100 CARDINAL PED PRESENTATIONS

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CASES 1-10

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

3 day old with vomiting

- 3 day old ex-full term (39 weeks) with vomiting x 1.5 days, abdominal distension, and streaks of blood in stools
- No fever, no ill contacts
- Born NSVD with Apgars 1 and 9 (required some PPV) to mom with adequately controlled diabetes, discharged at 28 hours of life after noted to be bottle-feeding well
- Temp 37.6, HR 170, RR 40, BP 65/35
- Abdomen soft, moderately distended, nontender, hypoactive bowel sounds, no masses palpable
- You obtain a screening KUB



https://pediatriceducation.org

3 day old with blood in diaper

- 3 day old male ex 36 week infant born NSVD to 38yo G2P2 mom, no complications
- Brought in because of concern that there is blood in diaper
- VS: temp 37.6, HR 140, RR 36, BP 72/34
- No fever, vomiting, diarrhea, blood in stool, apparent abdominal pain; breastfeeding avidly
- Stool guaiac is negative
- Diaper as shown



http://www.babycenter.ca/thread/ 2482927/blood-like-spot-in-diaperwith-pic

3 day old with vomiting, lethargy

- 3 day old ex-38 week infant born 2.9 kg, Apgars 9 and 10, +prenatal care, no complications, home yesterday
- Since home, not feeding well, increasingly lethargic, vomiting nonbilious, non-bloody
- VS: temp 37.6, HR 180, RR 60, BP 62/30, lethargic and minimally responsive, hypotonic; anterior fontanelle soft and flat, shallow tachypnea w/o increased work of breathing, cardiac exam no murmur, abdomen soft, nondistended
- Labs: pH 6.9, WBC 3,000 (50%N, 50%L), Hgb 9, Plt 21,000. Na 139, K 4.8, Cl 110, HCO3 5, anion gap 24, ammonia level 324 mcg/dL, UA 2 wbc, 0 rbc, 4+ ketones, bedside glucose 24

4 day old with rash

- 4 day old ex- full term 39 4/7 weeks infant, born NSVD to G3P3 mother, no prenatal or birth complications, home in 2 days
- Breastfeeding well, transitional stools, good urine output
- Mom noted rash after she had eaten peanuts and then breastfed baby
- Temp 37.6, HR 140, RR 30, BP 65/36
- Physical examination aside from rash unremarkable, no respiratory difficulty, no swelling; baby well appearing
- Starts with small papules, then yellowish papulovesicular appearing lesions surrounded by halo of erythema
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mddk.com



med.stanford.edu/newborns/

5 day old with conjunctivitis

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- Hospital records reviewed, received erythromycin eye ointment prophylaxis
- Parents wipe away discharge but it comes back right away
- Mom has h/o PID x 2 that was treated prior to this pregnancy, GBS negative prior to delivery, RPR and HIV negative during pregnancy
- VS: Temp 37.5, HR 140, RR 30, BP 68/38
- Eye exam as shown, remainder of physical exam noncontributory, no rash



http://iahealth.net/conjunctivitis/

5 day old with vaginal bleeding

- 5 day old girl ex-38 wk born NSVD to 28yo G2P2 mom, +prenatal care, no complications, home on 2nd day of life
- Feeding well, breastfeeding 15 min / side, q 2-3 hours
- Brought in due to concern of vaginal bleeding seen in both the diaper and at the vaginal introitus
- Not copious amounts
- No history of trauma, cared for by parents and doula
- Exam normal infant female genitalia, no active bleeding currently, no evidence of trauma

6 day old brought in by ambulance for BRUE

- 6 day old ex-37 wk born at 3.2 kg
- Born via c/s for repeat
- History of cleft palate only
- Using special bottle
- Parents have noticed baby is jittery
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APLS Slide Set

- VS temp 37.5, HR 140, RR 35, BP 65/33, wt 3.14 kg, O2 sat 99%, well appearing now
- Cleft palate, harsh 3/6 murmur LLSB, exaggerated Moro reflex with jitteryness
- What labs do you want to order?

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 Based of cord goopy and malodorous. Circumferential periumbilical erythema approximately 2cm diameter

8 day old with shock

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- Presents pale, dusky, lethargic, mottled
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10 day old male with poor growth, lethargy

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- Patient has been growing poorly despite frequent feeding, and then over past day increasingly irritable, lethargic, and not feeding
- Patient has been vomiting also, non-bloody, non-bilious; no fever
- Temp 37.6, HR 190, RR 40, BP 50/30
- Baby appears scrawny, lethargic, dehydrated with sunken eyes and fontanel, poor skin turgor, capillary refill 4 seconds
- CBC WBC 17.1, no left shift, Hgb 15, Hct 45, Plt 206, Na 125, K 7.0, Ca 9.6

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

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Necrotizing enterocolitis – suspect it

- Preterm infants at a few weeks of age (after starting enteral feeds)
- Term infants account for < 10% cases, typically age 1-3 days
- Can occur as late as 1 month of age
- Often risk factor for term infant, eg birth asphyxia, congenital heart disease, or decreased fetal gut blood flow from eg maternal diabetes, placental insufficiency, maternal cocaine abuse
- Vomiting (may be bilious), feeding intolerance (residuals), hematochezia, abdominal distension
- May rapidly progress to sepsis, shock, intestinal perforation, 15-30% mortality
- Bonus: suspect related disease ("neutropenic enterocolitis" or "typhlitis") in pediatric neutropenic oncology patient with abdominal pain, often RLQ, bloody diarrhea, vomiting

Necrotizing enterocolitis – diagnose and manage it

- Pneumatosis intestinalis = air in the bowel walls
- Seen in 50-70% of NEC cases, pathognomonic
- Look also for portal venous gas, free air
- Abdominal US in technically skilled may be diagnostic
- Treatment: NPO, NGT decompression, fluid resuscitation, full sepsis evaluation and broad-spectrum antibiotics (ampicillin, cefotaxime, metronidazole common regimen), consult pediatric surgeon, admit to NICU or PICU
- For more info:

https://radiopaedia.org/articles/necrotising-enterocolitis-1 and http://pedemmorsels.com/necrotizing-enterocolitis/ and http://pedemmorsels.com/pneumatosis-intestinalis/ and http://pedemmorsels.com/typhlitis/



http://acuclinic.com.au/pocit/NEC.htm

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Urate crystals, "pink diaper syndrome"

- Newborns excrete more uric acid
- Urate crystals can form and cause a reddish-orange (sometimes described as brick-colored) stain in the diaper, which can be mistaken for blood
- Common with mild dehydration, eg exclusive breastfeeding in first few days of life when breast milk is not fully in yet
- Colostrum has high protein content favoring acidic urine, which favors urate crystal formation
- Make sure infant adequately hydrated, reassure parents, and give follow up to monitor for good urine output (at least 5-6 wet diapers/day)

Other funny colored diapers

- Serratia marcescens in the urine can produce a red color that is mistaken for hematuria
- An inborn error of tryptophan absorption can result in "blue diaper syndrome"
- For more information:

http://informedparent.com/articles/view/whenblood-in-the-diaper-is-not-blood-urate-crystals and https://pedclerk.bsd.uchicago.edu/page/abnorma l-urine-colors and https://rarediseases.org/rarediseases/blue-diaper-syndrome/



http://www.bjmp.org/content/medicine -pictures-purple-urine-bag-syndrome

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What labs should you order?

Inborn error, organic acidemia - suspect it

- Often presents in first days of life; sometimes presents in older children, adolescence, adults
- Poor feeding, vomiting, lethargy progressing to coma, hypotonia, seizures, decompensation with acute illness
- Suspect inborn error? Order CBC, Chem panel (iStat if ill appearing or seizure), VBG, ammonia, lactate, pyruvate, bedside glucose, urine/serum ketones
- Anion gap metabolic acidosis, high ammonia (but < than urea cycle disorders), ketosis
- Also common: pancytopenia, hypoglycemia



Ruzkova et al. Ped Anesth 2015;25(8):807-817

PA: propionic acidemia MMA: methylmalonic acidemia IVA: isovaleric acidemia (sweaty feet odor) MSUD: maple syrup urine disease

Inborn error, organic acidemia - diagnose and manage it

- Send serum organic acids, urine and serum amino acids, free and total carnitine levels, but treat prior to confirmation
- NPO (take away protein substrate for pathway), IVF D10 ½ NS at 1.5x maintenance rate (goal glucose 8-10 mg/kg/min)
- May need insulin at some point to prevent hyperglycemia
- Treat any hypoglycemia with 2cc/kg D10, give bolus 20 cc/kg NS if dehydrated, perform r/o sepsis and give empiric antibiotics, supportive care as needed (airway, ventilation, shock management)
- Consider bicarb 0.5-1 mEq/kg if severely acidotic
- Carnitine 25-100 mg/kg/day divided into 3 daily doses IV
- Consult with genetics immediately and admit
- For more info: <u>https://www.ahcmedia.com/articles/136251-metabolic-emergencies</u>

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med.stanford.edu/newborns/

Erythema toxicum – suspect it

- Benign, self-limited, common (seen in approximately 50% newborns)
- Not commonly seen in preterm infants
- 98% were born at least 35 weeks gestation
- Not usually present in first few days of life
- Commonly presents age 3 days to 2 weeks





www.dermnetnz.org

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Erythema toxicum – diagnose it and manage it

- Diagnosis is a clinical diagnosis
- Lesions may be confused for urticaria
- If lesion contents are stained (Wright, Gram) and examined under a microscope, will see increased eosinophils, but this test is unnecessary
- If pustular lesions, may need other tests to r/o other pustular diseases or infections; if vesicular lesions must r/o HSV
- Self-resolves, typically over 2 weeks
- For more information on common newborn rashes: <u>http://med.stanford.edu/newborns/professional-education/photo-gallery/skin.html</u>

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http://iahealth.net/conjunctivitis/

Neonatal gonococcal conjunctivitis (ophthalmia neonatorum) – suspect it

- Profuse discharge and eyelid swelling
- 1st week of life, peak 2-5 days of age
- If clinically suspect, pursue diagnosis even if outside this age range as other factors may delay presentation (prophylaxis, inoculum size, transmission after delivery, etc)
- May occur despite receiving prophylaxis
- Resistance of organisms to erythromycin (23% in one study)
- Usually transmitted via vaginal delivery
- May have intrauterine infection after rupture of membranes (even if has c-section for failure to progress later)
- Increased risk if mother has risk factors for STDs, no prenatal care
- Note: chlamydia conjunctivitis much more common, presents 5-14 days of age, often milder presentation

Neonatal gonococcal conjunctivitis (ophthalmia neonatorum) – diagnose and manage it

- Gram stain of discharge shows gram-negative kidney bean shaped diplococci
- Culture on specialized Thayer-Martin media (obtain from lab)
- But, before confirmation, if clinically suspect, consult Ophthalmology emergently, IV cefotaxime 100 mg/kg (ceftriaxone 50 mg/kg also, but risk of hyperbilirubinemia), admit for frequent eye irrigation and continued antibiotics
- Risk for scarring, vision loss if inadequately treated
- Should also treat empirically for chlamydia infection and test for HIV
- Note: chlamydia is intracellular, so specimens must be conjunctival scraping
- Tx chlamydia with po erythromycin 50 mg/kg/day x 14 days +/- topical erythromycin ointment, need test of cure
- Pneumonia occurs in 10-20% infants with inadequately treated chlamydial conjunctivitis
- Evaluate and treat mother and partner(s)
- For additional information: <u>http://eyewiki.aao.org/Neonatal_Conjunctivitis</u>

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- Feeding well, breastfeeding 15 min / side, q 2-3 hours
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- Exam normal infant female genitalia, no active bleeding currently, no evidence of trauma

Neonatal estrogen effect and withdrawal bleeding – suspect it

- Neonates are exposed to mom's high estrogen levels via the placenta
- Levels slowly fall over first few months of life
- Female infants can have withdrawal vaginal bleeding similar to menses, usually starts in the first few weeks of life and goes away after a few months
- Both female and male infants can have breast swelling and even some fluid leakage ("witch's milk")
- Hormones can also produce neonatal acne, usually at age 2 weeks – 2 months



Neonatal estrogen effect and withdrawal bleeding – diagnose and manage it

- Clinical diagnosis based on appropriate age
- Management requires only explanation of cause and reassurance
- Make sure patient received Vitamin K prophylaxis at birth
- Advise parents to NOT squeeze the nipples or massage / frequently palpate the breasts can develop mastitis
- Mastitis requires rule out sepsis work-up, admission for IV antibiotics, consult with pediatric surgeon
- For more info: <u>https://medlineplus.gov/ency/article/001911.htm</u> and <u>http://www.pedchrome.com/2012/11/breast-engorgement-in-newborns.html</u> and on mastitis <u>https://pediatriceducation.org/2016/10/17/what-causes-neonatal-mastitis/</u> and on neonatal acne <u>http://www.dermatology.ca/skin-hair-nails/skin/acne/baby-acne/</u> with pictures at <u>http://pimplespictures.com/Neonatal-Acne-Pictures.php</u>

Absent thymus

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- Cleft palate, harsh 3/6 murmur LLSB (c/w VSD), exaggerated Moro reflex with jitteryness
- What labs do you want to order?

Neonatal seizures (DiGeorge syndrome hypocalcemia) – suspect it

- Spectrum of disorders (22q11 deletions): cleft palate, cardiac defects esp conotruncal, hypocalcemia from hypoPTH, absent thymus, immunodeficiency / recurrent infections, certain facial features
- CATCH-22: Cardiac, Abnl facies, Thymic aplasia, Cleft palate, Hypocalcemia
- Neonatal seizures can be subtle: jitteryness, bicycling, lip-smacking, chewing movements, eye-blinking
- Order ASAP: bedside glucose, iSTAT Na, Ca



Low set ears

telecanthus with short

palpebral fissures

Facial features of children with DiGeorge syndrome

https://www.slideshare.net/ghalan/14-primaryimmunodeficiency-diseases

Neonatal seizures (DiGeorge syndrome hypocalcemia) – diagnose and manage it

- Seizures with metabolic abnormalities correct the abnormality
- Hypocalcemia 10% calcium gluconate 100 mg/kg = 1 mL/kg
- Hypoglycemia D10W 2 cc/kg
- Hyponatremia NS 20 cc/kg or 3% saline, 3 cc/kg
- Other considerations: ECG (long QT), utox, consider inborn errors, consider non-accidental trauma, consider TORCH infections, consider meningoencephalitis
- Confirmed through genetic studies; consult geneticist
- Admit for stabilization of hypocalcemia and further w/u
- For more info: <u>http://pedemmorsels.com/neonatal-seizures-not-a-benzo-deficiency/</u>
- For info about DiGeorge: <u>https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4445702/</u>

7 day old with umbilical problem

- 7 day old ex-full term infant born NSVD and discharged home 2 days after birth with no complications and no maternal infections
- Doing well until evening prior to presentation when became fussier, not feeding well, and then began to develop erythema around umbilicus. No fever, vomiting. Parents have been cleaning cord with alcohol pad once a day as instructed; no other substances applied.
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 Based of cord goopy and malodorous. Circumferential periumbilical erythema approximately 2cm diameter

Omphalitis – suspect it

- Peak presentation at age 5 to 9 days in full term infants
- Association with "dry cord" care (not using antiseptic on cord at birth)
- May have symptoms c/w sepsis: irritability, lethargy, poor feeding, fever (or as in this case, hypothermia)
- Circumferential erythema (to ddx from irritation by cord pressed onto skin), warmth, tenderness, purulent umbilical discharge
- Unsure and baby well appearing? Consider leaving diaper off for 15-20 minutes – erythema from irritation should resolve



https://pedclerk.uchicago.edu/ page/omphalitis

Omphalitis – diagnose and manage it

- Clinical diagnosis
- R/o sepsis work-up: CBC, Blood cx, bedside glucose (all critically ill kids), consider CRP, LP, urine, CMP, DIC labs if septic
- Imaging: plain film to r/o subQ air of necrotizing fasciitis (10-16% progress to nec fasc), US to assess ddx cellulitis vs nec fasc
- Broad spectrum antibiotics to cover gram+ (staph esp) and gram-
- Oxacillin or nafcillin, vancomycin, gentamicin
- Consider clindamycin or metronidazole for anaerobic coverage
- Consult with ID, and surgery if suspect nec fasc or myonecrosis)
- Suspect if violaceous, bullae, peau d'orange, crepitus, petechiae, rapid progression despite antibiotics
- Admit all to ICU setting high mortality rate
- For more info: <u>http://pedemmorsels.com/omphalitis/</u>
- To learn about a benign diagnosis in the differential, umbilical granuloma: <u>http://pedemmorsels.com/umbilical-granuloma/</u>

Figure 9. Ultrasound image of necrotizing soft tissue infection. Gas with reverberation artifacts (arrow heads) can be seen within the subcutaneous tissues.



Image courtesy of Remedica Journals http://www.remedicajournals.com/CML-Dermatology/Browselssues/Volum sue-1/Article-Cutaneous-Abscesses-Current-Review-of-Microbiology-Sonog

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Ductal-dependent systemic flow in congenital heart lesion – suspect it

- First 1-2 weeks of life (typically near end of first week into second week)
- Shock and/or cyanosis
- Cyanosis does not improve with 02
- May or may not have a murmur
- HLHS, aortic coarctation, critical aortic stenosis
- Look for pulse / pulse ox differential between RUE and other extremities

DUCTAL DEPENDENT LESIONS



http://www.slideshare.net/dpark419/thecrashing-cardiac-baby

Ductal-dependent systemic flow in CHD – diagnose and manage it

- Consider and, if indicated, treat empirically for other common causes of critically ill neonate: sepsis, inborn error, hypoglycemia, volvulus
- Start PGE1 as soon as clinically suspect based on classic presentation, even before cardiology consult or confirmatory studies
- 0.05 to 0.1 mcg/kg/minute
- Side effects: apnea, hypotension
- If intubate, be careful; resuscitate to intubate, avoid RSI drugs that worsen hemodynamics (eg benzos worsen hypotension, ketamine increases SVR)
- Don't give high FiO2; oxygen constricts the ductus
- For more info: <u>http://www.emdocs.net/sick-neonate/</u> and <u>http://emergencymedicinecases.com/congenital-heart-disease-emergencies-2/</u> and <u>https://emdutch.com/2013/09/25/ductal-dependent-lesion-in-neonates/</u>

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Congenital adrenal hyperplasia w/saltwasting crisis – suspect it

- Girls born with CAH present with ambiguous genitalia with virilization
- Boys have normal appearing genitalia
- Salt-wasting form: present at 1-4 weeks of age with dehydration, irritability, lethargy, vomiting, failure to thrive, fluid-refractory shock
- Hypotension, hyponatremia, hyperkalemia, hypoglycemia (deficient in glucocorticoids and mineralocorticoid) may be present
- May have hyperpigmentation, best seen at genitalia and areolae



Infant presenting with ambiguous genitalia From the personal collection of Dr Ingrid Holm

https://online.epocrates.com/diseases/ 868/Genetic-disorders-of-sexualdevelopment

Congenital adrenal hyperplasia w/saltwasting crisis – diagnose and manage it

- Elevated 17-hydroxyprogesterone in classic 21-OH deficiency CAH
- Consult with endocrinologist regarding what labs to send (prior to treatment with any glucocorticoids)
- Cortisol level, aldosterone level
- Correct any hyponatremia, hyperkalemia
- Check bedside glucose and treat any hypoglycemia
- Normal saline fluid bolus for patients in shock, reassess and repeat fluid boluses as needed
- Stress dose hydrocortisone IV 1-2 mg/kg or 25 mg for an infant
- For more info: <u>http://pedemmorsels.com/congenital-adrenal-hyperplasia/</u> and also a sort of related interesting case <u>http://www.mededmasters.com/clinical-case-series/by-the-way</u>