‘J’ for JIA(Juvenile Idiopathic Arthritis)

What is JIA?

Juvenile idiopathic arthritis broadly refers to a group of heterogeneous diseases that share the common feature of chronic inflammatory arthritis of unknown cause lasting longer than 6 weeks with onset before 16 years of age. The frequency of occurrence differs but is estimated to be 1 in 1000 children.

The group of disorders under the term JIA except systemic JIA are considered as disorders of adaptive immunity occurring in genetically predisposed individuals whereas research suggest systemic JIA as a disorder of innate immunity and can be considered an autoinflammatory disease.

How is JIA classified?

Revised International League Against Rheumatism Classification (ILAR) Criteria for JIA Edmonton, 2001

JIA Type	**Diagnostic Criteria**
Systemic JIA 10-20%)	Fever of at least 2 weeks duration (daily for at least 3 days) and arthritis in one or more joints, plus one of the following: • Erythematous rash • Generalized lymph node enlargement • Hepatomegaly and/or splenomegaly • Serositis
Oligoarthritis (50-60%) Persistent & Extended	Arthritis affecting ≤ four joints during the first 6 months of the disease. If after 6 months more than four joints are involved the term extended oligoarthritis is used
Polyarthritis (20–30%) RF Negative	Arthritis affecting ≥ five joints during the first 6 months of the disease with rheumatoid factor negative
Polyarthritis (5–10%) RF Positive	Arthritis affecting ≥ five joints during the first 6 months of disease with rheumatoid factor positive on two occasions at least 3 months apart
Psoriatic arthritis (2-15%)	Arthritis and psoriasis or arthritis and at least two of the following:

	• Psoriasis in a first degree relative • Dactylitis • Nail pitting or onycholysis
Enthesitis -related arthritis (1–7%)	Arthritis and enthesitis** or arthritis or enthesitis with at least two of the following: • Presence/history of sacroiliac joint tenderness and/or inflammatory lumbosacral pain and HLA-B27 positive • Onset of arthritis in a male over 6 years of age • Acute (symptomatic) anterior uveitis • History of ankylosing spondylitis, enthesitis-related arthritis, sacroiliitis with inflammatory bowel disease or acute anterior uveitis in a first degree relative
Undifferentiated arthritis	a. Fits no other category b. Fits more than one categories

ILAR Exclusion criteria

a. Psoriasis or a history of psoriasis in the patient or first-degree relative.

b. Arthritis in an HLA-B27 positive male beginning after the 6th birthday.

c. Ankylosing spondylitis, enthesitis related arthritis, sacroiliitis with inflammatory bowel disease, Reiter's syndrome, or acute anterior uveitis, or a history of one of these disorders in a first-degree relative.

d. The presence of IgM rheumatoid factor on at least 2 occasions at least 3 months apart.

e. The presence of systemic JIA in the patient.

What are the clinical features of JIA?

While the presentation and evolution of each category of JIA differs from one another diagnosis is predominantly clenched with thorough history, clinical examination and pattern recognition of symptoms and signs (number and type of joints involved and constitutional and/or systemic features). The clinical features include arthralgia (pain, tenderness), arthritis (Pain, swelling, limitation of movement), morning stiffness or gelling, fever, rashes (systemic JIA), Uveitis (silent/chronic or acute/ painful red eye), extra articular features like hepatosplenomegaly, lymphadenopathy, growth retardation, anaemia.

Category	Age of Onset	Affected Joints	Systemic Features	Major Complications
Systemic	Throughout childhood	Any (Not necessarily at disease onset)	High fever, Rash, Polyserositis, Marked acute-phase response	Acute: Macrophage activation syndrome Chronic Growth disturbance, Amyloidosis
Oligoarticular persistent	Early childhood <6 yrs, ANA + in 60-65%, Girls>boys	Large joints, asymmetric (knee, ankle, wrist, elbow, temporomandibular, cervical spine)	No	Chronic/Silent uveitis- more in ANA + girls Local growth disturbances
Oligoarticular extended	Early childhood	Same as above, but more than four joints involved after the first 6 mo of disease	No	Chronic/Silent uveitis Local growth disturbances
Polyarticular RF negative	Throughout childhood	Symmetric polyarthritis with late onset Early onset: asymmetrical polyarthritis	Malaise	Chronic uveitis Local growth disturbances
Polyarticular RF positive	Adolescence Girls > Boys	Any but usually symmetric and involving small joints	Malaise	Local growth disturbances Articular damage
Psoriatic	Late childhood	Spine, lower extremities, distal interphalangeal joints, dactylitis	Skin manifestations of psoriasis +/-	Psoriasis Local growth disturbances
Enthesitis related	Late childhood	Spine, sacroiliac, lower extremities, thoracic cage joints	Inflammatory bowel disease	Acute symptomatic uveitis

How can we investigate further?

Often done to support the clinical diagnosis and look for complications.

- A complete blood count may demonstrate anemia of chronic inflammation and the C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) may show mild to modest elevations which may be used to guide therapy.
- In sJIA anemia, polymorphonuclear leukocytosis and thrombocytosis are classically seen.
- Rheumatoid factor (RF) should be performed in patients with polyarthritis as its presence helps to classify and prognosticate about severe erosive disease.
- The presence of a positive ANA identifies risk for the development of asymptomatic uveitis, particularly in those with OJIA, but should be performed in all children with JIA.
- Human leukocytic antigen (HLA) B27 should be tested in children who present with signs and symptoms consistent with ERA and can indicate susceptibility to the development of axial arthritis in future years.
- Plain X-rays may demonstrate effusions but erosions are rarely seen in childhood.
- Bedside ultrasonography is being increasingly used to evaluate joints at diagnosis and follow-up as a complement to clinical evaluation.

How is JIA managed?

Children with JIA are best managed in a specialist multi-disciplinary set up. While drug therapy is a crucial part of management, the role of the physiotherapist, occupational therapist, child psychologist and social worker is immense in supporting the physician to achieve maximum possible success.

Medication	Arthritis Subtype	Indication
NSAIDs (Naproxen, Ibuprofen preferred)	All types	Symptomatic: Pain, stiffness, serositis, antiinflammatory in mild cases
Intraarticular corticosteroids (Triamcinolone hexacetonide)	All types, mainly oligoarthritis	Injection of few active joints
Systemic corticosteroids	Systemic, polyarthritis	Fever, serositis, bridging medication, MAS
Methotrexate	All types; less effective for systemic and enthesitis-related axial disease	Disease modifying
Leflunomide	Polyarthritis	Disease modifying

Medication	Arthritis Subtype	Indication
Sulfasalazine	Oligoarthritis, polyarthritis, enthesitis-related peripheral disease	Disease modifying
Cyclosporine	Systemic	MAS
Thalidomide	Systemic	Biologic modifier
Anti-TNF (etanercept, infliximab, adalimumab, golimumab, certolizumab)	Polyarthritis, Enthesitis-related, uveitis (infliximab, adalimumab), less effective for systemic disease	Biologic modifier
Abatacept	Polyarthritis	Biologic modifier
Anti–IL-1 (anakinra, canakinumab, rilonacept)	Systemic	Biologic modifier, MAS
Anti–IL-6 (tocilizumab)	Systemic, polyarthritis	Biologic modifier
IVIG	Systemic	Steroid sparing, MAS

Take Home Messages

- **Age less,number of joints less , acute markers less- Juvenile oligo arthritis (ANA)**
- **Age more, number of joints more, acute markers more, upper and lower extremities- Juvenile Polyarthritis (RHEUMATOID FACTOR)**
- **Adolescent Boy ,lower extremities (Below the umbilicus joints), -Enthesitis RELATED ARTHRITIS(HlaB27)**
- **PUO(quotidian),SEROSITIS ORGANOMEGALY,RASH, ARTHRITIS,LNPATHY- Sjia(FERRITIN)**
- **DACTYLITIS of chronicity – Psoriatic Arthritis**

COMING UP NEXT - K for Kawasaki Disease….

*Dr Suma **BALAN***

Past Chairperson, PRSI, INDIA (2021-23)

Dr Vijay ViswANAthan

Chairperson PRSI, India (2023-25)

Dr Vighnesh PANdiArajan

Secretary PRSI, India (2023-25)

Dr Prajakta Joshi Ranade

Paediatric Rheumatologist

Sakra World Hospital,Bangalore

Dr UpendrA KiNjAwadekar

President CIAP (2023)