



ISSN: 0975-833X

Available online at <http://www.journalcra.com>

International Journal of Current Research  
Vol. 12, Issue, 02, pp.10319-10328, February, 2020

DOI: <https://doi.org/10.24941/ijcr.37847.02.2020>

INTERNATIONAL JOURNAL  
OF CURRENT RESEARCH

## RESEARCH ARTICLE

### THE VALUE OF ULTRASOUND IN THE DIAGNOSIS OF CONGENITAL RENAL DISCASES A REVIEW OF LITERATURES

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#### ARTICLE INFO

##### Article History:

Received 24<sup>th</sup> November, 2019  
Received in revised form  
10<sup>th</sup> December, 2019  
Accepted 09<sup>th</sup> January, 2020  
Published online 28<sup>th</sup> February, 2020

##### Key Words:

Normal Anatomy of the kidney  
Normal Ultrasonic Appearance  
Bilateral Renal Agenesis.

#### ABSTRACT

Anomalies of the urinary tract rank third to fourth among congenital malformations and are most frequently associated with other congenital malformations elsewhere in the body. This study is intended to shed light on the role of Ultrasound in the diagnosis of most of these diseases either alone or sometimes in conjunction with other tools of investigations in different age groups and in utero as “prenatal diagnosis”.

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Citation: Du'aa a. Hammody, Khulood Makki and Jazaer Jasim Mohammed, 2020. “The Value of Ultrasound in the Diagnosis of Congenital Renal Discases A Review of Literatures”, *International Journal of Current Research*, 12, (02), 10319-10328.

## INTRODUCTION

Anomalies of the urinary tract rank third to fourth among congenital malformations<sup>(3)</sup> and occur in about (10%) of the population.<sup>(3,17,46)</sup> These anomalies are generally the result of interaction between environmental factors, such as maternal illness and exposure to noxious substances, and genetic factors, including monogenic mutations, chromosomal anomalies or multigenic interactions<sup>(3)</sup>. Probably this high incidence is explained by complex embryology of the region<sup>(46)</sup>. As Edith L. Potter stated: “The more complicated an organ in its development, the more subject is to maldevelopment, and in this respect the kidney outranks most other organs”<sup>(11)</sup>. Frequently the urogenital anomalies are part of multiple congenital malformations involving other organs<sup>(3)</sup>, as the vertebrae, anorectal segment of the gut and the oesophagus<sup>(53)</sup>.

#### Classification<sup>(17)</sup>:

##### A - Anomalies in number:

- Supernumerary kidney
- A genesis and dysgenesis (unilateral or bilateral)

##### B - Anomalies in size:

1. The small kidney (hypoplasia).
2. Compensatory hypertrophy.

##### C - Anomalies in position:

1. Malrotation
2. Ectopia

##### D - Anomalies in form:

1. Fusion (crossed renal ectopia with fusion)
2. Hoarseshoe kidney

##### E - Anomalies in structure:

1. Renal dysplasia
  - a- Multicystic dysplastic kidney MCDK
  - b- Multiple cysts associated with lower urinary tract obstruction.
2. Polycystic kidney disease (PKD)
  - a- Autosomal recessive PKD.
  - b- Autosomal dominant PKD.
- C- Von Hippel Lindau syndrome.
- 3 - Medullary cystic diseases:
  - a- Medullary spong kidney.
  - b- Medullary cystic disease

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- C- Renal tubular ectasia
- d- Renal tubular ectasia with congenital hepatic fibrosis
- e- Juvenile nephronophthisis
- 4- Multilocular renal cyst (cystic nephroma)
- 5- Pelvocalyceal diverticula.
- 6- Congenital megacalyx.

**Embryology of the kidney:** The genital and urinary tract arise from the urogenital ridges on the posterior abdominal wall<sup>(46)</sup>. Three pairs of renal structures are formed in early fetal life, the pronephros (fore kidney) and the mesonephros (mid kidney) which involutes later on but the metanephros (hind kidney) which begins to form during the fifth week of life and persists to form definitive kidney which produces urine from the eleventh week of life.<sup>(46,10)</sup> The duct which drains the mesonephros "Wolffian duct" grows caudally, emptying into the " cloaca", and appears from its dorsal surface called ureteral bud". Its cranial end grows cephalad into a mass of undifferentiated mesoderm which is the metanephros and it will form the kidney proper.<sup>(17)</sup> The cephalad end of the ureteral bud starts to enlarge forming " the pelvis" then divides to form the " major and minor calyces"<sup>(10)</sup>. The renal cortex is formed from the renal mesenchyme which gives rise to the glomeruli and uriniferous tubules<sup>(17)</sup> which later unite with those derived from the ureteric bud .Kidneys are initially formed in the pelvis but later they migrate into the abdomen with medial rotation of the hilum, while in the pelvis, kidney derives its blood supply from the sacral and iliac vessels but as they ascend they derive their blood supply from the aorta<sup>(46)</sup>.

**Normal Anatomy of the kidney:** Kidney is retroperitoneal structure<sup>(46,49)</sup> lies under cover of costal margin and is obliquely on the posterior abdominal wall. The right is slightly lower, during inspiration both descend down by as much as 2.5cm.<sup>(49)</sup> The normal adult kidney varies from 9-12 cm length, 2.5-3 cm thickness and 4-5 cm in width<sup>(46,49)</sup>, a difference of more than 1.5-2 cm is significant. Kidney is surrounded by fibrous capsule called " true capsule", outside it is covering of perinephric fat surrounded by a perinephric fascia which encloses the kidney and adrenal gland is also called " Gerota fascia". On the medial border of each kidney lies the renal hilum which contains the renal vein, two branches of renal artery and the ureter<sup>(49)</sup>. Each branch of the renal artery supplies a corresponding segment of the kidney and breaks down to minute arterioles (interlobar artery), between the cortex and medulla. They are called "the arcuate vessels"<sup>(49)</sup>.

**Internal Appearance of the kidney:** Kidney is composed of internal medullary portion and external cortex. The medulla consists of series of striated conical masses "the renal pyramids". They vary from 8-18 in number with bases directed peripherally the apices converge toward the renal sinus where their prominent papillae project into the lumina of the minor calyces. Each unit is separated by prolonged cortical substance called the "renal column of Bertin". The four to thirteen minor calyces are cup-shaped tube which comes into contact with one or two or more central papilla<sup>(49)</sup>.

## PART 11

### RENAL ULTRASOUND

**Technique of Renal Ultrasound Examination:** Transducer: 3.5 MHz, but thick set patient may need 1.5 MHz only<sup>(16)</sup>.

•Position: The patient is usually examined in prone position. The supine position is useful for upper poles, otherwise it is obscured by the lung and ribs<sup>(16, 36)</sup>. The right kidney is frequently imaged through the liver<sup>(36)</sup> with the patient supine. Suspended respiration is a must because:

- (1) The kidney is pushed away from the ribs into view.
- (2) Fine details are not blurred by motion.
- (3) Longitudinal dimensions of the kidney are not spuriously altered by the kidney movement .

#### •Scanning planes<sup>(16)</sup>:

The simplest approach is to start with the longitudinal scan then the transverse is particularly useful in showing anteroposterior displacement of the kidney and cysts, then oblique scan with the long axis of the kidney

**Normal Ultrasonic Appearance:** The renal outline is produced by the echo that arise from the interface of renal capsule and surrounding tissue<sup>(36)</sup>. Cortical thickness is usually uniform but may be tabulated particularly in neonates<sup>(46)</sup>. The renal cortex has a fine homogenous hypoechoic relative to the hepatic and splenic parenchyma and may appear relatively echofree, and the renal parenchyma (medullary pyramids) are hypoechoic relative to the cortex. The cortico medullary junction is demarkated by the arcuate arteries which are seen as small echogenic foci<sup>(16,46)</sup>. The collecting system, vessels, and fibrofatty and lymphatic tissues at the centre of the kidney are seen as the echogenic " central echo complex" which is the most echogenic part of the kidney<sup>(46)</sup>. It is nonhomogenous flattened ellips<sup>(44)</sup> as in Figure (1)<sup>(46)</sup>. In premature and young infants, there is not yet enough peripelvic fibrofatty present to produce visible sinus echo<sup>(44)</sup>. The kidney of the preterm infant is more echogenic than the liver, the peri renal fat is not yet sufficiently developed in infants and young children to be sonographically visible and the renal fascia and capsule are directly apposed, so the kidney has in a very close proximity to the liver<sup>(44)</sup>.

**Renal Size:** The bipolar diameter is the most frequently used parameter measured from pole to pole. The greatest measurement is the closest estimation of the true renal length. The renal size is related to age, weight and body surface area .

#### A. Renal length<sup>(46)</sup>:

Baby < 1 year of age:

Average renal length =  $4.98 + (0.155 \times \text{age in months})$  cm

Children > 1 year:

Average renal length =  $6.79 + (0.22 \times \text{age in years})$  cm

#### B. Renal volume:

The renal volume is the most exact measurement of renal size<sup>(20)</sup>. Renal volume = width x length x thickness x  $0.5233 \text{ cm}^3$ <sup>(46)</sup>. The median renal volume are 146 cm in the left kidney and 134 cm in the right kidney<sup>(20)</sup>. In a study done on preterm and term infants, the kidney volume was measured by real time linear computerized Ultrasound Scanner, with 5 MHz transducer, a significant correlation was found between the volume of the kidney and either gestational age or birth weight or height and no difference were between males and females<sup>(44)</sup>.

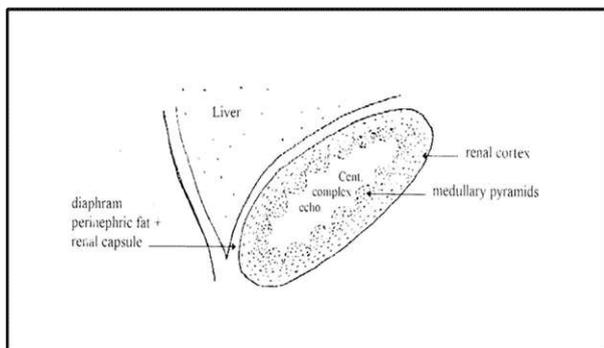


Figure 1. Right Kidney Sonographic Anatomy

Table 1. Renal length in cm

| Age     | Percentage |     |      |
|---------|------------|-----|------|
|         | 5%         | 50% | 95%  |
| Birth   | 4          | 5   | 6    |
| 1 year  | 5          | 6.5 | 8    |
| 5 year  | -          | 8   | -    |
| 10 year | 7          | 9   | 10.5 |
| adult   | 9.5        | 11  | 12.5 |

Table 2. Renal Volume

| Age     | Renal Volume        |
|---------|---------------------|
| Birth   | 20 cm <sup>3</sup>  |
| 1 year  | 30 cm <sup>3</sup>  |
| 18 year | 155 cm <sup>3</sup> |

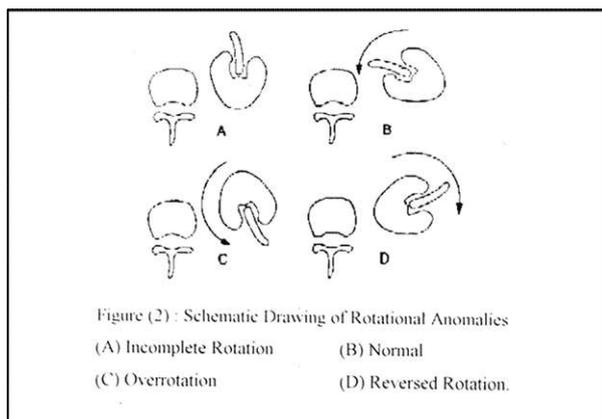


Figure (2): Schematic Drawing of Rotational Anomalies  
(A) Incomplete Rotation (B) Normal  
(C) Overrotation (D) Reversed Rotation.

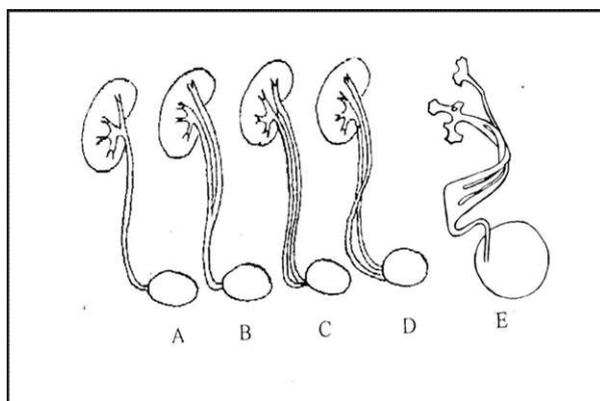


Figure 3. Schematic Drawing of:

## Different Types of Congenital Renal Diseases

### A. Anomalies in Number:

(1) Renal Agenesis: Ashley and Mastoti have suggested the following terminology: If no renal tissue is formed the

condition is called "agenesis". If the kidney is represented by, a nodule of tissue that lacks resemblance to normal renal parenchyma, it is called "dysgenesis". If the kidney is tiny but similar to normal organ, it is hypoplasia<sup>(17)</sup>.

**Bilateral Renal Agenesis:** This occurs in 0.3% of live newborns. It is incompatible with life<sup>(9)</sup>. It is associated with hypoplastic lungs, oligohydramnios, facial anomalies<sup>(9, 53)</sup>. It is due to failure of the ureteric bud to make contact with the nephrogenic blastema at the proper time<sup>(14)</sup> or due to failure of development of the ureteric bud from the Wolffian duct<sup>(53)</sup>.

**Unilateral Renal Agenesis:** This is uncommon congenital anomaly. It is about 1 case per 1000 population<sup>(9, 40, 40)</sup>. Ipsilateral ureter is absent in 90% of cases<sup>(9)</sup>, absent trigon in 50% of cases<sup>(9)</sup>, associated congenital anomalies are as much as seminal vesicle cyst, bicornuate, unicornuate uterus<sup>(46, 51)</sup>, uterine and vaginal septation<sup>(46)</sup>, vaginal agenesis<sup>(46)</sup>.

**Ultrasonic Appearance:** Ultrasound can outline the normal kidney and tell with certainty whether one kidney is absent or pathologically afflicted<sup>(49)</sup>. The solitary kidney is somewhat enlarged<sup>(9)</sup>. The renal fossa may be filled with bowel loops or pancreatic tail in the left side and with the duodenum, proximal small gut, hepatic flexure and liver in the right.

Certain structures may be mistaken for a kidney as an abscess, calculus or adrenal gland which is found to be large in neonates with renal agenesis<sup>(46)</sup>.

### (2) Free Supernumerary Kidney

Up to five free kidneys in one individual have been described.

**Embryogenesis:** Two ureteric buds arise from the Wolffian duct, the inferior kidney is the supernumerary which is usually smaller<sup>(17)</sup>.

**Ultrasonic Appearance:** Ultrasound may be able to outline two separate kidneys if they are within normal renal area but may overlook the extrasystem if they are in the pelvis<sup>(49)</sup>.

### B. Anomalies in Size

**Renal Hypoplasia:** It is incomplete development of the kidney<sup>(17)</sup>. Unilateral hypoplasia is more common than agenesis<sup>(46)</sup>. This is due to insufficient response of the metanephric mesoderm to the insufficient stimulus of the ureteric bud<sup>(17)</sup>.

**Uluson Meillilic:** The kidney is small in size, smooth in outline and more echogenic<sup>(44)</sup>. In posterior longitudinal scan: small oval echogenic with loss of corticomedullary differentiation<sup>(44)</sup>. Renal hypoplasia shows normal pelvis and calyces and even small if compared with that found in post obstructive atrophy

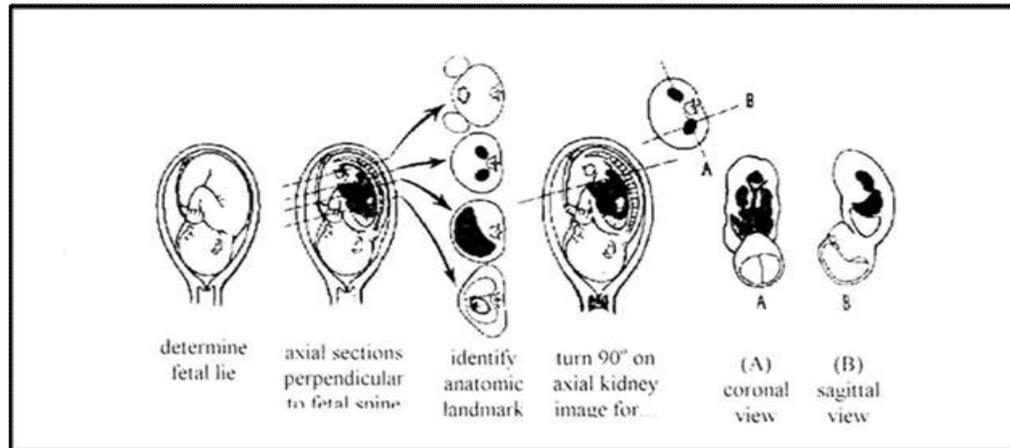
### C. Anomalies in Position:

#### (1) Malrotation

As the kidney ascends from the pelvis to the abdominal cavity, there is 90° rotation of the anteriorly located pelvis medially. If the kidney does not rotate or rotate less than 90° it is called "malrotation".

**Table 3. Mean Fetal Kidney Circumference / Abdominal Circumference Ratios and SD (Standard Deviation)**

|      | Gestational Age (wk) |                 |                |                 |                 |                 |
|------|----------------------|-----------------|----------------|-----------------|-----------------|-----------------|
|      | ≤16<br>(n=9)         | 17-20<br>(n=18) | 21-25<br>(n=7) | 26-30<br>(n=11) | 31-35<br>(n=19) | 36-40<br>(n=25) |
| Mean | 0.28                 | 0.30            | 0.30           | 0.29            | 0.28            | 0.27            |
| S.D  | 0.02                 | 0.03            | 0.02           | 0.02            | 0.03            | 0.04            |

**Figure 4. Identification of the Fetal Kidney**

If the kidney rotates more than 90° the renal pelvis will face posteriorly and called “ over rotation”, a rotation to the opposite side results in the renal pelvis facing laterally, it is “ reversed rotation”<sup>(9)</sup> as in Figure (2) .Malrotation may be unilateral, bilateral (17), may occur in normally located kidney or in the displaced or fused kidney and in the latter, it is more<sup>(9)</sup>.

**Ultrasonic Appearance:** Ultrasound can show very narrow kidney with no sinus echo by the longitudinal paravertebral scan, renal hila directed anteriorly instead of medially by the posterior transverse scan. Its diagnosis is very easy by C.t or MRI<sup>(44)</sup>.

## (2) Ectopia

The terms ectopia and dystopia describe kidneys that are congenitally located in abnormal position<sup>(17)</sup>. It is either:

### A. Simple ectopia:

#### **Embryogenesis:**

Simple ectopia is a kidney situated in its normal side but below its adult level result from failure of normal ascent<sup>(17,46,53)</sup>.

#### **Types:**

1- Pelvic (sacral kidney)

Kidney is located in the true pelvis<sup>(17)</sup>. It is most common ectopic with incidence, 1/700 of the population<sup>(9)</sup>.

2-Intrathoracic kidney

It is one of the least common positional anomalies<sup>(17)</sup>. The kidney is supradiaphragmatic mainly herniate through the foramen of Bockdalek during development (17.40.33), or less commonly resulted from migration of the primitive kidney above the level of diaphragm before this structure is completely formed<sup>(9,X)</sup>

It should be considered in a child with a mass at the base of the lung on CXR, I. V. U is indicated and it is diagnostic<sup>(6)</sup>.

### B. Crossed Renal Ectopia:

- (1) Crossed renal ectopia with fusion.
- (2) Crossed renal ectopia without fusion.
- (3) Solitary crossed renal ectopia.

The crossed ectopia with fusion is more common<sup>(9, 17)</sup>. The incidence of crossed renal ectopia is 1 in 7500 autopsy<sup>(58)</sup>. Several theories have been put to explain crossed renal ectopia<sup>(17)</sup>: The ureter somehow crosses the midline and induces formation of kidney from the contralateral, nephrogenic blastema. A normally developing kidney becomes attached to the kidney from opposite side and is dragged across the midline during ascent. Crossed renal ectopia may be associated with other congenital anomalies, as unilateral agenesis of fallopian tube and ovary<sup>(58)</sup>, unicornuate uterus, imperforate anus and scoliosis<sup>(19)</sup>.

**Ultrasonic Appearance:** Under no circumstance should a pelvic kidney be misdiagnosed as primary pelvic tumour<sup>(9)</sup>. Central sinus complex and corticomedullary differentiation should be looked for on all pelvic Ultrasound Examination<sup>(44)</sup>.

**Scrotal Kidney and Ureter:** Urinary tract herniation is not an unusual occurrence. It has been reported that 1 to 1000 of all large inguinal hernias contain bladder, however, ureteral herniation is rare. For the kidney to be in the scrotum it is mostly aptotic rather than being congenital anomaly<sup>(37)</sup>.

### D. Anomalies in Form

#### (1) Fusion

**Embryogenesis:** It has been postulated that renal fusion occurs if the nephrogenic tissues are pressed together by the umbilical arteries as the kidneys ascent out of the pelvis. This is of two types:

- 1 Crossed renal ectopia with fusion
- 2 Horseshoe kidney

**Horseshoe Kidney:** It is the most common type of fusion anomalies. It is characterized by fusion of either the upper or lower poles<sup>(17)</sup>, mostly the lower is in about 96% of cases<sup>(9)</sup>. It is arrested by the mesenteric vessel and they are lower in the abdomen than normal<sup>(46)</sup>. In most cases the pelvis is malrotated. Horseshoe kidney is the most frequent renal anomaly in Turner's syndrome<sup>(8)</sup>.

**Ultrasonic Appearance:** The horseshoe kidney should be evaluated from the supine since the kidneys generally appear lower in the abdomen and may be attenuated by the iliac crest in the prone position, the isthmus is best recorded with single sweep technique and is seen as a sonoduct band draping over the great vessels<sup>(9, 49)</sup>, which are sometime difficult to be differentiated from an enlarged lymph node. Real time equipments may show the isthmus connecting the two capsules. By the coronal scan, both kidneys imaged concurrently in the longitudinal view, the linear sonolucency between the kidneys is the aorta<sup>(44)</sup>. By the posterior transverse scan shows rotation of both renal axes with the hilum anterior, if the isthmus is visualised by the anterior scan, the diagnosis is settled<sup>(44)</sup>.

### **Pancake Kidney**

Fusion of both kidneys in front of the lower abdominal aorta and bifurcation results in flat nonreniform conglomeration of renal parenchyma which is called "pancake kidney".

## **(2) Duplication**

Duplication of the renal collecting system and ureter is a common finding affecting 1 in 70 of the population<sup>(46)</sup>. It is due to varying degree of division of the ureteric bud. There is a great variation in the degree of division which results in a spectrum of changes<sup>(46)</sup>, the affected kidney is larger

### **Types :**

#### **1 Bifid Renal Pelvis**

Common anomaly is considered as a normal variant<sup>(9)</sup>.

#### **2-Incomplete Duplication**

It occurs due to premature division of the ureteric bud during ascent. It may be confined to the renal pelvis or involved part of the ureter in a (Y) shape configuration<sup>(9, 53)</sup>.

#### **3-Complete Duplication**

This involves renal pelvis and whole length of the ureter. Both ureters have separate opening in the bladder<sup>(53)</sup>. The ureter drains the lower segment which inserts into the bladder more superiorly and laterally<sup>(9, 53)</sup> as in Figure (3)<sup>(9)</sup>.

- (A) Bifid Renal Pelvis.
- (B) Incomplete Duplication or (Y) ureter.
- (C) Incomplete Duplication when two ureters joint into a single ureteral orifice.
- (D) Complete Duplication when both ureters have separate opening in the bladder.

\*(E) Three major calyces connected in an extra renal pelvis (Artist's impression of "E")

**Unipapillary kidney:** An unbranched renal pelvis is extremely rare<sup>(9, 28)</sup>, it may be associated with other abnormalities more important than itself when the contralateral kidney is absent or abnormal<sup>(28)</sup>.

### **Ultrasonic Appearance:**

#### **(1) Incomplete Duplication**

By the posterior longitudinal paravertebral scan, "nose" of the parenchyma project from the posterior side into the renal sinus<sup>(44)</sup>.

#### **(2) Duplex kidney<sup>(44)</sup>**

- 1 Flank longitudinal scan shows: parenchymal bridge completely divides the renal sinus with smaller upper and larger lower poles.
- 2 Transverse upper abdominal scan show: hypoechoic area at the site of parenchymal bridge where the echogenic renal hilum or renal sinus would normally occur
- 3 Posterior longitudinal scan show continuous parenchymal bridge extends forward from the posterior side.

There is a correlation between congenital renal fusion and aortic pathology which may sometime aid in the diagnosis<sup>(43)</sup>.

## **E. Anomalies in Structure (Cystic diseases of the kidney)**

### **1- Multicystic Dysplastic kidney MCDK**

A condition in which the entire kidney is composed of numerous (10- 20) Variable sized (few mm-2 cm) cysts, resembling a bunch of grapes<sup>(17)</sup>. The cysts are not communicating, a normal uniform contour is absent<sup>(34, 47)</sup>, with small or absent pelvis and calyces, hypoplastic or absent renal artery<sup>(9)</sup>.

**Embryogenesis:** The MCDK and hydronephrosis are the two ends of spectrum of same embryogenesis. If there is a complete obstruction of the ureter (atresia) between (8-10 wk) of gestation, there is insufficient nephronic development too<sup>(9, 47)</sup>, that is "MCDK". If obstruction is incomplete and occurs after nephrogenesis is complete (36 th.wk), the pelvis and calyces will be dilated with no dysgenesis that is "internal hydronephrosis"<sup>(47)</sup>. If obstruction is incomplete between (10-36 th.wk) results in varying degrees of cyst formation, pelvic and calyceal dilatation and dysplastic parenchymal changes that is "hydronephrotic type of MCDK". MCDK is the second most common abdominal mass in neonates<sup>(2, 30, 34, 47, 49)</sup>.

### **Ultrasonic Appearance:**

#### **MCDK can be diagnosed by Ultrasound alone<sup>(2)</sup>**

- 1 Variable sized cysts with the largest cyst being peripherally located<sup>(9, 47)</sup>, with ragged inner cyst due to dysplastic tissue.
- 2 Absence of connection between the adjacent cysts on a good quality real time study<sup>(47)</sup>.
- 3 Absence of renal parenchyma surrounding the cysts<sup>(9, 47)</sup>.

- 4 Presence of echogenic areas indicating tiny cysts in an eccentric location<sup>(47)</sup>.
- 5 Renal pelvis and sinus are not seen<sup>(9)</sup>.
- 6 "Claw" like configuration or clump of echoes are seen between the cysts representing the septa<sup>(30)</sup>.

Vinocur et al, in their study, found that the MCDK may show change in size on follow up. Some may decrease in size and be even atrophied completely, others show no such changes (34) Such study and others, as that done by Strife et al, support the non surgical approach to the treatment of patients with MCDK<sup>(39, 52)</sup>.

## Internal Hydronephrosis

### Ultrasonic Appearance:

- 1) Visible renal parenchyma surrounding a central cysts component<sup>(47)</sup>.
- 2) Small peripheral cysts (calyces) budding off a large central cyst (the pelvis) forming a "glove like appearance"<sup>(47)</sup> that is medial localisation of the largest cysts<sup>(30)</sup>.
- 3) Visualisation of a dilated ureter in the region of the kidney in those cases with distal obstruction.
- 4) Single large cysts.

## 2 -Polycystic Disease of the Kidney

**Embryogenesis** :There is failure of connection of uriniferous tubules originating from nephrogenic blastema with the collecting tubules derived from the ureteric bud<sup>(17)</sup>.

**A. Autosomal Recessive PKD:** Incidence 1 in 6000 - 14000 births, it is of two forms distinguished by the age of presentation and predominance of renal over hepatic manifestation or vice versa .

### (1) Newborn Form

Renal disease predominates in neonatal period. The affected neonate has oligohydramnios sequence at death soon after birth<sup>(9, 11)</sup>.

### Ultrasonic Appearance:

- 1 Enlarged kidney<sup>(11, 53)</sup>, with maintain of the renal form configuration<sup>(11)</sup>
- 2 Diffuse increased echogenicity obscures the corticomedullary differentiation<sup>(11)</sup>.
- 3 Macrocysts appear as anechoic rounded area<sup>(11)</sup>.
- 4 Calyceal echoes are poorly seen because of similar echogenicity to renal parenchyma and small amount of renal sinus fat<sup>(11, 53)</sup>.
- 5 Renal margins are poorly outlined and radiolucent due to compressed cortex<sup>(11)</sup>.
- 6 Diffusely increased hepatic echogenicity and decreased visualisation of peripheral portal veins are due to fibrous tissue<sup>(11)</sup>.

**2 -Childhood Form:** Milder or less pronounced cystic changes and more hepatic fibrosis.<sup>(53)</sup> usually present at about the age of 3-5 years or later<sup>(9,33)</sup> usually with signs and symptoms of portal hypertension.<sup>(9, 11, 53)</sup>

## Ultrasonic Appearance

1 Enlarged kidney with increased echogenicity is mainly in the medulla<sup>(11)</sup>, with foci of very brightly increased echogenicity because of focal tubular cysts<sup>(11)</sup>.

2 Macrocysts may appear.

3 Liver may show enlargement with homogenous or heterogenous increase in echogenicity with decrease visualisation of peripheral portal venous vasculature<sup>(11)</sup>.

## B. Autosomal Dominant PKD

Incidence 1 in 1000 population<sup>(9)</sup>, although some authors as Kaplan et al, (1989) found that it ranges between 1 in 200 - 1 in 1000 population This makes it one of the common dominantly inherited conditions<sup>(11)</sup>. The age of presentation is dependant on the extent of abnormality .Bilateral renal enlargement, although unilateral cases have been postulated<sup>(7, 26)</sup>, with variable sized randomly scattered cysts in the cortex and medulla. Other organs cysts are common as liver and pancreas<sup>(9, 53)</sup>, nephrolithiasis may be associated with segmental obstruction due to calculus, cysts or blood clot, cysts walls may calcify<sup>(1)</sup>.

### Ultrasonic Appearance:

- 1) Kidney may be enlarged<sup>(11,49)</sup> or still normal in size with equal number of cysts in each kidney<sup>(11)</sup>.
- 2) Cysts can be diagnosed in the liver, pancreas and spleen<sup>(11,49,53)</sup>.
- 3) The most consistent renal ultrasonographic finding in the children with autosomal dominant PKD is renal enlargement with increased echogenicity<sup>(23)</sup>.

The quick diagnosis and non invasiveness of renal and extrarenal involvement makes Ultrasound superior to C.T. scan and makes it a procedure of choice for diagnosis, screening, follow up<sup>(54)</sup> and to detect any complication of autosomal dominant PKD<sup>(38)</sup>.

Identification of cysts in children of affected adults permits genetic counseling of children prior to procreation in addition to yearly screening which may determine the age at which the absence of cysts indicates that a patient will not develop autosomal dominant PKD<sup>(5)</sup>.

## (3) Medullary Cystic Diseases

Cystic disease of the medulla with rather normal size kidney<sup>(17)</sup>.

### A. Medullary Spong Kidney

Cystic changes of the medulla, partial or complete, sometime affect single papilla<sup>(9,53)</sup>, are associated with dilatation of the collecting tubules, intratubular calculi which are present in more than 50% of cases<sup>(9)</sup>. The uncomplicated disease is silent<sup>(4,53)</sup>. It sometimes causes urinary tract infection, unilateral limb hemihypertrophy may be seen on the ipsilateral side<sup>(9, 18)</sup>, congenital pyloric stenosis and Ehlers - Danlos syndrome have been described.

### Ultrasound appearance ,

1 The kidney is enlarged with localised or generalised increase in renal substance thickness<sup>(53)</sup>.

2Ultrasound shows a well-defined, highly echogenic pyramids due to multiple cysts which are too small to be individually identified<sup>(9)</sup>.

3Calculi may be detected by Ultrasound as hyperechoic areas are associated with acausticshadow<sup>(9)</sup>.

## B. Medullary Cystic Disease

### *Ultrasonic Appearance:*

1Irregular widened central echoes when small cysts are present and well defined cysts structure when larger medullary cysts are predominate<sup>(4)</sup>.

2Thin cortex

3The kidneys are normal in size or small and not large to differentiate it from autosomal recessive PKD<sup>(9)</sup>.

## C. Renal Tubular Ectasia with Congenital Hepatic Fibrosis:

### *Ultrasound Appearance*

1Higher than normal level echoes in the liver is due to the hepatic fibrosis, on A-mode examination. The hepatic echoes are normally one third to one half the height of echoes from the diaphragm, with fibrosis or inflammation the level of these echoes increase<sup>(4)</sup>.

2- Distorted echo pattern in the kidney is due to ductular ectasia<sup>(4,9)</sup>.

3- Nephromegaly.<sup>(4)</sup>

## (4) Congenital Mesoblastic Nephroma

Among renal masses in the first few months of life, the mesoblastic nephroma is the most common neoplasm<sup>(18,25)</sup>. It is a well defined mass within the renal parenchyma. It may show low level echoes or anechoic mass without acaustic enhancement if the tumour is solid, and may show complex echo pattern with several irregular anechoic areas within an echodens mass<sup>(118)</sup>.

## PART III

### PRENATAL DIAGNOSIS OF CERTAIN CONGENITAL RENAL DISEASES

Ultrasound has had a revolutionary impact on the detection of congenital malformations during the antenatal period<sup>(50)</sup>. Management of the pregnancy may be greatly altered depending upon the ultrasound findings ; first, early termination of pregnancy<sup>(33,42)</sup>, or fetal therapeutic procedures may be done. Second, the mode and timing of delivery may be changed<sup>(42)</sup>. Serial ultrasonic studies may be required, i.e. two ultrasonic examinations, one is at the "17 th." week, the other, at the " 32 nd" week as up to two third of congenital anomalies are still hidden by single examination<sup>(24)</sup>, or the abnormality may be associated with other congenital anomalies which at time of scanning not yet detectable<sup>(42)</sup>.

### **Ultrasonic Examination of the Urinary Tract in the Fetus:**

There are several important concepts the sonographer should be familiar with when evaluating the urinary tract<sup>(30)</sup>. The fetal

kidneys have assumed their recognisable adult form and position by approximately the tenth to twelfth post menstrual week. However, an accurate ultrasonic identification is not possible until 15th. week due to their small size and lack of perirenal fat<sup>(42)</sup>. However, between 17 and 22 weeks (the critical time for genetic counseling) one or both kidneys were seen in 90% of cases<sup>(42, 55)</sup>. The kidneys should be evaluated by assessing kidney's anatomy size, and texture. The normal anatomy is cortex, parenchyma, pyramids, calyces and pelvis. The texture, is the homogenous pattern of renal echoes<sup>(50)</sup>. The size of the fetal kidney: It can be assessed using the kidney circumference to abdominal circumference (KC/AC) ratio as it described by Grannum et al,<sup>(50)</sup> normally it is 0.27 - 0.30 . On transverse scan, the kidney circumference is obtained using the anteroposterior and transverse diameter, (similar technique to calculation of abdominal circumference ) the KC is divided by the AC (obtained at conventional level )<sup>(50)</sup> . Severe renal anomalies are almost always accompanied by significant Oligohydramnios, therefore, a careful assessment of amniotic fluid in these cases is mandatory<sup>(50)</sup>.

**Technique:** Once the fetal lie is located, sections perpendicular to the fetal spine are obtained. The relative craniocaudal location of the axial scan with the fetus is determined by identification of the major fetal landmarks: the heart (thorax) liver (upper abdomen), kidneys (mid abdomen), and bladder (pelvis)<sup>(55)</sup>. For a coronal or sagittal view, the patient or scanning arm is turned 90° from the plane used for the axial view, and the angle through the fetus is adjusted according to the landmarks identified as in Figure (4)<sup>(55)</sup>.

### **Ultrasonic Appearance of Different Congenital**

#### **Renal Disease:**

##### **(1) Renal Agenesis**

Bilateral agenesis should be suspected prenatally by Ultrasound, severe oligohydramnios<sup>(42)</sup> or complete absence of amniotic fluid will be diagnosed between 16th. - 28th. weeks gestation<sup>(42)</sup>. In early stages of renal agenesis, amniotic fluid may be visible as it is produced from other fetal sources<sup>(50)</sup>. The bladder will not be visualised and absent kidneys in the second trimester<sup>(40)</sup>. The use of fetal renal artery flow velocity waveform may be helpful in the prenatal diagnosis as it can not be visualized in the presence of such congenital anomaly<sup>(56)</sup>. Hill LM, (1993) emphasized an important pitfall in trans abdominal imaging in the presence of anhydramnios because sometimes structures that were thought to present fetal kidneys where they are shown either bowel or adrenal glands by using endovaginal sonography<sup>(15)</sup>.

##### **(2) Autosomal Recessive Polycystic Kidney Disease:**

Prenatal diagnosis may be feasible by Ultrasound<sup>(50)</sup>. Attempts of diagnosis in the second trimester (16-22th. week) were successful in 50% of cases, therefore, normal kidney in the early gestation in fetus at risk does not get the disease out<sup>(44)</sup>. The evident sonographic features become clear at 30 th. week of gestation<sup>(44)</sup>.

1Bilateral renal enlargement.<sup>(44, 34, 42, 50)</sup>

2Increased echogenicity is due to hundreds of tiny nonvisualized cyst .

3Performance of KC / AC i.e. kidney circumference over abdominal circumference may be increased more than 2 SD<sup>(31,50)</sup> (NR:0.27-0.30).

4Poor delineation of internal structures<sup>(11,31)</sup>.

5Identification of the renal pelvis and small atrophic bladder differentiates the condition from the dominant form of the disease.<sup>(11,31)</sup>

6Oligohydramnios and absent urinary bladder are good indicators of most severe cases<sup>(11, 31, 50)</sup>

**Autosomal Dominant Polycystic Kidney Disease:** The prenatal diagnosis of the dominant polycystic disease was first reported by Zerres et al, (1982). Prior to the 20th. week of gestation, the fetal kidney appears normal on sonography, however, repeated scannings early in the 3rd, trimester, may show it<sup>(91)</sup> as either or more of the following :

1Enlarged kidney<sup>(31,27)</sup>

2Increased echogenicity<sup>(34)</sup>

3Accentuation of the corticomedullary differentiation<sup>(27)</sup>

4Cystic changes<sup>(31, 27)</sup> .

### (3) Multicystic Dysplastic Kidney (MCDK) :

The earliest reported antenatal sonographic diagnosis was made at 18 week of gestation<sup>(31)</sup>.

1Renomegaly<sup>(44)</sup>.

2Multiple cystic lesion of variable sizes<sup>(31)</sup>.

3Not clearly defined renal outlines<sup>(50)</sup>.

4With bilateral renal involvement, the bladder will be atrophied and not visualized<sup>(42)</sup>.

5Oligohydramnios signals a grim prognosis<sup>(31, 50)</sup>.

6The contralateral kidney often shows congenital abnormality as hydronephrosis<sup>(42)</sup>.

It may be difficult to distinguish MCDK from congenital hydronephrosis in utero by Ultrasound<sup>(31, 30, 42)</sup>. But retrograde urography and percutaneous cyst puncture after birth may solve the problem<sup>(30, 42)</sup>. However, the radionuclide renal scanning is probably the most effective means to identify salvageable renal parenchyma<sup>(30)</sup>. MCDK may increase in size in utero then decrease in size, this size variance may be related to the function<sup>(34,12)</sup>. The color doppler Ultrasound used now in the differential diagnosis of unilateral cystic kidney abnormalities as MCDK (Multicystic dysplastic kidney) shows elevated resistive index (R.I.) (R. I.: 90-100%) but hydronephrosis shows slightly elevated R.I. (R.I.: 83%)<sup>(45)</sup>.

### (4) Hydronephrosis:

It is the most common cystic disease of the kidney which can be diagnosed by Ultrasound antenatally<sup>(42)</sup>. It may be unilateral or bilateral<sup>(42)</sup> its sonographic features:

1large dilated extrarenal pelvis<sup>(53)</sup>.

2Calyceal dilatation appears late<sup>(53)</sup>.

3Progressive narrowing of renal substance<sup>(53)</sup>.

4Oligohydramnios is not usually an associated feature<sup>(42)</sup>.

In 1975, Pais and Retik, showed that massive dilatation of the urinary tract in neonates resolved spontaneously in some cases<sup>(57)</sup>. Homsy (1986) in his study, recommended 3-6 month observation period for patients with hydronephrosis secondary

to ureteropelvic junction anomalies<sup>(57)</sup>. In ureterovascular hydronephrosis: the pelvis, an angulated upper segment of the ureter, and the blood vessel are entangled to produce hydronephrosis<sup>(22)</sup>. Ultrasound can play a role in the management of patient with hydronephrosis<sup>(29)</sup>

### Conclusions

- (1) Ultrasound is the most convenient diagnostic tool in the diagnosis of most of the renal malformation as MCDK (multicystic dysplastic kidney)<sup>(9)</sup> renal agenesis medullary is cystic disease and congenital hepatic fibrosis with renal ectasia<sup>(4)</sup>.
- (2) In the diagnosis of kidney cysts, it is recommended to begin with Ultrasound with thermography, in difficult cases use C.T.<sup>(41)</sup>
- (3) Ultrasound followed by nuclear scintigraphy, appears to be logical diagnostic sequence in the evaluation of neonates with uncharacterised flank mass<sup>(30)</sup>.
- (4) Gray scale Ultrasound has permitted a rapid identification of renal and hepatic changes in children of affected parents with dominant PKD and permitted genetic counseling of the children prior to procreation<sup>(5)</sup>.
- (5) Ultrasound, since it is independent of renal function, is an even sensitive indicator of urinary obstruction. It can be used as a good evaluator for the unilateral nonvisualized kidney by I. V.U.<sup>(29)</sup>
- (6) Differentiation of cystic renal enlargement in utero due to hydronephrosis from MCDK can not be done by Ultrasound and replaced by percutaneous cyst puncture<sup>(42)</sup>.
- (7) Ultrasound and nephrotomogram are accurate methods for the diagnosis of dominant PKD. However, the quick diagnosis and non-invasiveness make Ultrasound the procedure of choice for diagnosis, scanning and follow-up<sup>(54)</sup>.

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