Fluids

preemie 150cc/kg, neonate 90, < 30d 110
infant transfusion: 10-20cc/kg PRBC

Congenital/neonatal

sequestration:
  no airway communication
  systemic arterial blood supply
  most LLL
  extralobar: surrounded by separate visceral pleura
  extra associated with other anomalies: diaphragmatic hernia, congen ht dis
  complicated by infection, Rx resection
congenital cystic adenomatoid malformation:
  abnormal bronchi (communicate with airway v sequestration) and vasculature
  air trapping and mediastinal shift, usually lower lobes
  associated lung hamartomas
  reported malignancy, especially rhabdomyosarcoma
congenital lobar emphysema:
  hyperexpansion & mediastinal shift, mimics pneumothorax
  newborn, compromises good lung, resect
  first few days of life, LUL hyperinflation common, mediastinal shift
  Rx segmental lobectomy
bronchogenic cyst
  85% mediastinal, 15% intrapleural
  no communication with airway
  complicated by infection
diaphragmatic hernia
  Bochdalek hernia (90%)
    failure of closure of pleuropitoneal canal
    prenatal US can diagnose
    immediate postpartum distress
    posteromedial defect, scaphoid abdomen
    90% L side, < 1% bilateral
    major predictor of outcome is degree of bilateral lung hypoplasia
      medical emergency: hypoxia, hypercarbia, acidosis, pulmonary hypertension,
      persistent fetal circulation, shunt
      ECMO, nitric oxide to allow pulmonary vascular bed maturation
    aggressive resuscitation, NG, intubate
  Morgagni (5%)
    triangular space of Larrey
    90% R, M 4:1
    may be asymptomatic, incidental finding, constipation, strain
most diagnosed in childhood
abdominal approach, minimally invasive increasingly used
esophageal atresia/tracheoesophageal fistula (TEF)
   6 patterns: proximal pouch with distal tracheoesophageal communication most common
   (85%)
esophageal interruption with no communication second most common (10%)
multiple associated anomalies common
   omphalocele, gastroschisis, congenital diaphragmatic hernia
   VACERL defects
   V vertebral 2%
   A atresia/imperf anus 15%
   C cardiac 35%
   T T-E fistula
   R renal/lower GU 5%
   L limb 2%
gassless, flat abdomen if no distal TEF
distal TEF: abdomen distended, respiratory distress
   failure to pass NG to stomach (except H-fistula with continuous esophagus)
H-type: chronic cough, choking, recurrent pneumonia
R thoracotomy, retopleural dissection, single layer
   complications: leak, stricture, reflux
omphalocele
   defect through umbilicus, peritoneal covering
   55% associated anomalies/30% chromosome abnorm.: trisomy 13, 8 (lethal), 21, cardiac, GU
Beckwith-Wiedeman: macroglossia, islet cell hypertrophy, hemihypertrophy, Wilms kidney tumor, cardiac, GU
   severity of anomalies determine repair:
   mild: primary repair
   moderate: stage repair
   severe (lethal) palliate
staged closure
some associated malrotation
gastroschisis
   associated with young maternal age
defect to R of umbilicus, no peritoneal covering
   rare associated anomalies
   small defect bowel can compress blood supply causing intestinal atresia
   protect bowel: bowel bag, NG, enlarge defect laterally if necessary
   assess possibility of primary repair vs silo/staged reduction
   reduction = controlled compartment syndrome
   assess effect on respiration (most critical), renal perfusion
   some associated malrotation
meconium ileus/cystic fibrosis
   neonate meconium ileus, older fecal impaction
   soap bubble appearance, extraintestinal calcification if perforation

7 April 2009
mucomyst, fleets, gastrografin enema (not PO)

duodenal atresia/stenosis
  failure to recanalize @ 8-10w
  maternal polyhydramnios
  mucosal web with normal wall most common
  common association with trisomy 21, congenital heart disease, malrotation, second atresia
  85% distal to papilla
  bilious vomiting, double bubble sign on X-ray, no distal air
    (if distal air is present consider malrotation w volvulus v duodenal atresia, surgical emergency)
  Rx: duodeno-duodenostomy, instill saline distally to check for other atresias

malrotation/midgut volvulus
  80% manifest in neonatal period
  bilious vomiting
  Ladd’s bands compress duodenum
  UGI gold standard for Dx (94% sensitivity): corkscrew 4th portion duodenum, reversal
  SMA/SMV
  Rx: release bands, straighten duodenum, place colon L, small bowel R, appendectomy
  midgut volvulus surgical emergency, commonly age 3w

jejuno-ileal atresia
  few associations: Down’s, heart, duodenal atresia
  four types
    I web
    II fibrous cord
    IIIa discontinuity
    IIIb discontinuity, failure of SMA formation, entire small bowel fed by iliocolic
    IV multiple atresias
  in utero vascular accident, polyhydramnios prenatal US
  late in development, so few associated anomalies
  bilious emesis at 2d, distention, obstructive pattern
  barium enema microcolon (unused), no reflux into small bowel
  Rx: taper dilated proximal bowel to do anastomosis
    stomas, antegrade enemas to dilate bowel, conserve length, prevent short bowel

t necrotizing enterocolitis (NEC)
  95% in preemie, bowel necrosis, first week, of unknown etiology
  proposed etiologies: immature gut, premature feeding, colonized bacteria, stress
  early Dx: feeding intolerance, distention, hematochesia
  late Dx: shock, DIC, acidosis, thrombocytopenia, pneumatosis, fixed loop (dead), gassless
    abdomen, portal v gas
  non-operative management if non-toxic, no perforation
    temporize with peritoneal drain, may be definitive
    tenderness alone does not warrant surgery
  operation: perforation, fixed loop, worsening; most patients need OR

Hirschsprungs
  failure of distal colon innervation from neural crest cell migration
  normal innervation progresses from cranial to caudal, no skip areas
absence of ganglion cells Meissner’s, Auerbach’s plexi (large nerve trunks present), failure of distal colon propulsive wave, relaxation of internal sphincter (unopposed sacras parasympathetics), functional obstruction
rectosigmoid transition point most common
presentation at any age, male, assoc w trisomy 21
neonate acute obstruction, no meconium 1st 24h, feeding difficulty, distention
child: chronic constipation (1BM/2w)
BE: contracted rectum, transition zone, dilated proximal colon
full thickness trans-rectal suction Bx for Dx
eliminate non-ganglionated segment, intraop (FS) Bx for transition zone
leveling colostomy
Soave, Duhamel pull through of ganglionated bowel (laparoscopic)
primary pull thru less anastomotic disruption than staged?
pre-op complication: Hirschsprung’s associated enterocolitis (HAEC)
post-op complications: anastomotic leak, incontinence, stricture, constipation
most excellent results
can be complicated by enterocolitis
imperforate anus
VACTERL complex of anomalies
identify and treat life-threatening anomalies first
then anoplasty in neonatal period
colostomy rarely necessary
high (above levators) or low (through levators) rectal stump determines repair strategy
low stump: perineal anoplasty in the neonatal period
high stump: staged, colostomy followed by anoplasty
rectourethral/rectovesicle, vaginal, perineal fistula association
fecal incontinence
30% after treatment of Hirschsprungs, imperforate anus; also spina bifida
conservative bowel regimen often unsuccessful
antegrade colonic irrigation through appendiceal orifice
regain colonic tone
90% successful, 2/3 complete continence
if unsuccessful need end colostomy

Infants/children

circumcision
decreases HIV, UTI, penile cancer, cervical cancer (not chlamydia)
pyloric stenosis
4-6w, male 4:1, Caucasian, monozygous twins
postprandial non-bilious projectile emesis, failure to thrive, dehydration
FH 2X risk (mother also)
Dx: palpable olive sometimes, 65% Dx by US v clinical exam alone, UGI rarely used now
earlier referral pt in better shape: correct fluids and lytes before OR
Rammstedt pyloromyotomy (open or scope)
begin feeds 6h post op, progress over 24h
65% PO vomiting normal
intussusception
most common cause intestinal obstruction 6mo-2y
commonly occurs in 1st year, males 3:2
idiopathic, hypertrophic lymphoid tissue after viral infection
healthy infant with sudden progressive abdominal pain, distention, bloody/current-jelly stool (15%)
85% have mass, usually RLQ
95% occur at ileocecal valve
< 10% lead point: Meckels, polyp, lymphoma, foreign body
   (adult 90% lead point, 50% malignant)
Dx: US, BE
Rx
   if no shock or peritonitis, pt < 2: air or contrast enema reduction, 85% success
   if shock/peritonitis, failed enema, OR: push, don’t pull
   > 48h higher failure rate and perforation
colonoscopy not indicated
Meckels most prevalent congenital GI anomaly, 2%, M>F
most common source occult GI bleed child, young adult
juvenile polyps less common than Meckel’s
hernia/hydrocele
   60% of inguinal hernias detected 1st year, common in preemies
   no radiographic studies indicated
   Rx hi ligation of sac
   incarcerated: testicular atrophy most common complication
hydrocele: enlarged scrotum with flat inguinal canal
   90% resolve spontaneously, non-communicating < 12mo
undescended testicle
   3% higher incidence preemie, low birth weight
undescended, agenesis, retractile, ectopic, atrophy (vascular accident, injury during herniorrhaphy)
retractile 75% descend within 3 mo
most undescended in inguinal canal
reasons to do orchidopexy: cancer potential (40X), cosmesis, fertility, torsion, trauma
cancer risk unchanged after orchidopexy, but allows surveillance
testicular torsion
most common cause of acute scrotum in child
no radiographic studies indicated
may be associated with appendix epiploica testis (Mullerian and mesonephric remnants),
   blue dot sign on transillumination, assoc w hydrocele
surgical emergency: if in doubt, explore
testicular atrophy most common complication
biliary
choledochal cyst: jaundice, RUQ pain, mass
Dx: US, HIDA, ERCP
Rx: resection, Roux-Y hepaticojejunostomy
stones:
- bilirubinate/hemolytic anemia: spherocytosis, sickle cell
- cholesterol: obesity, TPN, cystic fibrosis, choledochal cyst
- sclerosing cholangitis associated with inflammatory bowel disease
- biliary atresia: untreated death < 2
  - Kasai procedure < 60d, 30% do not need transplant
- cirrhosis and portal hypertension remain problems

GERD
- common in infants, usually resolves by 15 mo
- vomiting, apnea (etiology of some sudden infant deaths?) most common symptoms
  - also asthma, pneumonia, failure to thrive
- pH monitor best diagnostic test for reflux

Rx
- conservative: position, medical Rx
- operative indications: near-SID episode, esophagitis, recurrent pneumonia, failure to thrive, do Nissen wrap

neck mass
- lateral: lymphadenopathy, branchial cleft cyst/fistula, salivary gland
  - Tb test, mono-spot or Ebstein-Barr titer, FNA, open Bx
- branchial cleft
  - 1 to external auditory canal to ant to ear
  - 2 thru carotid bifurcation into tonsilar fossa
- midline: lymph node, thyroglossal duct cyst, dermoid, thyroid
  - 1% of thyroglossal duct cysts have aberrant thyroid tissue
    - commonly 2nd decade/10-20
    - thyroid scan pre-op to R/O lingual thyroid
    - 25% may be lateral to midline
- posterior: lymph node, metastatic tumor, lymphoma (teen)

lymphangioma/cystic hygroma
- multiloculated cyst due to lymphatic malformation
- neck most common, then axilla, extremity, trunk
- most common complication infection
- Rx goal: excise without sacrificing vital structures

hemangioma
- most common soft tissue tumor of childhood (5-10% < 1Y olds)
- constellation of hamartomatous lesions arising from vascular tissue
- natural Hx: rapid growth, stable period, involution (most by 5y)
- pulse dye laser (target Hb) for superficial
- excise if complicated by rapid growth, bleeding, ulceration; failure of involution by school age

Malignancy: leukemia most common malignancy

solid tumors (Wilms and neuroblastoma the two major tumors of children)

Wilms (kidney)
- most common (10%) abdominal tumor of childhood, average age 3
asymptomatic abdominal mass, anorexia, fever, wt. loss, hematuria (10%, w trauma)
Dx: CT, assess contralateral (5%)
calyceal distortion on IVP
extension to IVC, 5% metastatic at Dx (lung)
majority are stage I or II and 90% have favorable histology
no Bx prior to definitive Rx
Rx: nephrectomy and chemo (doxycycline, actinomycin D, vincristine)
    radical nephrectomy for stage III, IV, unfavorable histology
spillage most significant factor for poor outcome
it is rarely necessary to resect other structures
favorable histology  overall survival 80% for all stages

neuroblastoma
most common solid tumor of childhood
80% younger than 2
younger better prognosis, increased VMA
    invade vertebral foramen
80% mets at Dx: bone, marrow, liver, nodes, lung
50% arise in adrenal, 24% paraspinal, 20% mediastinum, 4% cervical, 2% pelvis
tumor products (VMA), CT
no Bx prior to definitive Rx
multimodality therapy including marrow transplant
prognosis depends on age, stage, tumor biology
    young (< 1y) do better than older
stage    I  95% 5y
       II  80%
      III 35%
     IV 12%

hepatoblastoma more common than hepatic cell carcinoma, better prognosis

ovarian teratoma
most common ovarian tumor of childhood (most during reproductive age)
70% benign under age 30; 3 germ layers
cystectomy with ovarian preservation (open or laparoscopic)
4% of benign recur, 1% chance malignant transformation
follow child with annual ultrasound

sacroccygeal teratoma
3 germ layers
most benign (malignant endodermal sinus tumor, embryonal more in older child)
intrapelvic, may have intraabdominal component
surgical excision: control middle sacral a. first, preserve anus, excise coccyx (contains tumor)
follow-up for benign and malignant recurrence

myelomeningocele
lumbar neural and vertebral defects
Miscellaneous

appendicitis
most commonly misdiagnosed < 3 for mesenteric adenitis, gastroenteritis
CT less reliable in child, less fat stranding
scorpion bite child
autonomic nervous system activation, depolarization neuromuscular junctions
intense local pain, blurred vision, dyspnea, incontinence, M spasm
Rx: airway control, sedation, cardiac monitor for arrhythmias, calcium gluconate for M spasms,
(no narotics, exacerbate neurotoxic effects)
no tourniqued, debride
pediatric head injuries
leading cause of traumatic death in child, 40% of deaths < 1y
poor prognostic indicators: age < 1y, BP < 135, need for vasopressors, hi glucose, low bicarb, hi ICP, GCS < 8 1st 24h
hyperventilation no benefit
GCS rising to > 8 1st 6h marked increase in survival
complete androgen insensitivity syndrome (CAIS)
X-linked single gene causing androgen receptor gene mutation
46XY, but phenotype female
bilateral testes, breast development at puberty
testes often in hernia sac
remove testes @ puberty, cancer risk
no female organs, treat as female
hereditary spherocytosis
autosomal dominant, defect in RBC membrane structural protein spectrin
shortened RBC lifespan, osmotic fragility
anemia, reticulocytosis, jaundice, splenomegaly, pigment gallstones (94% by 13)
increased osmotic fragility, high sequestration, destruction by spleen
wait until at least age 4 before splenectomy, cholecystectomy at same time

Differential diagnosis

intestinal obstruction
air reaches the colon 6-12h post-partum
neonate double bubble: duodenal atresia, annular pancreas
3w double bubble: malrotation
3-12w vomiting: pyloric stenosis (male predominant, FH)
3w-18mo vomiting: intussusception
respiratory distress
diaph hernia
TEF
congenital lobar emphysema
solid tumor
  < 2 neuroblastoma
  > 2 Wilms

References: