Joint pain, Fevers, and Rashes the World of Pediatric Rheumatology and Beyond

Ricardo Guirola MD M Ed
Objectives

• Discuss clinical approach for 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
Famous Statements

• “Just because an 8-year-old boy has bilateral heel pain does not necessarily mean that he has an inflammatory arthritis”

Dr. Guirola discussion with Sports Med Fellow
Famous Statement # 2

Dr. House “ITS NEVER LUPUS”
How should we clinically approach these conditions???

Autoimmune Arthritis

What society thinks it is.  What people think we take.  What people think it feels like

What people think causes it.  What we really take.  What it really is.
Definitions

• Arthralgia
  – Artist formerly known as joint pain

• Arthritis (objective)
  – Warmth
  – Swelling
  – Stiffness
  – Decreased ROM
  – Erythema +/-
Tendonitis

- Patella
- Femur
- Tibia
- Fibula
- Inflamed patellar tendon

Anterior (front) view of knee
Inflammatory Joint Pain

REST WORSENS SYMPTOMS
Non Inflammatory Joint Pain

Activity Worsens Symptoms
Always ask the **extra** questions

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

- Acutely Swollen Joint
  - Warmth
  - Fever
- Until proven otherwise......
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 2nd 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
MSK EXAM

• Lack of confidence in MSK exam
  – Leads to unnecessary lab testing

• **Bottom line is practice makes perfect!**
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
Case # 1

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

- DX is Severs Disease
But wait a minute what about the ANA???
LAB GOLDEN RULES

1) KNOW THE LAB
2) KNOW HOW LAB IS PERFORMED
3) KNOW WHAT TO DO WITH LAB RESULT AND PATIENT
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
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**)
So just because a kid has a + ANA....

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

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

Autoimmune disorders in a nutshell.
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

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

- **HLA B 27**
  - + in 5% of Caucasians
  - Specific for Axial Arthritis
  - Not sensitive
My Joints Really Hurt....yet I **do not** have JIA or SLE!
Hypermobility Syndromes

- 3% of population
- Beighton Score
  - 6 and above
- Arthralgia frequent
  - Increased activity
- Connective tissue diseases?
- Management
  - Rest
  - ICE
  - PT
  - Analgesics
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
Overuse Injuries

• All the “tendinitis’s”
  – Patella
  – Achilles
  – Pes anserine
  – Low back pain
  – Throwing injuries
• Overtraining
• Poor Biomechanics
Poor Biomechanics

- Growth
- Decreased Proprioception
Traction Apophysitis

Sinding Larsen Johansson

Sever's Disease

- Tibia
- Fibula
- Achilles Tendon
- Calcaneus
- Growth Plate
- Pain

Osgood-Schlatter Disease

Cook Children's
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?**
Clinical Pearl # 2

- Malignancies common in children
  - Often have MSK complaints
    - **BONY PAIN**
- Ill appearing
- Cytopenias
  - Rare in JIA
Lets head into the Rheumatology realm!!!
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
Case 3

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

- Labs CBC nl
- ESR and CRP mildly elevated
- X rays mild knee effusion
Kids get arthritis too....

- Over 60,000 children in US
- Multi-disciplinary and multi-modal tx
- **Diagnoses of exclusion**
- **Juvenile Idiopathic Arthritis**
  - Unknown etiology
Criteria for Classification of JIA

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

- AT NO POINT IS THERE MENTION OF JOINT PAIN SOLELY
Juvenile Idiopathic Arthritis

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

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

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

• TREATMENT
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
Medications for JIA

- **NSAIDS**
- **DMARDS**
  - Methotrexate
    - Oral
    - SQ
    - IV
- **Corticosteroids**
  - Oral
  - IV
  - Intraarticular
Medications for JIA

• Biologics
  – TNF α (JIA)
    • Etanercept
    • Adalimumab
    • Infliximab
  – IL-1 (Systemic)
    • Anakinra
  – IL-6 (Systemic)
    • Tociluzumab
• Infections (TB)
• Malignancies ?
Anterior Uveitis

-Frequent screening:
ANA +, Oligoarticular JIA.
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
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
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\textsuperscript{nd} and 3\textsuperscript{rd} PIP’s bilaterally. Otherwise negative.
LABS for CASE 4

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

  – **ANA >1:1280 Homogenous**
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
SLE:
MSK Manifestations

- Non-erosive arthritis
  - > 2 joints
  - Symmetric
- **Fatigue**
- Weakness
- Myositis
- Avascular necrosis
SLE:
Skin and Mucosa

Prevalence of 76%

Butterfly rash
Discoid lesions
Photosensitivity
Oral/nasopharyngeal ulcers
SLE: Severe Organ Involvement

- Renal Disease (60%)
- Neuropsychiatric lupus
  - Central nervous system
  - Peripheral nervous system
- Pulmonary disease
- Cardiac disease
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
Henoch-Schönlein Purpura (HSP)

- Purpura
  - Petechiae
- Joint complaints
  - Soft tissue swelling
  - +/- arthritis
- Abdominal pain
  - Intussusception
- Renal involvement
Scleroderma Spectrum

- limited disease:
  - morphea
  - linear scleroderma
  - coup de sabre

More common than systemic sclerosis
Heart, GI tract, Lung involvement
So If I were a primary care doc...??

• History
  – Inflammatory
  – Non Inflammatory
  – Detailed history

• PE=
  – CHECK ALL JOINTS!

• MSK EXAM
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
So If I were a primary care doc...??

Imaging to get

- Immediately get an MRI or CT

- Basic X rays
  - Exclude trauma
  - Malignancy

- Concerns with inflammatory MSK condition
  - **CONTRAST**
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
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
Summary and Conclusions

• 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!
When necessary call your friendly Rheumatologist
Cassidy et al, Textbook of Pediatric Rheumatology 2011


