

CC: Bilateral foot swelling.

HPI:

- 6 yr old female with PMH anemia presents with approximately 6 weeks of swelling in feet, ankles, wrists.
- Refusing to walk secondary to pain and swelling.
- Joint swelling has been migratory and is usually associated with pain.
- No redness or warmth noted in joints when swollen.
- Night-time fevers for the past 2-3 weeks (100° – 103° ax) which occur twice weekly.
- Patient seems “tired out” and is no longer playing outside or participating in recess at school.
- 10 lb weight loss over the past 3 months.

Pertinent Negatives:

- **Denies rash**
- **Denies prior throat infection**
- **Denies previous URI**
- **Denies TB contacts or risk factors**
- **Denies vomiting/diarrhea**

PMH:

- **ANEMIA:** CBC drawn 3 months prior to admission by PMD to assess for cause of weight loss. Started on FeSO₄.
- **GASTRITIS:** 1 yr PTA, evaluated for abdominal pain. EGD normal, but was found to have H. Pylori gastritis. Also noted to have elevated ESR and CRP. Treated with triple therapy X 2 weeks with improvement noted.
- **BIRTH HX:** 36 WGA. BW 4lb 5oz.
- **DEV HX:** normal
- **FAM HX:** Negative
- **ROS:**
 - House built in 1970
 - Picky eater

PHYSICAL EXAM

- **Vitals: T 36 HR 100 RR 18 BP 98/57 SaO2 100% RA
Wt 16 kg (3rd percentile) Ht 120 cm (75%)**
- **General: Pale, diaphoretic, thin.**
- **HEENT: Sclera white; MMM; TM's normal; pale conjunctiva; palate normal; oropharynx clear; tonsils 1+ without exudate or erythema.**
- **CV: RRR. S1S2 normal. No murmur. 2+ femoral and radial pulses.**
- **Lungs: CTA bilaterally.**
- **Abd: +BS. Scaphoid. Soft, nontender. Liver palpable to 2 cm below RCM. Spleen palpable 3cm below LCM.**
- **Lymph: 1cm anterior cervical LAD and axillary LAD bilaterally.**

PHYSICAL EXAM:

- **MS: Thin extremities**

Swelling bilaterally in wrists, non-tender non-erythematous.
2nd right PIP swollen and tender.

Elbows without swelling, but pain with extremes of flexion bilaterally (flexion to 60°).

Left hip mildly painful with limited ROM.

Ankles swollen bilaterally, non-tender, non-erythematous.
Limitation in internal and external rotation of ankles.

- **SKIN: Diffusely dry and scaling skin. No rash or lesions.**
- **NEURO: negative**

Labs:

- **LABS:**

WBC 12.6/**Hg 7/Hct 24/Plt 750** (N80%, B1%)

MCV 70, RDW 16

BMP normal

Albumin 3.1

LFT's normal

UA unremarkable

CRP 143

ESR 108

Blood Culture drawn

Lead level

Fe-studies

Pre-albumin

- **FILMS: bilateral foot x-rays normal**

HOSPITAL COURSE:

- HD#1: Rheumatology, GI, HONC consulted. Transfused for symptomatic anemia. Hg increased to 10.
- HD#2: Blood cx positive for GPCC and chains. ID consulted and recommend ASO titer, antiDNase B, throat cx, HIV, Xray ankles, US left hip, repeat blood cx.

- HD#3: ASO 400 (<200).

Fe < 10; Ferritin 767; TIBC 144; transferrin 123.

ID service concerned for Rheumatic Fever → recommend PCN X 10 days, ASA, and echo.

Bone marrow Bx: No evidence of leukemia.

- HD#4: ANA positive.
Solumedrol burst initiated.
- HD#5: antiDNase B negative. Solumedrol #2. Discharged home.

Differential Diagnosis of Arthritis

- **Leukemia/Malignancy**
- **Infectious (lyme/viral/bacterial/osteo/septic joint/discitis)**
- **Systemic (SLE/Psoriasis/HSP/serum sickness)**
- **Reactive (Post-strep/Rheumatic Fever/post-enteric)**
- **Inflammatory (IBD/JIA/sarcoid)**

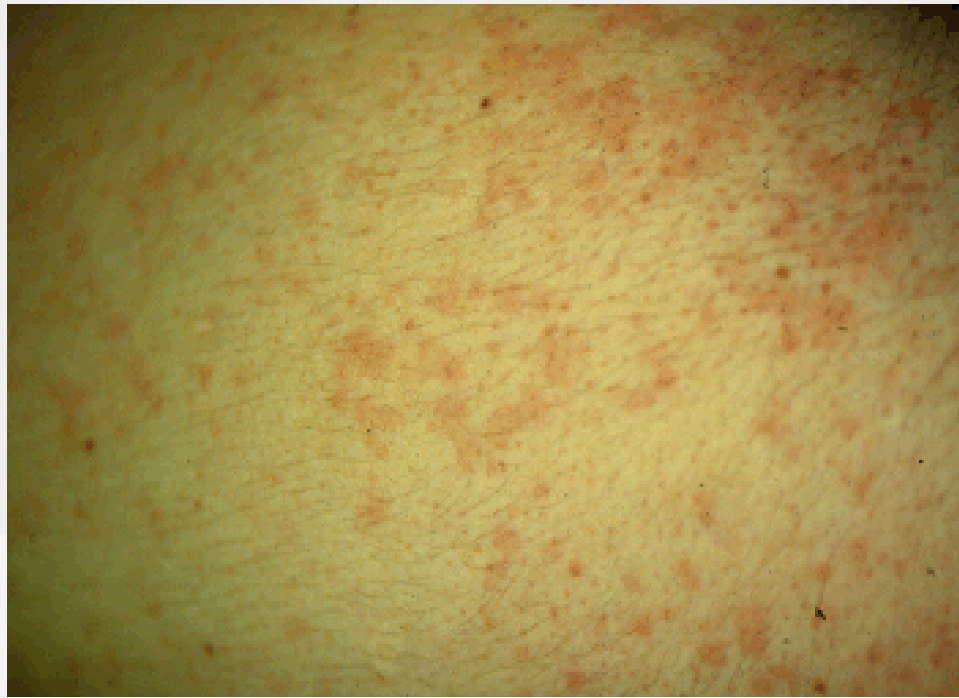
Acute Rheumatic Fever vs. Systemic JIA

	ARF	JIA
Onset	Arthritis develops 2-3 weeks after illness	Arthritis develops and must be present > 6 wks
Time Course	Migratory over weeks	Persists daily A.M. Stiffness
Location	Asymmetric Usually less than 5 joints	Symmetric Non-migratory Oligo to polyarthritis
Labs	Elevated ESR & CRP Elevated ASO & antiDNase B Positive β -strep	Elevated WBC Thrombocytosis Anemia +/- ANA and RF
Other Symptoms	Carditis, chorea, EM, nodules (MAJOR) Fever, arthralgia (MINOR)	Fevers Rash Pericarditis

Skin Findings: ARF vs Systemic JIA

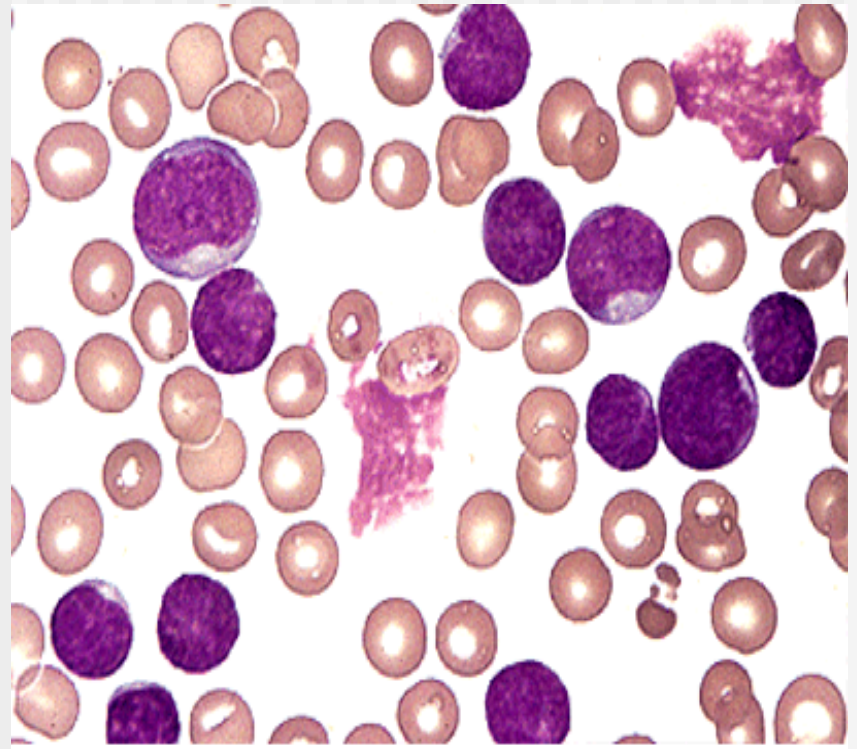


Skin Findings: ARF vs Systemic JIA



Systemic Onset JIA vs Leukemia

- Severe bone pain, night-time
- Fevers low grade
- Labs: Leukocytosis with blasts OR low WBC count
Thrombocytopenia
Elevated LDH
Bone marrow bx



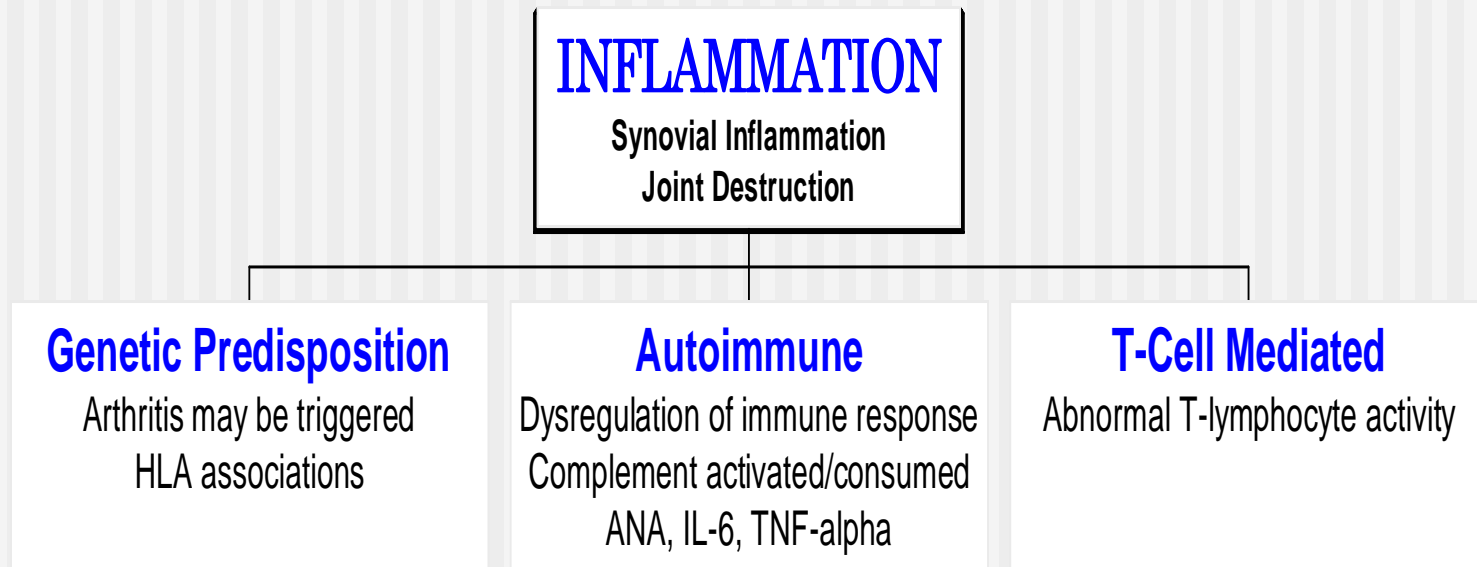
Juvenile Idiopathic Arthritis

- **Diagnosis of exclusion: arthritis in at least one joint > 6 weeks in patient 16 yrs old or younger.**
- **New classification system developed by ILAR**
- **Incidence of JIA ranges with ethnic populations with overall incidence of 1-22 per 100,000**
- **Decreased incidence in urban African-American children**

JIA ILAR Classification

ILAR Classification	Clinical Features	Associated Symptoms
Systemic Arthritis	Arthritis, fever 2 weeks ■ Rash ■ LAD ■ HSM ■ Serositis	Growth abnormalities MAS 25% - joint destruction
Oligoarthritis	Arthritis of 1-4 joints	Uveitis (ANA+) Onset at younger age
Polyarthritis	5 or more joints in the first 6 months	RH + / RH – subsets 10% - joint destruction
Enthesitis- related	Arthritis and/or enthesitis	■ Develop into juvenile spondyloarthropathies ■ Uveitis
Psoriatic	Arthritis and psoriasis OR arthritis AND 2 of: ■ Dactylitis, nail changes, fam Hx	Acute uveitis seen
Other	Unknown cause and greater than 6 weeks	

Theories of Pathogenesis



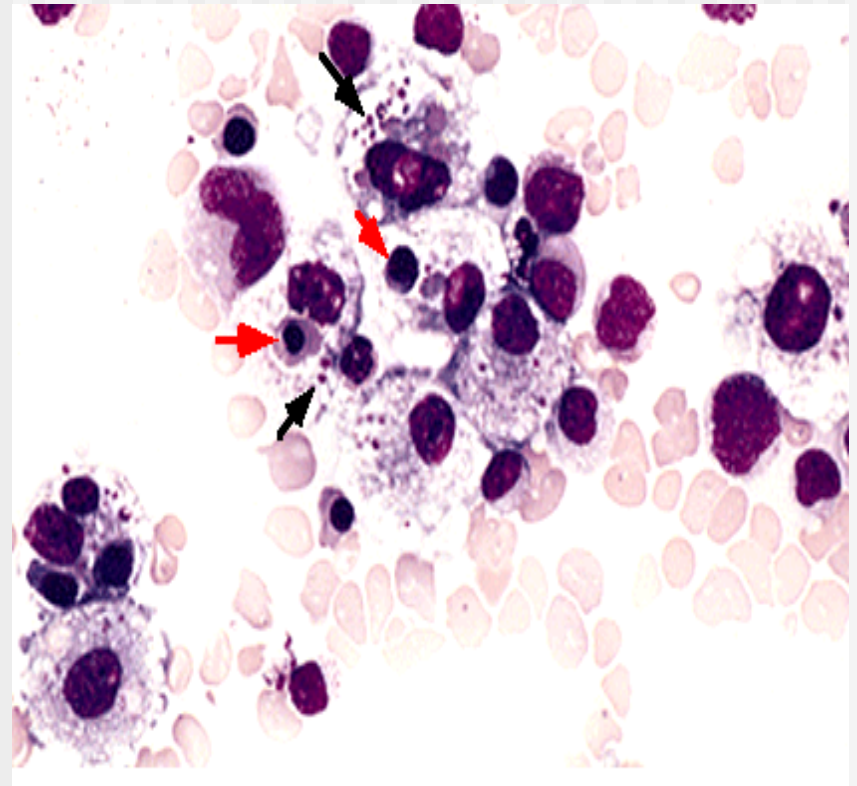
Systemic Onset JIA

- Approximately 10% of all JIA cases
- Affects any age, peak age 1-6 years
- Affects males and females equally
- Double quotidian fever for at least 2 weeks
- Arthritis symmetric of both large and small joints
- Morning stiffness
- Rash
- Systemic symptoms can precede arthritis by up to 6 months
- Rarely ANA+ and RF+
- ASO elevated in up to 30%

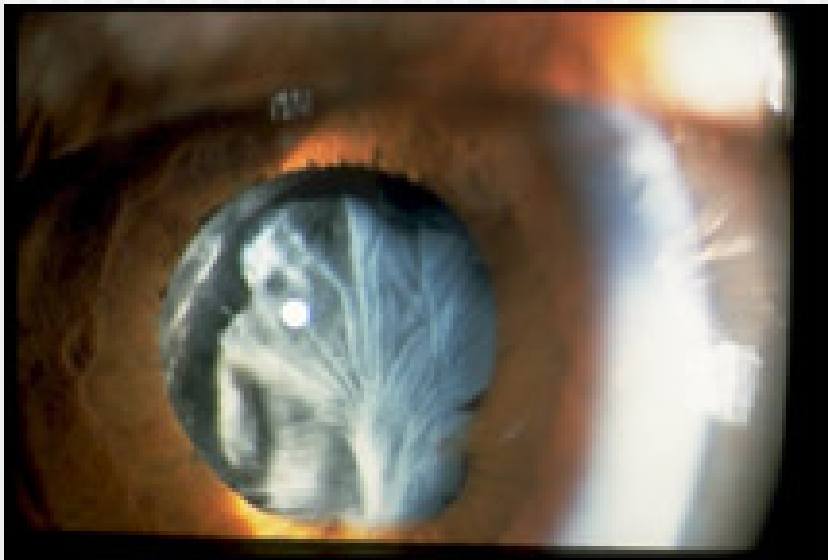


Complications of Systemic JIA

- Macrophage activation Syndrome (MAS)
 - High fever
 - Pancytopenia
 - Hepatosplenomegaly
 - Encephalopathy
 - DIC
 - Low ESR
- Possibly triggered by viral infection, NSAID, MTX, biologics.



Complications of JIA



Uveitis

- non-granulomatous chronic inflammation of anterior chamber
- **80% asymptomatic**
- Much increased risk in Oligo-JIA and ANA positive
- Screen annually in systemic JIA
- **Chronic uveitis can result in corneal clouding, cataract, glaucoma, vision loss**

Complications (cont.)

Growth Delay

- 2° to malnutrition and increased catabolic demands
- Steroids cause suppression of osteoblasts
- Overall growth retardation
- Local growth disturbances
- GH as treatment



TREATMENT OPTIONS

- **Multidisciplinary approach: MD's, PT, OT, SW, Nutrition**
- **NSAIDS: non-DMARD. Average time to improvement 1 month**
- **DMARD's: retard radiological progression of the disease.
Methotrexate/Sulfasalazine**
- **Biologics: TNF-antagonists, B-cell depleters**
- **Steroids: Provide symptomatic relief while other agents are initiated. Lots of adverse reactions**
- **Last resort: Joint replacement, Autologous Stem Cell Transplant**

Resources:

- Goldmuntz, E., White, P. Juvenile Idiopathic Arthritis: A review for the Pediatrician. *Pediatrics in Review* 2006;27: e24-e32.
- Weiss, J., Ilowite, N. Juvenile Idiopathic Arthritis. *Pediatric Clinics of North America*. (April 2005) Vol 52; No2; 413-442.
- Behrman: Nelson Textbook of Pediatrics, 17th ed., Copyright 2004. Saunders.
- Ravelli, A., Magni-Manzoni, S., Pistorio, A., *et al.* Preliminary Guidelines for macrophage activation syndrome complicating systemic juvenile idiopathic arthritis. *Journal of Pediatrics*. (2005) May; 146(5): 598-604.
- Bechtold, S., Ripperger, P., *et al.* Growth Hormone Improves Height in Patients with JIA: 4 year data of a controlled Study. *Journal of Pediatrics*. (2003) October; 143(4): 512-19.