BOARD REVIEW
FRIDAY, MARCH 27, 2015
HYPERMOBILITY SYNDROME

Aka

“Benign Joint Hypermobility Syndrome (BJHS)”
HYPERMOBILITY....

Joint that exceeds the normal range of motion...excessive ligamentous laxity
Differential Diagnosis

• Hereditary Connective Tissue Disorders
  – Marfan Syndrome
  – Ehlers-Danlos syndrome
  – Osteogenesis imperfect
  – Stickler

• Inflammatory, Infectious, Neoplastic, Traumatic, Biomechanical
Question 4

An 8-year-old boy is brought to you for pain in the knees and legs. Which of the following favors the diagnosis of hypermobility?

A. A grade II/VI diastolic murmur is heard in the second left intercostal space

B. After evening soccer practices, the patient is awakened with pain at night

C. Episodes of joint pain are associated with low-grade fever

D. A moderate amount of joint effusion without tenderness is noted in both knee joints

E. Point tenderness is observed on the medial aspect of both knees

Understand the relationship between hypermobility and joint complaints.
Classic Presentation- Hypermobility Syndrome

- Episodic joint pain
  - Late afternoons, evenings
  - Often follows excessive exercise
- Most common joint involved **Knees**
  - Other commonly involved joints include: **Ankles, hips, back**
- Symmetric or asymmetric?
- May awaken patient from sleep
- +/- periarticular swelling

Know the physical findings in hypermobility syndrome
Associations- Hypermobility Syndrome

Commonly associated with:
  • Congenital hip disorders
  • Delayed motor development
  • Anterior Knee pain syndromes
  • Back pain
  • Joint dislocation/subluxation
  • Pes Planus
  • Chronic pain syndromes
    • Fibromyalgia
    • Chronic fatigue syndrome

???

early osteoarthritis
Question 19

Which of the following statements regarding hypermobility is TRUE?

A. Caucasian children exhibit greater joint mobility than those of African descent
B. Hypermobility is a significant risk factor for mitral valve prolapse
C. Joint laxity observed in early childhood usually diminishes during late childhood and early adolescence
D. Males have a greater degree of joint laxity compared with females
E. The presence of hypermobility is usually indicative of a serious underlying connective tissue disorder
A 10-year-old girl of normal stature presents with a complaint of pain in her knees after cheerleading practice. There is no history of easy bruising or other skin abnormalities. Physical examination of her knees reveals hypermobility but no articular swelling. Findings on the remainder of the physical examination are normal except for multiple hyperextensible joints.

Of the following, the MOST appropriate next step in the management of this patient is:

A. Collagen analysis
B. Echocardiography
C. Measurement of plasma amino acids
D. Measurement of rheumatoid factor
E. No further testing
<table>
<thead>
<tr>
<th>Table. Diagnostic Criteria for Hypermobility</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Original Carter and Wilkinson Criteria</strong></td>
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<tr>
<td>Require more than three of five in joint pairs:</td>
</tr>
<tr>
<td>- Passive apposition of the thumb to the flexor aspect of the forearm</td>
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<tr>
<td>- Passive hyperextension of the fingers so that they lie parallel with the forearm</td>
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<tr>
<td>- Passive hyperextension of the elbows $&gt;10^\circ$</td>
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<tr>
<td>- Passive hyperextension of the knees $&gt;10^\circ$</td>
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<tr>
<td>- Excessive passive dorsal flexion of ankle and excessive foot eversion</td>
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<tr>
<td><strong>Beighton Scale</strong></td>
</tr>
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<td>Require four or more of nine tests (one point for each right and left side where applicable); varies by study:</td>
</tr>
<tr>
<td>- Passive dorsiflexion of 5th metacarpophalangeal joint beyond $90^\circ$ with forearm flat on table</td>
</tr>
<tr>
<td>- Passive apposition of the thumb to the flexor aspect of the forearm</td>
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<td>- Passive hyperextension of the elbows $&gt;10^\circ$</td>
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<td>- Passive hyperextension of the knees $&gt;10^\circ$</td>
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<tr>
<td>- Forward flexion of trunk, with knees straight, so that palms rest easily on the floor</td>
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<tr>
<td><strong>Revised Beighton (Beighton 1998)</strong></td>
</tr>
<tr>
<td>Require two major criteria, one major and two minor criteria, or four minor criteria</td>
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<tr>
<td><strong>Major Criteria:</strong></td>
</tr>
<tr>
<td>- Beighton score of 4/9 or greater</td>
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<tr>
<td>- Arthralgia for $&gt;3$ months in four or more joints</td>
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<tr>
<td><strong>Minor Criteria:</strong></td>
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<tr>
<td>- Beighton score of 1, 2, or 3/9</td>
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<tr>
<td>- Arthralgia ($\geq 3$ months) in one to three joints or back pain ($\geq 3$ months), spondylolysis, spondylolysis/spondylolisthesis</td>
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<tr>
<td>- Dislocation/subluxation in more than one joint or in one joint on more than one occasion</td>
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<td>- Soft-tissue rheumatism with three or more lesions (e.g., epicondylitis, tenosynovitis, bursitis)</td>
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<td>- Marfanoid habitus</td>
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<tr>
<td>- Abnormal skin: striae, hyperextensibility, thin skin, papyraceous scarring</td>
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<tr>
<td>- Eye signs: drooping eyelids, myopia, antimongoloid slant</td>
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<tr>
<td>- Varicose veins or hernia or uterine/rectal prolapse</td>
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</table>

Exclude other possibilities!
**Course/Treatment- Hypermobility Syndrome**

**Course:**
- Benign!
- Self-Limited

**Treatment:**
- Reassure the parents and the patient
- Educate
- Encourage Fitness, Physical Therapy, Pain Control
- +/- supportive devices
- No splinting!
- +/- NSAIDS
- Address comorbidities

Know that the treatment of hypermobility syndrome is by explanation (ie- counsel the patient to avoid excessive movement)
ARTHRITIS IN CHILDREN AND ADOLESCENTS

"I want you to take one of these every day until I think of something else."
A 6-month-old female infant presents to you with fever to 102°F, poor feeding, and decreased activity for five days. Her mother has noted that over the last seven days, she cries whenever her diaper is changed, and for the last two days, she has refused to move her left leg. On physical examination, you note a febrile infant who cries with passive movement of the left leg. Needle aspiration of the left hip demonstrates purulent fluid. Of the following, the organism that is MOST likely responsible for this illness is:

A. *E. coli*
B. Group A *streptococcus*
C. *H. influenza* type B
D. *Staphylococcus aureus*
E. *Streptococcus pneumonia*

Recognize the major clinical syndromes associated with *Staphylococcus aureus* including: pyogenic arthritis.
SEPTIC ARTHRITIS (PYOGENIC ARTHRITIS)
Septic Arthritis - Just Facts!

- “Pyogenic Arthritis”
- Most commonly affects what age group? 2-3 year olds
- **Males** or Females?
- Must common pathogen implicated in pediatric septic arthritis (not including neonates): **Staphylococcus aureus**
  - Most common pathogens in neonates: **Streptococcus, Staphylococcus aureus, GBS**
- Most Commonly affected joint? **Knees**
- Other common pathogens?
  - *Streptooccus pyogenes, Streptococcus pneumoniae, Neisseria gonorrhea, Neisseria meningitides, Salmonella species, (historically- H. influenza type B)*
  - *Kingella kingae* (Know the most common infections associated with Kingella kingae (ie pyogenic arthritis, osteomyelitis))
  - *Mycobacterium tuberculosis*: (Know the major clinical manifestations of M. Tb-arthritis)
  - *Mycoplasma pneumonia* (Identify the extrapulmonary manifestations (arthritis) of Mycoplasma infection)
Question 15

The most common mechanism of infection in septic arthritis is

A. Contiguous spread
B. Hematogenous seeding
C. Instrumentation
D. Intra-articular injections
E. Trauma

Lab findings (i.e. what should you order)?
Imaging findings?
Recognize the value of the examination of joint aspirate to distinguish between juvenile rheumatoid (idiopathic) arthritis and septic arthritis.
A 2-year-old boy who has been limping and has a T: 39°C presents with a swollen and warm left knee that is exquisitely tender with any movement. No other findings are of note on his physical examination. Magnetic resonance imaging of the knee reveals a large joint effusion but no bony or soft tissue involvement. Arthrotomy is performed for irrigation and debridement of the knee joint. The joint fluid contain 100 x 10³/mcL WBC’s with 80% polymorphonuclear leukocytes and 20% lymphocytes but the culture is negative after three days. The patient is not improving with intravenous vancomycin therapy.

Of the following, the next BEST step is to:

A. Add linezolid to the therapy
B. Add trimethoprim to the therapy
C. Change the therapy to cefotaxime
D. Change the therapy to Clindamycin
E. Continue vancomycin

Treatment:
- Empiric antibiotics
- Duration? 2-6 weeks
Question 9

Which of the following findings commonly occurs in both septic arthritis and toxic synovitis?

A. Effusion of a hip joint
B. Marked elevation of erythrocyte sedimentation rate
C. High fever
D. Inability to bear weight
E. White blood cell count higher than 12,000 /µL
TOXIC SYNOVITIS
Toxic Synovitis

• Most commonly affects: The hip.
• Most commonly affects what age group? 3-10 year olds.
• Males or Females?
• Setting? Prodromal or concurrent URI/AGE
Toxic Synovitis

Which of the following findings commonly occurs in both septic arthritis and toxic synovitis?

A. Effusion of a hip joint
B. Marked elevation of erythrocyte sedimentation rate
C. High fever
D. Inability to bear weight
E. White blood cell count higher than 12,000 /µL

Septic Arthritis

- Fever
- Inability to bear weight
- ESR > 40 mm/hr
- WBC > 12,000/mm³
- Constant Pain
- Synovial fluid: >50,000 WBC

Toxic Synovitis

- Absent or low-grade fevers
- Often limp, but can bear weight
- Normal to mildly elevated ESR
- Normal to mildly elevated WBC
- Bouts of improvement in pain
- Synovial fluid: 5,000-15,000 WBC
Toxic Synovitis- Treatment/Course

Treatment:
- Anti-inflammatory medications
- Limitation of activity

Course:
- Resolution of symptoms in days-2 weeks
- Self limited

Sequelae:
- Slightly higher risk of developing Legg-Calve-Perthes
Question 13

Which of the following is NOT a major manifestation of the Jones criteria for the diagnosis of acute rheumatic fever?

A. Arthralgia
B. Carditis
C. Erythema marginatum
D. Subcutaneous nodules
E. Chorea
ACUTE RHEUMATIC FEVER
Question 13

Which of the following is NOT a major manifestation of the Jones criteria for the diagnosis of acute rheumatic fever?

A. Arthralgia
B. Carditis
C. Erythema marginatum
D. Subcutaneous nodules
E. Chorea

**Major Criteria**
J - Joints (arthritis)
♡ - Carditis
N - Nodules (subcutaneous)
E - Erythema marginatum
S - Sydenham chorea
Table 3.

Jones Criteria for the Diagnosis of Acute Rheumatic Fever

<table>
<thead>
<tr>
<th>Major Criteria</th>
<th>Minor Criteria</th>
<th>Evidence of Recent GAS Infection</th>
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</thead>
<tbody>
<tr>
<td>Carditis</td>
<td>Fever</td>
<td>Positive throat culture or RADT</td>
</tr>
<tr>
<td>Polyaarthritis</td>
<td>Arthralgia</td>
<td>OR</td>
</tr>
<tr>
<td>Chorea</td>
<td>Elevated acute-phase reactants</td>
<td>Elevated or rising antistreptococcal antibody titers</td>
</tr>
<tr>
<td>Erythema marginatum</td>
<td>Prolonged PR interval</td>
<td></td>
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<tr>
<td>Subcutaneous nodules</td>
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</tbody>
</table>
A 10-year-old girl had recent pharyngitis with culture-proven group A *Streptococcus*. She was noncompliant with antibiotic therapy. She now presents with a fever to 102°F, a HR of 120 beats per minute, and a respiratory rate of 24 breaths per minute. She has no murmurs or gallop rhythm. She has a nonpruritic macular rash that appears as a serpiginous, erythematous circle surrounding normal skin. She also has an erythematous warm, swollen left knee and right ankle.

Of the following, the MOST appropriate diagnostic study for this patient is:

A. Chest radiography
B. Echocardiography
C. Left knee and right ankle radiography
D. Rheumatoid factor
E. Skin biopsy of the rash

Recognize the complications of post-streptococcal infection: rheumatic fever
A 12-year-old girl presents to your office for the first time with a swollen painful, erythematous right knee joint. She tells you that her left knee felt and looked similar yesterday but now feels normal. She also is easily fatigued and has had fever. On physical examination, she has a T 101.7°F, a HR of 125 beats per minute, a respiratory rate of 24 breaths per minute, and a blood pressure of 120/78 mm Hg. Her lungs are clear. On auscultation, you note a III/VI holosystolic murmur at the cardiac apex with radiation to the axilla. Of the following, the best plan for management of this patient’s joint swelling includes:

A. Antibiotic therapy with doxycycline
B. Anti-inflammatory therapy with aspirin
C. Aspiration of the right knee joint
D. Heat, elevation, and splinting of the right knee
E. Immunotherapy with azathioprine
Acute rheumatic fever

Plan the treatment of group A Streptococcal infection

Single dose of IM benzathine penicillin G
10 days of oral penicillin

Other treatment considerations:
 Anti-inflammatory
 Restriction of activity
 Antibiotic prophylaxis
Several weeks ago, a boy presented with fever, malaise, headache, and a skin lesion. The lesion began as a red papule and slowly enlarged to form a large annular ring with a flat, erythematous border. Today, he complains of recurrent joint pains that are particularly severe in his knees. Of the following, the MOST likely diagnosis is:

A. Epstein-Barr virus
B. Human parvovirus B19 infection
C. Lyme disease
D. Juvenile rheumatoid arthritis
E. Rocky Mountain spotted fever

Know that Lyme disease causes fever and arthritis that may be confused with JRA.
LYME DISEASE
Why are we talking about Lyme disease?

Bug?  *Borrelia burgdorferi*

Symptoms:  Rash- erythema chronicum migrans  
Fever  
Fatigue with myalgias  
Mono/oligoarthritis

Diagnose it?  Serologies*

Treat it?  Doxycycline OR Amoxicillin  
If arthritis present, plan for 28 days of antibiotics
A 16-year-old boy presents with a very swollen, painful right knee. He is a soccer player, but there is no history of recent injury. During the interview, you notice the boy has injected conjunctiva.

Of the following, further evaluation MOST likely will reveal:

A. Alopecia areata
B. Gottron’s papules
C. Kayser-Fleischer rings
D. A malar rash
E. Urethritis

What does he have?  
Reiter’s syndrome

Triad?
Conjunctivitis, arthritis, urethritis

Recognize a patient with post-infectious arthritis
REACTIVE ARTHRITIS

AKA

“Post infectious arthritis”
Reactive Arthritis

Arthritis associated with infection at a distant site

Identify the common illnesses associated with post-infectious arthritis

- *Chlamydia trachomatis*
- *Neisseria gonorrhea*
- *Shigella*
- *Salmonella*
- *Yersinia*
- *Campylobacter*
- *Streptococcus pyogenes*
- *Neisseria meningitidis*

HLA association?  HLA-B27

Clinical manifestations?

- Large joint arthritis, dactylitis, enthesitis, photophobia, skin changes, fever, weight loss,
Question 23

An 18-year-old boy has a swollen right wrist and left ankle with swelling over both Achilles tendons. He was treated 2 weeks ago with an antibiotic for a urethral discharge. His girlfriend also was treated. Physical examination reveals tenderness over both Achilles tendons, swollen painful joints (right wrist and left ankle), and limited forward bending at the waist.

The MOST appropriate therapeutic agent would be:

A. Ceftriaxone
B. Doxycycline
C. Prednisone
D. A nonsteroidal anti-inflammatory agent
E. Intravenous immunoglobulin

Plan the management of a patient with post-infectious arthritis
JUVENILE IDIOPATHIC ARTHRITIS...