Ain-Shams University
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CONGENITAL URETEROPELVIC JUNCTION OBSTRUCTION

ESSAY

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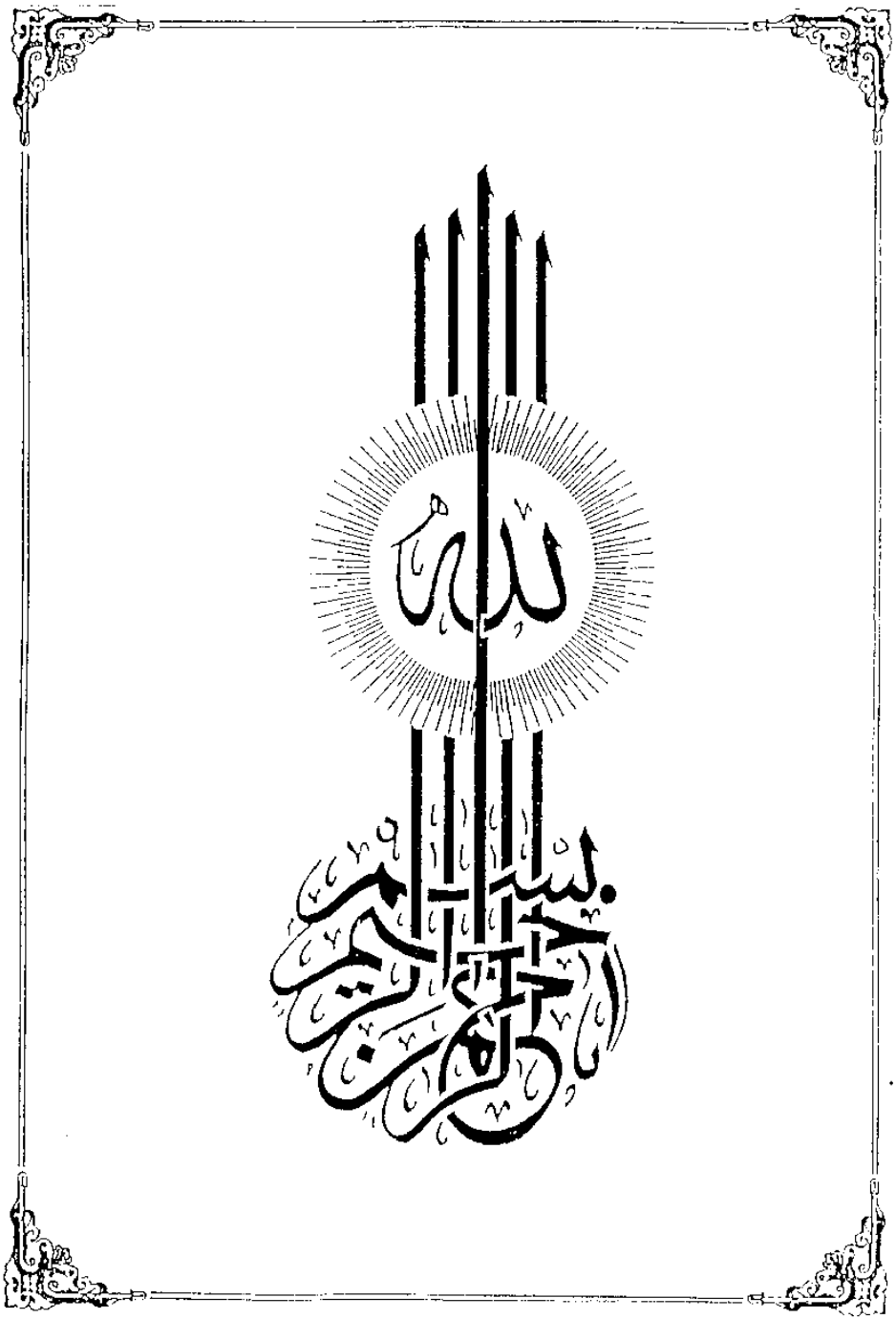

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INTRODUCTION

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Congenital ureteropelvic junction obstruction causing hydronephrosis is commonly encountered in infants and children and with proper early diagnosis and treatment the vast majority of hydronephrotic kidneys can be successfully repaired.

One of the fundamental principles of urologic surgery has been that relief of obstruction will prevent further deterioration of renal function. Recovery of some renal function occurs after release of obstruction.

This eassay discusses the embryology and physiology of the ureteropelvic junction.
Also, it discusses the pathology, diagnosis, indications of treatment and techniques of surgical correction of congenital obstruction.

EMBRYOLOGY

NORMAL DEVELOPMENT OF THE URETEROPELVIC JUNCTION

The definitive urinary system results from complex and poorly understood interactions of embryonic mesoderm with itself and with endoderm. This development is commonly viewed to repeat phases of development of lower vertebrates : the pronephros persists in adult cyclostomes (i.e., lamprey); the mesonephros is the functional kidney of most anamniotes (i.e., fish) ; and although the mesonephros functions temporarily , the metanephros is the definitive kidney of most amniotes (i.e. , birds and mamals). These phases are repeated in man : the pronephros is the first urinary tissue to appear, next the mesonephros, and finally the metanephros (Maizels, 1986).

Pronephros :

The pronephros appears in the cervical region of the 10 somite embryo between the second and sixth somites (Potter, 1972). It consists of several tubules formed from mesodermal cells of the intermediate cell

mass in the cervical region. Glomeruli do not develop and the tubules do not open into an excretory duct. The first formed tubules regress before the more caudally placed last ones are formed, and the entire pronephros disappears by the end of the fourth week (Snell, 1975).

Mesonephros :

The mesonephros, like the pronephros, consists of a number of tubules and is formed from the mesodermal cells of the intermediate cell mass in the thoracic and lumber regions. The medial end of each tubule enlarges and becomes pear-shaped, and its wall is invaginated by a cluster of capillaries which form a glomerulus. The capillaries are connected to a branch of the aorta. The lateral end of each tubule opens into a longitudinal collecting duct called the mesonephric (or Wolffian) duct. The mesonephric duct develops as a solid rod of cells in the intermediate cell mass. This canalizes and its caudal end grows to reach the lateral wall of the cloaca, which it perforates. Meanwhile each mesonephric tubule undergoes further development . With the formation of

the glomerulus, the indented medial end of the tubule forms the glomerular capsule and the glomerulus together are known as the mesonephric corpuscle. The increase in the length of the tubule causes it to become bent and S-shaped, and the equivalent of the proximal and distal convoluted tubules and the collecting tubule are formed. As the mesonephric tubules continue to form and grow , the mesonephros forms a spindle-shaped ridge, which projects into the coelomic cavity on each side of the midline. Later it becomes an ovoid body, which is suspended from the posterior abdominal wall by a thick mesonephric mesentery. During the second month, the mesonephros reaches its maximum degree of development and extends on each side from the region of the septum transversum down to the third lumber segment. After functioning for a short period, the tubules start to degenerate, and this proceeds in a craniocaudal direction. By the beginning of the third month, the majority of the mesonephric tubules and glomeruli have disappeared. However, a few caudal mesonephric tubules remain and become associated with the genital system in both sexes (Snell, 1975).

Metanephros :

The metanephros, or permanent kidney, develops from two sources : (1) the ureteric bud from the mesonephric duct and (2) the metanephrogenic cap from the intermediate cell mass of the lower lumber and sacral regions (Snell, 1975).

Development of the ureteropelvic junction :

After the mesonephric duct reaches and drains into the urogenital sinus, the ureteral bud appears. The ureteral bud originates from the mesonephric duct after 28 days of development as a diverticulum from the posteromedial aspect of the mesonephric duct at the point where the terminus of the duct bends to enter the cloaca. Potter (1972) has offered the most cohesive view of the later development of the ureteral bud. The initial four to six dichotomous branchings of the ampullae contribute to the renal pelvis. The metanoephrogenic blastema proliferates so as to remain apposed to all of the new ampullae of the new ureteral bud branches. The initial branch establishes the inferior and superior poles of the future

kidney. Branching near the poles of the kidney occurs faster than does branching near the midsection. Because of this asynchrony, by 7 weeks there are about six generations of ampullae at the poles of the kidney and only four generations of ampullae at the midsection of the kidney. The asynchronous branching helps maintain the reniform shape of the kidney (Hamilton and Mossman, 1976). This primitive network of ureteral bud branches dilates and creats 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. Ruano-Gil et al. (1975) presented a new view of the early development of the ureter. They studied human embryos and showed the patency of the ureter changes during development. Between 37 and 40 days of development, the lumen of the ureter is not apparent histologically except perhaps at the midportion. Intense elongate growth of the ureter may obliterate the lumen. At the end of this time interval, the lumen extends cranially and caudally from

the midportion of the ureter. After 40 days, the lumen is again apparent along the entire length of the ureter. These observations may handily help understand the genesis of congenital strictures at the ureteropelvic or ureterovesical junctions. The lumen of these sites would become patent last. Failure to become patent would cause a stricture. Thereby, by 8 weeks of development the ureter is a patent tube without muscle that has elongated pari passu with ascent of the kidney. After 10 weeks the epithelium of the ureter becomes two-layered, and by 14 weeks a transitional epithelium has appeared. By 18 weeks the ureter demonstrates relative intrinsic narrowings at the ureteropelvic junction , pelvic brim, and ureterovesical junction, and complementary intrinsic dilatations of the upper, middle and lower spindles of the ureter. 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 ist wall as pleats, the "fetal folds" of Ostling (1942).

Smooth muscle of the ureter first appears at the extravesical ureter after about 14 weeks (Felix , 1912) ; muscularization gradually extends toward the kidney and is completed by about 18 weeks. The ureteropelvic junction becomes well apparent by 18 weeks after fertilization.

Postnatally, the infant's growth rate exceeds that of the ureter. The ureter may lose its tortuosity and unfolds its pleats (Ostling, 1942). However, a pleat that intrudes into the lumen of the ureter and that is fixed by adventitia of the ureter may obstruct unine drainage as a valve (Maizels and Stephens, 1980).