بسم الله الرحمن الرحيم

" قالوا سبحانك لا علم لنا الا ما

علمتنا انك أنت العليم الحكيم"

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Role of MRI in Evaluation of Urogenital Congenital Anomalies

Essay

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Introduction

Urogenital abnormalities are among the commonest disorders seen in the perinatal period and account for almost 20% of all prenatally diagnosed anomalies. In view of the common origin of large parts of the urinary and genital systems from the urogenital ridge, it is not surprising that malformations of the urinary and genital tract occur together at least as frequently as separately. In many instances it is important to discuss to which system a specific malformation belongs, as in hypospadias or a vesicovaginal fistula **(Jenny Yiee et al., 2008)**.

The most frequent genitourinary anomalies are renal, testicular and urethral, respectively; about 10% of the population has some kind of genital or urinary system anomaly. Although urogenital anomalies usually occur in isolation, these defects can form part of more complex syndromes or chromosomopathies. In addition, for reasons not explicable on a basis of development from the same rudiments, urogenital malformations are often associated with malformations elsewhere in the body **(K. Lakhoo et al., 2009)**.

Ultrasound study has a fundamental role in this investigation, since it is an accessible method of real-time diagnosis without the use of ionizing radiation, which is an important factor, especially in this pediatric age bracket. But this ability of US to detect fetal abnormalities is limited in cases such as maternal obesity, oligohydramnios, and in certain fetal positions. Thus MRI is indicated to diagnose these anomalies and to rule out associated

abnormalities. MR enables high-quality fetal images to be acquired regardless of the mother‘s physical condition or fetal position, respectively MRI notably avoids exposure to ionizing radiation **(C. Martín et al., 2004)**.

Fetal MR imaging often poses a diagnostic challenge for the radiologist. Both fetal anatomy and pathology differ decidedly from pediatric and adult MR imaging. Magnetic resonance imaging (MRI) is likely to emerge as the imaging modality of choice for children with complex genitourinary pathology being capable of acquiring both morphologic and functional data of the genitourinary system in one study. It combines characteristics appreciated in computed tomography (CT) and nuclear diuretic renal scintigraphy (DRS), such as excellent anatomic visualization and multiplanar three-dimensional reconstruction capability **(Thomas M. Dykes et al., 2007)**.

Many diagnostic techniques such as ultrasonography, intravenous urography, voiding cystourethrography, and radionuclide scintigraphy are used in assessing urogenital anomalies. However, it is not always possible to reach a definite diagnosis with a single imaging method. However, magnetic resonance imaging may be a useful adjuvant in circumstances that other conventional methods fall short. MRI reduces the need for radiation exposure and invasive procedures as a diagnostic imaging modality **(Martha Hanemann et al., 2009)**.

The aim of this study is to high light the role of MRI in evaluation of urogenital congenital anomalies.

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Pathology of urogenital congenital anomalies

The urinary Tract

Congenital anomalies of the kidney

About 10% of all people are born with potentially significant malformations of the urinary system. Renal dysplasias and hypoplasias account for 20% of chronic renal failure in children. Autosomal-dominant polycystic kidney disease, a congenital anomaly that becomes apparent in adults, is responsible for about 10% of chronic renal failure in humans. Congenital renal disease can be hereditary but is most often the result of an acquired developmental defect that arises during gestation. All except horseshoe kidney are uncommon **(Charles E. Alpers et al, 2004)**.

Disorder of the number of kidneys

Agenesis of the Kidney

Renal agenesis is failure of development of the renal anlage (uretic bud), resulting in a complete absence of the kidney. Renal agenesis may be bilateral or unilateral.

Bilateral renal agenesis is a rare anomaly resulting in death in utero or soon after delivery. Infants have renal failure associated with characteristic facial features: wide-set eyes and prominent inner canthi, a broad, flattened nose, large and low-set ears, and a receding chin (Potter facies). It is often associated with many other congenital disorders (e.g., limb defects, hypoplastic lungs) and leads to early death **(T.W. Sadler et al, 2006).**

Unilateral renal agenesis is more common, occurring in 0.1% of the population. Unilateral agenesis is compatible with normal life if no other abnormalities exist **(Fig. 4-1**). The opposite kidney is usually enlarged as a result of compensatory hypertrophy. Some patients eventually develop

progressive glomerular sclerosis in the remaining kidney as a result of the adaptive changes in hypertrophied nephrons **(Charles E. Alpers et al, 2004).**

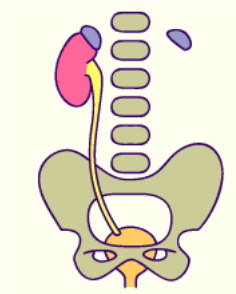

Fig. 4-1. Unilateral renal agenesis **(Hidaka S. et al, 2002).**

Too many kidneys (doubling)

This congenital anomaly is extremely seldom. Even if a doubling of the urinary tract occurs frequently, that of the kidneys is extraordinarily rare. Involved is an independent kidney with its own vascular supply, a capsule and its own urinary tract. The cause is a very early division of the ureter anlage before the invasion into the metanephric blastema **(Pohi M. et al, 2000).**

Abnormal kidney size

Hypoplasia

Renal hypoplasia refers to failure of the kidneys to develop to a normal size. This anomaly may occur bilaterally, resulting in renal failure in early childhood, but it is more commonly encountered as a unilateral defect. True renal hypoplasia is extremely rare; most cases reported probably represent acquired scarring due to vascular, infectious, or other parenchymal diseases rather than an underlying developmental failure. Differentiation between congenital and acquired atrophic kidneys may be impossible, but a truly hypoplastic kidney shows no scars and has a reduced number of renal lobes and pyramids, usually six or fewer. In one form of hypoplastic kidney,

oligomeganephronia, the kidney is small but the nephrons are markedly hypertrophied **(Charles E. Alpers et al, 2004)**.

Aplasia. In an aplastic kidney, a fibroused kidney anlage with its own derivates of the mesonephric duct (Wolffian duct) is present. It represents the extreme form of a renal dysplasia and differs from agenesia, in which absolutely no kidney anlage exists.

Disorder of the ascent of the kidneys

Ectopic Kidneys

The development of the definitive metanephros may occur in ectopic foci, usually at abnormally low levels. These kidneys lie either just above the pelvic brim or sometimes within the pelvis **(Fig. 4-2**). They are usually normal or slightly small in size but otherwise are not remarkable. Because of their abnormal position, kinking or tortuosity of the ureters may cause some obstruction to urinary flow, which predisposes to bacterial infections **(Clive R. Taylor et al, 2001).**

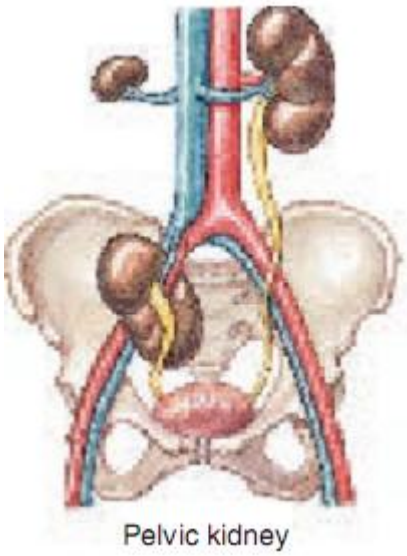

Fig. 4-2.Pelvic kidney **(Quoted from Van De Graaff, Human anatomy, 2001).**

Horseshoe Kidneys

Fusion of the upper or lower poles of the kidneys by a broad band of renal tissue produces a horseshoe-shaped structure that is continuous across the midline anterior to the great vessels **(Fig. 4-3**). This anatomic anomaly is common and is found in 1/600 of individuals, 90% of such kidneys are fused at the lower pole, and 10% are fused at the upper pole. The ureters pass anterior to the isthmus of the horseshoe kidney and may be narrowed. Most patients are asymptomatic; there is a higher incidence of urinary infection and renal calculi **(Charles E. Alpers et al, 2004)**.

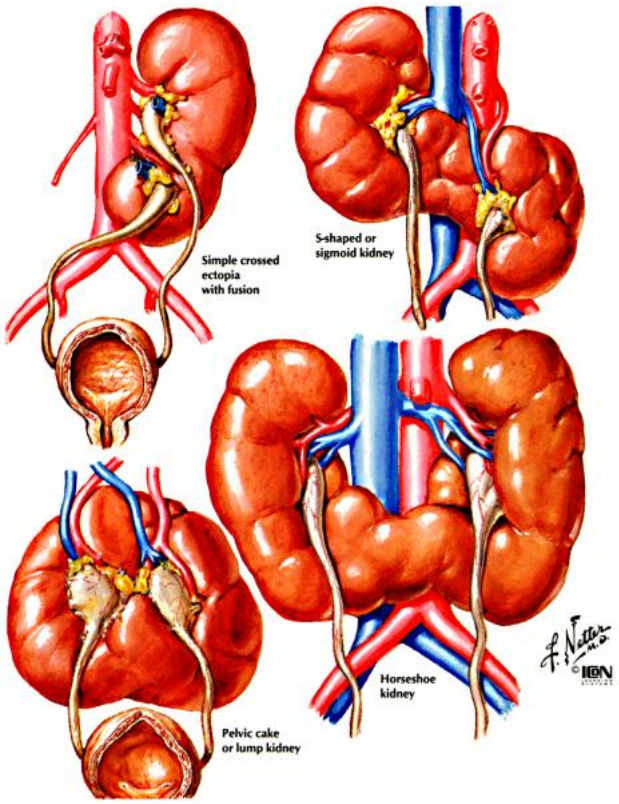

Fig. 4-3.Congenital anomalies involving the kidneys **(Quoted from Netter's Clinical Anatomy, 2005).**

Crossed ectopia a kidney migrates to the other side. Its ureter crosses the midline and inserts normally into the bladder. The crossed ectopia can occur unilaterally or bilaterally. In the case of a unilateral crossed ectopia a fusion of

the two kidneys often occurs. Ectopias are normally asymptomatic **(Hidaka S. et al, 2002)**.

Structural anomalies

Polycystic kidneys

A **renal dysplasia** must be distinguished from a simple hypoplasia in which the histological structure is normal. Renal dysplasia is characterized by a congenital anomaly of embryonic renal tissue development. The kidneys develop large epithelial cysts that are localized in the renal parenchyma and lead to the loss of the functional tissue, which can end in renal insufficiency **(Fig. 4-4**). The cysts are fluid-filled spaces, usually formed by dilation of some part of the tubule that has lost its communication with the rest of the tubule. Most of the cysts are lined by flattened tubular epithelium **(Bhatnagar V. et al, 2002).**

One distinguishes among various forms of dysplasia:

The autosomal recessive polycystic disease is a rare with an incidence of 1/50'000 births, manifested as severe renal failure in infancy. The cut surface of the kidney shows innumerable radially oriented fusiform cysts lined by cuboidal epithelium. There is no normal renal parenchyma. Infantile polycystic disease is believed to result from failure of communication between the nephron and the pelvicaliceal system during development.

The **autosomal dominantly inherited renal polycystic dysplasia** is more common (1/500 to 1/1,000 births) but less progressive than the autosomal recessive disease **(T.W. Sadler et al, 2006)**.

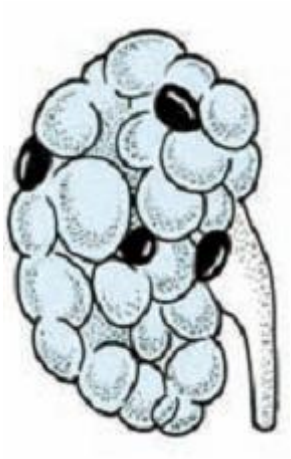

Fig. 4-4. Polycystic kidney **(Harold Ellis, Clinical Anatomy, 2006).**

The **multicystic renal dysplasia** is an anomaly that normally appears on only one side (in contrast with polycystic kidneys which are always found on both sides). The bilateral form is not compatible with survival. The cause of this illness is a differentiation disorder of the fetal kidney, leading to an atrophy of the renal parenchyma, which is replaced by cysts with a yellowish fluid. The ureter is atretic, the vessels are thin and the kidney consists only of a mass of cysts. Usually, over time, the cysts atrophy and for this reason no treatment is required. When the other kidney is normal, this has no consequences **(Hidaka S. et al, 2002)**.

Vascular anomalies of the kidney

Accessory renal arteries are common; they derive from the persistence of embryonic vessels that formed during ascent of the kidneys **(Fig. 4-5)**. These arteries usually arise from the aorta and enter the superior or inferior poles of the kidneys **(T.W. Sadler et al, 2006)**.

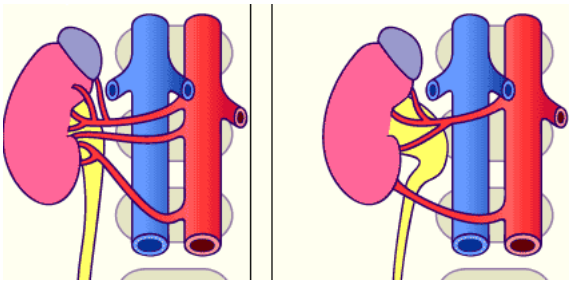

Fig. 4-5. Accessory renal arteries **(Bhatnagar V. et al, 2002).**

Congenital Diseases of the Ureters

Congenital anomalies of the ureters occur in about 2% or 3% of all autopsies. Although most have little clinical significance, certain anomalies may contribute to obstruction to the flow of urine and thus cause clinical disease.

Course anomalies of the ureter

Retrocaval retroiliac ureter

In this abnormality the right ureter traces out an "S" at the L4 level behind the vena cava (retrocaval ureter) **(Fig. 4-6)**. Responsible is a developmental disorder of the inferior vena cava. In a similar fashion the ureter can also run behind the common iliac artery at the L5 level (retroiliac ureter) **(Pohi M. et al, 2000)**.

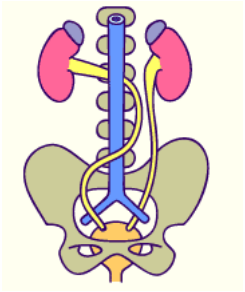

Fig. 4-6. Retrocaval/ retroiliac ureter **(Pohi M. et al, 2000).**

Abnormal number of ureters

Double ureters (derived from a double or split ureteral bud) are almost invariably associated either with totally distinct double renal pelves or with

the anomalous development of a large kidney having a partially bifid pelvis terminating in separate ureters **(Fig. 4-7)**. Double ureters may pursue separate courses to the bladder but commonly are joined within the bladder wall and drain through a single ureteral orifice. In rare cases one ureter opens into the bladder, and the other is ectopic, entering the vagina, urethra, or vestibule. The majority of double ureters are unilateral and of no clinical significance **(T.W. Sadler et al, 2006).**

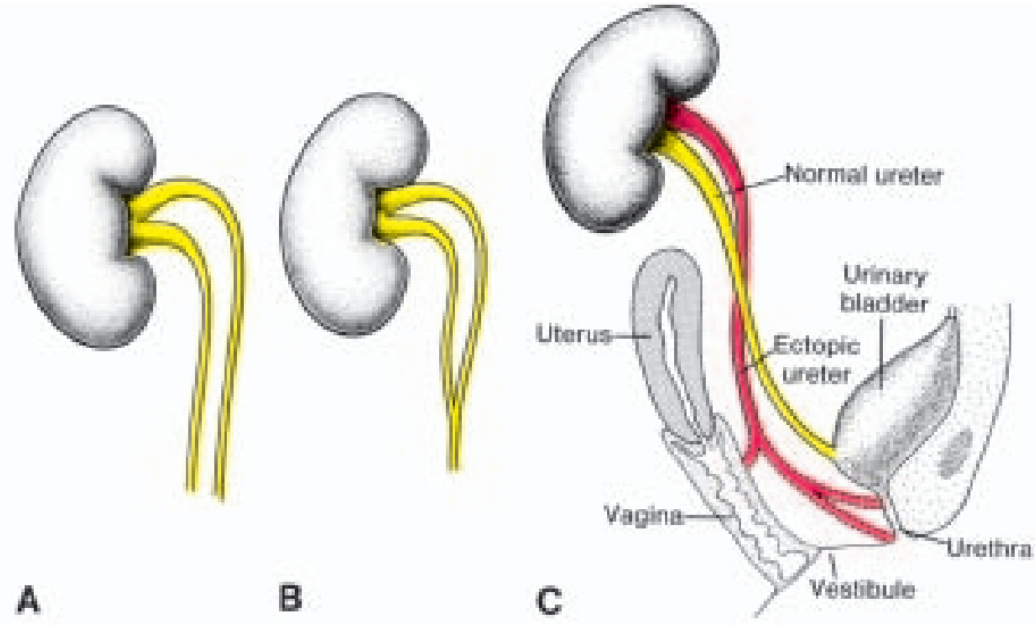

Fig. 4-7. A and B. A complete and a partial double ureter. C. Possible sites of ectopic ureteral openings in the vagina, urethra, and vestibule **(Quoted from Langman's medical embryology, 2006)**.

Anomalies of the ureteral diameter

Dilation **(hydroureter)**, elongation, and tortuosity of the ureters may occur as congenital anomalies or as acquired defects. **Congenital hydroureter** is thought to reflect some neurogenic defect in the innervation of the ureteral musculature. Massive enlargement of the ureter is known as **megaloureter** and is probably due to a functional defect of ureteral muscle **(Fig. 4-8)**. Hydronephrosis and decreased renal function results if the lesion goes