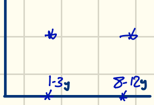
Clinical Dx.

2 peaks
♀ > ♂



JIA
Juvenile idiopathic arthritis

Several different forms of Chronic arthritis in children.
(joint effusion + · limitation of ROM · tenderness · hotness)

* JIA → ≥ 1 joint at least 6 ws in child < 16 ys.

* Diagnosis of exclusion.
Exclude ⇒ Infections, Malignancy, Trauma, Reactive arthritis, SLE & other CT dis.

* Multifactorial → Genetic, Environmental
+ psychological / trauma / Hormonal / infectious trigger

* لو ما تعالج منيح ⇐ significant morbidity
- leg length discrepancy
- joint contractures
- permenant joint destruction
- Blindness from chronic uveitis.

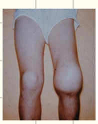
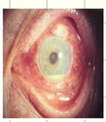
Pathology :-
• cell mediated + humoral (↑ autoantibodies, ↑ ANAs circulating Ig.)
• Chronic synovitis (· thickened · Hypervascular)
+ lymphocyte
+ cytokines [TNF-α, IL-6, IL-1, IL-17.]

Clinical picture :- (common symptom in all subtypes :-)
- joint swelling
- morning stiffness
- limbing
- pain
- spiking fever
- Rash
- eye involve (uveitis, iritis) Red eyes
- unspecific symptoms (lethargy, ↓ activity, Poor appetite.)

Oligoarthritis MC

- often mild
- 1-4 joints during 1st 6 m.
- medium → large sized joints
- asymmetrical ✗/✓

Good prognosis
10% → symmetric polyarthritis!

DDx:- trauma

mostly → no laboratory abnormalities.
70% → +ve ANA Do → SLE

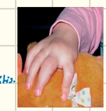
2nd MC Polyarthritis = pJIA
polyarticular juvenile idiopathic arthritis

≥ 5 joints in 1st 6 months
oligo ⇒ poly → [extended oligoarthritis]
Syst. inflammation > oligo أكثر من
(↑ESR CRP, malaise, low grade fever, anemia of chronic disease)

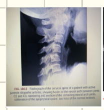
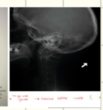
RF
absence → Seronegative (MC)
present → Seropositive

Seronegative		Seropositive
✓ or ✗	symmetrical?	✓
1-4 / 6-12	peak	9-12
✓	♀ > ♂	✓
chronic uveitis	extra-articular manifestation	rheumatoid nodules on extensors (elbow, achilles)

sausage finger (Dactylitis)
DDx:- psoriasis / SCD / arthritis.

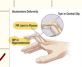
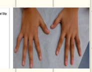


Long-term Complications:-

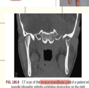



Cervical spine
- Subluxation
- Fusion
- loss of lordosis

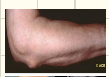
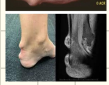
Boutonnière finger

Micrognathia
undersized jaw



TMJ involvement + loss of lordosis
⇓
intubation & anesthetic difficulty.

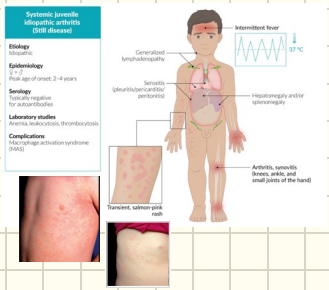
Systemic onset JIA = Juvenile form of Still's dis.

♀ = ♂ 2-4 yrs.

10% ⇒ preceding syst. inflammation ① > arthritis ②
6 wks – 6 mo
- polyarticular
- extensive & resistant to Tx.

Intermittent spiking fever
- ≥39
- ≥1 per day
- ≥2 weeks duration

- Salmon-pink macules
- Hepatosplenomegaly
- Generalized lymphadenopathy
- Serositis ↳ pleuritis, pericarditis



Dx of exclusion.

prognosis depend on arthritis severity

MAS macrophage activation syndrome

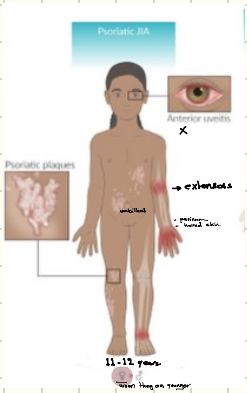
↳ mortality < 0.3% → + infections 2ry to ↓immunity

high fever / hepatosplenomegaly / ↓ Liver insufficiency / Neuro-abnormalities / bleeding diathesis

DIC

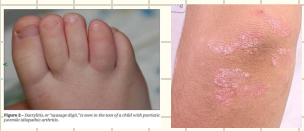
Physical findings	Bruising, purpura, mucosal bleeding Enlarged lymph nodes, enlarged liver and spleen
Laboratory findings	Elevated: AST, ALT, PT, PTT, fibrin degradation products, [ferritin, triglycerides] ⇐ liver insufficiency
	Decreased: white blood cell and platelet counts, erythrocyte sedimentation rate, fibrinogen, clotting factors ⇐ infiltration of BM
Bone marrow	Active phagocytosis by macrophages and histiocytes
Treatment	Intravenous glucocorticoid, cyclosporine

✓ corticosteroid or cyclosporine

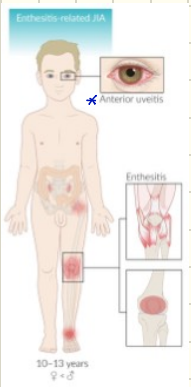


Psoriatic arthritis

arthritis + psoriatic rash or ≥ 2 of:
- dactylitis
- nail pitting
- onycholysis
- psoriasis in 1st degree relative



Enthesitis Related arthritis



Tenderness at the insertion of tendon, ligament, fascia to bone, joint capsule

♂ > 6 years.

linked to ⇒ • ankylosing spondylitis
• IBD.

RF / ANA -ve

HLA B27 +ve

asymmetric / lower limb

Achilles MC

Undifferentiated arthritis

arthritis that fulfils criteria in no category or ≥2 of above categories

Complications :-

Joint
- contractures ⇒ Loss of function
- bony fusions
- loss of joint space

Eye
- uveitis → blindness (cataract)

Long term
- anemia of chronic dis.
- permanent joint damage
- ↓ growth - disproportionate limbs
- pericarditis, pleuritis.

Table – Differential diagnosis of childhood arthritis

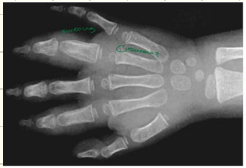
- **Reactive:** postviral, reactive arthritis, rheumatic fever, poststreptococcal arthritis
- **Inflammatory:** juvenile idiopathic arthritis, inflammatory bowel disease, sarcoidosis
- **Infection:** septic, osteomyelitis, Lyme disease, viral, bacterial sacroiliitis, diskitis
- **Systemic:** Kawasaki disease, Behçet disease, Henoch-Schönlein purpura, serum sickness, systemic lupus erythematosus, dermatomyositis, progressive systemic sclerosis
- **Malignancy:** leukemia, neuroblastoma, malignant bone tumors (osteosarcoma, Ewing sarcoma, rhabdosarcoma), benign bone tumors (osteoid osteoma, osteoblastoma)
- **Trauma:** accidental, nonaccidental, foreign-body synovitis

Poly + Syst → anemia of chronic dis.
leukocytosis
thrombocytosis.
↑ ESR + CRP

anti-CCP (cyclic citrullinated peptide) → early onset adult RA.
poor prognosis

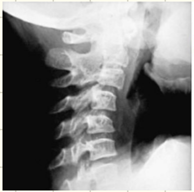
Synovial fluid analysis → WBC < 50k-100k (lymphocyte > neutrophil) → Suppurative arthritis.

> 50 → septic



Early(6mo duration) radiographic changes of JIA. Soft-tissue swelling and periosteal new bone formation appear adjacent to the 2nd and 4th proximal interphalangeal joints.

erosion → late.



Radiograph of the cervical spine of a patient with active JIA, showing:
1- fusion of the neural arch between joints C2 and C3,
2- narrowing and erosion of the remaining neural arch joints,
3- obliteration of the apophyseal space, and loss of the normal lordosis

* ما بقدر المريض يعمل complete extension.

Treatment

#Non pharmacological
Joint rehabilitation

① NSAIDs
- Naproxen
- ibuprofen
- indomethacin

② DMARD
- hydroxy chloroquine
- Sulphalazine.
- [methotrexate] drug of choice ⇉ poly, syst.
  - ↳ B.M supression.
  - hepatotoxicity

③ Biologic agent
risk of infection & malignancy.

∀ TNFα
- etanercept
- infliximab
- adalimumab.

④ IL-1 R antagonist — Anakinra. ✓syst

⑤ syst. steroids
- prednisone
- prednisolone.

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✓ in severe syst. with int. organ involvement
by bridging therapy.

Table 89-1 Features of Juvenile Idiopathic Arthritis Subgroups

FEATURE	OLIGOARTICULAR	POLYARTICULAR	SYSTEMIC ONSET
No. joints	<5	≥5	Varies, usually ≥5
Types of joints	Medium to large (also small in extended oligoarthritis)	Small to medium	Small to medium
Gender predominance	F > M (especially in younger children)	F > M	F = M
Systemic features	None	Some constitutional	Prominent
Eye disease	+++ (uveitis)	++ (uveitis)	+ (uveitis)
Extra-articular manifestations	None	None	Systemic features
ANA positivity	++	+	—
RF positivity		+ (in older children with early-onset RA)	
Outcomes	Excellent, >90% complete remission	Good, >50% complete remission, some risk of disability	Variable, depends on extent of arthritis

- **American College of Rheumatology (ACR) criteria for complete remission are as follows :**

- No inflammatory joint pain
- No morning stiffness
- No fatigue
- No synovitis
- No progression of damage, as determined in sequential radiographic examinations
- No elevation of the erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) levels