Board Review
Juvenile Idiopathic Arthritis
A 4-year-old white girl has had joint swelling in multiple joints for over 6 months. She is slow to move in the morning and moves as if stiff for the 1st hour of the day. Thereafter, she is a very active child. She has no rash and very little limitation of range of motion. Her erythrocyte sedimentation rate (ESR) is 4 mm/hr. The most likely diagnosis is:

A. Hypermobility syndrome
B. Dermatomyositis
C. Systemic lupus erythematosus
D. Juvenile idiopathic arthritis
E. Henoch Schlein Purpura
JIA at a Glance

- Broad term for several forms of chronic arthritis
- Most common rheumatologic disease in children
- Understand that juvenile idiopathic rheumatoid arthritis is often a disease of exclusion, and know the differential diagnosis.
  - What must you rule out?

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<th>Reactive</th>
<th>Poststreptococcal Rheumatic fever Serum sickness “Reiter syndrome”</th>
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<td>Inflammatory</td>
<td>Juvenile idiopathic arthritis Inflammatory bowel disease Sarcoidosis</td>
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<td>Septic joint Postinfectious: toxic synovitis</td>
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<td>Immunodeficiency</td>
<td>Common variable immunodeficiency</td>
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<td>Trauma</td>
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JIA at a glance

• Intra-articular swelling or the presence of 2 or more of: limitation in range of motion, tenderness or pain on motion, increased heat or erythema.
  – Must be present to make diagnosis of JIA!!!

• Initial symptoms may be subtle or acute, often include morning stiffness with a limp or gelling after inactivity.

• Inflammation may cause elevations in ESR and CRP → it is not unusual for both to be normal.
You are evaluating a 10-year-old girl for joint pain that has been present for approximately 2 months. She has no fever but complains of pain and swelling in her hands and feet, which is worse in the morning. On physical examination, she has evidence of symmetric swelling of all proximal interphalangeal joints in her hands and feet and pain over her temporomandibular joint. The remainder of the examination is normal. Which of the following is the most likely diagnosis?

A. Enthesitis-related arthritis
B. Oligoarticular juvenile idiopathic arthritis
C. Polyarticular juvenile idiopathic arthritis
D. Psoriatic arthritis
E. Systemic-onset juvenile idiopathic arthritis

Question #12
Recognize the major presentations of juvenile rheumatoid (idiopathic) arthritis
JIA Types

• 50% of cases of chronic arthritis in children!

Identify the typical patient with pauciarticular (oligoarticular) juvenile rheumatoid arthritis –
- Well appearing, typically affects lower extremities, may present with limp, joint is warm and swollen but not very painful/tender

- Small joint involvement of both hands and feet, cervical spine and temporomandibular joints
- Rheumatoid nodules and joint deformities
- Must rule out Lupus

Recognize Lyme disease as a mimic of (oligo-) juvenile rheumatoid arthritis

- Early childhood; peak at 2–4 years
- F >> > M
- Recognize Lyme disease as a mimic of juvenile rheumatoid arthritis
- 3:1

- 5:1

- Small joint involvement of both hands and feet, cervical spine and temporomandibular joints
- Rheumatoid nodules and joint deformities
- Must rule out Lupus
Mary Jones has a recent diagnosis of juvenile rheumatoid arthritis. She is 13-years-old and has multiple small and large joints affected by her arthritis. She has been diagnosed as having polyarticular JRA. Her rheumatoid factor (RF) is pending.

Which of the following is true regarding her disease?

A. If the RF is negative, she will likely have more aggressive disease
B. If the RF is positive, she will likely have more aggressive disease
C. She is likely to have a negative ANA if her RF is positive
D. She is likely to have a negative ANA if her RF is negative
E. If the RF is negative, this usually indicates an HLA association
JIA RF

- **Recognize that rheumatoid factor is usually negative in juvenile rheumatoid (idiopathic) arthritis**
  - RF is a component of polyarticular arthritis
  - 5-10% of patients
  - Typically seen in females in late childhood or early adolescence
  - Seropositive patients have similar presentations to adults with RA
  - Associated with a more aggressive course
JIA Types

- Peak onset 1-5 years of age

- **Identify the patient with systemic juvenile rheumatoid (idiopathic) arthritis**
  - *Serositis, lymphadenopathy, increased leukocyte count, and anemia, thrombocytosis, elevated LFTs/APRs, ANA titer usually negative*

- Distinct from other types due to high-spiking fevers of at least 2 week’s duration
  - Fever is Qday or BID (late afternoon/evening), rapid return to baseline

- Salmon-colored macular rash with febrile periods, nonpruritic
Systemic JIA

- **Distinguish between juvenile rheumatoid (idiopathic) arthritis and rheumatic fever**
  
  **Acute Rheumatic Fever**
  
  Fever: ___**Persistent**___
  
  Arthritis: ___**Migratory and asymmetric**___
  
  Cardiac involvement: ___**Often associated with endocarditis**___
  
  Rash: ___**Erythema Marginatum** → **associated with expanding margins**
• large and small joints

Boys >8 yoa (M:F 7:1)

• Inflammation at sites of tendon insertions onto bone

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Figure 5. Swelling of left third proximal phalangeal joint with “sausage” appearance of finger in a patient with psoriatic arthritis. (Courtesy of Charles H. Spencer [http://www.rheumatlas.org].)
A 7-year-old girl has developed a limp and complains of pain in her right knee, which is warm and swollen. Although she is afebrile in the office, her parents say she had a fever at home. You suspect oligoarticular JIA but have concerns about infection. Which of the following tests would give you a definitive answer?

A. ANA
B. Erythrocyte sedimentation rate
C. Rheumatoid factor
D. Synovial fluid analysis
E. White blood cell count

Recognize the value of the examination of joint aspirate to distinguish between juvenile rheumatoid (idiopathic) arthritis and septic arthritis
Which of the following medications used in the treatment of rheumatic disorders is NOT disease modifying?

A. Celecoxib
B. Cyclosporin
C. Methotrexate
D. Sulfasalazine
E. Azathioprine
JIA Treatment

• **Recognize the need for a comprehensive program (including physical therapy) for the management of juvenile rheumatoid (idiopathic) arthritis**

• **Understand the pharmacologic treatment of juvenile rheumatoid (idiopathic) arthritis**
  – First line: NSAIDs (ibuprofen, naproxen, indomethacin)
    • Decrease both pain and acute/chronic inflammation
  – Disease modifying agents: required in 2/3 of patients
    • Most commonly used?
    • Methotrexate- requires 6-12 weeks to see effects; major side effects are GI \( \rightarrow \) decreased if given folic acid
    • Entanercept/infliximab/adalimumab block TNF-a
  – Steroids for systemic manifestations

• **Recognize the complications of nonsteroidal anti-inflammatory drug therapy in juvenile rheumatoid (idiopathic) arthritis: bleeding, gastritis, abnormal liver function test results, encephalopathy, Reye-like syndrome**
  – What would you use in a patient with arthritis associated with IBD?
    • COX 2 inhibitors
You are evaluating a 13-year-old boy who has had intermittent low-back pain and stiffness for the past 3 months. He also has some ankle pain but no fever or other complaints. He plays soccer but does not recall any particular injuries. On physical examination, you note some tenderness of his sacroiliac joints. He has normal lower extremity strength, sensation, and reflexes and negative results on a straight leg raising test. There is moderate tenderness to palpation over the insertion of his Achilles tendon. He has no skin or eye findings and no other joint findings. Spinal radiographs are read as normal.

Of the following, the BEST next step is
A. Acetaminophen use as needed
B. Corticosteroid injection in the Achilles tendon
C. Evaluation for spondyloarthropathies
D. Heat to his back and bedrest
E. Screening for sexually transmitted infection

Question #16
The spondyloarthritides, also referred to as enthesitis-related arthritides, include all of the following except:

- Rheumatoid factor-positive juvenile idiopathic arthritis
- Ankylosing spondylitis
- Arthritis with inflammatory bowel disease
- Reactive arthritis secondary to diarrhea
- Reactive arthritis secondary to genitourinary infection
Enthesitis-related (10% of JIA)

• Collectively referred to as spondyloarthropathies
  – Ankylosing spondylitis, arthritis associated with IBD and psoriasis, reactive arthritis following GI or GU infections.
• Enthesitis-mechanical stress or infl reaction resulting in fibrosis or calcification
• Are the most common causes of chronic low back pain in adolescents
  – Spondylolysis (defect of pars interarticularis)
  – Spondylolisthesis (forward slippage of one vertebra over another, L5 on S1)
  – Ballet, gymnastics, wrestling require hyperextension of spine
• Asymptomatic or associated with pain that can be mild or severe/disabling
  – Lower limbs
  – May involve synovial, cartilaginous or fibrosis, or extra-articular areas
Enthesitis-related: Diagnostics

- Laboratory evidence of systemic inflammation with elevation of ESR/CRP is variable
  - May not be present at onset
- RF and ANA are negative except in those with psoriatic arthritis
  - Of which as many as 50% are ANA positive
- HLA-B27 (+) >90% of kids with AS, 7% in healthy individuals
- MRI is most common modality but is nonspecific
  - Bone changes may be a late finding
- US with Doppler: high sensitivity and specificity for diagnosing spondyloarthritis
Enthesitis-related: Treatment

- Eyeball exam!!!
- Pain is most likely to respond to NSAIDs
- Modification of activity rather than bed rest - pain is often worse with rest and any decrease in activity
- Corticosteroids may increase the risk of rupture
- Reiter syndrome: sacroiliitis, arthritis, conjunctivitis, urethritis, and skin rash (keratoderma blenorrhagica) associated with HLA-B27, sometimes results from CZ

Of the following, the BEST next step is
A. Acetaminophen use as needed
B. Corticosteroid injection in the Achilles tendon
C. Evaluation for spondyloarthropathies
D. Heat to his back and bedrest
E. Screening for sexually transmitted infection
Which of the following statements regarding JIA is true?

A. African American children more often have systemic-onset JIA than other subtypes

B. Associated with HAL-B27 positivity is typical in enthesitis-related arthritis

C. Oligoarthritis occurs most commonly in adolescents

D. Polyarthritis occurs more commonly in male subjects

E. Psoriatic arthritis is not associated with ophthalmologic disease

A. AA typically present at older age and have higher rate of RF+ disease

C. But, we already said it occurs in those 2-4 years of age

D. But we already know that females predominate in all types, except in systemic JIA, where they are equal

E. But it is!
You treat a 15-year-old girl in your practice who has juvenile idiopathic arthritis. She is brought in by her mother today with complaints of a low-grade fever and diffuse pain. On physical examination, she has a T 38 and a HR of 100 bpm. As she sits on the examination table, she leans forward. During auscultation of her lungs, she complains of pain with deep inspiration.

Of the following, the MOST likely explanation for her symptoms is:

A. Costochondritis
B. Gastritis
C. Pericarditis
D. Pneumonia
E. Pulmonary embolism

Question #3
JIA Complications

• **Identify a patient with cardiac complications of systemic juvenile rheumatoid (idiopathic) arthritis**
  – Concerning chest pain is usually substernal, positional, severe if pericardial

• Without aggressive treatment, JIA may result in significant morbidity such as leg-length discrepancy, joint contractures, permanent joint destruction, or blindness from chronic uveitis
  – If knees: accelerated growth of the affected leg
  – If ankles/feet or wrists/hands: local growth retardation in affected joint
All of the following juvenile idiopathic arthritis subtypes have a moderate to high risk for the development of and require screening for iritis EXCEPT:

★ A. Systemic juvenile idiopathic arthritis
B. Rheumatoid factor-positive, polyarticular juvenile idiopathic arthritis
C. Rheumatoid factor-negative, polyarticular juvenile idiopathic arthritis
D. Enthesitis-related arthritis
E. Oligoarticular juvenile idiopathic arthritis
JIA Complications

- **Know the ocular complications of juvenile rheumatoid (idiopathic) arthritis**

- **Identify ANA as a marker for eye disease in juvenile rheumatoid (idiopathic) arthritis**
TODAY’S GRAND TOTAL: 29!