Paediatric Rheumatology & Immunology

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Dr.Singaram

Contributing Editor

Dr. Singaram is MD in Paediatrics from Calicut Government Medical College and MBBS from PSG Institute, Coimbatore. His academic achievemen...

PAEDIATRIC RHEUMATIC DISORDERS

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Approach to Arthritis in Childhood
4.7 50 Min video

2. 

Juvenile Dermatomyositis
4.6 22 Min video

3. 

Juvenile Idiopathic Arthritis
4.5 55 Min video

4. 

Hemophilic Arthritis
4.7 28 Min video

IMMUNOLOGY AND ALLERGIC DISORDERS

5. 

Approach to a child with Allergic Disorder
4.4 19 Min video

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Allergic Rhinitis
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Atopic Dermatitis
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Urticaria, Angioedema and Anaphylaxis
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Primary Immunodeficiency Disorders
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Hematopoietic Stem Cell Transplantation
4.6 91 Min video

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

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- 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.

Active space

- 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%

Active space

JUVENILE DERMATOMYOSITIS

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

Introduction

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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

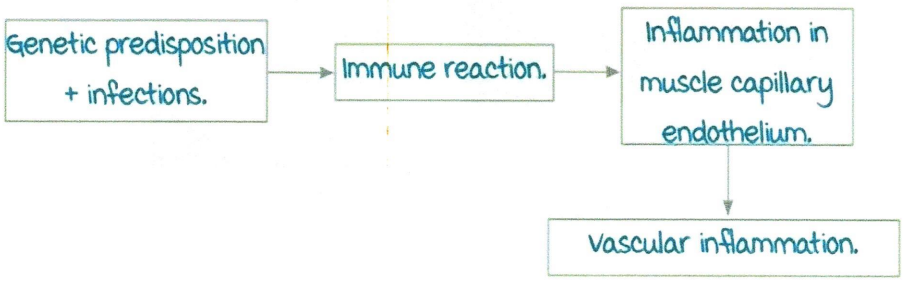
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Genetic :

- 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 :

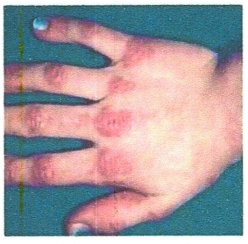
- Involves nasolabial fold.

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

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/m^2/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 DRB1, 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.

Active space