PEDIATRIC RHEUMATOLOGY

- JUVENILE IDIOPATHIC ARTHRITIS (JIA)
- SYSTEMIC LUPUS ERYTHEMATOSUS (SLE)
- HENOCH-SCHONLEIN PURPURA (HSP)
- KAWASAKI DISEASE (KD)

PEDIATRIC RHEUMATOLOGY

JUVENILE IDIOPATHIC ARTHRITIS (JIA)

Introduction

- Most common rheumatic diseases of childhood.

- It is characterized by an idiopathic synovitis of the peripheral joints, associated with soft tissue swelling and effusion.

Definition and Etiology

Definition

- Chronic arthritis that persists for a minimum of 6 consecutive weeks in one or more joints, commencing before the age of 6 years and after active exclusion of other causes.

Etiology

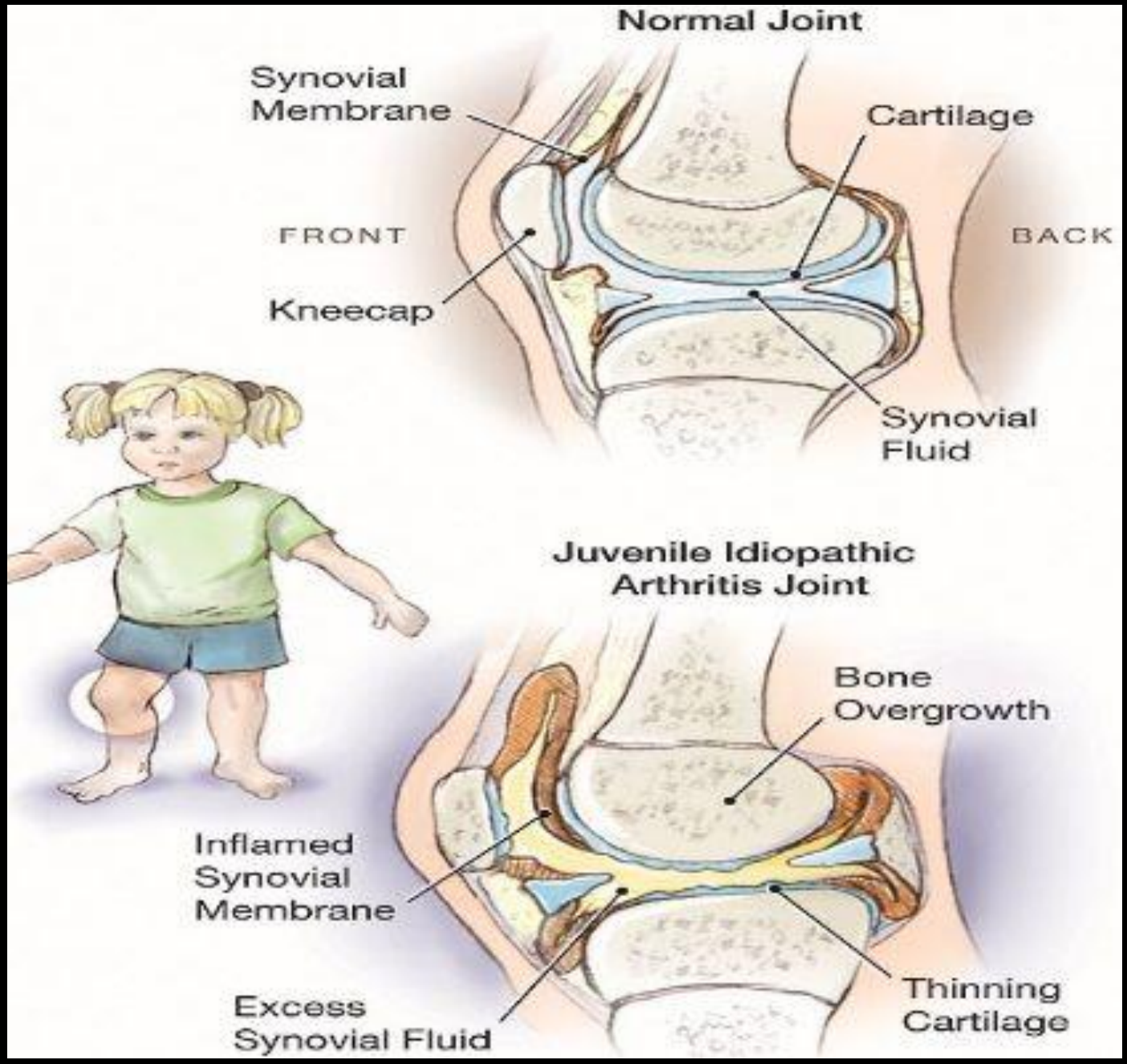
- The etiology of JIA is unknown. Two events are considered:
  - Immunogenetic susceptibility.
  - Environmental triggers.
- Certain viruses like parvovirus 819, rubella, Epstein-barr virus are also responsible

Epidemiology

- Although difficult to determine with precision, the incidence of JIA is approximately 13.9/100,000 children/year among children 15 years or younger, with an overall prevalence of approximately 113/100,000 children.

Pathogenesis

- Synovitis of JIA is characterized by villous hypertrophy and hyperplasia with edema of subsynovial tissue.
- Infiltration of mononuclear and plasma cells and pannus formation occur in advanced disease and results in progressive erosion of articular cartilage and bone.


Normal Joint
Synovial Membrane
Cartilage
FRONT
BACK
Kneecap
Synovial Fluid
Juvenile Idiopathic Arthritis Joint
Bone Overgrowth
Inflamed Synovial Membrane
Thinning Cartilage
Excess Synovial Fluid

Classification

- Systemic onset disease.
- Poly-articular onset.
  - Rheumatoid factor negative.
  - Rheumatoid factor positive.
- Oligo-articular onset.

Investigations

- There is no diagnostic test for JIA.
- ESR is high.
- Hemoglobin is low. There is normochromic normocytic anemia. White blood cell count is raised (neutrophil leucocytosis).
- Platelets are raised (> 400 x 10^6/L).
- In oligoarticular JIA, CBC and platelets are normal.

Investigations

- IgM rheumatoid factor is *negative but may be positive* in polyarticular onset disease.
- Anti-nuclear antibodies (ANA) are *negative but are* occasionally positive in polyarticular disease and frequently positive in oligoarticular disease.
- HLA-DR 4 is frequently present in RF-positive polyarticular disease and HLA-A2, DR5 and DRB are present in oligoarticular disease.
- Radiographically, there are early changes of affected joints.

Management

- The main *objectives of treatment are to:*
  - Restore function.
  - Relieve pain.
  - Maintain joint motion.
- Physiotherapy is important to maintain joint's mobility and muscle function. Range of motion and muscle strengthening exercise should be taught.

Management

- Bed rest is not needed except in the most acute stage.
- Non-steroidal anti-inflammatory drugs (NSAIDs) to control pain, inflammation and *fever.*
  - Aspirin 10-15 mg/kg/dose *every 6 hours.*
  - Naproxen 7.5 mg/kg twice daily.
  - Ibuprofen 10 mg/kg four times daily.
  - Diclofenac 1 mg/kg twice daily.
  - Celecoxib (new *selective cyclo-oxygenase-2* inhibitors).

Management

- Methotrexate is the second-line medication of choice especially for arthritis. Symptomatic response usually occurs within 3-4 weeks. Dose is 5-10 mg/m2/wk as a single dose.
- Corticosteroids are used in severe disease, either pulsed intravenous, single daily dose or given on alternate days.

Management

- Cyclosporin for systemic features.
- Splinting is used to prevent deformity.
- Surgical intervention such as replacement arthoplasties is often required.
- Frequent ophthalmological assessment (every 3-6 months) is needed to prevent and treat iridocyclitis.

Prognosis

- There may be exacerbations and remissions.
- There may be alteration in growth of affected limb.
- The younger the age of onset, the greater the risk of poor growth.
- Iridocyclitis is bilateral in 2/3 cases. Its prognosis depends on early detection and good management.

Systemic Onset Disease

- It is manifested by arthritis and prominent visceral involvement.
- There is high remittent fever and rash with one or more of the following:
  - Hepatomegaly
  - Splenomegaly
  - Generalized lymphadenopathy
  - Serositis (occasionally pericarditis)

Systemic Onset Disease

- Arthritis may be absent at the onset, but myalgia or arthralgia is usually present.
- Usually begins before 5 years of age, but can occur throughout childhood into adult life.
- It is present equally in boys and girls less than 5 years old. There is female predominance after 5 years of age.

Clinical features Systemic Onset Disease

- High once-daily spikes of fever for> 2 weeks.
- Myalgia.
- Arthralgia.
- Malaise.
- Rash-salmon pink or red maculopapular eruption.
- Lymphadenopathy-cervical, epitrochlear, axillary, and inguinal.
- Hepatosplenomegaly.
- Serositis (occasionally pericarditis).
- Hepatitis.
- Progressive anemia.
- Disseminated intra-vascular coagulation.
- Arthritis-knees, wrists and carpi, ankles and tarsi, neck, followed by other joints

Poly-Articular Disease

- Five or more joints are involved in onset period, usually somewhat insidiously and symmetrically.
- May be further divided by the presence or absence of IgM rheumatoid factor (RF).
  - RF-negative disease can occur at any age, occasionally before one year of age.
  - RF-positive disease 'occurs after 8 years of age.
- There is female predominance.

Clinical Features Poly-Articular Disease

- Poly-arthritis can affect any joint. Most commonly affected joints are the knees, wrists, ankles and proximal and distal inter-phalangeal joints of the hands.
- Metacarpo-phalangeal joints are often spared.
- Limitation of neck and temporo-mandibular movement is common. Cervical spine involvement increases the risk of atlantoaxial subluxation.

Clinical Features Poly-Articular Disease

- There is flexor tenosynovitis.
- Occasionally, there is low-grade fever.
- Occasionally there is mild lymphadenopathy and hepatosplenomegaly.
- In RF positive disease, there are rheumatoid nodules on the pressure points, particularly elbows.

Investigations Poly-Articular Disease

- ESR is raised.
- Hemoglobin may be low.
- There may be mild neutrophil leucocytosis.
- There is moderate thrombocytosis.
- IgM rheumatoid factor may be negative or positive.
- HLA DR4 is frequently present in RF- positive disease.
- ANA are occasionally positive.
- Radiographically there are early changes of affected joints, particularly of hands and feet.

Management Poly-Articular Disease

- Physiotherapy is very important to maintain and improve joint and muscle function.
- Splinting is used to prevent deformity.
- NSAIDs are used to control pain and inflammation.
- Methotrexate is very effective and can be used early to prevent deformity.
- Surgical intervention such as replacement arthroplasties is often required.

Pauciarticular Disease

- It is the commonest form with four or fewer joints involved, particularly knees and ankles.
- Usually occurs under 6 years of age.
- Girls are affected more than boys.

Clinical Features Pauciarticular Disease

- Mainly large joints (knee, ankle, elbow) are affected.
- Risk of chronic iridocyclitis in the first 5 years of disease is 1:3. This is ANA associated.
- If after 6 months assessment there are still four or fewer joints involved, it is called persistent oligoarticular disease.
- If more than four joints are involved, this is called extended oligoarticular disease.