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Represented by

Abd El-Wahab Abd El-Aziz Abd El-Wahab Wafy

M. B. B. Ch.

Supervised by

Prof. Dr. Abd El-Wahab El-Kasaby

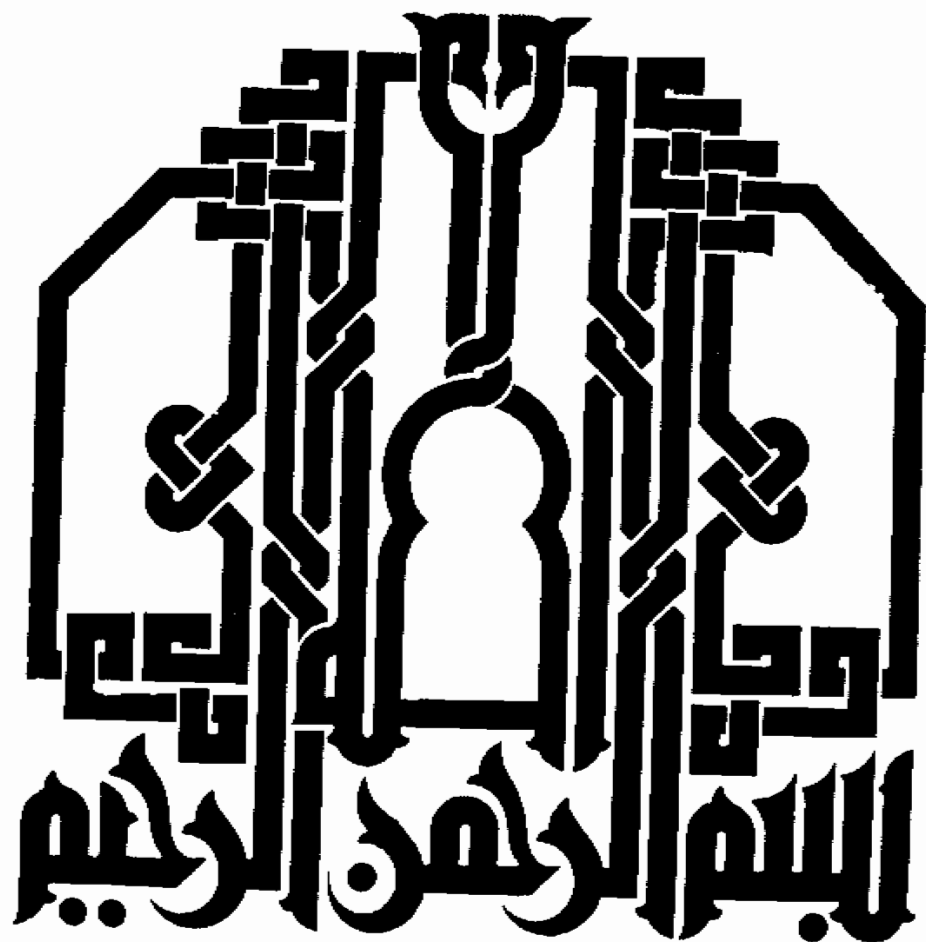
Professor of urology
Faculty of Medicine
Ain Shams Univeristy

Dr. Sherein Ragy

Lecturer of urology
Faculty of Medicine
Ain Shams Univeristy

Ain Shams Univeristy

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بِسْمِ اللَّهِ الرَّحْمَنِ الرَّحِيمِ



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INTRODUCTION

Bladder exstrophy is one of the most devastating anomalies, that affect the lower urinary tract, genital tract, bony pelvis and musculoskeletal system, there is a spectrum of associated anomalies of the osseous, soft tissue and gastro-intestinal systems. The classical bladder exstrophy is more common than the cloacal type, in which the infraumbilical portion of the abdominal wall, and the anterior bladder wall are absent with widely separated pubes and short epispadiac penis.

These anomalies, most probably result due to the wedge effect of the abnormal large or persistent cloacal membrane that prevents medial ingrowth of lateral mesoderm.

Great strides have been made in the last decade regarding the general management but further advances are needed. and success is not assured, correction obviously must be individualized but most patients require urethral lengthening to gain more penile lengthening, a staged urethroplasty is often required. When all else fails, perhaps one must consider uretero sigmoidostomy but today is seldom used because of the high risk of late development of colon carcinoma. Exstrophy has challenged the skill and ingenuity of the urologists for generations and will continue to do so in the foreseeable future (king, 1984).

CHAPTER 1

EMBRYOLOGY OF THE URINARY

BLADDER AND URETHRA

EMBRYOLOGY OF THE URINARY BLADDER

AND URETHRA

INTRODUCTION :-

When the sperm fertilizes an ovum, the combined cell, "The zygote" begins its development by division and cleavage into about 30 smaller cells, "blastomeres", which arrange themselves as a hollow sphere, "The blastula", which is about the same size of the parent zygote, and gives the embryonal tissues.

By two weeks the inner cell mass flattens, a separate sheet of cells, "The endoderm", appears on the under surface of the inner cell mass, and the cells that oppose the endoderm compose the ectoderm. A cleft appears above the ectoderm and enlarges as the amnion cavity.

At the third week, cells migrate between the ectoderm and the endoderm to streak the smooth surface of the blastoderm; the two layered inner cell mass, these cells develop into mesoderm.

In the caudal end of the primitive streak, the endoderm remains opposed to the ectoderm without intervening mesoderm, this area of opposition persists during later stages of development, and called the cloacal membrane.

The following time table showing events of developments of urinary system of human body in relation to gestational date. According to Campbell's

urology . 5th ed. page 1639.

Days after Gestation	Events During Development
<hr/>	
14 days	Cloacal membrane at caudal end of primitive streak.
20 "	Tail end of embryo folds to creat cloaca.
22 "	Pronephric duct present.
24 "	Pronephros di sappears Mesonephric ducts and tubles appear.
26 "	Caudal protion of the wolffian ducts end blindly short of the cloaca.
28 "	Wolffian duct has fused to the cloaca. Ureteral buds appear. , Septation of the Cloaca begins.
41 "	Mullerian ducts appear, cloaca partitioning
44 "	Urogenital sinus separate from rectum , with ureters drain separately into it.
12 weeks	External genitalia become distinctive for sex, male penile urethra forming, prostate appears, Cowper's gland and skenes gland apappears.
13 "	Bladder becomes muscularized.
Up to 42 "	Further growth and development.

Development of the Cloaca :-

During the period of laminar embryo, the cloacal membrane lies in the caudal region. Where the ectoderm oppose the endoderm without any intervening mesoderm, as the mesenchyme grows near the cloacal membrane, it lifts the tail end of the embryo off of the blastoderm, this lifting folds the tail end and creates the cloacal chamber.

The cloaca, a sacculated, dilated, caudal terminal end of the hindgut, which forms later on the future bladder, is lined with endoderm and the cloacal membrane is covered by an ectoderm.

Further growth of the tail fold flexes it, and the cloacal membrane become on the ventrum of the embryo.

Growth of the mesenchyme lateral to the cloacal membrane elevates the tissue as labioscrotal swellings, and the growth of mesenchyme cranial to the cloacal membrane forms the genital tubercle, the primordium of the phallus.

Cloacal septation begins at 28th day, (4 mm stage)[stephens 1983] when the uro-rectal septum, "Tourneux's fold" , extends coronally towards the cloacal membrane forming the uro-rectal fold, at the same time Rathke's plicae appears as 2 tissue folds from the lateral aspects of the hindgut, to meet each other in the midline to complete the septation of the cloaca, resulting in a separate rectum and primitive urogenital sinus by the end of seventh week (44 days) and before the rupture of the cloacal membrane, so each one opens to the outside separately.

The part of the urogenital sinus cranial to the mesonephric ducts is the vesico-urethral canal, and the part caudal to it is the definitive urogenital sinus. [Hamilton and Mossman 1976 - b].

The urogenital sinus is cylindrical and continuous cranially with the allantois and receives the two meso-nephric ducts., The caudal end of the mesonephric ducts distal to the ureteric bud, which arises from it, is progressively absorbed into the urogenital sinus. So by the end of the seventh week both the meso-nephric ducts and the ureteral buds open independently into the bladder resulting in formation of an island of mesodermal tissue among the surrounding endoderm lining the urogenital sinus, this forms later on the Trigonal structure. [Max Maizels] [Fig. 1].

Development of the Trigone and the Bladder:-

By the 28th day of embryonal development, the mesonephric duct reached and fused with the uro-genital sinus, by the coalescence of the epithelium of the uro-genital sinus. (endodermal in origin) with that of the mesonephric duct (mesodermal structure)., at the same time the ureter originates from the mesonephric duct, the segment of it distal to the origin of the ureteral bud dilates and forms the common excretory duct which is absorbed into the urogenital sinus either directly or after looping of its terminus. [Hamilton and Mossman 1976 - b].

The epithelium of the duct diffuses medially to meet that of the other side forming the trigonal area, and the terminus of the ureters joins the bladder now directly.

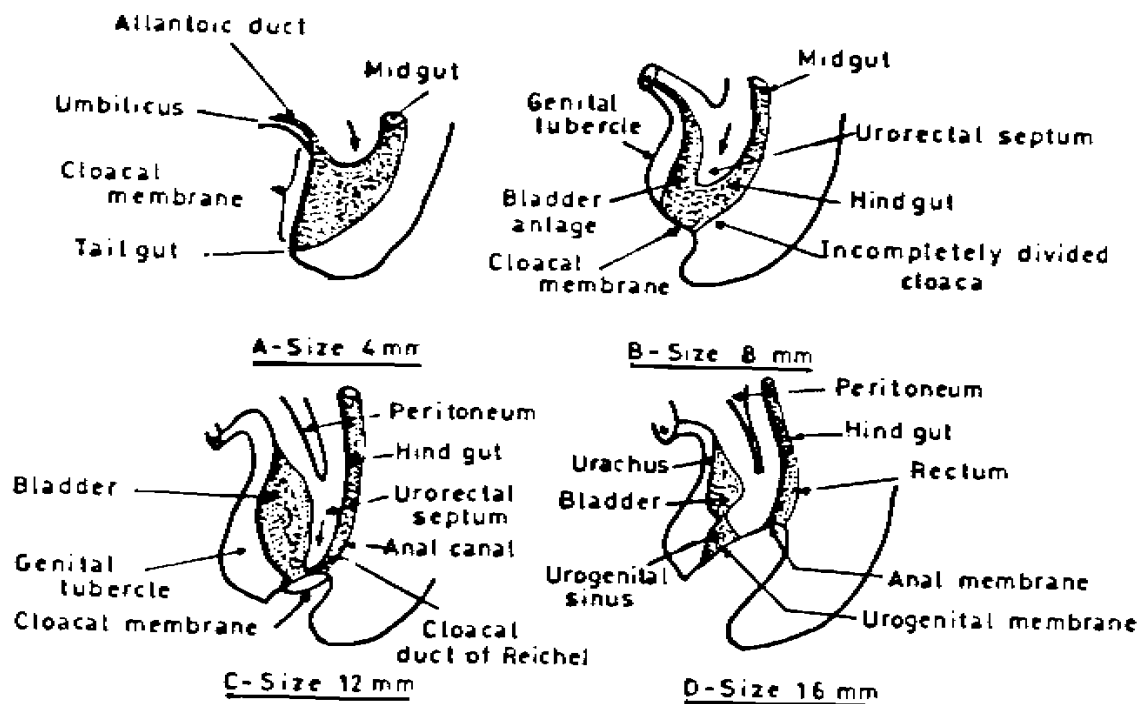


Fig. 1. Schematic illustrations of the division of the cloaca into an anterior bladder anlage and posterior hindgut by the formation and caudal growth of the urorectal septum (Muecke, E.C., 1978).

The orifice of the mesonephric duct migrates caudally and the ureteric orifices migrate cranially and displaced laterally due to the continued growth of the epithelium of the absorbed common excretory duct forming the Triangular trigone. Because of the separate growth of the trigone and the bladder, the muscle lamina of the trigone is continuous with that of the ureter and not with the bladder detrusor, even they response differently to the pharmacological stimuli. [Max Maizels].

Due to the migration of the mesenchyme of each common excretory duct medially, there is no bladder endodermal mucosa in the midsection of the trigone. [Hamilton and Mossman 1976 - b].

By the end of 10th. week, the bladder is acylindrical tube lined by connective tissue [Lowsley 1912] and its wall over the trigone is twice as thick as elsewhere.

The apex of the vesio-urethral tube tapers as the urachus which is continuous, but not formed by, the allantoic duct. [Felix 1912 , and Arey 1974] .

At the end of 12th. week this part of the bladder involute and lies between the bladder and the umbilicus thickend and fibrosed forming. The median umbilical ligament. [Moore 1977].

Bladder muscularization starts by the 13th. week as three muscle coats which are interlacing each other. [Stephens 1983].

The musculature of the ureter is continuous with that of the trigone. The bladder base is the most abundant muscularization area of the bladder up to 5 times as thick as elsewhere. Bladder muscularization is completed at 16th week. The internal sphincter is formed from a mass of the circular muscle layer.

The bladder at term is of emptying capacity of 16 ml. and this is done every 90 minutes as detected by the ultra sonic studies.

[Abramovich et. al. 1979].

Bladder muscularization starts by the outer longitudinal layer starting over the fundus as a wide curves of muscle fibres leaving a broad sheath of uncovered bladder mesenchyme specially on the anterior surface [Felix 1912].

Trigonal muscularization starts too later (100 mm stage) as a single syncytium muscle mass limited superiorly by the dense circular muscle layer of the inter-ureteric ridge and inferiorly by the dense bladder neck. [Mueck 1978].

Development of the Urogenital sinus :-

It appears at the 6th week, its cranial narrow portion forms the prostatic and membranous urethra, its caudal phallic part forms the phallus urethra, which is not differentiated sexually until 5-10 weeks. The endodermal portion of the urogenital membrane thickens and extends distally towards the genital tubercle.

Mesenchyme along the lateral margins of the phallic portion of the urogenital sinus raises the overlying ectoderm to create the primitive urethral groove. Then the urogenital membrane ruptured at the 7th week resulting in exposure of the phallic portion of the urogenital sinus which appears as a sac lined by endoderm.
[Fitz Gerald 1978].

At 8th week , the thickened endodermal plate disintegrates giving the superficial primary and deep secondary urethral groove, the later is lined by ectoderm.

Sexual differentiation appears at 10th week.

In male :-

The phallic portion of the urogenital sinus gives the bulber 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 and migrate caudally and fuse to form the base of the penis.

The glandar urethra appears at the 4th month where the glandar ectoderm proliferates and creates a plug which burrows into the glans to meet penile urethra, then it cavitates as glandar urethra.

Pelvic part of the urogenital sinus gives the lower portion of the prostatic urethra and the membranous urethra.
[Hamilton & Mossman 1976].