**Cases of the week, from Blair Hammond, MD, Mt. Sinai, NY**

**This week's case is thanks to our teaching resident, Theresa Zhou!**

**History:** 4 y F with painful lesion to right third finger x 2 days. Patient also has crusted lesions around right eye…

## Diagnosis: Herpes simplex virus type 1 infection – manifesting as herpetic whitlow & eczema herpeticum

**Etiology:** Inoculation of HSV-1 at mucosal or skin sites, which permits entry of virus into epidermis, dermis, and eventually sensory and autonomic nerve endings

**Primary Infection:**

- Onset is usually sudden, with appearance of multiple characteristic vesicular lesions superimposed on

an inflammatory, erythematous base

- Primary infection may also be associated with systemic symptoms – fever, malaise
- In general, severity and number of lesions is considerably less with reactivation
- Lesions can be painful and last for 10-14 days
- Vesicles usually grouped at single anatomic site; however, autoinoculation can occur

**Differential Diagnosis:**

Skin / nail infections

- Herpetic whitlow – HSV infection of the finger
    - Can occur as complication of primary oral or genital herpes by inoculation of the virus through a break in the skin
    - Majority of children with herpetic whitlow also have oral lesions
    - Untreated, gradually heal over 2-3 weeks, but have potential to recur
    - Usually appear as one or grouped vesicles on an erythematous base
    - Usually monolateral, more painful than pruritic
- Cellulitis / abscess
    - Bacterial infection of skin, usually caused by local flora that is inoculated through break in the skin
    - Important to distinguish this from herpetic whitlow, as herpetic whitlow does NOT require antibiotics or I&D
- Acute paronychia
    - Bacterial infection of nail fold, usually manifests as swelling, erythema, and purulent collection on contiguous with lateral nail, most often caused by Staph aureus
    -

**Diagnostic Approach:**

- Viral culture – unroof an alcohol-wiped vesicle and use sterile swab to place fluid from inside vesicle in viral medium. Vesicles contain highest titres of virus within first 24-48h.

- Serology – IgM antibodies may be useful in diagnosing neonatal infections; these appear during first 4 weeks of infection and persist for months. Less useful in this case.

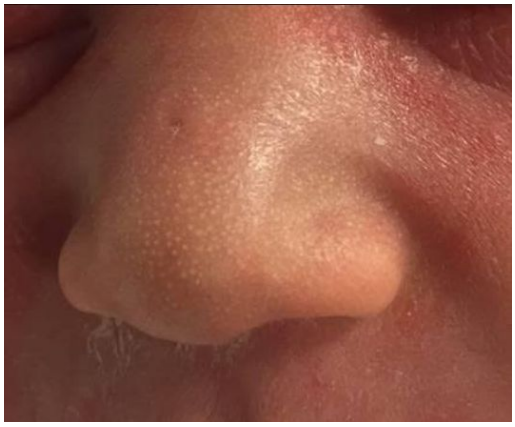
- PCR – more sensitive than viral culture, and has become gold standard for establishing diagnosis of HSV-1 infection in CSF specimens in patients with symptoms of encephalitis. Not necessary in this case.

- Tzanck smear – classic, but limited utility b/c only helpful if positive. Also will not determine if it is HSV-1 vs HSV-2 or other serotypes, unlike a viral culture.

**Treatment:** Acyclovir is useful if begun early during primary HSV infection, or if symptoms are severe. Treatment with oral acyclovir results in earlier disappearance of fever, shorter duration of lesions, decreased duration of odynophagia, and reduced viral shedding. Prompt initiation within 72 hours is important to obtain maximal clinical benefit. Usual duration of treatment is 7 to 10 days. In severe cases of odynophagia, patients may require IV acyclovir and hydration. Involvement of areas like the eye are also indications for IV vs po therapy.

**Update on our patient:** She received IV acyclovir given the proximity of her eczema herpeticum to the right eye, though evaluation by ophthalmology showed no ocular involvement (phew). The herpectic whitlow improved over a few days with treatment. Her eye initially improved after 24h of IV acyclovir, but still had continued redness of her eyelid on HD#2, and wound culture was found to be growing Staph aureus = bacterial superinfection. Clindamycin was added, and she was able to be transitioned to po acyclovir and antibiotics to continue treatment at home.

**References:** UpToDate: Clinical manifestations and diagnosis of herpes simplex virus type 1 infection *and* Treatment of herpes simplex virus type 1 in immunocompetent patients



# Diagnosis: Nasal Dermoid Sinus Cyst

**Discussion:** Dermoid cysts and dermal sinuses are congenital midline nasal malformations that are grouped under common and inclusive designation of **nasal dermoid sinus cyst (NDSC)**

**Etiology:**

- Due to abnormal embryologic development of nasofrontal region
    - Nasofrontal fontanelle connects anterior cranial fossa with the nasal bones and a transient prenasal space. A projection of the dura mater reaches the prenasal space and comes into contact with the overlying skin of the nose for a short lapse of time – creating a dural diverticulum
    - Normally, the dural diverticulum quickly involutes and retracts back into cranium, the nasofontanelle is obliterated and leaves behind the foramen cecum of the skull
    - When the dura mater does not entirely separate from the skin, the cutaneous elements are dragged back into the prenasal space. The tract can follow the usual pathway of involution, with resulting involvement ranging from very superficial to very deep (with intracranial extension)
- NDSCs have the features of normal skin:
    - Cavity lined by stratified squamous epithelium with hair follicles,

sebaceous glands

- **Hair protruding through sinus opening is pathognomonic for NDSC** (but not a constant feature) – isolated non-hairy pits are more easily overlooked!

- Filled with keratinous material (products of skin desquamation and glandular production)

*From Cambiaghi et al 2007:*

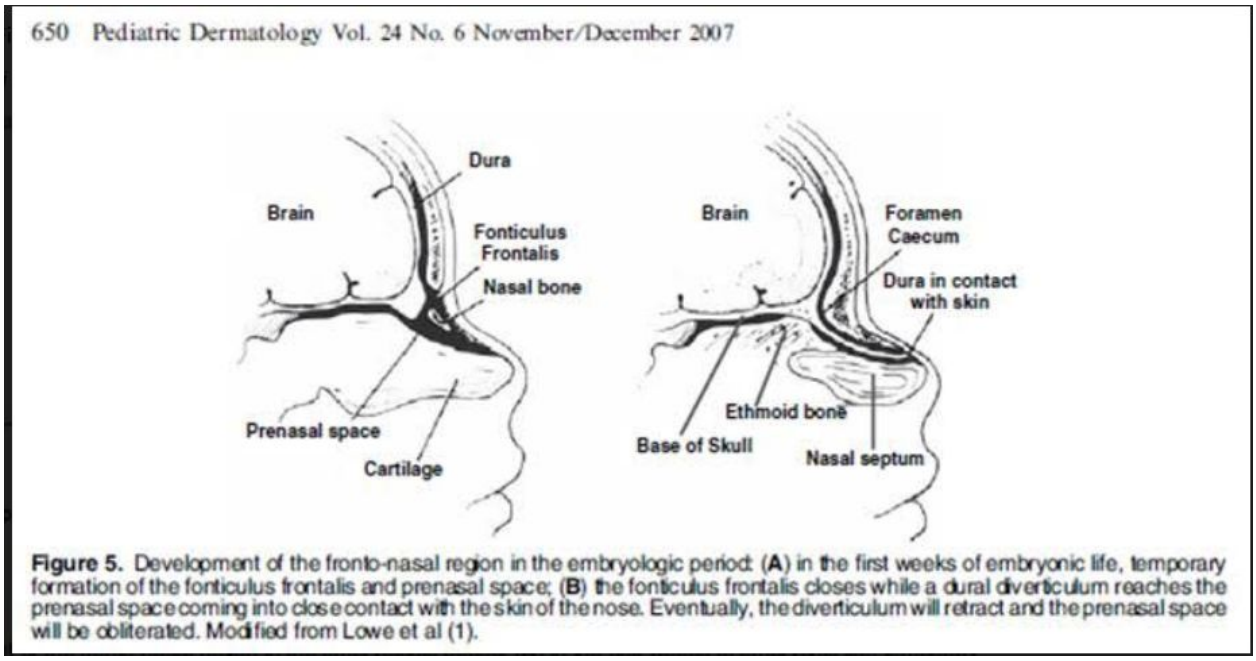



**Figure 5.** Development of the fronto-nasal region in the embryologic period: (**A**) in the first weeks of embryonic life, temporary formation of the fonticulus frontalis and prenasal space; (**B**) the fonticulus frontalis closes while a dural diverticulum reaches the prenasal space coming into close contact with the skin of the nose. Eventually, the diverticulum will retract and the prenasal space will be obliterated. Modified from Lowe et al (1).

**Epidemiology:**

- Incidence of congenital midline nasal masses is 1/20,000 – 1/40,000
    - Includes NDSCs, gliomas, encephaloceles
- Usually sporadic but isolated case reports of familial clustering
- No association of a syndrome with NDSC and no described genetic transmission
- Usually diagnosed in the first 3 years of life, but some progress to adulthood

**Clinical features:**

- Sinus ostium may open anywhere on midline of the nose, usually at the distal 23

of the dorsum of the nose

- Varying dimension and deepness, from a small pit or blind sinus, to a fistula extending into nasal structures

    - Majority have a short sinus and confined to superficial tissues

    - Some have associated dermoid cyst, may penetrate nasal septum and base of skull, reaching intracranial structures

    - Localization of lesion is not reliable in predicting degree of penetration, but one series suggested that having a sinus ostium on the dorsum of the nose is at higher risk for intracranial involvement (vs dermoid cyst without sinus opening)

- Can have a hairy tuft at site of sinus ostium

**Complications:**

- Can have recurrent infections and intermittent discharge of sebaceous material

    - Local abscesses, periorbital and mid-facial cellulitis, osteomyelitis

    - Skeletal distortion from recurrent inflammation and cystic lesion expansion

- More severe risk exists in intracranial connection: meningitis, meningoencephalitis, seizures

**Diagnosis/Treatment:**

- MRI +/- CT studies to confirm clinical suspicion and determine extent of involvement, look for possible intracranial extension

- Surgical excision, with ideal age considered to be around 2 years old by some

- Incomplete excision may result in recurrence and complications; biopsy should be avoided until intracranial connection has been excluded.

**Take home message:**

**A midline nasal lesion without hairs could be a nasal dermoid sinus cyst malformation; if hairs are present, it is definitely such a malformation. Given the potential of intracranial extension, it should be worked up appropriately with cranial/sinus imaging,**

**and in consultation with dermatology and other surgical subspecialties as warranted.**

**Our Patient:**

Following the diagnosis of a NDSC, an MRI was performed under sedation. The study demonstrated no intracranial extension. His parents were reassured and referred for out-patient management with our colleagues in pediatric plastic surgery.

**Acknowledgements:**

Kind thanks to Dr. Shanna Kowalsky for catching this lesion, and to Dr. Lauren Geller and her team for their consultation and for sharing the picture.

**References:**

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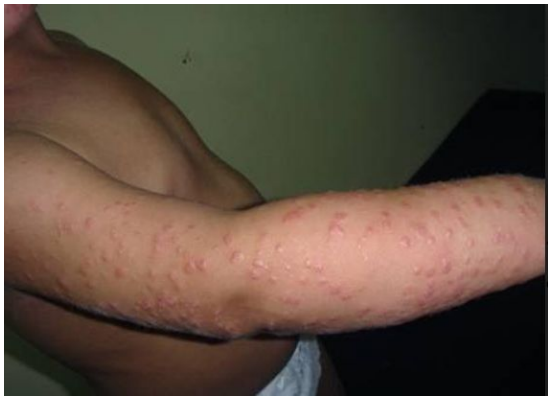
Post G, McMains KC, Kountakis SE. Adult nasal dermoid sinus cyst. Am J Otolaryngol 2005;26:403-405.

Tatar EC, Selcuk OT, Saylam G, et al. The management of rare nasal mass-nasal dermoid sinus cysts: open rhinoplasty. Rare Tumors 2009;1(2):e40.

Tolkien, JRR. The Lord of the Rings. 1959.

**This week's case is thanks to our awesome teaching residents, Kate Schwartz and Risa Hoshino!**

**First a brief pearl from Dr. Lisa Forman (a clinician-educator/general pediatrician at Elmhurst and sister of Joel, daughter of Ed) about our Gianotti Crosti case last week...**



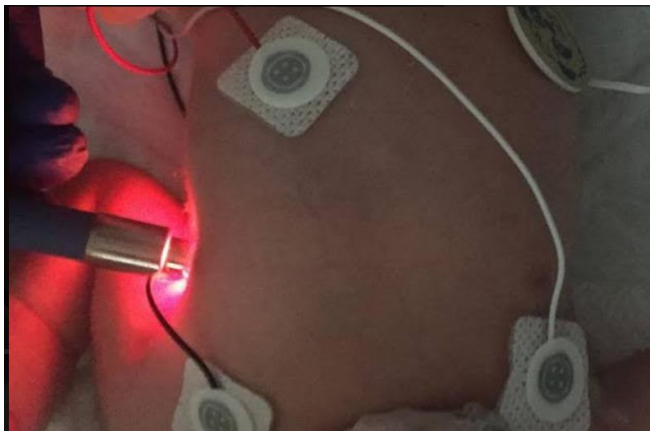
The lesions can be quite erythematous and even be scabbed when there is scratching. In atopic, itchy kids, the rash can be extremely pruritic. Also, during the course of the disease, new lesions appear as old ones regress. I know from personal (my son's) experience!

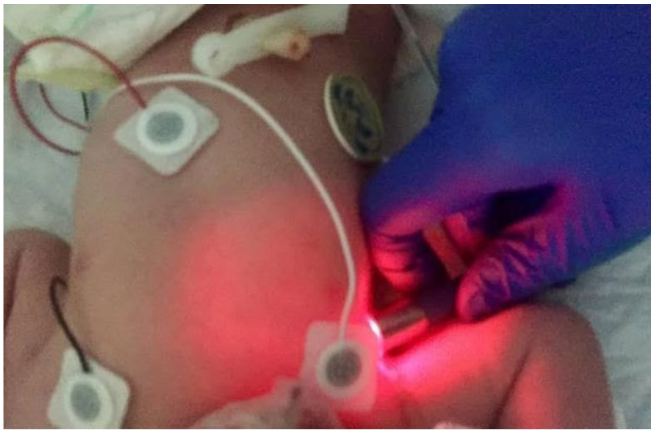
**Onto this week's case of impressive transillumination!**

**HPI:** You are called to evaluate a male infant born at 36+4weeks gestational age via uncomplicated NSVD for respiratory distress at approximately 30 minutes of life. The pregnancy was complicated by IUGR and gestational diabetes, but fetal echo was normal. Baby was born vigorous with spontaneous cry, and Apgar's were 8 at 1 minute of life and 9 at 5 minutes of life. Baby did not require any resuscitation in the delivery room.

On your exam, the baby is tachypneic with grunting and retractions, and O2sats in the mid-80s on blow-by O2. You bring the baby to the NICU and start a rule-out-sepsis work-up and order a CXR. While waiting for the CXR, your fellow whips out a handy fiberoptic light, and you observe the following:

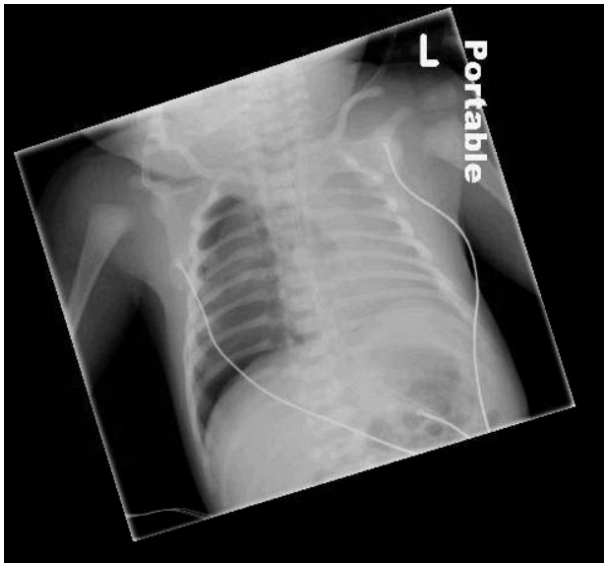
**Picture:**





**Diagnosis: Right-sided Symptomatic Spontaneous Pneumothorax demonstrated on transillumination of the chest! Picture courtesy of Dr. Louisa Keith**

**Discussion:** Air leak syndromes are more common during the newborn period than at any other time of life. Air leak syndromes are the result of rupture of over-distended alveoli, with resulting extra-pulmonary air dissecting into various tissues. Air leak syndromes include pneumothorax (most common), pneumomediastinum, pulmonary interstitial emphysema, pneumopericardium, and, more rarely, pneumoperitoneum and subcutaneous emphysema.




**Epidemiology:**

- Symptomatic spontaneous pneumothorax occurs in 0.05-1% of all term newborns.
    - True incidence is probably higher as cases of spontaneous pneumothorax can be asymptomatic.

- Incidence rises to around 4.1% in infants with birthweight between 500-1500g.
- Risk factors include:
    - Prematurity
    - Underlying lung disease (including RDS)
    - Pulmonary hypoplasia
    - Pneumonia
    - Meconium aspiration syndrome (can complicate 10-30% of MAS cases)
    - Transient tachypnea of the newborn
    - Mechanical ventilation (including delivery room resuscitation)

**Pathogenesis:**

- Rupture of overdistended alveoli.
- Overdistention can be caused by air trapping or uneven distribution of gas.
- Spontaneous pneumothorax may be due to high transpulmonary pressures that are generated with an infant's first breaths.
- Some familial cases and cases with autosomal dominant inheritance have been reported.

**Clinical Manifestations:**

- Patients with symptomatic spontaneous pneumothorax typically present within a few hours of birth.
    - Many cases may be asymptomatic!
- Symptoms of all air leak syndromes include tachypnea, grunting, cyanosis.
- Physical exam can reveal chest asymmetry, decreased breath sounds on the affected side in cases of pneumothorax, and hypoxemia.
- Severe cases can present with hemodynamic compromise.

**Clinical Course:**

- Many asymptomatic and mildly symptomatic cases may resolve without intervention.
- Large lesions can lead to tension pneumothorax and resulting hemodynamic compromise.
- Pneumopericardium may result in cardiac tamponade.
- Pulmonary interstitial emphysema may resolve over time.
- Pneumomediastinum usually resolves spontaneously.

**Diagnosis:**

- Main diagnostic modality is CXR for all air leak syndromes.
    - May need multiple views.
- Transillumination of the chest can also be useful to diagnose pneumothorax (as in above pictures).
    - Side with pneumothorax will light up.
    - Faster than CXR.
    - Pneumopericardium may also transilluminate, may flicker with heart rate.
- Pneumothorax can also be diagnosed with ultrasound.

**Treatment:**

- Asymptomatic lesions or mildly symptomatic lesions may be closely observed without intervention.
- Supplement O2 for low O2 sats.
    - Hyperoxia was used in the past as it was thought to lead to faster rate of resolution, but this has not been shown on recent studies and is now uncommon.

- For mechanically ventilated infants, settings should be optimized to minimize mean airway pressure, PEEP, and peak inspiratory pressure.

- Symptomatic pneumothorax can be treated with thoracentesis or chest tube placement (definitive treatment).

    - Thoracentesis is ideal for non-mechanically ventilated infants, but may only be a temporizing measure if patient is mechanically ventilated.

- Symptomatic pneumomediastinum or pneumopericardium may require percutaneous drainage.

- Pulmonary interstitial emphysema has no definitive treatments, but decreasing ventilator settings to minimize mean airway pressure can help, as can high-frequency ventilation. Other interventions include affected-side-down positioning, minimal chest PT, and minimal ETT suctioning.

- Severe cases of pulmonary interstitial emphysema may require selective collapse of the affected lung.

**Prognosis:**

- Depends on severity and type of lesion, can range from complete resolution without intervention to shock, hemodynamic compromise, and death.

    - Mortality of pneumopericardium can be up to 80%.

**Our Patient:**

Upon arrival to the NICU the baby was placed on CPAP, which was discontinued when CXR confirmed large right-sided pneumothorax. The patient subsequently did well on room air, without respiratory distress or hypoxia, and he did not require thoracentesis or chest tube placement. Serial CXRs were performed, which demonstrated almost complete resolution of the pneumothorax by 6 days of life. Rule-out-sepsis work-up was negative. The patient was discharged home on day of life 7.

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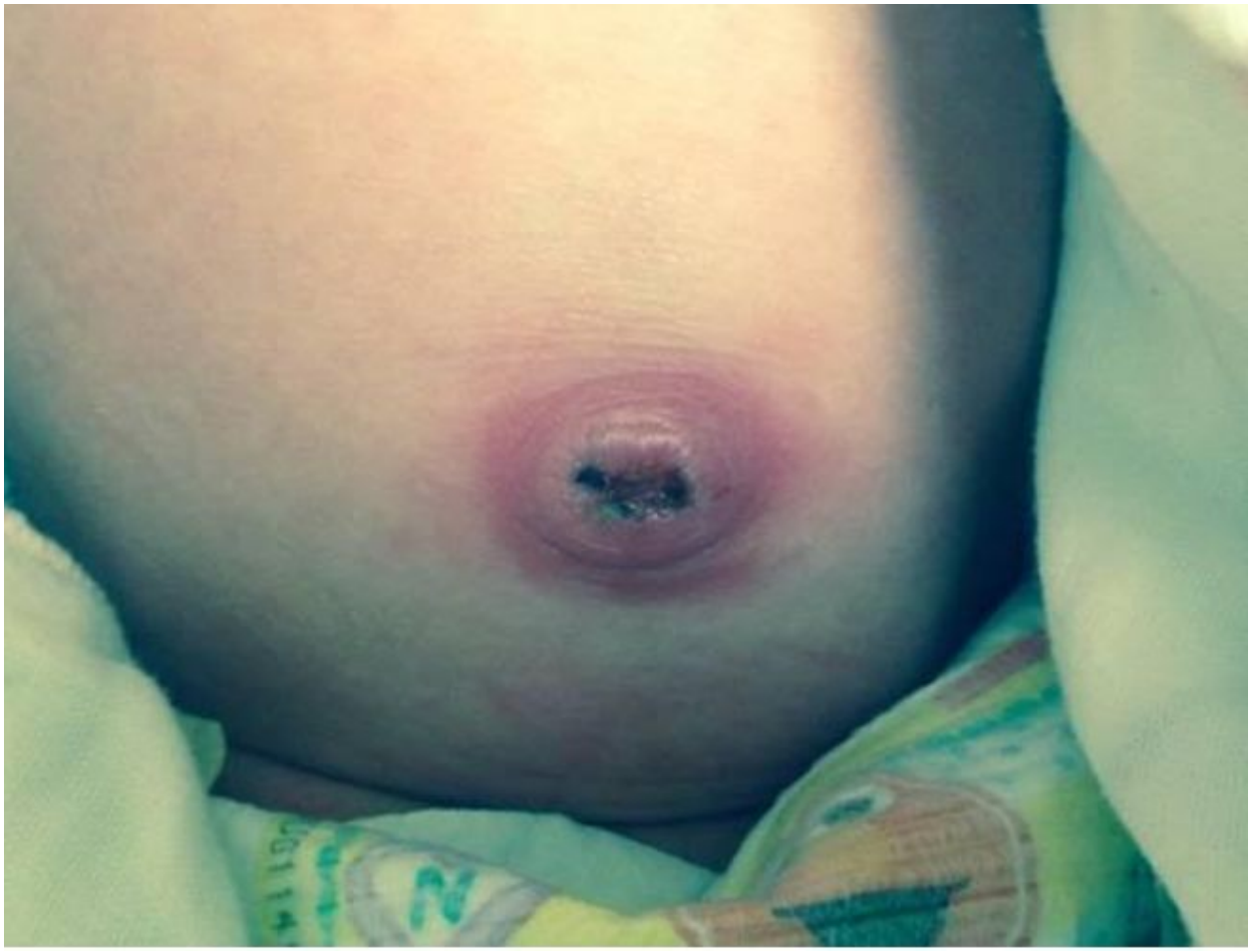
- Cools B, Plaskie K, Van de Vijver K, Suys B. Unsuccessful resuscitation of a preterm infant due to a pneumothorax and a masked tension pneumopericardium. Resuscitation. 2008;78(2):236.

**This week's case is thanks to our wonderful teaching resident, Sarah Milburn!! With a special historical follow up from our former great teaching resident, Tom Hays, and legendary Dr. Irwin Gribetz!**

Presentation:

A 15 day old female, born full term via normal spontaneous vaginal delivery, presented to our ED for abdominal redness. The pregnancy was complicated by a maternal papular rash of unclear etiology, which began at 4 months gestation and improved after delivery with the help of steroids. Otherwise, there was no other significant medical history during the pregnancy, delivery, or newborn period and the umbilical cord separated at 7 days of life. Patient presented to our Peds ED with worsening abdominal erythema and irritability of two days duration. On exam the following was noted:

Picture:



# Diagnosis: Omphalitis!

Definition:

- An infection of the umbilicus and/or surrounding tissues
- Presents with:
    - purulent or foul smelling discharge
    - Periumbilical induration, erythema, and tenderness
    - May have signs of systemic infection

- Typically presents in the first two weeks of life

Epidemiology

- Rare in developed countries --> incidence 0.7%

- In developing countries, incidence as high as 8% in hospitals and 22% for those born at home

- Risk factors:

    - Low birth weight, prolonged labor, prolonged rupture of membranes, maternal infection, non-sterile delivery, umbilical catheterization, home birth

    - Improper cord care such as non-sterile clamping

    - Immune system abnormalities i.e. defects in leukocyte adhesion, neutrophil or natural killer lymphocyte function, interferon production

Etiology

- Immediately after birth, the umbilicus becomes colonized with staph and other gram positive cocci within hours, followed by enteric organisms

- Devitalized tissues of the umbilical stump provide excellent growth medium for bacteria

- Thrombosed blood vessels within the umbilical stump provide an entry for microorganisms into the bloodstream which can result in sepsis

Complications

- most common: sepsis

- less common: septic umbilical arteritis, portal vein thrombosis, liver abscess, peritonitis, intestinal gangrene, small bowel evisceration, necrotizing fasciitis

- Mortality: **between 7-15%**

Bacterial species involved → usually polymicrobial!

- Staph aureus

- Strep pyogenes

- Gram negative bacteria: E. coli, klebsiella, proteus mirabilis

- Anaerobic bacteria: clostridium perfringens, clostridium tetani → especially common in infants born to mothers with chorioamnionitis

Management/Antibiotic choice

- Whenever possible, obtain cultures of the discharge prior to starting antibiotics

- Blood and CSF cultures should also be obtained in any infant with systemic signs (i.e. fever, irritability, etc) as they are more likely to be septic or develop meningitis

- Systemic antibiotic treatment is a requirement

- Multiple antibiotics needed as polymicrobial infection

    - Staph coverage -> Vancomycin

    - Anaerobic -> clindamycin or flagyl

    - Typically 10-14 day course of IV antibiotics

Of Note:

- Mild discharge from the umbilical stump in absence of inflammatory signs can be a normal occurrence, even when there is some odor

- No evidence that topical alcohol, bacitracin, or mupiricin is of any benefit in preventing omphalitis

Back to our patient:

Although our patient was afebrile initially in the ED, as she had evidence of infection with systemic signs (i.e. irritability) she underwent a full rule out sepsis with blood, urine, and CSF studies. A wound culture was also obtained from expelled discharge. Our ID team was consulted who recommended empiric treatment with vancomycin (especially with the strange history of maternal rash concerning for MRSA), cefotaxime, and flagyl. A bedside ultrasound was performed which did not show any abscess formation. The patient was admitted to the floor, and shortly thereafter developed a fever to 38.2. The wound culture grew citrobacter freundii (an anaerobic gram negative rod) as well as MSSA, all other cultures were negative. Due to slightly expanding erythema several days after admission, she underwent an operative I&D by Peds Surgery. Following I&D, the patient improved and was able to be discharged home 2 days later

on PO Bactrim and Flagyl.

References:

Bugaje, Mairo Adamu; et al. "Omphalitis". *Paediatric Surgery: A Comprehensive Text For Africa*. Retrieved 23 July 2013.

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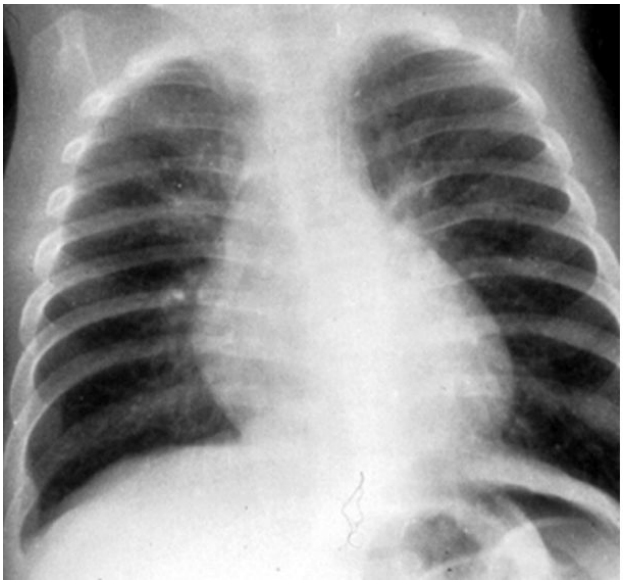
Brook I. Cutaneous and subcutaneous infections in newborns due to anaerobic bacteria. J Perinat Med 2002; 30:197.

Cases from Adam Weinstein, MD, Darthmouth-Hitchcock

## A case of RSV Bronchiolitis

A 6 month old boy presents to his pediatrician with 2-3 days of runny nose and wet cough, and last night was coughing, wheezing, and having trouble breathing (parents describe he was breathing really fast and hard and couldn't get comfortable). He is cranky and not eating as well but is still making urine normally. No vomiting or diarrhea.

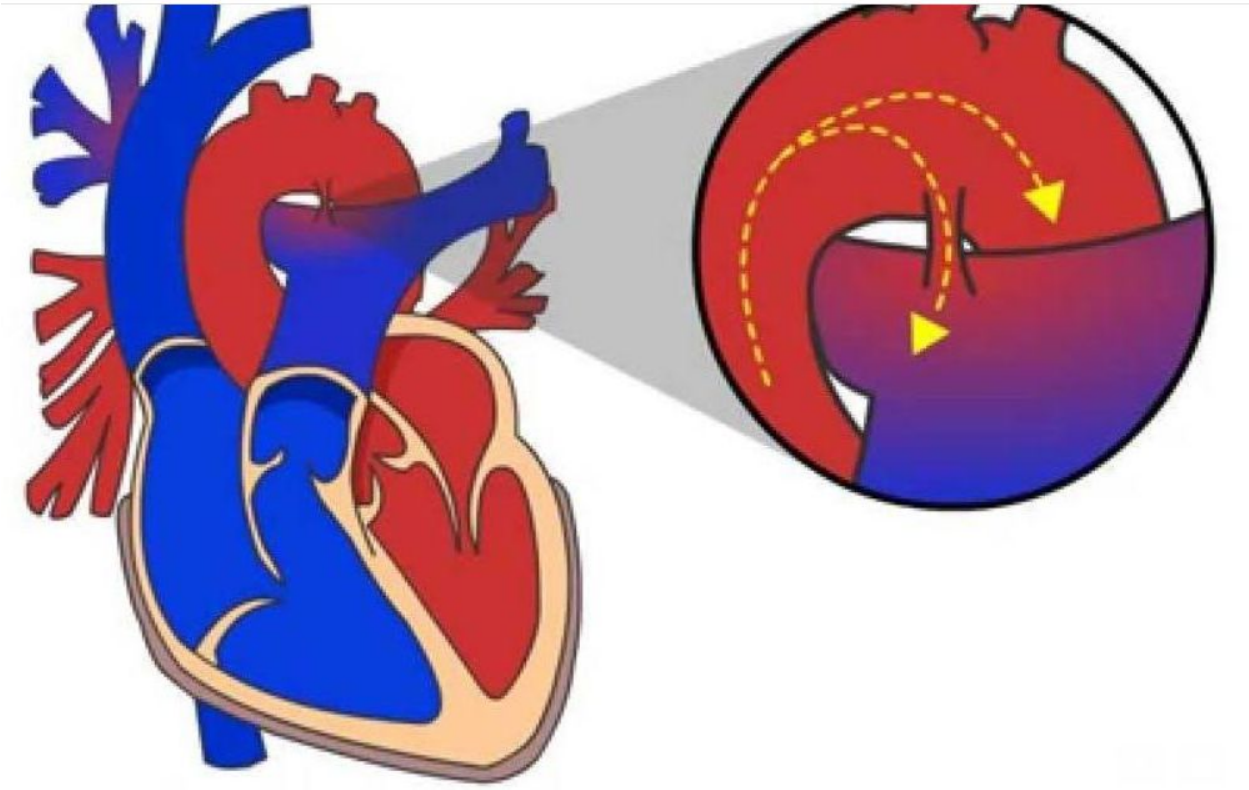
On exam Temperature is 100 degrees F, Heart rate is 150, BP is 90/50, and RR is 50, Oxygen saturation is 90% on Room Air. He looks like he is breathing hard, and you note intercostal retractions and coarse breath sounds on your exam. He had a chest x-ray done and it appears as follows:



Please list the pathophysiologic mechanisms by which RSV causes "Bronchiolitis" and accordingly, for each mechanism list treatment(s).

## Case 2

What makes premature neonates at risk for this finding below?



## Case 3

# 16 month old with seizure

An otherwise healthy 16 month old girl presents to her local emergency room.

She was at daycare and at around 10:30am, her caregivers saw her eyes roll, and extremities stiffen and jerk repeatedly for about 60 seconds. EMS was called and took her to the ER along with her parents.

She arrives to the ER around 11am and her parents note she has had some mild upper resp symptoms but has otherwise been acting herself. You see her and she is sleeping, well-perfused. You note her temperature of 102.6 F

and Heart Rate of 140 bpm.  As you are taking her history, she has another episode consistent with a generalized seizure.

Come up with a list of 1) steps to make sure she's ok in the immediate moment; 2) history, exam, diagnostic tests to evaluate the cause; and 3) factors that will effect what you might share as the long-term prognosis.

Case 4

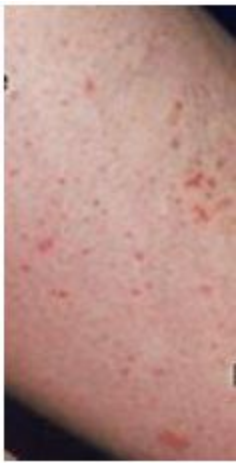
# Covered in Spots!

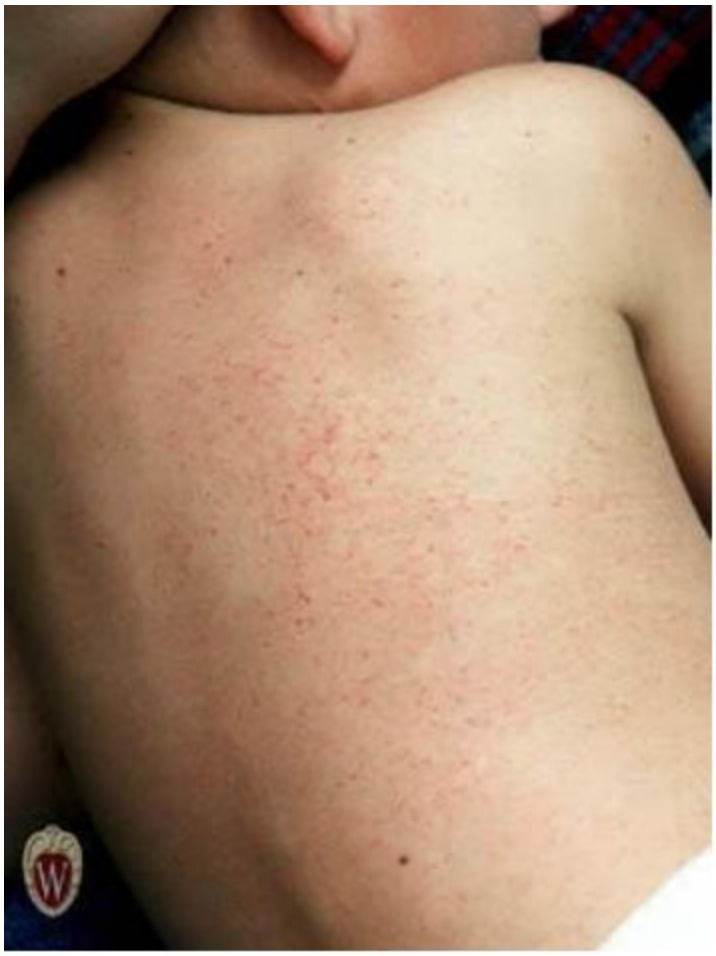
A mother brings her 3 year old son in because he's covered in red spots.  They do not blanch on palpation.

Name at least three diagnoses that can present with red spots that do not blanch *in a 3 year old* --be specific (e.g. don't say "infection"--name the infection)

For each diagnosis, discuss how you would clinically (by history, exam, or diagnostic testing) distinguish it from the other diagnoses.

Lastly-- if the child is completely and otherwise well, acting totally normal, no other symptoms or findings on exam, what do you think the most likely cause would be?





Case 5

## Can't stop moving!

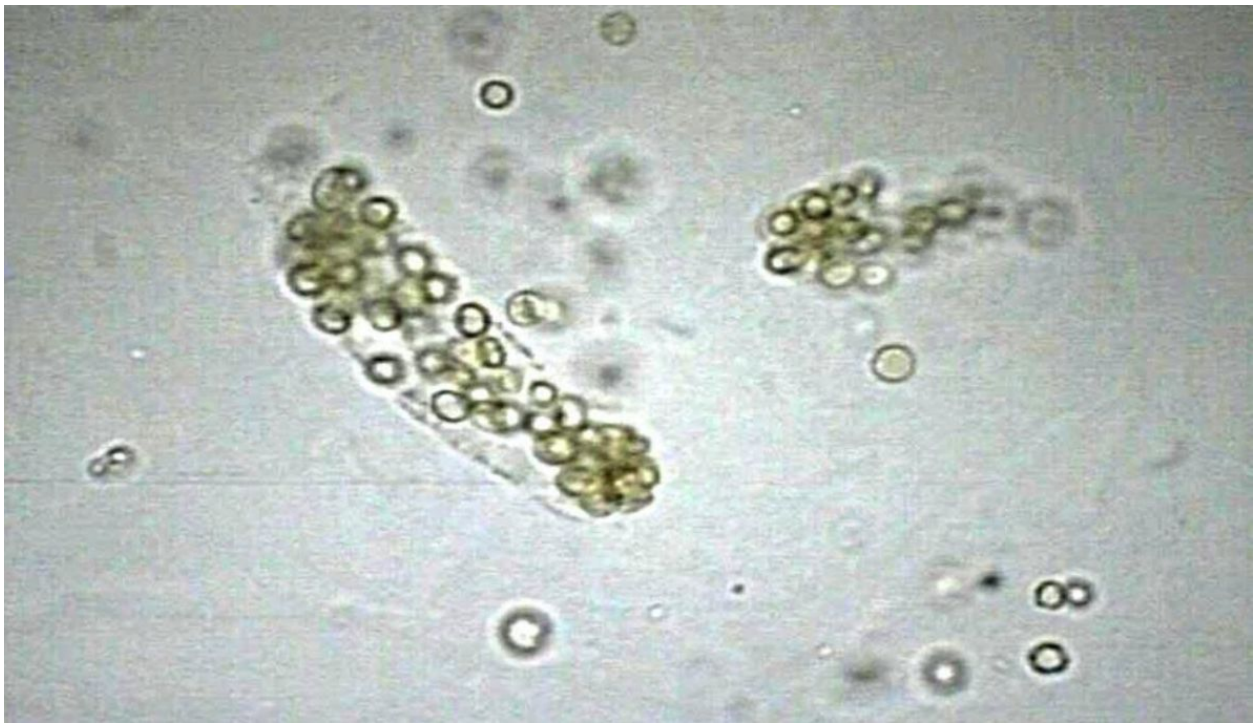
ADHD is one of the more common diagnoses (and "chronic conditions") you'll see in pediatric care settings. Stimulants are the mainstay of pharmacologic therapy.  Name some *Non-pharmacological" means to treat ADHD. (that is, other than playing Pokemon Go).

Case 6

## Eyes swollen shut!

The parents of a 3 year old bring him to the Emergency Dept because he's been getting progressively more swollen and he woke this morning with his eyes swollen shut.  He has no other symptoms.He looks well, and comfortable, just swollen.  His vitals are normal including a normal blood pressure but his weight is increased compared to

baseline. The astute medical student on her/his ED rotation grabs a urine sample and it has lots of protein in it, and under the microscope it looks like this:



Based on the presentation and factoring in the urinalysis and microscopy, list the three most likely diagnoses. List some lab tests you'd like to get. And what treatment, if any, you'd start (even before the lab results come back).