

# SEMINAR ON JIA

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# Juvenile idiopathic arthritis (JIA)

1. Definition
2. Etiology
3. Pathogenesis
4. Pathology
5. Classification
6. Clinical picture
7. Investigations
8. Differential diagnosis
9. Complications
10. Treatment
11. Prognosis



# Definition

- Juvenile idiopathic arthritis is not a single disease, but a term that includes all forms of arthritis that begin before the age of 16 years, persist for more than 6 weeks after exclusion of other causes of arthritis.



# Etiology

- Etiology of JIA is unknown.
- As with all autoimmune disease there is a complex interplay between genetic risk factors and environmental triggers.  
**Genetic:** MHC class II loci DR, DQ, and DP are particularly associated.
- Early childhood onset oligoarthritis associated with HLA A2, DR5, DR8, and DPB1\*0201
- In older boy oligoarthritis is associated with HLA-B27
- Polyarticular JIA with a positive rheumatoid factor is associated with HLA DR4  
**Environmental:** Possible external triggers include viruses (parvovirus B19, rubella, Epstein-Barr virus),  
This occurs due to:
- Host hyperreactivity to specific self antigens (type II collagen) and enhanced T-cell reactivity to bacterial proteins.

# PATHOGENESIS.

T-cell activation



B-cell activation



Complement consumption



Immune-complex formation



Release of (TNF)- $\alpha$ , IL-6, IL-1



severe articular and systemic disease.

# Pathology

- **The synovitis of JRA is characterized pathologically by**
  1. Villous hypertrophy
  2. Hyperplasia with hyperemia and edema of the subsynovial tissues.
  3. Vascular endothelial hyperplasia is prominent and characterized by infiltration of mononuclear and plasma cells .
  4. **Pannus formation:** which is an inflammatory exudate over the synovial lining, occurs in advanced uncontrolled disease and results in progressive erosion of articular cartilage and contiguous bone.

## Inflamed Joint

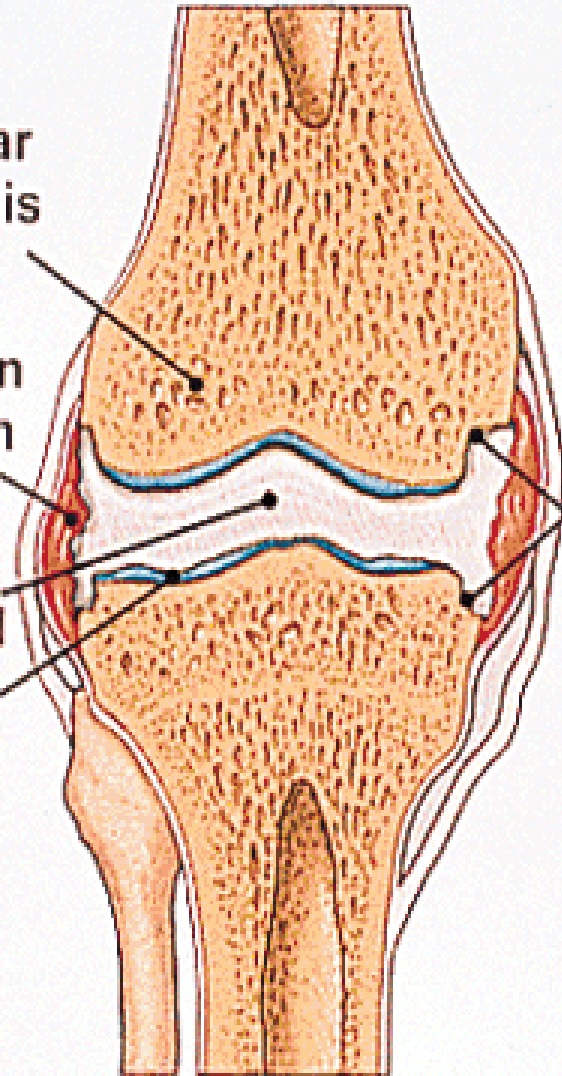
## Damaged Joint

Periarticular osteoporosis

Inflammation of synovium

Increased synovial fluid

Loss of cartilage



Degeneration of cartilage

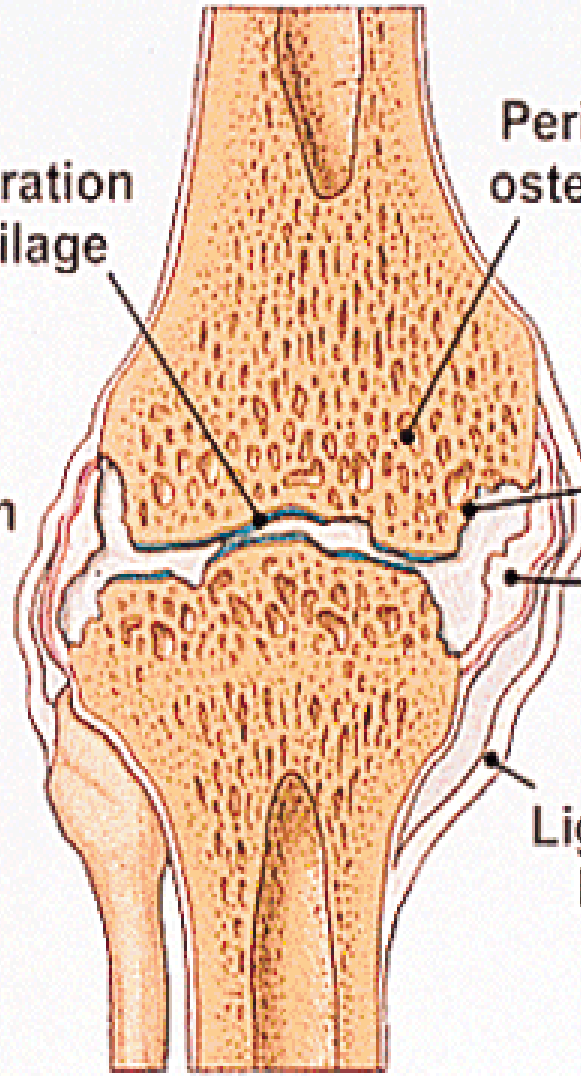
Erosion

Periarticular osteoporosis

Erosion of bone

Fibrosis of synovium

Ligament laxity



# Classification of juvenile Arthritis

- Juvenile rheumatoid arthritis(**ACR**)
- Juvenile chronic arthritis(**EULAR**)
- Juvenile idiopathic arthritis(**ILAR**)





# Classification

*The International League of Associations for Rheumatology (ILAR) has provided the most recent classification.*

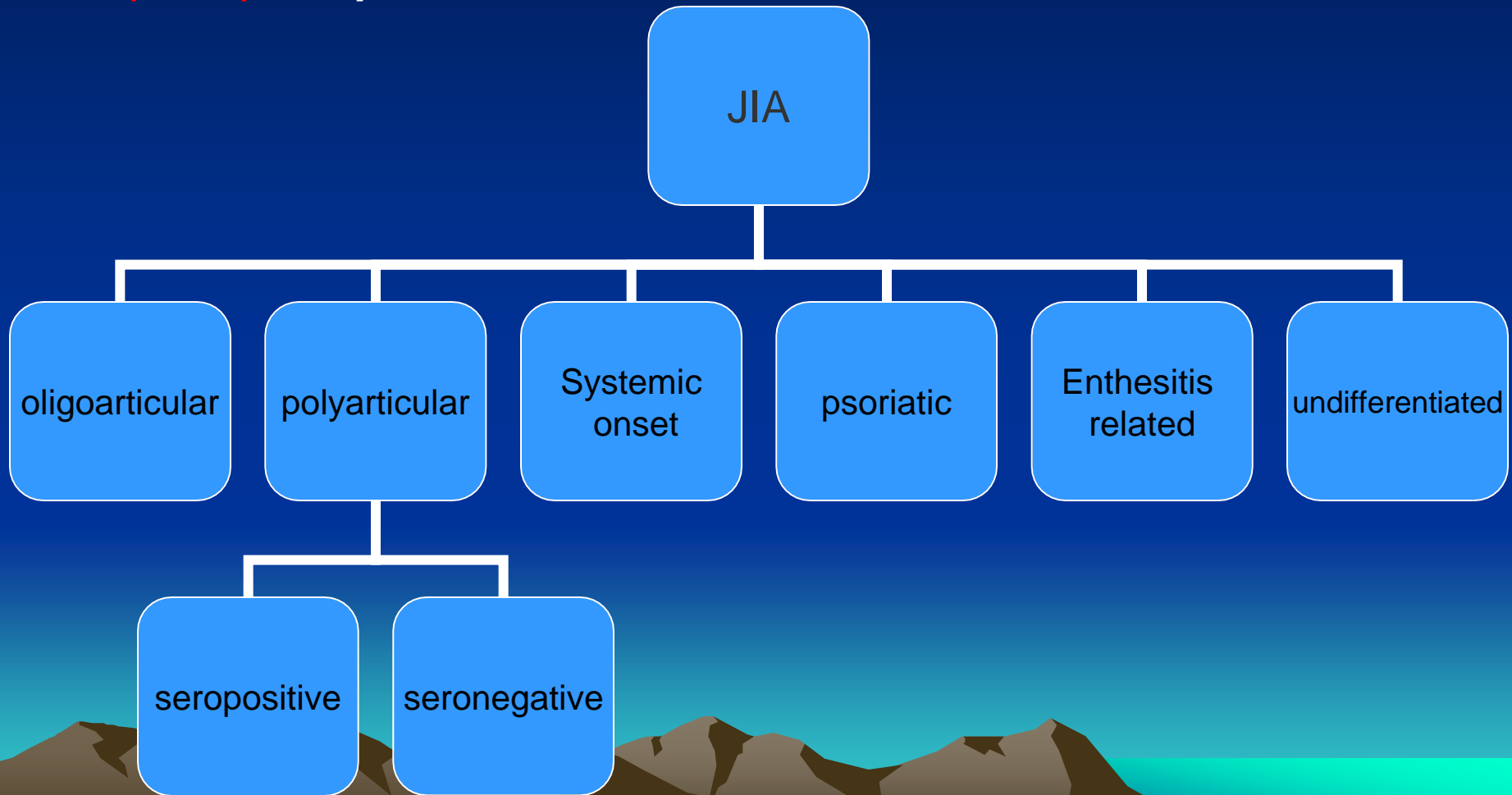


Table 11-1

## Comparison of Classifications of Childhood Arthritis

ACR <sup>1</sup>	EULAR <sup>2</sup>	ILAR <sup>3,4</sup>
Juvenile rheumatoid arthritis	Juvenile chronic arthritis	Juvenile idiopathic arthritis
Systemic	Systemic	Systemic
Polyarticular	Polyarticular JCA	Polyarticular RF-negative
	Juvenile rheumatoid arthritis	Polyarticular RF-positive
Pauciarticular	Pauciarticular	Oligoarticular
		Persistent
		Extended
	Juvenile psoriatic arthritis	Psoriatic arthritis
	Juvenile ankylosing spondylitis	Enthesitis-related arthritis
		Other arthritis

ACR, American College of Rheumatology; EULAR, European League Against Rheumatism; ILAR, International League of Associations for Rheumatology; RF, rheumatoid factor.

# Oligoarthritis

Age of onset: children 1-7 years

Gender prevalence: girls>boys

Articular symptoms:

- Morning stiffness
- Asymmetric
- Not more than 4 joints affected
- The affected joints include:

Knee most common one

Others: ankle, wrist, elbow

Hidden sites: atlantoaxial, TMJ

# Oligoarthritis cont...

- Presentation: Oligoarthritis in preschool children is often insidious, the child may limp especially after rest. the joint appears swollen and warm, but rarely painful until passively moved through its full range.
- Examination: of knee joint may reveal a palpable effusion.
- Hip joint involvement is rarely a presenting sign



# Oligoarthritis cont...

## Extrarticular symptoms:

- Iritis especially in ANA positive patients, it may be asymptomatic and identified at a screening slit lamp examination
- Chronic anterior non granulomatous uveitis and iridocyclitis

## Fate:

- a) Polyarticular JIA: If progress to involve 5 joints within first 6 months
- b) Extended oligoarticular: If progress to involve 5 joints more than 6 months after diagnosis.
- c) Persistent oligoarticular: If not progress.

# Polyarthrititis

- Definition: Arthritis of five or more joints within the first 6 months.
- Subdivided: according to presence of RF.
  - a)RF positive: RF positive disease rare but Equivalent to 'adult' rheumatoid arthritis.
  - b)RF negative.



# Polyarthritis cont...

## Seropositive:-

Age of onset: children 8-12 years

Gender prevalence: girls>boys

Articular symptoms:

- Symmetric
- Affect small joints of hands and feet.
- It is more likely to be aggressive and erosive disease.
- Rheumatoid nodule occur in 5-10% of patients, on the extensor surfaces of the elbows and over the Achilles tendons

Extrarticular symptoms:

- Felty syndrom, Rheumatoid vasculitis, Rheumatoid lung disease.
- Iritis is uncommon

Fate:

It may continue into active disease in adulthood.

# Polyarthritis cont...

## Seronegative:-

Age of onset: occur in teenage

Gender prevalence: girls=boys

Articular symptoms:

- Symmetric or asymmetric
- Synovial swelling affects large joints knees, ankles.
- No erosive changes.
- Joints may be simply tender with loss of function without gross synovial swelling or palpable effusion( so-called 'dry synovitis').

Extrarticular symptoms:

- No associated extrarticular symptoms.
- Iritis is rare.

Fate:

The course is variable and transition to seropositive polyarticular can occur




# Systemic Arthritis(Still's disease)

Age of onset onset: at any age

Gender prevalence: girls=boys

Extrarticular symptoms:

1. The hallmark of SOJIA are daily spiking fever in quotidian or “rabbit ear” pattern.
  2. Skin rash characterized by being salmon coloured, macular, evanescent, appear on trunk and extremities, present only during fever.
  3. Hepatomegally and/ or splenomegaly
  4. Generalized lymphadenopathy: Occurs in majority and enlarged nodes are painless, rubbery and mobile.
- 

# Systemic arthritis cont...

5. Serositis: Pericardial effusion ( useful to aid diagnosis), Pleural effusion(may co-exist with Pericardial effusion )
6. Iritis is rare
7. Children appear acutely ill with significant fever,fatigue,painful myalgia.

## Articular symptoms:

- True arthritis may not manifest until months after onset of fever which may make early diagnosis difficult.
- The pattern of arthritis associated with systemic JIA is variable.
- Approximately one third develop a polyarticular course with joint destruction within 2 years of disease onset .





# Systemic arthritis.cont...

## Fate:

- Three patterns of disease progression have been described:
  - a) Monocyclic
  - b) Intermittent
  - c) Persistent disease with systemic and polyarticular phases.

## Prognostic factors

- a) Risk factors at disease onset:  
Hepatosplenomegaly , serositis and low serum albumin
- b) Later risk factors for an adverse outcome:  
Thrombocytosis, persistent fever or steroid dependancy at 6 months.

# JRA by the Type-of-Onset

<u>Characteristic</u>	<u>oligoarticular</u>	<u>Polyarticular</u>	<u>Systemic</u>
Gender (F:M)	(5:1)	(3:1)	(1:1)
# Joints	≤ 4	≥ 5	Variable
Age at onset	1-7 yr	8-12 years	Any age
Systemic involvement	Iritis, uveitis (++)	Felty syndrom , rheumatiod vasculitis	Fever, rash, HSM, lymph node enlargement
Chronic Uveitis	5-15%	5%	Rare
Prognosis	Excellent except for eyesight	Guarded to moderately good	Moderate to poor

# Psoriatic arthritis

Definition: Arthritis and psoriasis OR arthritis with at least two of :

- Dactylitis,
- Nail pitting
- Onycholysis
- Psoriasis in first degree relative.

Age of onset: 10 years

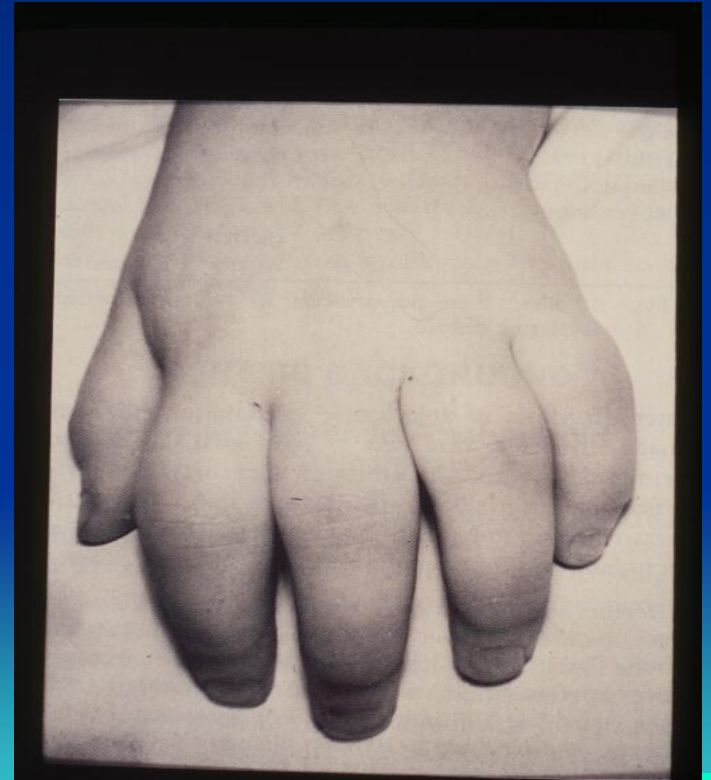
Gender prevalence: girls > boy

Articular symptoms:

1. Arthritis of one or more joints, asymmetrical involvement of large and small joints, it may be erosive

# Psoriatic arthritis cont...

2. Sacroilits
3. Distal interphalangeal joint synovitis.
4. Dactylitis: Dactylitis is a hallmark feature ( diffuse swelling of a finger or toe joint and periarticular tissue).
5. Sausage digits,nail pitting



# Psoriatic arthritis cont...

## Extrarticular symptoms:

- Psoriatic rash which can precede arthritis in 75% of cases.
- Acute anterior uveitis with photophobia and painful red eye.
- Chronic painless iritis more in patients with positive ANA.

## Fate:

Variable course with periods of remission with exacerbation, it may continue to active disease in adulthood





# Enthesitis-related Arthritis

**Definition:** Enthesitis refers to inflammation at site of attachment of tendon/ligament to bone, often affecting the insertion of Achilles tendon into the calcaneum and severely painful, causing significant functional impairment.

**Age of onset:** 8-12 years

**Gender prevalence:** boys > girls

**Type of patient:** It occurs in patient with juvenile onset spondylitis, reactive arthritis, arthritis associated with IBD.

# Enthesitis related arthritis cont...

## Articular symptoms:

- Synovitis in peripheral joints, mainly lower extremities.
- Asymmetrical distribution.
- Pain, stiffness, loss of flexibility in lumbar spine may occur in older children.
- Symptomatic sacroiliitis.

## Extrarticular symptoms:

- IBD is a common association.
- Heel pain, oral ulcers, acute painful uveitis.
- Rash including pyoderma gangrenosum.

## Fate:

Prognosis is quite good in these children if enthesitis is recognized and treated to reduce pain and resultant disability

# Undifferentiated Arthritis

- This category includes patients whose arthritis either doesn't fit into any of other categories or whose arthritis symptoms fit into more than one of these categories.

