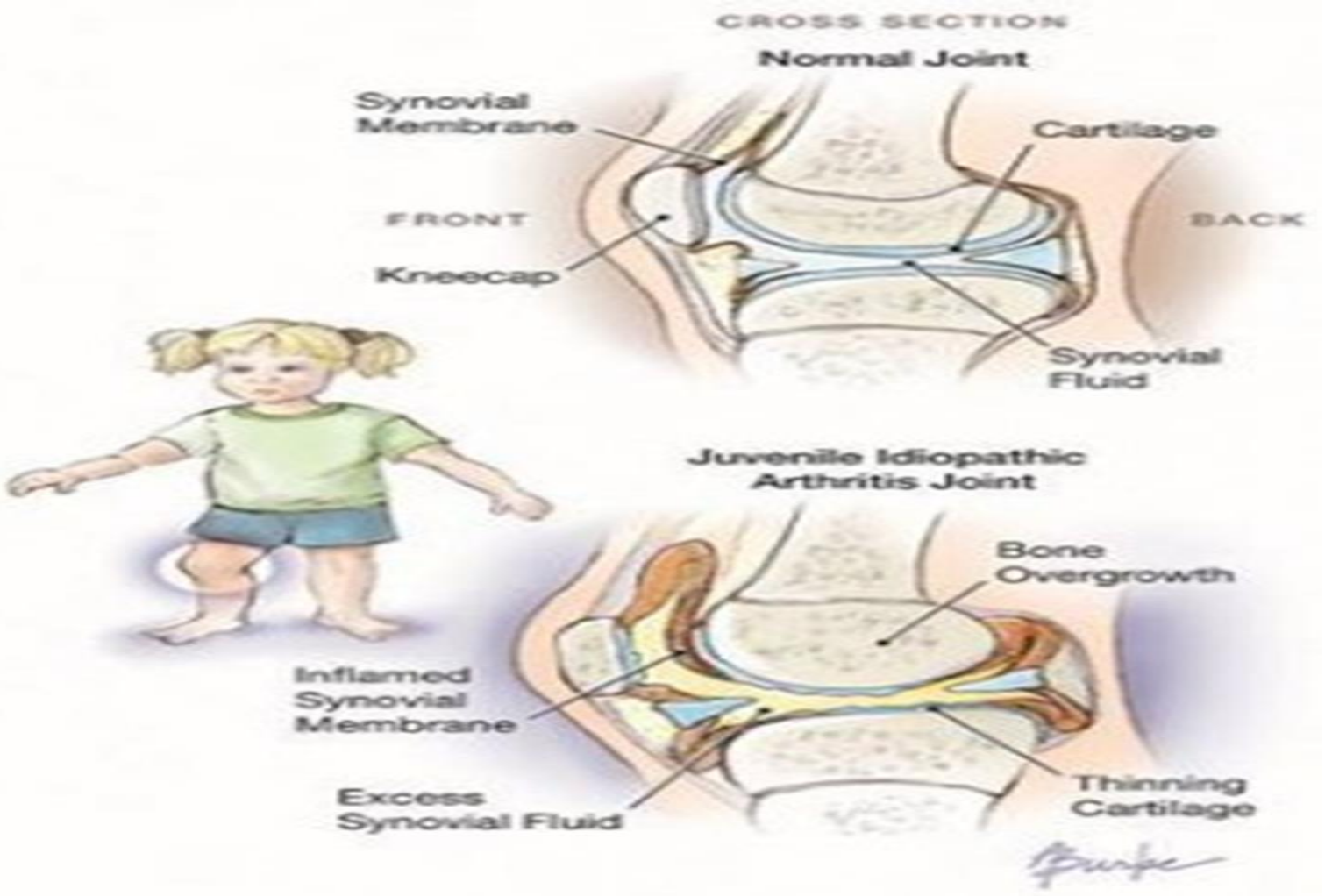
Juvenile Rheumatoid Arthritis

- Etiology:
- Pathogenesis
- Clinical manifestation
- Investigation
- Treatment
- Prognosis

- JIA is the most common rheumatic disease in childhood and a major cause of chronic disability.

- Etiology: Unknown, but may be due to immunogenetic susceptibility with an external trigger.

- Pathogenesis: JIA is an autoimmune disease associated with infiltration of mononuclear cells in the affected joint → villous hypertrophy & hyperplasia with hyperemia & edema of synovial tissue. Advanced uncontrolled disease leads to progressive erosion of articular cartilage and bone.


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

Clinical manifestation:

Initial symptoms may be subtle or acute :

- morning stiffness with limp or gelling after inactivity with easy fatigability and poor sleep quality.
- Involved joints are often:

i. Swollen
ii. Warm
iii. Painful on movement or palpation
iv. Reduced range of motion
v. Usually not erythematous

OLIGOARITHRITIS

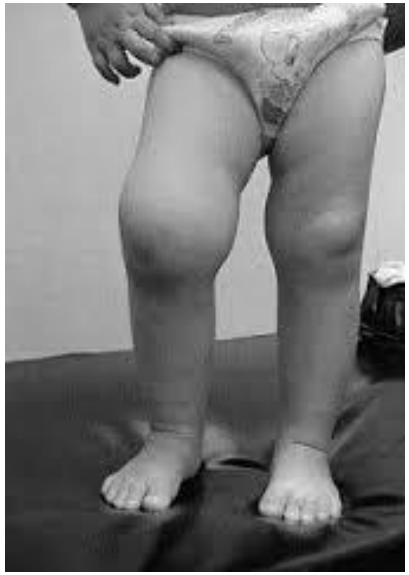
- ≤4 inflamed joints
- affect the large joints of the lower extremities e.g. knees and ankles
- hip is rare

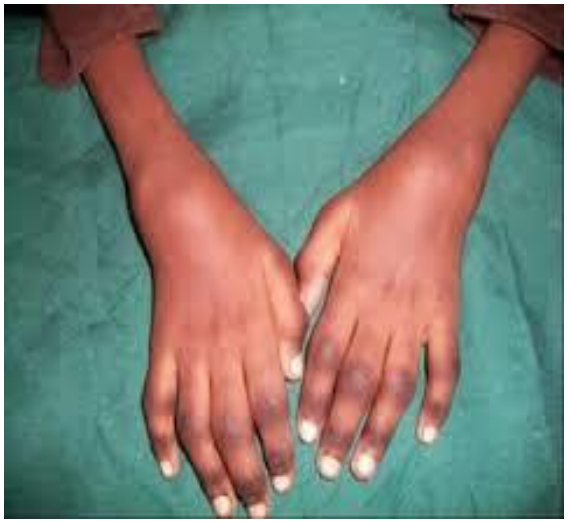
POLYARITHRITIS

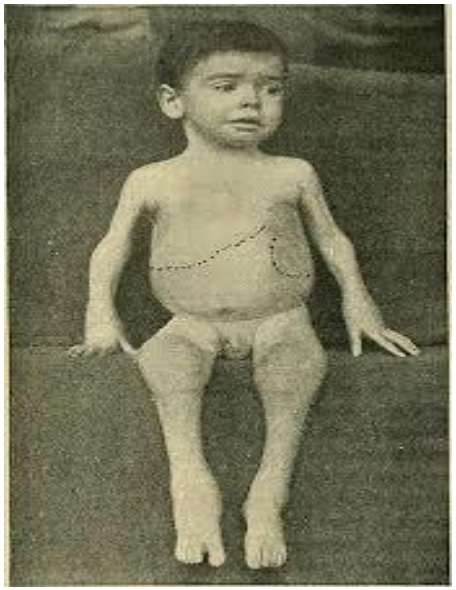
- ≥5 inflamed joints
- affect both upper and lower extremities.
- Micrognathia reflects chronic TM joint disease.
- Cervical spine involvement manifested as ↓ neck extension, with the risk of atlantoaxial subluxation and neurologic sequelae

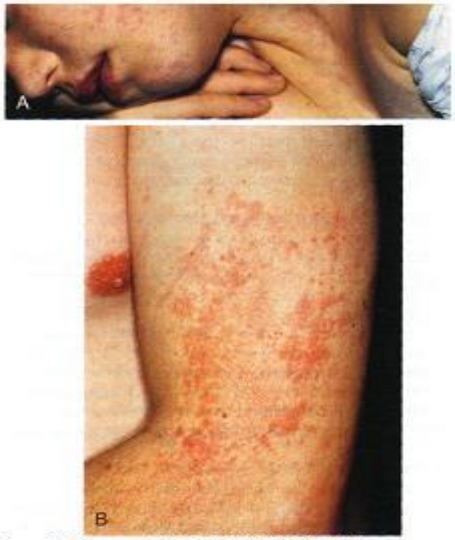
SYSTEMIC ONSET

- systemic manifestations e.g. fever, HSM, LAP, and serositis (pericarditis)
- present as FUO.
- The fever is ≥39 C & spiking, especially in evening, for at least 2 wk; it is accompanied by faint, erythematous, macular rash "Salmon-rash" which is nonpruritic, migratory, & lasting <1 hr.





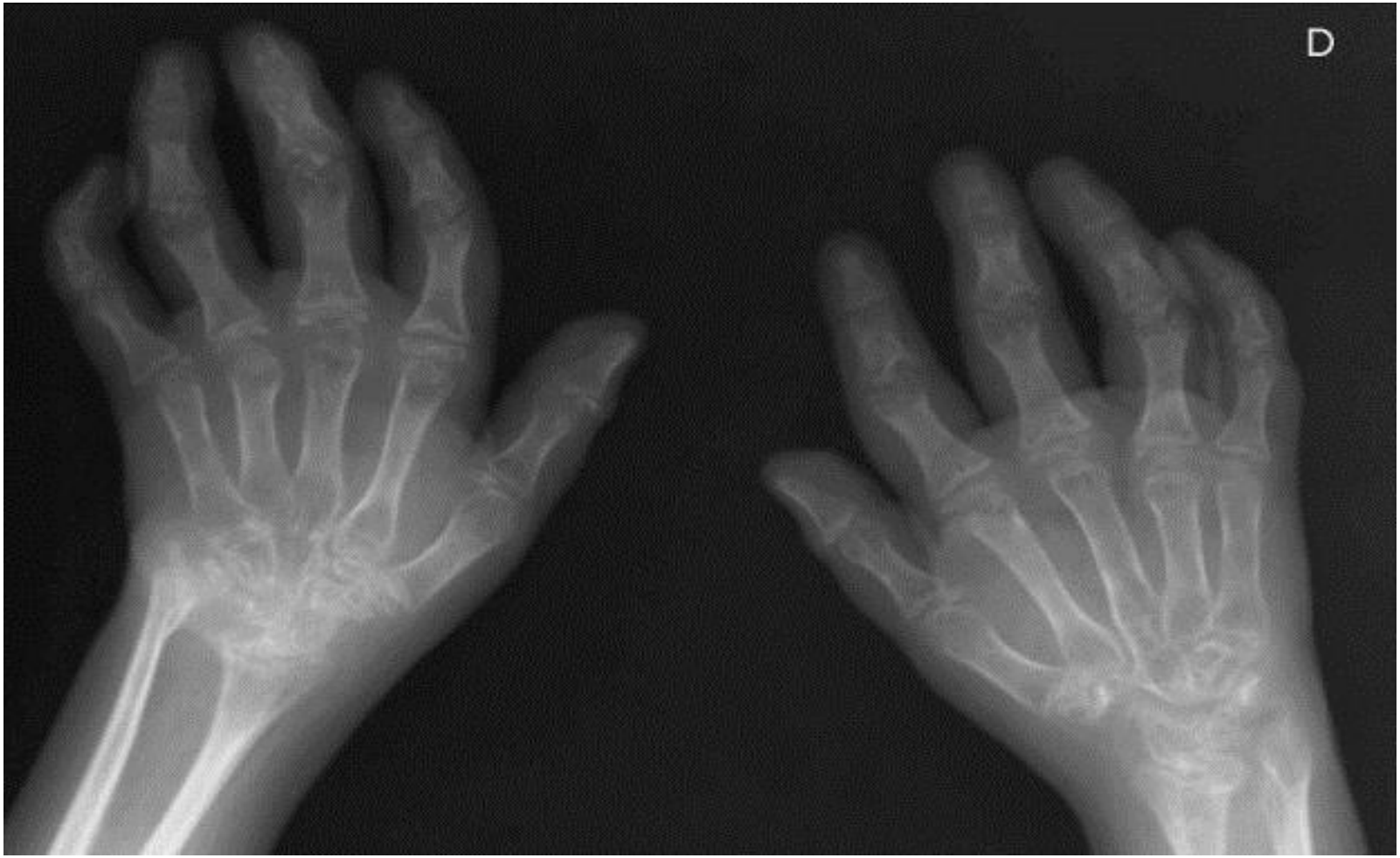




Juvenile Rheumatoid Arthritis Skin Rash 1

Invistigation:

- **X-ray of joints** in early disease shows soft tissue swelling, periarticular osteoporosis and periosteal new-bone apposition. Continued active disease may cause subchondral erosions & loss of cartilage with bony destruction.
- **MRI** is more sensitive to early changes than radiography.
- **CBP** show anemia of chronic disease, leukocytosis, & thrombocytosis.
- Inflammatory markers are ↑ e.g. **ESR, CRP**
- **ANA is +ve** in 40-85% of patients with oligo- & polyarticular arthritis; it is associated with ↑ risk for chronic uveitis
- **RF is +ve** in only 5-10% of patients with polyarticular arthritis which indicate a bad prognosis
- **Anti–Cyclic Citrullinated peptide (CCP) antibody**; it is similar to RF in that it is a marker of more aggressive disease



- Radiograph of the hands reveals joint space narrowing and erosions of the intercarpal joints, right worse than left.

Treatment:

- **NSAI agents** e.g. Naproxen, Ibuprofen.
- **Intra-articular injection of Corticosteroids**
- **Methotrexate** (which may take 6-12 wk for its effects), Sulfasalazine
- **Systemic corticosteroids** may be recommended for management of severe systemic illness or for control of uveitis (periodic slit lamp ophthalmologic examination of all pts is required to monitor asymptomatic uveitis.)
- **Dietary therapy** include: adequate intake of calcium, vit D, protein, and calories.

Note: Oligoarthritis is usually responding to NSAIs & IAI of corticosteroids, whereas Polyarthritis & Systemic-onset diseases are usually required MTX & other agents.

Prognosis:

- **Children with oligoarticular** disease esp girls with age at onset <6 yrs are at risk to develop chronic uveitis.
- **The child with polyarticular** disease often has a more prolonged course of active joint inflammation which requires early and aggressive therapy. Predictors of severe and persistent disease include: young age at onset, presence of RF or anti-CCP antibodies, rheumatoid nodules, and large numbers of affected joints.
- **Systemic-onset disease** is often the most difficult to control in both articular inflammation and systemic manifestations. Poorer prognosis is related to polyarticular distribution of arthritis, fever lasting >3 mo, and increased inflammatory markers (e.g. platelet count and ESR) for >6 mo

Thank you