Pediatric Puzzles
What is this Rash?

Renee Howard MD
Director of Dermatology
UCSF Benioff Children’s Hospital Oakland
Professor of Dermatology, UCSF
Renee.howard@ucsf.edu
Disclosures

I declare that neither I, nor any immediate member of my family, have a financial arrangement or affiliation with any corporate organization offering financial support or grant monies for this continuing medical education activity. In addition, I do not intend to include information or discuss investigational or off-label use of pharmaceutical products or medical devices.
What we will do today

• Present cases of mystery skin rashes and lesions
  – Special sites
  – Imitators of common skin rashes
  – Skin nodules and red papules through the age spectrum
Case 1: Cause?

- 8 year old ballerina
- Sudden onset itchy rash
Case 1: Cause?

1. Mango
2. Makeup
3. Poison oak
4. Lime juice
5. Fragrance
Case 1: Cause?

1. Mango
2. Makeup
3. Poison oak
4. Lime juice
5. Fragrance (# 2)
Challenge: facial dermatitis

- Skin is thin and sensitive
- Exposed to a lot
- Topical steroid can cause Periorifical dermatitis
Perioral dermatitis
DDx Facial dermatitis

- Perioral dermatitis
- Irritant contact dermatitis
  - Drool & pacifier
  - Lip licking
- Atopic dermatitis
- Allergic contact dermatitis
  - Mango, poison oak
Mango peel allergy

Derm Resident answer: off the top of my head: impetigo, herpes, phytophoto, sensitivity to a sunscreen or other lip balm, or sun sensi from isotretinoin/doxycycline?
Rhus (poison oak) allergy

Mom’s photos from the ED
Challenge: facial dermatitis

- Put patient on “skin care diet”
  - Water cleansing
  - Petrolatum
- Taper topical steroids
- Be ready to use topical antibiotics
Case 2: Diaper rash that will not budge
Case 2: First line treatment

a) Terbinafine
b) Nystatin
c) Hydrocortisone 2.5%
d) Tolnaftate
e) Desonide
Case 2: First line treatment

a) Terbinafine
b) Nystatin
c) Hydrocortisone 2.5%
d) Tolnaftate
e) Desonide
Candida Diaper Dermatitis

Beefy red confluent in folds

KOH here
Psoriasis in the Diaper Area

Confluent, pink, in folds; asymptomatic

Doesn’t budge with barriers, 1% hydrocortisone or antifungal
GIFT for diagnosis
Glean Information From The

- Scalp - scaling
- Nails – pits, onycholysis
- Intertriginous-erythema
  - Intergluteal crease
  - Umbilicus
  - Axillae
Goals for Rx of Psoriasis

• Control of symptoms not clearing
• Look for and treat triggers
• Watch for comorbidities
Approach to stubborn diaper dermatitis

- Cover Candida, irritant
- Culture
- Consider rare causes
Infected irritant dermatitis

High water mark

Staph aureus +
Irritant dermatitis
Fig. 1. Scaly pink papules coalescing into plaques that correspond with the green dye in the diapers. These findings are suggestive of allergic contact dermatitis from the diaper dye.

Christopher Klunk, Erik Domingues, Karen Wiss

An update on diaper dermatitis


http://dx.doi.org/10.1016/j.clindermatol.2014.02.003
Allergens in infants?

- Fragrance
- Preservatives
- Topical antibiotics
- Textile dyes
Approach to stubborn diaper dermatitis

- Culture
- Brown diapers, water wipes
- Barrier q diaper change
- 2.5% HC or desonide ointment
- Nystatin ointment or imidazole cream
Case 3: Diagnosis?

1. Lymphoma
2. Lipoma
3. Follicular cyst
4. Pilomatrixoma
5. Branchial cleft cyst
Case 3: Diagnosis?

1. Lymphoma
2. Lipoma
3. Follicular cyst
4. Pilomatrixoma
5. Branchial cleft cyst
Case 3: Nodule on the Neck

Wondering if you need to see her...9yo has this for a month, no discomfort/itch. Cystic, bluish, superficial, mobile, less than 1 cm. Not a lymph node or along chain. Don't think it's a congenital structure.
Pilomatrixoma

- Benign neoplasm
- Derives from hair matrix
- Can self resolve
- Can rupture, get inflamed
Multiple Pilomatrixomas
Multiple Pilomatrixomas

• Familial
  – Isolated
  – Myotonic dystrophy
  – Familial adenomatous polyposis
Nodules in Teens, Adults

- Follicular cyst
- Lipoma
Nodules in infants

- Dermoid cyst
- Deep hemangioma
Case 4: Bleeding red papule

- Present for one month
- Profuse bleeding
- ED visit
Case 4: Diagnosis?

1. Spitz nevus
2. Pyogenic granuloma
3. Hemangioma
4. Melanoma
Case 4: Diagnosis?

1. Spitz nevus
2. Pyogenic granuloma
3. Hemangioma
4. Melanoma
Case 5: Bleeding red papule

- 3 month old
- Present for 1 month
- Profuse bleeding
- ED visit
- “Bandaid sign”
Case 5: Best next step?

1. Timolol
2. Emergency room
3. Imiquimod
4. Plastic surgery
5. Dermatology
Case 5: Best next step?

1. Timolol
2. Emergency room
3. Imiquimod
4. Plastic surgery
5. Dermatology
Why Dermatology?

Electrodesiccation and Curettage

- Only indicated for low-risk lesions

- PROS: minimal blood loss, ease, convenience for the patient

- CONS: no specimen for pathology, clinician experience influences cure rate

Images courtesy of Margaret Mann, M.D.
New onset red papules

ALL SEEN IN UBCHO PEDIATRIC DERMATOLOGY CLINIC IN ONE DAY!

Pyogenic granuloma

Infantile Hemangioma

Spitz nevus
Dermoscopy
Dermoscopy of Spitz

Dotted blood vessels

Residual pigment

Actas Dermosifiliogr 2012;103:357-75 - Vol. 103 Num.5 DOI: 10.1016
**CLINICAL DATA:**  RED BLEEDING PAPULE X 1 MONTH

**DIAGNOSIS:**  PYOGENIC GRANULOMA (LOBULAR CAPILLARY HEMANGIOMA)  
(LEFT CHEEK)  
(L98.0)

**SPECIMEN SITE:**  LEFT CHEEK

**GROSS DESCRIPTION:**
Skin segment measuring 0.3 x 0.2 x 0.1 cm. Submitted intact. PL/PL

**MICROSCOPIC DESCRIPTION:**
There is a protuberant lesion formed by a lobular proliferation of capillaries and venules within an expanded papillary dermis.
Approach to red papules

• Consider DDx as not always vascular

• Tissue should be sent for pathology when removed

• Follow size and shape with serial photography if not removed
Case 6: Facial Pustules
Diagnosis?

1. Pyoderma faciale
2. Acne conglobata
3. Gram negative infection
4. Pityrosporum
5. Staphyloococcal infection
Diagnosis?

1. Pyoderma faciale
2. Acne conglobata
3. Gram negative infection
4. Pityrosporum
5. Staphylococcal infection
Acne conglobata
Acne DDx evolves
DDx Acne in Teens

- **Folliculitis**
  - Staphylococcal
  - Gram negative
  - Pityrosporum

- **Rosacea**
  - Central face
  - Background of erythema
Case 7: Suddenly scaly scalp
Case 7: Suddenly scaly scalp

Most likely etiologic organism?

a) Microsporum canis
b) Malassezia furfur
c) Trichophyton mentagrophytes
d) Trichophyton tonsurans
e) Microsporum gypseum
Case 7: Suddenly scaly scalp

Most likely etiologic organism?

a) Microsporum canis
b) Malassezia furfur
c) Trichophyton mentagrophytes
d) Trichophyton tonsurans
e) Microsporum gypseum
Case 6: Suddenly scaly scalp

Dad’s neck
Case 7: Suddenly scaly scalp

- 2 month old developed crusty rash on face diagnosed as hand foot and mouth last month
- Hair started falling out
- Sister with tinea capitis, on griseofulvin, dad, brother tinea corporis
Tinea capitis

**DDx Noninflammatory**
- Seborrheic, atopic dermatitis, psoriasis

**DDx Inflammatory**
- Folliculitis decalvans, staphylococcal abscess
Tinea facei

**DDx Noninflammatory**
- Atopic, seborrheic dermatitis

**DDx Inflammatory**
- Cutaneous lupus, psoriasis
KOH positive for spores around hair
Dermoscopy as Diagnostic Tool for Tinea

Clinical Trial

Trichoscopy as a useful method to differentiate tinea capitis from alopecia areata in children at Zagazig University Hospitals

Mohamed Amer MD, Afaf Helmy MD, Amin Amer MD

First published: 22 September 2016
DOI: 10.1111/ijd.13217

Dermoscopy
not just for nevi
Dermatophyte infections in infants

- Rare-case reports
- Tinea faciale, capitis
- Oral griseofulvin 20mg/kg
- No safety data or monitoring guidelines


Hairdresser’s occupational hazard?

- Consider occupational as well as leisure activity exposures when considering source of infection
  - Petting zoo, pet rat in classrooms, animal adoption fairs
- Neonatal dermatophyte infection very rare but can occur with family exposure
- May treat systemically if necessary
What we did today

• Present cases of mystery skin rashes and lesions
  – Special sites
  – Imitators of common skin rashes
  – Skin nodules and red papules through the age spectrum
Acknowledgements

- UCSF Benioff Children’s Hospital Oakland Dermatology
  - Renee Howard MD, Director
  - Anjali Washington, PA-C
  - Nelly Gutierrez, Office Assistant

- UCSF Pediatric Dermatology
  - Kelly Cordoro, Ilona Frieden, Erin Mathes, Anu Mathur, Sonal Shah
Neurology
Unknowns- Is this a seizure?

Audrey Foster-Barber, MD, PhD
Disclosure

- I declare that neither I, nor any immediate member of my family, have a financial arrangement or affiliation with any corporate organization offering financial support or grant monies for this continuing medical education activity. In addition, I do not intend to include information or discuss investigational or off-label use of pharmaceutical products or medical devices.
What is a seizure?

• Paroxysmal event- cognitive, motor, autonomic
• Distinct on and off, evolves
• Associated with sudden surge of synchronized electrical activity in the brain
• Generalized, Focal/Localized
Generalized Seizures

- Absence
- Tonic clonic
- Myoclonic
- Tonic
- Clonic
- Atonic
Focal or Localized

- Focal Motor
- Autonomic
- With Dyscognitive features
- With secondary generalization
Other Unique Pediatric Seizures

- Epileptic spasms
- Unique Neonatal seizures
  - apnea
  - repetitive mouth movement
  - pedaling
  - tonic
  - clonic
  - myoclonic
- unable to mount full generalized tonic-clonic
Video 1
Question 1

Most seizures in children are provoked by?
1) Flashing lights
2) Fever
3) Stress
4) None of the above
Increase risk of seizures

- Sleep-wake transitions
- Sleep deprivation
- Fever, illness
- Flashing lights
- Alcohol intoxication
- Drug use (THC in marijuana)

- Hormonal changes, pubertal and perimenstrual
- Low blood sugar
- Medications- robinul, demerol, ocp, antibiotics
Most seizures are unpredictable, unprovoked

Reflex Epilepsies

- Very rare, triggered epilepsies <5% of all Epilepsy
- Also unprovoked seizures in these patients
- Mostly visual stimuli (flash)
- Also sensory, auditory, somatosensory, olfactory
- Very rare-Reading Epilepsy, Music Epilepsy, Praxis
- Mostly genetic in origin, generalized epilepsy syndromes
Video - Is this a seizure?
Question 2

Which best distinguishes between a true Epileptic seizure and a Nonepileptic seizure?

1) Nonrhythmic movements involving trunk
2) Presence or absence of Epilepsy history
3) Presence of anxiety or mood disorder
NonEpileptic Seizure

• Predictable trigger or location for event
• More prominent proximal and truncal movements, Pelvic thrust
• Horizontal head movement
• Bilateral motor activity with preserved consciousness
• Eye closure with resistance to opening
• Variable rate, nonrhythmic
• Stuttering or stop and start course
• Long event with no postictal period
• 8:1 female to male
• Often personal or family history of epilepsy
• Stress, anxiety triggers more common than abuse (though mental health comorbidities common in epilepsy)
• Prolactin elevation, 2x increase over baseline- thought to be a surrogate marker for seizure
  - specificity 96%
  - sensitivity for GTC 60%, focal dyscognitive seizure 46%
  - similar elevations in syncope
  - data suggests no elevation in NES
Video-Induced Seizure?
Question 3

Which feature best distinguishes between syncope and a convulsion?

1) Mouth laceration
2) Presence or absence of abnormal movements
3) Urinary incontinence
4) Autonomic symptoms preceding the event
5) Post-ictal period
Fits vs Faint

- Common to have short, nonrhythmic movements after syncope
- Postictal fatigue or confusion is present in some patients after syncope (10% or less)
- Syncope - more reliable trigger (vasovagal, cardiogenic), prodrome of dizzy, sweaty, heart palpitation, vision darkening
- Tongue lacerations can occur in either syncope (TIP) or seizure (SIDE)
- Urinary incontinence can be present in either (unreliable)
Video- Is this a seizure?
Breath-holding spells

- 5% of the pediatric population
- Cry from pain or upset, exhale and breath-hold and lose consciousness
- Often decerebrate posture with extensor stiffening
- Sometimes irregular flexor spasms
- Rare true brief anoxic seizure as a result
- Recovery immediately most often, sometimes sleeps after
- More common with iron deficiency anemia
Video- Is this a seizure?
Infantile self gratification

- Masturbation
- Seem distracted or uncomfortable
- More girls than boys
- Often when bored
- Often when strapped into seat
- More in young children and kids with developmental delay
Video- Is this a seizure?
Paroxysmal tonic upgaze of infancy

- Movement disorder
- Upgaze attacks - short or long, often with additional movements as a reaction to this
- Brief, < 1 min, often with or after an illness
- EEG negative, MRI negative
- Often associated with mild Learning Disability, mild ataxia or incoordination
Video
Night Terrors

• A parasomnia
• Sudden arousal from non-REM sleep, usually in first 1/3 of the night
• Autonomic and behavioral- sweating, increased HR, fear, crying, screaming, thrashing
• No response to stimuli or minimal
• Upon awakening confusion or amnesia to the event
• Not stereotyped, not repeated in the night
Mimics of Seizures

- Behavioral- NES, daydreaming, self gratification, breath holding spells
- Cardiac- syncope, v tach
- GI- Sandifer syndrome
- Movement Disorder- tic, dystonia, paroxysmal dykinesia, PTUI, hyperekplexia, episodic ataxia
- Migraine Syndromes- hemiplegic migraine, basilar migraine, paroxysmal vertigo or torticollis
- Sleep related- benign sleep myoclonus, narcolepsy, night terrors, PLMS or RLS
Video- Is this a seizure?
Seizures commonly missed

- Absence seizure
- Gelastic seizure
- Nocturnal Frontal Lobe seizure
- Infantile spasms
How to identify a seizure

• If dramatic- distinguish between convulsion, NES, faint (trigger, duration, postictal period, injury)
• If more subtle- some pattern recognition for common mimics
• Often the clinical context and frequency are most important
• Sometimes you just need an EEG
  -Ideal if you capture a seizure- inpatient admission or portable EEG
  -Otherwise catch epileptiform discharges (sleep-wake transitions)
Thank you!
Audrey Foster-Barber, audrey.foster-barber@ucsf.edu
Pediatric Brain Center 1-855-PBC-UCSF
MISSION
Caring
Healing
Teaching
Discovering

VISION
To be the best provider of health care, the best place to work, and the best environment for teaching and research.

VALUES
Professionalism
Respect
Integrity
Diversity
Excellence
Information for Physicians

CHILDREN’S HOSPITAL ACCESS CENTER

A single point of access available 24/7 for neonatal, pediatric and maternal transfers, transports and admissions, as well as outpatient referrals and telephone consultations with any of our pediatric specialists.

Phone: (877) UC-CHILD / (877) 822-4453
For more information, visit www.ucsfbenioffchildrens.org/accesscenter.

PHYSICIAN LIAISON SERVICE

Provides assistance and information to referring physicians, medical groups and health plans.

Phone: (800) 444-2559
Fax: (415) 353-4395
Infectious Disease Case Studies

Meg Fisher, MD
Medical Director

The Unterberg Children’s Hospital
at Monmouth Medical Center
Barnabas Health
Disclosures

I have no disclosures

I may be mentioning off label uses of drugs
Objectives

• Manage a febrile child
• Care for a child with bronchiolitis
• Assume care for an immigrant teen
• Evaluate a newborn with microcephaly
Change in Practice

• Become familiar with practice guidelines
• Use guidelines to manage common infections
Case 1 - February

• 20 month old with fever
• URI ten days ago
• Improved and returned to child care
• Fever onset 3 days ago
• Less playful today

Advice?
My Thoughts

- Timing of fever suggests complication of URI
- In child care so could be virus #2
- Still febrile so I am considering UTI
- Might opt to see patient
Next day

- Less playful; comes to office
- PE: lethargic, febrile
- No source for fever identified
- Listless but cries when examined

Now what?
My Thoughts

- Listless and lethargic make me twitch
- Meningitis
- Urinary tract infection
- Influenza still possible
Petechiae identified

Photograph from CDC website

Differential diagnosis?

Now what?
My Thoughts

- Very likely needs hospitalization
- How will I get the patient to the hospital: car or ambulance or walk over
- Which laboratory studies do I want: spinal fluid, blood, urine
Laboratory Results

- Hgb 9.9 gm/dl, wbc 12,200, 68% neut, 27% lymphs, 488,000 platelets
- CSF: 1270 wbc, 90% neutrophils, glucose 37 mg/dl (blood 100), protein 66
- Electrolytes normal

Most likely pathogen?
My Thoughts

- Gram stain can be your friend
- Gram negative diplococci = think meningococcus
- Gram positive diplococci = think pneumococcus, ? vaccine failure
- Gram negative pleomorphic rods = think *Haemophilus*, vaccine failure
Admitted

- Empiric antibiotics?
- Dexamethasone?
- Recommendations for contacts?
My Thoughts

- Antibiotics – ceftriaxone, vancomycin
- Dexamethasone – for me yes and before antibiotics; others will say no
- Contacts – for meningococcosis yes: rifampin 2 doses/day for 2 days; or ceftriaxone, ciprofloxacin or azithromycin
- Child care center will be a nightmare

2015 Red Book. Meningococcal Infections
More dilemmas

• Mother goes into labor. What do you recommend for the newborn?

• Older sibling just left for college – what about her?
My Thoughts

- There are no specific recommendations for the baby BUT…
- Mother needs prophylaxis
- College age sibling was immunized but still needs prophylaxis
Case 2 - January

• A 10 month old boy presents with cough and rhinorrhea for 2 days
• His older sister has a “cold”
• Past medical history is unremarkable
• Family history negative for asthma
Case 2 - PE

- T 100.8, HR 105, RR 55, BP 80/45
- Weight 70\textsuperscript{th} %, length 60\textsuperscript{th} %, HC 50\textsuperscript{th} %
- Cloudy nasal discharge, no flaring
- Mild retractions
- Scattered wheezes, prolonged expiration
Most likely diagnosis?

a) Sinusitis
b) Bronchiolitis
c) Heart failure
d) Bacterial pneumonia
e) Mycoplasma pneumonia
My Thoughts

• Sinusitis – no, only 2 day illness
• Bronchiolitis – of course
• Heart failure – not likely
• Bacterial pneumonia – not by exam
• Mycoplasma pneumonia – wrong age
What tests are needed?

a) Chest radiograph
b) Pulse oximetry
c) Complete blood count
d) Viral panel
e) None
My Thoughts

• Chest radiograph - no
• Pulse oximetry - no
• Complete blood count - no
• Viral panel - no
• None – yes, Just say No!

What treatment is needed?

a) Trial of nebulized albuterol
b) Amoxicillin
c) Prednisone, oral or IM
d) Nebulized hypertonic saline
e) None of the above
My Thoughts

• Trial of nebulized albuterol - no
• Amoxicillin - no
• Prednisone, oral or IM - no
• Hypertonic saline – not in your office
• None – Yes, Just say NO

What preventive measures

a) Palivizumab as AAP recommended
b) Hand hygiene
c) Counsel about avoiding smoke
d) Encourage breastfeeding
e) All of the above
My Thoughts

• Palivizumab – yes, as recommended
• Hand hygiene – always
• Counsel about avoiding smoke - sure
• Encourage breastfeeding – of course
• All of the above – Yes, Just do It

Reference

Case 3: Julius

Julius, a 13 year old Guatemalan who came to the United States in the summer of 2014 with other teens from his home, is now living with an aunt in your town. You are seeing him for the first time.

What else do you want to know?
Case 3: Julius

- He has no complaints. The family says he had a negative skin test at a holding center in Texas. He was in the center for 3 weeks; then with other relatives. He got to your town 3 weeks ago.
- He received a Tdap at the center.
- His weight and height are at the 25th percentile. Exam is normal.

Now what?
Case 3: Julius

- He needs screening tests: Serology for HBV, HCV, syphilis, HIV 1 and 2, Chagas disease, CBC, 3 stool samples for ova and parasites, 1 stool for *Giardia intestinalis* and *Cryptosporidium* antigen
- TST or IGRA
- What about immunizations?
Case 3: Julius

- It is highly unlikely you will find any immunization records. He got Tdap so tetanus and diphtheria serology should be positive and won’t help. You will get hepatitis serology. You could get measles, mumps, rubella and varicella serology.

Which vaccines will you give today?
Vaccines for Julius

- HBV: await serology
- Rotavirus: not needed
- Td: a dose now, another in 6 months
- HIB: not needed as he is 13
- PCV13: not recommended
- Hepatitis A: a dose now, repeat in 6 months
Vaccines for Julius

- IPV: now, 1 month, 7 months
- Influenza: now
- MMRV: now, 1 month
- Human papillomavirus: now, 1 mo, 6 mo
- Meningococcal vaccine: now
Screening Results on Blood

- HBsAg +, anti-HBsAg -, anti-HBcAg +
- HCV negative
- RPR and FTA non-reactive; HIV 1 and 2 neg
- *Trypanosoma cruzi* antibody negative
- Hgb 13.9, wbc 12, 45% N, 40% L, 15% E, platelets 380

Explain the results
Screening Results on Blood

- HBV: acute or chronic hepatitis B infection
- HCV: negative
- Syphilis: negative
- HIV: negative
- Chagas disease: negative
- CBC: eosinophilia
Screening Results on Stool

- Ova and parasites: *Ascaris lumbricoides*, *Entamoeba coli*
- Stool Antigen: negative for *Giardia*; negative for *Cryptosporidium*

Now what?
Screening Results on Stool

- *Entamoeba coli* – not a pathogen
- *Ascaris lumbricoides* – a round worm which is likely the reason for his eosinophilia. He should be treated with albendazole or mebendazole
Skin Test Result

- Julius returns for the skin test reading at 72 hours. Your nurse measures 14 mm induration; you measure 11 mm.

Now what
# Positive Tuberculin Skin Test

<table>
<thead>
<tr>
<th>Reaction</th>
<th>Patient characteristics</th>
</tr>
</thead>
<tbody>
<tr>
<td>5mm or more</td>
<td>Close contact of contagious case; Suspected infection; Immunosuppressed</td>
</tr>
<tr>
<td>10 mm or more</td>
<td>Age &lt; 4 yr; Chronic illness; Increased likelihood of exposure (birth, travel, adult contacts at risk)</td>
</tr>
<tr>
<td>15 mm or more</td>
<td>Low risk people over age 4 yr</td>
</tr>
</tbody>
</table>

So, Julius’ test is positive – What next?
Boosted or Seroconversion?

- Boosting: increase in reaction to TST in patients with prior BCG or past infection
- Interferon gamma release assay (IGRA): T-cells of the patient exposed to antigens of *Mycobacterium tuberculosis*
- IGRA more specific than TST in patient who has received BCG
Case 3: Julius

• You decide to send an IGRA and get a chest radiograph. The IGRA is negative and the chest radiograph is normal. You breathe a sigh of relief.

• You have already referred him to a gastroenterologist or hepatologist for HBV
References

• www.cdc.gov/immigrantrefugeehealth/index.html
• www.cdc.gov/immigrantrefugeehealth/guidelines/domestic/domestic-guidelines.html
• www.cdc.gov/vaccines
Case 4 - November

- You are called to the nursery to examine a newborn with microcephaly.
- Mother recently moved to your area; she received no prenatal care.
What do you ask

a) Where did you live before
b) Where have you traveled
c) Where has your sexual partner lived or traveled
d) All of the above
Zika in the US - Florida

As of 10/20/16
Microcephaly

- Incidence in US: 2-12/10,000 births
- Infectious causes: Zika virus, rubella, toxoplasmosis, cytomegalovirus
- Other causes: severe malnutrition, alcohol, drugs, toxins, interruption of blood supply to the fetal brain
Evaluation of the Baby

a) My hospital is a “Zika center”
b) I am aware of current CDC recs
c) I know where to find current recs
d) I would transfer the baby stat
e) I have no idea what I would do
Current Recommendations

Newborn, Mom + Zika

• Comprehensive physical examination
• Neurologic assessment
• Postnatal head ultrasound
• Standard hearing screen
• Zika virus testing of infant

Newborn, confirmed Zika

• Ophthalmologic exam within a month
• Hearing by auditory brainstem response before a month
Follow up

• Depends on findings
• Abnormalities consistent with Zika: microcephaly, intracranial calcifications, other brain or eye abnormalities
• US Zika Pregnancy Registry
Zika Abnormalities

- Consultations: neurologist, infectious diseases, ophthalmologist, endocrinologist, geneticist
- MCF opinion: palliative care essential
- Lab: blood counts, metabolic panel, liver enzymes and function
Zika Abnormalities

• Feeding difficulties: refer for lactation, OT, speech therapy, nutrition, GI as needed for poor suck, swallowing, reflux and aspiration
Zika +, Long Term

- Multidisciplinary team, medical home
- Neurology: sleep, irritability
- Eye and hearing repeat exams: 3, 4-6 mo
- Hypothalamic dysfunction: thyroid screen (TSH, free T3, T4) at 2 wk and 3 mo
- Development screen, early intervention
Mom Zika, Baby Negative

• Routine care
• Monitor head circumference, wgt, lgth
• Developmental screening
• Vision screening
• Hearing at 9 months or sooner
Change in Practice

- Become familiar with practice guidelines
- Use guidelines to manage common infections
Thank you
Pediatric Knee Pain Unknowns…

Coleen S. Sabatini, MD, MPH
Medical Director and Chief, Orthopaedic Surgery
UCSF Benioff Children’s Hospital Oakland
“I declare that neither I, nor any immediate member of my family, have a financial arrangement or affiliation with any corporate organization offering financial support or grant monies for this continuing medical education activity. In addition, I do not intend to include information or discuss investigational or off label-use of pharmaceutical products or medical devices”
Pediatric Knee Pain

- Patellofemoral Syndrome
- Osgood-Schlatter’s
- Sinding-Larsen-Johannsen
- Jumper’s Knee
- Osteochondritis Dessicans
- Trauma
- Blount’s Disease
- SCFE
- Infection
- Tumor
Knee - ACL Function

- **Anterior Cruciate Ligament (ACL)** – prevents the tibia from moving forward independent of the femur, cutting/pivoting.
Knee - PCL Function

- Posterior Cruciate Ligament (PCL) – prevents the tibia from moving backward independent of the femur, “dashboard injury”
Knee - MCL / LCL Function

- Medial and Lateral Collateral Ligament (MCL / LCL) – prevents the tibia from moving medial or lateral independent of the femur
Knee - Meniscus Function

- **Meniscus** – shock absorber, dissipates force, “cushion”
Key History Questions

- Insidious and dull vs. *traumatic and sharp*
- Diffuse vs. *localized* pain
- Pain before / after sports vs. *during sport*
- Normal gait vs. *locking, instability, limping*
Knee Exam 101: The Basics

- Lateral Meniscus / LCL
- Medial Meniscus / MCL
- Pes Anserine
- Patella-Femoral
- Osgood-Schlatter
- IT Band
- Lateral Meniscus / LCL
Effusion = Fluid in the Knee Joint

- Preadolescents (7-12)
  - ACL injuries 47%
  - Meniscal tears 47%
  - Osteochondral fractures 13%

- Adolescents and Adults (> 13)
  - ACL injuries 65%
  - Meniscal tears 45%
  - Osteochondral fractures 5%
Physical Exam: Range of Motion
Physical Exam: Meniscus

- Tests to evaluate for possible meniscal pathology
  - McMurray’s Test
  - Thessaly Test
  - Apley Compression Test

McMurray’s Test
Physical Exam: ACL

Lachman’s Test
Physical Exam: PCL

Posterior Drawer
Physical Exam: MCL / LCL

Valgus Stress – MCL

Varus Stress - LCL
Additional Aspects of Physical Examination

- Can they extend their knee?
- If not, extensor mechanism is not intact
  - Patella fracture
  - Patella sleeve fracture
  - Patella tendon or Quad tendon tear
  - Tibial Tubercle Avulsion Fracture
Radiographic Evaluation

- Imaging:
  - X-ray (AP/Lat/Notch/Merchant)
    - Evaluate for fractures. Rule out physeal fractures, epiphyseal injuries, tumors, tibial spine fracture
  - MRI
    - Obtain if traumatic effusion is present
    - Evaluates soft tissue injuries, meniscus
    - Associated injuries
Imaging: Radiographs for Bone

1. Fractures / Dislocations
2. Alignment
3. Arthritis
4. Tumors
Imaging: MRI for Soft Tissue

1. Ligament Tears
2. Tendon Tears
3. Cartilage Tears
4. Loose Bodies
Discoid meniscus, ACL tear, meniscus tear
Case #1

- 12 year old female who participates in PE at school, but no other sports activities
- 3 months of knee pain when she goes up stairs or gets up after sitting for a long time.
- No major episode of trauma
- States that the pain is “all around the front” and motions around the patella

Exam:
- Negative Lachman
- Negative Posterior Drawer
- No instability
- Positive patella grind
- No other specific areas of tenderness
- Valgus at knee with single leg squat
Case #1 – Audience Response

- What is the diagnosis?
  1. Meniscus Tear
  2. ACL Tear
  3. Osgood-Schlatter’s Syndrome
  4. Patellofemoral Syndrome
  5. Osteochondritis Dissecans
Patellofemoral Pain

- Anterior knee pain – “behind the kneecap”
- Athletes and non-athletes
- Often worse with going up and down stairs, running and squatting down

- Due to overuse and/or poor tracking of the patella in the trochlear groove of the femur.
- Malalignment
- Muscle imbalance/weakness
- Poor technique with certain sports/activities
Case #2

- 14 year old male with 6 weeks of pain in the knee, particularly towards the end of soccer practice/games.
- No trauma that he can recall, although did slide tackle another player around the time that the pain started.

- Points to an anterior bump on the proximal tibia as the site of pain.

On Examination:

- Normal ROM, some pain anteriorly with forced flexion.
- No instability
- Negative Lachman and Posterior Drawer
- Tender to palpation over the tibial tubercle
Case #2 – Audience Response

What is the diagnosis?

1. Meniscus Tear
2. Sindig-Larson-Johansson Syndrome
3. Osgood-Schlatter’s Syndrome
4. Discoid Meniscus
5. Osteochondritis Dissecans
Osgood-Schlatter’s “Disease”

**History:**

- Pain at the tibial tubercle
- Repetitive traction leads to inflammation, swelling and pain.
- Improves with rest, worsens with running/jumping/sports
- May notice that prominence on anterior knee is “getting bigger”
Osgood-Schlatter’s Disease

- **Treatment**
  - Rest
  - Stretching
  - NSAIDs
  - Ice
  - Immobilization (rare)

- **Symptoms will resolve by end of growth (bump does not)**

- **Can pre-dispose to tibial tubercle avulsion fractures**

Sinding-Larsen-Johansson Syndrome

- Active Adolescents between ages 10-14
- Pain at inferior pole of patella
- Local inflammation, pain and swelling
- Worsened with running/jumping
- Treatment: Rest, strengthening/flexibility
Jumper’s Knee

- **Patellar Tendonitis/Tendinopathy**
- Not unique to adolescents – occurs in adults since not a growth plate issue
- Inflammation of the patella tendon itself (not it’s origin or insertion)
- Exacerbated by jumping/landing/cutting
  - Seen in sports with repetitive jumping (basketball, high-jump, gymnastics, volleyball)
Case #3

• 13 year old female presents with acute onset of right knee pain after twisting her knee playing soccer.

• Had swelling of the knee immediately.

• Could bear some weight, but needed help off the field

On Examination:

• Pain with range of motion

• Too much guarding on examination to assess Lachman and Posterior Drawer

• Large effusion on examination
Case #3 – Audience Response

- What is the most likely diagnosis given the clinical scenario?
  1. Meniscus Tear
  2. ACL Tear
  3. Tibial Spine Fracture
  4. Osteochondritis Dissecans
  5. MCL Sprain
What should you order?

1. Knee Xray and labs
2. Knee Xray and Hip Xray
3. Knee Xray and PT if negative for fracture
4. Knee Xray and MRI if negative for fracture
5. Knee Xray and Bone Scan if negative for fracture
ACL INJURIES

250,000 ACL Injuries / Year in the US
How Do You Tear Your ACL?

- Contact
- Non-contact
- Deceleration
- Internal rotation
- Hyper-extension
- Heard a “pop”
- Acute painful swollen knee
ACL Injuries
Natural History: Non-Op in Kids


**Patellar tendon graft reconstruction for midsubstance anterior cruciate ligament rupture in junior high school athletes. An algorithm for management.**

McCarron JR, Shelbourne KD, Porter DA, Rettig AC, Murray S.

Department of Research and Education, Methodist Sports Medicine Center, Indianapolis, Indiana 46202.

---

*Arthroscopy: The Journal of Arthroscopic & Related Surgery*  
*Volume 5, Issue 3, September 1989, Pages 197-200*

**Anterior cruciate ligament injury in children and adolescents**

Kevin R. Angel, David J. Hall

Department of Orthopaedic Surgery, Adelaide Children's Hospital, North Adelaide, South Australia, Australia.

---


**Anterior cruciate ligament tears in skeletally immature patients: meniscal pathology at presentation and after attempted conservative treatment.**

Graf BK, Lange RH, Fujisaki CK, Landry GL, Saluja RK.

Division of Orthopedic Surgery, University of Wisconsin Hospital and Clinics, Madison 53792.

---

Instability, Swelling, Pain, & Meniscal Injury
Conservatively Treated Tears of the Anterior Cruciate Ligament

Long-Term Results*

By Pekka Kannus, M.D.†, and Markku Järvinen, M.D.§, Tampere, Finland

From the University Central Hospital of Tampere, Tampere

Fig. 1-A

Fig. 1-B
Recommend ACL Reconstruction in Patients Under Age 30
So I Tore My ACL Yesterday: Can I Get It Fixed Tomorrow?

• Pre-op goals with PT prior to OR:
  • Full ROM
  • WBAT (no crutches)
  • No swelling
  • Leg control

Surgery is NOT an emergency!!
ACL is **REPLACED** not **REPAIRED**
MENISCUS TEARS
Key History

- Acute vs. chronic
  - Acute painful swollen knee
  - Locking / catching
  - Associated with ACL
  - Pain with deep squats
  - Joint line pain and tenderness
My Meniscus is Torn: What to Do?
My Meniscus is Torn: What to Do?

• Yes…Fix Me!!
  • Acute
  • Mechanical symptoms
  • Severe Pain
  • Young (Ped/Adol)

• No…Let Me Keep Playing!!
  • Chronic
  • Occasional Pain
  • Older
My Meniscus is Torn: Repair vs. Remove

Tears in the periphery have better blood supply and can be repaired, otherwise remove.
Case #4

11 year old female who was playing basketball in PE when she came down and landed awkwardly – Her knee bent inwards and she had significant, immediate pain in her knee.

Felt “something shift”

Was able to get up with help and walk, but needed assistance.

On Examination:

- Pain with range of motion
- Large effusion on examination
Case #4 – Audience Response

- What is the most likely diagnosis given the clinical scenario?
  1. Meniscus Tear
  2. ACL Tear
  3. Tibial Spine Fracture
  4. Osteochondritis Dissecans
  5. MCL Sprain
Traumatic Patellofemoral Dislocation
Background

- Majority non-contact twisting injury
- 10% direct trauma
- 29 per 100,000 (ped) to 5 per 100,000 (adult)
- Family history
- Most spontaneously reduce
  - Extension
Physical Exam

- Physical Exam (Acute)
  - (Obvious deformity if not yet reduced)
  - Effusion
  - Block to range of motion
  - TTP medial patella facet / lateral femoral condyle
Physical Exam

- Physical Exam (Recurrent)
- Femoral anteversion
- Genu valgum
- Tibial torsion
- Ligamentous laxity
- Tracking
- Patellar apprehension
- +/- Q angle
Imaging

- We look for things you have to deal with now
  - Fractures / dislocations

- And for things we have to deal with later
  - Proximal Soft Tissue
  - Distal Bone
  - Trochlear Dyplasia
  - Patella Alta
MRI = Osteochondral Fractures + MPFL + Trochlear Dysplasia
Treatment for 1st Time Dislocator

- Osteochondral Injury = Arthroscopy +/- ORIF
- No Osteochondral Injury = Physical Therapy
Surgical versus non-surgical interventions for treating patellar dislocation

No difference in 1st time dislocators
Recurrent Dislocation

- **Trochlear Dysplasia** = more PT

- **Patella Alta** = PT + later tubercle tx

- **TT –TG (proximal or distal)**
  - < 20 mm = MPFL reconstruction
  - > 20 mm = PT, wait until skeletal maturity, tubercle tx
OCD: History

- +/- Trauma
- Vague and poorly localized knee pain, swelling, and stiffness in varying degrees
- Worse with twisting / cutting movements
- “Locking” or “catching” may occur
OCD: Physical Exam

- Effusion
- Decreased ROM
- Tender over the femoral condyles
OCD: X-Ray

Notch View
OCD: MRI

Looking for fluid behind lesion!!
Case #5

11 year old patient presents with 6 weeks of right knee pain that started one day while running in PE.

There has not been much improvement since that day.

Mother notes that child limps and the leg is rotated outwards more than it was before.

Points to distal anterior and medial thigh as the location of pain.

They have been icing the knee and taking NSAIDs, but this doesn’t help much.
Audience Response – Case #5

What is the most important sequence of steps to take in evaluating this patient:

1. Inspection, Palpation, and ROM of the knee followed by Knee Xray

2. Inspection, Palpation, and ROM of knee followed by Xray and MRI of knee, given that the symptoms have been going on for over 6 weeks.

3. ROM of the Knee and Hip, Xray of the Pelvis if limited ROM of the hip

4. ROM of the Knee and Hip, Xray of the Pelvis if limited ROM of the hip and MRI of the right hip.

5. Xray of the knee and hip, laboratory evaluation.
Obligate external rotation with flexion
12 year old with left knee pain
Slipped Capital Femoral Epiphysis (SCFE)

- Most common hip disorder in adolescents
- Displacement of femoral neck relative to head
- Most common in ages 10-15
- In acute setting, analogous to SHI fracture of the proximal femoral physis.
SCFE – Epidemiology

- Common problem with serious consequences
- Annual incidence - 2 to 13 per 100,000
- Increased risk in certain groups
  - Male
  - Obese
  - Peripubertal
  - Polynesian
Presentation and Workup

• Complaints of groin/thigh/knee pain + / - trauma

• May or may not be ambulating

• May complain of only knee pain!!

• Check Hip Motion

• AP and frog lateral pelvis x-ray

• MRI of hip if seems like SCFE, but xray equivocal
Slipped Capital Femoral Epiphysis

- Consider endocrinopathy in patients younger than 10 with SCFE or thin child

- Classification:
  - Stable: able to bear weight
  - Unstable: unable to bear weight

- Unstable SCFE have much higher risk of osteonecrosis
Radiographs
Slipped Capital Femoral Epiphysis

- Ortho Consult; do not send home
- [Consider Endocrine Consult]
- Keep NWB – put in wheelchair (avoid crutches if possible and do not allow to walk)
- Will go to OR with Ortho for in-situ pinning.
Initial Treatment

• Prevent further slip progression
• Restore proximal femoral anatomy
Goals of SCFE Treatment

• Prevent further slip progression

• Restore proximal femoral anatomy
Post-Op
ID Unknowns

Meg Fisher, MD
Medical Director

The Unterberg Children’s Hospital
at Monmouth Medical Center
Barnabas Health
Disclosures

I have no disclosures

I may be mentioning off label uses of drugs
Objectives

• Select appropriate antibiotics for a variety of infections
• Name the bug to pick the drug
Case 1: September

26 mo white girl with swollen glands noted on returning from grandparents. No history of fever or other complaints. Grandmother coughing for 6 mo so she stopped smoking. Exam: 2 cm minimally tender node – submandibular area; molars emerging, rest of exam normal
Most likely diagnosis?

A. Teething
B. Epstein-Barr virus infection
C. Streptococcal pharyngitis
D. Tuberculosis
E. Some other viral illness
Do you start antibiotics?

A. Yes
B. No
C. Only if the mother insists
D. Only if the rapid strep test is positive
E. Only if I have a sample in the office
Case 1 Continues

Over the next two weeks the node increases in size; skin becomes violaceous. No history of fever. Area now a bit tender. Grandmother still coughing; her PPD is positive and her chest radiograph is negative.
Case 1

Photograph will be shown
Most likely pathogen?

A. *Haemophilus influenzae*, type b
B. *Haemophilus influenzae*, nontypable
C. *Staphylococcus aureus*
D. *Streptococcus pyogenes*
E. Atypical mycobacteria
More of Case 1

You place a PPD skin test on the child; at 72 hours, there is a 13 mm area of induration and about 18 mm of erythema.
Likely cause of + skin test

A. Tuberculosis from grandmother
B. Tuberculosis from someone else
C. Prior immunization with BCG
D. Exposure to tuberculosis
E. Infection with atypical mycobacteria
Rx for TB lymphadenitis?

A. Excision
B. Isoniazid for 9 months
C. No therapy needed
D. Azithromycin
E. Combination therapy
Rx for atypical mycobacteria

A. Excision
B. Incision and drainage
C. No therapy needed
D. Isoniazid, rifampin, ethambutol and pyrazinamide
E. Azithromycin or clarithromycin + ethambutol or rifampin
Case 2: August

13 yr Asian boy interrupted a dog and cat fight. He was bitten on his arm by his Yorkie. Wound is 1 cm by 5 mm; he cleaned it at home. The dog is immunized but the cat missed her appointment. The boy is immunized.
Best treatment?

A. Clean wound with povidone iodine or chlorhexidine
B. Clean + oral amox/clav
C. Clean + intramuscular ceftriaxone
D. Clean + Tdap
E. Clean + RIG + rabies vaccine
When do you suture?

A. If the bite involves the face
B. Bite of limb longer than 3 inches
C. Bite involves the finger
D. None of the above
E. More than one of the above
Most likely cause of infection

A. *Staphylococcus epidermidis*
B. *Eikenella corrodens*
C. *Corynebacterium bovis*
D. *Pseudomonas aeruginosa*
E. *Pasteurella multocida*
Drug of choice for infection

A. Amoxicillin/clavulanate
B. Ampicillin/salbactam
C. Clindamycin
D. Azithromycin
E. Linezolid
Most likely to transmit rabies

A. Dog
B. Cat
C. Bat
D. Snake
E. Raccoon
Case 3: Any Season

- The local urgent care center starts your 3 year old patient on cephasomething for otitis media. Now she has diarrhea and it is bloody.
- Diet: picky eater but nothing special
- Exposures: gymnastic classes, no travel
- Pets: older dog, cat, canary, gerbil and mice
Case 3

- Medications: cephahsomeone; H2 blocker
- Stool: loose, bloody, lots of it, stool smell
- Exam: afebrile, cranky but not toxic, well hydrated, ears look just fine, mild abdominal distention

What is the likely diagnosis?
Likely diagnosis

A. Norovirus
B. Clostridium difficile
C. Entamoeba histolytica
D. Shigella dysenteriae
E. Salmonella
My Thoughts

A. Virus not likely to cause bloody diarrhea
B. Best bet given the story
C. No travel so not likely
D. No travel so not likely
E. Possible, but we already talked about that
Case 3 Continues

- You suspect antibiotic associated diarrhea
- You send stool for toxin assay

Do you do anything else?
Anything else?

A. Stool culture for *Yersinia*
B. Start metronidazole
C. Stop the cephasomeone
D. Start loperamide, anti-motility agent
E. Start oral vancomycin
My Thoughts

A. Wrong story for *Yersinia*
B. Not approved for this use; may not need it
C. Excellent idea; always the first step
D. No, anti-motility drugs contraindicated
E. Not approved for children; may not need it
Case 3: The Plot Thickens

- Your patient recovers when you stop the antibiotic.
- Unfortunately, her 6 yr old brother now has bloody diarrhea. He is ventilator dependent and severely impaired following near drowning.
- His diet: enterostomy tube feedings
Case 3 - Brother

- Exposures: cared for at home, multiple hospitalizations
- Pets: the dog and cat interact with him
- Medications: seizure meds, asthma meds, H2 blocker, amoxicillin in a month ago
- Stool: loose, bloody, mucus, stool smell
Case 3 - Brother

- Exam: afebrile, nontoxic, ventilator settings unchanged, well hydrated, no new findings except diaper area rash
- Stool toxin assay positive for *C. difficile*

What to do?
What to do?

A. Hope his specialists will handle it
B. Start metronidazole
C. Start vancomycin
D. Arrange for a stool transplant
E. Call your favorite ID doctor
My Thoughts

A.  Hope springs eternal but don’t count on it
B.  Not a bad idea; off label
C.  More expensive and may not need it; off label
D.  Not the first step
E.  Not my favorite call but not a bad idea
Case 3 Continues

- He gets better but soon gets an antibiotic and once again develops bloody diarrhea.

Your thoughts?
Your thoughts

A. Not surprising; relapses are common
B. Wish I had started metronidazole with the ab
C. Time to retreat with metronidazole
D. Time to switch to vancomycin
E. More than one of the above
My Thoughts

A. Relapses are very common (25%)
B. Might have worked
C. Makes sense
D. Still expensive
E. Absolutely
**Clostridium difficile**

- **Illness:** antibiotic associated diarrhea to colitis
- **Spores common in the environment, resistant to cleaning, easy to transmit**
- **Colonization rates:** adults less than 5%, newborns up to 50% in some series
- **Dx:** toxin assay: many methods
Clostridium difficile

- Don’t test children < 1 yr; no test of cure
- Rx: stop antibiotics whenever possible; metronidazole or vancomycin (both off label in children); probiotics may help
- Control: wash hands, infection control, judicious use of antimicrobials

Relapses of *C. difficile*

- First one: retreat with same medicine
- Next one: vancomycin, tapering dose
- Other interventions: not proven
  
  Probiotics: better for primary prevention
  
  Immune globulin: IV or po
  
  Restore normal flora

Fecal Transplants

• Logical to restore normal flora
• First reported in 1958 in Denver
• Minimal studies in children
• Yuk factor is major but should be overcome
• In adults, superior to other therapies – 90%
• Routes: enema, nasogastric tube, capsules
Fecal Transplants

- Multiple recipes available online
- Clinical trials in progress
- Informed consent needed
- Donor stool must be screened and processed appropriately
- Issues: pathogens, “fat flora”, immune system

Kelly CP. *NEJM* 2013;368:474-5
Case 4: Spring

- Your 6 yr old African American boy with sickle cell disease is very excited about getting a pet for his birthday. He wants an iguana.

What advise do you give?
A. Sorry, but you can’t have any pet ever
B. Would you consider a snake instead
C. Don’t you think turtles are cuter
D. Let’s discuss this with your parents
E. Iguanas make wonderful pets
My Thoughts

A. Not a very caring approach
B. Snakes are colonized with *Salmonella*
C. Turtles are colonized with *Salmonella*
D. Great approach for child and family
E. Iguanas are colonized with *Salmonella*
Salmonella species

- Illness: diarrhea with rotten egg smell, green, abdominal pain, enteric fever
- Bacteremia and focal infections
- Increased risk in sickle cell disease
- Food: chicken, turkey, eggs, need $10^6$ orgs
- Pets: lizards, chicks, dogs, cats, turtle, etc
Salmonella

- **Dx:** stool culture, blood, urine, etc
- **Rx:** gastroenteritis: supportive; extraintestinal: TMP/SMX, other
- **Control:** wash hands, cook food, beware of pets, avoid cross contamination
- **Typhoid vaccine for travelers**
Quick Shots

4 month old, fever, hepatosplenomegaly, irritable, anemic

Photograph will be shown
Quick Shots

14 month old has been on cephasomething for 2 weeks

Photograph will be shown
Quick Shots

10 year old returned from a month with his cousins who live in Georgia, this itches

Photograph will be shown

Courtesy of Dr. Mancini
Quick Shots

2 1/2 year old back from North Carolina with fever, progressive rash and irritability. Photograph will be shown.
Quick Shots

The entire family is itching.
Steroids aren’t helping

Photographs will be shown

Courtesy of Dr. Mancini
Practice Changes

I encourage you to incorporate these changes in your practice:

• Name the bug before choosing a drug
The End
Rheumatology Unknowns

*What tests should I order?*

Nicole Ling, MD, MAS
Pediatric Rheumatology

12/3/2016
Disclosures

None
Learning Objectives

- Define Juvenile Idiopathic Arthritis (JIA)
- Differentiate arthritis & arthralgia
- Recognize broad differential for arthritis in children
- Understand the heterogeneity of JIA and its implications
What is JIA?

Juvenile Idiopathic Arthritis

- Onset before age 16
- Diagnosis of exclusion
- Persistent >6 weeks
**Nomenclature:**

**Arthralgia**
- “Joint pain”
- Used to describe pain without objective findings

**Arthritis**
- “Joint inflammation”
- Swelling or limited range of motion of joint accompanied by:
  - Warmth
  - Pain/tenderness
Anatomy/Pathology

- Synovial proliferation
- Increased synovial fluid
- Bone osteopenia/erosion
- Joint space narrowing -> ankylosis
- Cartilage destruction
- Joint mal-alignment/deformity/subluxation
- Periosteal reaction
- Growth disturbance

http://www.rheumatology.org/practice/clinical/patients/diseases_and_conditions/juvenilearthritis.asp
Is there arthritis?

- **Physical Examination**
  - Swelling
  - Decreased range of motion
  - Warmth
  - Tenderness
  - Growth disturbance
  - Extra-articular manifestations
Screening for arthritis

- Pediatric gait arms legs spine (pGALS)
- Pediatric regional examination of the musculoskeletal system (PREMS)


Arthritis Care & Research
Volume 55, Issue 5, pages 709-716, 29 SEP 2006 DOI: 10.1002/art.22230
http://onlinelibrary.wiley.com/doi/10.1002/art.22230/full#fig2
Extra-articular manifestations

- Uveitis
- Nodules
- Nail changes
- Rash
Anatomy

Uvea

- Iris
- Ciliary body
- Choroid
Uveitis in JIA

- Anterior, non-granulomatous inflammation
- Insidious onset, chronic
- Usually asymptomatic
- Can cause permanent vision loss
Complications of uveitis

- Synechiae
- Band keratopathy
- Glaucoma
- Macular edema
Clinical Pearl

Children that you are worried about having JIA should be screened for uveitis by an ophthalmologist.
Extra-articular manifestations

- Uveitis
- Nodules
- Nail changes
- Rash

RF + Poly JIA
Commonly found:
- Elbow
- Dorsum of hand - MCP’s

Nail Dystrophy
- Pitting
- Discoloration
- Early onycholysis
- Psoriatic subtype of JIA

American College of Rheumatology: image bank
Petty, Textbook of Pediatric Rheumatology, 7th ed.
Rash and JIA

- Rash present in two subtypes of JIA

Psoriatic JIA:
Silver scale
Dry, red patches

Systemic JIA:
Macular, salmon colored, evanescent

American College of Rheumatology: image bank
Petty, Textbook of Pediatric Rheumatology, 7th ed.
Is there arthritis?

- Physical Exam
- Radiographic Imaging
  - Magnetic Resonance Imaging
  - Ultrasound
  - Radiographs
Magnetic Resonance Imaging

- Order with and without contrast
- Visualize inflammation & damage
  - Hypertrophic, inflamed synovium
  - Effusion
  - Bone Marrow edema
  - Erosions
  - Ligaments, tendons, tendon sheaths
Ultrasound

- Synovial thickening
- Effusion
- Vascularity
  - Doppler sonography
- Guide injections
Radiographs

- Not always sensitive for effusion
- Assessment of damage:
  - Osteopenia
  - Erosion
  - Joint space narrowing
  - Joint subluxation/misalignment
  - Ankylosis
Is there arthritis?

- Clinical diagnosis made by physical exam findings and imaging.
- There is no lab test that indicates presence/absence of arthritis.
What is JIA?

Juvenile Idiopathic Arthritis

Onset before age 16

Diagnosis of exclusion

Persistent >6 weeks
Cases
Case 1

- 13 y/o M p/w acute onset of R elbow pain
  - 6 mo prior: T 99-100 x4 days after summer camp at Martha’s Vineyard
    - Developed blotchy, circular rash on trunk, HA
  - 4 days ago: R elbow pain, swelling
  - No preceding illness/trauma
- PE: effusion, warmth, tenderness, unable to fully flex/extend R elbow
- Labs: 9.2>13.5<340, CRP 22.4 (0-8), ESR 27
13 y/o M p/w acute onset of R elbow pain
• 6 mo prior: T 99-100 x4 days after summer camp at Martha’s Vineyard
• Developed blotchy, circular rash on trunk, HA
• 4 days ago: R elbow pain, swelling
• No preceding illness/trauma
PE: effusion, warmth, tenderness, unable to fully flex/extend R elbow
Labs: 9.2>13.5<340, CRP 22.4 (0-8), ESR 27

Diagnosis?
A. Inflammatory Bowel Disease  B. JIA
C. Lyme arthritis                D. Reactive Arthritis
Lyme Arthritis

- Spirochete B. Burgdorferi (tick: Ixodes)
- Disease: cutaneous, articular and neurologic
- Late finding
- Typically responsive to antibiotics
- Labs:
  - 50%: ESR elevated
  - Two tier testing:
    - Enzyme immunoassay (high sens, low spec)
    - Immunoblot (high specificity, confirmatory)
Case 2

- 8 y/o F p/w 2.5 wks swelling/pain knees, ankle, feet
- 2.5 wk ago jumped into van and hit L foot, neg XR
- 3 d later: swelling L knee, foot, then R foot, ankle, knee
- Unable to walk + fever for 9 days, waking from sleep
- No GI sx, no preceding illness, no rash (sm white pustule 1 wk ago, popped)
- PE: tachy, febrile, not moving, swollen wrists/hands, R knee, b/l ankles, diffuse pain
- Labs: 19.8>9.4<219, 17% bands, LDH 454, ESR >120, CRP 24
8 y/o F p/w 2.5 wks swelling/pain knees, ankle, feet

2.5 wk ago jumped into van and hit L foot, neg XR

3 d later: swelling L knee, foot, then R foot, ankle, knee

Unable to walk + fever for 9 days, waking from sleep

No GI sx, no preceding illness, no rash (sm white pustule 1 wk ago, popped)

PE: tachy, febrile, not moving, swollen wrists/hands, R knee, b/l ankles, diffuse pain

Labs: 19.8>9.4<219, 17% bands, LDH 454, ESR >120, CRP 24

Diagnosis?

A. Septic Arthritis  
B. JIA

C. Inflammatory bowel disease  
D. Cancer
Septic Arthritis

- Infectious agent found in synovial space
- Bacteria, virus, spirochete, fungi
- Most common in < 2 yrs old
- Joint pain, severe, can have pseudoparalysis
- Typically monoarticular
- Swelling, warmth, sometimes erythema
- Typically with systemic sx: fever, HA, vomiting
- Culture blood and other sites of possible infection, tap joint
Case 3

- 15 y/o M p/w foot and knee pain/swelling x 3 wk
- 5 wk ago: 1 mo: bloody diarrhea, preceded by trip to MX
- Tx with flagyl, Stool O&P, cx negative
- Diarrhea resolved, decreased PO, no abd pain, ↓16 lbs
- 3 wk ago: B/l red eyes, discharge, dysuria x 1 wk
- PE: ↓: lateral flexion, extension of neck, L shoulder ext rotation. Effusions in knees, L>R, pain with MTP squeeze L > R esp at 5th MTP L.
- 10.3>12.3<315, CRP 159, ESR > 100, ALT 107, ASO nl, tap: neg
15 y/o M p/w foot and knee pain/swelling x 3 wk
5 wk ago: 1 mo: bloody diarrhea, preceded by trip to MX
Tx with flagyl, Stool O&P, cx negative
Diarrhea resolved, decreased PO, no abd pain, ↓16 lbs
3 wk ago: B/l red eyes, discharge, dysuria x 1 wk
PE: ↓: lateral flexion, extension of neck, L shoulder ext rotation. Effusions in knees, L>R, pain with MTP squeeze L > R esp at 5th MTP L.
10.3>12.3<315, CRP 159, ESR > 100, ALT 107, ASO nl, tap neg

Diagnosis?

A. Inflammatory Bowel Disease  B. JIA
C. Lyme arthritis         D. Reactive Arthritis
Reactive Arthritis

- Related to enteric or genitourinary bacterial infections
  - Yersinia, Salmonella, Shigella, Camplylobacter
  - Chlamydia trachomatis
- Predominantly HLA B27 + individuals
- Triad: arthritis, conjunctivitis, urethritiscervicitis
- Arthritis: lower limb, asymmetric, oligoarticular
- Preceding infection: clinically within 4 weeks, or if no symptoms, need lab confirmation.
Infections cause arthritis

- Septic
  - Bacterial, viral, spirochete, fungi
- Post infectious
  - Strep (arthritis and rheumatic fever)
  - Reactive arthritis
  - Transient synovitis
Case 4

- 8 y/o M p/w ankle, heel and thumb pain x 2-3 wks
  - 2 mo ago:
    - red, painful bumps on legs x 2 weeks
    - N/v, decreased PO, fatigue, slightly loose stools
    - Mouth sores
  - 66th to 34th percentile, weight over last 10 mo
- PE: mild R ankle swelling, MTP tenderness
- Labs: 10.6> 9.6 <Plt 749, Albumin 2.5, ESR 34, CRP 4.4
8 y/o M p/w ankle, heel and thumb pain x 2-3 wks

- 2 mo ago:
  - red, painful bumps on legs x 2 weeks
  - N/v, decreased PO, fatigue, slightly loose stools
  - Mouth sores

- 66th to 34th percentile, weight over last 10 mo

PE: mild R ankle swelling, MTP tenderness

Labs: 10.6> 9.6 <Plt 749, Albumin 2.5, ESR 34, CRP 4.4

Diagnosis?

A. Inflammatory Bowel Disease  
B. JIA  
C. Cancer  
D. Reactive Arthritis
IBD associated arthropathy

- Arthralgia or arthritis
- Peripheral or axial (including SI)
- Supporting lab evidence
  - Elevated ESR, CRP, acute phase reactants
  - Low albumin
  - Stool testing: FOBT, fecal calprotectin
Case 5

- 16 y/o F, recently diagnosed with Hashimoto’s
  - Intermittent fever, adenopathy
- Joint swelling for the last month: knees, knuckles
- Edema in ankles, hair loss
- PE: nasal ulceration, hepatomegaly, mild adenopathy, pitting edema to shins, swelling b/l knee, L elbow, decreased ROM elbow, MCP’s, PIPs
- Labs: 5>9.4147, ESR 71, CRP 2.8, albumin 3, Cr 1
- CXR: small to moderate L pleural effusion
16 y/o F, recently diagnosed with Hashimoto’s
- Intermittent fever, adenopathy
- Joint swelling for the last month: knees, knuckles
- Edema in ankles, hair loss
- PE: nasal ulceration, hepatomegaly, mild adenopathy, pitting edema to shins, swelling b/l knee, L elbow, decreased ROM elbow, MCP’s, PIPs
- Labs: 5>9.4<147, ESR 71, CRP 2.8, albumin 3, Cr 1
- CXR: small to moderate L pleural effusion

Diagnosis?

A. Systemic Lupus Erythematosus  
B. Systemic JIA  
C. Cancer  
D. Reactive Arthritis
Arthritis and other rheumatic disease

- SLE
- Juvenile Dermatomyositis
- Mixed connective tissue disease
- Systemic Sclerosis
- Behcet’s
- Kawasaki
- Henoch-Schonlein purpura
- Periodic fever syndromes
- Sarcoidosis
Case 6

- 12 y/o M p/w 2.5 mo R knee pain
- 6 weeks ago, XR showed effusion
- Developed pain: b/l elbow, wrist (possible swollen), ankle pain
- No fever, 15 lb weight loss, no GI sx
- Difficulty sleeping due to pain
- PE: Diffuse arthritis: shoulder, elbow, wrist, hand, knee, ankle
- Labs: 4.5>11.3<177, CRP 5.38 (<0.8), ESR 86
- 5d later, unable to ambulate, crying in pain all day
- 12 y/o M p/w 2.5 mo R knee pain
- 6 weeks ago, XR showed effusion
- Developed pain: b/l elbow, wrist (? swollen), ankle pain
- No fever, 15 lb weight loss, no GI sx
- Difficulty sleeping due to pain
- PE: Diffuse arthritis: shoulder, elbow, wrist, hand, knee, ankle
- Labs: 4.5>11.3<177, CRP 5.38 (<0.8), ESR 86
- 5d later, unable to ambulate, crying in pain all day

**Diagnosis?**

A. Inflammatory Bowel Disease  
B. Systemic JIA  
C. Cancer  
D. Lupus
Cancer causes arthritis

- Leukemia
  - Result of leukemic infiltration of the synovium
  - Usually oligoarticular rather than polyarticular
  - Relative thrombocytopenia

- Neuroblastoma

- Malignant bone tumors
  - Osteosarcoma, Ewing sarcoma, rhabdosarcoma

- Benign bone tumors
  - Osteoid osteoma, osteoblastoma
Ddx arthritis

- **Infection**
  - Septic arthritis
    - Bacteria, virus, spirochete, fungi

- **Autoimmune/Autoinflammatory**
  - SLE
  - Juvenile Dermatomyositis
  - Mixed connective tissue disease
  - Systemic Sclerosis
  - Behcet’s
  - Kawasaki
  - Henoch-Schonlein purpura
  - Periodic fever syndromes
  - Sarcoidosis
  - Inflammatory bowel disease

- **Reactive/Postinfectious**
  - Post-streptococcal arthritis
  - Rheumatic fever
  - Reactive arthritis
  - Toxic synovitis

- **Malignancy**
  - Leukemia
  - Neuroblastoma
  - Malignant and benign bone tumors

- **Other**
  - Trauma (+/-) hemophilia
  - Serum Sickness
  - Common variable immunodeficiency
Are we there yet?

Juvenile Idiopathic Arthritis

- Onset before age 16
- Diagnosis of exclusion
- Persistent >6 weeks
Juvenile Idiopathic Arthritis

- Seven different subtypes
  - Oligoarticular (persistent vs extended)
  - RF + Polyarticular
  - RF – Polyarticular
  - Systemic onset
  - Psoriatic
  - Enthesitis related
  - Undifferentiated
**Oligoarticular**

1-4 joints: 1st 6mo

Most common, age 2-4y, caucasian, large joints, not hips, F>>M

incr risk for uveitis if ANA + and <7 yrs at dx.

**RF + Polyarticular**

5+ joints: 1st 6mo
RF +

Like adult RA, late adolescent F>>M

CCP confirms RF status

**RF - Polyarticular**

5+ joints: 1st 6mo

2nd most common, age 2-4, 6-12yrs, F>>M

**Psoriatic**

Arthritis + [Psoriasis or 2/3: dactylitis, nail pitting, +fhx psor]

Age 2-4 or 9-11 yrs F>M, periph or axial

HLA B27

**Enthesitis Related**

Arthritis &/or enthesitis +2/5: SI/back pain, + HLAB27, onset male>6yrs, acute ant uveitis, + fhx

Late childhood/early adolescence M>>F, periph or axial

HLA B27

***Systemic Onset**

Arthritis + 2wk fever + 1/4: rash, LAD, HSM, and/or serositis

F = M

Biggest morbidity

incr LFTs ferritin, ESR, CRP
Macrophage Activation Syndrome

- Excessive activation and expansion of macrophages and T cells
- See with SJIA, SLE, & Kawasaki
- Potentially fatal
Macrophage Activation Syndrome

Signs/Symptoms:
- Persistent Fever
- HSM
- LAD
- Liver dysfunction
- Coagulopathy c/w DIC
  - Rash, epistaxis, GI bleed
- Mental status changes

Lab findings:
- Cytopenia (2/3)
- Fall in ESR
- Hypofibrinogenemia
- Elevated AST/ALT, bilirubin, LDH, TG, ferritin
<table>
<thead>
<tr>
<th>Component</th>
<th>Notes</th>
<th>Most common, age 2-4y, caucasian, large joints, not hips, F&gt;&gt;M</th>
<th>incr risk for uveitis if ANA + and &lt;7 yrs at dx.</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Oligoarticular</strong></td>
<td>1-4 joints: 1st 6mo</td>
<td>Most common, age 2-4y, caucasian, large joints, not hips, F&gt;&gt;M</td>
<td>incr risk for uveitis if ANA + and &lt;7 yrs at dx.</td>
</tr>
<tr>
<td><strong>RF + Polyarticular</strong></td>
<td>5+ joints: 1st 6mo</td>
<td>Like adult RA, late adolescent F&gt;&gt;M</td>
<td>CCP confirms RF status</td>
</tr>
<tr>
<td><strong>RF - Polyarticular</strong></td>
<td>5+ joints: 1st 6mo</td>
<td>2nd most common, age 2-4, 6-12yrs, F&gt;&gt;M</td>
<td></td>
</tr>
<tr>
<td><strong>Psoriatic</strong></td>
<td>Arthritis + [ Psoriasis or 2/3: dactylitis, nail pitting, +fhx psor ]</td>
<td>Age 2-4 or 9-11 yrs</td>
<td>HLA B27</td>
</tr>
<tr>
<td><strong>Enthesitis Related</strong></td>
<td>Arthritis &amp;/or enthesitis +2/5: SI/back pain, + HLAB27, onset male&gt;6yrs, acute ant uveitis, + fhx</td>
<td>Late childhood/early adolescence</td>
<td>HLA B27</td>
</tr>
<tr>
<td>*<strong>Systemic Onset</strong></td>
<td>Arthritis + 2wk fever + 1/4: rash, LAD, HSM, and/or serositis</td>
<td>F = M</td>
<td>incr LFTs, ferritin, ESR, CRP</td>
</tr>
<tr>
<td><strong>Undifferentiated</strong></td>
<td>Arthritis that fills no or 2+ categories</td>
<td>F = M</td>
<td>incr LFTs, ferritin, ESR, CRP</td>
</tr>
</tbody>
</table>
So if lab tests do not diagnose arthritis, are they worth sending?

- Yes.
  - CBCd, ESR, CRP, CMP

- Depends on clinical scenario
  - If suspect SJIA, additionally: ferritin, fibrinogen, LDH, coags, TG
  - If suspect SLE: U/A with micro, ANA
  - Others include: Lyme, ASO, Anti-DNAse B, or other tests if other rheum diagnoses are suspected

- Call us!
Take home points

- Arthritis is a clinical diagnosis made by physical exam and sometimes with imaging.
- There is no lab test that will tell you if the patient has arthritis or not.
- Children that you are worried about having JIA should be screened for uveitis by an ophthalmologist.
Take home points

- Systemic JIA is the only subtype where we typically see fever, rash, and arthritis, and these patients can be pretty sick.

- There is a long differential for arthritis in children, navigating nuances can be difficult – call us!
References

- American College of Rheumatology, Image bank:
  http://images.rheumatology.org/

- American College of Rheumatology, patient fact sheet:
  http://www.rheumatology.org/practice/clinical/patients/diseases_and_conditions/juvenilearthritis.asp


- Rheum Dis Clin North Am 33:3, 441-70, vi
