PITFALLS IN PEDIATRIC RHEUMATOLOGY

Mary D. Moore, MD

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1

OBJECTIVES

- To review the differential diagnosis of musculoskeletal conditions in children
- To briefly discuss the evaluation and management of rheumatic disorders
- Present an overview of the complications of childhood rheumatic disorders

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CASE 1 - DR

- 8 week old infant- admitted locally for irritability, fracture of arm noted. Full term to 18 yo G1, no perinatal problems. On admission - aseptic meningitis, hemolytic anemia, and ESR 145. Treated for 10 days with ampicillin and ceftriaxone.
- age 10 weeks - ESR 112, WBC normal H/H 8/26, PTH, fibroblast cultures normal. Bone survey - multiple fractures. ESR decreases to 50 and infant is sent home to foster care.

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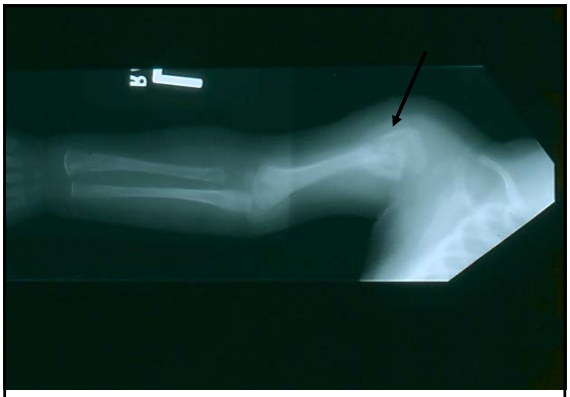

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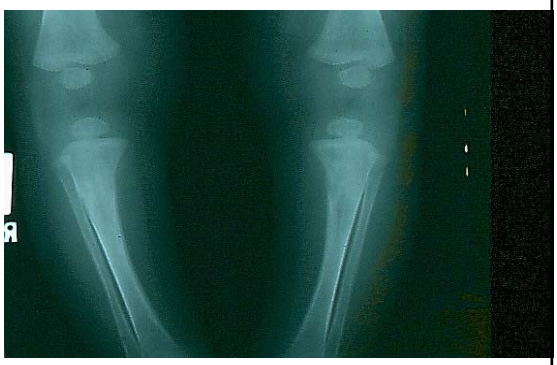

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Diagnosis – congenital syphilis

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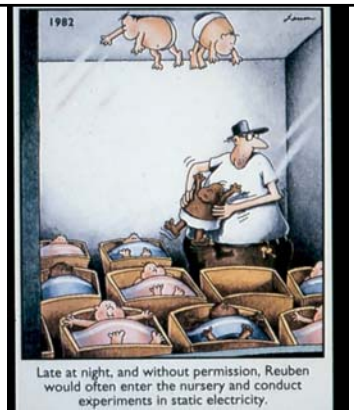

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DIFFERENTIAL DIAGNOSIS – NEONATES & INFANTS

- congenital infections
- musculoskeletal infection
- trauma
- immune deficiency
- systemic disease

8

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Musculoskeletal infections -neonate

- More difficult to diagnose
- May be no obvious joint swelling or fever
- Higher incidence of coexisting osteomyelitis and septic arthritis
- Pathogens are different (GBS, gram negative organisms, candida)
- Inflammatory markers may not be very elevated

9

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CASE 2 - MF

- 18 month girl presents with a swollen knee for 3 days. ESR 40, ANA 1:40, RF 1:10, radiographs normal, normal CBC, started on NSAID
- symptoms worsen over next 11 days, stops walking. ESR 120, H/H 9/30, platelet 800,000
- PMH: chickenpox 2 weeks prior to onset of knee swelling
- LABS: synovial WBC 79,800, gram stain negative

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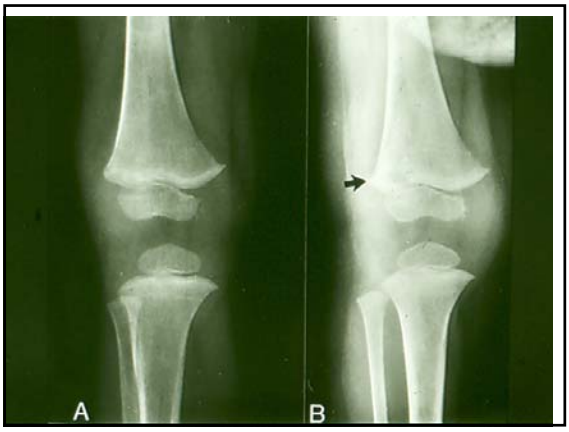

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Diagnosis - osteomyelitis

12

CASE 3 - TW

- 3 yo male referred for persistent symptoms following Kawasaki Disease.
- 2 months earlier - fever, vomiting, abdominal pain. 2 doses of amoxicillin, rash, red eyes, red lips, swollen hand. Admission blood culture + staph aureus, second culture is negative.

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CASE 3 - TW

- Receives IVIG, fever and rash defervesce. later, peeling of hands and thrombocytosis
- 2 months later - ESR 40-60's, child awakens at night with abd pain, refuses to jump, otherwise normal.
- Exam normal, blood culture negative, plain x-rays and abdominal ultrasound are normal, ESR 50.

14

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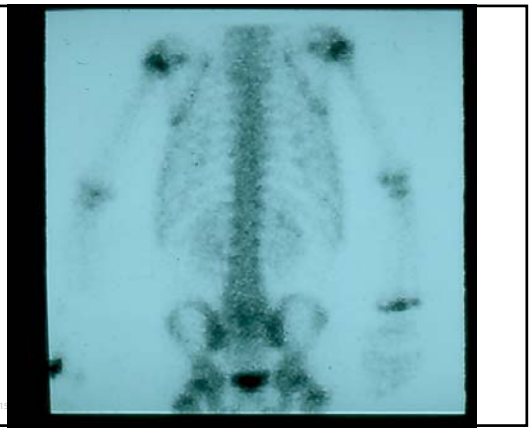

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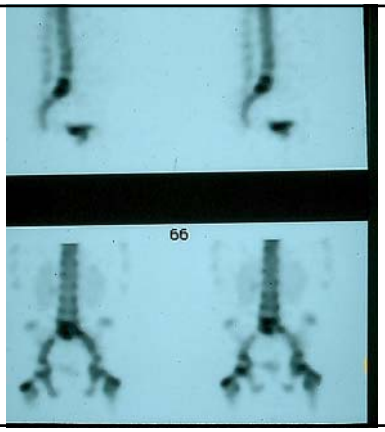

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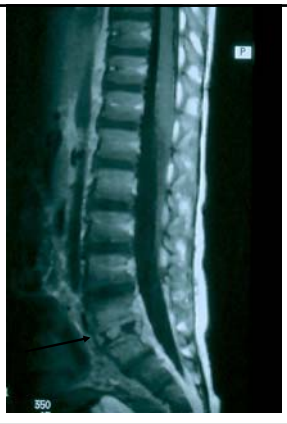

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Diagnosis – discitis due to staph aureus

18

TAKE HOME POINTS

- Staph aureus is never a contaminant
- The parent is usually right
- Meeting criteria for a specific disorder does not necessarily mean the patient has that disorder
- When in doubt – repeat basic studies, re-examine the patient, get further history, consider subspecialty consultation

19

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DIFFERENTIAL DIAGNOSIS – PRESCHOOL AGE

- musculoskeletal infection
- trauma - child abuse
- growing pains
- Kawasaki Disease
- Henoch Schonlein purpura
- Rheumatic disorder (JIA)
- immune deficiency
- systemic disease/malignancies

20

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Criteria for the diagnosis of growing pains (based on Naish & Apley, 1951)

- At least a 3-month history of pain
- Intermittent pain with symptom-free intervals of days, weeks, or moths
- Pain late in day or awakening child at night
- Pain not specifically related to joints
- Pain of significant severity to interrupt such normal activity as sleep
- Normal PE, labs & radiographs

21

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Findings suggesting benign limb pain

LIKELY BENIGN LIMB PAIN

- Shin pain
- Paroxysmal night time occurrence
- Episodic w asymptomatic periods
- Well otherwise
- Normal physical examination

UNLIKELY BENIGN PAIN

- Joint pain
- Daytime pain
- Constant/fluctuating
- Systemic symptoms
- Findings on physical examination

22

CASE 4 - AR

- 11 yo girl sees her physician for 2-3 weeks of fatigue, poor appetite, Hb 10.1, WBC 3.1, normal differential, ferritin 500
- Returns two weeks later with joint symptoms, swollen knees, wrists, hands, elbows, Hb 9.6, WBC 4.1, normal differential, plt 245,00, ESR 102, RF+16, CRP 8.5, neg monospot, ferritin 541

23

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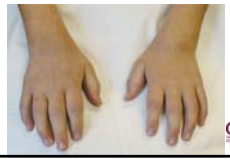
CASE 4 - AR

- Started on NSAID, joint pain and swelling persists, rib pain, and pain in right shin
- On exam, temp 99.6, polyarthritis 2-3+ swelling elbows, wrists, hands, knees, ankles



24

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CASE 4 - AR

QUESTIONS:

1. Diagnosis?
2. Further testing?
3. Management?

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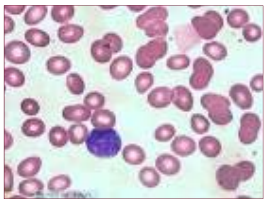
CASE 4 - AR

WBC 3.4, Hb 7.6, plt 158,000, retic 2.5%

Uric acid 3.4

LDH 474



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26

DIFFERENTIAL DIAGNOSIS – SCHOOL AGE CHILD

- musculoskeletal infection
- Trauma
- orthopedic conditions
  (Perthes, SCFE, patellofemoral syndrome)
- pain amplification syndromes
  (RSD, fibromyalgia)
- Rheumatic disorders
  (JIA, spondyloarthropathy, SLE, etc.)
- Malignancy, systemic disease, immune deficiency

27

27

Malignancy & musculoskel con

- <1% of children with MSK complaints have malignancy
- Most common malignancies – leukemia, lymphoma, Ewing sarcoma & neuroblastoma
- RED FLAGS:
  - pain disproportionate to exam, night pain, migratory
  - Arthritis – atypical onset & sites, no morning stiffness
  - Systemic symptoms – night sweats, fever, weight loss

28

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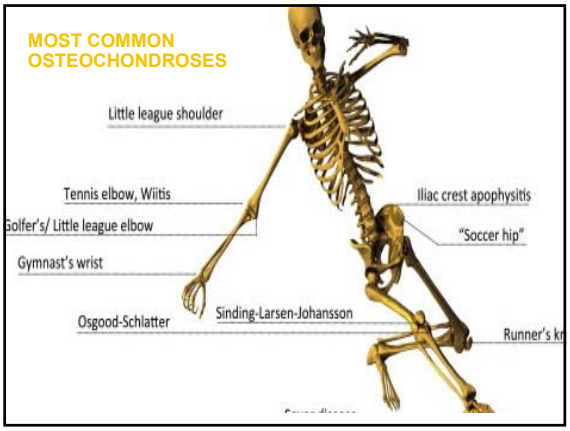

29

PEDIATRIC RHEUMATOLOGY

- JA (JRA, JIA JCA)
- SLE
- OTHERS (dermatomyositis, scleroderma, vasculitis)
- PAIN AMPLIFICATION SYNDROMES
- SPONDYLOARTHROPATHY
  - Reiter syndrome
  - psoriatic arthritis
  - Ankylosing spondylitis
  - IBS associated
  - undifferentiated spondy

30

30

Pain amplification syndromes – screening clues

- La belle indifference – incongruent affect
- High pain tolerance
- Enmeshment "our pain"
- Lack of sleep hygiene
- Allodynia

- 1/3 of school age children report pain lasting longer than 6 months, half have musculoskeletal pain
- Highest prevalence in young teen girls

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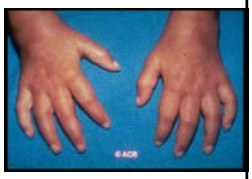
POLYARTICULAR JIA



- 40% of JA
- Older children
- F>M
- Half have rheumatoid factor
- Involvement of more than 4 joints
- Usually symmetric polyarthritis
- Worse prognosis

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OLIGOARTICULAR JIA

- 50% of JA
- Younger children
- F>M
- negative rheumatoid factor
- Involvement of fewer than 4 joints
- High chance of iritis
- Good prognosis
- 75% have +ANA

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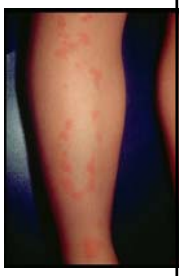
SYSTEMIC JIA



- 10% of JA
- Younger children
- F=M
- negative rheumatoid factor and ANA
- Usually polyarthritis
- High fever, rheumatoid rash
- Prognosis variable

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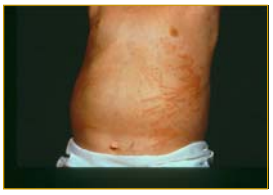

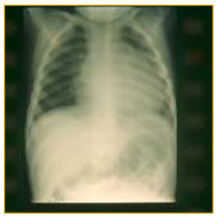

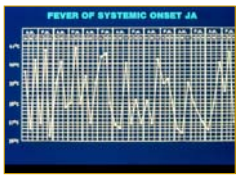

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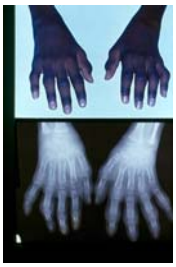
JA - complications



- Musculoskeletal
- joint destruction
- Atrophy
- Contractures
- bony overgrowth
- C-spine involvement - spinal cord compression

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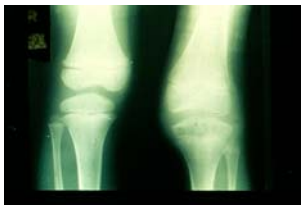
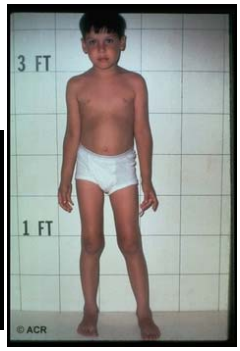
Bony overgrowth in JA





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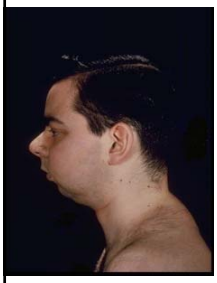
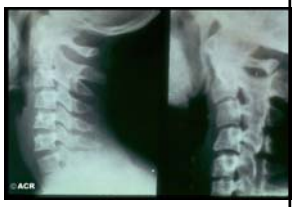
37

micrognathia & C-spine disease in JIA





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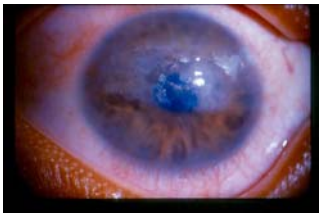
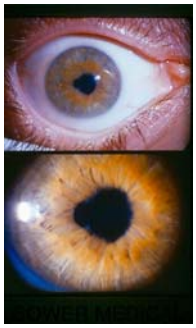
38

UVEITIS - risk of vision loss, catarct, glaucoma





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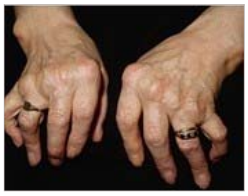

COMPLICATIONS OF JA
SEVERE DISABILITY
amyloidosis
SYSTEMIC JRA - anemia, infections, mortality 0.1%.

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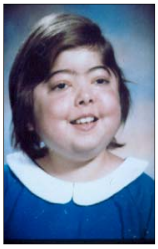
PREDNISONE / CORTICOSTEROIDS

3 RD GRADE

4 TH GRADE



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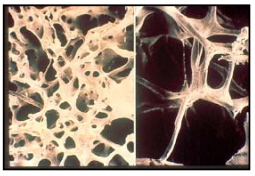
PREDNISONE / CORTICOSTEROIDS

5TH GRADE



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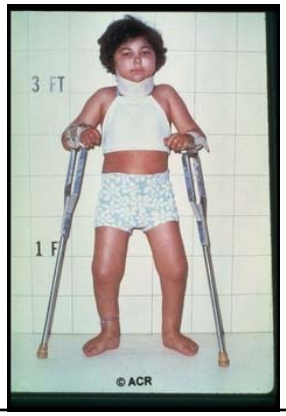

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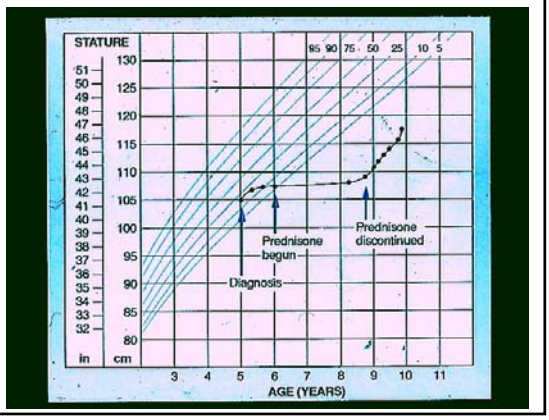

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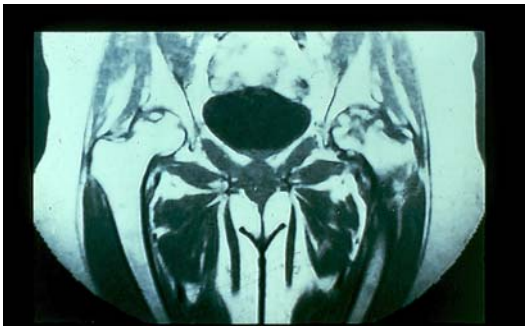

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Table 51–2. CONSIDERATIONS PRIOR TO THE USE OF GLUCOCORTICOIDS AS PHARMACOLOGIC AGENTS*

1. How serious is the underlying disorder?
2. How long will therapy be required?
3. What is the anticipated effective steroid dose?
4. Is the patient predisposed to any of the known hazards of glucocorticoid therapy?
   - Diabetes mellitus
   - Osteoporosis
   - Peptic ulcer, gastritis, or esophagitis
   - Tuberculosis or other chronic infections
   - Hypertension and cardiovascular disease
   - Psychological difficulties
5. Which glucocorticoid preparation should be employed?
6. Have other modes of therapy been utilized to minimize the glucocorticoid dosage and to minimize the side effects of glucocorticoid therapy?
7. Is an alternate-day regimen indicated?

*Modified from Thorn, G. W.: N. Engl. J. Med. 274:775, 1966.

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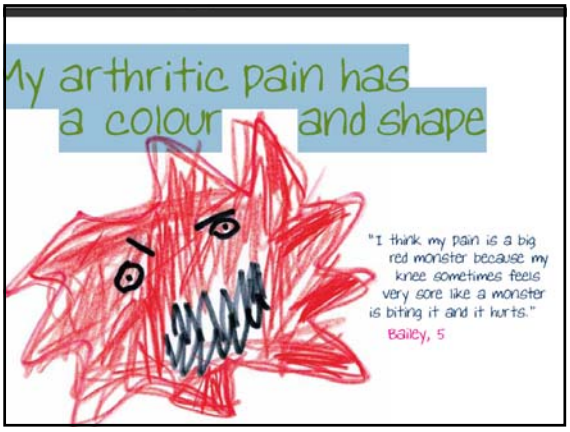

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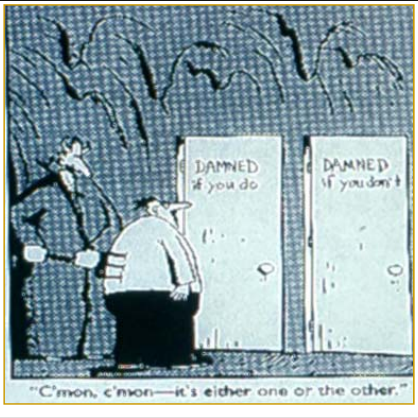

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QUESTIONS?