

REVIEW
ON
CONGENITAL ANOMALIES OF UPPER
URINARY TRACT

Submitted in Partial Fulfilment
For the Degree of M. Sc.
(UROLOGY)



616-63

E. H.

By

EDRIES HUSSEIN ASKALANY
M. B., B. Ch. (Assiut Univ.)



Supervised by

Prof. Dr. ALY MAGED

Professor of Urology

Faculty of Medicine, Ain Shams University

Faculty of Medicine

Ain Shams University

1984

ACKNOWLEDGEMENT

I would like to express my deepest gratitude to Prof. Dr. ALY MAGED , the Head of Urology Department, Faculty of Medicine Ain Shams University for his kind guidance, sincere help, fatherly encouragement and supervision throughout the course of this work.

EDRIS HUSSEIN

1984



C O N T E N T S

	Page
INTRODUCTION.....	1
EMBRYOLOGY OF URINARY TRACT.....	2
ANATOMY OF UPPER URINARY TRACT.....	17
CLASSIFICATION OF RENAL ANOMALIES - ANOMALIES OF NUMBERS.....	35
ANOMALIES OF VOLUME AND STRUCTURE.....	45
ANOMALIES OF ASCENT.....	100
ANOMALIES OF FORM AND FUSION.....	106
ANOMALIES OF RENAL VASCULATURE AND CONGENITAL HYDRONEPHROSIS.....	117
URETERAL ANOMALIES.....	131
Anomalies of Number.....	133
Anomalies of Structure.....	140
ANOMALIES OF TERMINATION AND POSITION.....	148
SUMMARY.....	165
REFERENCES.....	166
ARABIC SUMMARY.	

INTRODUCTION

Congenital anomalies of urinary tract are not uncommon the commonest malformation are the bifid pelvis, duplicated ureter (occur in 4 %), ectopia and anomalies of renal vessels, the presence of a third kidney is very rare. The anomalies may be on both sides such as polycystic kidney. The anomalies of upper U.T. may associated with other anomalies such as unilateral renal agenesis Hypoplasia are often seen in association with imperforate anus some of these anomalies are of nosignificance and does not lead to complications, on other hand some cases produce or predispose to serious complication such as infection, stone formation and obstruction, some are even incompatible with life as in bilateral renal agenesis others of polycystic kidneys always end by renal failure.

Therefore review of congenital anomalies of upper U.T. is very important because many of such abnormalities can be corrected and its complications can be treated, but others can not be treated except by removal of the affected organ such as complicated ectopic kidney.

EMBRYOLOGY OF URINARY TRACT

EMBRYOLOGY OF URINARY SYSTEM

The intra-embryonic mesoderm of each side of the embryo was described as separating into a medial, paraxial mass which becomes segmented to form the somites; lateral plate which remains unsegmented and forms the lining of the coelom; and a segmented junctional region, the intermediate mesoderm lying between and connecting these two parts. By longitudinal fusion this intermediate mesoderm gives rise to a nephrogenic cord from which most of the excretory system develops.

Embryology of Kidney:

The nephric system develops progressively as three distinct entities: pronephros, mesonephros and metanephros.

Human pronephros = Before kidney

The human pronephros is vestigial, it consists of several pairs of rudimentary pronephric tubules arising as dorsolateral sprouts from the longitudinally fused nephrotomes (the nephrogenic cord) of each side. The total extent of the pronephrosis in the region of

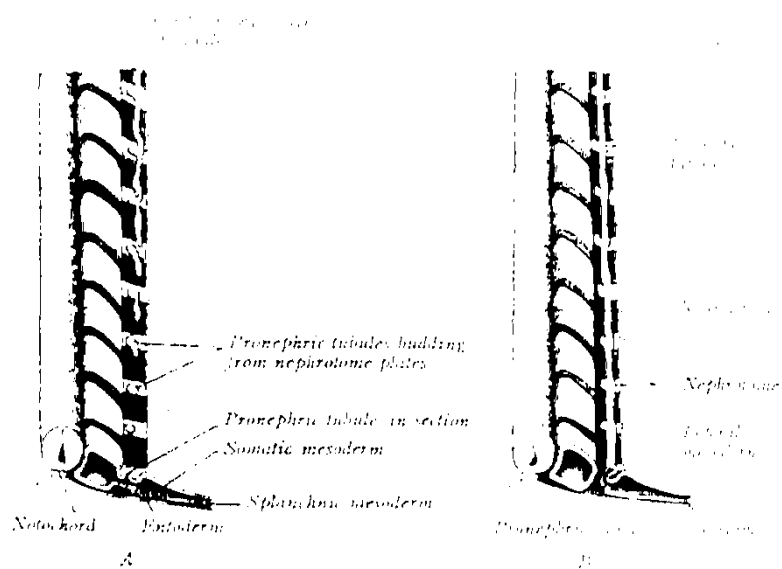


Fig. 1: Development of the pronephric system.

junction between the future neck and thorax. All the tubules are not present at the same time, for most cephalic have appeared, differentiated and disappeared the caudal ones are in the process of development. The first pair appears at nine somite stage. Each tubule fuses with the succeeding one so a longitudinal collecting duct is formed (pronephric or wolffian duct). The wolffian duct opens into the cloaca. The pronephric tubules communicate medially with the coelomic cavity by way of a ciliated nephrotome, laterally they open into the pronephric duct. The degeneration of pronephric tubules is complete at about 5mm stage but the main duct persists and becomes the mesonephric duct.

Mesonephros: Wolffian body - middle kidney:

It is larger than the pronephros, not only does it contain more tubules, but also these are longer and more complicated. It is located farther caudal. The first pair of mesonephric tubules makes its appearance in the 2.5 mm stage at the level of the thirteenth somite. From this point, new tubules appear in a cephalic direction reaching the nine somite level at the 5mm stage, and in a caudal direction reaching the twenty-sixth somite level at the 7.5mm stage. The tubules begin to take origin from spherical masses of

cells brought into existence by subdivision of nephrogenic cord. Each mass hollows and the vesicle, so formed, sends out a solid extension which unites with the mesonephric duct nearby to complete a mesonephric tubule there is further canalization, growth with S-shaped bending. The free end of the tubule enlarges and becomes thin walled. When a knot of blood vessels indents one side Bowman's capsule is formed. Capsule and glomerus together comprise a unit known as the mesonephric corpuscle. Next in order comes a thicker, lighter staining secretory segment of the tubule and then a thinner, darker - staining collecting segment which, in turn, opens into the mesonephric duct.

When the developing tubules begin to enlarge there is no room for them in the body wall and they bulge ventrolaterally into the coelom. On each side of the dorsal mesentery there is thus produced a longitudinal urogenital ridge soon after its formation this common fold subdivides longitudinally into a more medial genital ridge and a lateral mesonephric ridge.

During the fifth week the cranial nephrons start degenerating while the caudal nephrons are still differentiating. Remnants of a number of the caudal mesonephric tubules, however do persist to become part of

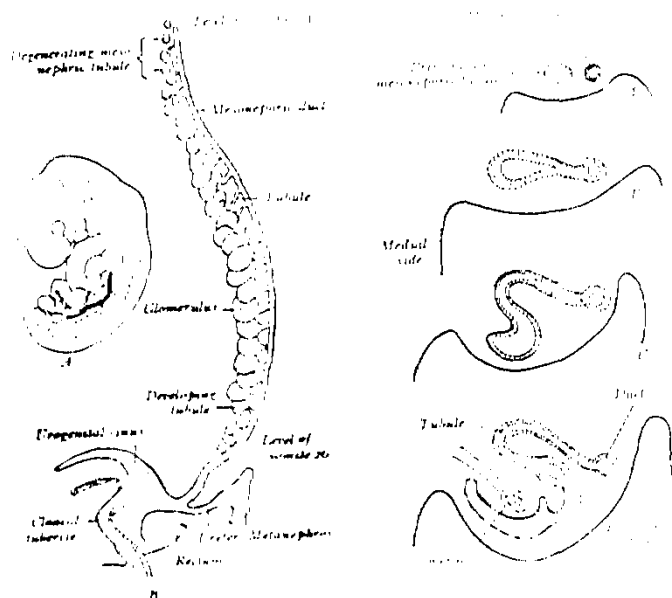


Fig.2: Location, composition and stages in the differentiation of a human mesonephros.

the duct system of the testis, these persistent tubules may be divided into a cranial, epigenital group and a caudal, paragenital group. In the epigenital portion the number of tubules varies from (5-12) and these excluding the first two form the efferent ductules of the testis, the first two epigenital tubules persist as the (superior aberrant ductules). Some of the lower paragenital tubules become separated from the mesonephric duct to form the paradidynis in the male and the paroophoron in the female. Those which remain attached to the duct become (the inferior ductules). The main or the mesonephric duct is transformed into the (vas deferens).

Metanephros = After kidney = third kidney:

It has a dual origin. The distal or collecting portion develops from a diverticulum of the mesonephric duct, while the proximal or secretory part develops from the nephrogenic cord caudal to mesonephros. The ureteric primordium arises at the 4-5mm stage from the dorsal and medial portion of the mesonephric duct just before it joins the cloaca. The ureteric bud as soon as it is formed it encounters the caudal end of the nephrogenic cord which is now called the "metanephrogenic blastema". The first change that occurs in

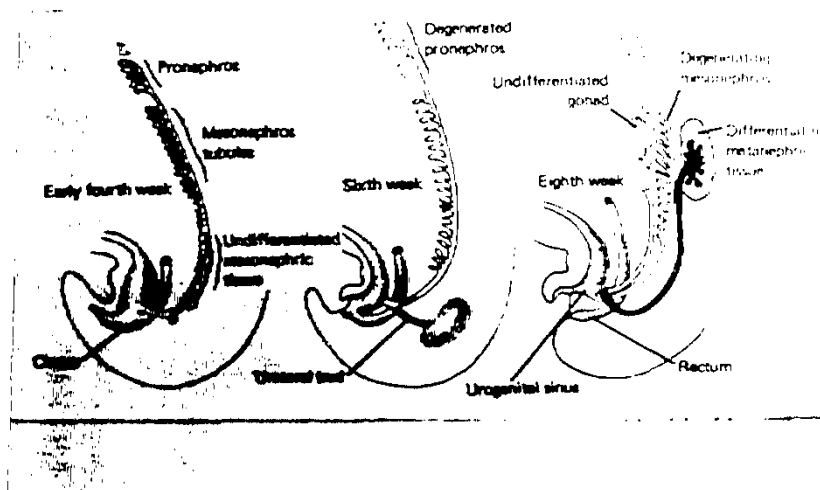


Fig.3: The development of Nephritic system.

the ureteric bud is a terminal flattening which results in the formation of a primary renal pelvis. At the 10mm stage, the primary renal pelvis gives rise to two renal tubules, one present in the cranial and one in the caudal portion. After short time two other tubules appear in the mid portion of the primary renal pelvis each primary renal tubule then forms a terminal ampulla from which a series of tubules arise in a dichotomous manner. The division is progressive so that by the 5th month 12 generations of tubules have formed and by 9th month 20 generations have appeared. The caudal and cranial primary tubules form the major calyces and the secondary tubules along with the third and fourth generations form the minor calyces, the 5th generation forms the papillary ducts of the pyramids, and those above the 5th generation form the medullary collecting tubules.

Metanephrogenic blastema:

The presence of the ureteric bud in the metanephrogenic tissue causes the latter to differentiate into two layers, an external which forms the connective tissue and capsule of the kidney and an internal which forms the secretory tubules. As the ureteric bud divides and subdivides the surrounding metanephrogenic

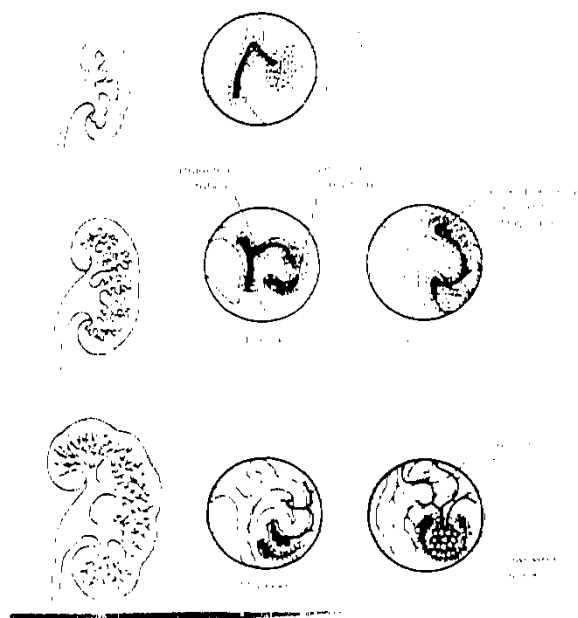


Fig.4: Progressive stages in the differentiation of the nephrons.