

Congenital Renal Anomalies in Indian Population

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ABSTRACT

The congenital anomalies of kidney are not uncommon. The incidence of renal anomalies varies from 1:400 for horseshoe kidney to 1:10,000 for bilateral renal agenesis. The most of this available data is from western populations. Little or no data is available on incidence of renal anomalies in Indian population. Our objective was to determine the incidence of congenital renal anomalies in Indian population. A retrospective analysis of observations made from 1,900 consecutive autopsies for presence of congenital renal anomalies was done. In 1,900 autopsies, there were nine cases of congenitally anomalous kidneys (42%). These anomalies included three cases of horseshoe kidney (16%), two cases of extrarenal calyces (12%), and one each of renal ectopia with fusion, trilobar kidney, polycystic kidney and unilateral renal agenesis. The incidence of renal anomalies in Indian population is relatively less as compared to their western counterparts.

Keywords: Horseshoe kidney, Incidence, Renal anomalies.

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INTRODUCTION

The congenital anomalies of kidney are not uncommon. Approximately, 30% of the children born with congenital anomalies have urogenital anomalies.¹ Some of the renal anomalies remain asymptomatic throughout the life without altering the lifespan of subjects, while others are associated with considerable morbidity and early death. The most common anomalies reported are horseshoe kidney, polycystic kidney, renal ectopia, unilateral renal agenesis, crossed ectopia with fusion, and bilateral renal agenesis, etc. The incidence of renal anomalies has been reported based on autopsy studies^{2,3} and radiological data.⁴ The incidence of renal anomalies varies from 1:400 for horseshoe kidney² to 1:10,000 for bilateral renal

agenesis.⁵ Most of this available data is from western populations. Little or no data is available on incidence of renal anomalies in Indian population. The aim of our study is to determine the prevalence of renal anomalies in north-western Indian population.

MATERIALS AND METHODS

This is a retrospective study based on data from 1,900 consecutive medicolegal autopsies carried out in our institute over period of last two decades. These autopsies were conducted on patients died of road traffic accidents, trauma, homicidal poisoning, etc. These patients hailed from north-western part of Indian subcontinent. The ages of the deceased varied from 2 to 100 years. They were apparently normal before having had met with an accident. In none of the cases reported below, the cause of death was the anomalous kidney they carried.

OBSERVATIONS

In 1,900 autopsies, we came across nine cases of congenitally anomalous kidneys (47%). These anomalies included three cases of horseshoe kidney (16%), two cases of extrarenal calyces and one each of renal ectopia with fusion, trilobar kidney, polycystic kidney, a pair of hyperblastic and hypoblastic kidneys, and unilateral renal agenesis.

In all of three horseshoe kidneys (Figs 1 to 3), isthmus was connected with lower poles. In two of the cases, isthmus was quite bulky while it was thin in the third case. In all of the cases, the isthmus was lying anterior to the aorta and inferior vena cava (IVC) just below the origin of inferior mesenteric artery. The calyces were normal in number and position in all but one kidney in which the calyces were extrarenal bilaterally (Fig. 3). The ureters were inserted high on renal pelvis and were medially placed in all cases except one with extrarenal calyces in which the calyces of the right kidney were laterally directed and the right ureter was forming at the lateral edge of the kidney near the inferior pole (Fig. 3). In two of the cases, ureters crossed anteriorly over the isthmus. In the third case of horseshoe kidney, right ureter turned laterally over renal parenchyma before descending downwards while left ureter crossed the isthmus anteriorly. One of these horseshoe kidneys (Fig. 1) had single right and left renal arteries (RAs) while other two had accessory RAs. In one case, there were four accessory RAs from aorta, two arising as ventral branches and bifurcating to supply the isthmus

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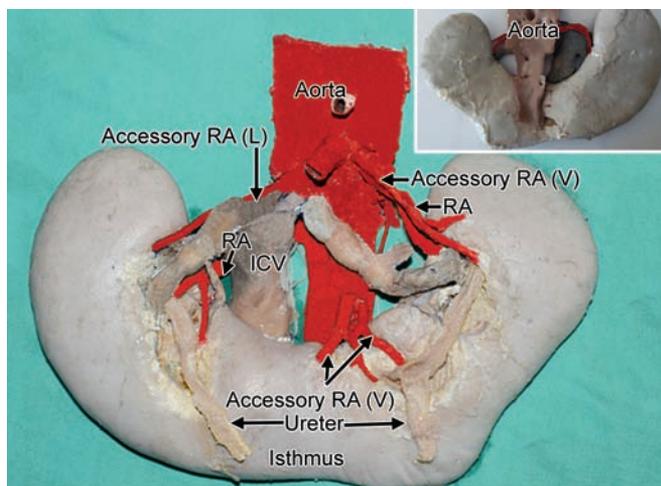


Fig. 1: A case of horseshoe kidney with accessory renal arteries. Posterior aspect of the specimen is shown in the inset. RA: renal artery; V: ventral; L: lateral; ICV: inferior vena cava

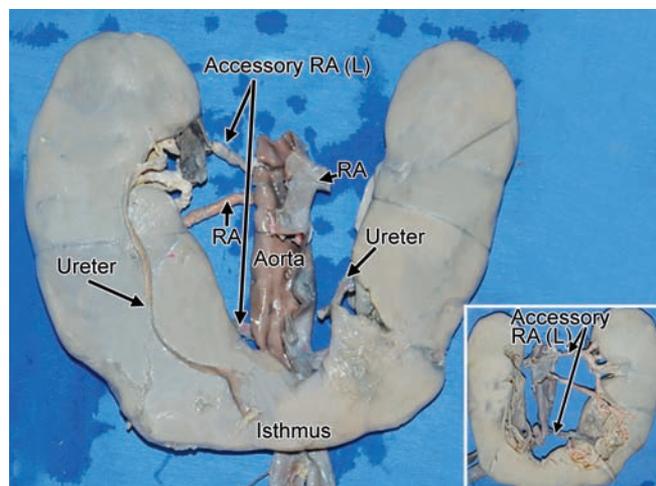


Fig. 2: A case of horseshoe kidney with two lateral accessory renal arteries seen on the right side. Posterior aspect of the specimen is shown in the inset. RA: Renal artery; L: Lateral

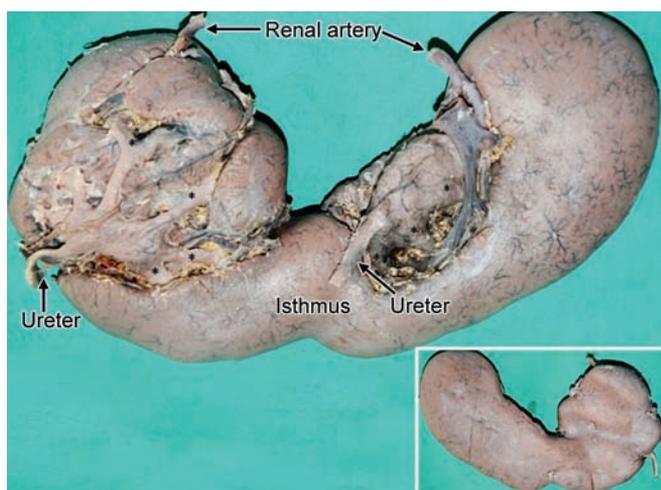


Fig. 3: A case of horseshoe kidney. Bilateral extra renal calyces (*) were seen on the anterior aspect. Right ureter was present along the lateral border of the kidney near the lower pole. Posterior view of the specimen is shown in the inset

and adjacent renal parenchyma, while one ventral accessory RA supplied the left kidney and one lateral accessory RA was entering the right kidney. In second case (Fig. 2), there were two accessory RAs, one on the right side and other on the left; all three were lateral branches from the aorta.

In one of the cases, there was enlarged right kidney with single RA. The left kidney, ureter and RA were absent (unilateral renal agenesis) (Fig. 4A). The left suprarenal gland was normal in size and position. In another case (Figs 4B and C) of unilateral enlarged kidney, hyperplastic kidney was present on the right side weighing 350 gm with left hypoplastic kidney of 7 gm.

A single case of crossed ectopia with fusion was found (Fig. 5). The fused kidneys were situated on the right side at the level of aortic bifurcation. The left kidney was fused with the right kidney just above the hilum of

right kidney. The pelvis of the crossed kidney was placed anteriorly with its separate ureter. Both the kidneys had independent blood supply. The RAs took their origin from the aorta close to its bifurcation.

Two cases of extrarenal calyces were found (Figs 6A and B). In both cases, the extrarenal part was comprised of major calyces. In one of the cases (Fig. 6A), the right kidney was lobulated and was lying in lower lumbar region. Three RAs were supplying the kidney. The upper two arteries took origin from aorta while lower one took origin from right common iliac artery (CIA). The left kidney was normal in size, shape, and position. In other cases of extrarenal calyces (Fig. 6B), the major calyces could be seen from the posterior aspect of the right kidney.

One case of polycystic kidney was found (Fig. 7). In this, both the kidneys were studded with thin walled multiple cysts varying in size from 0.3 to 1.7 cm. The kidneys were enlarged with irregular surface. The pelvicalyceal system and vessels were normal.

We also came across one case of right-sided trilobar kidney with an extrarenal pelvis (Figs 8A and B). This kidney had an anterior, a posterior, and a superior lobe. The hilum was "Y" shaped situated between anterior and posterior lobes with single pelvis and single RA. The left kidney was normal.

DISCUSSION

In our study, congenital anomalies were found in 8 out of 1,900 autopsies. The commonest anomaly in our series was horseshoe kidney (3:1,900). Two of three cases were male. This incidence is slightly less than reported in previous studies (1:400).² The male to female ratio in our series is same as reported earlier (2:1). The age of our cases was 21, 28, and 32 years. This differs from higher incidence in children as reported by Segura et al.³



Figs 4A to C: (A) Hyperplastic right kidney with left renal agenesis. The left suprarenal gland was normal in size and position. Right suprarenal gland is not seen in the figure; (B) Case of unilateral right hyperplastic kidney with left hypoplastic kidney; and (C) Specimen of the figure B is shown after longitudinal renal splitting

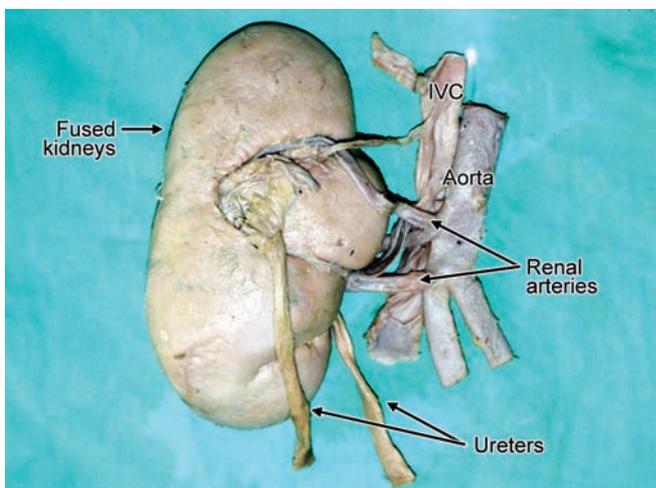
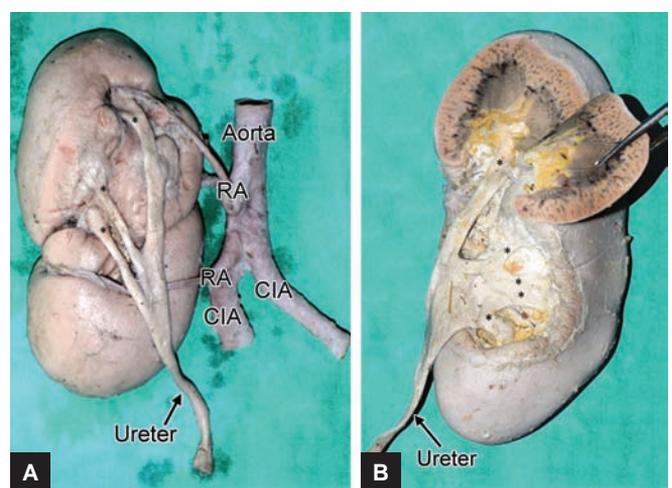


Fig. 5: A case of crossed ectopia with fusion. The fused kidneys were situated on the right side with separate ureters. IVC: Inferior vena cava



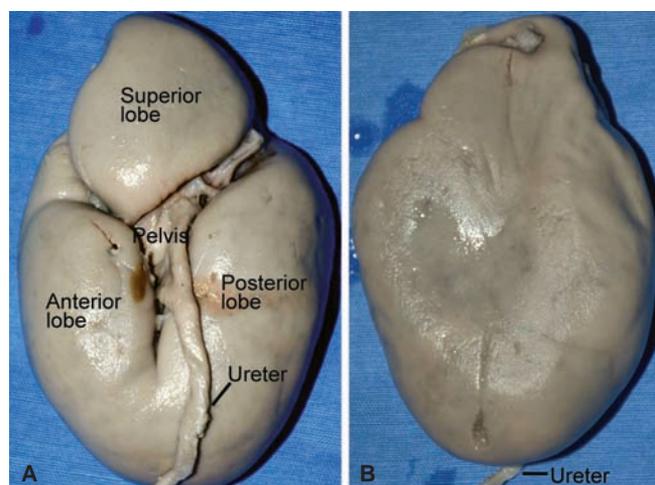
Figs 6A and B: Two cases of extra renal calyces (*). (A) Major calyces seen exposed on the anterior aspect of the right kidney. Three renal arteries were found to be supplying the kidney; and (B) Major calyces seen on the posterior aspect of the right kidney. Upper end of the kidney was excised to show the interior. CIA: Common iliac artery; RA: Renal artery



Fig. 7: Bilateral polycystic kidneys seen from the anterior aspect

In all cases of horseshoe kidney, isthmus was located at lower pole. This finding has been reported in 95% of the cases.⁶ Two of our cases had bulky isthmus and in one isthmus had minimal parenchyma. This is same as reported by Love and Wasserman⁶ and Glenn.⁷ In our cases, isthmus was lying anterior to the aorta and the IVC. The same is reported by Dajani⁸ and Jarman.⁹ The blood supply of these kidneys has been reported to be quite variable.¹⁰ Glenn⁷ reported a single RA to each kidney in 30% of his cases, while in others, there were duplicate, triplicate, or asymmetric RAs. The blood supply to isthmus has been reported to be very variable which may be from RAs or independently arising from aorta, inferior mesenteric artery, external iliac, or common iliac arteries.¹¹ Our findings of single RA in 33% cases, additional blood supply to isthmus in 33% and additional RA in remaining 33% cases corresponds with the reported incidence.⁷ During fetal life, as kidneys ascent from pelvis to posterior abdominal wall, they are supplied by segmental mesonephric arteries at different levels. In ectopia, unaccented kidney retains the vascular pattern corresponding to the developmental stage at which their ascent is arrested.¹²

In our series, one case of unilateral agenesis was found in 1,900 autopsies (1:1,900). Autopsy incidence of this anomaly has been reported to be 1:1,100¹³ and 1:1,500.¹⁴ Embryologically, the fault is at the level of ureteral bud, complete absence of bud or aborted ureteral bud development prevents the maturation of metanephric blastema into an adult kidney.¹⁵ In such cases, gonad was normal while ipsilateral ureter and vas were absent.¹⁶ Ashley and Mustofi had reported absence of ipsilateral ureter in more than half of the cases. In our series, renal agenesis was associated with absent ipsilateral ureter. The ipsilateral suprarenal is normal in cases of renal agenesis as its embryological development is independent of kidney.



Figs 8A and B: A case of right-sided trilobar kidney with an extrarenal pelvis. (A) Three lobes with Y-shaped hilum seen on the medial aspect; and (B) Lateral aspect of the same kidney

Concurrent absence of suprarenal has been reported in less than 10% of autopsy cases.¹⁶ Renal agenesis might be associated with the autosomal dominant, the autosomal recessive, as well as, certain X-linked syndromes.¹²

The autopsy incidence of crossed ectopia with fusion has been calculated at 1 in 2,000.¹⁷ There is a slight male predominance (3:2) and a left-to-right crossover occurs somewhat more frequent. In our series, incidence of crossed ectopia with fusion has been 1:1,900. Our single case of crossed ectopia with fusion does not fit into any of the categories described by McDonald and McClellan.¹⁸ The commonest described crossed ectopia is a unilateral fused kidney with inferior ectopia; others being sigmoid kidney, lump kidney, L-shaped kidney, disk kidney, and unilateral fused kidney with superior ectopia.¹⁷ Our case is an intermediate type of crossed ectopia with incomplete development of crossed kidney.

Extrarenal calyces are an uncommon congenital anomaly of the kidney. This entity was originally reviewed by Eisendrath.¹⁹ There are only about 20 cases reported in the literature.²⁰ In our series, two cases of extrarenal calyces were found. One was isolated anomaly of calyceal system; other was ectopic lobulated kidney with extrarenal calyceal system. It has been hypothesized that this anomaly could be due to a disparity resulting from slow development of metanephric tissue and to a relatively rapid development of ureteric bud. The slow development of metanephric tissue could delay its attachment to the collecting system, leading to development of first and second order collecting systems outside the renal parenchyma. Conversely, with the rapid development of ureteric bud, calyceal system could well develop prior to its coalescence with the nephrogenic mass. This might be the possible mechanism for mal-development in our first case. It is also hypothesized that failure of ureteric bud

to indent the nephrogenic mass fails to provide stimulus for rotation and ascent, accounting for association of extrarenal calyces with renal ectopia and non-rotation. This pathogenesis might have led to the renal development in the second case.²¹⁻²³

The kidney is one of the most common sites in the body for cyst formation.¹⁵ Renal cystic diseases include a broad spectrum of congenital or acquired conditions that have, in common, the presence of cysts in one or both kidneys. Cystic kidneys of different etiologies may appear morphologically similar, whereas the same etiologic entity may cause a wide spectrum of renal abnormalities. Autosomal-dominant, polycystic kidney disease is by far the most common inheritable form of renal cystic disease with an incidence ranging from 1 in 400 to 1,000 live births.^{24,25} In our series, prevalence of polycystic kidney was 1 in 900 autopsies.

The persistence of foetal lobulated contour of kidneys into adult life is not uncommon. However, degree of irregular contour may vary from notch deformities to diffuse multilobulated kidneys. The previous studies have suggested that irregularity of contour occurs more commonly on the left side.^{26,27} A single local lump occurs most commonly on lateral border.²⁷ In our case, it was right kidney which was having three well-defined lobes.

CONCLUSION

The congenital anomalies of kidney are not uncommon. These anomalies can pose diagnostic dilemma to the radiologists and difficulties in treatment planning to surgeons. Therefore, knowledge of these anomalies can help the radiologists and surgeons in improving the diagnosis and treatment of diseases in patients with such anomalies.

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