Recent Trends in Diagnosis & Management of Congenital Hydronephrosis

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SUMMARY

Hydronephrosis refers to the accumulation of fluid (urine) in the drainage system of the kidney (the calyces and renal pelvis). It may be due to obstruction of the normal flow of urine or backflow of urine from the bladder, a condition termed reflux.

Congenital hydronephrosis giving an overall prevalence of 11.5 cases per 10,000 births, the large majority of cases were livebirths 96% of total.

The effects of hydronephrosis on the kidney are variable. In many cases mild and sometimes even moderate degrees of hydronephrosis may cause no detectable alteration in kidney function. More severe cases may have caused or may lead to some degree of kidney dysfunction. In some cases the kidney may have been or could be destroyed by the severity of the condition.

Congenital hydronephrosis may cause no symptoms, even when it is severe. If the condition leads to urinary infection the child may have fever and pain related to the infection.

The common causes of congenital hydronephrosis are: Pelviuretric junction obstruction (PUJ), Vesicoureteral reflux (VUR), Posterior urethral valves (PUV), Ureterovesical junction (UVJ) obstruction, Primary nonrefluxing megaureter, Ureterocele, , Ectopic ureter,

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List of Abbreviations

ADPKD	Autosomal dominant polycystic kidney disease
AF	Amniotic fluid
ANH	Antenatal hydronephrosis
APD	Anteroposterior diameter
ARPKD	Autosomal recessive polycystic kidney disease
BMP-7	Bone morphogenetic protein-7
CT	Computed Tomography
DMSA	Dimercaptosuccinic acid
DTPA	^{99m}Tc-diethylenetriaminepentaacetic acid
FGF-2	Fibroblast growth factor-2
GDNF	Glial-derived neurotrophic factor
IVP	Intravenous pyelogram
MAG3	^{99m}Tc-mercaptoacetyltriglycine
MCDK	Multicystic dysplastic kidney
MGU	Megaureter
MRI	Magnetic Resonance Imaging
NAG	N-acetyl-β-D-glucosaminidase
PBS	Prune-belly syndrome
RTT	Renal transit time
$t_{1/2}$	half-time
UPJ	Ureteropelvic junction
UTI	Urinary tract infection
UVJ	Uretrovesical junction
VCUG	Voiding cystourethrography
VUR	Vesicoureteral reflux

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DEDICATION

I would like to Dedicate this work to the memory of my Parents, ***IBRAHIM EL-KASHORY & NAHED HANEM RAGHEB,*** *my lovely parents who support me to the end.*

My special Dedication to my lovely wife, ***MAI AHMED EISSA****, for everything.*

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AIM OF THE WORK

This work aims at giving highlights about Current Diagnosis and Management of congenital hydronephrosis. Special emphasis will be given for recent trends in management of that congenital hydronephrosis whether conservative or surgical.

INTRODUCTION

Congenital hydronephrosis giving an overall prevalence of 11.5 cases per 10,000 births, the large majority of cases were livebirths 96% of total. Almost all livebirths were alive 1 week after birth. Boys accounted for 72% of all cases. A high proportion of the cases (86%) had an isolated renal malformation. There were large regional differences in prevalence of congenital hydronephrosis ranging from 2 to 29 per 10,000 births. There was little regional variation in the prevalence of postnatally diagnosed cases while there were large regional differences in prevalence of prenatally diagnosed cases **(Ester Garne et al, 2009).**

Diagnosis of hydronephrosis

The threshold for the diagnosis of hydronephrosis is based on the recognition that renal pelvic diameter may vary with gestational age. There is considerable variation in the definition of prenatal hydronephrosis in the literature. Siemens and colleagues found that an anteroposterior diameter (APD) of the fetal renal pelvis measuring greater than 6 mm at less than 20 weeks, greater than 8mm at 20 to 30 weeks, and greater than 10 mm at greater than 30 weeks was associated with persistent postnatal renal abnormalities **(Siemens et al, 1998).**

Differential diagnosis of congenital hydronephrosis:

Pelviuretric junction obstruction (PUJ)

Vesicoureteral reflux (VUR)

Primary nonrefluxing megaureter

Ureterocele

Ureterovesical junction (UVJ) obstruction

Ectopic ureter

Posterior urethral valves

Megacystis megaureter

Physiologic dilatation

Multicystic dysplastic kidney

Autosomal recessive polycystic kidney disease

Exstrophy

Prune belly syndrome

The management of congenital hydronephrosis can be divided into conservative and surgical treatment, and this determinate by the degree of hydronephrosis and complications that could be happen due to it, surgical treatment include Fetal intervention and surgical intervention after delivery **(Ester Garne et al, 2009).**

EMBRYLOGY OF UROLOGY SYSTEM

Functionally the urogenital system can be divided into two entirely different components: the urinary system and the genital system. Embryologically and anatomically, however, they are intimately interwoven. Both develop from a common mesodermal ridge (intermediate mesoderm) along the posterior wall of the abdominal cavity, and initially the excretory ducts of both systems enter a common cavity, the cloaca **(Sadler, 2000)**.

KIDNEY SYSTEMS

Three slightly overlapping kidney systems are formed in a cranial to caudal sequence during intrauterine life in humans: the pronephros, mesonephros, and metanephros. The first of these systems is rudimentary and nonfunctional; the second may function for a short time during the early fetal period; the third forms the permanent kidney **(Sadler, 2000)**.

Pronephros

At the beginning of the fourth week, the pronephros is represented by 7 to 10 soild cell groups in the cervical region. These groups from vestigial excretory units, nephrotomes, that regress before more caudal ones are formed. By the end of the fourth week, all indications of the pronephric system have disappeared **(Sadler, 2000)**.

Mesonephros

The mesonephros and mesonephric ducts are derived from intermediate mesoderm from upper thoracic to upper lumber (L3) segments. Early in the fourth week of development, during regression of the pronephric system, the first excretory tubules of the mesonephros appear. They lengthen rapidly, from an S-shaped loop, and acquire a tuft of capillaries that will form a glomerulus at their medical extremity. Around the glomerulus the tubules from Bowman`s capsule, and together these structures constitute a renal corpuscle. Laterally the tubule snters the longitudinal collecting duct knows as the mesonephric or wolffian duct **(Sadler, 2000)**.

In 6 weeks the mesonephros forms a large ovoid organ on each side of the midline. Since the developing gonad is on its medial side, the ridge formed by both organs is known as the urogenital ridge. While caudal tubules are still differentiating, cranial tubules and glomeruli show degenerative changes, and by the end of the second month the majority have disappeared. In the male a few of the caudal tubules and the mesonephric duct persist and participate in formation of the genital system, but they disappear in the female **(Sadler, 2000)**.

Metanephros: The Definitive Kidney

The third urinary organ, the metanephros, or permanent kidney, appears in the fifth week. Its excretory units develop from metanephric mesoderm in the same manner as in the mesonephric

system. The development of the duct system differs from that of the other kidney systems.

Collecting system. Collecting ducts of the permanent kidney develop from the ureteric bud, an outgrowth of the mesonephric duct close to its entrance to cloaca. The bud penetrates the metanephric tissue, which is molded over its distal end as a cap. Subseqently the bud dilates, forming the primitive renal pelvis, and splits into cranial and caudal portions, the future major calyces **(Sadler, 2000)**.

Each calyx forms two new buds while penetrating the metanephric tissue.These buds continue to subdivide until 12 or more generations of tubules have formed. Meanwhile, at the periphery more tubules form until the end of the fifth month. The tubules of the second order enlarge and absorb those of the third and fourth generations, forming the minor calyces of the renal pelvis. During further development, collecting tubules of the fifth and successive generations elongate considerably and converge on the minor calyx, forming the renal pyrmid. The ureteric bud gives rise to the ureter, the renal pelvis, the major and minor calyces, and approximately 1 million to 3 million collecting tubules **(Sadler, 2000)**.

Excretory System. Each newly formed collecting tubule is covered at its distal end by a metanephric tissue cap. Under the inductive influence of the tubule, cells of the tissue cap form small vesicles, the renal vesicles, which in turn give rise to small S-shaped tubules. Capillaries grow into the pocket at one end of the s and diffrerentiate