Genitourinary Disorders

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Disclosures

The following speaker of this CME activity has no relevant financial relationships with commercial interests to disclose:

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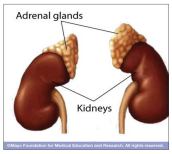
Objectives

Discuss normal embryologic development of genitourinary system
Describe common genitourinary disorders, clinical presentation, and management strategies
Describe radiographic findings of common genitourinary disorders

Fetal Kidney Development



3 Successive Phases

Pronephros – immature form of kidney - develops 4 weeks then degenerates

Mesonephros – appears 5 to 12 weeks - develops glomeruli and tubules, degenerates by 4th month

Metanephros – final kidney, develops by 5 weeks, functioning by 9 weeks, first nephrons formed by week 8

These structures are key to development of final kidney

Fetal Kidney Development

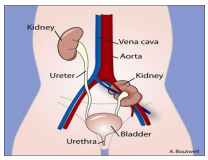
Kidneys start in pelvis, migrate upward, rotate 90 degrees

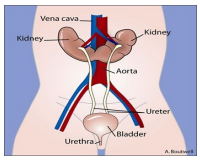
Failure to migrate up = pelvic kidneys

Abnormal ascent and rotation = horseshoe kidneys (kidneys pushed together and fused)

Adult kidney - size computer mouse

Bladder size - 32 weeks (10mls), term (40mls)





Ascension

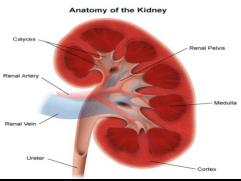
Renal Anatomy

Cortex - outermost portion - glomeruli, proximal & distal tubules, collecting ducts of nephrons

Medulla - middle section - renal pyramids, straight portions of tubules, loop of Henle, vasa recta, terminal collecting ducts

Renal sinus and pelvis - innermost portion - renal pelvis and calyces, branching of renal vessels, nerves

Ureter - excretory duct, transports urine to bladder



Ascension

Major Functions of Kidney

3 major functions: filtration, reabsorption, and secretion

- ✔ Removal toxic waste products from blood
- ✔ Regulate electrolyte & water balance
- ✔ Regulate blood pressure
- ✔ Regulate body's pH
- ✔ Process vitamin D
- ✔ Produce erythropoietin to stimulate bone marrow
- ✔ Produce prostaglandins

Urine Production

Begins at 9-10 weeks - major component of amniotic fluid

Fetus produces 2mls/hr at 20 weeks, 10mls/hr at 30 weeks, 17 mls/hr at 35 weeks

Fetal swallowing, breathing and urination regulate amniotic fluid

Polyhydramnios - usually caused by GI anomalies - renal dysplasia, nephrotic syndrome, inherited tubular defects

Oligohydramnios - renal agenesis or dysplasia, polycystic kidneys, low urinary tract obstruction

Specific gravity usually low (1.002-1.010)- inability to concentrate urine

In utero homeostasis is regulated by what?

What is normal urine output after birth?

Nephron

Nephron formation begins - 8 weeks

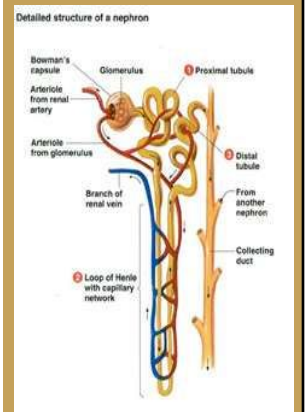
Nephrons develop until 35-36 weeks

Maturation of nephrons continue into infancy

~1 million nephrons in adult kidney

Preemie - shortened tubules - continue to grow after birth

Filtrate flows through kidney - modified in various tubule segments (excretion/absorption fluid & electrolytes)



Kidney Function

GFR= Glomerular Filtration Rate

GFR - reflective of renal function

GFR - low initially, increases postnatally

25% cardiac output filtered by kidneys (adult)

Everything that affects cardiac output (CO) - affects kidneys (kidneys are sensitive indicators)

Infants less than 34 weeks - low GFR

Kidney Function

Blood flow = filtration, driven by BP

Blood flows through capillaries

Plasma filtered through capillary walls to

Bowman's Capsule -collects filtrate & enters tubules

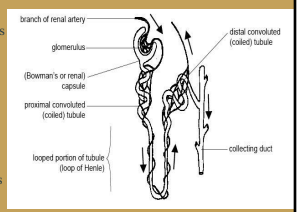
Proximal tubule -60-70% reabsorb Na/water

Loop of Henle: reabsorption of Na, water, K+, bicarb, calcium (Loop Diuretics)

Distal Tubule: reabsorption Na/Aldosterone- causes water/Na reabsorption, K excretion

Collecting tubules : ADH, reabsorption of water, urine concentrates

Specific gravity: 1.002-1.010



Acid Base Balance

Extracellular buffers: bicarbonate/ammonia

Intracellular buffers: protein/phosphate

Acid-base balance: **LUNGS**

- pH < 7.35 = acidosis: body (lungs) responds by increasing RR to decrease CO2
- pH > 7.45 = alkalosis: opposite

KIDNEYS - response to acidosis: tubules **reabsorb** bicarbonate, collecting duct **secrete** hydrogen

KIDNEYS -response to alkalosis: **excrete** more bicarbonate by decreasing hydrogen ion secretion from tubules

Hypertension

Defined as blood pressure >2 SD above normal or systolic BP >95th %ile

Term systolic > 90 mmHg, Preterm > 80 mmHg

Renal causes - hypertension:

- Polycystic kidney disease
- Mechanical compression kidneys (hydronephrosis)
- Acute tubular necrosis
- Renal vein/artery thrombosis (history umbilical lines)
  - thrombocytopenia
  - hematuria
  - hypertension
- Infection - UTI

Diagnosis : doppler ultrasound

Hematology workup - if suspect thrombosis -coagulation studies, sepsis evaluation (UTI)

Treatment RVT- anticoagulant therapy (heparin) and monitoring levels

Types of Acute Renal Injury

Prerenal - decreased blood flow to kidneys, kidneys normal (dehydration, asphyxia, hypotension, sepsis)

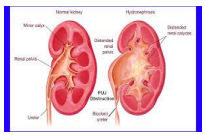
Intrinsic - injury to kidneys glomeruli and tubules - inflammation, toxins, drugs, damage from reduced blood flow (thrombosis), congenital anomalies

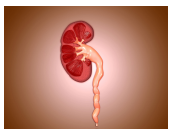
Postrenal - blockage below kidneys causing obstruction urine

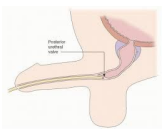
- Bilateral ureteropelvic junction (UPJ): involves ureters and renal pelvis
- Bilateral ureterovesical obstruction (UVJ): involves ureters and bladder
- Posterior urethral valves (PUV) - distal bladder, urethra

Labs - serum Cr >1.5 mg/dl, elevated BUN, hyperkalemia, metabolic acidosis

Treatment - (underlying cause)







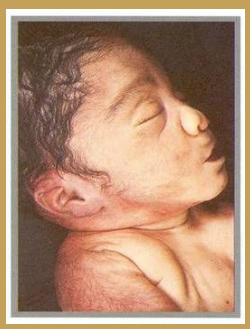
Potter's Sequence (*Oligohydramnios syndrome*)

Classic Potter's - bilateral renal agenesis, severe respiratory distress, fatal

Potter's sequence (triad)

- Clubbed feet
- Pulmonary hypoplasia
- Cranial anomalies - related to oligohydramnios

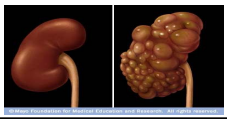
Treatment - support respiratory status, follow renal function, dialysis, kidney transplant



Multicystic Dysplastic Kidney

Multicystic Dysplastic Kidney (MCDK) - congenital anomaly - renal cortex replaced by numerous cysts

- usually unilateral disorder
- dysplastic parenchyma with cysts - resembles bunch of grapes
- majority undergo partial or complete spontaneous involution over time
- nonfunctional kidney
- bilateral condition - incompatible with life
- normal kidney grows larger (compensatory hypertrophy) - allowing child to maintain normal renal function



Polycystic Kidney Disease

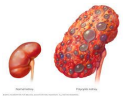
Affects both kidneys, contain many small cysts

- usually autosomal dominant - one parent carries gene - 50% chance baby developing disorder
- occurs in both children and adults - more common in adults (symptoms occurring middle age)

Autosomal recessive - appears infancy or childhood, very serious, progresses rapidly, results in end-stage kidney failure, generally causing death shortly after birth

- liver enlarged and fibrotic
- bile duct proliferation
- scattered cysts
- renal insufficiency

** An increase number of infant's are surviving with dialysis and transplant

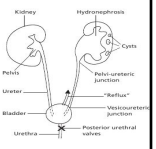


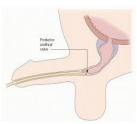
Hydronephrosis

Hydronephrosis - distention of renal pelvis, obstruction of urine from kidney

Posterior urethral valves - occur only in boys

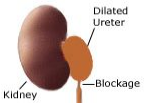
- congenital membrane obstructs posterior urethra - affects urethra, bladder, ureters & kidneys
- most common cause - lower urinary tract obstruction
- palpable distended bladder/poor urinary stream
- VCUG - diagnostic
- Initial treatment- urinary catheter/vesicostomy, later primary ablation of valves





Ureteropelvic junction (UPJ) obstruction

- most common cause fetal hydronephrosis
- more common males - associated congenital anomalies, syndromes, other genitourinary malformations
- intrinsic narrowing of proximal ureter
- treatment - surgical repair



Vesicoureteral reflux (VUR)

Retrograde passage urine from bladder into ureter & collecting system

- reflux occurs if bladder outlet obstruction causes - increased intravesical pressures
- common cause - ureteropelvic junction (UPJ) - (obstruction/narrowing where ureter meets bladder)
- common cause UTI

Diagnosis - Renal ultrasound - hydronephrosis

VCUG (Grading scale - UTD P1 to P3 - >48 hours)

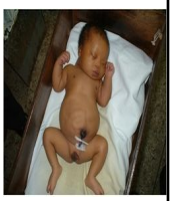
UTD 1 (<15mm renal pelvis or central dilated calyces)

UTD 2 (>=15mm renal pelvis or peripheral dilatation calyces or >4=4mm dilated ureter

UTD 3 (>10mm renal pelvis or any dilatation of calyces/ureter with oligo, abnormal bladder/parenchyma

Treatment - antibiotics, surgery

Prune Belly Syndrome (Eagle-Barrett Syndrome)



Characterized by triad of findings

1) bilateral undescended testicles

2) partial/complete lack abdominal muscles

3) urinary tract abnormality - large ureters, distended bladder, Vesicoureteral reflux (VUR)

Frequent UTIs, dislocated hips

Incidence: 1/35,000-50,000 live births, 95% males

Etiology - unknown, theory in-utero urinary tract obstruction or mesodermal injury during 4th- 10th week gestation

Treatment- optimize urinary drainage (vesicostomy) and later bladder reconstruction, manage renal insufficiency & antibiotic prophylaxis, respiratory support

Bladder Exstrophy

Occurs 4-5 weeks - failure of abdominal wall to close

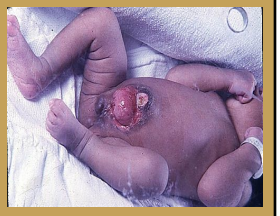
No known genetic link - if mother has child with it, chances increase further pregnancy

Extremely rare

Commonly have other GU anomalies, pelvic malformations

Treatment - surgical repair

Outcomes - frequent UTIs, urinary incontinence, and vesicoureteral reflux



Hernias and Such

Testes descend during 7th month

Inguinal Hernia - protrusion of abdominal-cavity contents through inguinal canal

Most common surgical condition in children, ↑ incidence in premature infant

Hydrocele - fluid-filled sac surrounding testicle - swelling of scrotum

- 10% male infants have hydrocele at birth
- most disappear without treatment - within first year of life

Differentiation made by transillumination

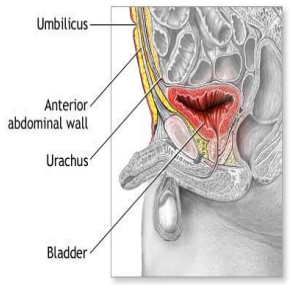
Testicular torsion

Painful, blue or erythematous, edematous scrotum

Doppler ultrasound

Emergent surgery

Patent Urachus



ADAM.

Fibrous remnant of canal - drains bladder & runs within umbilical cord

Leaking urine from umbilicus

High risk of UTI

Diagnosis - abdominal ultrasound, VCUG

Treatment - surgery (ligation) and antibiotics

Hypospadias

Urethral opening located on ventral (underside) surface

Incidence: 1/300 live births

10-15% have first degree relative

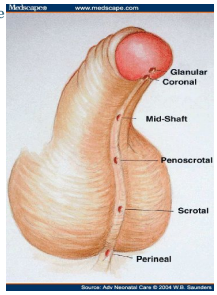
Increases in severity when closer to body wall

Main types - glanular, penile, penosacral, perineal

Epispadias - very rare (dorsal side)

Chordee commonly present (downward curving, bending shaft)

Treatment - no circumcision, surgery - outpatient Urology



Ambiguous Genitalia

Sexual organs develop from same fetal tissue

Presence of male sex hormones causes male organs to develop

Absence of male hormones causes female organs to develop

Characteristics in females

- an enlarged clitoris, or what appears to be small penis
- concealed vagina

Characteristics in males

- hypospadias
- abnormally small penis - urethral opening near scrotum
- absence both testicles in what appears to be scrotum

Ambiguous Genitalia

Possible causes in females:

- congenital adrenal hyperplasia (CAH) - cause adrenal glands make excess male hormones (androgens)
- mother taking progesterone (early stages of pregnancy to stop bleeding)
-

Possible causes in males:

- impaired testicle development - genetic abnormalities or unknown causes
- impairment testosterone production (Leydig Cell Aplasia)
- androgen insensitivity syndrome
- enzyme deficiency - impairs male hormone production (5 Alpha-Reductase Deficiency)
- mother taking estrogens (during pregnancy)

Practice

Ultra filtration in the kidneys of a neonate is driven by:

1) Pressure
2) Chemicals
3) Osmosis

Practice

Urine formation occurs in which part of the neonate's kidneys?

1) The ureter
2) The nephron
3) The loop of Henle

Practice

A significant drop in arterial pressure may result in the activation of :

1) The parasympathetic nervous system
2) The renin-angiotensin-aldosterone system
3) The renal prostaglandin system

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