Paediatric Rheumatology & Immunology

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Contributing Editor

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PAEDIATRIC RHEUMATIC DISORDERS

1  **Approach to Arthritis in Childhood**
4.7 50 Min video

2  **Juvenile Dermatomyositis**
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3  **Juvenile Idiopathic Arthritis**
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IMMUNOLOGY AND ALLERGIC DISORDERS

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APPROACH TO ARTHRITIS IN CHILDHOOD

Definition 00:00:35

Arthritis is characterized by intra-articular swelling or any two of the following :

- Pain or tenderness on moving the joint.
- Limitation of ROM.
- Warmth

Approach :

- Monoarthritis.
- Polyarthritis.

Monoarthritis - Acute & Chronic 00:01:30

- Acute : Duration < 3 wks.
- Chronic : > 6 wks.

Acute monoarthritis :

1. Causes :
   - Septic arthritis.
   - Reactive arthritis.
   - Hemarthrosis, traumatic joint effusion.
   - Tumors : Leukemia, bone tumors (malignant like Ewings or osteosarcoma; benign like osteoid osteoma).
2. Approach to well child :

With h/o significant trauma	No h/o significant trauma
• Fracture.	• Hemarthrosis • Neoplasia. • Regional pain syndromes : Osteochondrosis, slipped capital femoral epiphysis.

Active space

3. Approach to sick child :

Fever present	No fever
• Septic arthritis. • Part of systemic infection : Leptospirosis, HBV, mycoplasma, Chikungunya. • Vaculitis : Kawasaki, HSP. • Post infective : Post strept reactive arthritis, acute rheumatic fever.	• Partially treated septic arthritis. • Neoplasia.

Chronic monoarthritis :

Sick child	Well child
• Septic arthritis. • Infections : TB, lymes disease, brucellosis. • Vasculitis : PAN, microscopic polyangitis. • SLE. • Other chronic diseases : IBD.	• Juvenile idipathic arthritis (JIA) : Oligoarticular JIA (mc), enthesitis related JIA, psoriatic JIA. • Pigmented villonodular synovitis.

Polyarthritis 00:14:27

With fever	Without fever
• JIA : Polyarticular JIA, systemic JIA. • Reactive arthritis. • Vasculitis. • Sarcoidosis.	• JIA : Enthesitis related JIA. • Dermatomyositis. • Immunodeficiencies. • Malignancy.

Septic Arthritis 00:17:24

- mc organism : S. aureus. Other organisms : Streptococci, Pseudomonas, E. Coli.
- Features : Prominent localizing features will be present.
- most consistent sign : Pain on passive movement of joint.

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KOCHER's criteria for septic arthritis of hip :

1. Fever > 38°C.
2. Inability to bear weight.
3. WBC > 12,000/mm^3.
4. ESR > 40 mm/hr.
5. CRP +ve.

5th criteria is added by Children's hospital of Philadelphia.

Investigations :

- Usual investigations : Synovial fluid analysis + culture. Blood culture to rule out septicemia.
- Radiological investigations :
  - X-ray : Normal at early stages (unreliable).
  - USG can be useful.
  - Bone scan : It can detect changes within 2-3 days of disease onset. Disadvantage : High radiation exposure (not preferred).
- Gold standard imaging investigation : MRI.

Treatment of septic arthritis :

- Surgical debridement & removal of pus.
- IV antibiotics :
  - Initial antibiotics : IV 3rd generation Cephalosporins + Vancomycin (for MRSA).
    As the patient improves : Oral antibiotics.
  - Duration : 6 weeks.
- Steroids (Dexa) for 4 days : Rapid decrease in fever, decreases inflammation.

Transient Synovitis of Hip 00:26:26

- DD for septic arthritis.
- Seen in toddlers, upto 8 yrs of age.
- Mild condition : Well child; fever usually absent.
- C/F :
  - Initially URI → After 5-7 days, synovitis of hip.
  - Characteristic presentation : Painful limp.
- Mildly increased ESR.
- Responds well to NSAIDS x 2-3 wks.

Active space

Reactive Arthritis 00:29:17

- D/t enteric infection : Shigella, Salmonella, Campylobacter.
- Reiter's syndrome : Arthritis + urethritis + conjunctivitis.
- Usually mild course of arthritis that settles in few days to few weeks.
- If HLA B27 +ve or family h/o spondyloarthropathy : Prolonged course of illness.

Post-infective conditions :

- Post streptococcal reactive arthritis (PSRA).
- Acute rheumatic fever (ARF).

	PSRA	ARF
Onset of arthritis after infection	1-2 weeks	2-3 weeks
Pattern	Additive	Fleeting/migratory
Duration	Few months	Days - week
NSAID response	Less/No response	Excellent
ESR elevation	Mild	Significant
Carditis association	Rare	Significant
Antibiotic prophylaxis	Needed for 1 yr	Needed for long time

Malignancy Associated Arthritis 00:34:55

- Can be acute or chronic.
- Can be monoarthritis or polyarthritis.

Malignancies associated :

- Blood : Leukemia.
- Bone : Ewing's sarcoma, osteosarcoma.

C/F :

- Sick child.
- Nocturnal pain.
- Arthritis on some joint, periarthritis in other joints.
  Periarthritis : Painful restriction of rotation of joints; but X-ray will be normal.

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- Associated features :
  - Generalized lymphadenopathy.
  - Hepatosplenomegaly.
  - Bone tenderness.
  - X-ray : Increased risk of lytic lesions of the bone.

Osteoarticular TB 00:38:30

- Presents as chronic form of monoarthritis.
- Occurs in 5% of children with extrapulmonary TB.

Pathogenesis : Direct invasion from an adjacent site of osteomyelitis.

Poncet's disease :

- Reactive/allergic phenomenon in TB.
- It is characterized by arthritis at a distant site from primary TB.
- Polyarticular in nature.

Osteoarticular TB - features & diagnosis :

- Chronic disease of insidious onset with a slow progression.
- Constitutional symptoms like fever, night sweat, weight loss. They are not reliable.
- Extreme pain : Unusual in JIA.

Investigations :

- Synovial fluid culture : Low yield.
- Specific test : PCR testing for M.TB.
- Synovial biopsy : Presence of caseating granulomas.

Pigmented Villonodular Synovitis 00:43:07

- Chronic arthritis.
- Benign synovial hypertrophic condition.
- F > M.

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- Recurrent in nature.
- C/F : Painless joint effusion.
- Synovial fluid analysis : Blood in joint space.
- Best IOC : MRI of joint shows hemosiderin deposits. It also shows post-gadolinium enhancement in MRI.

Synovial Fluid Characteristics 00:45:33

	Normal	Non-inflammatory	Inflammatory	Septic	Hemorrhagic (Hemarthrosis)
Clarity	Transparent	Transparent	Translucent	Opaque (pus)	Bloody
Colour	Clear	Yellow	Yellow	Dirty/ Yellow	Red
Viscosity	High	High	Low	Variable	Variable
WBC/mm^3	<200	200-2,000	2,000-10,000 (upto 100,000)	>80,000	200-2,000
PMNs%	<25%	< 25%	>50%	>75%	50-75%

JUVENILE DERMATOMYOSITIS

Introduction 00:00:11

Features :

most common inflammatory myositis in children.

Proximal muscle weakness and a characteristic rash.

more common in females.

Age group : 4 to 10 years.

Pathogenesis 00:00:47

Genetic :

- HLA predisposition : B8, DRB1*0301, DQA1*0501.

Environmental :

- Infection : Diarrhoea, pneumonia.

Clinical features 00:02:38

Muscle weakness seen in 90 to 100%.

Cutaneous manifestations seen in 85 to 100%.

Muscle weakness :

- Insidious onset.
- Progressive muscle weakness and pain.
- Predominantly proximal muscles (Neck flexors, hip flexors and shoulder girdle) affected.
- Presentation : Difficulty in walking, combing hair, climbing stairs.
- Gowers sign : Using hands while standing from a sitting posture.

----- Active space -----

- Other muscle groups include pharyngeal, palatal (Aspiration) and respiratory (Hypercarbia).

Cutaneous manifestations :

- In 50% cases, skin manifestation is the first symptom.
- In other 50%, it may appear either simultaneously or soon after the onset of muscle weakness.

Heliotrope rash :

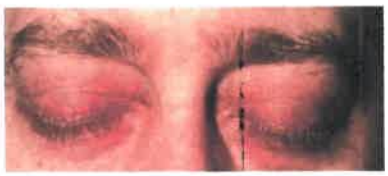
- Violaceous or blue-violet discolouration surrounding the eyelids with periorbital edema.

Facial erythema :

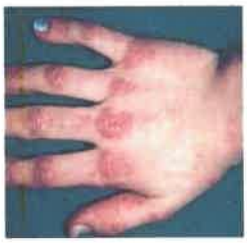
- Involves nasolabial fold.

Gottron papules :

- Bright pink, shiny, thick plaque over PIP/DIP joints.



Heliotrope rash.



Gottron papules.

Nail fold (Periungual) abnormalities :

- Capillary loops (Telengiectasia) which are tortuous/dilated.
- more readily seen under capillaroscopy or with a magnifier.

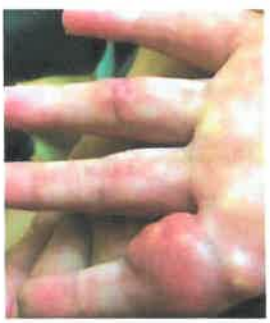
Other features :

Mechanic's hands :

- Thick, erythematous and scaly rash.
- Associated with anti-Jo I antibody.
- Photosensitivity in the neck and arm → Shawl sign.

In inadequately treated patients :

- Calcinosis cutis.
- Lipodystrophy.



Calcinosis cutis.

Diagnosis

00:13:04

----- Active space -----

Bohan and Peter's criteria (1975) :

Classical rash (Essential feature) : Heliotrope/gottron papules.

Any 3 of the following features :

- Weakness : Symmetric, proximal.
- Muscle enzyme elevation (≥1) : CK, AST, LDH, aldolase.
- EMG changes : Short, small, polyphasic potential/fibrillations/+ve sharp waves.
- Muscle biopsy : Necrosis/inflammation.

MRI :

- Current preferred investigation of choice.
- T2W MRI : Symmetrical muscle edema.

Labaratory findings :

Muscle enzymes :

- 20 to 40 times elevated.
- CK → Rises late and first to normalize.
- LDH : Best correlates with disease activity.

Auto-antibodies :

- m/c is ANA (80%).
- MAA (SS-A and SS-B) : Non specific.
- MSA (Anti Jo-1 and anti Mi-2) : Specific.

Treatment

00:19:06

First line : Steroids.

- methylprednisolone I/V 30 mg/kg/day for 3 to 5 days.
- Prednisolone 2 mg/kg/day for 4 weeks.

Steroid sparing agent : Methotrexate 15 to 20 mg/m²/week.

Maintenance therapy : Steroids 1 mg/kg/day tapered over 2 years.

Other measures :

- High-SPF sunscreen daily.
- Vitamin D and calcium supplements.

JUVENILE IDIOPATHIC ARTHRITIS

Definition 00:00:17

most common rheumatic disease in childhood.

Definition (ILAR) :

- Arthritis starts before 16 years & last for atleast 6 weeks.
- Unknown etiology.
- Diagnosis of exclusion.

Pathogenesis :

- Autoimmune disease.
- Target of autoimmune attack : Synovium.
- Affected synovium shows :
  1. Infiltrating inflammatory cells.
  2. Fibrosis.
  3. Increased vascularity.
- Both T cell and B cell mediated disease, but major damage is by T lymphocytes.
- T lymphocytes produce inflammatory mediators like cytokines and result in joint damage.
- B cells are involved in production of auto antibodies :
  1. Rheumatoid factor (RF).
  2. Anti CCP.
  3. Anti Nuclear Antibody (ANA).
- Strong evidence for a genetic predisposition (HLA DR8I, HLA DR4, HLA B27).
- Environmental factors like infection has also been proposed.

Arthritis in JIA :

- Intra articular swelling (OR) any two of the following :
  - Limitation in the range of movement.
  - Tenderness/pain on joint movements.
  - Warmth of joint.

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- Presentation :
  - Early morning stiffness.
  - May present with delayed walking as a manifestation.
  - Gelling phenomenon : Stiffness in joints following prolonged periods of inactivity.
  - Some present in the late stage with complications :
    1. Short stature (leg length discrepancy).
    2. Delayed puberty.
    3. Contractures.

Types

00:08:22

1. Oligoarticular JIA : ≤ 4 joints (10 %).
2. Polyarticular JIA : > 4 joints (10 %).
   - Rheumatoid factor(RF) positive JIA.
   - Rheumatoid factor(RF) negative JIA.
3. Systemic JIA (sJIA) (30 %).
4. Psoriatic JIA (5 %).
5. Enthesitis related JIA (m/c subtype in India : 40 %).
6. Others/unclassifiable (< 5 %).

Generally : females > males.

- Systemic JIA : females = males.
- Enthesitis related JIA : males > females .

Oligoarticular JIA :

- ≤ 4 joint affected in the first 6 months of disease.
- Subdivided into :

Persistent oligoarthritis	Extended oligoarthritis
Throughout disease ≤ 4 joints.	> 4 joints affected after 6 months of disease.
High chance of remission.	Chances of remission is low.
	Risk factors : ↑ ↑ ESR/ CRP, upper limb joints.

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- Joints affected : Lower limb joints (knee/ankle). Hip is uncommon.
- Extra articular findings : Chronic uveitis(asymptomatic).
- Serology : ANA positive (high risk of developing chronic uveitis).

Polyarticular JIA :

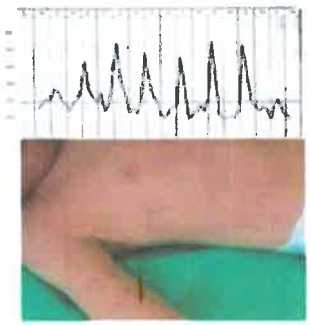
5 or more joints involved in the first 6 months of disease.

	RF negative	RF positive (Atleast 2 occasions 3 months apart)
Clinical picture	Large joints.	Large joints and small joints.
Genetic predisposition	HLA DR B1.	HLA DR 4.
age	Bimodal : 2 to 4 years/ adolescent.	9 to 12 years.
Symmetry	Asymmetrical involvement.	Symmetrical involvement.
Association	ANA positive → Chronic uveitis.	Systemic features (fever, rash, serositis). Rheumatoid nodules (subcutaneous nodules over extensor surfaces).

Systemic JIA

00:19:21

Clinical features :



- Arthritis :
  - Large + small joints.
- Fever :
  - At present /previous.
  - Atleast 2 weeks.
  - Daily intermittent fever (quotidian) documented (atleast 3 consecutive

Active space

days).

- Rash :
  - Salmon colored, macular rash.
  - Trunk, proximal extremities.
  - Evanescent : fever spike +.
  - Non pruritic rash
  - Migratory rash (< 1 hour).
  - Koebner's phenomenon : superficial trauma induces development of lesion.

Criteria for diagnosis :

1. Fever ≥ 2 weeks, quotidian in pattern (≥ 39° C ,atleast once a day and returns to ≤ 37 ° C), documented daily for ≥ 3 days.
2. Arthritis in ≥ 1 joint (for ≥ 6 weeks).
3. Any one of the following :
   - Evanescent erythematous rash.
   - Generalised lymph node enlargement.
   - Hepatomegaly and /or splenomegaly.
   - Serositis (pleuritis/pericarditis).

Complications :

MAS (macrophage activation syndrome) : 2° hemophagocytic syndrome/ Hemophagocytic lymphohistiocytosis (HLH).

Life threatening complication of systemic JIA.

Pathogenesis :

- Trigger unknown (infection) → Uncontrolled hyperactivation of macrophages → Increased cytokine production (IL 1 β, IL 6, IL 18, TNF α , IFN γ) : Cytokine storm → Hemophagocytosis (Bone marrow/ reticulo endothelial organs).

Suspicion :

- Change in pattern of fever (intermittent to continous) and rash (evanescent to persistent).
- Drop in cell counts (especially thrombocytopenia).
- Low ESR (decreased fibrinogen levels).

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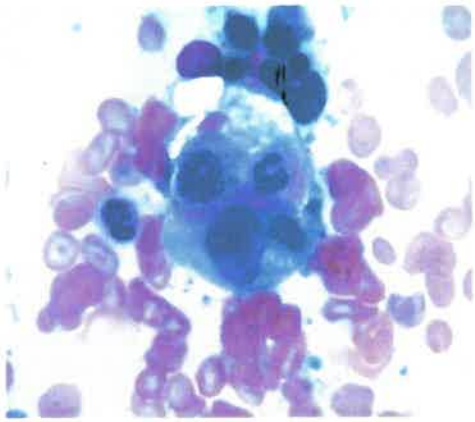
Criteria for diagnosis :

1. Fever (persistent).
2. Increased ferritin : > 684 ng/mL.
3. And any one of the following criteria :
   - Platelet count : < 181 x 10^9/L.
   - AST level : > 48 /L.
   - Triglyceride level : > 156 mg/dL.
   - Fibrinogen level : < 360 mg/dL.

Bone marrow biopsy : Hemophagocytosis.

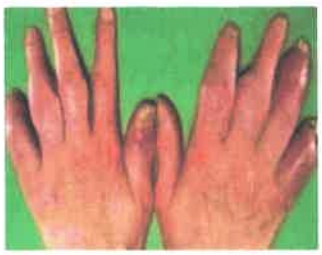


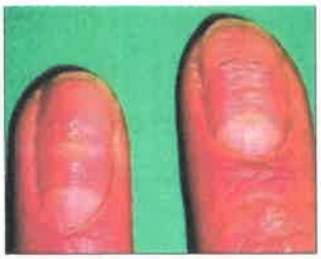
Psoriatic JIA 00:35:20

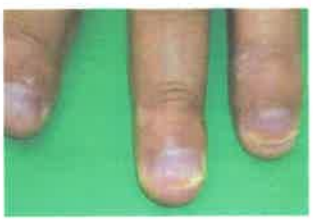
Psoriasis + arthritis /

Psoriasis (or) arthritis + any 2 of the following :

- Dactylitis : Sausage digits.
- Nail pitting or onycholysis.
- First degree relative with psoriasis.







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Enthesis related arthritis :

- most common type.
- Males > females.
- Affects large joints of lower limbs (Hip involvement common).
- HLA association : HLA B27 + → Spine involvement.
- Criteria for diagnosis :
- Arthritis or enthesitis of ≥ 6 weeks duration in children aged < 16 years, (or) arthritis or enthesitis + any two of the following :
    1. Sacroiliac tenderness and/or lumbosacral pain.
    2. HLA B27 positivity.
    3. Onset of arthritis in boys aged > 6 years.
    4. Acute anterior uveitis.
    5. Family history of HLA B 27 associated disease.

JIA associated uveitis :

- Associated with all types (rare in RF + polyarticular JIA and uncommon in sJIA).
- Acute or chronic type :
    1. Chronic : Asymptomatic
    2. Acute : Painful red eye (enthesitis related arthritis).

Uveitis may precede development of arthritis.

Screening :

- Slit lamp examination : Frequency : Every 2 months for first 6 months, later every 3-4 months. Continued till age of 12 years.

Investigations 00:43:01

JIA is a clinical diagnosis without any diagnostic laboratory tests.

Role of investigations :

1. Exclude alternative diagnosis.
2. Assess disease severity.
3. Asses complications of medications.

Blood counts :

- Anemia of chronic disease.
- ↑WBC, ↑Platelets.

Inflammation evidence :

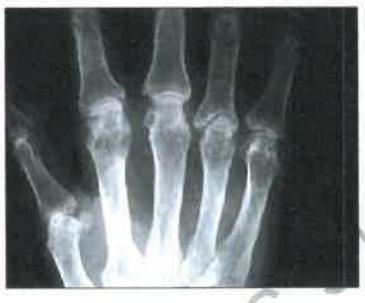
- ↑ ESR, CRP +ve.

Antibodies :

- ANA, RF, Anti CCP.

Radiological investigations :

- MRI : most sensitive/best investigation.
- USG : Synovial fluid aspiration.
- X rays :
  - Soft tissue swelling around joint.
  - Periarticular osteopenia.
  - Periosteal new bone apposition.



Differential Diagnosis 00:48:22

- Monoarticular presentation :
  1. Septic arthritis.
  2. Tubercular arthritis.
  3. Malignancy.
- Limp and isolated hip pathology :
  1. Perthe's disease.
  2. SCFE.
- Polyarticular presentation :
  1. Infections : streptococcal, mycoplasma, lyme's disease, leptospirosis.
  2. Systemic disorders : SLE (thrombocytopenia, anti ds DNA).
- migratory polyarthritis : Acute rheumatic fever.
- Systemic JIA :

Active space