Juvenile Arthritis

Sarah Merrill, MD

ACTIVITY DISCLAIMER

The material presented here is being made available by the American Academy of Family Physicians for educational purposes only. Please note that medical information is constantly changing; the information contained in this activity was accurate at the time of publication. This material is not intended to represent the only, nor necessarily best, methods or procedures appropriate for the medical situations discussed. Rather, it is intended to present an approach, view, statement, or opinion of the faculty, which may be helpful to others who face similar situations.

The AAFP disclaims any and all liability for injury or other damages resulting to any individual using this material and for all claims that might arise out of the use of the techniques demonstrated therein by such individuals, whether these claims shall be asserted by a physician or any other person. Physicians may care to check specific details such as drug doses and contraindications, etc., in standard sources prior to clinical application. This material might contain recommendations/guidelines developed by other organizations. Please note that although these guidelines might be included, this does not necessarily imply the endorsement by the AAFP.
DISCLOSURE

It is the policy of the AAFP that all individuals in a position to control content disclose any relationships with commercial interests upon nomination/invitation of participation. Disclosure documents are reviewed for potential conflict of interest (COI), and if identified, conflicts are resolved prior to confirmation of participation. Only those participants who had no conflict of interest or who agreed to an identified resolution process prior to their participation were involved in this CME activity.

All individuals in a position to control content for this session have indicated they have no relevant financial relationships to disclose.

The content of my material/presentation in this CME activity will not include discussion of unapproved or investigational uses of products or devices.

Sarah Merrill, MD

Assistant Professor, Department of Family Medicine and Public Health, University of California, San Diego School of Medicine

Dr. Merrill is a board-certified family physician who specializes in sports medicine. In addition to diagnosing and treating injuries associated with athletics, she provides primary care for patients of all ages, including preventive care and treatment of acute and chronic diseases. An avid yoga practitioner and certified yoga instructor, she has a special interest in rehabilitating individuals who have yoga injuries. She also enjoys performing ultrasound-guided diagnostics and procedures. Recently, she published two chapters in the clinical reference text The 5-Minute Sports Medicine Consult, 3rd Edition.

Dr. Merrill instructs students, residents, and fellows at UC San Diego School of Medicine, is an assistant program director for the UCSD Family Medicine Residency Program, and is the medical director of UC San Diego Health’s Scripps Ranch Family Medicine Center. She also provides care throughout the San Diego community and for many sporting events, serving as the team physician for the University City High School and Scripps Ranch High School football teams; medical director for the California State Games; medical team captain for San Diego Rock ‘n’ Roll Marathon; and event physician for the BMX World Championships. Dr. Merrill completed a sports medicine fellowship at UC San Diego School of Medicine, where she also completed a residency in family medicine with an additional certification in integrative medicine. She earned her medical degree from Loyola University Chicago Stritch School of Medicine in Illinois. She is a member of numerous professional associations, including the American Medical Society for Sports Medicine (AMSSM), the American Medical Association (AMA), and the AAFP.
Learning Objectives

1. Describe diagnostic principles, appropriate pre-referral evaluation, and red flags in pediatric rheumatologic diseases.

2. Discuss treatment options, including side effects of common medications and what contraindications to immunizations exist for patients in active treatment.

3. Define the subtypes of Juvenile Idiopathic Arthritis (formerly Juvenile Rheumatoid Arthritis) and identify common signs and symptoms of each subtype.

Audience Engagement System

Step 1

Step 2

Step 3
Juvenile Arthritis: Definition

- Variety of autoimmune and inflammatory diseases that affect the joints of children 18 years and younger\(^3\)

Pathogenesis

- Genetic and environmental factors\(^3, 4\)
- Stress
- Trauma
- Gut microbiome
- History of infections\(^4\)
Juvenile Idiopathic Arthritis

- “Group of inflammatory disorders that begins before the 18th birthday and persists for at least 6 weeks with other known conditions excluded”

Juvenile Idiopathic Arthritis: Categories

- Systemic JIA
- RF+ JIA
- Enthesitis/spondylitis related JIA
- Early onset ANA+ JIA
- Other JIA
- Unclassified JIA
Systemic JIA

**Fever** of unknown origin for > 3 days and recurring for at least 2 weeks

- **Major criteria**: erythematous rash and/or arthritis
- **Minor criteria**: generalized LAD/hepatomegaly/splenomegaly, serositis; arthralgia without arthritis; leukocytosis with neutrophilia

Need fever + 2 major or 1 major and 2 minor

Systemic JIA

- Leukocytosis, hypochromic microcytic anemia, thrombocytosis, elevated acute phase reactants$^{4,13}$
- **ANA and RF negative**$^{4,13}$
Systemic JIA

- Salmon colored rash on trunk and proximal extremities
- Can be polyarticular in both small or large joints

Poll Question #1

Which of the following is not required for the diagnosis of systemic juvenile idiopathic arthritis?
A. +ANA
B. Fever of unknown origin
C. Arthralgia
D. A and C
E. A and B
**+Rheumatoid Factor JIA**

- Arthritis > 6 weeks and 2 +RF tests or +CCP\textsuperscript{13}

**+Rheumatoid Factor JIA**

- **Worst prognosis**
- Female predominance
- Late onset
- Symmetric and progressive
-Predominantly affects wrists and small joints hands and feet\textsuperscript{13}
Enthesitis/spondylitis-related JIA

- Peripheral arthritis and enthesitis
- Arthritis or enthesitis + ≥3 months of inflammatory back pain and sacroiliitis on x-rays
- Arthritis or enthesitis + two of the following
  - SIJ TTP
  - Inflammatory back pain
  - +HLA-B27 antigen
  - Acute symptomatic anterior uveitis
  - H/o SpA in 1st degree relative

Early onset ANA+ JIA

- Arthritis ≥ 6 weeks
- Early onset (< 6 years)
- Two +ANA at least 3 months apart (titer > 1/160)
Other JIA

- Arthritis > 6 weeks
- Does not fit criteria for previous categories

Unclassified JIA

- Arthritis > 6 weeks
- Fits > 1 previous disorder
Poll Question #2

A 13 year female with a 8 week history of joint pain and swelling in bilateral wrist and toes would most likely have the following lab values?

A. -ANA
B. -RF and -ANA
C. +RF and +CCP
D. +HLA-B27
E. Type O blood

Work Up

- **No antibody panels** unless positive ANA and evidence of rheum disease!²
- Initial lab testing: ANA, CBC, RF, ESR and CRP¹
- Initial imaging: MRI or ultrasound preferred⁸
Complications

- Uveitis

Complications

- Increased risk of developing CVD\textsuperscript{4,11}
- Increased systolic and diastolic pressures\textsuperscript{11}
- Increased aortic stiffness\textsuperscript{11}
Complications

- Sleep and psychosocial factors

Complications

- Growth retardation
- Macrophage activation syndrome
- Multi-organ insufficiency
- Osteoporosis
Treatment

• Involves multidisciplinary team\textsuperscript{4, 9}
  – PCP
  – Pediatric rheumatologic
  – Ophthalmologist
  – Pediatric psychiatrist
  – Physical therapist
  – Sports med/ortho

Treatment

• NSAIDs
• DMARDs
• Corticosteroids
• Biologics
Treatment

- Monitor pain, sleep and psychosocial indicators for both parents and patients\(^6\)

Prognosis

- FHx disease, early ankle or hip joint involvement, erosions on xray and higher number of joints affected are poor prognostic indicators\(^4, 8\)
- Remission rate increased and rate of joint damage decreases with early initiation of treatment\(^4\)
Follow Up

• Psychosocial monitoring
• Cardiac screening
• Eye screening

Poll Question 4

You are evaluating a 3 year old female with fever of unknown origin daily for 2 weeks and joint swelling. You suspect JIA, although are still waiting on lab results and imaging. Which is the most appropriate next step?
A. Refer to sports medicine/ortho
B. Discuss patient’s sleep habits with patient
C. Refer to ophthalmology
D. Order EKG
E. Order bone marrow biopsy
Practice Recommendations

• JIA is a broad diagnosis of exclusion and does not necessarily require arthritis/arthralgia
• Practitioners need to be aware of presentations in order to start workups quickly – better outcomes are associated with early treatment
• Initial lab work should include ANA, RF, CBC, ESR, CRP, HLA-B27 and imaging of affected joints
• Inclusion of multidisciplinary team is key early in diagnosis and treatment
• Uveitis is the primary extra-articular complication of JIA and needs to be screened frequently
• NSAIDs and DMARDs are first line therapies

Questions
Resources


Contact Information

Sarah Merrill, MD
UC San Diego Health Systems
semerrill@ucsd.edu