Update on Juvenile Idiopathic Arthritis (JIA)

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Objectives

• Know the definition of arthritis
• Recognize the subtypes of juvenile idiopathic arthritis (JIA)
• Know when to order and how to interpret common rheumatologic labs in a child with joint pain
Etiology of Joint Complaints in Childhood

- Infection
- Post-infectious/Reactive
- Orthopedic/Trauma
- Rheumatic diseases
- Psychogenic/Behavioral
- Hematologic
- Genetic/metabolic
- Malignancy
- Immunodeficiency
**Arthritis: Definition**

- **Objective signs** of inflammation within the joint
  - Swelling or an effusion
  - OR
  - Limitation in range of motion plus:
    - Tenderness to palpation (small joints) OR
    - Pain with movement

Inflammatory Arthritis

- Pain (Arthralgia) ≠ Arthritis

- Arthritis is:
  - Inflammation of the joints.
  - Characterized by swelling, warmth, redness, stiffness, decreased range of motion, pain.
  - Multiple causes: infection, trauma, systemic autoimmune disease, idiopathic
Joint Complaints: History

• Is the complaint "mechanical" or "inflammatory"?
  – Mechanical: no AM stiffness, pain with activity, improves with rest, no or minimal swelling
  – Inflammatory: AM stiffness, pain improves with activity, associated with swelling, warmth

Arthritis!
Juvenile Idiopathic Arthritis (JIA)

- Heterogeneous group of inflammatory disorders
- Characterized by chronic arthritis
- Clinical diagnosis
Definition of JIA

1. Arthritis in one or more joints for at least 6 weeks
   • Swelling or effusion
   • Limited Range of Motion plus either:
     • Tenderness to palpation    OR
     • Pain with Range of Motion

2. Age of onset <16 years

3. Diagnosis of exclusion- other identifiable causes have been ruled out

Symptoms of JIA

• Indolent onset
• Stiffness, especially in AM or after a sedentary period
  – Limping or reluctance to walk
  – Difficulty with buttons, snaps, zippers, opening jars and bottles, writing
  – Improves as day progresses or with activity
• Discomfort without severe or acute pain
• Swelling (not always noticed)
Symptoms of JIA
McGhee et al. Pediatrics 2002

• Isolated MSK pain without other signs or symptoms is almost never the presenting complaint of chronic arthritis in kids

• JIA usually presents as joint swelling or limp

• Children referred with +ANA and/or RF were no more likely to have a chronic inflammatory disease than those who did not include such results as a reason for referral.
When do I draw antibodies to diagnose arthritis?

NEVER - Arthritis is a clinical diagnosis!

Rheumatoid Factor (RF):

- Negative in 95% of children with arthritis.
- Positive in many conditions including infections, lupus, liver disease, and malignancy.
- Most common cause of + RF in children is viral infection.
When do I draw antibodies to diagnose arthritis?

Antinuclear Antibody (ANA):

– Does NOT help with diagnosis of JIA, only to determine risk of uveitis
– Sensitive for SLE only
– Positive in liver disease, infections, malignancy, and well children
– Only 5% of +ANA tests are due to lupus – many are in healthy people
# History of JIA Classification

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<tr>
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JIA Subtypes

Oligoarticular (<5 joints), about 50%
  • Persistent or Extended

Polyarticular (≥5 joints)
  • RF negative or positive

Psoriatic

Enthesitis Related

Systemic

Undifferentiated
Oligoarticular JIA

• Most common form
• Girls: Boys 3-5:1, peak age 1-4 years
• Northern European
• Most common presentation: single swollen knee
  – Limp, worse in AM or after nap
  – May not complain of any pain
  – Otherwise well-appearing
Complications

- Flexion Contracture
- Leg Length Discrepancy
- Atrophy
Uveitis and JIA

- Usually asymptomatic until damage occurs
  - Chronic in most subtypes
- Risk factors: younger age, +ANA
- Up to 20% of oligoarticular JIA
- Complications: cataracts, glaucoma, blindness
Polyarticular JIA

• Girls: Boys 3:1

• 2 age peaks:
  – Age 1-3: RF negative
  – Later childhood/ adolescence: RF positive

• Symmetrical, small joints, C-spine, TMJ disease

• +/- mild systemic disease: anemia, fatigue, ↑ ESR

• More aggressive, requires more medication
Spondyloarthropathies

- Enthesitis Related Arthritis
- Psoriatic Arthritis
- Reactive Arthritis
- IBD Associated Arthritis
- Undifferentiated Spondyloarthropathy
JIA Subtypes

Oligoarticular (<5 joints), about 50%
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Psoriatic

Enthesitis Related

Systemic

Undifferentiated
Common Features

- Peripheral (esp. knees, hips and ankles), sacroiliac and spine involvement potentially
  - Enthesopathy is common (Achilles, patellar)
  - “Sausage” digits or Dactylitis
- Older age, Boys > Girls
- Acute anterior uveitis
- HLA B27-related
Inflammatory Eye Disease

• Conjunctivitis
  – Transient

• Anterior Uveitis
  – **Acute** onset, unilateral\(\rightarrow\)bilateral, painful, red eye
  – Can cause permanent vision loss if not treated
  – Does not always parallel arthritis activity
Psoriatic Arthritis

• Often precedes the onset of psoriasis in children

• Clues to diagnosis:
  – 1\textsuperscript{st} degree relative with psoriasis
  – Nail pitting
  – DIP involvement (33%)
  – Dactylitis (sausage digit)

• Small joints hands/feet

• Aggressive, erosive disease
Psoriatic Arthritis (PsA)

• Plaque psoriasis (psoriasis vulgaris) is most common skin phenotype in PsA

• 15% develop psoriasis AFTER onset of arthritis
  – Often within 2 years in kids
  – Some pts may be unaware they have it
    • Scalp, behind ears, navel, nail changes

• Little relationship between skin severity and arthritis severity
Enthesitis-Related Arthritis

- Male > Female
- Strong association with HLA-B27
  - Arthritis and enthesitis
  - OR:
  - Arthritis or enthesitis with 2 of the following:
    - SI joint tenderness, +HLA-B27, arthritis in a boy 6 years of age or older, acute anterior uveitis, or 1st degree relative with spondyloarthritis
- Peripheral → axial disease
Systemic Onset JIA: An autoinflammatory disease

Clinical Manifestations:

• Arthritis (may be absent at onset)
• High grade spiking fever
• Evanescent rash
• +/- Adenopathy
• +/- Hepatosplenomegaly
• +/- Pleural/pericardial effusion
Differential Dx of SOJIA

• Primarily a diagnosis of exclusion
• Infection
• Inflammatory bowel disease
• Malignancy
• Systemic Rheumatic disease – lupus, dermatomyositis
• Acute Rheumatic Fever
• Kawasaki’s disease
SOJIA Prognosis

• Past:
  – steroid dependant $\rightarrow$ toxicity/growth failure

• Current:
  – More targeted therapy: IL-1 and IL-6 blockade
Macrophage Activation Syndrome (MAS)

A life-threatening complication of SOJIA

• Excessive/Unregulated expansion of Macrophages/ T cells produces a cytokine storm.

• Increased recruitment and activation of macrophages leads to tissue damage and organ failure
MAS

- Acutely ill with purpura, mucosal bleeding
- Sustained fever
- Hepatosplenomegaly, adenopathy
- CNS dysfunction: Lethargy with irritability, disorientation, headache, seizures, coma
- Liver dysfunction: AST, ALT, PT, PTT elevated
- Pancytopenia
- Low ESR due to low fibrinogen and liver dysfunction.
JIA Treatment

• Goals of treatment: control inflammation to prevent damage:
  – erosions, contractures, deformities.
• Once there is damage we have little power to reverse it.
JIA Treatment

Medications

- NSAIDS for mild disease
- Systemic Steroids: try to avoid, do not affect long term outcome
- Intra-articular steroids
- Disease modifying agents (DMARDS): methotrexate
- Biologics: target specific inflammatory mediators
  - TNF alpha, IL-1, IL-6, Tcell costimulatory molecules, others
Changes to consider in practice

- A careful history and exam trump labs in diagnosing arthritis.
- ANA and RF are *prognostic*, not *diagnostic*, tests in JIA.
- Avoid starting systemic steroids if you do not know the diagnosis.
References


• Rheumatology Image Library. American College of Rheumatology
