Gastrointestinal Disorders

Genitourinary Disorders

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Functions

- Absorption and digestion of nutrients
- Maintenance of fluids and electrolytes
- Protection from toxins and pathogens
Embryology

- Intestine is present at week 4
- Reenters abdominal cavity at week 9–10
- Meconium is present at week 16
- Peristalsis begins at week 26
- Sucking and swallowing become coordinated at week 34

Assessment

- History
  - Family history
  - Genetic syndromes
  - Prenatal ultrasound
- Exam
  - Assessment - Size, shape, muscle development, umbilical cord, bowel loops, veins
  - Auscultation – bowel sounds
    - Increased bowel sounds with malrotation, Hirschsprung’s; Decreased with ileus
  - Percussion – size of organs, masses, fluids
  - Palpation – abdominal tone, masses, fluid, organ size (liver, spleen)
- Diagnostic Tests
  - Gastric aspirate
  - Apt test
  - Stool exam
  - pH probe
  - X-ray
  - Ultrasound
Case 1
- Prenatal ultrasound
- Congenital anomaly with herniation of abdominal viscera into umbilical cord
- May be associated with other anomalies

Abdominal Wall Defects: Care
- Goals
  - Prevent hypothermia
  - Maintain sterile environment
  - Maintain perfusion
  - Cover with warm, moist dressings (NS), or bowel bag
  - NPO, NG
  - Place on side or support defect
  - Primary or staged repair
Obstructions of the GI Tract

- History
  - Polyhydramnios
  - Failure to pass meconium (24-48 hours)
  - Abdominal distention
  - Bilious vomiting
- Preop management
  - NPO/NG
  - IV, replace losses
  - Antibiotics

Case 3

- 23 year old g1 p1 with polyhydramnios
delivered by c-section due to non-reassuring fetal heart rate. Baby active and
crying at delivery. Placed on radiant warmer and suctioned for copious amounts
of frothy white secretions. Color improved without oxygen. Required continuous
suctioning.

EA/TEF

- EA: proximal and distal portions of the esophagus are not connected
- TEF: abnormal communication between esophagus and trachea
- Presentation: secretions, drooling, can’t pass NG
- Surgery
  - Primary or staged
  - Depends on distance between esophageal pouches
Case 4

- 32 week infant born by C-section due to maternal PIH. Baby is now 36 weeks gestation and had been doing well. Began to have projectile vomiting within 20-30 minutes after feedings. Baby is then hungry and wants to eat.

Pyloric stenosis

- Hypertrophy of the pyloric musculature
- Usually occurs at 3-4 weeks of age
- Nonbilious vomiting – becomes projectile
- Diagnosis: UGI with contrast
- Surgical repair
Case 5

“Hello - so glad to have found you all! I am 34 weeks along and at 30 weeks, Doctors told me they suspect our son has ________. There is not a clear “double bubble” but there does look to be a narrowing and his tummy is bloated. We had an amnio at 14 weeks due to test results and the tests came back negative.”

http://www.shareyoustory.org

Duodenal atresia

- Obstruction of the duodenum
- May be associated with intestinal malrotation (50%), Down’s syndrome (25%), prematurity, congenital heart disease, TE abnormalities
- Bilious vomiting, distention
- History of polyhydramnios
Double bubble on x-ray
- Surgery to remove atretic portions
- Post op:
  - NPO
  - Decompression
  - Pain management
  - TPN
  - Antibiotics

Jejunal or ileal atresia
- 1/3 have malrotation
- Present with bilious vomiting, distention
- X-ray: dilated loops of bowel, multiple air fluid levels
- Surgery – location
  - NPO until normal bowel function (3-7 days)
  - Elemental formula

- NPO
- Decompression
- Pain management
- TPN
- Antibiotics
Malrotation
- Intestines do not rotate and/or fixate properly
- Bilious vomiting, distention
- Surgery
  Ladd procedure

Case 6
- Baby boy G was born at 25 weeks gestation. He required intubation at delivery and has received 2 doses of surfactant.
- At about 24 hours of age, he was noted to have a very full and distended abdomen. He had bilious drainage noted in his OG tube and had not stooled. A KUB was done and showed bowel gas throughout with some distention and a soap bubble appearance.
Meconium Ileus/Plug Syndrome

- Nonsurgical approach
  - GI priming alternating mucomyst
  - Mucomyst enemas alternating with glycerin chips
  - Gastrografin enemas

- Surgical
  - Intestine that is compromised is resected
  - End to end anastomosis
  - Ostomy

Case 7

- Baby boy W was born at 38+6 weeks gestation by vaginal delivery. To NICU at 30 hours of age due to abdominal distention and bilious aspirates. Placed on intermittent OG suction and had 9.3 ml green mucoid aspirate/24 hours. Given 2 glycerin suppositories in 12 hours without results, then had a smear of a stool. KUB with dilated loops of bowel, fixed dilated bowel loop in the left quadrant. Abdominal girth increased from 35-36.5cm. Gastrografin enema done which revealed no peristalsis.
Or...Case 8

- Baby girl B born by vaginal delivery at 40 weeks. Infant with distended abdomen, and no stool since birth. Bilious emesis at ~24 hours of age in newborn nursery. KUB with large distended loops of bowel and no air in rectum. 18 month old sibling and father of infant have Hirschsprung's disease and are followed by UNC.

Treatment: Hirschsprung's disease

- Surgery – ostomy, then bring normal bowel down to the anus
- Confirmed by rectal biopsy–no ganglionic cells

Imperforate anus

- Anorectal malformation
- Different levels
  - High-more difficult to repair
- Surgery
Case 9 & 10

- Baby E was born at 25 weeks gestation. At 36 days of life, she presented with apnea, pallor, abdominal distention, emesis, and bloody stool.
- Baby D was also born at 25 weeks. She was placed NPO at 15 DOL due to a 5 ml residual. KUB with stacked loops of bowel and pneumatosis. Baby began having bloody stools later that day and had persistent pneumatosis on x-ray x 2.

Necrotizing enterocolitis

- Acquired disease - areas of necrotic bowel
- Most important risk factor: prematurity
- Etiology unclear and multifactorial
  - Intestinal ischemia
  - Bacterial colonization
  - Enteral feedings
- Onset – usually days 3-10
Presentation: residuals, distention, bilious vomiting, bloody stools
- Labs: acidosis, thrombocytopenia, leukocytosis
- X-ray: pneumatosis, persistent loop of dilated bowel
- Medical or surgical treatment

Short Bowel Syndrome
- Syndrome of malabsorption and malnutrition due to loss of bowel
- Infants require elemental formula and TPN
- Infants can survive with 15 cm of small bowel if ileocecal valve intact; 30-45 cm if not
  - Delays intestinal transit time
  - Prevents colonic bacterial overgrowth in small intestine

Case 11
- Baby boy S developed jaundice at 2 ½ weeks of age. He also had light colored stools and dark urine.
Treatment

- Surgery
  - resection of atretic areas, anastomosis
  - Kasai – hepatic portoenterostomy
    - intestinal conduit between liver surface and small intestine
  - liver transplant
- Complications
  - Less than 20% of patients who have the Kasai procedure survive to adulthood without a liver transplant.

Cholestasis

- Impaired bile flow
  - 7-50% associated with prolonged TPN
- Presentation
  - Hepatomegaly
  - Clay colored stools
  - Direct bili >2
- Management
  - Decrease TPN protein, increase enteral feeds
  - Phenobarb – decreases bile acid pool size
  - Actigall
Case 12
- Baby boy R weighed 738 grams at 25 weeks. As he was approaching full feedings, he began to have desaturations associated with his feedings. He also had some spits after the feeds.

Treatment
- Feed slowly - small and frequent feeds
- Prone or left lateral positioning
- Thickened feeds (rice cereal, AR formula)
- Reglan

Prognosis
- Resolves in most by 12-18 months
- 10-15% require prolonged medical management
- 10-15% require surgery (Nissan, fundoplication)

Case 13
- Prenatal ultrasound at 19 weeks shows laxity of abdominal musculature
Prune belly syndrome

- Congenital anomalies – absence of abdominal musculature, GI abnormalities, undescended testes
- Treatment
  - Surgery to repair urinary and renal problems
  - May require Crede to empty bladder
  - Binders and reconstructive surgery for abdominal musculature
  - Constipation is a problem

Case 14

- Baby girl C was born at 38 weeks gestation by scheduled C-section. She cried at delivery, but had increased work of breathing, and a scaphoid abdomen. Bowel sounds were decreased.
- Treatment
  - Intubation
  - NG suction – gastric decompression
- Primary closure if possible
- Prognosis related to degree of pulmonary hypoplasia

Hyperbilirubinemia
- Jaundice is visible when the bili is 5-7
- 80% of preterm infants due to
  - Increased bili load to liver
  - Shorter life span of RBCs
  - Increased reabsorption of bilirubin from the intestine
- Peaks on day 3 in term infant, day 5-6 in preterm
Breast milk jaundice
- AAP does not encourage interruption of breast feeding
- Phototherapy, supplement bottle feeding, or temporarily stop breast feeding

Kernicterus
Treatment
- Phototherapy – blankets, lights (blue)
- Exchange transfusion – rate of rise, gestational age

Case 15
- Twin A born at 27 weeks. Birth weight was 1137 grams. Twin B weighed 660 grams. Baby had generalized edema and ascites. Given 2mg/kg of Lasix. Mild fluid restriction of 70 ml/kg/day. Baby had intermittent episodes of duskiness and appreciable differences in pre and post ductal saturations and decreased oxygenation. Echo showed severe biventricular dysfunction and left ventricular enlargement. Moderate MR and severe TR.

Hydrops
- Generalized edema
- Alloimmune – maternal antibodies cross placenta and destroy fetal RBCs
- Nonimmune – other causes (cardiac, chromosomal, infection, hematologic, etc)

Treatment
- Antenatal – intrauterine transfusion, maternal and/or fetal medication
- Neonatal – may require resuscitation, para/thoracentesis, partial exchange
Renal and Genitourinary Disorders

Embryology
- Kidney development begins at 3-4 weeks gestation
- Nephrons begin forming at 8 weeks; continue through 34 weeks
- Urine production begins at 9-10 weeks
- Vascular pattern of the kidney by 14-15 weeks
Hemodynamics

- Renal blood flow rate determined by:
  - Cardiac output
  - Renal to systemic pressure
- Low renal blood flow in the neonate
  - High renal vascular resistance
  - Low perfusion pressure
- Hormonal regulation
  - Renin-angiotensin-aldosterone system
    - Regulates systemic blood pressure, sodium, potassium
    - Prostaglandins

Physiology

- After birth:
  - GFR doubles in the first 2 weeks of life to 30-40 ml/min and increases to adult levels of 100-120 ml/min by 1 to 2 years of life
  - Sodium excretion decreases (increased tubular excretion)
  - Increased ability to concentrate urine
  - Renal vascular resistance decreases; blood flow increases

- Babies born < 34 weeks have low GFR (nephrogenesis is not complete)
- Tubular function is altered – tubules regulate fluids and electrolytes
- Preterm baby can dilute urine, but concentrating ability is limited
- Acid base balance
Acute Renal Failure

- Causes:
  - Prerenal – hypoperfusion - hemorrhage, sepsis, CHF, dehydration, nec, RDS, hypoxia, drugs (indomethicin)
  - Intrinsic – damage to the kidney – asphyxia, sepsis, congenital anomalies, thromboembolic disease, infection, acute tubular necrosis
  - Postrenal – obstruction to flow of urine beyond the kidney - urethral valves, UPJ obstruction, neurogenic bladder

- Clinical signs:
  - Oliguria: < 1 ml/kg/hr
  - Azotemia – BUN > 20mg/dl or rising >10mg/dl/day
  - Serum creatinine > 1.5mg/dl or rising more than 0.2 mg/dl/day

- Diagnostic workup:
  - urinalysis
  - BUN, creatinine, lytes
  - renal ultrasound
  - VCUG

Congenital Adrenal Hyperplasia (Adrenogenital Syndrome)

- Results from lack of an enzyme used by the adrenal cortex to produce cortisol and aldosterone
- Autosomal recessive
- 90% caused by 21-hydroxylase deficiencies
  - Salt losing
    - Cortisol and aldosterone production blocked
    - ↑ potassium and ↓ sodium levels – may not be evident for 1 week
  - Non-salt losing
    - Cortisol production blocked
    - Mild hyponatremia
CAH (cont)
- Females will have virilization; males at highest risk for misdiagnosis
- All infants with ambiguous genitalia should be screened
- Elevated levels of CAH by 24-36 hours of age
- If not treated: weakness, vomiting, dehydration, hyponatremia, hyperkalemia, hypoglycemia (carbohydrate metabolism)

CAH - treatment
- Hydrocortisone (replaces cortisol)
- Mineralocorticoids for salt-losing
- Sodium supplements as needed
- Girls – deal with issues of ambiguous genitalia

Case 16
- Baby born at 34+6 weeks. B/P at 3 days of age was 91/68. Blood pressures on the day before were 76/54, 83/46 and 90/59. Urine output is normal. Perfusion good. Baby with history of respiratory depression at birth. BUN <5 and creatinine 0.6.
Causes of Hypertension

- Renovascular anomalies
  - Thromboembolism
  - Renal artery stenosis
  - Renal vein thrombosis

- Intrinsic renal disease
  - Polycystic kidneys
  - UPJ obstruction
  - Acute tubular necrosis

- Endocrine disorders (CAH)

- Medications:
  - Maternal cocaine or heroin; Dexamethasone, Caffeine, theophylline, Pancuronium

Hypertension

- > 90/60 in term, 80/50 preterm
  - Use appropriate size cuff!

- Identify cause

- Treat pain, volume overload

- Antihypertensives:
  - Hydralazine
  - Propranolol
  - Captopril

Complications of Hypertension

- CHF
- Left ventricular hypertrophy
- Intracranial hemorrhage
- Cerebrovascular accident
- Encephalopathy
Case 17

- Baby A was a 498 gram female born at 25 weeks via C-section for cord prolapse. Oligohydramnios was noted at 18-20 weeks in utero. She had multiple malformations - ? positional from oligohydramnios. Pulmonary hypoplasia suspected when infant developed respiratory failure and airleak.

Case 18

- 23 year g3 p1 mom delivered at 32 weeks. History of previous 34 week infant that died within 24 hours due to bilateral polycystic kidneys, hypoplastic lungs, and microcephaly. Baby with a lusty cry at delivery, but had dysmorphic features including microcephaly.
Case 19

- Delivered at 32 weeks via C-section due to twin gestation. Systolic BP 90 to 110 on day 18. Echocardiograms have shown mildly diminished function with fraction shortening of 25%, mild TR. Given cardiac findings and hypertension, a renal ultrasound was done with doppler studies to determine if possible renal disease was present that may be contributing to cardiac function. Renal ultrasound showed cysts in both kidneys. Differential diagnosis included both autosomal recessive and autosomal dominant polycystic kidney disease, glomerulocystic disease and other less common diagnosis such as cystic disease of the renal medulla or multiple malformation syndromes.

Hydronephrosis

- Dilation of the pelvis and calyces of the kidney(s) due to obstruction of urine flow
- Most common congenital condition picked up by prenatal ultrasound
- Diagnostic studies – renal ultrasound, VCUG
- Complications – UTI, hypertension, damage to renal parenchyma
### Hydronephrosis – Differential Diagnosis

- **Obstructive**
  - UPJ obstruction
  - Multicystic dysplastic kidney disease
  - Ureterocele
  - Duplicated collecting system
- **Nonobstructive**
  - Physiologic dilation
  - Vesicourethral reflux
  - Prune belly syndrome

### Hydronephrosis - Treatment

- **UPJ obstruction**: antibiotics and then follow up renal work-up in 3 months
- **Pyeloplasty** if obstruction and compromised renal function
- **Posterior urethral valves**
  - Catheterize initially
  - Surgical repair if high degree of reflux
- **Vesicourethral reflux**:
  - Antibiotics
  - Cystogram in 12-18 months to determine resolution

### Renal Vein Thrombosis

- **Symptoms**
  - Hematuria
  - Flank mass
  - Thrombocytopenia
  - Hypertension
  - Oliguria
- **Diagnostic studies**
  - Renal ultrasound with Doppler flow
  - Renal scan
- **Treatment**
  - Treat underlying cause – thrombolytic therapy
Patent Urachus
- Communication between the bladder and the umbilicus
- Wet umbilicus or urine noted
- Spontaneous closure may occur if defect is small
- Surgical closure of the urachal duct

Hypospadias
- Urethral meatus on ventral surface or under penis
- Do not circumcise
- Surgical repair to move meatus distally
Exstrophy of the Bladder

- Bladder is exposed on the abdominal wall
- More common in males
- Care: Cover exposed area with sterile saline/clear plastic wrap
- Surgical closure within 48 hours
- Antibiotics
- 75-85% rate of continence
Undescended testes

- Caused by endocrine dysfunction, abnormal epididymal development, anatomic abnormality
- Complication: testicular torsion
- Treatment: surgery

Case 20

- Baby boy G delivered at 36-5 weeks via elective C-section due to fetal anomalies (skeletal dysplasia- Campomelic dysplasia). 4 vessel cord and ambiguous genitalia with bifid scrotum noted at delivery.
Ambiguous Genitalia
- Medical and psychosocial emergency
- Diagnose congenital adrenal hyperplasia
- Designate correct sex
- Correct problems early to promote normal body image and gender
- Provide genetic counseling
- Identify increased risk for gonadal tumor

Circumcision
- No absolute medical indications!
- Perform on preterm infants when they meet discharge criteria
- Complications – bleeding, infection, injury to glans, meatal stenosis, adhesions
- EMLA cream, dorsal nerve block should be used
References

- Up To Date.