ORIGINAL RESEARCH PAPER

Radiodiagnosis

A STUDY OF CONGENITAL FETAL RENAL MALFORMATIONS / MANIFESTATIONS BY ANTENATAL ULTRASOUND SCAN IN PREGNANT WOMEN FOR A PERIOD OF TWO YEARS

KEY WORDS: Prenatal ultrasound, antenatal hydronephrosis

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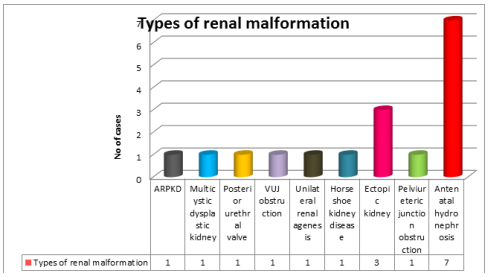
ABSTRACT

Congenital abnormalities /manifestations of the kidneys and urinary tract are one of the most common sonographically identified malformations / manifestation in the prenatal period. The aim of prenatal diagnosis is to detect those anomalies having impact on the prognosis of the affected child and requiring early postnatal evaluation or treatment to minimize adverse outcomes.

In this paper, We present 10 cases of congenital renal malformations and 7 cases of renal manifestations in rural population.

Out of 17 cases of renal malformations/manifestation in the rural area ,1 case is of ARPKD ,1 case of multicystic dysplastic kidney ,1 case of posterior urethral valve ,1 case of vesicoureteric junction obstruction ,1 case of Unilateral renal agenesis ,1 case of horse shoe kidney disease,3 cases of ectopic kidney ,1 case of pelviureteric junction obstruction ,and 4 cases of mild antenatal hydronephrosis,2 cases of moderate antenatal hydronephrosis and 1 case of severe antenatal hydronephrosis.

	N = 17
TYPE OF RENAL MALFORMATION /MANIFESSTATION	**NO OF CASES**
ARPKD	1
Multicystic dysplastic kidney	1
Posterior urethral valve	1
Vesicoureteric junction obstruction	1
Unilateral renal agenesis	1
Horse shoe kidney disease	1
Ectopic kidney	3
Pelviureteric junction obstruction	1
Antenatal hydronephrosis	7



Introduction

The incidence of congenital abnormalities of the kidneys and urinary tract is 3 in 1000 pregnancies . As such they represent 15 % of all prenatal diagnosed congenital anomalies

Embryology

The urinary tract develops from the third week of embryonic life from the intermediate mesoderm (kidneys and ureters) and from the urogenital sinus (bladder and urethra). Renal development passes through three stages: the pro- and mesonephros (both transitory), and the metanephros, which develops into the definitive kidney.

Normal appearance of fetal kidney and urinary bladder.

From about 9 to 12 weeks of pregnancy, the fetal kidneys and adrenal glands can be visualized at both sides of the lumbar spine. They are usually easy to identify because of their relatively hyperechogenicity. The antero-posterior diameter of the renal pelvis (APPD) should be less than 4 mm in the second trimester and less than 7mm in the third trimester .The fetal bladder should be visualized from 13 weeks on. Its identification is easy because of its pelvic location between the umbilical arteries . Fetal urinary production starts at 9 weeks of pregnancy and increases significantly beyond 16 weeks.

The ultrasound examination of the normal urinary tract consists of the assessment of the presence, location and size of both kidneys and the evaluation of their structure and echogenicity. In addition, the presence, size and shape of the fetal bladder are examined.

Below are the antenatal sacn images of fetus showing right and left renal artreies arising from the aorta and supplying both the kidneys (Fig 1) and the normal echoegnicity of both kidneys which are noted anterior to the lumbar spine(Fig 2). Urinary bladder is seen as an anechoic area in the pelvis with two umbical arteries on either side(Fig 3 and 4)

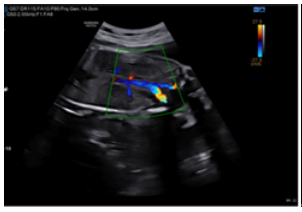


Fig 1:Antenatal scan showing right, left renal arteries

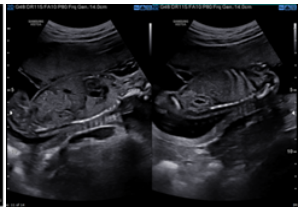


Fig2:Antenatal scan showing normal echogenicity of kidneys

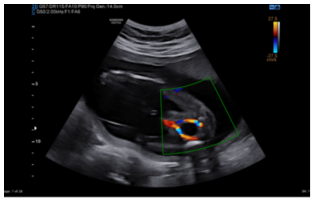


Fig 3:Antenatal scan showing normal urinary bladder

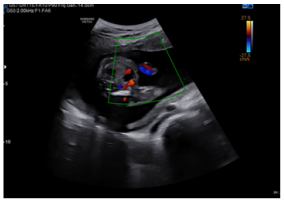


Fig 4:Antenatal scan showing normal urinary bladder

Renal abnormalities
Abnormalities in number and location
Unilateral renal agenesis

Unilateral renal agenesis refers to congenital abscense of one kidney. It occurs in approximately 1/1,300 pregnancies, the majority of which are probably cases of renal aplasia. Although unilateral renal agenesis is mostly isolated and sporadic, it might be part of a genetic syndrome, or occur in association with chromosomal or developmental defects (VACTERL association) and genital abnormalities.

On ultrasound examination the lumbar fossa is empty and the adrenal gland appears elongated ("lying down adrenal sign").The amount of amniotic fluid will be normal.

Bilateral renal agenesis is a lethal condition with an incidence of 1/4,000 pregnancies. Ultrasound features are early anhydramnios from 16 weeks of gestation onwards, absence of bladder filling and empty lumbar fossae. Renal artery color Doppler and Magnetic Resonance Imaging (MRI) may be helpful in confirming the diagnosis.

Below are the antenatal scan images of two fetuses showing empty right renal fossa (Fig 5 and 8) and colour Doppler imaging showing absence of right renal artery in both the fetuses(Fig 6 and 7)

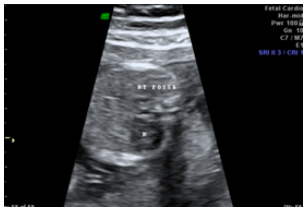


Fig 5:Antenatal scan showing empty right renal fossa.

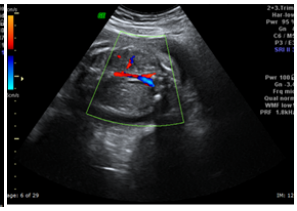


Fig 6:Antenatal scan absent right renal artery

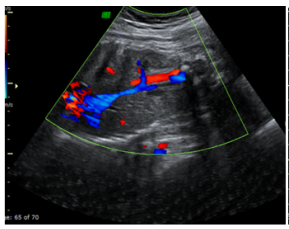


Fig 7 :Antenatal scan of other patient showing empty right renal fossa.

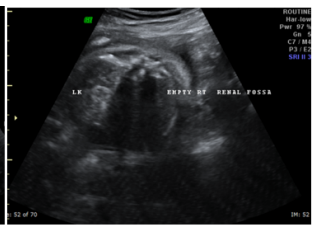


Fig 8:Antenatal scan absent right renal artery.

Ectopic kidney:

Ectopic kidneys occur in about 1/1,000 pregnancies .They are usually smaller and may be malrotated. The pelvic location is the most common, but horseshoe kidneys, crossed (fused) ectopia and even intrathoracic kidneys have been described. Uncomplicated renal duplication should be considered as a normal variant. However, renal duplication is often associated with ureteral abnormalities and hydronephrosis.

Below are the antenatal scan images of a fetus showing empty left renal fossa(Fig 9) Another image of the same fetus showing ectopic left kidney in the pelvis(Fig 10).Antenatal scan image of other fetus showing low lying right adrenal gland (Fig 11) with empty right renal fossa.

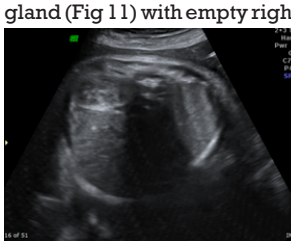


Fig 7 :Antenatal scan of other patient showing empty right renal fossa.

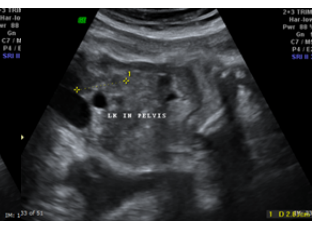


Fig 8:Antenatal scan absent right renal artery.

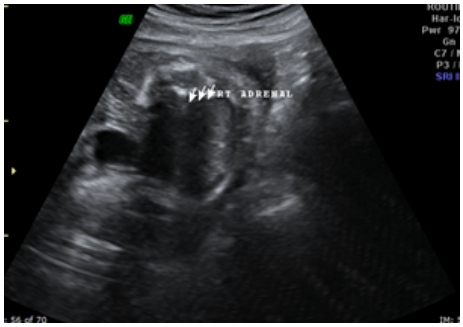


Fig 11 :Antenatal scan of other patient showing right adrenal gland in pelvis.

Horse shoe kidney:

Horseshoe kidney is the most common fusion anomaly of the kidney that occurs with an incidence of approximately one in 400 people .On prenatal ultrasound, the horseshoe kidney shows a fused lower pole and abnormal longitudinal axis. The lower renal margin is not well-defined and the long dimension of the kidney is usually shorter than normal. The fused isthmus is the most conclusive finding, but surrounding bowel echoes can obscure the isthmus in many fetal cases.

Below are the antenatal scan images of a fetus showing fusion of the lower poles of both the kidneys anterior to the spine (Fig 12)

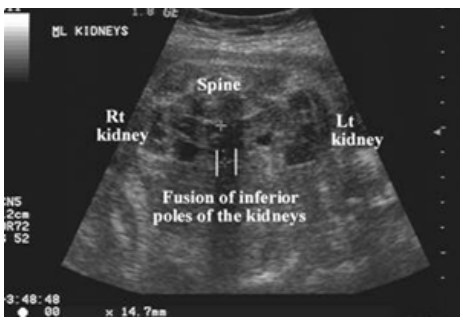


Fig 12: Antennal scan showing the horseshoe kidney with fused lower poles and abnormal longitudinal axis.

Abnormalities in renal size, structure and echogenicity

Hyperechoic kidneys are normal in premature baby's and infants up to 6 months. However, they are an indicator of significant renal pathology in the pediatric population. Fetal hyperechoic kidneys present an etiological diversity with an outcome that is speficic for each of the conditions.

Non-hereditary, fetal hyperechoic kidneys can result from various causes such as an obstructive dysplasia, bilateral multicystic kidney disease, nephroblastomatosis, renal vein thrombosis, ischemia, infectious and metabolic diseases, nephrotic syndrome and aneuploidy. In case of an underlying genetic syndrome and in polycystic kidney disease, the recurrence rate is high. The presence of hyperechoic enlarged kidneys without associated malformations is most frequently associated with polycystic kidney disease (both autosomal recessive and dominant) (Chaumoitre In case of bilateral, isolated hyperechogenic kidneys without family history or cysts, the diagnosis of the underlying etiology

Autosomal recessive polycystic kidney disease (ARPKD)

Autosomal recessive polycystic kidney disease (ARPKD) occurs with an incidence of 1 in 20,000 live births .Ultrasound scans can be normal up to 20 weeks of pregnancy. Kidneys will be markedly enlarged (+4-15 SD) and hyperechoic without (or with reversed) corticomedullary differentiation and with a hypoechoic outer cortical rim .

Below is the antenatal scan images of a fetus showing enlarged and hyperechoic kidneys without corticomedullary differentiation (Fig 13)

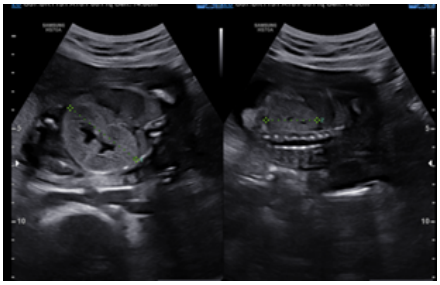


Fig 13:Antenatal sac showing markedly enlarged and hyperechoic kidneys without corticomedullary differentiation

Multicystic kidney disease (MCKD)

Multicystic kidney disease (MCKD) is a developmental disorder of the kidney, in which the normal renal parenchyma is replaced by multiple, non-communicating cysts of varying size. The renal outline is difficult to delineate and can be irregular. The incidence is about 1/4,300 pregnancies. In 25-40% the contralateral kidney will also be abnormal, reflux being the most frequently associated anomaly. Bilateral multicystic kidney disease occurs in about 10-20% of cases and is a lethal condition.

Below are the antenatal scan images of a fetus showing left fetal kidney with multiple, non-communicating cysts of varying size (Fig 14 and 15)

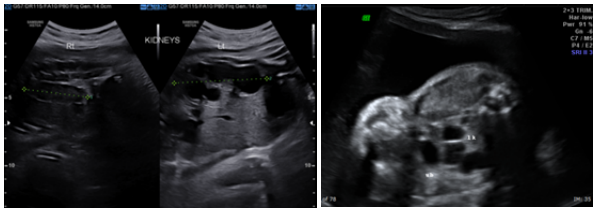


Fig 14 and 15:Antenatal scan showing left kidney with multiple, non-communicating cysts of varying size.

Antenatal hydronephrosis

Pyelectasis is defined as a dilatation of the renal pelvis, whereas hydronephrosis consists of a dilatation of the renal pelvis and the calyces. Both are very common findings on prenatal ultrasound.In fetuses of gestational age 16-27 weeks 4 to 7 mm and 7 to 15 mm, the pelvic dilatation is classified as mild and moderate respectively. When the pelvic dilatation is more than 15 mm, hydronephrosis is called severe. Whereas in fetuses of gestational age >27 weeks ,pelvis dilatation > 7 mm is considered as mild hydronephrosis.

The incidence of hydronephrosis varies between 0.6-4.5% in a non selected population .In about 20-40% the dilatation will be bilateral, and boys are affected twice as much as girls. The severity of renal pelvic dilatation can be assessed using different grading systems. Measuring the maximal antero-posterior diameter of the renal pelvis (APPD) on a transverse scan of the fetal abdomen is the generally accepted method.

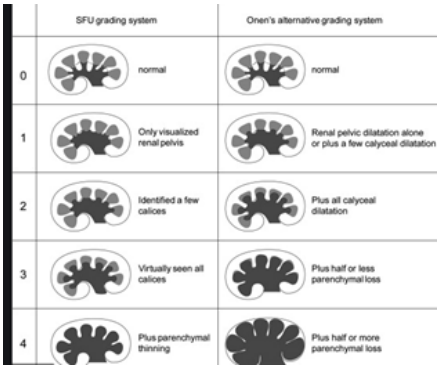


Diagram showing grading of fetal hydronephrosis by society for fetal urology.

The etiology of antenatal hydronephrosis is very diverse .The majority of cases are transient (48%) or physiological (15%). Only in a minority of cases, a significant underlying pathology of the urinary tract is found. Rare causes are an ectopic ureter, prune belly syndrome, urachal cysts and urethral atresia.

The majority (35-50%) of cases of mild antenatal hydronephrosis are transient dilatations. The etiology is unclear, possible causes are insufficient maturation of the pelvi-ureteral or vesico-ureteral junctions associated with increased urinary production or fetal ureteric folds. Physiological hydronephrosis can be secondary to a large, unobstructed renal pelvis and/or an extrarenal pelvis.

Below are the antenatal scan images of a separate fetuses showing mild dilatation of the bilateral pelvicalyceal systems (Fig 15 and 16) and moderate dilatation of the pelvicalyceal system in right kidney(Fig 17) and severe dilatation of the pelvicalyceal system in left kidney(Fig 17)

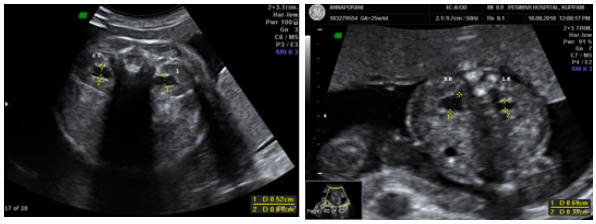


Fig 15 and 16 :Antenatal scan showing mild dilataion of bilateral renal pelvis .

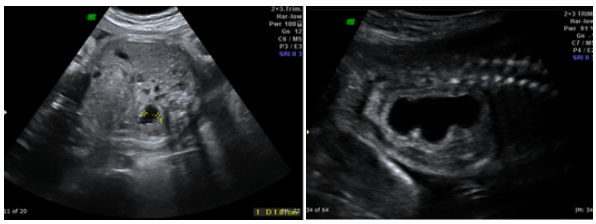


Fig 17 and 18 :Antenatal scan showing moderate dilataion left renal pelvis and severe dilataion of calyceal system respectively.

Pelvi ureteric junction (PUJ) stenosis

Uretero-pelvic junction (UPJ) stenosis occurs in about 1/500 live births (Liang et al., 2002). The male:female ratio is 3:1 and cases are usually sporadic. Bilateral renal involvement can be present in 10-40% of cases (most frequently vesico-ureteral reflux) and 10% of fetuses have extrarenal anomalies. The obstruction is either anatomical or more frequently, functional; the stenosis can be partial or complete. Causes are an intrinsic stenosis or the presence of valves (75%), an insertion anomaly of the ureter, peripelvic fibrosis or crossing vessels (20% are associated with an accessory renal artery). A severely dilated renal pelvis can rupture and evolve to perinephric urinoma and urinary ascites.

Below are the antenatal scan images of a separate fetuses showing significant dilatation of the bilateral pelvicalyceal systems causing thinning of renal parenchyma . No dilatation of the ureters seen. (Fig 19)

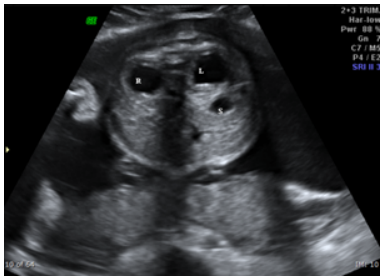


Fig 19 :Antenatal scan showing significant dilatation bilateral renal pelvis and calyces due to bilateral pelviuretric junction obstruction

Posterior urethral valve:

Posterior urethral valves have an incidence of 1/5,000-8,000 neonates and they are the most important cause of severe, bilateral obstructive uropathy. The ultrasound features of this disease are a dilated proximal urethra (keyhole sign), a megabladder with a thick, hyperechogenic bladderwall and severe bilateral hydro-ureteronephrosis or signs of obstructive renal dysplasia (hyperechogenic appearance of the renal parenchyma with cortical cysts)

Below are the antenatal scan image of a fetus showing significant dilatation of the urinary bladder with thick and hyperechoic wall (Fig 20). Significant dilatation of the ureter noted(Fig 21) The antenatal scan image of other fetus showing key hole appearance of the urinary bladder(Fig 22)

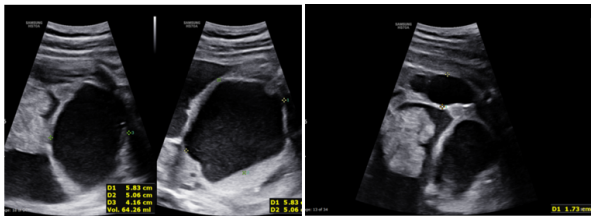


Fig 20 and 21: Antenatal scan shows dilated megabladder with a thick, hyperechogenic bladderwall and severe bilateral hydro-ureteronephrosis

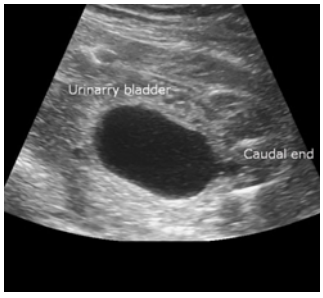


Fig 22: Antenatal scan shows dilated megabladder with a key hole appearance.

CONCLUSION:

Prenatal diagnosis of renal and urinary tract malformations improves perinatal management and the prognosis of the affected child. The objective of prenatal ultrasound is to describe the type of anomaly as accurately as possible, to exclude associated malformations and to screen for parameters predictive of bad renal function, allowing for a multi disciplinary perinatal approach.

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