# Joint pain, Fevers, Rashes and + ANA's….The World of Pediatric Rheumatology and Beyond

## Ricardo Guirola MD M Ed

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# Objectives

- Discuss clinical approach for Pediatric patients with suspected rheumatic diseases
- Brief overview of some common conditions with MSK complaints
- Brief overview of some common rheumatic conditions
- Discuss utility of lab testing in the diagnosis of patients with suspected rheumatic disease

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Famous Statements

Dr. House "ITS NEVER LUPUS"



How should we clinically approach these conditions???

## Definitions

- Arthralgia
  - Artist **formerly** known as joint pain
- Arthritis (objective)
  - **Warmth**
  - **Swelling**
  - **Stiffness**
  - Decreased ROM
  - Erythema +/-

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## Tendonitis

Patella

Femur

Tibia

Fibula

Inflamed patellar tendon

Anterior (front) view of knee
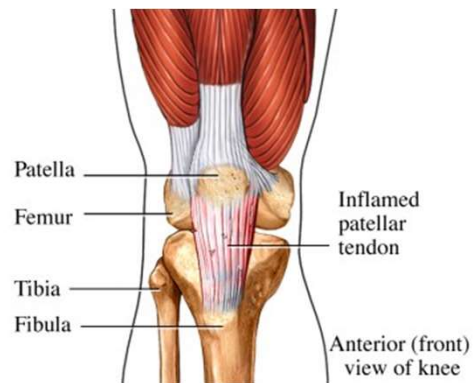
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# Enthesitis
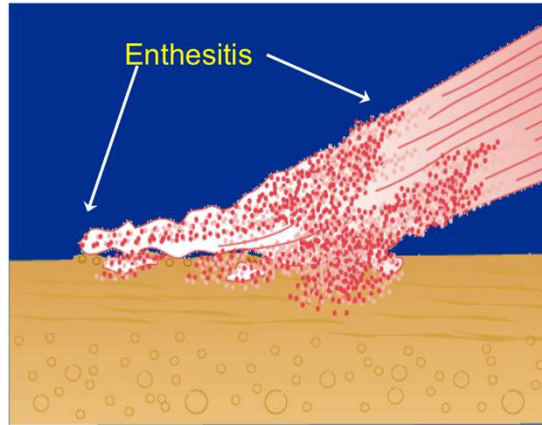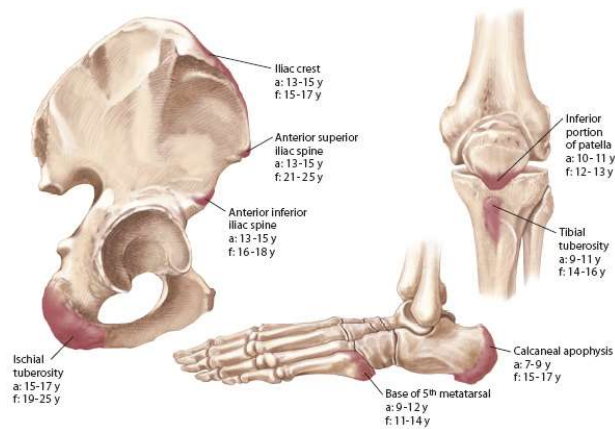


# Apophysitis

# Inflammatory Joint Pain



**REST WORSENS SYMPTOMS**

# Non Inflammatory Joint Pain
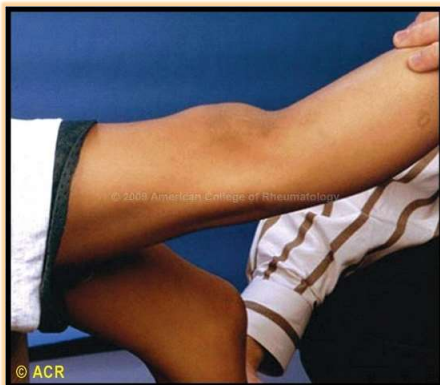


**Activity Worsens Symptoms**
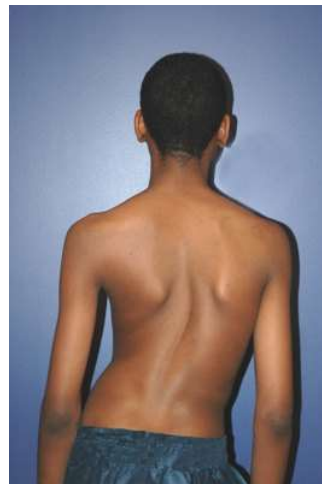
# Always ask the **<u>extra</u>** questions

- Antecedent history (trauma, infection)?
  - Acute vs Chronic
- **<u>Systemic Symptoms</u>**
  - Fevers
  - Rashes
  - Weight loss
- Extracurricular Activities
  - Athletics
    - Hours per week
  - Previous injuries



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# **<u>Clinical Pearl</u>**





- Acutely Swollen Joint
  - Warmth
  - Fever
- Until proven otherwise……

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# Bottom Line, Why do we Need to Know all this Stuff ??
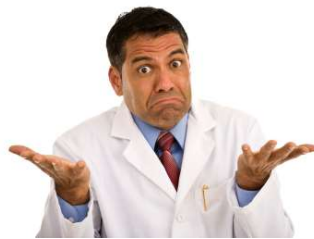


# Importance of MSK Medicine

- MSK Complaints in children and adolescents **30%** percent of visits
- National Ambulatory Survey MSK complaints 2$^{nd}$ to **cough** and **acne**
- Deficiencies in MSK medicine at resident and student level
- **2000-2010 WHO Bone and Joint Decade**

# Importance of MSK Medicine

- MSK Symptoms are **Common**?!
- 97% of complaints: NON-RHEUMATIC
  - **YET: 300,000 children in US with arthritis-spectrum disorders**
    - 24,000-30,000 in TX?
    - Rheumatic disorders chronic and require early diagnosis
    - MSK pain **not** universal in children with juvenile arthritis



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# MSK EXAM

- Lack of confidence in MSK exam
  - Leads to unnecessary lab testing
- Bottom line is practice makes perfect!



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# pGALS



- Screening tool

- Designed by expert panel

- Questionnaire

- Pictorials

- **Takes 3-4 minutes**

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# Case # 1

- 8-year-old male competitive baseball player for the last 6 weeks has been having daily exquisite discomfort and tenderness to bilateral feet after baseball practice, pain is very severe, causes remarkable limp, in addition some difficulties during school days, due to pain and discomfort, no swelling reported, no stiffness reported, evaluated by PCP, x-rays of feet were negative, labs were normal except for mildly positive **ANA of 1:80 speckled**

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# Case # 1

- ROS WNL
- PE- VS WNL
- EXAM WNL except  for *magical* test called **heel squeeze**



- **DX is Severs Disease**

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# But wait a minute what about the **ANA**???



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# LAB GOLDEN RULES



1) KNOW THE LAB
2) KNOW HOW LAB IS PERFOMED
3) **KNOW WHAT TO DO WITH LAB RESULT AND PATIENT**

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# Rheumatologist=Vampire

- Basic labs much more relevant
  - **Complements**
    - SLE
- **ANA and RF**
  - Poor sensitivity for pediatric population

- Labs helpful when you are sure about the diagnosis



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# A few words about ANA's

- Can be positive in :
  - **normal person – up to 15-30 %**
  - other autoimmune diseases
  - chronic liver disease
  - neoplasms
  - TB, malaria, SBE, EBV, HIV, Mycoplasma

- **Flip a coin!!!** (positive 40-50% in **JIA**)

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# So just because a kid has a + ANA….

- **Does not exactly mean**
  - **Autoimmunity**
- ELISA/IFA
  - Commercial labs
  - ACR recs…

- **Unfortunately if SLE**
  - **100 %**



Intruder! DESTROY!

Wh-what?!

But I'm one of you!!

Don't listen to her lies.

Autoimmune disorders in a nutshell.
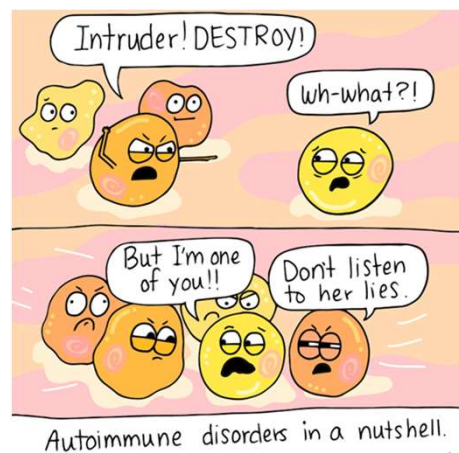
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# Use caution when ordering…



# The Infamous Rheumatoid Factor

- Immunoglobulin against FC portion of IgG
  – IgM
- **Only positive in 3-5% of all JIA!!!!**
- Positive
  – 1-4% of population
  – Viral Infections
  – TB
  – Hepatitis
  – Malignancy
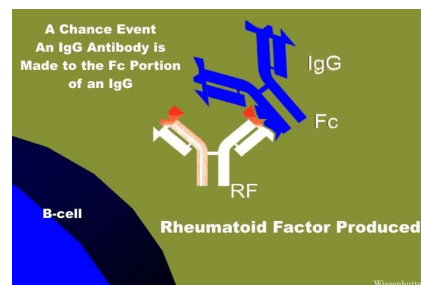
# Other more "sophisticated" tests

Axial arthritis        Peripheral arthritis
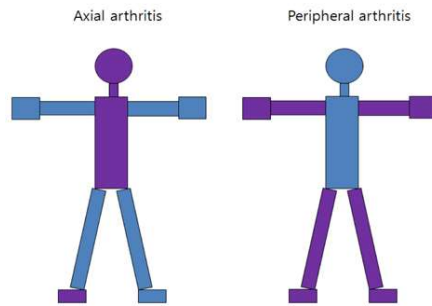
- CCP
  - Very specific to inflammatory arthritis
    - RF + JIA and adult RA

- HLA B 27
  - + in 5 % of Caucasians
  - Specific for Axial Arthritis
  - Not sensitive

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# My Joints Really Hurt….yet I **do not** have JIA or SLE!

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# Hypermobility Syndromes

- 2-3 % of population
- Beighton Score
  - 6 and above
- Arthralgia frequent
  - Increased activity
- Management
  - Rest
  - ICE
  - **PT**
  - Analgesics



# Hypermobility Syndromes



**You Know You Have EDS When...**

You do something that should hurt you, but it doesn't... YAY!!

Then you hurt yourself getting excited that you did it without getting hurt...

FB you know you have EDS when
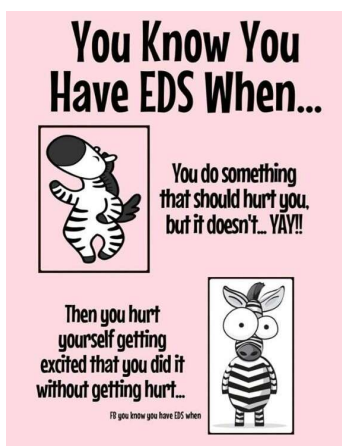
- Spectrum?
  - EDS?
  - Genetics?
- POTS/Dysautonomia
- Chronic Pain
- GI dysfunction
- MAST Cell

## Growing pains



- Prevalence of 4-37%
- 2-12 yrs of age
- Classic presentation
  - Pain in PM or evening
  - Increased activity
  - Resolves by next day
  - Analgesics or comfort
  - **PE nl**
  - **Labs nl**

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## Overuse Injuries

- All the "tendinitis's"
  - Patella
  - Achilles
  - Pes anserine
  - Low back pain
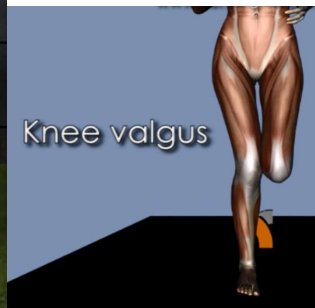  - Throwing injuries
- Overtraining
- Poor Biomechanics



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# Poor Biomechanics



- Growth
- Decreased Proprioception

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# Traction Apophysitis

Sinding Larsen Johansson



Sever's Disease

Tibia

Fibula

Achilles Tendon

Pain

Calcaneus

Osgood-Schlatter Disease

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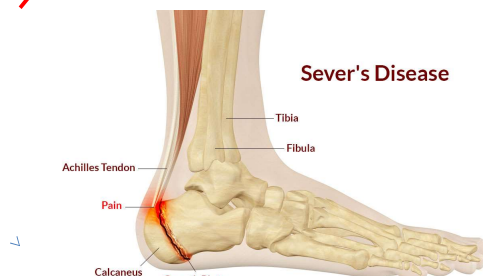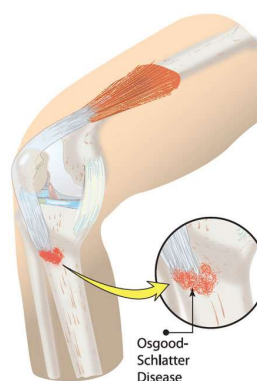
## ISELIN DISEASE

# Case #2

- CC: "All my joints hurt"
- HPI:
  - 7 yr. old boy, arthralgias
    - LE
  - 2 months duration
  - No AM stiffness
  - No limp or swelling
  - Fatigue, no weight loss

- **Wakes up in pain 5/7 nights**
- ESR 47, CRP 7.5 (elevated)
- Hgb 10.9 (MCV 80)

- Sent to ER due to persistence of symptoms

## Case 2

- VS tachycardia
- GEN: mild pallor
- MS: mild reduced flexion of left wrist, no effusions or other joint abnormalities
- **Biggest concern?**



T1 coronal    T1 coronal    T1 post-sagittal

**Leukemia**
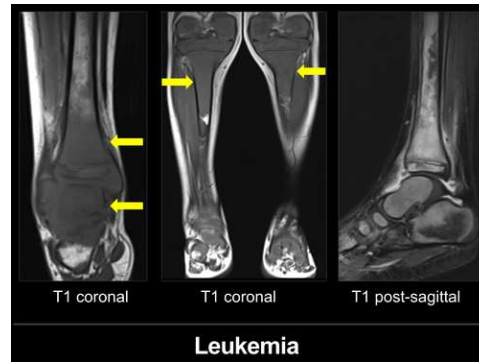
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# Clinical Pearl # 2



DANGER

DO NOT MISS THIS ONE!

- Malignancies common in children
  - Often have MSK complaints
  - **BONY PAIN**
- Ill appearing
- Cytopenias
  - Rare in JIA

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# Lets head into the Rheumatology realm!!!



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# Case 3

- 2 yr old
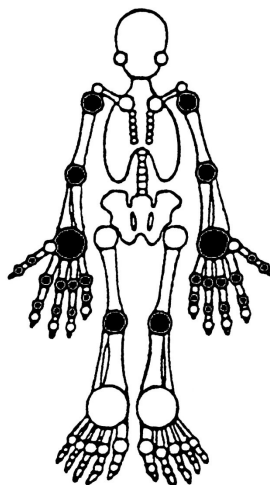  - Limp x 2 months
    - Left leg??
  - Worse in am
  - Better in afternoon
  - Worse during naps??

- Told growing pains??
- SENT TO ER



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## Case 3

- Exam
- VS   stable <u>No fever</u>
- **<u>Left knee 2 + effusion with warmth lacks full extension</u>**
- Rest of exam nl

- Labs CBC nl
- ESR and CRP mildly elevated
- X rays mild knee effusion



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## Kids get arthritis too....

- Over 60,000 children in US
- Multi-disciplinary and multi-modal tx
- **<u>Diagnoses of exclusion</u>**

- **<u>Juvenile Idiopathic Arthritis</u>**
  - Unknown etiology



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# Criteria for Classification of JIA


Kids get Arthritis too
Arthritis Foundation®

- Age at onset <16yr
- Evidence of arthritis
- 6 wks
- EXCLUSION diagnosis

- **AT NO POINT IS THERE MENTION OF JOINT PAIN SOLELY**

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# Juvenile Idiopathic Arthritis

- Oligoarthritis
  – Persistent
  – Extended
- Polyarthritis
  – RF (-)
  – RF (+)
- Systemic Onset
- Psoriatic Arthritis
- Enthesitis Related Arthritis



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# JIA oligoarticular



- Younger children
- < 4 joints affected
- Chronic Uveitis common
- Pain not severe
- **ANA +**
  - **30 % UVEITIS**

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# Polyarticular JIA

- Teenagers or older kids
- Usually affecting hands
  - Multiple joints
- >5 joints
- **RF +/-**
  - **RF + = RA**
    - **CCP**
  - **RF –**

- **TREATMENT**





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# Systemic JIA

- Daily Fever
  - Quotidian pattern
- Salmon like rash
  - Dermatographism
- **Arthritis**
- EXCLUSION
- Macrophage Activation Syndrome
  - **Ferritin**
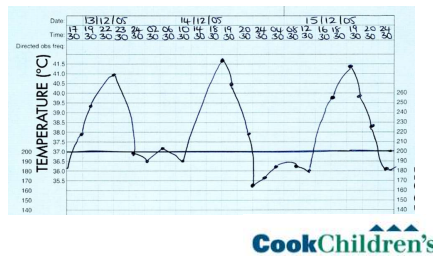


# Enthesitis Related Arthritis



- Stiffness
- Enthesitis
- Attention to SI joint
  - Axial Skeleton
- Juvenile Ankylosing Spondylitis
  - Radiographic
- HLA B 27

# Psoriatic Arthritis

- Classic Rash
  - 30-40 % absent
- First line relative
- Asymmetric small joints
- Uveitis
  - 30%
- Dactylitis
- HLA B27

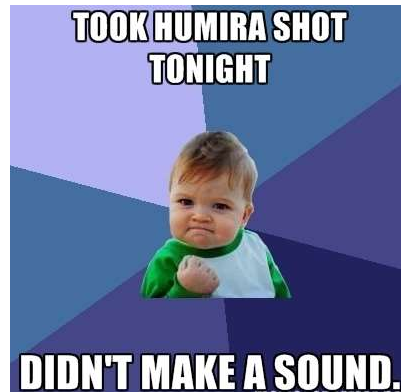**Cook**Children's.



# Medications for JIA



- NSAIDS
- DMARDS
  - Methotrexate
    - Oral
    - SQ
    - IV
- Corticosteroids
  - Oral
  - IV
  - **Intraarticular**

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# Medications for JIA

- Biologics
  - TNF α (**JIA**)
    - **Etanercept**
    - Adalimumab
    - Infliximab
  - IL-1 (**Systemic**)
    - **Anakinra**
    - **Canakinumab**
  - IL-6 (**Systemic/Poly JIA**)
    - Tociluzumab
- **Infections (TB)**
- **Malignancies** ?



TOOK HUMIRA SHOT TONIGHT

DIDN'T MAKE A SOUND.

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# Anterior Uveitis
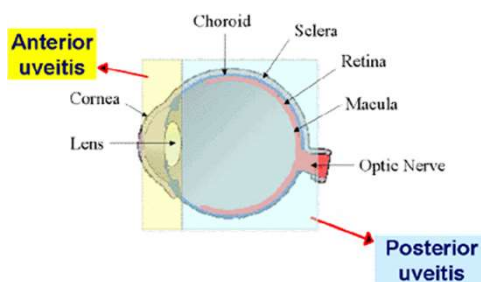


-**Frequent screening**:

ANA +, Oligoarticular JIA .

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# Case 4

- 13 year old female previously well until the last **3 months** when she developed
  - **30 lb weight loss**
  - **Fatigue**
  - decreased appetite noted by parents
    - intentional or unintentional?
  - dancer
  - honor roll student



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# Case 4

- PE:
  - VS: T 98°F, HR **100→125, BP 130/70, RR 12**
  - Weight 101 lbs, Height 5"4
  - Skin: No rashes, **hair thinning particularly over temporal regions bilaterally**, eyebrows intact
  - HEENT: **palatal ulcer**, gingival inflammation
  - Neck: No thyromegaly
  - CVS: tachycardia with mild activity otherwise negative
  - Lungs: CTA
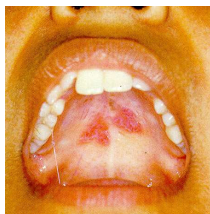


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# Case 4

- ABD: quiet BS, no masses palpated
- **Extremities: mild pitting edema of lower extremities to mid shins**
- Neuro: CN II-XII intact, normal gait, 2+DTR's, normal strength in all extremities
- **MSKE: Pain with flexion and extension of wrists, unable to perform pad to palm or make fists bilaterally. Swelling of 2<sup>nd</sup> and 3<sup>rd</sup> PIP's bilaterally. Otherwise negative.**

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# LABS for CASE 4

- Labs
  - UA: microscopic **hematuria and 3+ proteinuria**
  - **+ hemolytic anemia**, mild thrombocytopenia
  - Elevated AST and ALT

  - **ANA >1:1280 Homogenous**
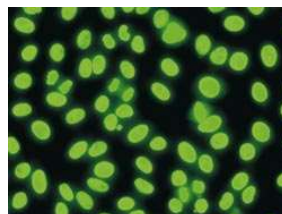
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# How do patients with SLE present?



# SLE: Etiology & Pathogenesis...

# SLE : Classification Criteria ACR (1997)

**Total 11 :**

**3 skin** (malar rash, discoid, photosensitivity)

**3 itis** (oral ulcers, arthritis, serositis)

**3 major organs** (renal, neurologic, hematological)

**2 labs**  (ANA, specific auto antibodies including APLA)
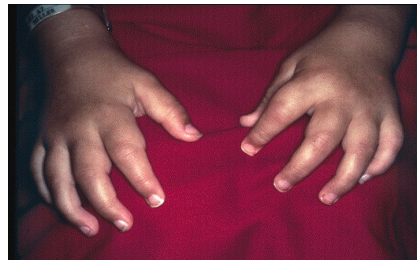
**Joint pain and + ANA……**

**4 out of 11 criteria provide a sensitivity of 96% and a specificity of 96%.**

**SLICC CRITERIA 2012**

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# SLE:
# MSK Manifestations

- Non-erosive arthritis
  - ≥ 2 joints
  - Symmetric
- **Fatigue**
- Weakness
- Myositis
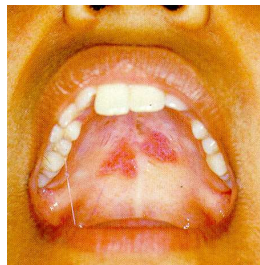- Avascular necrosis



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# SLE:
# Skin and Mucosa

**Prevalence of 76%**

**Butterfly rash**
**Discoid lesions**



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**Photosensitivity**
**Oral/nasopharyngeal ulcers**



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## SLE:
## Severe Organ Involvement

- **Renal Disease (80%)**

- Neuropsychiatric lupus
  - Central nervous system
  - Peripheral nervous system
- Pulmonary disease

- Cardiac disease
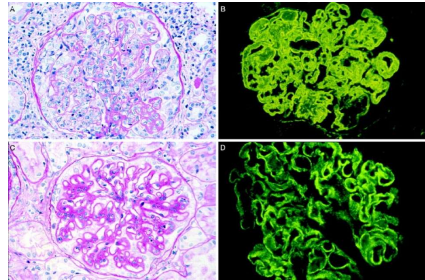


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# Dermatomyositis



- Proximal muscle weakness
- Heliotrope rash
- Gottron's papules
- **Labs (elevated):**
  - AST, ALT
  - CK, aldolase, LDH
  - **ANA ???**

MRI- diffuse muscle inflammation

Muscle biopsy: inflammation, atrophy

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# Henoch-Schönlein Purpura (HSP)
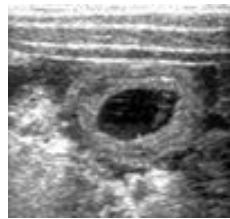




- Purpura
  - Petechiae
- Joint complaints
  - Soft tissue swelling
  - +/- arthritis
- Abdominal pain
  - Intussuception
- Renal involvement

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# Scleroderma Spectrum





-**limited disease:**

**morphea**

**linear scleroderma**

**coup de sabre**

**More common than systemic sclerosis**

**Heart, GI tract, Lung involvement**

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# So If I were a primary care doc...??

- History
  - Inflammatory
  - Non Inflammatory
  - Detailed history
- **PE=**
  - **CHECK ALL JOINTS!**
- **MSK EXAM**



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# So If I were a primary care doc...??
## Labs to get



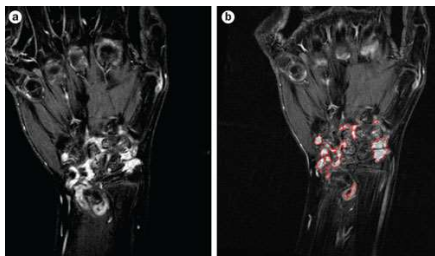- Basic labs
  - CBC
  - CMP
  - UA
  - CRP
  - ESR
- Infection
  - **Blood Cx**
  - **Gram Stain and Cx**
- Malignancy
  - **Peripheral smear**
  - **LDH/URIC ACID**

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## So If I were a primary care doc…??
## Imaging to get



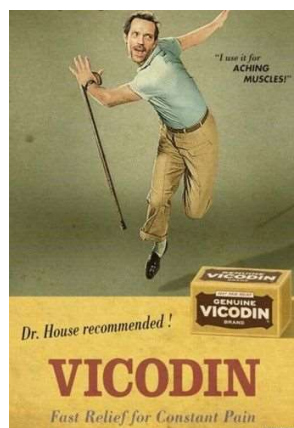- Immediate get an MRI or CT

- Basic X rays
  – Exclude trauma
  – Malignancy
- Concerns with inflammatory MSK condition
  – **CONTRAST**

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## So If I were a primary care doc…??
## Treatment and Management

- NSAIDS OK to use
  – Unless contraindicated
  – LIVER
  – KIDNEY
  – GI
- Naproxen
  – 10-20mg/kg BID
- **Careful with opiates!**
  – **Amplified Pain**



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## So If I were a primary care doc…??
## Treatment and Management



- Don't use corticosteroids
  - Until knowing diagnosis
  - Exclusion
    - Malignancy
    - Infection
- **When necessary call your friendly Rheumatologist**

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# Summary and Conculsions

- Kids do get arthritis but…
  - **Inflammatory arthritides are diagnoses of exclusion**
- MSK complaints may be features of systemic Dx
- **Trust your Hands, Eyes, and Ears (H + P)**
  - **MSK EXAM**
- **Labs mostly useless, unless…**
  - **You know what you are looking for!**

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# When necessary call your friendly Ped Rheumatologist

- ● Pediatric Rheumatology Centers with Board Certified Pediatric Rheumatologists
- ● Pediatricians or Rheumatologists treating Children with Rheumatic Diseases (w/o Center)
- ● Pediatric Rheumatology Centers with Board Certified Pediatric Rheumatologists and Approved Fellowship Training Program
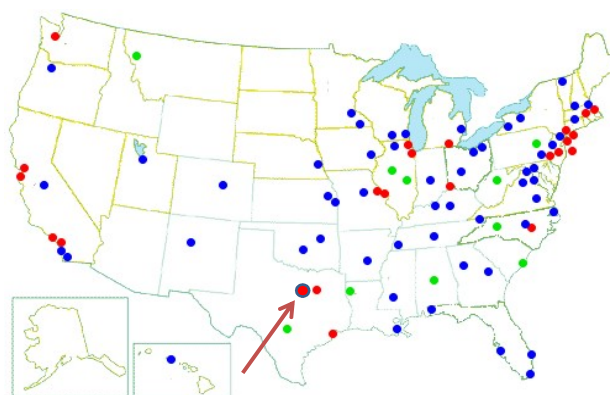


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*Jandial S, Myers A, Wise E, Foster HE,* **Doctors** **likely to encounter children with musculoskeletal complaints have low confidence in their clinical skills.** J Pediatr. 2009 Feb;154(2):267-71. Epub 2008 Sep 27.

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