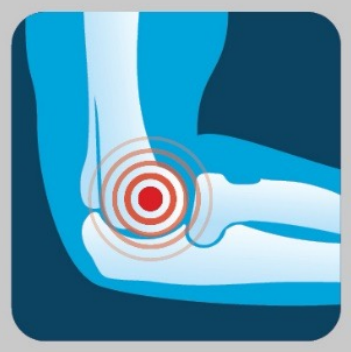
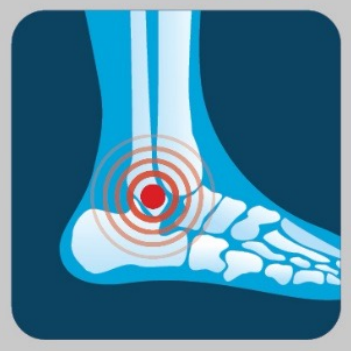




COMPLIMENTARY CME/CE

Thriving Through Transitions in JIA Care: The Vital Role of Advanced Practice Providers

Developed in Collaboration

Med-IQ

National Association of Pediatric Nurse Practitioners℠

AAPA

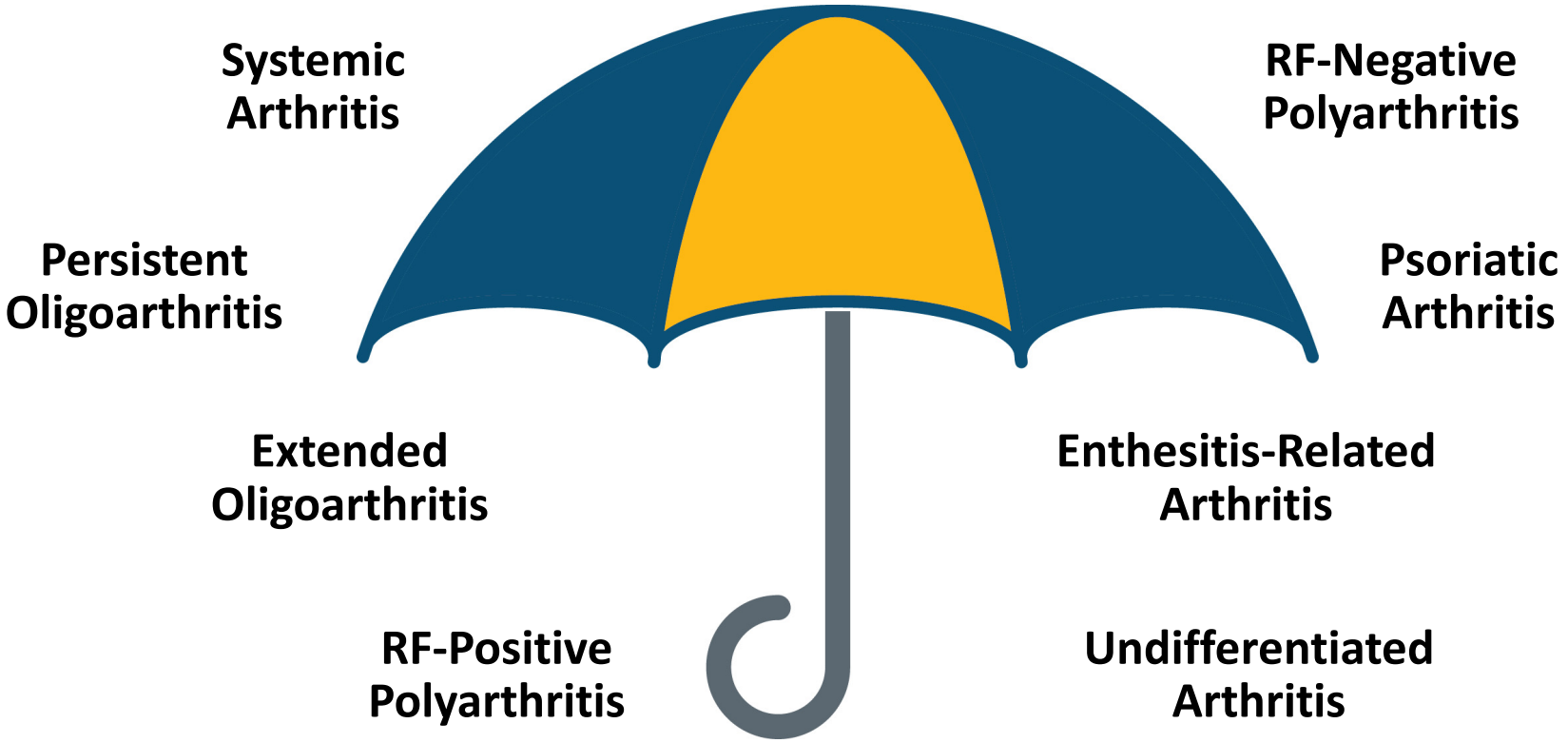
Introductions

Video 1: Brief JIA Background

- Most common rheumatic disease in childhood
- Incidence and prevalence vary by gender, race, and geographic location
- Incidence: about 11 per 100,000 person-years
- Prevalence: about 45 per 100,000 persons
- Approximately 300,000 children in the US
- Onset before age 16 years
- Arthritis in one or more joints that lasts 6 weeks or longer
- Other etiologies excluded such as trauma, local infection, systemic infection, malignancy, post-vaccination, other connective tissue diseases

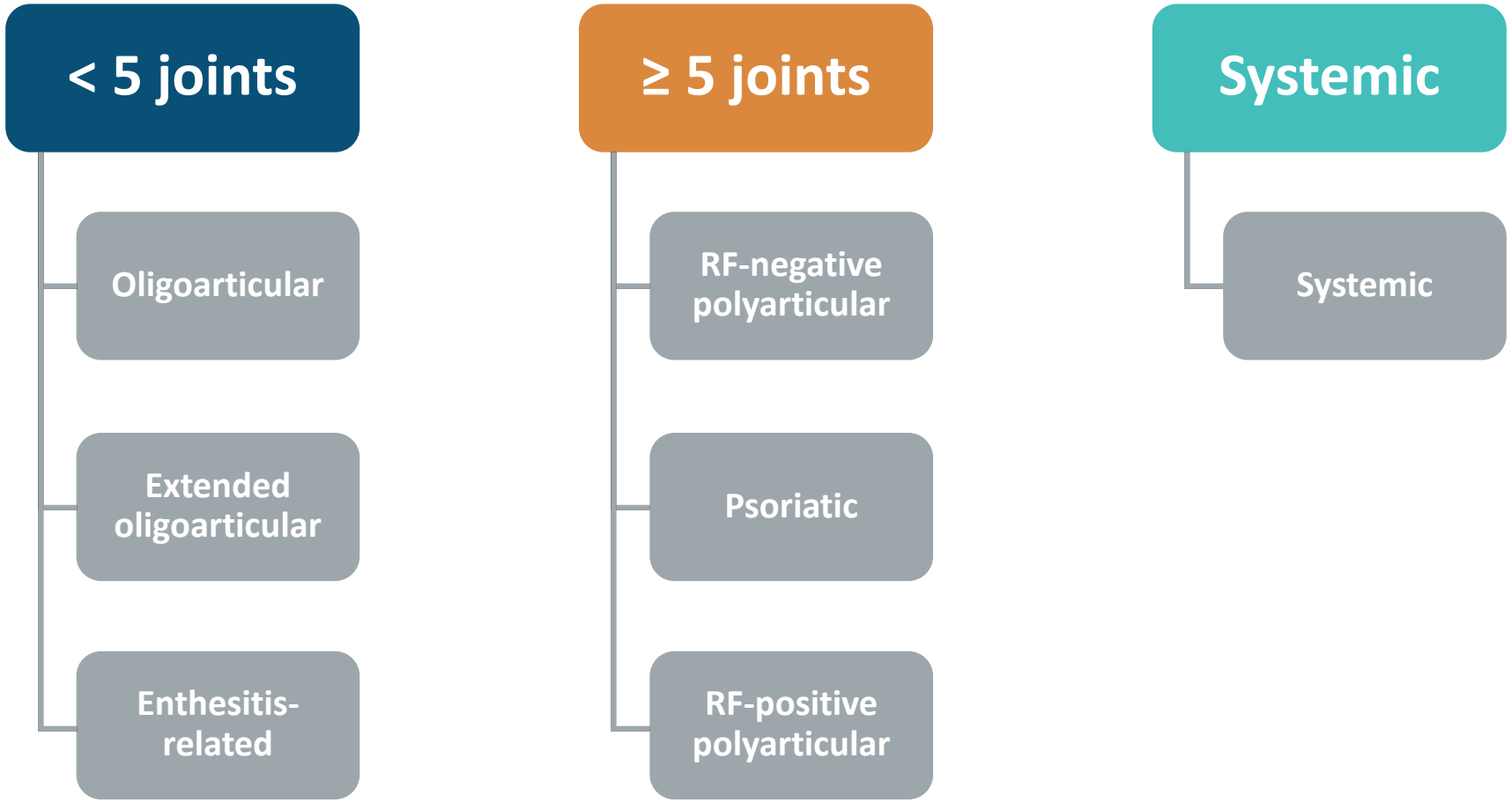
Thierry S, et al. Joint Bone Spine. 2014;81:112-7;
Harrold LR, et al. *J Rheumatol.* 2013;40:1218-25;
Prakken B, et al. *Lancet.* 2001;377:2138-49.

ILAR Classification



Petty PE, et al. *J Rheumatol.* 2004;31:390-2;
Gowdie PJ, et al. *Pediatr Clin N Am.* 2012;59:301-27.

Classification



Petty R. *Textbook of Pediatric Rheumatology.* 7th ed.

Cases: Video/Discussion Based

Will demonstrate different clinical characteristics as well as available/optimal treatments

Case 1

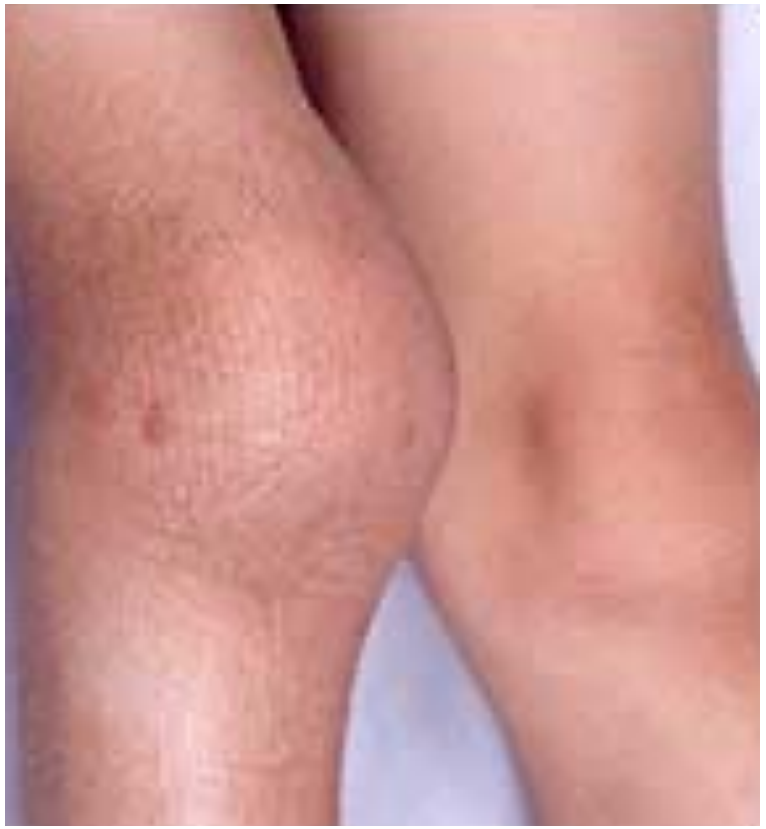
- 4-year-old White female
- Fell off bike 2 months ago
- Two days later, mom noticed swelling of the right knee; since then, the patient has been limping (worse in the morning and after naps) and has had difficulty straightening the leg; she has not had any fevers or rash, but does have mild warmth at the joint
- Evaluated by PCP
  - PE: swelling of right knee with ↓ROM
  - Bilateral leg x-rays WNL; labs showed normal CBC, ESR, UA, CRP, ANA+

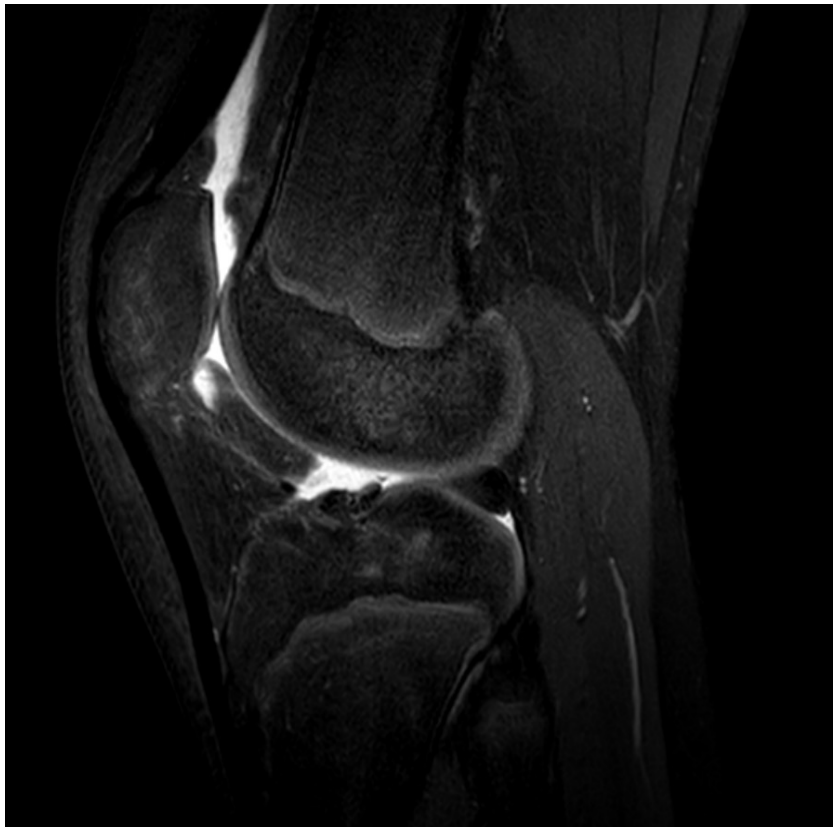
Case 1

- Referred to orthopedics
- MRI of right knee reveals:
  - Moderate joint effusion
  - Synovial thickening
  - No internal derangement
- Referred to rheumatology

Case 1

- HX:
  - Morning stiffness 60 min
  - Stiff after naps
  - Limp
  - No fever, no preceding infection
- PE:
  - Tense swelling of the right knee
  - Limited to full flexion, mild flexion contracture
  - LLD R > L





Oligoarthritis

3:1 Female:Male

Age of Onset: 1-4 yo

Accounts for 50% to 80% of JIA cases

Mild trauma is often a red herring

Risk for leg length discrepancy

Often ANA+ with increased risk of chronic uveitis

Arthritis affecting 4 or fewer joints in the first 6 months

Often a monoarticular arthritis; knee > ankle > elbow

Extended oligoarthritis—develops arthritis in more than 4 joints 6 months or more after onset

Management

- Counseling on diagnosis
- Consultation with physical therapy
- Referral to ophthalmology
  - Up to 20% of patients with JIA will develop uveitis
- Discuss treatment options
  - NSAIDs
  - Intra-articular corticosteroid injection

Angeles-Han ST, et al. *Pediatric Rheumatol Online J.* 2015;13:19.

Non-Steroidal Anti-Inflammatory Drugs (NSAIDs)

- First line for patients with limited arthritis
- Naproxen, meloxicam, celecoxib
- Must be dosed appropriately (anti-inflammatory dosing), taken at the correct interval for at least 6 to 8 weeks; take with food
- Common side effects: stomach upset, ulcer, liver and renal toxicity
- Monitor: CBC, liver enzymes, creatinine, UA every 6 months

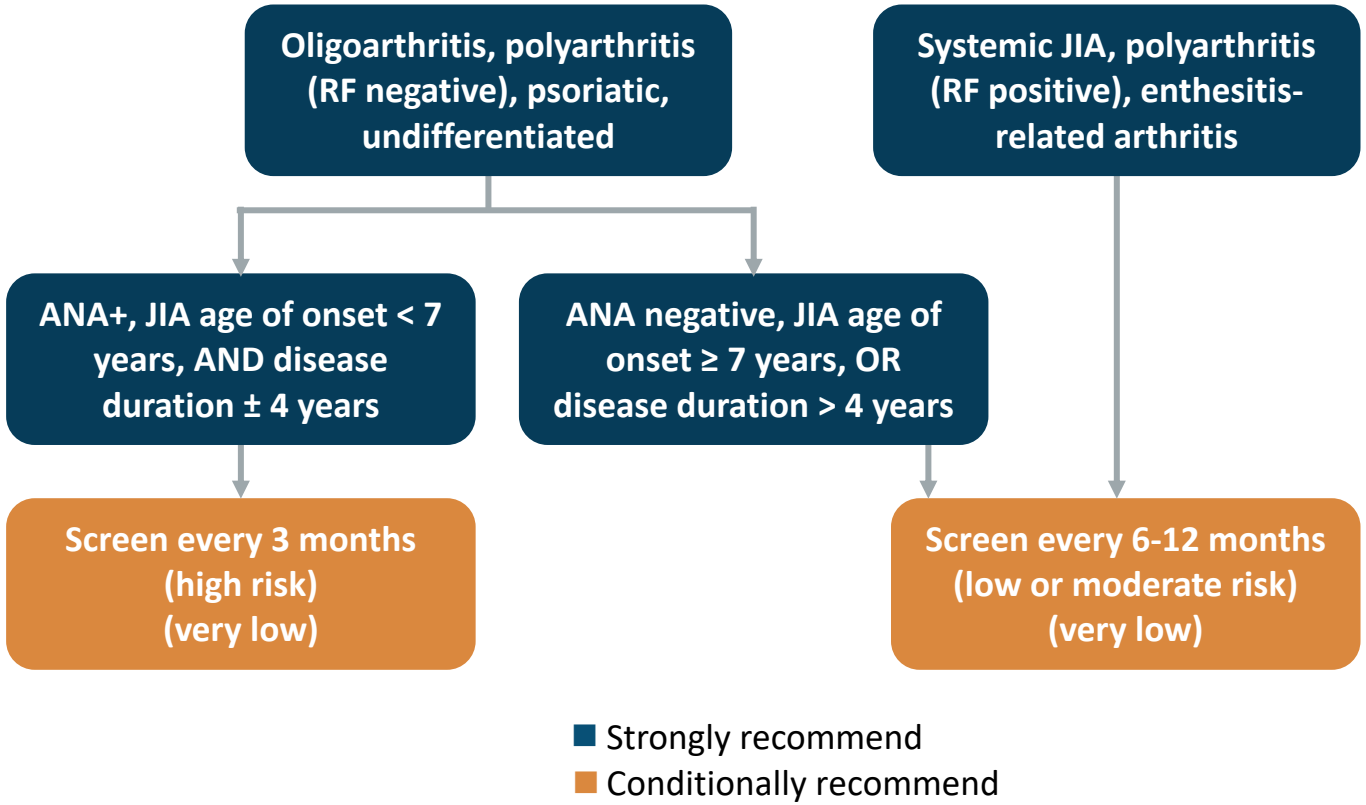
Prescribing information; efficacy data

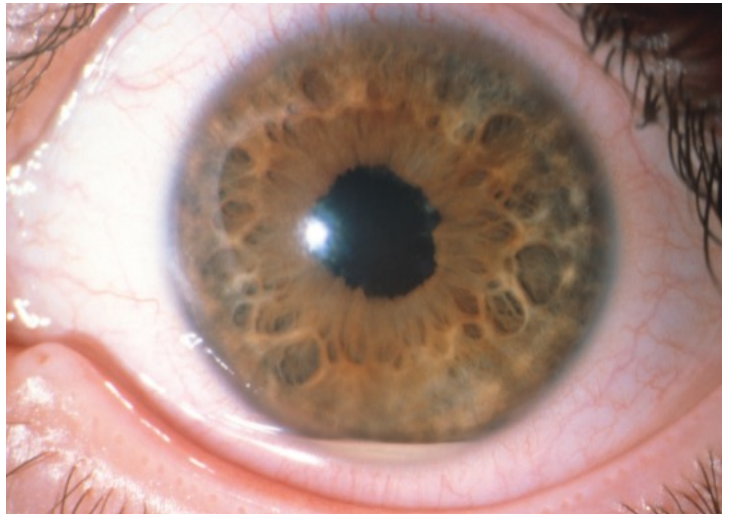
Uveitis

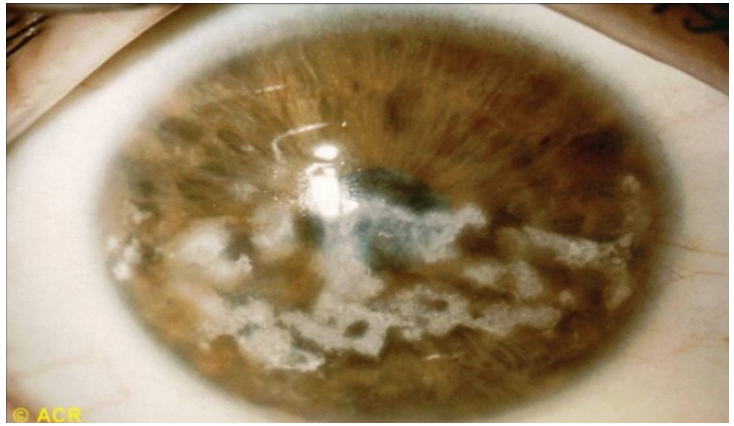
- Usually asymptomatic with insidious onset
- Highest risk:
  - JIA onset before age 7 years
  - Early on in disease course (< 4 years)
  - Oligoarticular subtype
  - ANA+
- Acute uveitis can be seen in JIA patients with ERA, juvenile ankylosing spondylitis, juvenile psoriatic arthritis
- Risk for vision loss, tissue scarring (synechia, band keratopathy), cataracts/glaucoma (long-term topical steroid eye drops)

Cassidy J, et al. Pediatrics. 2006;117:1843-5;
Hayworth JL, et al. *Joint Bone Spine*. 2019;86:685-90.

Current Screening Guidelines







Angeles-Han ST, et al. *Arthritis Care Res (Hoboken).* 2019;71:703-16.
Photo credits: American College of Rheumatology From ACR Image Bank: Top photo: Synechiae; Bottom photo: Band Keratopathy

Case 2

- A 14-year-old White male presents with a 4-month history of left ankle and foot swelling
- He is having trouble playing basketball
- About 2 months ago, he also developed left sided low back pain; he thought he pulled a muscle lifting weights
- Maternal uncle has history of Crohn’s disease

Case 2

- HX:
  - Morning stiffness for 60 min
  - Limp
  - Pain in left lower back with bending forward and squats
  - No hx for abdominal pain, diarrhea, bloody stools, weight loss, mouth ulcers
- PE:
  - Diffuse swelling of the left foot and ankle
  - Left ankle and subtalar limitation
  - Positive FABER on the left
  - Left SI tenderness

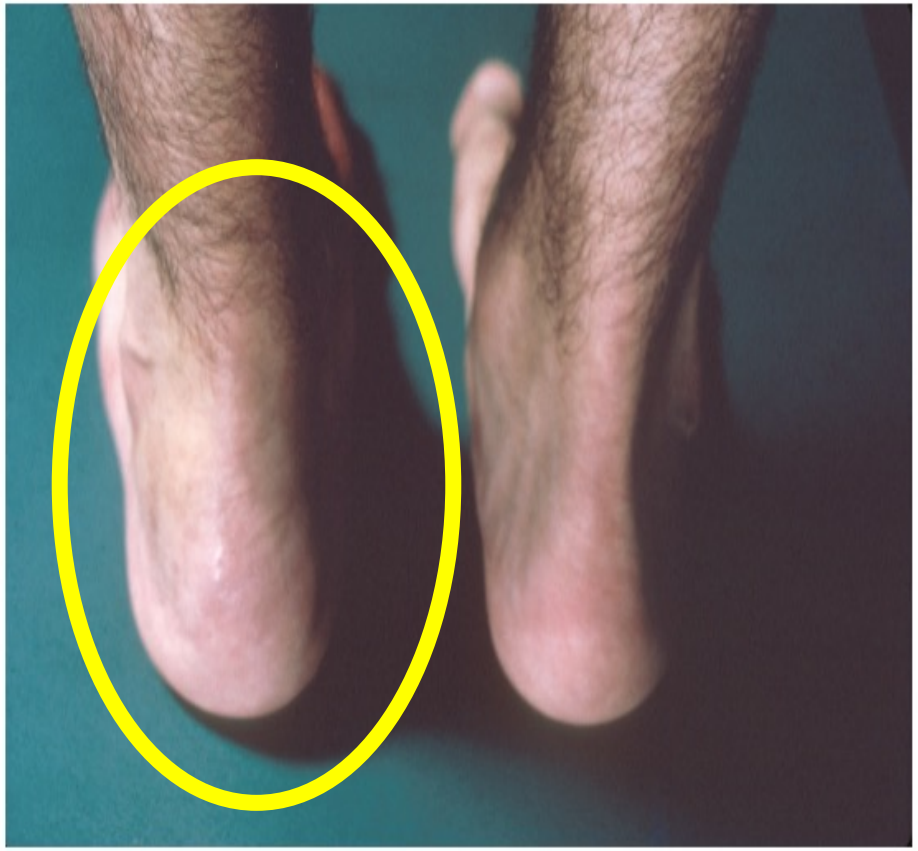
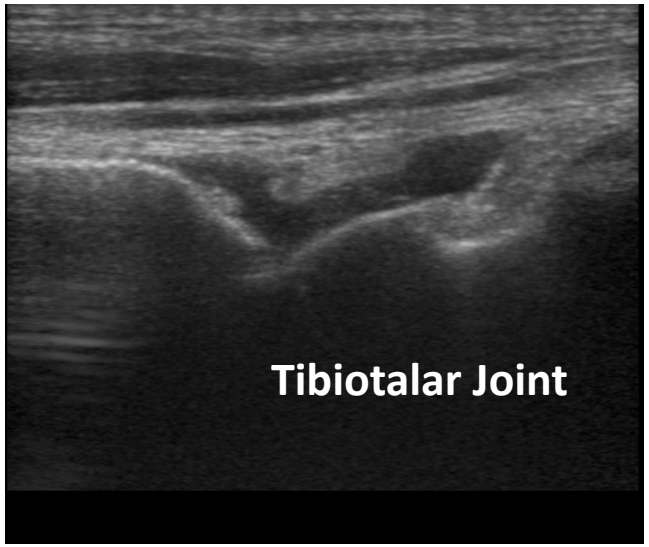
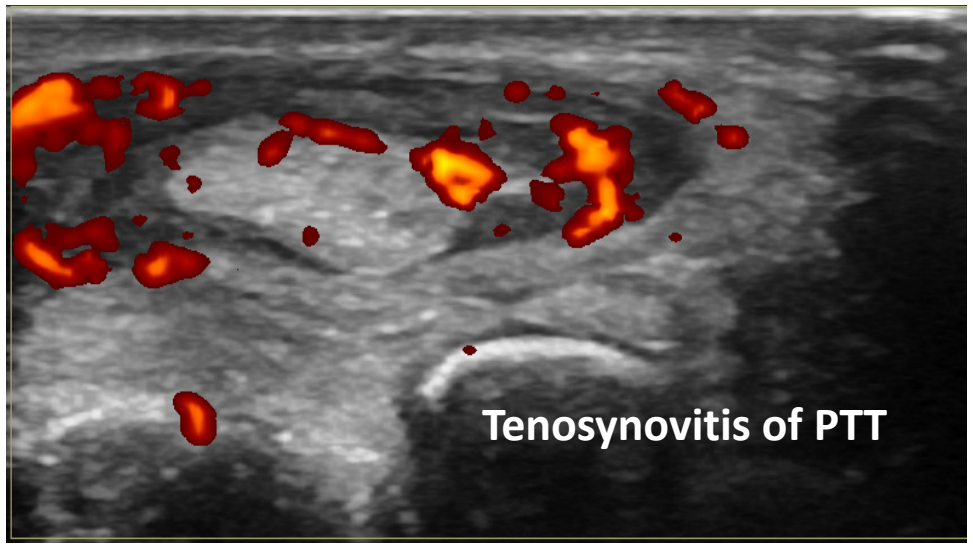


Photo credit: American College of Rheumatology

Patient Work-up

- Labs:
  - ESR 45
  - HLA-B27 positive
- Imaging:
  - US of right ankle
  - MRI of the SI joints





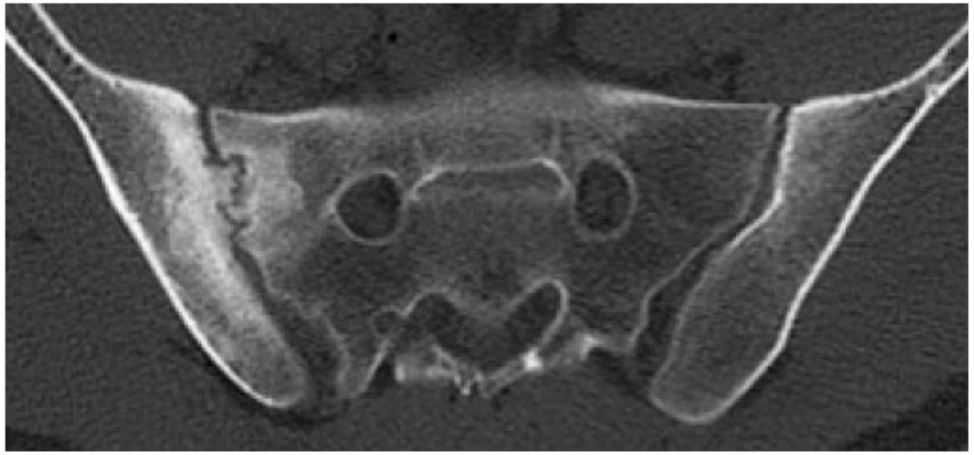


Photo credit: American College of Rheumatology

Case 2: Enthesitis-Related Arthritis

4:1 Male:Female

Age of Onset: 9 to 12

Accounts for 10% to 20% of JIA cases

Arthritis is usually oligoarticular at onset; typically large lower-extremity joints

Prognostic Indicator: HLA-B27+

Risk for acute uveitis

Risk for sacroiliitis

Enthesitis: inflammation where ligaments, tendons, fascia, capsules attach to bone

Management

- Counseling on diagnosis
- Consultation with physical therapy
- Referral to ophthalmology
  - Up to 20% of patients with JIA will develop uveitis
- Discuss treatment options
  - NSAIDs
  - DMARD: methotrexate
  - Biologic and small molecule DMARDs

Angeles-Han ST, et al. *Pediatr Rheumatol Online J*. 2015;13:19.

Juvenile Psoriatic Arthritis (ERA subtype)

Female > Male

Age of Onset: bimodal—1-3 and 9-11

White > Hispanic, African American, Asian

Morning stiffness, painful/swollen joints, psoriasis or family history of psoriasis

Chronic painless uveitis

Nail pitting, +/- psoriatic rash; dactylitis is also common

Usually asymmetric joint pattern

Dactylitis then develops additional large joint swelling; small and large joints

DIP involvement

Petty R. *Textbook of Pediatric Rheumatology*. 7th ed.

Juvenile Psoriatic Arthritis

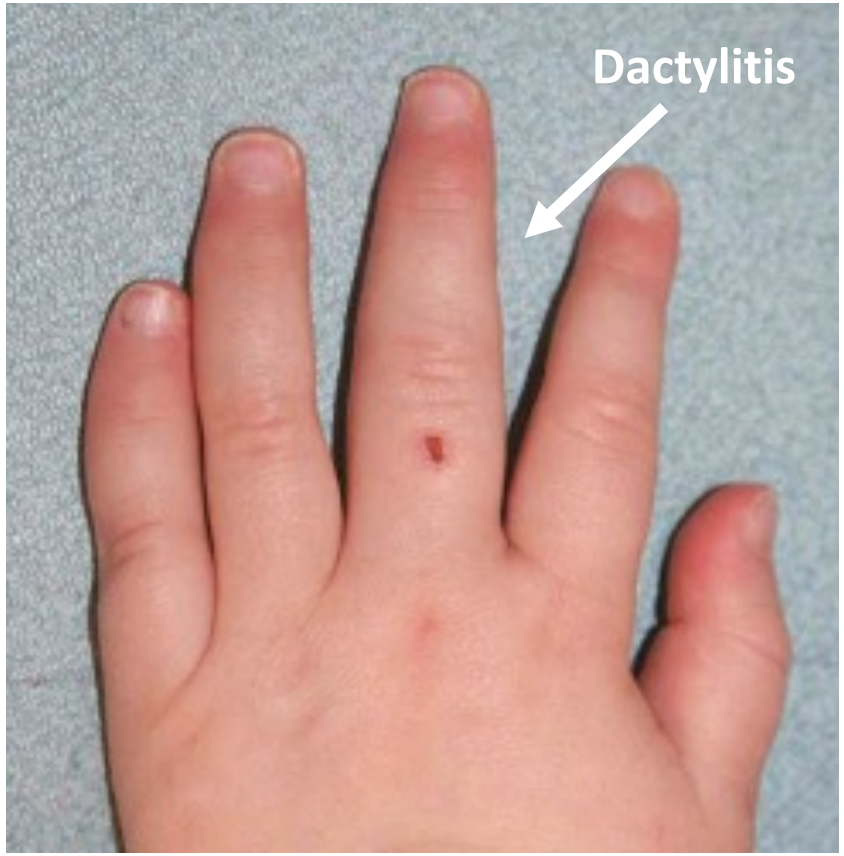


Photo Credit: Texas Scottish Rite Hospital
Petty R. *Textbook of Pediatric Rheumatology*. 7th ed.
Photo courtesy of Lorien Nassi, MD.

Case 3: History and Physical

- **14-year-old Hispanic girl**
- Jammed her right 3rd and 4th fingers while playing basketball 2 weeks ago
- Evaluated in ED with normal x-rays
- Right **finger joint swelling** and now some left finger joints look swollen, too, in a **symmetric distribution**
- **Physical exam: 12 swollen joints**, mainly **small** or **upper-extremity joints**

Case 3: Labs

Lab	Results
CBC	**Normocytic anemia, elevated platelets**
ESR	**Mildly elevated**
Urinalysis	Normal
Albumin	Low normal to normal
Creatinine	Normal
Autoantibodies	**ANA positive 1:320** **RF positive > 14** HLA-B27 negative

Case 3: Polyarthritis, RF Positive

5:1 Female:Male

Age of Onset: 9-16; mean age, 12

Hispanic, African American > White

Severe morning stiffness, fatigue, malaise, multiple swollen, tender joints in **symmetrical** pattern

Involving **small joints** of hands and feet and joints of **upper and lower extremities**

Arthritis affecting 5 or more joints during the first 6 months

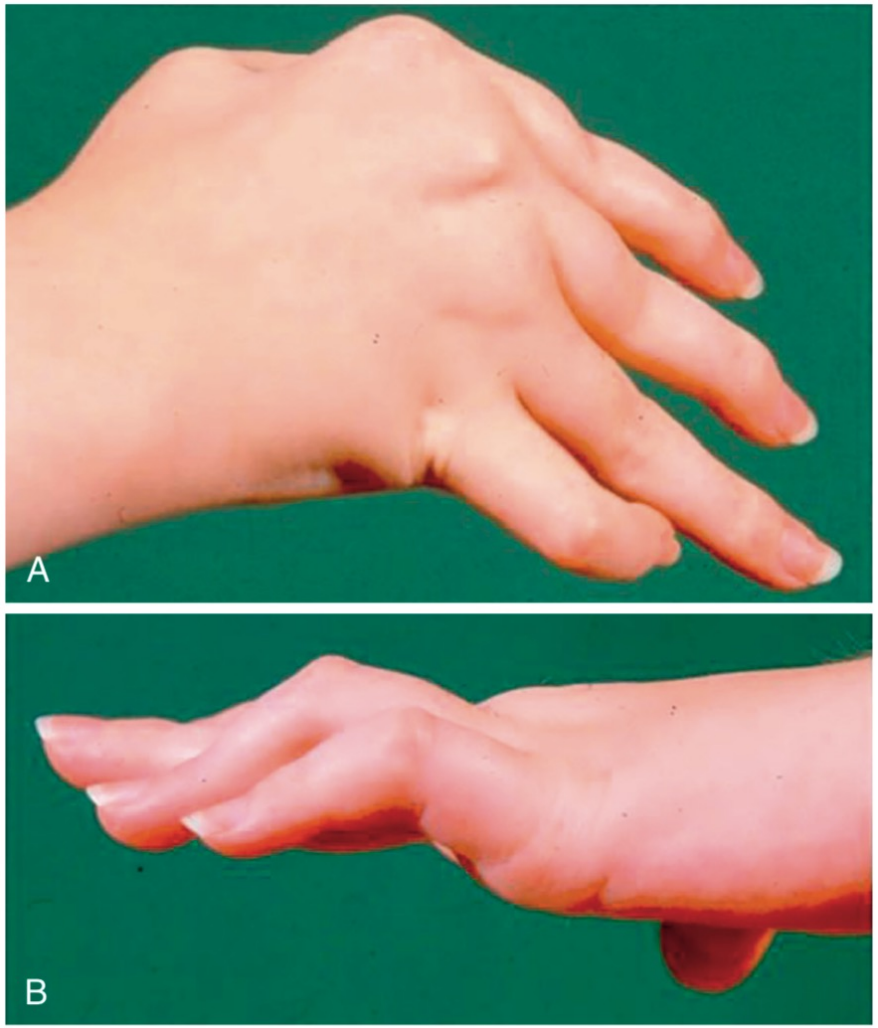
2 positive rheumatoid factors at least 3 months apart

Deformities, bony erosions, subcutaneous nodules

Polyarthritis—shoulders, elbows, wrists, MCPs, PIPs, hips, knees, ankles, and toes
Note: spares the DIPs

Petty R. *Textbook of Pediatric Rheumatology*. 7th ed.

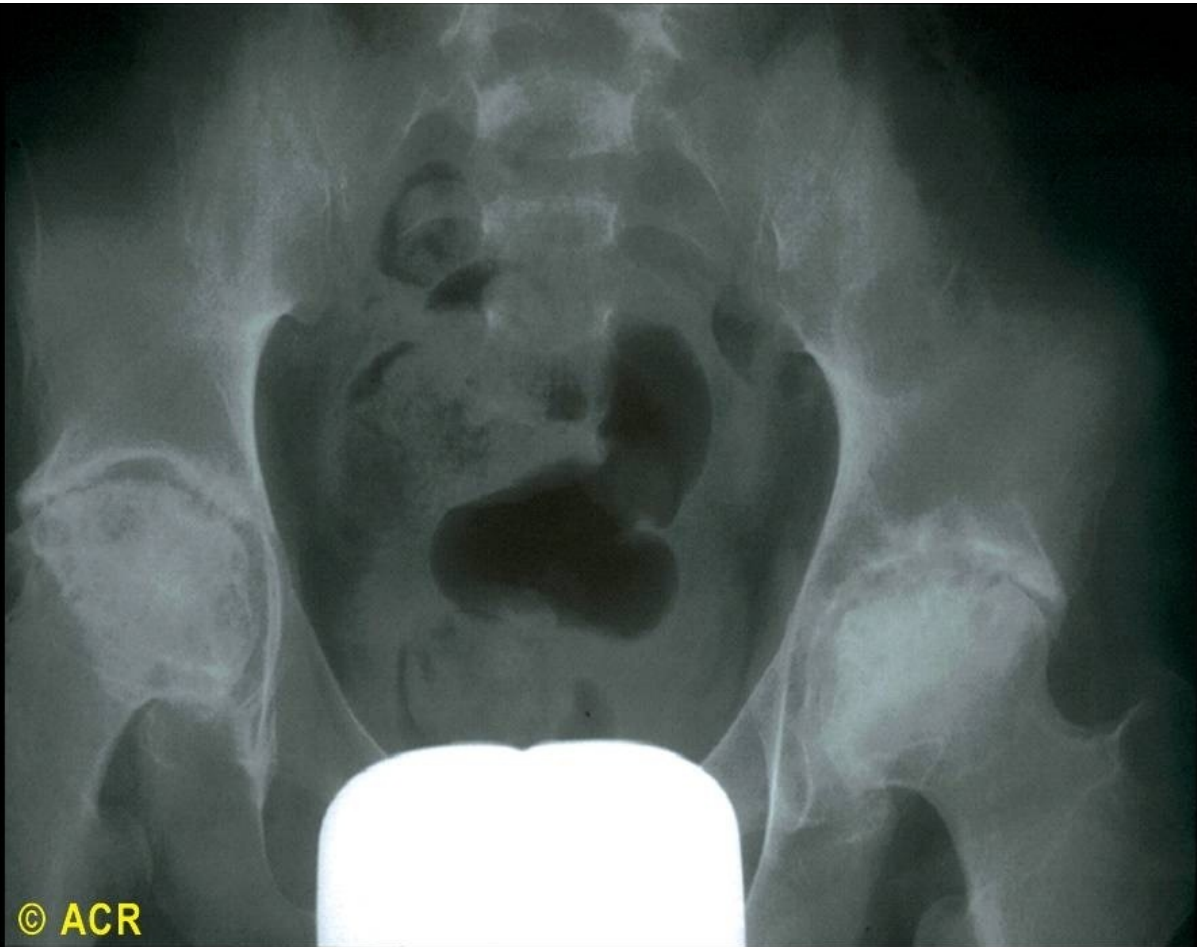
RF-Positive Polyarticular JIA



- Top photograph shows Swan neck deformities in the second through fourth digits with extension at PIPs and flexion at DIPs; ulnar deviation at MCPs can occur
- Bottom photograph shows Boutonniere deformities in fourth and fifth digits with flexion at PIPs and extension at DIPs

Petty R. *Textbook of Pediatric Rheumatology*. 7th ed.

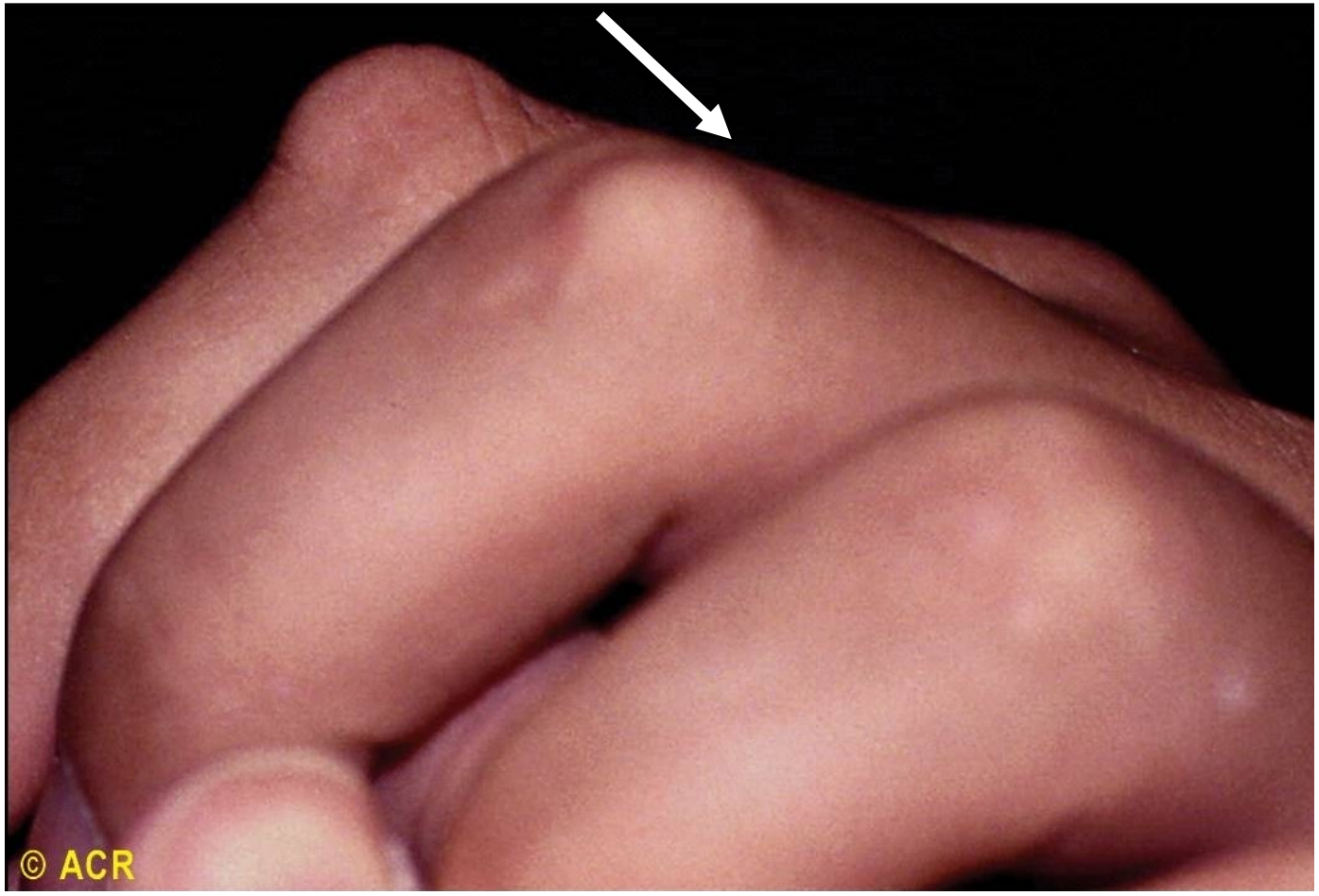
RF-Positive Polyarticular JIA



- X-rays of a 15-year-old boy with a 4-year course of incompletely controlled inflammatory bilateral hip arthritis
  - Bilateral hip joint-space narrowing
  - Erosive changes of the femoral heads and acetabula

Photo credit: American College Rheumatology
Petty R. *Textbook of Pediatric Rheumatology*. 7th ed.

RF-Positive Polyarticular JIA



- Rheumatoid nodules are seen in the seropositive polyarticular subgroup
- Subcutaneous nodules are present overlying the proximal interphalangeal joints of this child with polyarticular JIA

Photo credit: American College Rheumatology
Photo courtesy of Lorien Nassi, MD, ACR.

RF-Positive Polyarticular JIA: Prognosis

- **< 5%** chance of spontaneous remission
- **Severe progressive** disease with extensive radiologic changes (eg, bony erosions)
- Requires early, **aggressive treatment**
- May require **escalation of treatment**

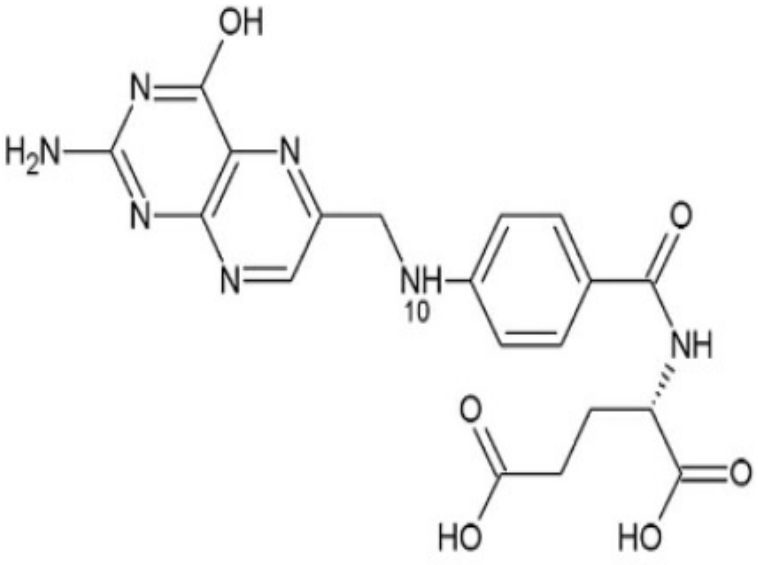
Petty R. *Textbook of Pediatric Rheumatology*. 7th ed.

RF-Positive Polyarthritis Treatment

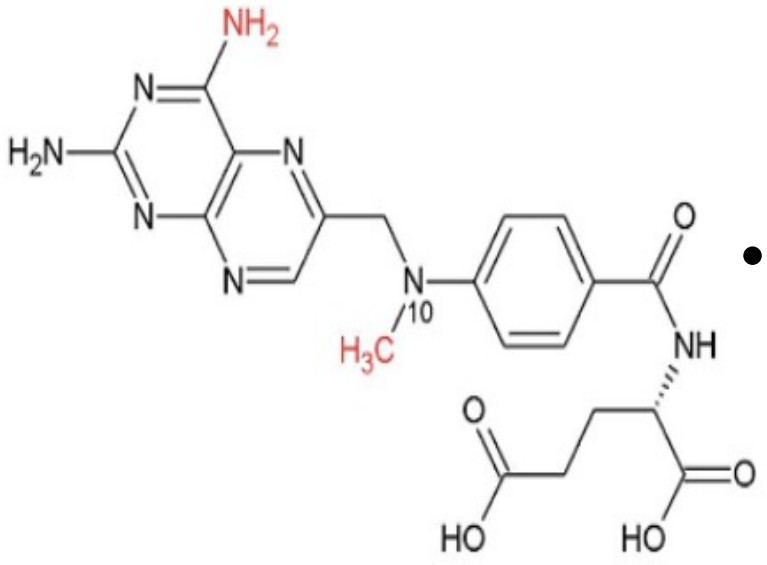
- While waiting for her rheumatology appointment in 2 weeks, she is started on an appropriate anti-inflammatory dose of **naproxen**
- She is finally seen at her rheumatology appointment; she had **14 joints** that were swollen and limited

What medication would be the *next step*?

Methotrexate (MTX)
Disease-Modifying Antirheumatic Drug (DMARD)



Folic Acid



Methotrexate

- Methotrexate is a folic acid analogue and a potent competitive inhibitor of several enzymes in the folate pathway
- We often tell patients to take folic acid on the days they are not taking methotrexate to help with common side effects

Petty R. *Textbook of Pediatric Rheumatology*. 7th ed.

DMARDs

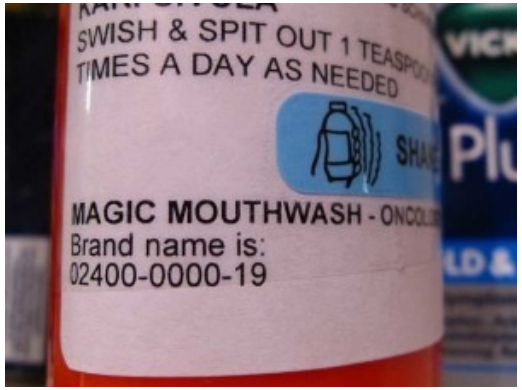
- MTX:
  - Well-established efficacy
  - Used in relatively low doses →
    - 10 to 15 mg/m^2, once weekly
  - When will we see improvements?
    - 6 to 12 weeks
- Recommended in combination with other biologic DMARDs

Petty R. *Textbook of Pediatric Rheumatology*. 7th ed;
American College of Rheumatology. www.rheumatology.org/Practice-Quality/Clinical-Support/Clinical-Practice-Guidelines/Juvenile-Idiopathic-Arthritis.

Helping With Possible Side Effects

Injection-site reaction →

Mouth sores →



Nausea →

Polyarthritis, RF Negative

3:1 Female:Male

Age of Onset: biphasic 1-4 yo; late childhood/early adolescence

White > Hispanic, African American, Asian

Multiple symmetrical large joints
- Can often have TMJ, C-spine involvement

Chronic uveitis ~5 %

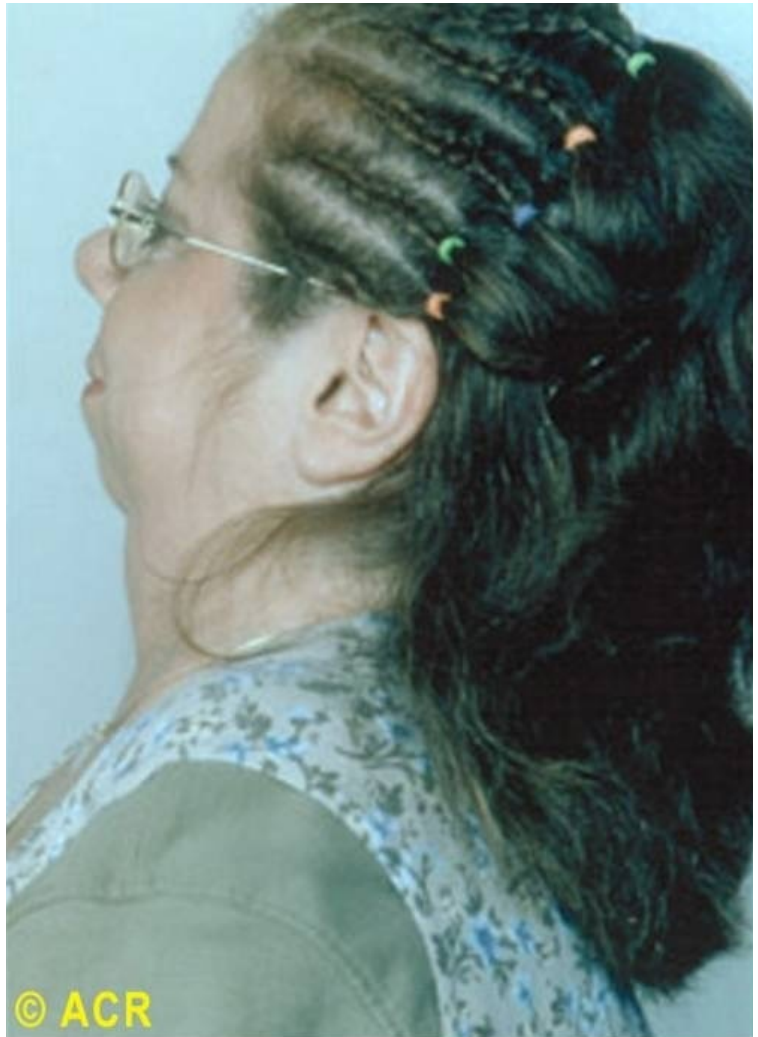
Arthritis affecting 5 or more joints

TMJ involvement—long term causes **micrognathia**

Polyarthritis—C-spine, shoulders, elbows, wrists, MCPs, PIPs, hips, knees, ankles, and toes

Petty R. *Textbook of Pediatric Rheumatology*. 7th ed.

RF-Negative Polyarticular JIA



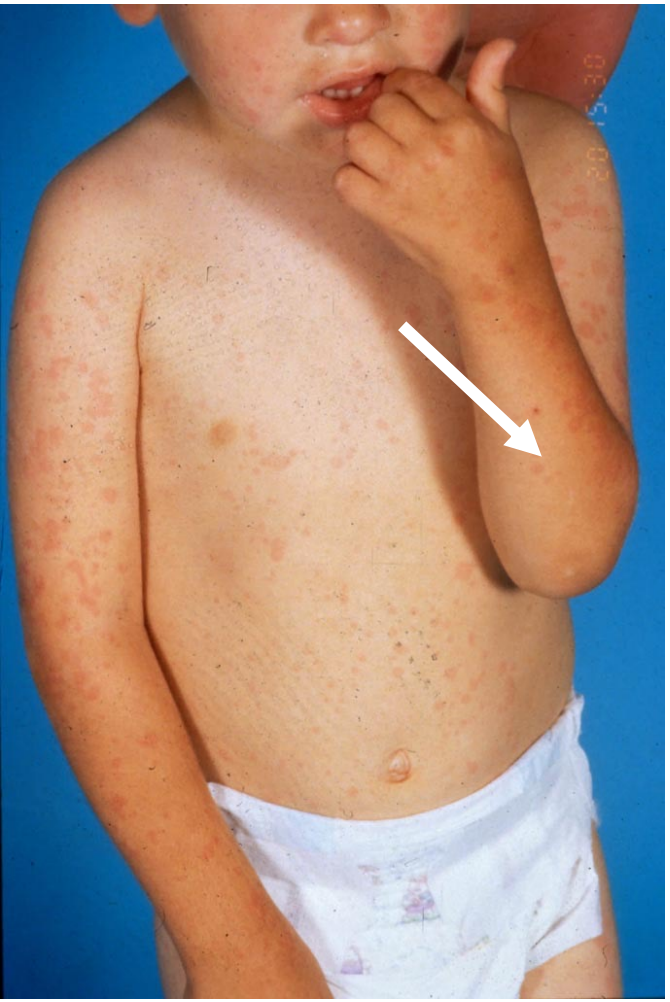
- This patient had RF-negative polyarticular JIA with TMJ arthritis and micrognathia, or underdeveloped jaw

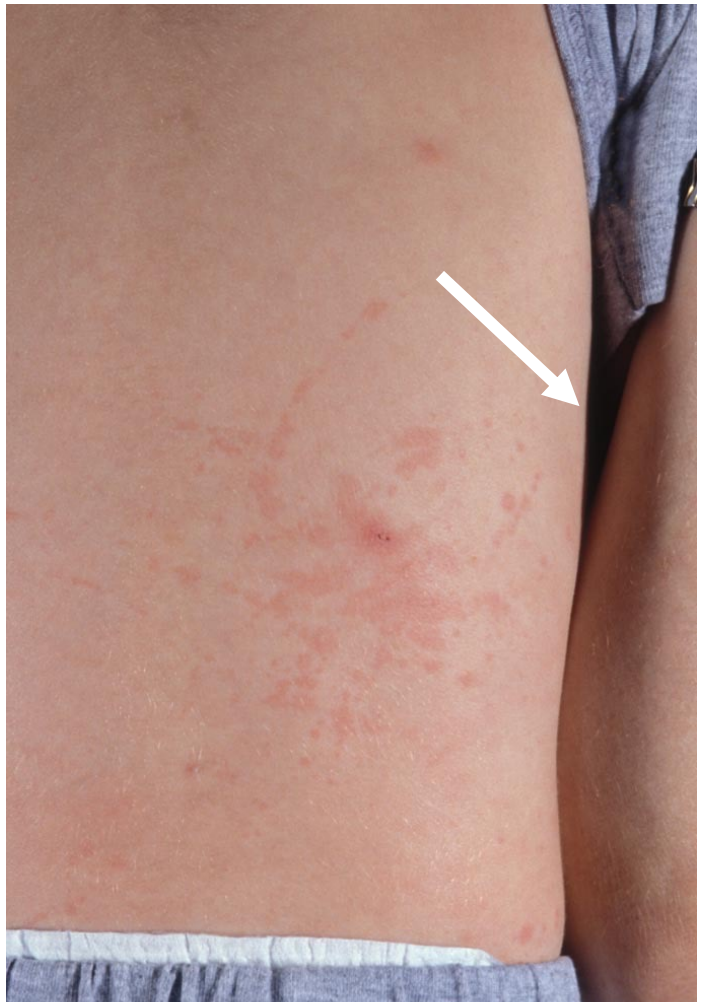
Photo credit: American College Rheumatology
Photo courtesy of Marilynn Punaro, MD.

Case 4: Systemic JIA

- 2-year-old White male, vaccinated, recently started daycare
- **Daily, high-spiking fever** to 105°F for 7 days with mild cough, stopped walking
- **Pale pink, salmon-colored rash, migratory** all over his body→ more on his trunk, **only during fevers and disappears after a few hours**
- Seen by PCP who refers to local emergency department for concern of Kawasaki disease
- Patient is admitted; rheumatology is consulted because of swollen hands/feet

Case 4: Physical





Physical exam:

- Febrile, temperature 102.2°F
- Pale, fussy
- Slightly tachycardic
- Mild cervical lymphadenopathy
- Salmon-colored, patchy rash on trunk and extremities with erythematous macules ~ 2- 5 mm; a few linear streaks
- Limitations and swelling in bilateral elbows, knees
- Bilateral moderate effusions on hip US

Photo Credit: Texas Scottish Rite Hospital

Case 4: Labs

Lab	Results
CBC	Very **high WBC > 20** Normocytic anemia Very **high platelets > 700,000**
ESR	**90 (high)**
Urinalysis	Normal
Albumin	**2.9 (low)**
Creatinine	Normal
Autoantibodies	ANA negative RF negative HLA-B27 negative
Ferritin	**5,000 (high)**

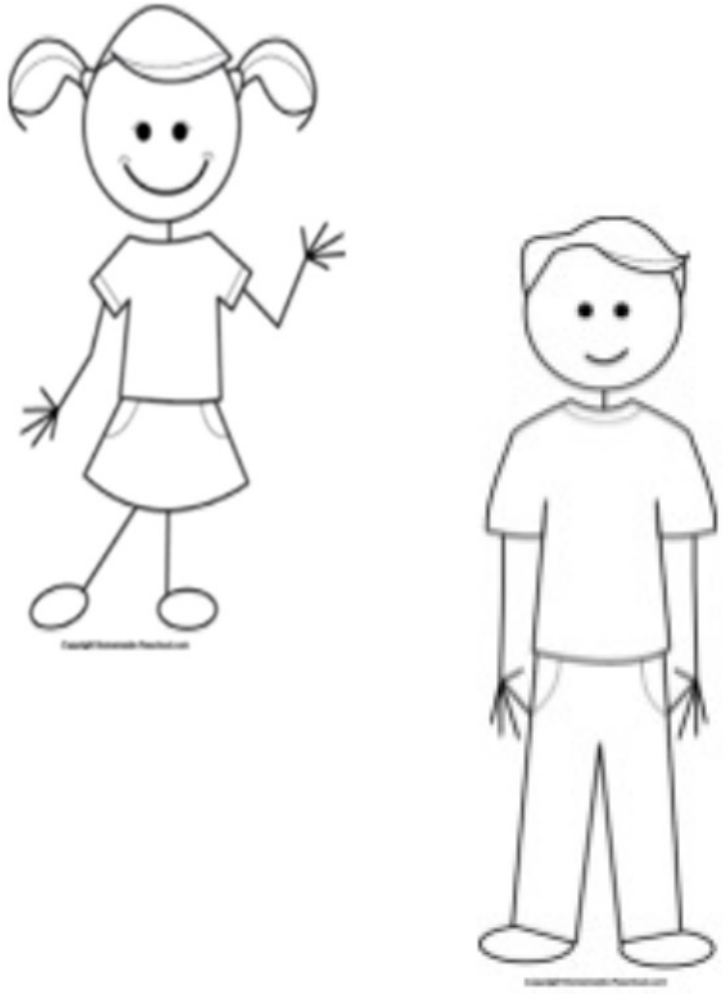
Systemic JIA
"Still Disease" Female = Male

Age of Onset: affects any age; (caution in diagnosing younger than 6 months)

White > Hispanic, African American, Asians

Joint involvement: variable; may have arthralgias or multiple swollen joints; arthritis may not develop for months later

No autoantibodies→ more autoinflammatory disease and no strong genetic predisposition



Fever for at least 2 weeks:

- Daily (usually evenings) or twice daily high grade (usually 102°F or more) for at least 3 days
- Rapid return to baseline or sub-baseline between fevers

Plus arthritis and at least one of the following:

- Rash (80%): evanescent rash

Other systemic features:

- Generalized lymphadenopathy
- Splenomegaly
- Hepatomegaly
- Serositis

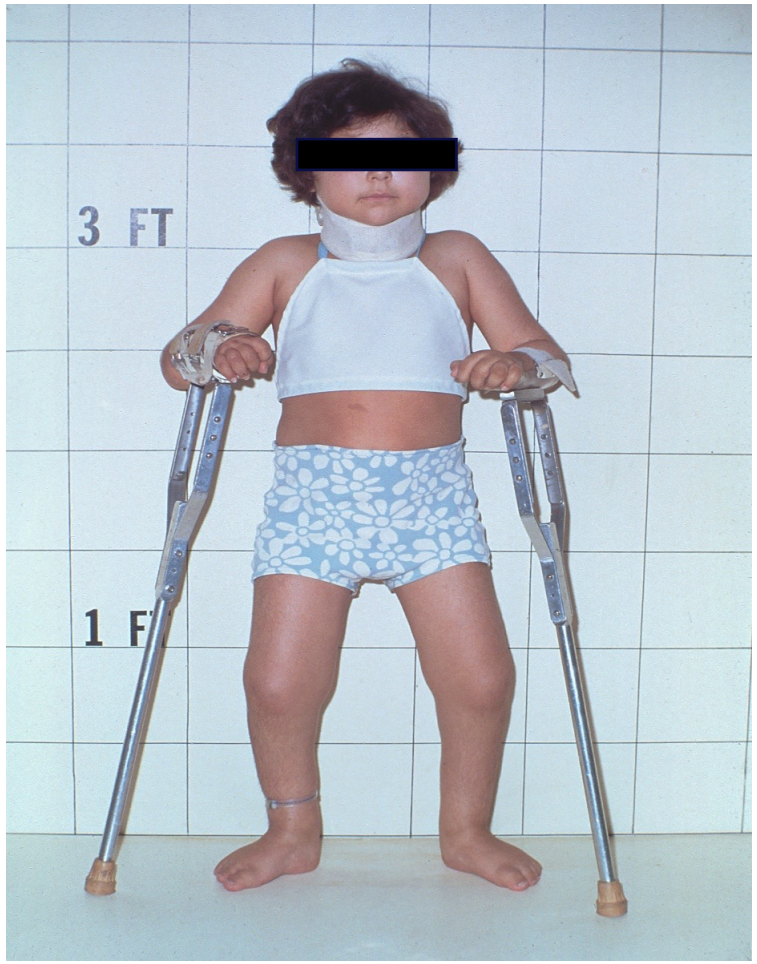
Petty R. *Textbook of Pediatric Rheumatology*. 7th ed.

Systemic JIA Treatment

- NSAIDs
- Methotrexate
- Combination with biologic DMARDs
- Tofacitinib – although not included in 2019 guidelines, what role is it playing in your practice?

Petty R. *Textbook of Pediatric Rheumatology*. 7th ed.

Systemic JIA: Prognosis



- **14-year-old girl** with **inadequately treated systemic JIA** (Still disease)
  - Especially in very early sJIA before 18 months:
    - Severe growth retardation
    - Permanent deformities
    - Multiple contractures
    - Risk of interstitial lung disease

Pascual V, et al. *J Exp Med*. 2005;201:1479-86.
Photo credit: American College Rheumatology

Goals of Treatment

- Radiologic joint damage occurs in most patients with systemic and polyarthritis within 2 years and in oligoarthritis within 5 years
  - Slow radiologic joint damage
  - Maintain low disease activity
- Features of poor prognosis → early aggressive therapy
  - Polyarthritis, positive rheumatoid factor (RF), antibodies to cyclic citrullinated peptides (anti-CCP), presence of human leukocyte (HLA)-DR4, nodules, early onset symmetric small joint involvement
- Low, moderate, and high disease activity based on subset of JIA
- Treat-to-target approach—our goal is clinical remission, which may require biologic switching

Beukelman T, et al. *Arthritis Care Res (Hoboken).* 2011;63:465-82;
Ringold S, et al. *Arthritis Rheum*. 2013;65:2499-512; Ravelli A, et al. *Ann Rheum Dis.* 2018;77:819-28.

Video: Considerations for Transitioning Your JIA Patients to Adult Care

Key Components to Successful Transitions of Care

- Identify a healthcare professional who can assume responsibility for current care, care coordination, and future care planning and who is attentive to the challenges of transition
  - The pediatric healthcare provider should speak directly with the clinician who will assume care and provide a written copy of the medical summary
- Prepare and maintain an up-to-date medical summary that is portable and accessible
- Create a written healthcare transition plan, ideally by age 14 years
  - The adolescent and family should help create the transition plan
  - At a minimum, the plan should include the services that need to be provided, the identification of the person who will provide the services and how they will be financed, details regarding the educational/vocational transition, and who will be responsible for ensuring the services are provided

Conti F, et al. *Clin Exp Rheumatol*. 2018;36:1086-94.

Challenges With Transition of Care

- Rheumatology workforce shortage!
- No national standardized policy—highly variable depending on location
- Insufficient training and limited resources (reimbursement, time, personnel)
- Ideal transition process should be coordinated and adapted to the psychological and social features of the patient, including readiness
- Lack of universal electronic health record
  - May lead to loss of some information or access challenges
- Adherence concerns
- Transitions in medical insurance

Conti F, et al. *Clin Exp Rheumatol*. 2018;36:1086-94; Cruikshank M, et al. *Clin Rheumatol.* 2016;35:893-9.

Resources for Transition of Care: American College of Rheumatology—Transition Toolkit

- Tool for the Pediatric Rheumatologist
  - Transition policy
  - Readiness assessment
  - Transfer letter
  - Medical summary of JIA
- Tools for the Adult Rheumatologist
  - JIA fact sheet
  - Medical summary of JIA
  - Welcome letter
  - Adult self-assessment tool

- **We also developed** a patient checklist and a discussion guide that you can download and provide to your patients to help them during transition

https://www.rheumatology.org/Practice-Quality/Pediatric-to-Adult-Rheumatology-Care-Transition
https://www.gottransition.org

Other Resources for Transition of Care

- American College of Physicians Pediatric to Adult Care Transitions Initiative
  - www.acponline.org/clinical-information/high-value-care/resources-for-clinicians/pediatric-to-adult-care-transitions-initiative
- National Alliance to Advance Adolescent Health
  - www.gottransition.org/families-resources
- National Center on Secondary Education and Transition
  - www.ncset.org

Video Discussion:

Why are advanced practice providers in such a unique position to treat patients with JIA (and in rheumatology more generally)?

Video: Conclusions