Vesalius SCALpel™ : Pediatrics (see also: pediatric surgery folios)

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

congentital 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 pleuroperitoneal 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, gastroschesis, 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, retropleural 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

gastroschesis
- 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 v 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

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

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
sclersoing 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

sacrococcygeal 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:

Warner B. Whats new in pediatric surgery. JACS, 199(2), Aug. ’04: 273-284.