

ROLE OF DIAGNOSTIC IMAGING IN THE DIAGNOSIS
OF CONGENITAL UROPATHIES

Thesis submitted for the partial fulfilment of
the Master Degree in Radiodiagnosis

by

KARIMA MOUSTAFA MAHER
(M.B., B.Ch.)

616.0757
K.M

SUPERVISORS

44032

Dr. NAWAL ZAKARIA

Professor of Radiodiagnosis (Ain Shams University)

Dr. AHMED EL DORRY

Assistant Professor of Radiodiagnosis (Ain Shams University)

FACULTY OF MEDICINE
AIN SHAMS UNIVERSITY

1992



**To Mahmoud,
our smile and sunshine**



Acknowledgement

During the course of this work I was offered a lot of help. As I would have never been able to accomplish my thesis without this assistance, I feel, that the least I can do to express my thanks and deep gratitude, is to mention them by name.

First of all I would like to mention Prof. Dr. Nawal Zakaria, Professor of Radiodiagnosis, Ain Shams University, for her continuous encouragement, valuable comments and never ending patience throughout this work. I would also like to thank Dr. Ahmed El Dorry, Assistant Professor of Radiodiagnosis, Ain Shams University, for his support.

Dr. Anne Hollman, Consultant Radiologist at the Royal Hospital For Sick Children in Glasgow, provided me with most of the best references in this thesis and taught me a lot of the techniques advised in this work. She did not only provide me with scientific support, but also with inestimable moral encouragement. I do owe her a lot. I would also like to thank Dr. Ruth Mac Kenzie, Consultant Radiologist at the same hospital and all the staff and personnel of the Department of Radiodiagnosis at the Royal Hospital For Sick Children.

My mother has taken the burden of typing most of my work upon her shoulders and spent a lot of effort in the lay-out and helping me print it, as well as my father, who has assisted me in everything concerning the Arabic language. Last but not least, I mention my husband, Hesham El Safoury, Assistant Lecturer of Pediatric Surgery, who has provided me with a lot of references and photos, he took himself.

I wish to thank all my colleagues and friends who have also participated either actively or with advice.

C O N T E N T S

	Page
I. Introduction And Aim Of The Work	1
II. Anatomical Considerations	2
a) Embryology Of The Genitourinary Tract	2
b) Anatomy Of The Genitourinary Tract	10
III. Pathological Considerations	24
IV. Material And Methods	48
V. Results And Illustrated Cases	59
VI. Discussion	109
VII. Summary And Conclusion	131
VIII. References	134
IX. Arabic Summary	143

I. INTRODUCTION AND AIM OF THE WORK

CONGENITAL UROPATHIES are congenital disorders of a certain part of the genitourinary tract, namely the parts responsible for draining the urine: the pelvicalyceal system, the ureters, the urinary bladder and the urethra.

With the improvement of scanning techniques and the increased awareness of both pregnant mothers and obstetricians to the importance of routine sonographic evaluation of the fetal outcome, a remarkable rise in the incidence of prenatally diagnosed congenital uropathies has been reported. Prenatal diagnosis of congenital malformations of the genitourinary tract is now considered a routine. Michael Carr and James Mandell, 1992, have reported fetal hydronephrosis in 0,2% of all prenatal ultrasonographic scans.

The incidence of ureterovesical junction obstruction rose sharply from 8% in the pre-ultrasound era (1947-1977) to 23%, while that of pelviureteric junction obstruction rose from 22% to 41% (Lebowitz, R.L.; Mandell, J., 1986). This has offered the chance for early intervention and treatment, before it is too late.

The aim of this work is to evaluate the role of different diagnostic procedures in the perinatal diagnosis of congenital uropathies and in what way the timing affects the prognosis of different cases.

II. ANATOMICAL CONSIDERATIONS

a) Embryology Of The Genitourinary Tract

In the course of this thesis a variety of imaging modalities for congenital uropathies will be discussed. For the proper understanding of the subject and accurate interpretation of the images, a brief review of the embryology of the genito-urinary tract is first given, emphasizing on details of the points which are important in the pathogenesis of the diseases dealt with later.

The nephric system develops progressively as 3 distinct entities: pronephros, mesonephros, and metanephros (which gives the ureteral bud)

Pronephros

During the fourth week of development, condensed mesoderm adjacent to the midline divides into blocklike units, the somites. The pronephros appears in the cervical region of the 10-somite embryo between the second and sixth somites (Potter, 1972).

The pronephros consists of 6-10 pairs of tubules, it is a vestigial structure that disappears completely by the fourth week of embryonic life (Emil A Tanagho, 1975).

Mesonephros

The mesonephric duct descends from the cervical somites to the caudal region of the embryo. During its descent, the mesonephric duct induces about 40 pairs of tubules and becomes in contact with the urogenital sinus, into which it later drains (Potter, 1972).

By 37 days the mesonephroi are fully developed, and they appear as paired retroperitoneal organs close to the midline that bulge into the peritoneum.

After 10 weeks of development, many mesonephric nephrons degenerate (Stephens, 1983), but some tubules persist and become incorporated into the genital duct system.

Development Of The Ureteral Bud

After the mesonephric duct reaches and drains into the urogenital sinus, the ureteral bud appears. The bud originates as a diverticulum from the posteromedial aspect of the mesonephric duct at the point where the end of the duct bends to enter the cloaca. The ureteral bud is thought to pass further through four periods during its development.

Period I (5th-14th week)

Asynchronous branching of the ampullae helps maintain the reniform shape of the kidney (Hamilton and Mossman, 1976). This primitive network of ureteral bud branches dilates and creates the appearance of the pelvis and calyces. Because the dilatation is not uniform, the appearance of the pelvis and calyces in the newborn may be varied.

The subsequent three to five generations of branchings contribute to the calyces. The next five to seven generations of branchings of the ampulla of the ureteral bud create the collecting tubules.

Period II (15th-22nd week)

During this period the ampullae divide infrequently and a family of nephrons is induced by a single ampulla. As each ampulla extends, a family of about four (range of two to eight) nephrons is laid down in an arcade manner.

The central zone, the medulla, contains the mass of collecting tubules; the peripheral zone, the cortex, contains the nephrons, which are attached to collecting tubules.

Period III (22nd-36th week)

This period of development creates nephrons whose glomeruli lie in the outer half of the cortex. About 75 % of the complement of glomeruli are induced during Periods 2 and 3 of development.

Period IV (32nd-36th week to adulthood)

During this period the collecting ducts elongate, the proximal tubules convolute, and the loops of Henle penetrate deeper into the medulla. A typical collecting duct drains about nine to eleven nephrons.

Nephron induction by a branching ureteral bud causes the surface of the kidney to appear lobular at birth (Hamilton and Mossman, 1976).

Ascent Of The Kidney

The renal blastema originates at the level of the upper sacral segments.

The final position of the kidney at the level of the upper lumbar vertebrae is attributed to ascent of the renal blastema.

Active elongate growth of the ureter into the metanephrogenic blastema causes ascent in the pelvis. The bud migrates cranially and dorsally, and the kidney reaches the umbilical artery. That is when the intrinsic growth and moulding of the renal parenchyma hurdles the kidney over the umbilical arteries.

By 58 days the kidney fixes to the tissues of the retroperitoneum, which permits axial growth of the spine to elevate the kidney to its final position. In addition to scaling the retroperitoneum, the kidney also rotates around its longitudinal axis to cause the pelvis to face the spine (Friedland and De Vries, 1975).

Development Of The Ureter

The ureteral bud originates from the mesonephric duct after 28 days of development, it then elongates as a hollow tube to ascend the pelvis. The patency of the ureter changes during development. This patency may relate to the hydrostatic pressure generated by the mesonephric urine, which fills the ureter since the cloaca is still imperforate at this stage. Intense elongate growth of the ureter may obliterate the lumen. After 40 days, the lumen is again apparent along the entire length of the ureter. These observations may help understand the genesis of congenital strictures at the ureteropelvic or ureterovesical junctions, as the lumen at these sites becomes patent last.

With further development however, the ureter may elongate in excess of that needed to accompany ascent of the kidney. To absorb the excess length, the ureter may become tortuous or invaginate its wall as pleats, called the "fetal folds" of Ostling (Stephens, 1983).

Postnatally, the infant's growth rate exceeds that of the ureter, so the ureter may lose its tortuosity and unfolds its pleats. These pleats are not ordinarily obstructive. However, **a pleat that intrudes into the lumen of the ureter and that is fixed by the ureteric adventitia may obstruct urine drainage as a valve (Maizel's and Stephens, 1980).**

Development Of The Cloaca

During the period of the laminar embryo, the cloacal membrane is in the caudal region of the embryo (where ectoderm opposes endoderm without any intervening mesoderm). Growth of mesenchyme near the cloacal membrane lifts the tail end of the embryo away from the blastoderm. This lifting creates a chamber, the cloaca, a dilated terminal portion of the hindgut.

Further growth of the tail fold flexes the tail end of the embryo so that the cloacal membrane lies on the ventrum of the embryo. Growth of mesenchyme lateral to the cloacal membrane elevates the tissue as labioscrotal swellings; growth of mesenchyme cranial to the cloacal membrane becomes confluent as the genital tubercle, the primordium of the phallus.

Septation of the cloaca begins at about 28 days (Stephens, 1983). The urorectal septum extends in the coronal plane toward the cloacal membrane. At the same time, Rathke's plicae appear as two tissue folds from the lateral aspects of the hindgut. These plicae meet each other in the midline to complete the septation of the cloaca. A separate rectum and primitive urogenital sinus appear by 44 days of development.

The portion of the primitive urogenital sinus cranial to the mesonephric ducts is the vesicourethral canal, that caudal to the mesonephric ducts is the definitive urogenital sinus (Hamilton and Mossman, 1976).

Development Of The Trigone And Bladder

By 28 days of development, the mesonephric ducts have already reached and fused with the urogenital sinus. At this time the ureter originates from the mesonephric duct.

The segment of the mesonephric duct distal to the site of origin of the ureteral bud dilates as the common excretory duct, the precursor of the trigone.

The right and left common excretory ducts are absorbed into the urogenital sinus with the orifice of the duct migrating caudally and the orifice of the ureteral bud migrating cranially.

Continued growth of the epithelium and mesoderm of the absorbed common excretory duct separates the orifices of the ureters laterally and establishes the framework of the primitive trigone.

The mesonephric ducts also migrate caudally and flank the paramesonephric ducts at the level of the urogenital sinus. This is the site of the future verumontanum.

The period when the ureter attains the oblique submucosal course of the newborn bladder is uncertain, but occurs after 14 weeks.

By 10 weeks, the bladder is a cylindrical tube lined by connective tissue. The wall of the bladder over the trigone is twice as thick as elsewhere. But by the 13th week rapid muscularization causes the wall of the trigone to become 5 times as thick as elsewhere.

The apex of the vesicourethral canal tapers as the urachus.
(Arey 1974)

The bladder lumen narrows at the internal vesical sphincter.

The internal sphincter forms from a mass of circular fibres that surrounds the neck of the bladder.

Development to term results in further increase in the size of the muscles of the detrusor, trigone, and sphincter.

As imaged by ultrasonography, the fetal bladder varies in size, emptying about 16 ml every 90 minutes near term (Abramovich et al., 1979).

Development Of The Urogenital Sinus

By 6 weeks of development the urogenital sinus is apparent. The portion of the sinus near the bladder is narrow (pelvic urethra), and the portion near the urogenital membrane is expanded (phallic urethra).

Between 5 and 10 weeks, the phallic portion of the definitive urogenital sinus is sexually indifferent. After 10 weeks of development, the external genitalia of the male and female appear different.

Male Urethra:

The phallic portion of the urogenital sinus develops into the bulbar and penile urethra.

The genital tubercle grows and elongates into a cylindrical phallus. The endodermal edges of the secondary urethral groove fuse to tubularize the penile urethra. The ectodermal edges of the groove fuse as the median raphe.

The scrotal swellings round, migrate caudally and fuse to form the scrotum at the base of the penis.

During the fourth month: the glandular urethra appears. First, the glandular ectoderm proliferates to create a plug of tissue; next, the plug burrows into the glans to meet the penile urethra; and finally, the ectodermal plug cavitates as the glandular urethra.

The glandar ectoderm disintegrates similiarly to the urethral plate ectoderm. The pelvic part of the urogenital sinus develops into the lower portion of the prostatic urethra and the membranous urethra (Hamilton and Mossman, 1976).

Female Urethra:

The pelvic part of the urogenital sinus develops into the lower portion of the definitive urethra and vagina (Hamilton and Mossman, 1976).

The phallic portion of the urogenital sinus remains as a vestibule because the urethral plate does not extend as far to the genital tubercle as in the male.

The Urethral Valves

The Wolffian (mesonephric) ducts, which enter the anterior wall of the cloaca, recede to the level of the verumontanum (an elevation of the posterior wall of the prostatic urethra where the seminal ducts enter) in the posterior wall of the urinary compartment of the newly divided cloaca. Hence, the posterior wall of the urethra has two normal folds, called the "urethrovaginal folds" or "plicae colliculi".

They are considered remnants of the cephalad migration of the wolffian ducts and extend longitudinally from Müller's tubercle (a prominence located between the entrance of the two müllerian ducts in the urogenital sinus) to the origin of the Cowper or Bartholin glands. So urethral valves are of heterogenous embryologic origin.

Some valves (type I) seem to result from an exaggerated development of the urethrovaginal folds with an abnormal insertion of the distal end of the wolffian ducts. Other valves (type III) develop because of abnormal canalization of the urogenital membrane. (Kelalis PP, et al., 1985).

b) Anatomy Of The Genitourinary Tract

The kidneys are paired, retroperitoneal, bean shaped organs lying on each side of the vertebral column.

The surface of the kidney is invested by a thin but strong fibrous capsule. External to it is a considerable quantity of fat tissue known as the adipose capsule [Gerota's capsule] (Meschan I., 1975). All surfaces are usually smooth and convex though traces of lobulation, normal in the foetus, are often seen.

The kidneys lie high up on the posterior abdominal wall behind the peritoneum, largely under cover of the costal margin. Each kidney lies obliquely, with its long axis parallel with the lateral border of psoas major (Last R.J., 1986).

The right kidney is usually slightly smaller than the left. Discrepancies between the two kidneys of greater than 1 cm are of considerable significance.

The mean kidney length in children varies from 6 cm with a body height of 24 inches to 12 cm with a body height of 72 inches.

The size of the kidney depends largely on the size of the child (Meschan I., 1975).

Each kidney moves in a vertical range of almost 1 inch (2 cm) during full respiratory excursion of the diaphragm (Last R.J., 1986).

A maximal excursion of 5 cm (or 1,5 vertebral bodies) occurs in the change from the recumbant to the erect position (Meschan I., 1975).

The medial border of the kidney is concave and contains a slitlike aperture, the hilus. This is the orifice of a cavity called the renal sinus and contains: