EMBRYOLOGY AND CONGENITAL ANOMALIES OF THE KIDNEY

AN ESSAY

SUBMITTED IN HARTIAL TULFILMENT OF THE MASTER DESCREE IN UROLOGY

BY

NADI SHAWKY EKLADIOUS

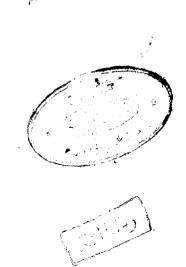
M.B., B.Cb.

SUPERVISED BY

Prof. Dr. IBRAHIM RAG.
Prof. & Hecd of
Department of Urology

FACULTY OF MEDICINE
AIN SHAMS UNIVERSITY

1987



ACKNOWLEDGMENT

I am greatly indebted to professor Dr. Ibrahim

Ragi , professor and head of depatment of Urology,

Ain-Shams University , for his faithfull supervision ,

guidance and encouragement throughout the complishment

of this work.

I also acknowledge with profound gratitude the cooperative collaboration and meticulous supervision of Dr. Shereen Ragi , lecturer of Urology , Ain-Shams University.



CONTENTS

		Page
-	INTRODUCTION	•• 1
-	EMBRYOLOGY OF THE KIDNEY	2
	* Pronephros	6
	* Mesonephros	8
	* Metanephros	21
-	CONGENITAL ANOMALIES OF THE KIDNEY	38
	A. Agenesis	
	1- Bilateral Renal Agenesis	41
	2- Unilateral Renal Agenesis	. 44
	B. Supernumerary Kidney	48
	* Anomalies Of Volume And Structure	
	A. Hypoplasia	51
	B. Renal Dysplasia:	62
	C. Polycystic Kidney	
	1- Infantile Polycystic Kidney	76
	2- Adult Polycystic Kidney	86
	D. Other Cystic Diseases Of The Kidney	86
	E. Medullary Sponge Kidney	87
	F. Medullary cystic Disease Of The Kidney.	89
	* Anomalies Of Ascent	
	A. Simple Ectopia	92
	B. Cephalad Ectopia	. 96

C. Thoracic Kidney 97

	¥	Ar	nomalies	of Form	and	Fusio	n		Page ———
		Α.	Crossed	l ectopia	with	and	withou+	fusion	. 99
			1- Unil	ateral f	used l	kidney	(inferio	or ectopia	.).101
			2- Sign	oid kidne	у		• • • • • • •		. 102
			3- Lum	kidney .					. 102
			4- L-sh	naped kid	ney				. 103
			5- Disc	e kidney	• • • • • •		• • • • • • •		. 103
			6- Unil	ateral f	used 1	kidney	(inferio	or ectopia	1).103
		В.	Horsesh	noe Kidne	у		• • • • • • • •		105
	¥	A	nomalies	of Rota	tion				
		Α.	Incompl	Lete					. 110
		В.	Excess	ive	• • • • • •			· · · · · · · · · · · ·	. 111
		С.	Reverse	· · · · · · · · ·	• • • • •				. 111
	¥	Ai	nomalies	of Rena	l Vas	culatu	re		
		Α.	Aberran	nt , acces	sory,	or mu	ltiple ve	essels	113
		В.	Renal A	Artery and	urysms	• • • • •			. 114
		С.	Arterio	ovenous f	istula	• • • • •	• • • • • • • • • • • • • • • • • • • •		. 116
	X	A	nomalies	of the	Colle	cting	System		
		Α.	Anomali	ies of Ca	alyx a	nd In	fundibul	am	. 119
		В.	Anomal	ies of t	he Re	nal F	elvis		126
_	SUI	VTVF A	RY						. 128
_									1) (
-	A.	RAB	IC SUMM.	ARY	• • • • • •		• • • • • • • •		. 147
-	H	RRA	TA	• • • • • • • • •			• • • • • • • •		. 148

INTRODUCTION

Abnormalities of embryogenesis account for many of the congenital anomalies of the urinary tract that require recostructive procedures.

An understanding of the embryogenesis of these anomalies is often helpful to the physician confronted with these congenital anomalies, and a brief discussion of the embryology and the commoner congenital anomalies of the kidney is appropriate (Steckel and John, 1977).

Nearly all types of congenital anomalies of the kidney are of clinical importance, and this is apparent when we consider the large number of individuals presenting for treatment who are found to have some such anomalies. (Badenoch, 1974).

EMBRYOLOGY

Origin :

The intra-embryonic mesoderm of each side of

the embryo (Fig. 1 - 1) was described as separating

into a medial, paraxial, mass which becomes segmented

to form the somites; a lateral plate which remains

unsegmented and forms the lining of the celom; and

a segmented junctional region, the intermediate

mesoderm, lying between and connecting these two

parts.

The intermediate cell mass (nephrotome) forms during the fourth week of gestation .

By longitudinal fusion this intermediate mesoderm gives rise to a nephrogenic cord (ridge) from which most of the exoretory system, transitory or permanent, develops (Hamilton and Mossman, 1972).

Each ridge extends from the region of the cervical somites right down to the caudal end of the embryonic celomic cavity. On the lateral and ventral aspects of the ridge a longitudinal duct develops. This is the mesonephric or wolffian duct.

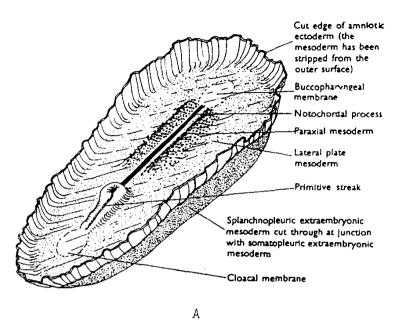
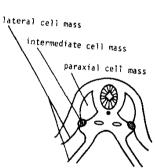
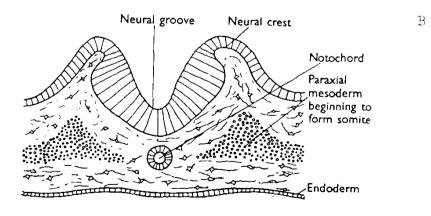


Fig. 1-1. The intraembryonic mesoderm of each side of the embryo is separated into paraxial, lateral, and intermediate cell masses (Beck et al., 1973).





-4-

In the male, it plays an important role in the development of the genital system and in both sexes its lower end will form part of the bladder (Beck et al., 1973).

Succesion :

The pattern of development of the urinary tract is unusual in that the appearance of the definitive excretory system is preceded by two "false starts" in the form of evolutionarily more primitive excretory systems, which begin to form then promptly undergo degeneration (James , 1976).

These three kidneys (Fig. 1-2) develop succesively and overlapping, one cauded of the other in the time and place order named (Arey, 1974).

The most cranial segments collectively constitute the pronephros, the intermediate segments, the mesonephros, and the most caudal segments, the metanephros or definitive kidney (Hamilton and Mossman, 1972).

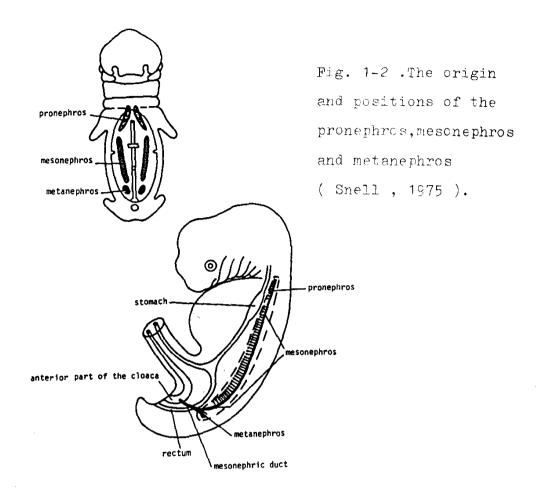
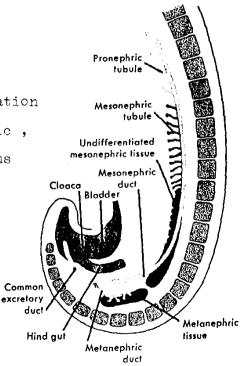


Fig. 1-3. Schematic representation of the development of the pronephric, mesonephric, and metanephric systems and related structures (James, 1976).



Comparison between the three stages :

All three bodies are made up of nephric tubules opening into a common excretory (wolffiam) duct (Fig. 1-3). While in the cervical region the tubules are simple and retain their segmental arrangement, in the dorsal and lumbar region they multiply in number and complexity; in the sacral region they become exceedingly numerous and massed around a diverticulum from the wolffian duct - which forms the primitive ureter (Keith, 1980) (Fig. 1-4).

PRONEPHROS

The condensed nephrogenic mesoderm opposite 7th to 14th somites, mesodermal segments, in the cervical region of the embryo becomes segmented to form a series of nephrotomes, each of which gives rise to two pronephric buds (Fig. 1-5). Each bud, in turn, grows dorsomedially (away from the celom) and may or may not canalize to form a pronephric tubule.

The formed tubule may open into the celom (by way of the nephrostome). The nephrostome is the

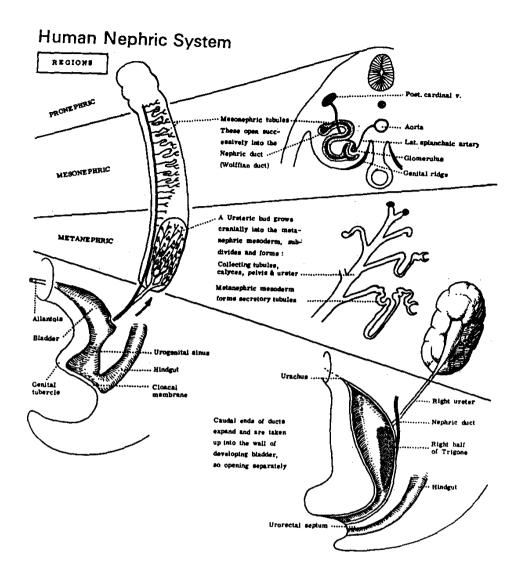


Fig. 1-4. Human nephric system regions in the cervical, lumbar, and sacral regions (Williams and Wendell, 1969).

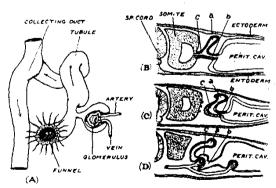


Fig. 1-5. Composition and A. Diagram of an isolated nephric tubule. Composition and Origin of Nephric Tubules.

- A. Diagram of an isolated reprire cubule.
 B. Showing the manner in which the intermediate cell mass (a, b, c) gives origin to the nephric tubule (a), peritoneal funnel (b) and the nephrocele (c).
 C. The isolation of these parts from the somite and their union to form a system.
 D. The origin of a glomerulus in the wall of the nephrocele (c).

(Keith, 1980).

- 8 -

cavity. Whereas the dorsal ends of adjacent buds or tubules join to form the pronephric duct.

An arterial tuft, linking aorta and postcardinal vein, projects into the celom and forms a primitive pronephric glomerulus (external glomerulus).

The pronephros of each side is rudimentary in the human subject (Snell , 1975).

MESONEPHROS

IN the second month of human development the wolffian body is at the hight of its development and in the later part of this month and in the early part of the third month is actually functional (excretory) (Keith, 1980).

During the 5th week the cranial nephrons start degenerating while the caudal nephrons are still differentiating (Hamilton and Mossman , 1972)(Fig. 1-6).

Fig. 1-6. The cranial nephrons start degeneration while the caudal nephrons are still differentiating (Arey, 1974).

