Embryology of the GU Tract

DEVELOPMENT OF THE KIDNEY AND URETER

The embryonic kidneys are, in order of their appearance, the ***pronephros*, the *mesonephros*, and the *metanephros*.** The first two kidneys regress in utero, and the third becomes the permanent kidney. All three kidneys develop from the intermediate mesoderm.

The first evidence of pronephros is seen in the 3rd week, and it completely degenerates by the the 5th week. Although renal maturation continues to take place postnatally, nephrogenesis is completed before birth (at 36 Wk).

Between the 6th and 9th weeks, the kidneys ascend to a lumbar site just below the adrenal glands.

Urine production in the human kidney is known to begin at about 10 to 12 weeks.

From ureteric bud which appears at 4^{th} wk .The bifurcation of the ureteric bud determines the eventual pelvicalyceal patterns and their corresponding renal lobules.

BLADDER DEVELOPMENT

Formation of the Urogenital Sinus (Ectoderm)

The bladder and posterior urethra fromed from the cephalad part of Urogenital Sinus(vesicourethral canal) while the caudal part of urogenital sinus forms the penile urethra for males and the distal vaginal vestibule for females.

Formation of the Trigone (Mesoderm)

By day 33 of gestation, the common excretory ducts (the portion of the mesonephric ducts distal to the origin of the ureteric buds) dilate and become absorbed into the urogenital sinus. The right and left common excretory ducts fuse in the midline as a triangular area, forming the primitive trigone and bladder neck.

GENITAL DEVELOPMENT

During the 5th week, primordial germ cells migrate from the yolk sac along the dorsal mesentery to populate the mesenchyme of the posterior body wall near the 10th thoracic level. In both sexes, and form a pair of ***genital ridges*** just medial to the developing mesonephros. During this time, a new pair of ducts, called the ***paramesonephric (müllerian) ducts*,**

Between the 8th and 12th weeks, testosterone secretion by Leydig cells stimulates the mesonephric ducts to transform into the vas deferens. The cranial portions of the mesonephric ducts degenerate, leaving a small remnant of tissue protrusion called the *appendix epididymis*, and the region of mesonephric ducts adjacent to the presumptive testis differentiate into the epididymis.

The seminal vesicles sprout from the distal mesonephric ducts, whereas the prostate and bulbourethral glands develop from the urethral epithelium (11-16 wk)

The testicles descend to the level of internal inguinal ring by the 3rd month and complete their descent into the scrotum between the 7th and 9th month

The ovaries also descend and become suspended within the broad ligaments of the uterus

Development of Female Genital Structures

In female embryos, the primitive sex cords do not contain the Y chromosome, do not elaborate SRY protein, and therefore do not differentiate into Sertoli cells. In the absence of Sertoli cells and SRY protein, MIS synthesis, Leydig cell differentiation, and androgen production do not occur.In the absence of MIS, the mesonephric (wolffian) ducts degenerate and the paramesonephric (müllerian) ducts give rise to the fallopian tubes, the uterus, and the upper two thirds of the vagina. The remnants of mesonephric ducts are found in the mesentery of the ovary as the epoöphoron and paroöphoron, and near the vaginal introitus and anterolateral vaginal wall as Gartner's duct cysts.

CONGENITAL ANOMALIES OF THE KIDNEY

1) ANOMALIES OF NUMBER

1-Agenesis: Absence of the renal tissue either bilateral (only about 500 cases having been reported and it is incompatible with live) or unilateral Renal Agenesis(unilateral agenesis occurs once in 1100 births)

2- Supernumerary Kidney : 3-4 separated kidneys

2) ANOMALIES OF VOLUME AND STRUCTURE

Hypoplasia**,** Multicystic kidney**,** polycystic kidney (Infentile and Adult type), Medullary cystic disease.

3) ANOMALIES OF ASCENT

1- Simple Renal Ectopia: refers to a low kidney in proper side that failed to ascend (pelvic kidney, or at pelvic brim) 2- Cephalic Renal Ectopia 3- Thoracic Kidney

4) ANOMALIES OF FORM AND FUSION

1- Crossed Renal Ectopia With and Without Fusion, Inferior Ectopic Kidney, Sigmoid or S-Shaped Kidney, Lump Kidney L-Shaped Kidney, Superior Ectopic Kidney

2-Horseshoe Kidney about 1 in 400 persons. The horseshoe kidney is probably the most common of all renal fusion anomalies. 2 kidneys fused at lower pole by Isthmus (fibrous or renal tissues) and are stopped during ascend by inferior mesenteric artery so it will be lower than normal and not completed normal rotation and the axes of the 2 kidneys will be parallel to the spine . If there is no obstruction and asymptomatic patient no need for any treatment.

5) ANOMALIES OF ROTATION; The adult kidney, as it assumes its final position in the "renal" fossa, orients itself so that the calyces point laterally and the pelvis faces medially. When this alignment is not exact, the condition is known as ***malrotation***

6) ANOMALIES OF RENAL VASCULATURE

1- Aberrant, Accessory, or Multiple Vessels

"aberrant vessels" should be reserved for those arteries that originate from vessels other than the aorta or main renal artery**.** The term **"accessory vessels"** denotes two or more arterial branches supplying the same renal segment.

2-Renal Artery Aneurysm 3-Renal Arteriovenous Fistula

7) ANOMALIES OF THE COLLECTING SYSTEM

Calyceal Diverticulum, Megacalycosis, Bifid Pelvis

CYSTIC DISEASE

The kidney is one of the most common sites in the body for cysts. Although the lesions themselves in the various cystic conditions are histologically similar (i.e., microscopic or macroscopic sacs lined with epithelium), their number, location, and clinical features are different. The terms multicystic and polycystic should not be confused, even though both terms literally mean "many cysts." Multicystic refers to a dysplastic entity, and polycystic refers to a number of separate entities, most inherited, all without dysplasia and all with nephrons throughout the kidney

Polycystic Kidney Diseases

1) Autosomal Recessive ("Infantile") Polycystic Kidney Disease

ARPKD has been reported as a rare disease affecting about 1 of every 40,000 .

ARPKD has a spectrum of severity, the most severe forms appearing in the neonate. If it is mild the disease will become apparent later in childhood (up to age 13 years or, rarely, up to age 20 years) and is bilateral. All patients have varying degrees of congenital hepatic fibrosis as well. the genetic locus has been identified at chromosome 6 .

The diagnosis may be suspected from in utero ultrasound examination and may be associated with oligohydramnios, a finding secondary to low urinary output. In both fetus and newborn,

sonography identifies very enlarged, homogeneously hyperechogenic kidneys, especially when compared with the echogenicity of the liver.
Intravenous urography with delayed films may show functioning kidneys with characteristic radial or medullary streaking (sunburst pattern) caused by dilated collecting tubules filled with contrast medium.

Treatment

No cure has been found for ARPKD. Respiratory care can ease or extend the child's life. Patients who survive may require treatment for hypertension, congestive heart failure, and renal and hepatic failure.

2) Autosomal Dominant ("Adult") Polycystic Kidney Disease

The autosomal dominant form of polycystic kidney disease (ADPKD) is an important cause of renal failure. Its incidence is approximately 1 in 500 to 1000. Two genes for ADPKD have been localized, PKD1 on chromosome 16 and PKD2 on chromosome 4. most cases are identified when the patients are between 30 and 50 years of age, but renal failure is seldom seen before the age of 40 years. the typical age at diagnosis will decline as more members of families at risk for the trait are screened by genetic testing and by ultrasound examination. A number of associated anomalies are common, including cysts of the liver, pancreas, spleen, and lungs; aneurysms of the circle of Willis (berry aneurysms); colonic diverticula; and mitral valve prolapse
it appears that tubular epithelial cell hyperplasia is the major component of cyst development, although possibly not the first.
The renal cysts range from a few millimeters to a few centimeters in diameter and appear diffusely throughout the cortex and medulla with communications at various points along the nephron.
the incidence of RCC in patients with ADPKD is no higher than that in the general population.

Clinical Features

Typically, symptoms or signs first occur between the ages of 30 and 50 years. These include microscopic and gross hematuria, flank pain, gastrointestinal symptoms (perhaps secondary to renomegaly or associated colonic diverticula), and renal colic secondary either to clots or stone. As blood pressure screening has become more widespread, hypertension more than hematuria has become the principal form of presentation.
Approximately 10% to 40% of patients have berry aneurysms, and approximately 9% of these patients die because of subarachnoid hemorrhages.
On intravenous urography, the calyces may be stretched by cysts. However, the picture may simulate that of ARPKD. A CT scan or MRI (or both) may be helpful in some cases and often is superior to sonography for detecting cysts in organs other than the kidney
Because ADPKD is an autosomal dominant condition, 50% of the children of affected adults will also be affected. Therefore, when the disease is diagnosed, the patient's children should be examined by ultrasound and genetic studies.

Treatment and Prognosis

The aim of treatment is to preserve renal function as long as possible by controlling hypertension and treatment of infection. Men tend to have more renal involvement than women, manifesting with hypertension and renal insufficiency earlier than in women . progressive renal failure require dialysis and ultimately renal transplant. The pain can be colicky, acute, or chronic. Colicky pain occurs secondary to the passage of either stones or clots. Acute pain may be secondary to infection or hemorrhage into a cyst or to subcapsular bleeding.Surgical unroofing the cysts to relieve the pain (Rovsing operation). laparoscopic unroofing of cysts as an alternative to an open procedure in order to reduce the incidence of morbidity. Upper tract urinary tract infections are common in patients with ADPKD, especially women and the only dependable antibiotics were those that were lipid soluble, namely, trimethoprim-sulfamethoxazole, fluoroquinolones, and chloramphenicol. Most patient may need Renal Transplant

Simple Renal Cyst

A simple cyst is a discrete finding that may occur well within a kidney or on its surface. It is usually oval to round in shape, has a smooth outline bordered by a single layer of flattened cuboidal epithelium, and is filled with transudate-like clear or straw-colored fluid. It is not connected to any part of the nephron, although it may originate initially from a portion of the nephron. Simple cysts may be singular or multiple, unilateral or bilateral. may be congenital or acquired. Simple cysts may manifest at any time
Simple cysts vary in size, ranging from less than 1 cm to greater than 10 cm. The majority are less than 2 cm in diameter.

Clinical Features
In both children and adults, cysts rarely call attention to themselves. Instead, they are discovered incidentally on sonography, CT, or urography performed for a urinary tract or other pelvic or abdominal problem. However, cysts can produce an abdominal mass or pain, hematuria secondary to rupture into the pyelocalyceal system, and hypertension secondary to segmental ischemia

Differential Diagnosis
RCC, ADPKD, Complex renal cyst(that contain blood, pus or calcification),Hydonephrosis, Hydatidcyst of kidney, Renal carbuncle.

The diagnosis of a classic benign simple cyst by sonography when the following criteria are met: (1) absence of internal echoes; (2) sharply defined, thin smooth wall and margin (3) good transmission of sound waves through the cyst with consequent acoustic enhancement behind the cyst; and (4) spherical or slightly ovoid shape.
If all of these criteria are satisfied, the chance that malignancy is present is negligible

The CT criteria for a simple cyst are similar to those used in sonography: (1) sharp, thin, distinct, smooth walls and margins; (2) spherical or ovoid shape; and (3) homogeneous content.t(The density ranges from -10 to +20 HU, similar to the density of water), and (4) no enhancement should occur after the intravenous injection of contrast medium

Treatment
No treatment if asymptomatic cyst
Symptomatic cyst may need percutanous drainage and injection of scleroting agents(Tetracycline or Alcohol) and some time may need surgical excision.
Suspicious cyst may be treated as cystic Tumour.