small cysts were present on both sides.

Fig.11: Single Simple Cyst of Kidney present bilaterally.



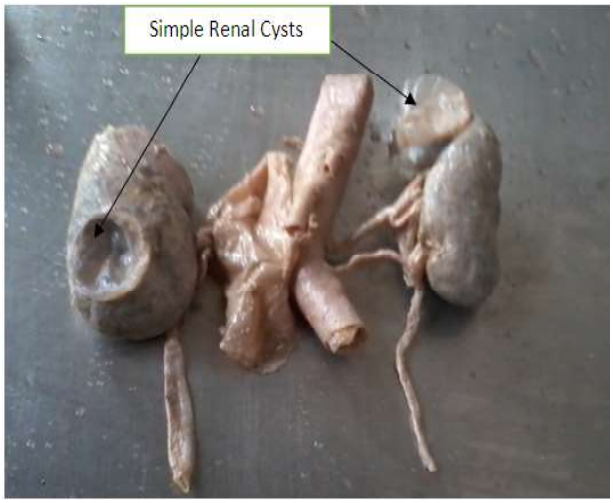
Multiple cysts present unilaterally on left kidney.

Fig. 12: Unilateral Multiple Cysts of Left Kidney.



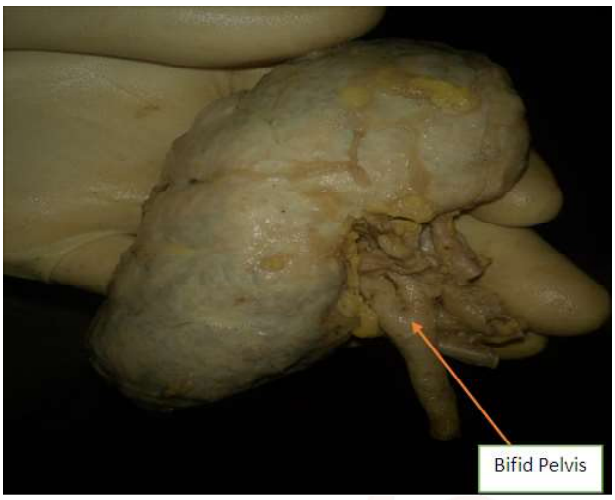
Bilateral simple renal cysts were present in this case with abnormal vasculature.

Fig. 13: Simple Renal Cyst present bilaterally.



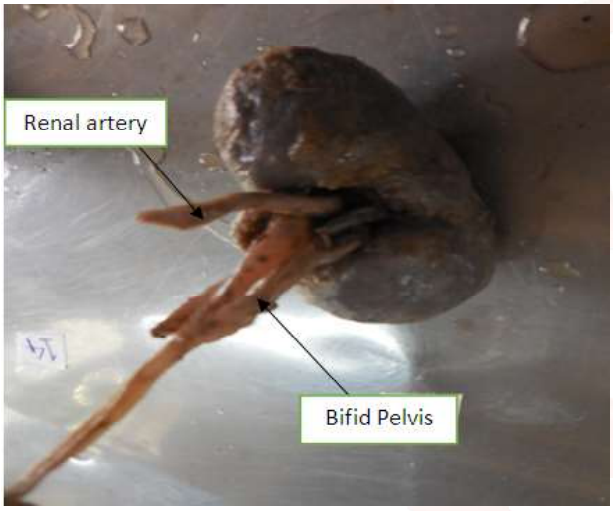
Bifid Pelvis: We found 2 bifid pelvis in 2 male cadavers, 1 on right side and other on left. In this case bifid pelvis was found in a male cadaver on right side which united just below the hilum.

Fig.14: Bifid Pelvis.



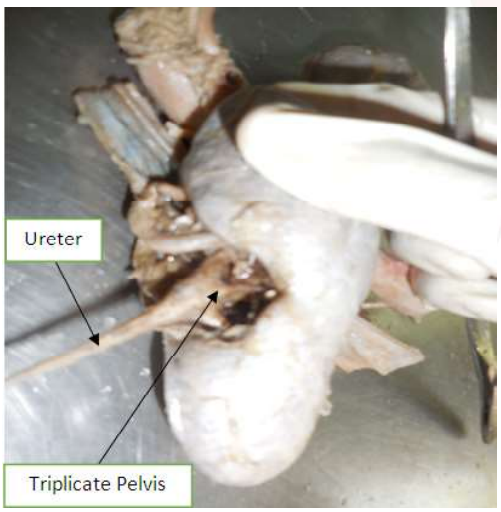
Left kidney is showing bifid pelvis uniting at the lower border of kidney.

Fig.15: Bifid Pelvis.



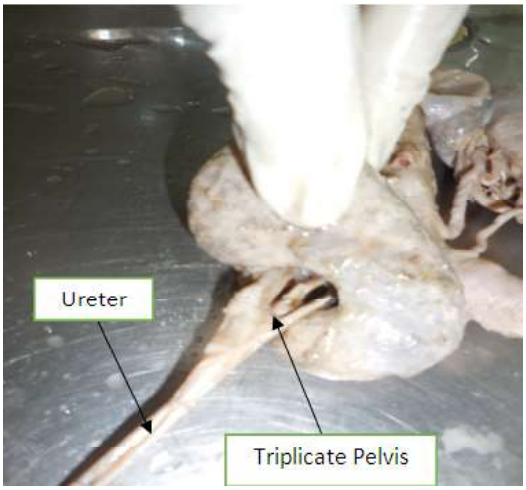
Triplicate Pelvis: We found 3 cases of triplicate pelvis unilaterally, 1 in male and 2 in female cadavers; 1 on right side and 2 on left side. In this case, triplicate pelvis is uniting as a single ureter at the lower border of hilum.

Fig.16: Triplicate Pelvis.



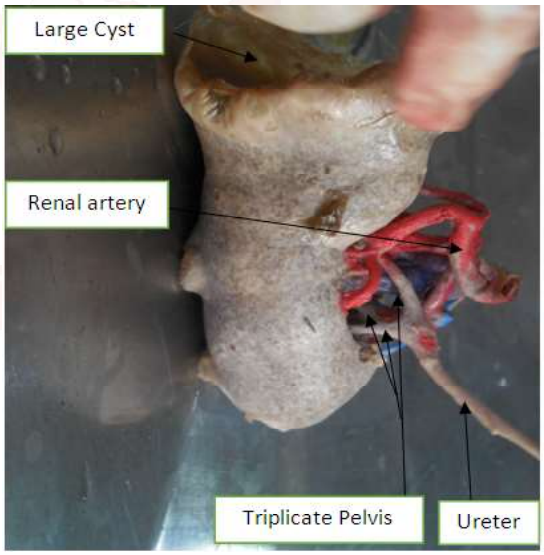
Triplicate pelvis uniting as a single ureter below the lower border of hilum.

Fig. 17: Triplicate Pelvis.



Triplicate pelvis uniting as a single ureter at the lower border of hilum. This case also shows a large renal cyst at upper pole and renal artery divided early as many segmental branches.

Fig.18: Triplicate Pelvis in a Cystic Kidney.



RESULTS

The total anatomical variations pertaining to kidney and ureter were found to be in 27 cases i.e. 27.27%, out of 99 kidneys, studied in 50 cadavers, as one cadaver had renal agenesis. Variations in male were found in 15 cadavers out of 31 i.e. 48.33% and that in female were found in 09 cadavers out of 19 i.e. 47.36%. This concludes that no significant difference was found in males and females, though some of the variations like renal agenesis, atrophic kidneys, lobulated kidneys, cystic kidneys, bifid pelvis were found to be more in male cadavers, which

is consistent with previous studies. Total variations found on right side i.e. 15 out of 50 kidneys (30%) were more than that on left side i.e. 10 out of 49 kidneys (20.41%). In our study we found, 01.01% of renal agenesis, 01.01% of fused pelvic or pancake kidney, 01.01% of malrotated kidney, 02.02% of unascended kidneys, 05.05% hypoplastic or atrophic kidneys, 07.07% lobulated kidneys, 05.05% % polycystic kidneys, 02.02% of bifid pelvis and 03.03% of triplicate pelvis.

DISCUSSION

There are multitude of congenital anomalies involving the kidney and ureter. Congenital disorders are conditions present since birth. About 15 to 20% of anomalies are due to chromosomal factors or single gene factors, 8 to 12% are said to be due to environmental factors, 25% are due to multifactorial inheritance, 40 to 60% are of unknown origin [24,25]. Congenital anomalies are seen in 2% of population as major abnormality. There are two types of abnormalities namely malformation, where growth disturbance occurs during embryogenesis and other is deformation, which is a late change that appears in a structure which was normal earlier [26].

Molecular Development of Renal system [27]:

Transcription factor 'WT1' produced by the mesenchyme of the metanephric blastemal helps in the epithelialization of ureteric bud. Production of Glial Derived Neurotrophic Factor (GDNF) and Hepatocyte Growth Factor (HCF) are regulated by WT1. Ureteric bud produces PAX2 and WNT4 that helps in epithelization of mesenchyme to form and differentiate into excretory tubules. The growth factor known as FGF2 and BMP7 stimulates the proliferation of mesenchyme and WT1 expression [28]. Agenesis of kidneys is failure of interaction between metanephric mesoderm and ureteric bud. It also occurs when there is mutation of genes that regulates the expression of signaling of GDNF1.

In our study we studied 50 cadavers and found a few variants of kidney and ureter:

Renal agenesis: Renal agenesis may be unilateral or bilateral. Unilateral renal agenesis (URA) is usually asymptomatic when it occurs as an isolated anomaly [7]. URA is 4 to 8 times more common than bilateral renal agenesis. Bilateral renal agenesis is a rare anomaly incompatible with life and approximately 20 to 36% of it presents as familial recurrence. It is 2.5 times more common in males than in females [29]. Renal agenesis is associated with other congenital anomalies like maldevelopment of Mullerian duct. Associated anomalies are more common in females as Mullerian system develops at a later stage in embryogenesis than Wolfian duct [30]. Adrian S Woolf et al [16] (2006) observed in his study that if solitary kidney is of normal size, it is either hypoplastic or dysplastic. According to him, a solitary functional kidney is always hypertrophied. Manisha S More et al [7] found URA in one cadaver and cited that in one-third to two-third of cases of URA, the opposite kidney has been found to be diseased, usually secondary to chronic pyelonephritis. Renal agenesis has been reported to be linked with Kallman's syndrome, Trisomy 21,22,7 etc. In present study URA is found in one kidney of left side with compensatory hypertrophy of existing kidney, which is in accordance with the findings of Adrian S Woolf[7] et al, Manisha S More et al [7].

Embryological basis [7]: Renal agenesis occurs when there is:-

1. Absence of metanephric blastemal
2. Maldevelopment of ureteric bud.
3. Lack of induction of metanephric blastemal by ureteric bud.

Genetic basis: Renal agenesis could be due to the absence of transcription factor WT1 that influences growth factor FGF-2 and BMP-7 to prevent apoptosis of metanephric cells or failure to convert metanephric cells into nephric epithelium by regulatory genes PAX2 and WNT4 from ureteric bud [31].

Fused Kidney: Renal fusion anomalies are defined as the congenital fusion of the kidneys in early embryonic period either partially or completely. Partial fusion anomalies include horseshoe kidney and crossed fused renal ectopia; and complete fusion represented by 'cake' kidney or fused pelvic kidney [9]. These renal anomalies exhibit abnormalities of position (ectopia), migration, rotation and vascular

supply. They occur more frequently in males [10]. Wilmer in 1938 was the first to describe the logical categorization of fusion anomalies of the kidney, while McDonald and McClellan in 1957 refined and expanded the classification given by Wilmer. There are two types of renal ectopia- simple renal ectopia and crossed renal ectopia. Crossed renal ectopia is again classified as crossed renal ectopia with fusion, crossed renal ectopia without fusion, solitary crossed renal ectopia, bilateral crossed renal ectopia.

Mechanism of fusion anomalies [32]: Several theories have been put forward to explain the anomaly. The Mechanical Theory proposes that during cephalad migration, the kidneys pass through the fork between the two umbilical arteries – and any positional change in these arteries squeeze the kidneys close together allowing their fusion (result in horseshoe kidney). Fusion of both nephrogenic blastemas with early arrested migration result in completely fused pelvic kidney. Abnormal position of umbilical artery can result in abnormal migration of of a renal unit to the contralateral side following the path of least resistance (crossed renal ectopia). The Theory of Abnormal Caudal Rotation proposes that fusion occurs due to lateral flexion and rotation of the caudal end of the embryo disturbing the relative position of the nephrogenic blastemal and ureteric bud [33]. [Cook WA, Stephens FD 1977]. The distal curled end of the vertebral column permit one ureter to cross the midline and enter the opposite nephrogenic blastemal or transplant the kidney and ureter to the opposite side during ascent. Association of scoliosis with crossed renal ectopia supports this theory. The Ureteral Theory states that cross over is strictly a ureteral phenomenon with the developing ureteral bud wandering to the opposite side and inducing the differentiation of the contralateral metanephric blastemal and it is assumed that that the metanephric tissue which does not receive a ureteric bud regresses. According to Teratogenic Theory, horseshoe kidney (HSK) results from abnormal migration of posterior nephrogenic cells due to teratogenic insult forming a parenchymal isthmus [34,35].

The increased incidence of malignancies and other organ system anomalies associated with HSK possibly supports this theory[36]. Finally Genetic Theory suggests that genetic influence may play a role because some renal fusion anomalies have been reported to occur in identical twins and siblings within the same family. It is suggested that the sonic hedgehog gene signal is critical for kidney positioning along the mediolateral axis and its disruption will result in renal fusion[37]. [Shapiro E et al]. McPherson suggested that HSK may occur as a previously undescribed autosomal dominant condition[38]. Analysis of patients with Turner's syndrome revealed that 33% of patients presented some renal malformations with HSK occurring in 7.1% of these patients, which renders support to the Genetic Theory [39].

Incidence: Renal fusion anomalies are reported in 0.1 to 0.2% of all live births [11]. Horseshoe kidney is the most common renal fusion anomaly found more commonly in men than in women with a ratio of 2:1. It accounts for 90% of all fusion anomalies and occurs in about 0.25% of the population [9,40]. Crossed fused renal ectopia (CRFE) is the second most common renal fusion anomaly with an estimated incidence of 1:1300 to 1:7500 [11,10]. It is both a fusion and ectopic anomaly and occurs in about 0.08 to 0.01% case. The prevalence was estimated to be 1 in 1000 live births[41]. Cake kidneys or fused pelvic kidney is a very rare congenital anomaly with a few more than 20 cases described in the literature [11,42]. Cake kidney accounts for only about 2% of all fused kidney types. The estimated incidence is 1/65000 to 1/375000 cases [43]. It is more common in males with a male to female ratio of about 2-3:1 [40].

Embryological Basis: The embryological development of the kidney results from the interaction between the mesonephric duct derived ureteric bud, and the metanephros, the caudal most part of the nephrogenic cord. Development begins early in the fourth week of gestation and during the sixth and eighth weeks, the lobulated embryonic kidneys ascend from the pelvic region upwards along the posterior abdominal wall to their normal position and undergo a 90^0 axial rotation from horizontal to medial. During the process of ascent from the

pelvis, the kidneys derive their blood supply sequentially from vessels that are closest to them; initially median sacral, then common iliac and inferior mesenteric and finally the abdominal aorta. Anomalies can occur due to abnormalities of development, migration and rotation. An ectopic kidney results from incomplete, excess or abnormal ascent. If during the process of ascent, the kidneys come into contact, horseshoe kidney or crossed renal ectopia will result [44].

Clinical Significance: Many fusion anomalies remain asymptomatic and are incidentally detected at autopsy, surgery or radiological investigations. Less frequently they may be associated with anomalies of skeletal, cardiovascular, genitourinary and gastrointestinal systems [39]. [Kaufman MH]. Presence of such renal fusion anomalies poses difficulties and complications during abdominal aortic aneurysm surgery, retroperitoneal and pelvic surgeries, renal transplantation and interventional procedures [32].

Cake Kidney or Fused Pelvic Kidney: Cake kidney is a very rare congenital anomaly used to describe completely fused renal mass located in pelvic cavity and drained generally by two ureters, which do not cross the midline. Very rarely, a single ureter is found draining the cake kidney. This anomaly also known as 'pancake', 'lump' or 'disc' kidney shold not be confused with with crossed fused renal ectopia (CRFE) type of 'lump' or 'disc' kidney, in which the ureter of the ectopic kidney crosses the midline. Moreover, CFRE type of 'lump' kidney is generally located at a higher level lying on one side of the midline. Developmentally, when the renal analgens fail to ascend and remain in the pelvic cavity extensively fusing with each other, a cake kidney is formed retaining the primitive vascular supply. Vascular supply may be derived from a single renal artery (from distal aorta or common iliac) and a single renal vein (draining into IVC or common iliac vein). The single renal vascular supply of cake kidney is at increased risk of damage by pelvic trauma, pregnancy or space occupying lesions [45].

Clinical Significance: Generally cake kidney remains asymptomatic and may be detected at any age. This condition may be present associated with other congenital anomalies like anomalies of testicular descent, anomalies of vas deferens, vaginal agenesis, bicornuate or unicornuate uterus, sacral agenesis, caudal regression syndrome, tetralogy of Fallot, and Spina bifida [46-48].

Pancake kidney exposes to urinary infections and renal calculi probably due to rotational abnormalities and short length of ureter, which favour obstruction and stasis. Various imaging modalities used to investigate renal fusion anomalies include sonography, intravenous pyelography, computed tomography, renal scintigraphy, MDCT angiography and MRI. It is described in the literature that upon surgery of a pancake kidney, division of the parenchyma presents potential problems such as renal vascular damage and postoperative renal failure [49]. In the literature, most cases describe a conservative management of incidentally diagnosed pancake kidney [40,50,51].

Looney and Duke (1926) were first to describe the pancake kidney [52]. Alok Kumar Tiwari et al (2014) [50] reported a case of pancake kidney detected incidentally while treating a female patient for urinary tract infection. They revealed a fused mass situated in the pelvic cavity with a short course uncrossed ureters opening separately into the urinary bladder. Jolio Slongo, Lucas R Weigand (2017) [53] presented a case of a 28 year old male with symptomatic obstruction of a non-functioning moiety of a pancake kidney with right hydroureter and calculus at right uretero-vesicle junction. In present study, we found a case of fused pelvic or pancake kidney just below aortic bifurcation, which is the arresting the ascent of kidney. Hilum is placed anteriorly with renal artery above, renal vein below, and a single ureter emerging further inferiorly. This pancake kidney is supplied by a single renal artery arising from right common iliac artery. The renal artery is further dividing into two, which is separately draining into right and left half of the pancake kidney.a single renal vein is draining the pancake kidney into left common iliac vein. A single ureter arising from the hilum anteriorly, below renal artery and vein, is draining into the urinary bladder at right vesico-ureteric opening, which is very rare, else two ureters are present draining separately. In

the literature, most cases are reported in males, but in our case, it was found in a female cadaver incidentally with no associated anomaly.

Malrotated kidneys: Rotational anomaly of kidney is a rare entity [12,54] and have been cited in a very few of the embryology text books. Though rare this type of anomaly has wide implications in the context of advanced surgical procedures and diagnostic evaluation of kidney donors. Anomalies of renal rotation are associated with renal ectopia and the fusional abnormalities but may be exhibited by the kidneys which are otherwise normally placed [13].

Embryological Basis: During ascent, the hilum of the kidney is directed ventrally. On reaching the definitive position it undergoes 90^0 medial rotation around the vertical axis such that the hilum directs medially [55]. The exact cause of malrotation of the kidneys is not known but it is believed that malrotation occurs due to abnormal insertion of ureteric bud into an abnormal region of metanephros. When associated with renal ectopia, malrotation probably occurs due to incomplete medial rotation brought about by the aberrant vessels [14]. Four types of rotational anomalies have been identified [56,57]:

-In non-rotation, the renal pelvis presents itself ventrally in relation to the kidney mass.

-In incomplete rotation, it presents itself ventromedially.

-In complete rotation, the renal pelvis presents itself laterally.

-In the more rare severe and excessive rotation, the renal pelvis presents itself in a position depending upon the number of degrees through which rotation has occurred [58].

This process occurs during the ascent of kidneys which occurs between 38 to 49 days of development. Renal rotation takes place before definitive vascularization [59]. Rotational anomalies are often caused by or related to aberrant renal vessels [54,56,60].

Clinical Significance: Rotational anomaly though rare, has important implications from surgical point of view, as it may be mistaken for some more serious condition on IVP. It may erroneously be attributed to displacement by para vertebral mass. Pelvis and calyces may look peculiar though normal. Commonly lower pole causes deviation of the course of the crossing ureter. Usually such deviation is anterior and lateral sometimes creating an impression that lower pole mass is present. It assumes a great importance in the context of present day surgical procedures like percutaneous nephrectomy and preoperative diagnostic evaluation of the kidney donors etc.

Though the association of malrotation of kidney and anomalies of renal vessels have frequently been quoted, Das and Amar (1984) [12] in the management of their 27 patients of ureteropelvic junction obstruction with associated renal anomalies observed only one case of malrotation. Ingole IV, Ghosh SK [13] found a right unascended and laterally rotated kidney with hilum facing completely laterally at the level between 3^{rd} and 4^{th} lumbar vertebra. Dr. Sushil Kumar et al [14] studied a case in which they reported a right kidney extending from lower border of L_2 to upper border of S_1 with hilum directed ventrally at the level of body of L_4. In present case, amongst 50 cadavers we found one case of malrotated right kidney extending from L_2 to S_1 with hilum facing ventro-medially at the level of L_3-L_4 interphase.

Unascended or Ectopic Kidney: Kidneys are situated behind the peritoneum on either side of vertebral column, extending from upper border of T_{12} to L_3. Right kidney is slightly lower in position than the left due to presence of liver [61]. Failure of the kidney to ascend into the renal fossa in utero results in ectopic kidney. Such kidney is often found in the pelvis, however it may be placed higher up in lower lumbar region. Generally defects in ascent of kidney are associated with variations in branching pattern of abdominal aorta [14]. Ectopic kidney has a reported frequency of 1:500 to 1:110; ectopic thoracic kidney 1:1300; solitary ectopic kidney 1:1000; solitary pelvic kidney 1:22000; one normal and one pelvic kidney 1:3000; and crossed renal ectopia 1:7000 [62]. Unilateral renal ectopia is most frequent with incidence ranging from 1:1000 to 1:300 births [62,63].

Embryological Basis: At first the permanent kidney derived from metanephros lies in the sacral region. Later due to differentiated growth of posterior abdominal wall and reduction in the

flexed curvature of the fetus, the kidneys ascend to the thoracolumbar region. During ascent, the kidneys pass through the arterial fork formed by umbilical arteries and one of them may fail to ascend because of the obstruction of sickle shaped fold of peritoneum projecting from the lateral pelvic wall containing the umbilical artery [14]. During embryological ascent, the metanephros derives its blood supply from median sacral artery initially followed by common iliac and inferior mesenteric and lastly from aorta. During the ascent, the former vessels degenerate. Failure of degeneration of these vessels results in origin of accessory renal arteries [64,655].

Clinical Significance: Kidneys in ectopic position are dysplastic and often non-functional. They may go undetected in life and get noticed only after death either in autopsy or during dissection. Often they are diagnosed for presence of a pelvic mass or on pyelogram. Ectopic or congenital unascended kidney has to be carefully differentiated from acquired nephroptosis where the length of the ureter is normal. Symptoms due to ectopic kidney may vary from none to pain, hydronephrosis, pyelonephritis, renosigmoid fistulae or lithiasis [66]. In case of females, the pelvic kidney may result in obstetric complications [67]. [Banner 1965]. Since it has an atypical location, ectopic kidney is more prone to trauma and may cause hydronephrosis due to obstruction of flow through the ureter, most common being at pelvi-ureteric junction. Since intravenous pyelography may erroneously mistaken an ectopic kidney for paravertebral mass, CT and MRI are best diagnostic tools for ectopic kidney [68,63].

Cases of ectopic kidney, unilateral or bilateral have been reported in the literature regularly [58,64,66,69]. Belsare SM et al [65] studied a case where kidney was not entirely pelvic, its upper pole being at the level of L_5 due to halt in its ascent during development. They also noted a series of other anomalies associated like spleen without renal impression, sigmoid colon and mesocolon pushed to right side, ovary found in rectouterine pouch. They also cited that unilateral ectopic kidney is commoner than bilateral. It is also found that congenital pelvic kidney is commoner on left side than on right. In their case, ectopia was unilateral and on left side in accordance with the findings of others. The frequency is quoted to be higher in males than in females. But they found it in a female cadaver. Meril Ann Soman et al [70] reported an unascended right kidney lower down anterior to the bodies of L_5, S_1, S_2, S_3 vertebrae. Dr.Sushil Kumar et al [14] reported a case of right kidney extending lower border of L_2 to upper border of S_1 with hilum directed ventrally at the level of body of L_4. There were multiple renal arteries seen arising from the abdominal aorta and multiple renal veins draining into inferior vena cava. In present study, we found two unascended kidneys: one case of right kidney extending from the lower border of L_2 to upper border of S_1 in a male cadaver, which is in accordance with the findings of previous studies to be commoner in males, unilaterally, and on right side. In another case, kidney was found to be pelvic in location, below the bifurcation of abdominal aorta and was fused to the other kidney, which is discussed later in detail under fused kidney.

Hypoplastic or atrophic kidney: The diminutive kidney is one weighing less than 100 gms occurs frequently enough to present a problem of diagnosis and treatment both to general practitioners and urologists. There are several causes: Hypoplasia, in which the kidney is miniature or rudimentary at birth due to arrested development; Aplasia, in which there is no true kidney, only remnants of parenchyma and vascular pedicle; Pyelonephritic atrophy results from infection or obstruction and atrophy due to nephrofibrosis usually takes place in a kidney of normal size at birth although may occur also in a hypoplastic kidney [18]. Hypoplasia usually occurs due to inadequate ureteral bud branching and results in a small kidney with histologically normal nephrons, though few in number [71].

Pabbati Raji Reddy et al [71] found one case of renal hypoplasia among 50 cadavers studied. Rubinstein et al [72] found an incidence of 2.5% of true hypoplasia. Ramzan Davran et al [17] studied 2417 cases, amongst which 1.3% cases had left renal atrophy and 0.2% cases had right renal atrophy. They concluded that left renal atrophy may be significantly higher than the right side. In present study, we found 5 atrophic

kidneys, one left weighing 16 kgs other right weighing 35 kgs with both cadavers showing compensatory hypertrophy of fellow kidney, which is in accordance with the study done by Geraghty and Plaggemeyer [73] in 1913, and Charles Pierre Mathe [18] in 1956.

Aortic pressure induced flow disorders in the left renal vein, structural anomalies of left renal vein including Nutcracker syndrome, passage behind the aorta and possibly the higher arterial pressure of the left kidney due to shorter distance from the heart as an underlying etiology of endothelial damage and atherosclerosis may be some of the possible causes [17]. As a rule only one kidney is hypoplastic and since compensatory hypertrophy is present in the other, nephrectomy can be done if necessary for the relief of pain, chronic infection and hypertension. The function of hypoplastic kidney alone is inadequate to eliminate the waste products of the body, should the other be removed , the patient would die of uremia [18]. In 1913, Gerraghty and Plaggemeyer [73] advised that separate phenosulfonphthalein function studies be carried out on each kidney. Although the appearance time of the dye may be normal, the hypoplastic kidney usually eliminates one-quarter to one-fifth the amount of that secreted by its hypertrophied mate. A hypoplastic kidney is prone to infection and stone formation, often scarring with fibrosis, cystic degeneration and round cell infiltration are present. Other congenital anomalies of the genitourinary tract frequently are associated with renal hypoplasia. Hypoplasia is to be differentiated from pyelonephritic atrophy in which the kidney is reduced in size by infection [18].

Embryological basis: The kidney develops from the wolffian body through the stages of pronephros, mesonephros and metanephros. An insufficient blood supply occurring at any of these stages could cause arrest in the development [74,75].

Clinical significance: Congenital hypoplasia should be differentiated from atrophic pyelonephritis.in atrophic pyelonephritis, there is reduction in the size of the kidneys, width of the cortex is reduced and the cortical and medullary markings are obliterated, the pyramids are white and show less defined radial striation, the pelvis may retain its normal size but usually is enlarged [18].

Lobulated kidney: It is the result of fetal lobulation that persists into adulthood. Typically, the fetal kidneys are subdivided into lobes by grooves that disappear by the end of the fetal period. It occurs due to incomplete fusion of developing renal lobules. It is discovered incidentally and carries no clinical significance but during imaging it is important to distinguish between lobulation and scarring, which can occur reflux or chronic infection. Lobulation can be seen on CT or USG as indentations that occur between the medullary pyramids, compared with renal scars, which are located overlying the medullary pyramids [7].

Embryological basis: Embryologically, the kidney develops in several distinct lobules that fuse as they grow. Incomplete fusion of these renal lobules can persist postnatally and may be observed in 7% of adults. After 28th week of gestation, varying degrees of assimilation of independent 14 renal lobes occur (8-16). Normally this lobulated structures of kidney remains apparent at birth and it gradually disappears during infancy as the nephrons increase and grow and fully disappear over the first 5 years of life as kidney grows [64].

Clinical significance: It is recognized incidentally on imaging studies as smooth regular indentations in the renal contour without parenchymal thinning or abnormalities in the underlying calyx. Often this is a normal variation but should be distinguished from inflammatory scarring of kidney, renal infarcts and tumours. It can mimic as a pseudotumour [7]. [Manisha S More et al]. It can be distinguished from other causes of of irregularities of outline by its symmetry, shape and absence of any deformity of underlying calyces. A cortical defect opposite a calyx represents pathological loss of lobar tissue. On IVU, fetal lobulations appear as smooth regular indentations in between the renal calices without parenchymal thinning or abnormalities in the underlying calyx. Whereas in vesicoureteric reflux, the scars occur over calyces which are abnormally clubbed. Inflammatory scars are deeper and typically associated with an abnormal underlying calyx. Renal infarcts are generally random in distribution and cause a broad

flat depression in the outline [7]. Patil et al [76] reported a rare congenital condition of kidney where bilateral lobulation and malrotation were were observed in association with the open hilar structure of kidney. Manisha S More et al [7] found unilateral lobulated kidney in 6 cadavers. According to them, lobulations can be seen in 5% of right kidney and 10% of left kidney. Choudhary U et al [77] found 6.25% lobulation in left kidney and 3.12% showed bilateral lobulations. In our study 7 kidneys out of 99 were lobulated, out of which 3 cadavers had bilateral lobulations and 1 had unilateral lobulation in left kidney.

Cystic Kidney: Polycystic kidney disease is a genetic disorder characterized by thhe growth of numerous cysts in the kidney. Cysts can profoundly enlarge the kidneys while replacing much of the normal structure, resulting in reduced kidney function and leading to kidney failure [78]. Multiple cystic disease has has an incidence of 1 per 400-1000 persons among whites and accounts for 8-10% of all cases of end stage renal disease. Simple cysts are the most common cystic renal lesions present in 5% of general population increasing in frequency to 25-33% of patients older than 50 years and account for 65-70% of renal masses [79].

Formation of Cyst: Cysts arise anywhere along the nephron, they may be 3-4cm in diameter and may compress the adjacent parenchyma. In late disease interstitial inflammation and fibrosis occurs. The primary cilium in tubular epithelial cells function as a mechanosensor to monitor changes in fluid flow and shear stress. These sensors regulate ion flux in response to external forces. Mutated proteins affect second messengers and influence proliferation, apoptosis, extracellular matrix interactions and secretory function, leading to formation of tubular cysts [80].

Genetic Basis: Adult polycystic kidney disease (ADPKD) is an autosomal dominant disease with high penetrance caused by mutations in one of the two genes, PKD1 and PKD2. 85% of cases are due to mutation in PKD1 (chromosome 16p13.3). PKD1 encodes polycystin 1, a large (460 KD) protein that localizes to tubular epithelial cells and has domains that are usually involved in cell-cell and cell-matrix interactions. PKD2 encodes polycystin 2, a cation channel, mutations of which disrupts the regulation of intercellular calcium. Polycystin 1 and 2 are both localized to the primary calcium. They form a complex that regulates intracellular calcium in response to fluid flow [21].

Embryological Basis: Definitive human kidney is derived from two sources. Collecting part of kidney is derived from the ureteric bud, which arises from the caudal part of mesonephric duct and secretory part is derived from metanephros-metanephric blastema [64]. Failure of the ureteric bud to integrate and branch appropriately into the metanephros during development of kidney resulted in multiple cystic disease of kidney [81]. Eswari AK et al [82] discussed and concluded that time of formation of cysts is not known but tubular and glomerular microcysts are seen as early as 12 weeks of gestation.

Cysts could be developmental or inherited or acquired. Factors responsible for cyst formation are [83] - Increased production of cells lining the cyst wall.

-Increased production of fluid by the cells.

-Abnormal basement membrane structure and function.

Clinical Significance:

The major extrarenal complications of ADPKD include cerebral aneurysms, hepatic cysts, pancreatic cysts, cardiac valve disease, colonic diverticula and aortic root dilatation [82]. Autosomal dominant PKD occurs in both children and adults. Symptoms often do not appear until middle age. An autosomal recessive form of PKD appears in infancy or childhood. It is less common but it tends to be very serious and gets worse quickly. It can cause serious lung and liver disease, end-stage kidney disease, and it usually causes death in infancy or childhood [84]. Acquired cysts are common in older persons. Multiple cysts may be seen in association with potassium deficiency, congenital disorders, metabolic diseases and toxic renal injury. Acquired polycystic disease occurs in the setting of chronic progressive renal scarring due to diabetes mellitus, chronic glomerulonephritis, or other renal disorders that lead to azotemia. It is seen most commonly in patients undergoing dialysis and is discovered inciden-

tally in most instances. The nephrons that survive the underlying renal disease are stimulated to grow and accumulate abnormal amounts of fluid. In contrast to patients with ADPKD and ARPKD, those with acquired cystic kidney disease are more likely to have solitary or multicentric adenocarcinomas [85].

In a study undertaken by Jaswinder Kaur, 2013 [86], out of 30 cadavers, multiple cysts were seen in 4(13.3%) cases in male cadavers. The cysts were seen more on right kidney as compared to left. Eswari AK et al (2015) [82] observed multiple cysts on the surface of both the kidneys. Sagittal section of the kidney also revealed the presence of cysts. Histological section showed the presence of cysts of various shapes and size close to the cortex. They concluded that in ADPKD, cyst can develop from any part of the nephron [87].

In a study conducted by J Athiban Raj et al [88], out of 10 cadavers 1 had polycystic kidney, which signifies that over 10% of the normal person have polycystic kidney and mainly occurs in old aged persons. Kaur Manpreet et al [89] observed and reported that multiple cysts were present only on left side. They concluded that unilateral polycystic kidney is seen not so frequently. They found unilateral cystic kidney in one male cadaver. Sathialakshmi V et al [90] studied diseases in various organs and reported bilateral polycystic kidney in a male cadaver. Dr. Shroff Gautam A et al [81] studied ultrasonography of patients between 25-29 weeks gestation for kidney development. They found unilateral multicystic dysplastic left kidney, right hypoplastic kidney with left lobulated kidney, right kidney agenesis and left compensatory enlarged kidney with cystic mass. Adrian S Woolf et al [16] have described three types of diseases associated with glomerular cysts. One of them is urinary obstruction. The author cited studies which described that normal glomeruli can become cystic after birth. The Bowman's space becomes enlarged after collapse of glomerular tufts secondary to glomerulosclerosis or mesangiolysis or hemolytic uremic syndrome. When cysts form in the kidney, they are filled with fluid. Cysts can enlarge in the kidneys while replacing much of the normal structure, resulting in reduced kidney function leading to kidney failure[78]. It was reported that cystic kidneys were slightly more severe and common in males than in females[91]. Sujatha K et al[21] found cystic kidneys in 7 cadavers out of 30 and observed that cysts were present in male cadavers more than in females. Number of cysts were more in right than in left kidneys. In present study, we found cystic kidneys in 4 cadavers out of 50 with 5 cystic kidneys, bilaterally in one cadaver, 4 on right side and 1 on left side. This is in accordance with the previous studies, being more common in males and on right side.

Ureteral Duplication: Ureteral duplication may be incomplete or complete. If there are two separate pyelocalyceal systems and they join at the ureteropelvic junction (UPJ), it is considered a bifid pelvis. If there are two separate ureters at the proximal aspect and they join at any point below UPJ but before entering into the urinary bladder, it is considered bifid ureter. Complete ureteral duplication is when there are two separate ureters that continue and enter the urinary bladder separately. Double ureter is more common in girls than in boys in a ratio of 6:1 [5].

Embryological Basis: Incomplete duplication of the ureter may be due to some fallacy or disturbance in development of ureteric bud which arises from mesonephric duct. Duplication of the ureter is a consequence of early splitting of the ureteric bud. Splitting may be partial or complete, and metanephric tissue may be divided into two parts, each with its own renal pelvis and ureter. One of the buds generally has a normal position, whereas the abnormal bud moves down together with the mesonephric duct. Thus it has a lower abnormal entrance into the bladder, urethra, vagina or epididymal region [31,64].

Clinical Significance: Girls may have urinary incontinence when the ectopic ureter opens in the urethra distal to the sphincter. A duplex system can be associated with other renal complications such as obstruction, reflux and infection. If the obstruction is maintained for some time, the kidney can become hydronephrotic. When the infection becomes persistent, it can also lead to severe chronic pyelonephritis, which ultimately produces chronic renal disease [5]. According to Lowsly et al [92], out of 4215

cadavers studied, 18 showed duplication of ureter. Out of 18, 7 had unilateral incomplete duplication, 2 were bilaterally incomplete and 8 had unilateral complete duplication [92]. Russel et al showed an average of 3% showing ureteral duplication [93]. Duplex systems are the most commonly encountered congenital abnormalities of the renal tract, with a reported incidence of 0.8%. Asakawa M et al [94] reported 5 cases of double pelvis and ureter among 340 cadavers (1.47%, 1.8% Right, 0.3% Left). Standring S et al [95] has described the incidence of unilateral bifid as 1 in 1251. Choudhary U et al [77] found 2 out of 32 cadavers with unilateral incomplete duplication (6.25%, 3.12%Right, 3.12%Left). In our study, we found 2 bifid pelvis on 2 separate male cadavers, 1 on right side and 1 on left side.

R Scott [96] in 1970 reported a case of triplication of the upper part of the left ureter, which united at the transverse process of L_5. Varma Kakarlapudi [97] in 2011 reported a rare observation of presence of 3 accessory renal arteries, a duplicate renal pelvis of the right kidney, and a triplicate renal pelvis of the left kidney in a 60 year old male cadaver. In the present study, out of 50 cadavers we found 3 cases of triplicate pelvis, a very rare finding, 1 in male cadaver on left side and 2 in female cadavers both on right side.

CONCLUSION

Renal anomalies are one of the common congenital anomalies inherited by the offspring. Many times it may remain unnoticed till adulthood. Studies have isolated the gene responsible for the heredity of congenital malformations of the kidneys [71]. These anomalies of renal system have profound embryological and clinical importance. It gives lot of information regarding variations in the development of kidneys to the Anatomists and Clinicians. These anomalies can be detected through ultrasound and other latest investigating tools [27]. Treatment is mainly based on the functional capacity of the kidney; nephrectomy being done on non-functional kidneys and corrective procedures forming the mainline of treatment for the functional kidneys [65]. A greater number of such studies should be conducted to enhance the diagnosis and precision of surgery in this region and limit the avoidable complications arising out of these variations [20].

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