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What is this lecture about?

- Pediatric – exclusive to or commonly seen in kids
- Not a Zebra (ie something I've either seen during my career or know has been seen in our PED)
- Not a horse either – ie something you may make it through training without seeing
- Emergency practitioner can make the diagnosis or at least suspect it
- Emergency practitioner *should* make the diagnosis or at least suspect it, and can make a difference by either getting diagnostic studies, appropriate consultations, and starting initial management or by not doing unnecessary work-up

Quick info in 3 slides

- Classic case – build on illness scripts to reinforce when you should suspect this entity
- What should make you suspect this diagnosis
- Basics of diagnosis and ED management
- You can look it up for more detail, but you can't look it up until you at least suspect it
- FOAM resources for additional readings

How to use this lecture

- After the initial case presentation, think about the differential diagnosis
- Helpful framework: SPIT
  - *What is the most Serious diagnosis?*
  - *What is the most Probable diagnosis?*
  - *What is the most Interesting diagnosis?*
  - *What is the most Treatable diagnosis (ie what diagnosis should the EP do something about ASAP)?*
- Write down what you think is the diagnosis – commit!
  - *At the end, see how many you got right*

12 day old trisomy 21 patient with abdominal distension and constipation

- 12 day old trisomy 21 ex-38 week gestation born NSVD, no complications, no cardiac problems, home at 48 hours old
- + prenatal care, no infections, no complications other than trisomy 21
- Passed meconium on day of discharge
  - *Since then, passing small smear of stool only every 3-4 days and increasing abdominal distension*
  - *No vomiting, fever, decreased po intake*
- VS: temp 37.5, HR 144, RR 28, BP 72/36, O2 sat 99% room air
- PE positive only for abdominal distension, nontender, no mass palpable, no hepatosplenomegaly, digital rectal exam results in explosive bowel movement, no hard stool

2 week old with jaundice

- 2 week old ex-38 week infant born NSVD to 32yo G1P1 mom, +prenatal care, no complications, discharged at 36 hours of life
  - *Patient is exclusively breastfeeding*
  - *Mom blood type A, baby blood type unknown*
- Parents note increasing jaundice over last few days, light-colored stools and dark urine
- VS temp 37.5, HR 140, RR 32, BP 68/38, O2 sat 100%
- Alert, anterior fontanelle soft and flat, icteric, jaundiced
- Lungs clear, heart regular rate & rhythm, no murmur
- Abdomen soft, nontender, liver edge palpable 4cm below SCM
- Total bilirubin 10.5 mg/dL, conjugated (direct) 6 mg/dL

3 ½ week old with vomiting

- 3 ½ week old male ex-full term infant born NSVD with no complications
- Brought in because vomits non-bilious after every feed
  - *Baby eats eagerly and seems hungry soon after vomiting*
  - *No fever, diarrhea, apparent colicky abdominal pain*
- History of presumed chlamydial conjunctivitis at age 10 days, treated with po azithromycin
- VS temp 37.5, HR 140, RR 32, BP 68/34, O2 sat 100%, abdominal exam soft nondistended nontender
  - *Baby is observed to vomit in the ED*

Video can be found at https://www.youtube.com/watch?v=JKmVHusL4Ms

4 week old with diarrhea, lethargy, and cyanosis

- 4 week old ex-full term infant with 2 days of diarrhea, watery and yellow, non-bloody, became increasingly lethargic and cyanotic over today
  - *Birthweight 2.78 kg, poor weight gain, on cow's-milk formula*
  - *No vomiting, ill contacts: parent with bloody diarrhea & low-grade fever x 3 days*
- Temp 39, HR 170, RR 60, BP 64/40, O2 sat 81% on room air
- Dry mucous membranes, sunken fontanel, sunken eyes, poor skin turgor, capillary refill 3-4 seconds
- Lungs clear to auscultation, cardiac regular rate & rhythm, no murmur
- Abdomen soft, nondistended, nontender
- Cyanotic, with improvement of O2 sat to only 84% with O2

5 week old with cough and cyanosis

- 5 week old with paroxysms of cough and perioral cyanosis
- One week of viral URI symptoms including runny nose and cough, no fever
  - *+ ill contacts, college student uncle with prolonged cough illness*
  - *Everyone in family has immunizations up to date*
- Temp 37.7, HR 170, RR 30, BP 74/35, O2 sat 97% on room air
- Well-appearing when not coughing, no respiratory distress
- Paroxysms of cough, non-productive, not barking, no whoop, associated with perioral cyanosis, no apnea

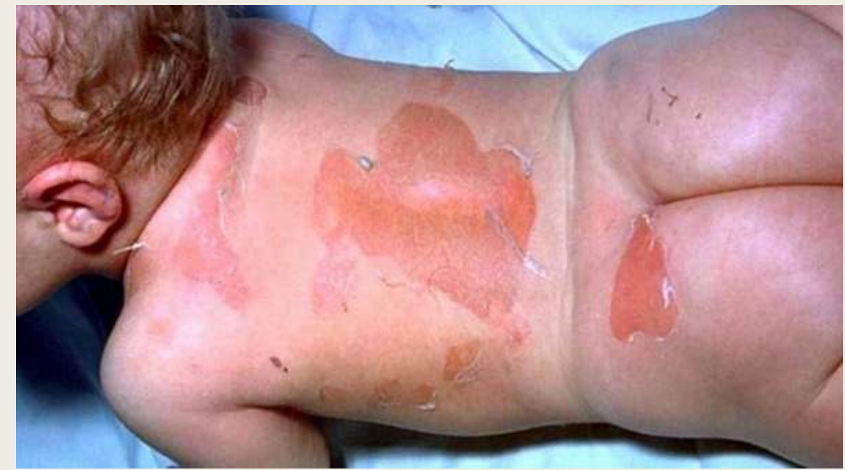
6 week old with vomiting

- 6 week old female, ex 38 week infant born NSVD to 28yo G1P1 mom, no complications
- Brought in for vomiting x 3 that day
  – *Vomit is on the baby's blanket and seen here*
- No diarrhea, hematochezia, hematemesis
- VS temp 37.7, HR 140, RR 32, BP 74/34
- Parents also feel that the baby's abdomen appears mildly distended, but it is soft and nontender, no discoloration or ecchymoses

3 month old with SOB

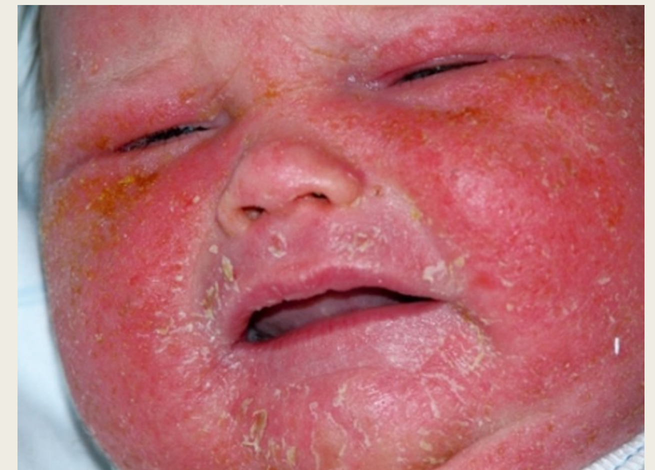
- 3 month old ex-full term infant, born NSVD with no complications, home in 2 days
- Over the last week, infant has been having more difficulty breathing and breathing faster. Also, the infant feeds more slowly, taking 40 min to feed 2 ounces.
- VS 37.7, 165, 60, 75/35, O2 sat 95% room air
- Alert, tachypneic, mild intercostal and subcostal retractions, lungs with coarse crackles diffusely, heart regular rate and rhythm, 3/6 harsh murmur LLSB
- What other physical exam finding do you want to check?

3 month old with fever and rash



www.247wellness.org

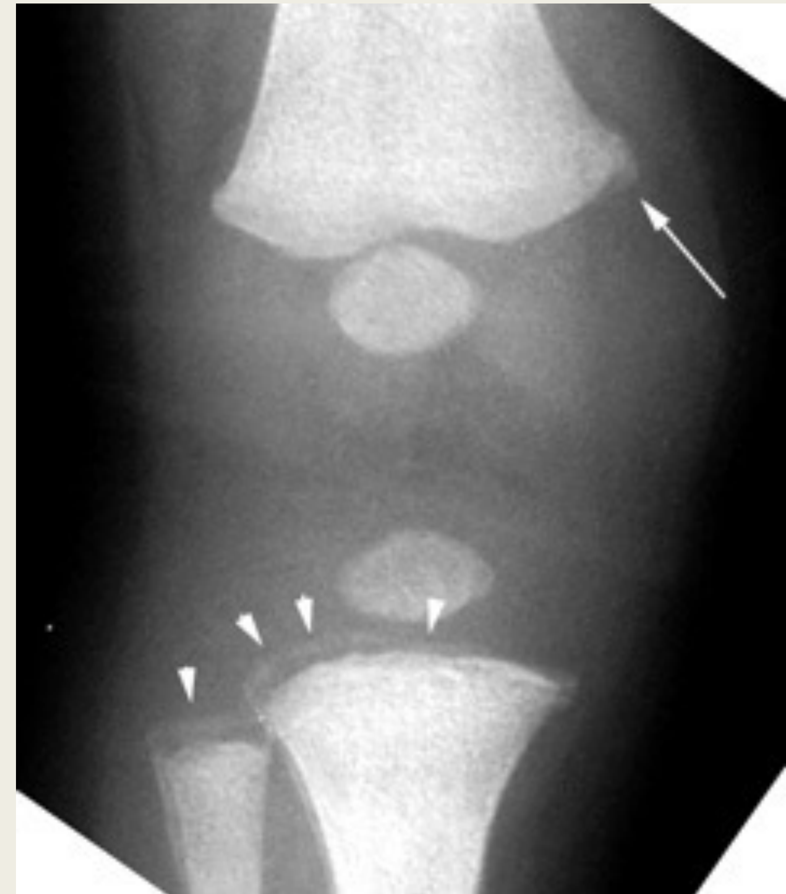
- 3 month old ex-full term infant born NSVD with no complications
- Seen in ED yesterday for erythematous skin rash to underarms and face, low grade fever
  – *Discharged with diagnosis viral exanthem*
- Returns today due to spread of rash with crusting on face, and blistering and peeling on trunk
- Temp 39, HR 170, RR 30, BP 70/40
- Toxic appearing, rash seems tender
  – *Touching rash causes top layer of skin to peel off*



www.hxbenefit.com

3 month old with decreased right leg movement

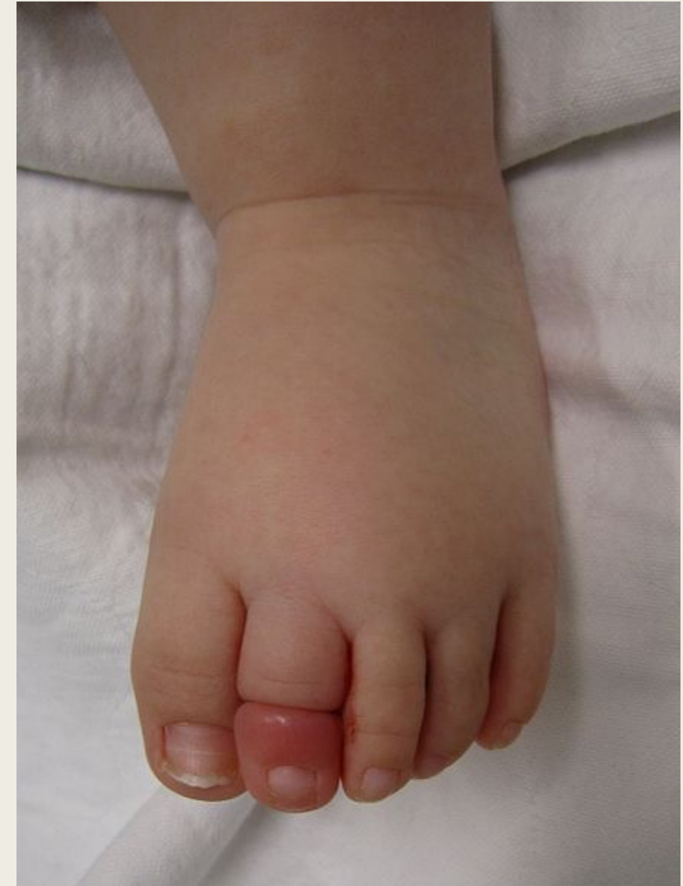
- 3 month old brought in for decreased movement of right leg x 1 day
- Was being carried by 4yo sibling and was dropped 2 feet onto carpeted floor
  - *No loss of consciousness, no vomiting*
  - *Since this fall, however, not moving right leg much, is more fussy*
- Ex full term NSVD no complications
- VS temp 37.5, HR 144, RR 28, BP 74/43
- Swelling around right knee and apparent pain with passive ROM at right knee
- No evidence of head trauma, no bruising, no deformity



http://www.meddean.luc.edu/

4 month old with swollen toe

- 4 month old ex-full term infant born by c/s for failure to progress to 34yo G1P1 mom with no complications
- Brought in for swollen, red 2nd toe, noted for one day
- No fever, no known trauma although patient is in daycare
- VS temp 37.6, HR 140, RR 32, BP 76/34, remainder of exam unremarkable



Did you write down what you thought the answers were? Answers on following slides

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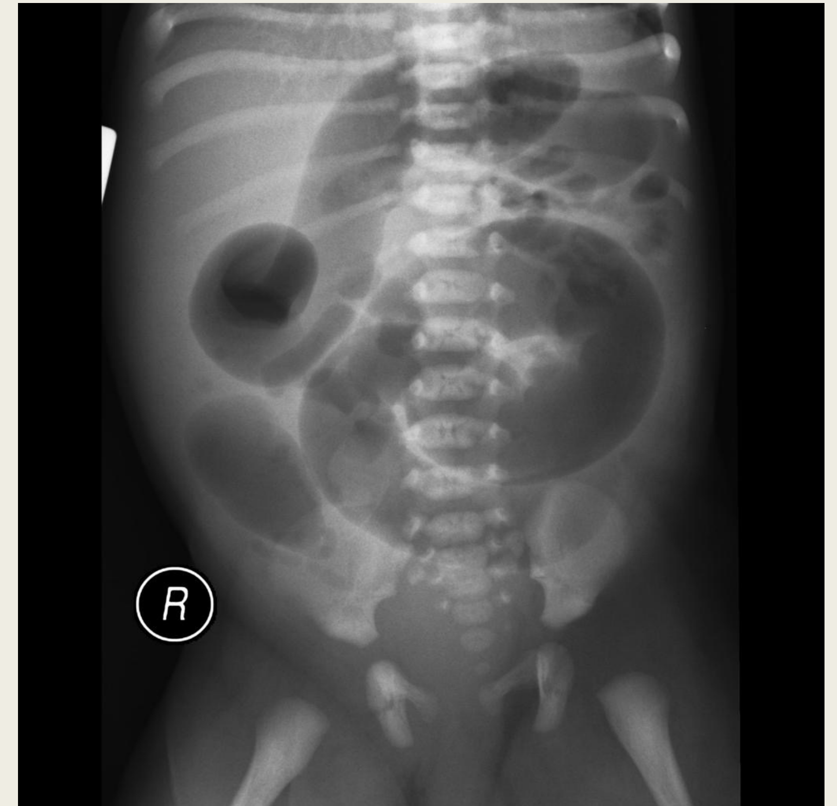
Hirschsprung disease – suspect it

- Presents as constipation, occasionally as enterocolitis, toxic megacolon
- Neonatal presentation
  - *Delayed passage of meconium at > 24 hours of life in 90%*
  - *Abdominal distension, feeding intolerance, bilious vomiting may occur*
  - *Digital rectal exam may produce explosive stool / gas expulsion*
- Childhood (sometimes even adulthood) presentation
  - *Chronic constipation (may develop at time of weaning from breast milk)*
  - *May have partial aganglionosis*
- 10% present with enterocolitis: fever, abdominal distension, diarrhea
  - *May be severe, progress to toxic megacolon*
  - *May develop even after surgical repair of Hirschsprung disease*
- Hirschsprung disease associated with many syndromes and congenital anomalies, particularly trisomy 21

Hirschsprung disease – diagnose and manage it

- Plain film KUB may show dilated loops of bowel, paucity of gas in rectum
  - *Neither sensitive nor specific*
- Water-soluble contrast enema demonstrates “transition zone” between normally innervated colon and aganglionic region
- Anorectal manometry, biopsy may be performed by GI specialist
- Definitive treatment is surgical
- Enterocolitis: stabilize first with NPO, fluid resuscitation, NG tube decompression, IV antibiotics, rectal irrigations
- For more info: http://pedemmorsels.com/hirschsprungs-disease-hd-and-enterocolitis/ and https://radiopaedia.org/articles/hirschsprung-disease



radiopaedia.org

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Biliary atresia – suspect it

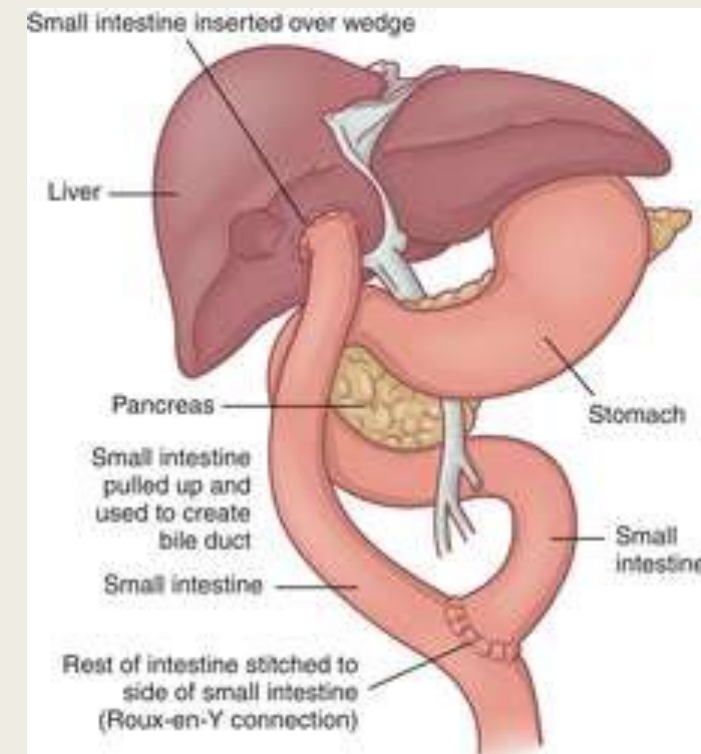
http://getdocsays.com/neonatal-jaundice/

- Physiologic jaundice rarely persists beyond 2 weeks
- Breast milk jaundice begins at the end of the 1st week of life (days 4-7) and may persist for several weeks
- Direct (conjugated) hyperbilirubinemia ≥ 2 mg/dL or 20% of total bilirubin is ALWAYS abnormal
  - *Think biliary atresia, although there are other less common causes*
  - *The earlier biliary atresia is diagnosed, the better the outcome*
  - *Conjugated (direct) bilirubin may be normal or only slightly elevated if checked soon after birth*
  - *In jaundiced infant ≥ 2 weeks old, check total **and** direct bilirubin*
  - *Elevated GGT harbingers biliary atresia diagnosis*
- 1:10,000-15,000 live births, increased in Asians, esp Chinese

Biliary atresia – diagnose and manage it

http://medical-dictionary.thefreedictionary.com/portoenterostomy

- Biliary ultrasound has 80-90% sensitivity
- Consult pediatric gastroenterologist and pediatric surgeon, admit for further work-up
- May need hepatic scintigraphy, liver biopsy, intraoperative cholangiogram
- Treatment: Kasai procedure or liver transplant
- Pearl: post-Kasai patient presenting with fever needs to have blood culture and be admitted and treated with IV antibiotics to r/o cholangitis (occurs in 30-60%)
- For more info: http://pedemmorsels.com/biliary-atresia/
- For more on neonatal jaundice: http://empem.org/2011/06/neonatal-jaundice/ and http://pedemmorsels.com/hyperbilirubinemia/



Kasai procedure

3 ½ week old with vomiting

Projectile vomiting

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Pyloric stenosis – suspect it

- Typically presents at 3-6 weeks of age, rarely at > 12 weeks
- More common in males, first-born, pre-term, if mom smoked during pregnancy, if treated with po macrolide at ≤ 14 days old
- Projectile vomiting, non-bilious, immediately after feeding, hungry after vomiting
- Dehydration over time, but typically presents earlier when not yet dehydrated
- May be associated with hyperbilirubinemia
- Maintain a high index of suspicion in infants with vomiting after every feed and appropriate age
  - *Observe feeding and aftermath in ED*

Pyloric stenosis – diagnose and manage it

- Classically, palpable “olive” mass in RUQ of abdomen
  - *Helpful if infant quietly sucking on gloved finger or pacifier + sucrose*
- Classically electrolytes show hypochloremic metabolic alkalosis from prolonged vomiting (lose HCl of gastric acid)
- Currently, patients usually present earlier and these classic findings may not be seen / appreciated
- Diagnosis made by ultrasound
- Treatment: NPO, IVF rehydration, consult pediatric surgeon
- For more information: http://pedemmorsels.com/pyloric-stenosis/ and http://dontforgetthebubbles.com/pyloric-stenosis/ and for infant vomiting ddx http://pemplaybook.org/podcast/vomiting-in-the-young-child-nothing-or-nightmare/

4 week old with diarrhea, lethargy, and cyanosis

- 4 week old ex-full term infant with 2 days of diarrhea, watery and yellow, non-bloody, became increasingly lethargic and cyanotic over today
  - *Birthweight 2.78 kg, poor weight gain, on cow's-milk formula*
  - *No vomiting, ill contacts: parent with bloody diarrhea & low-grade fever x 3 days*
- Temp 39, HR 170, RR 60, BP 64/40, O2 sat 81% on room air
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- Lungs clear to auscultation, cardiac regular rate & rhythm, no murmur
- Abdomen soft, nondistended, nontender
- Cyanotic, with improvement of O2 sat to only 84% with O2
  - *CXR and EKG are normal*

Methemoglobinemia – suspect it

https://www.medvet.umontreal.ca

- Infant's Met-Hb level was 31%
- Ferrous (Fe++) iron in hemoglobin oxidized to Ferric (Fe+++) form, which can't bind oxygen
- Commonly seen after exposure to oxidizing agents, especially benzocaine (topical for ENT procedures or for teething)
- Young infants with diarrhea: bacteria can reduce nitrate to nitrite, which then induces methemoglobinemia
  - *At higher risk also because fetal Hb is more easily oxidized and infant has alkalotic gut environment conducive to gram negative organism growth*
  - *History c/w milk protein intolerance (poor weight gain) -> leads to gut inflammation and increased risk as well*
- Blood dark red or chocolate in color, doesn't change when exposed to O2

Methemoglobinemia – diagnose and manage it

- O2 sat often plateaus around 85% no matter what the PaO2 is
- Diagnose by obtaining blood gas with co-oximetry to measure methemoglobin level
- Discontinue offending agent
- Treat dehydration with NS bolus(es)
- Give O2 to ensure full saturation of available normal hemoglobin
- Treat levels > 20% and symptomatic with methylene blue 1%, 1 to 2 mg/kg IV over 5 min
  - *Should see improvement in next 15-30 minutes, repeat as needed*
- Admit for continued monitoring, supportive care, and repeat doses as needed
- For more info: http://lifeinthefastlane.com/ccc/methaemoglobinaemia/ and http://toxicology.ucsd.edu/art%202%20methemoglobin.pdf

5 week old with cough and cyanosis

- 5 week old with paroxysms of cough and perioral cyanosis
- One week of viral URI symptoms including runny nose and cough, no fever
  - *+ ill contacts, college student uncle with prolonged cough illness*
  - *Everyone in family has immunizations up to date*
- Temp 37.7, HR 170, RR 30, BP 74/35, O2 sat 97% on room air
- Well-appearing when not coughing, no respiratory distress
- Paroxysms of cough, non-productive, not barking, no whoop, associated with perioral cyanosis, no apnea

Pertussis – suspect it

- Classic pertussis 3 phases
  - *Catarrhal stage viral URI 1-2 weeks*
  - *Paroxysmal stage 1-6 weeks (up to 10 weeks or longer), paroxysms of cough with whoop at end (children < 6 months old often no whoop)*
  - *To hear a whoop: http://www.whoopingcough.net/symptoms.htm*
  - *Convalescent stage 1-2 weeks*
- Infants can have apnea, cyanosis & hypoxemia, post-tussive emesis, choking spells, shorter catarrhal stage
  - *Mortality from pertussis is primarily in infants*
- Adolescents and adults with waning immunity have only a prolonged cough illness and serve as a reservoir of infection for infants

Pertussis – diagnose and manage it

- Suspect based on clinical features
- CBC often shows high WBC count with lymphocytosis
- PCR nasopharyngeal swab or aspirate confirms pathogen
  - *Most common Bordatella pertussis*
  - *Less commonly Bordatella parapertussis*
- If apnea, cyanosis, hypoxemia with paroxysms, admit to monitored setting for close observation
  - *High index of suspicion to admit for infants < 3 months old*
- Macrolide antibiotic if suspect even before diagnostic testing confirmation
  - *If given early in disease, may limit duration*
  - *Treatment recommended later to limit spread of disease, even if patient will not benefit*
- For more info: http://www.emdocs.net/wp-content/uploads/2015/03/Pertussis-Chase-.pdf and http://pedemmorsels.com/pertussis-still-a-problem/

6 week old with vomiting

- 6 week old female, ex 38 week infant born NSVD to 28yo G1P1 mom, no complications
- Brought in for vomiting x 3 that day
  - *Vomit is on the baby's blanket and seen here*
- No diarrhea, hematochezia, hematemesis
- VS temp 37.7, HR 140, RR 32, BP 74/34
- Parents also feel that the baby's abdomen appears mildly distended, but it is soft and nontender, no discoloration or ecchymoses

Bilious emesis

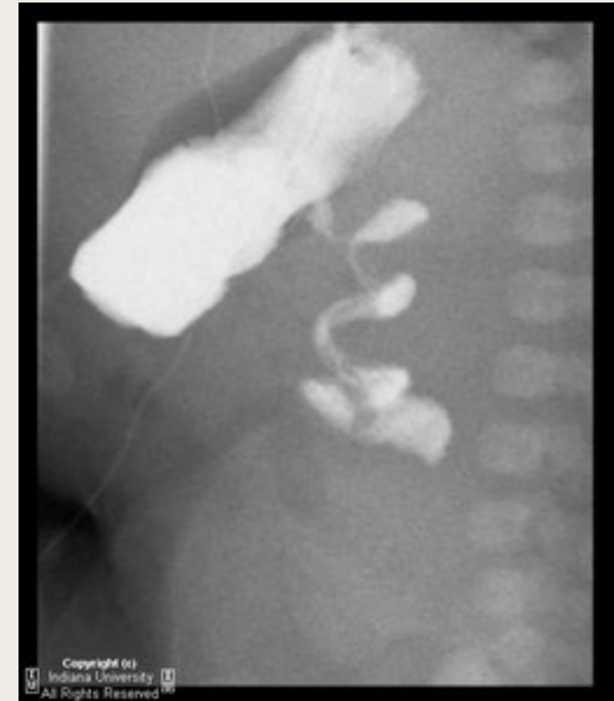
http://pedsurgzone.blogspot.com/2010/09/how-often-is-bilious-emesis-in-new-born.html

Midgut volvulus – suspect it

- Bilious vomiting in an infant – pediatric surgery consult to r/o midgut volvulus
  - *Presentation can be insidious, and baby may appear well initially and decompensate rapidly*
- Classically, majority present at < 1 year of age, especially < 1 month, but may present throughout life including in adulthood
  - *Older children and adults may have a more chronic picture of abdominal pain and vomiting that may not be bilious*
- Later, as ischemia occurs, may have abdominal distension, tenderness, signs of peritonitis, hematochezia, shock
- Associated anomalies common

Midgut volvulus – diagnose and manage it

- Maintain high level of clinical suspicion = key
- If ill-appearing, NPO, IVF, empiric antibiotics for possible intestinal perforation, emergent pediatric surgery consult for surgery
- If stable, consider KUB/XTL (not highly sensitive, can look for perforation)
- Upper GI contrast study 96% sensitivity (corkscrew sign)
- For more information: http://pedemmorsels.com/malrotation/ and on infant vomiting in general http://pemplaybook.org/podcast/vomiting-in-the-young-child-nothing-or-nightmare/



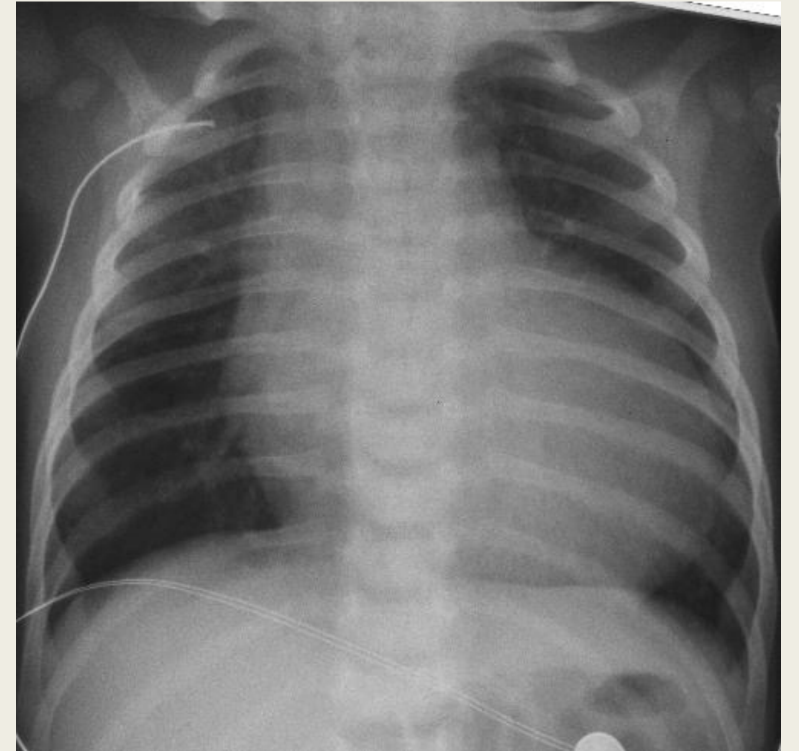
https://www.mypacs.net/cases/31711482.html

3 month old with SOB

- 3 month old ex-full term infant, born NSVD with no complications, home in 2 days
- Over the last week, infant has been having more difficulty breathing and breathing faster. Also, the infant feeds more slowly, taking 40 min to feed 2 ounces.
- VS 37.7, 165, 60, 75/35, O2 sat 95% room air
- Alert, tachypneic, mild intercostal and subcostal retractions, lungs with coarse crackles diffusely, heart regular rate and rhythm, 3/6 harsh murmur LLSB
- What other physical exam finding do you want to check?
- Hepatomegaly

Congestive heart failure due to congenital heart disease – suspect it

- Left to right shunt with increased pulmonary flow
- Commonly presents at 2 to 6 months of age
- VSD, PDA, AV canal
- SOB, tachypnea, irritability, poor feeding / sweating with feeding, weight loss are common complaints
- Tachypnea and tachycardia, crackles or rales, murmur, hepatomegaly

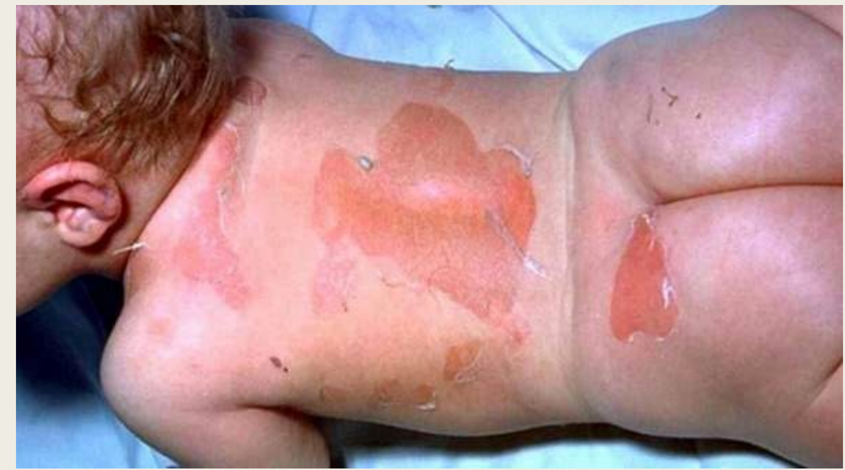


http://www.hawaii.edu

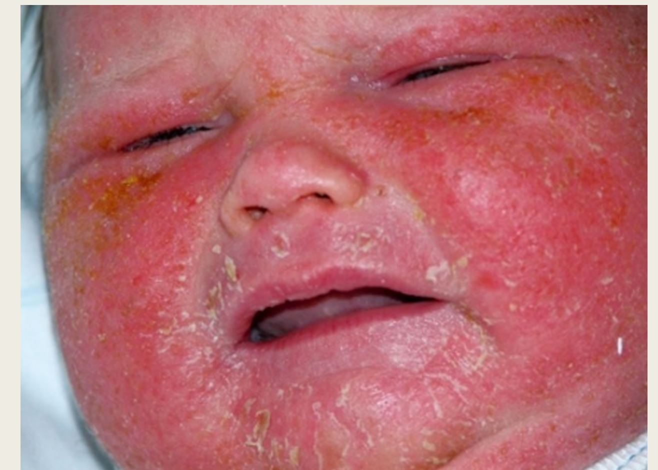
Congestive heart failure – diagnose and manage it

- Diagnosis: CXR and EKG, echocardiogram for definitive diagnosis
  - *CXR cardiomegaly, pulmonary congestion*
  - *EKG may show ventricular hypertrophy*
- Check electrolytes, calcium, and glucose and correct as needed
- Furosemide 1 mg/kg IV
- Dopamine or dobutamine or milrinone (in consultation with PICU, cardiology) for hypotension
- CPAP or BiPAP may be helpful for significant respiratory distress
- For more info: http://pedemmorsels.com/subtle-signs-of-pediatric-heart-failure/ and http://emergencymedicinecases.com/congenital-heart-disease-emergencies-2/

3 month old with fever and rash


www.247wellness.org

- 3 month old ex-full term infant born NSVD with no complications
- Seen in ED yesterday for erythematous skin rash to underarms and face, low grade fever
  - *Discharged with diagnosis viral exanthem*
- Returns today due to spread of rash with crusting on face, and blistering and peeling on trunk
- Temp 39, HR 170, RR 30, BP 70/40
- Toxic appearing, rash seems tender
  - *Touching rash causes top layer of skin to peel off*


www.hxbenefit.com

Staph scalded skin syndrome – suspect it

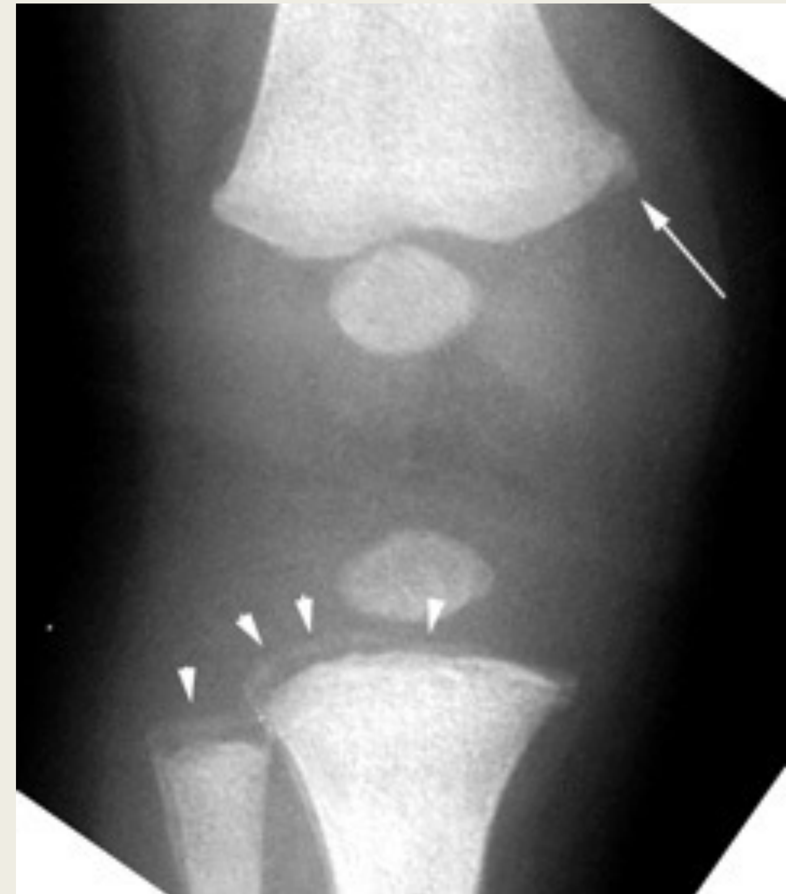
- Young children aged < 5 years typically, most common in $\leq$ 3 months old
- Fever, irritability, redness of skin that looks like a sunburn
  - *Commonly affected areas: armpits, groin, around nose and mouth, diaper area or periumbilical for neonates*
  - *Over 24-48 hours rash spreads, blisters, and peels*
  - *Nikolsky sign: gentle stroking of the skin results in sheets of skin exfoliating*
- Due to infection with a toxigenic strain of *Staphylococcus aureus*
- AKA Ritter disease, Lyell disease in newborns / infants
- Treated appropriately, heals without scars within 1 week usually

Staph scalded skin syndrome – diagnose and manage it

- SSSS is a clinical diagnosis
  - *Differentiate from toxic epidermolysis necrosis (which is treated differently) – SSSS has no mucous membrane involvement*
  - *Skin biopsy may eventually be performed to confirm diagnosis*
- CBC, blood culture, basic metabolic panel (at risk for dehydration)
- Consult with ID expert, dermatologist
- Admit as inpatient, anti-staphylococcal antibiotic eg oxacillin or nafcillin; vancomycin if at risk for MRSA; alternatives: cefazolin, ceftriaxone
  - *Clindamycin sometimes added in toxin-mediated staphylococcal disease due to possible antitoxin effect – unclear if beneficial in SSSS*
- For more information http://pedemmorsels.com/staph-scalded-skin-syndrome/ and http://www.dermnetnz.org/topics/staphylococcal-scalded-skin-syndrome/

3 month old with decreased right leg movement

- 3 month old brought in for decreased movement of right leg x 1 day
- Was being carried by 4yo sibling and was dropped 2 feet onto carpeted floor
  - *No loss of consciousness, no vomiting*
  - *Since this fall, however, not moving right leg much, is more fussy*
- Ex full term NSVD no complications
- VS temp 37.5, HR 144, RR 28, BP 74/43
- Swelling around right knee and apparent pain with passive ROM at right knee
- No evidence of head trauma, no bruising, no deformity
- Radiograph shows distal femur chip fracture, proximal tibia and fibula avulsion fractures



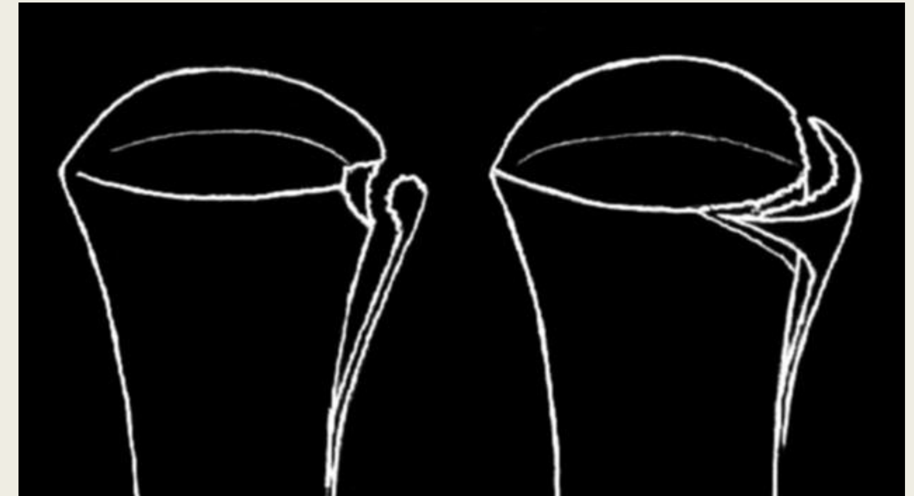
http://www.meddean.luc.edu/

Classic metaphyseal lesion (bucket handle and corner fractures) – suspect it

- Pathognomonic for physical abuse
  - *Thought due to jerking or twisting of extremity, or shaking of child*
- If exam concerning for fracture, obtain radiographs even if history not supportive, as caregivers may be falsifying history
- Concerning for abuse
  - *Blaming mechanism of injury on sibling*
  - *Child is preambulatory and has lower extremity fractures*
  - *Multiple injuries*
  - *Delay in seeking care*
  - *Caregiver keeps changing history*
  - *Evidence of injury and no history of accidental trauma*

Classic metaphyseal lesion (bucket handle and corner fractures) – diagnose it and manage it

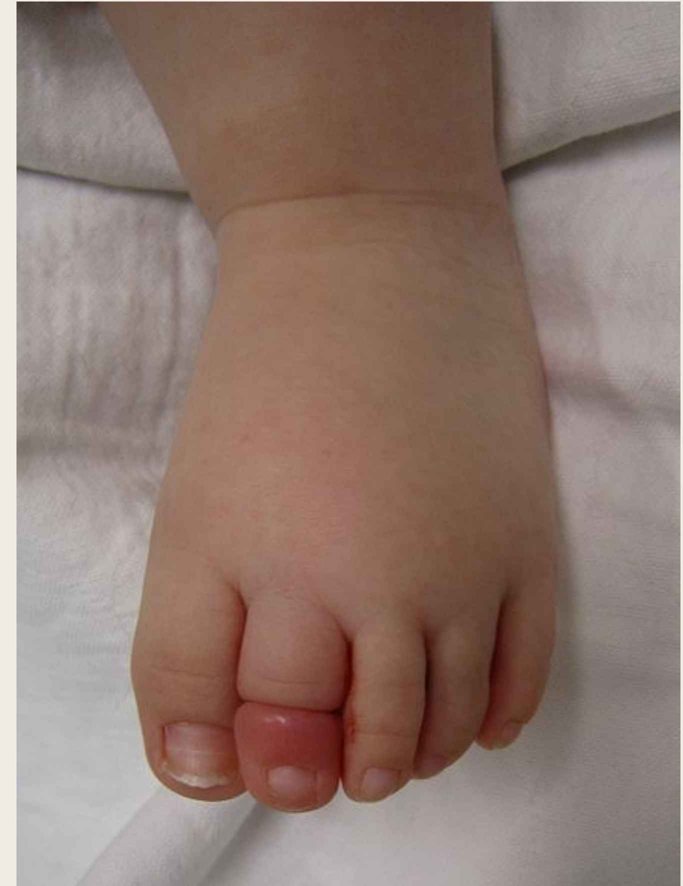
- Classic metaphyseal lesion appears as a corner-shaped chip avulsion fracture in one view, and as a thin rim of avulsion fracture (similar to a bucket handle of an upside-down bucket) in a perpendicular view
- Consult orthopedics and child abuse specialist
- Perform skeletal survey to look for additional fractures
- For more info: http://www.hawaii.edu/medicine/pediatrics/pemxray/v4c02.html and https://emrems.com/2015/10/14/student-corner-pediatric-non-accidental-trauma/



http://www.hawaii.edu/medicine/pediatrics/pemxray/v4c02.html

4 month old with swollen toe

- 4 month old ex-full term infant born by c/s for failure to progress to 34yo G1P1 mom with no complications
- Brought in for swollen, red 2nd toe, noted for one day
- No fever, no known trauma although patient is in daycare
- VS temp 37.6, HR 140, RR 32, BP 76/34, remainder of exam unremarkable



https://en.wikipedia.org/wiki/Hair_tourniquet

Hair/thread tourniquet – suspect it

- Hairs, usually from parent/caretaker with long hair, or threads from socks, mittens, sheets, blankets, get wrapped around appendage
  - *Toes, fingers, penis common, less commonly clitoris, uvula*
  - *Toes, penis more commonly hair, fingers more commonly threads*
- All ages, but typically presents in infants, median age 4 months
- Swelling may make it difficult to see the hair or thread
  - *If untreated, hair or thread may cut into skin with re-epithelialization over the top, obscuring hair/thread completely*
  - *Occasionally can cut down to bone*

Hair/thread tourniquet – diagnose and manage it

- Clinical diagnosis
- Pain management – consider digital block or even procedural sedation
- If able to unwind hair/thread, do so to remove
- Try to lift or grasp hair/thread and cut it
  - *May need consultation with surgeon, obtain micro-instruments and magnification*
- If hair, can try depilatory cream (should work in 10 minutes)
- May require incision perpendicular to the hair/thread (ie long axis of finger/toe) – incise dorsal finger or toe to avoid flexor tendon, neurovascular structures
- Post removal, apply antibiotic ointment and follow up for wound check
- For more information: http://pedemmorsels.com/hair-tourniquet/