JUVENILE IDIOPATHIC ARTHRITIS (Juvenile Rheumatoid Arthritis)

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.

Clinical manifestation:

Initial symptoms may be subtle or acute e.g. morning stiffness with limp or gelling after inactivity with easy fatigability and poor sleep quality. Involved joints are often swollen; warm, painful on movement or palpation with reduced range of motion but usually not erythematous

- Oligoarthritis [≤4 inflamed joints] is usually affect the large joints of the lower extremities e.g. knees and ankles, whereas the hip is rare.

- Polyarthritis [≥5 inflamed joints] is usually affecting both upper and lower extremities. Micrognathia reflects chronic temporomandibular joint disease. Cervical spine involvement manifested as ↓ neck extension, with the risk of atlantoaxial subluxation and neurologic sequelae.

- Systemic-onset disease: is characterized by arthritis with systemic manifestations e.g. fever, HSM, LAP, and serositis (e.g. pericarditis); or it may be initially present as FUO. The fever is $\geq 39^0$C & spiking, especially in evening, for at least 2 wk.; it is classically accompanied by the characterized faint, erythematous, macular rash called "Salmon-colored lesions" which is nonpruritic, migratory, & lasting <1 hr.

Investigations

- 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

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. There is no ass between severity of arthritis and 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.