Question #1

- Parent brings 4 year old child
  - CC: Nighttime pain in knees, ankles
  - No limping, swelling, functional impairments
  - Responds to massage, occasional NSAIDS
- PE:
  - Benign
  - Healthy happy child
- Question:
  - What is most appropriate next step?

- A. Imaging of knees and ankles
- B. Lab work
- C. Trial 6 weeks of consecutive NSAIDS
- D. Ophthalmology evaluation for NSAIDS
- E. Reassurance

Growing Pains

- Common benign syndrome of recurrent discomfort affecting young children
  - Common age of onset 3-6 years
  - No increased risk of inflammatory joint condition
- Not an inflammatory or dangerous condition
  - Diagnosis is based on clinical history
  - 80-90% with bilateral discomfort
  - Resolves with analgesics, massage
  - Systemic symptoms should prompt additional evaluation
  - Resolves without lasting sequelae
A 2 year old male (pictured at left) presents with his mother who complains that his left knee is swollen and he is limping for the past 6 weeks. He is not in any distress and not in significant pain but is having difficulty walking up and down stairs. He has had no preceding trauma or infection. The complication most likely associated with this process is:

a. Disability, contractures  
b. CNS involvement  
c. Cardiac arrhythmia  
d. Uveitis  
e. A, b, C

**Differential Diagnosis of Joint Swelling**

- **Acute**
  - Infection related arthritis
  - Septic
  - Reactive-Lyme
  - Malignancy
  - Leukemia
  - Neuroblastoma
  - Hemophilia
  - Trauma

- **Chronic**
  - JIA
  - TB
  - Sarcoidosis

**Juvenile Idiopathic Arthritis (JIA)**

- Synovial inflammation leading to bone/joint erosion
- Morning stiffness, limp, or falling often
- Easy fatigability
- Joint swelling
- Minimal pain
- Joint NEVER red or exquisitely tender
- Alteration of activities
- Loss of function
Juvenile Idiopathic Arthritis (JIA)

- Oligoarticular
  - Persistent
  - Extended (>4 joints after 6 months)
- RF Positive Polyarticular
- RF Negative Polyarticular
- Systemic Onset
- Psoriatic Arthritis
- Enthesitis-related Arthritis
- Other

Pauci JRA

- 4 or fewer joints
  - Large joints: knees, ankles, wrists
  - NOT HIP
- Serology
  - Positive ANA
  - Negative RF
- Main morbidity
  - ASYMPTOMATIC ANTERIOR UVEITIS
  - (associated with positive ANA)
  - Can lead to blindness
Poly JRA

- 5 or more involved joints
  - Small and large joints
  - PIP, MCP, wrist
- Rheumatoid nodules
- ANA may be positive
- RF may be + or –
  - If + then worse prognosis
Question #3

- A 4-year-old female presents with 3 weeks of non-remitting fevers, bilateral knee, ankle, and unilateral elbow pain and swelling, as well as a salmon-colored rash on her trunk that fluctuates with her fever. What is the test most likely to be negative/normal in this patient?

- A. ESR
- B. Ferritin
- C. Hemoglobin
- D. Platelet count
- E. ANA

Systemic JIA

- Males = Females
- Quotidian fever
- Arthritis
- Visceral involvement
  - HSM
  - LAD
  - Serositis
- Leukocytosis
- Rash
  - Evanescent, salmon-colored
  - ANA and RF negative

"Fleeting salmon-color rash"
- Macular or wheal-like
- Not pruritic
- Irregular
- May coalesce with fever
Question #4

A 10 year old patient presents with a history of 4 weeks of a swollen middle toe without any noted trauma. On examination you note the following:

• What statement is most likely true in this patient?
  ▫ A. This is likely traumatic
  ▫ B. Patient likely has osteomyelitis of the toe
  ▫ C. Soaking in Epsom salts or warm water will be beneficial for this patient
  ▫ D. Patient will likely have nail pitting or onycholysis
  ▫ E. Surgical intervention is necessary to help this patient prevent loss of their toe

Psoriatic Arthritis

• ILAR Criteria
  ▫ Arthritis and Psoriasis
  ▫ OR
  ▫ Arthritis and two of the following:
    ▪ Dactylitis
    ▪ Nail pitting or onycholysis
    ▪ Family history of psoriasis in a first degree family member

• Epidemiology
  ▫ 2-15% of patients with chronic arthritis
  ▫ Age of Onset: Bimodal: preschool years and middle to late childhood
  ▫ Sex Ratio
    ▪ F slightly more predominant than M
Psoriatic Arthritis

- Arthritis
  - Predominantly asymmetric large and small joints
  - Predilection for small joints of hands and feet
  - Dactylitis
    - "sausage digits"
    - Swelling in one or more digits that extends beyond the joint margin

Psoriatic Arthritis

- Rash:
  - Well demarcated, erythematous, scaly lesion
  - Occurs over extensor surfaces of elbows, forearms, knees and interphalangeal joints
  - 80% of patients with psoriatic arthritis

Psoriatic Arthritis

- Nail Pitting
  - Small, round, shallow and dimple like
- Onycholysis
  - Complete nail dystrophy
Spondyloarthritis

- Enthesitis-related JIA
- Enthesis: insertion of ligaments and tendons into bone
- Asymmetrical arthritis affected 4 or fewer joints
- Male predominance

Ankylosing spondylitis

- Enthesitis of axial skeleton and sacroiliac joints.
  - Present with back pain
  - Loss of lumbar sacral mobility
- Oligoarthritis of joints of lower extremities
- Common presentation
  - Male with back pain, morning stiffness that is relieved w/ exercise
- Labs
  - HLA-B27 positive
  - Increased ESR
  - ANA and RF are NEGATIVE
- Radiology
  - Bamboo Spine
Question #5

3 year old boy admitted with right ankle swelling, high fever, and GI symptoms (vomiting, abdominal pain, and bloody stools). Labs show leukocytosis with left shift, elevated inflammatory markers, and anemia. Stool culture reveals a diagnosis of shigella and appropriate antimicrobial therapy is started.

Which statement is most correct regarding this patient?

A. Antibiotics will most likely help resolve the swelling
B. HLB27 is likely to be positive
C. Intra-articular steroid injection will be needed
D. Long term antimicrobials and surgical intervention will be necessary

Post infectious/Reactive Arthritis

- Common cause of lower limb oligoarthritis
- Part of spondylo-arthritis family
- HLAB27+(50x greater risk)
- Diagnosis:
  - Temporal link with infectious trigger
  - Management of infectious trigger important
  - Linked usually to GI and GU bugs
  - Natural history of independent of antibiotic therapy

Post infectious/Reactive Arthritis

- Reiter's syndrome:
  - ACR definition: peripheral arthritis >1 month with urethritis or cervicitis
  - Classic Definition: urethritis, conjunctivitis and arthritis secondary to infectious dysentery
- Recovery
  - Typically spontaneous within 3-12 months
  - Some develop chronic arthritis
A 16-year-old Hispanic female presents to your office after returning from summer vacation in the Caribbean for the past 2 months. She notes that over the past 2 months she has developed diffuse joint pains and swelling around her ankles which have progressed to involve her mid-calf. She also notes the development of an erythematous rash on her face and neck, as well as generalized fatigue and tactile temperatures. On examination, her joint pain is greater in her lower extremities, and her BP is 140/80. She has a malar rash, oral ulcers and 2+ pitting edema to her mid-calf. Initial labs reveal:

- Sodium: 137 mEq/L
- Potassium: 4.7 mEq/L
- BUN: 40 mg/dL
- Creatinine: 2.1 mg/dL
- Albumin: 1.8 g/L
- Hgb: 8.7 g/dL
- UA: 3+ protein, 2+blood
- ANA: 1:2560
- dsDNA: 1:1280

The most important subsequent test to help guide treatment is:

- A. MRI brain
- B. Kidney biopsy
- 24 hour urine collection
- EMG
- Bone marrow aspiration

Systemic Lupus Erythematosus

- Multisystemic autoimmune disease of unknown etiology
- More common in females
  - Prepubertal: 4:1
  - Postpubertal: 8-9:1

SLE Criteria ≥ 4/11

- **4 Skin**
  - Malar Rash
  - Discoid Rash
  - Photosensitivity
  - Oral Ulcers
- **2 Immunologic**
  - ANA
  - dsDNA, anti-Smith, antiphospholipid antibodies
- **5 Organ Systems**
  - CNS
  - Seizure
  - Psychosis
  - Serositis
  - Kidney
  - Proteinuria
  - Arthritis
  - Non erosive
  - Hematologic
  - Lymphopenia (<4,000)
  - Lymphopenia (<1,500)
  - AIHA
  - Thrombocytopenia (<100,000)
Discoid Lupus

Well-circumscribed, red-purple, elevated plaques

Malar Rash

Spares nasolabial folds

Oral Ulcers
Question #7

- You are called to evaluate a full-term newborn born three days ago to a healthy 27 year old G1P1 mother, who developed the following rash after treatment for hyperbilirubinemia with phototherapy. What is the most appropriate management of the rash?
  - A. Referral to dermatology for KOH scraping
  - B. Treatment with steroid topical cream
  - C. Advice the family on sun protection and reassure them that this is self-limited
  - D. Refer to hematology
  - E. LP and antibiotics

Neonatal Lupus

- Maternal Transfer of Antibodies
  - Anti-Ro (SS-A)
  - Wane at 6 mo
  - Even with asymptomatic mom’s

- Complications
  - Rash
    - Heart block – usually 3rd degree
    - Damage and scarring during 2nd trimester
    - Not reversible
  - Hepatitis
  - Neutropenia/thrombocytopenia
  - Hydrops fetalis

- Treatment
  - Supportive
  - May need cardiac pacing

Neonatal Lupus

Raccoon Eyes

Annular Scaling

Annular plaques
Congenital Heart Block

Drug Induced Lupus

- D-SLE
  - D = Drugs for the Heart (procainamide)
  - S = Sulfonamides
  - L = Lithium
  - E = Epilepsy medications (anticonvulsants)

- Others
  - INH
  - Minocycline

- Most often reversible
- ANA typically positive, but transiently
- Anti-histone Ab

Question #8

- A 9 year old girl presents with fevers, progressive fatigue and proximal muscle weakness. She has the following rashes on presentation. The test most likely to confirm the diagnosis is:
  - A. Slit lamp examination
  - B. Lab testing
  - C. MRI thighs
  - D. Chest X ray
  - E. Muscle Biopsy
Juvenile Dermatomyositis (JDM)

- Myopathy and Vasculopathy
- Myopathy ➔ Symmetrical proximal muscle weakness
- Vasculopathy ➔ Skin Manifestations

JDM: Clinical Manifestations

- Insidious in onset
- Constitutional Symptoms
  - Fatigue
  - Fever
  - Weight loss
  - Muscle weakness
- Physical Findings
  - Heliotrope Rash
  - Photosensitive rash – upper torso, extensor surfaces of arms/legs
  - Nail fold telangiectasias
  - Gottron papules
  - Gower’s sign
  - Dysphagia/dysphonia/dyspnea
  - Nodular calcifications

Heliotrope Rash

- Violaceous hue
- Periorbital edema
- Malar rash
Gottron’s Sign
Pathognomonic for JDM

Red, thickened, scaly skin overlying PIPs

Calcinosis
Striae
Nail fold dilation and loops
Photosensitive Rash

JDM: Work Up & Treatment

- Labs
  - Increased CK, Aldolase, LDH, AST, ALT
  - Increased vWF Antigen
  - Usually nl ESR/CRP
  - + ANA at times
- Radiology
  - Increased T2 signal on MRI b/l thighs
- EMG
  - Normal NCS, increased muscle irritability and discharge
- Treatment
  - Sunscreen
  - Steroids
  - Methotrexate
  - IVIG
- Complications
  - At high risk of gastric perforation

Complications
Question #9

- A 6 year old boy with recent strep pharyngitis presents to his pediatrician's office with intermittent cramping abdominal pain for the past 2 days. He has had decreased food intake secondary to abdominal pain, and his mother reports that he has been complaining of leg pain. On examination, he is well appearing with a rash as shown below, and has a left swollen and tender ankle and knee.

Of the following the most important laboratory investigation to order next is:

A. CBC
B. IgA level
C. ANCA levels
D. Urinalysis with microscopy
E. BMP

Henoch-Schonlein Purpura (HSP)

- IgA mediated leucocytoclastic vasculitis
- Most common small vessel vasculitis in children
- Usually preceded by URI or Strep infection
- Age: 2-13 years old
- Usually self limited

HSP: Clinical Manifestations

- Rash
  - Palpable purpura
  - Angioedema
- Abdomen
  - Colicky pain (may precede skin rash)
  - Intussusception - currant jelly stool
  - Ileoileal
- Arthritis
  - Large joints: knees, ankles, wrists
  - Periarticular therefore no damage to joint
- Renal
  - Hematuria
  - Proteinuria
Palpable Purpura

- May have some superficial ulcerations
- Concentrated on buttocks and lower extremities

![Image of palpable purpura]

HSP: Laboratory Evaluation

- **NORMAL PLATELETS**
- Mild/mod ↑ WBC
- Urinalysis
  - Range from normal to nephritic picture
- ↑ ESR
- ANA/RF negative
- C3, C4 normal
- ANCA negative

HSP: Clinical Course

- Usually self-limited disease in childhood
- Resolution of symptoms in 6-8 weeks
- Recurrence in 33% within the first few months
- Prognosis dependent upon renal involvement
  - Ranges from GN to Renal insufficiency
- Monitoring
  - Weekly Uas and BP measurements with active lesions then monthly x 6 months
HSP: Treatment

- Supportive therapy for joint and abdominal complaints

- NSAIDs may aggravate abdominal complaints
  - Avoid in pts with renal manifestations

- Controversial role of steroids
  - Inability to hydrate
  - Renal disease
  - Severe arthritis

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Scleroderma

- Scleroderma
  - Systemic
  - Localized
    - Multisystemic Disease
    - Morphea
    - Linear

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Linear Scleroderma

- Linear bands of hard, translucent, shiny skin
- Flexion contracture
- Muscle atrophy
Morphea
- Flesh colored, erythematous or purplish patches
- Ivory plaque

Raynaud’s Phenomenon
- Sharp demarcation

CREST
- Sclerodactyly
- Calcinosi
- Telangiectasia
- Raynaud
Question #10

- A 2 year old boy presents with 6 days of fever to 40 degrees C, significant irritability, conjunctival injection, and diffuse maculopapular rash on his torso and lower extremities. On physical examination, he is noted to be irritable, febrile to 39 degrees, and with a strawberry tongue, unilateral cervical adenopathy, injected conjunctivae that are non-exudative and limb-sparing. Laboratory investigations reveal mild anemia, elevated ESR and CRP, and mildly elevated transaminases. He is admitted to the floor for treatment. The intervention most likely to prevent morbidity related to this disease is:
  • A. Aspirin therapy
  • B. Antibiotics
  • C. IVIG therapy
  • D. Motrin
  • E. Steroids

Kawasaki disease

- AKA Mucocutaneous Lymph Node Syndrome

  - Definition:
    ▫ Self-limiting Medium Size Vasculitis
    ▫ Etiology
      ▪ Unknown
      ▪ Maybe immune mediated in a genetically predisposed child
    ▫ Affects mainly young children
      ▪ Peak 2-3 years, 80% are less than 5 years old
      ▪ All races, although more common in Asians, and least common in African Americans
    ▫ Complication
      ▪ Coronary Artery aneurysms develop in 15-25% of untreated children and may lead to ischemic heart disease or sudden death
      ▪ Most common cause of acquired heart disease in the US

Kawasaki Disease

- Fever ≥ 5 days plus 4/5
  ▫ Rash (not vesicular)
  ▫ b/l non exudative bulbar conjunctivitis (limbic sparing)
  ▫ Oral mucus membrane changes
  ▫ Single unilateral anterior cervical lymph node enlargement ≥ 1.5 cm
  ▫ Hand/foot changes (edema, erythema, peeling)
KD: Laboratory Studies

- Inc WBC (PMN predominance)
- Inc Platelet count > 7 days
- Anemia for age
- Inc AST and bilirubin
- Low Albumin
- Hyponatremia
- Inc ESR/CRP
- Sterile pyuria

Kawasaki Disease

- Treatment
  - IVIG
    - Only proven therapy to decrease coronary artery abnormalities
  - ASA therapy
    - High dose initially
    - Low dose after IVIG treatment until CRP normalizes
  - Cardiology evaluation

Question #11

An 8 year old female presents with 1 week of high grade fevers, and migratory joint pain ~8 days following reported sore throat and abdominal pain. Management of this patient includes all of the following except:

- A. EKG
- B. Echo
- C. Throat Culture
- D. Upper GI
- E. ESR, CRP
Acute Rheumatic Fever

- Dramatic decline since the 1940’s with recent resurgence
  - Outbreaks in poor, overpopulated communities
- Complications
  - Rheumatic Heart disease: Major cause of acquired heart disease in the world

Acute Rheumatic Fever: Modified Jones Criteria (1992)

- Major Criteria:
  1. Migratory arthritis (counter clockwise) of large joints
     - Most frequent and least specific
     - Large joints: typically first in lower extremities and then upper
     - Painful polyarthritis
     - Transient, self limited (1-3 days/joint, 2-3 weeks total)
     - Very responsive to ASA and NSAIDS

- Carditis
  - Endocarditis (most common)
    - Asymptomatic or with new heart murmur
  - Myocarditis
    - Typically presents with heart failure
  - Pericarditis
    - Chest pain, discomfort, pleurisy, friction rub

- Valvular disease
  - Mitral > Aortic > Tricuspid > Pulmonary Valve
Acute Rheumatic Fever: Modified Jones Criteria (1992)

- Major Criteria
  - Sydenham’s Chorea
    - Abrupt, purposeless, nonrhythmic involuntary movements
    - Muscular weakness
    - Emotional disturbance
    - May be bilateral or unilateral
    - Longer latency period, late isolated finding
    - Self limited: 2-3 months
    - 5% of ARF patients

- Major Criteria
  - Erythema Marginatum Rash
    - Erythematous rings on the trunk
    - Fluctuant over the course of weeks to months

- Major Criteria
  - Subcutaneous nodules
Acute Rheumatic Fever: Modified Jones Criteria (1992)

- **Minor Criteria**
  - Fever
  - Arthralgias
  - Elevated inflammatory markers (CRP, ESR)
  - Prolonged PR
- All in the presence of evidence of recent strep infection

Management of ARF

- **General Measures**
  - Hospitalize
  - Bed rest
  - If carditis, typically 4 weeks of rest from activity

- **Antimicrobial therapy**
  - Eradication of strep pharyngitis
  - Does NOT alter course, frequency or severity of cardiac involvement

Management of ARF

- **Suppression of Inflammatory Response:**
  - ASA 100mg/kg/day
    - Toxicity issues
    - No good controlled studies comparing ASA and NSAIDS
  - NSAIDS
    - Used in cases of ASA intolerance or allergy
    - Meta analysis of ASA and Steroids
  - Duration
    - Clinical response
    - Normalization of acute phase reactants
Management of ARF

- **Secondary Prevention**
  - **Purpose**: prevention of recurrent RF
  - **Antibiotic choices**
    - IM benzathine Benzylpencillin q3-4 weeks (q4 weeks in low risk areas, or low risk patients)
    - Oral PCN
      - Compliance risk

Management of ARF

- **Duration of Secondary Prophylaxis**

<table>
<thead>
<tr>
<th>Category of patient</th>
<th>Suggested duration of secondary prophylaxis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient, without proven carditis</td>
<td>For 6 years after the last attack, or until 18 years of age whichever is longer.</td>
</tr>
<tr>
<td>Patient, with carditis, recent mitral valve surgery or healed carditis</td>
<td>For 6 years after the last attack, or at least until 25 years of age whichever is longer.</td>
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</tbody>
</table>

Question #12

- A 16 year old female patient presents with wide spread pain to all of her joints and muscles, associated with fatigue and non-restorative sleep for the past 4 months. She demonstrates a significant response to both non painful and painful stimuli, but has no objective findings on examination.

- What is the most likely diagnosis of this patient?
  - A. Depression
  - B. Juvenile Fibromyalgia
  - C. Juvenile dermatomyositis
  - D. Conversion disorder
Juvenile Fibromyalgia

- Chronic pain condition
  - Widespread MSK pain
  - Allodynia
  - Hyperesthesia
  - Fatigue
  - Sleep disturbances
  - HA
  - IBS
  - Mood disorders
  - F>M