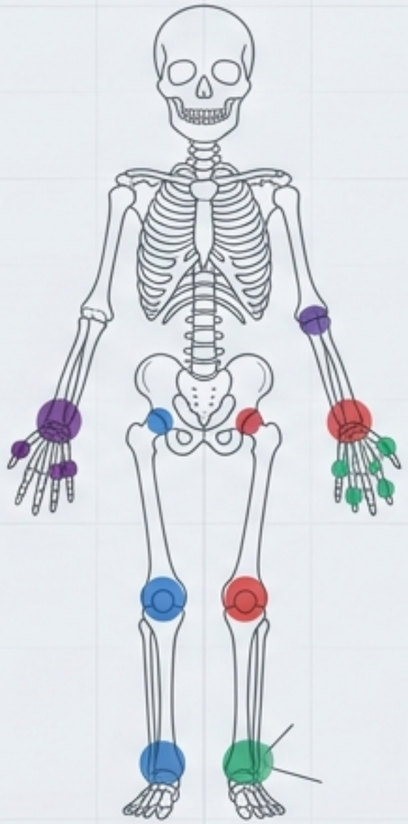
Evidence-Based Rapid Reference | Adapted from Med2Date Australian Guidelines



Juvenile Idiopathic Arthritis

A Clinical Blueprint for Diagnosis, Stratification, and Targeted Intervention

CORE DEFINITION

JIA is the most common chronic rheumatic disease in children.

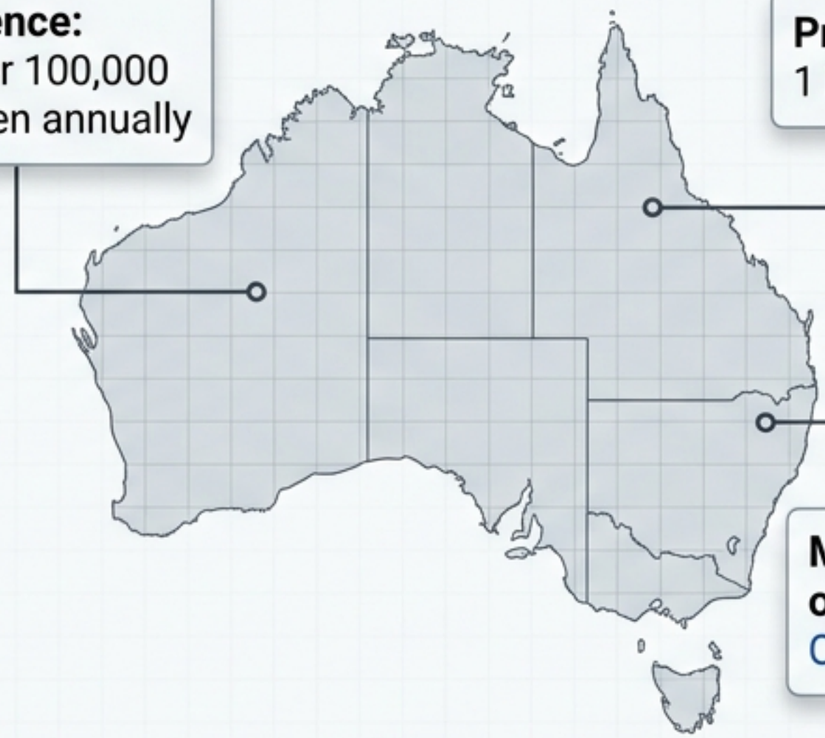
ILAR absolute diagnostic criteria
✔ Onset <16 years of age
✔ Arthritis in ≥1 joints
✔ Duration ≥6 weeks

⚠ **Diagnosis of exclusion:** Other known causes must be ruled out.

AUSTRALIAN EPIDEMIOLOGY

Incidence:
2-4 per 100,000 children annually

Prevalence:
1 in 1,000 children

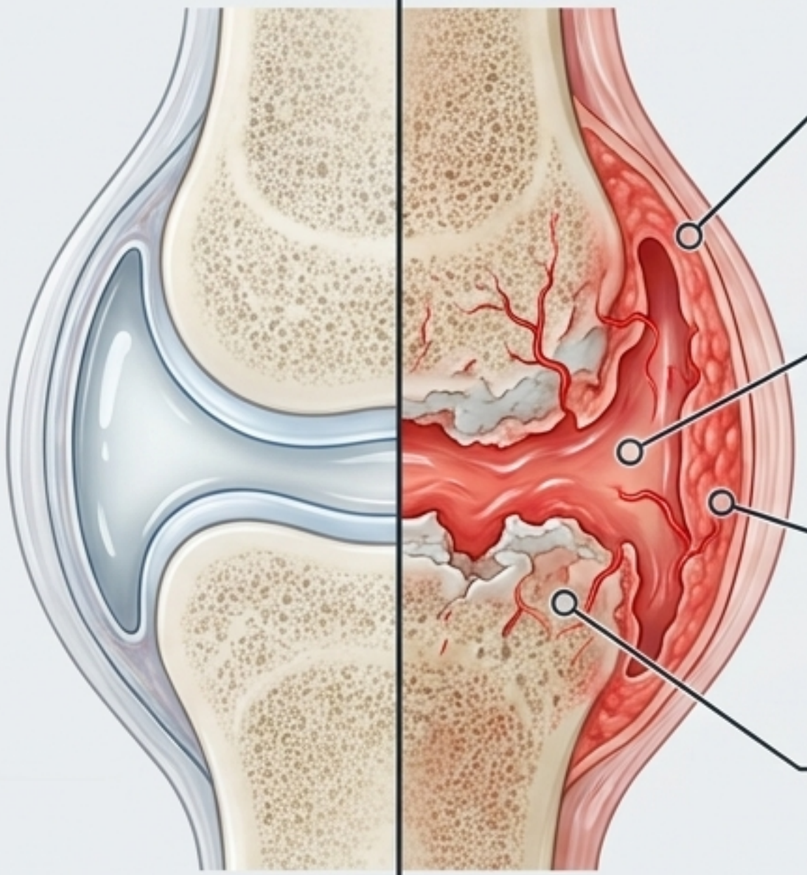


Most common overall subtype:
Oligoarticular JIA

Note: Higher incidence and greater disease severity noted in Aboriginal and Torres Strait Islander populations.

THE JOINT DESTRUCTION CASCADE

HEALTHY JOINT | ACTIVE PATHOLOGICAL CASCADE



STEP 1 (Infiltration): T-cells, B-cells, and macrophages breach the synovial membrane.

STEP 2 (The Storm): Pro-inflammatory cytokines (TNF-α, IL-1, IL-6, IL-17) flood the joint space.

STEP 3 (Hyperplasia): Synovial lining thickens with neovascularization.

STEP 4 (Destruction): Cytokines drive irreversible cartilage degradation and bone erosion.

KEY TAKEAWAY: EARLY, AGGRESSIVE DMARD THERAPY TARGETS THESE SPECIFIC CYTOKINES TO PREVENT IRREVERSIBLE PHYSICAL DESTRUCTION, GROWTH DISTURBANCE, AND PERMANENT DISABILITY.

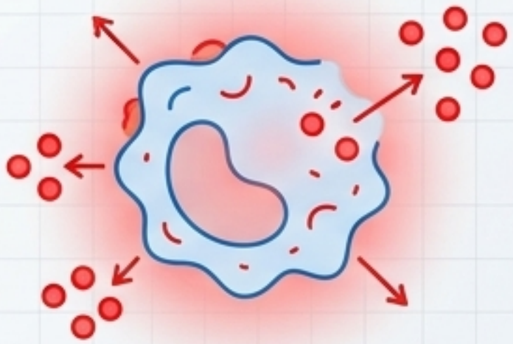
THE BIFURCATED PATHOLOGY

Autoinflammatory Pathway (Systemic JIA)

Mechanism: Prominent innate immune dysregulation.

Drivers: Massive overproduction of IL-1β and IL-6.

Clinical Translation: Drives systemic features (fever, rash) and responds specifically to direct cytokine blockade (Anakinra, Tocilizumab).

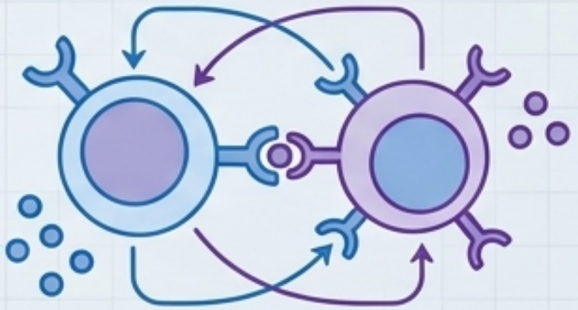


Autoimmune Pathway (Oligo/Poly JIA)

Mechanism: T-cell and B-cell driven immune failure.

Drivers: HLA associations (HLA-DR5, HLA-B27) + TNF-α and IL-17.

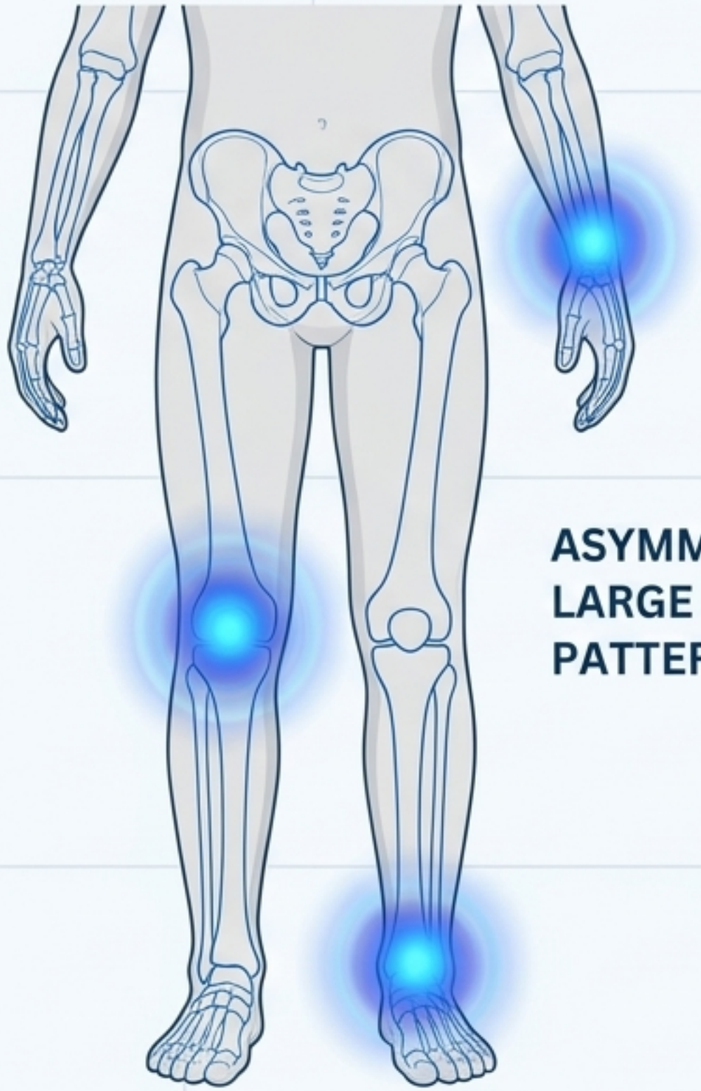
Clinical Translation: Drives localized joint damage and responds to broad immunosuppression (Methotrexate) or TNF inhibition (Etanercept).



THE JIA SUBTYPE DIAGNOSTIC MATRIX

	Oligoarticular	Polyarticular RF-	Polyarticular RF+	Systemic	ERA
Age of Onset	2-4 yrs	2-4 yrs & 6-12 yrs	≥9 yrs	1-5 yrs & late childhood	≥6 yrs
Sex Ratio	Strong Female	3:1 Female	9:1 Female	Equal	8:1 Male
Joint Pattern	Asymmetric large	Symmetric small/large	Symmetric small, erosive	Variable	Asymmetric lower limbs
Systemic Features	None	Mild	Mild	Quotidian fever, rash	None
Key Markers	ANA+ (60-80%)	ANA+ (40-50%)	RF+	Elevated Ferritin/CRP	HLA-B27+
Uveitis Risk	High, chronic silent	Moderate, chronic	Low	Rare	Moderate, acute

OLIGOARTICULAR JIA: CLINICAL BLUEPRINT



ASYMMETRIC LARGE JOINT PATTERN

CORE DEFINITION

≤4 joints in the first 6 months. Accounts for 50-60% of all cases. Peak onset 2-4 years (Female dominant).

SUB-CLASSIFICATIONS

- **Persistent:** Remains ≤4 joints.
- **Extended:** Progresses to >4 joints after 6 months (occurs in up to 50%).

CLINICAL CLUES

Painless limp, joint swelling, potential leg length discrepancy.

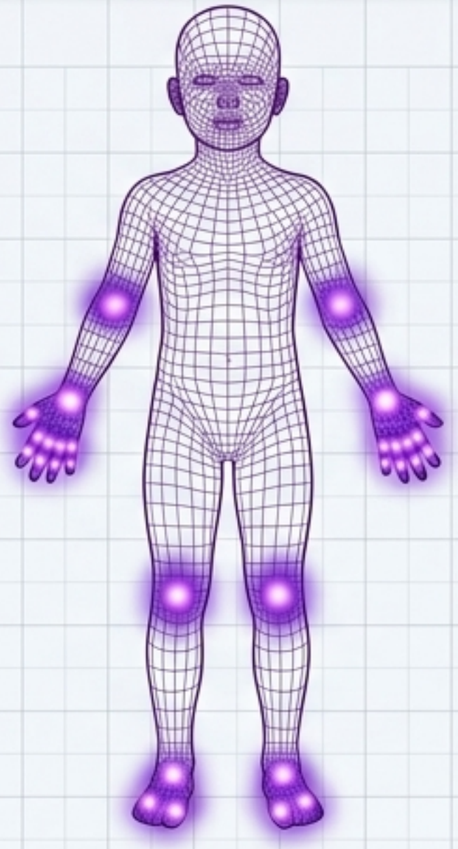
RED FLAG WARNING

Asymptomatic Chronic Anterior Uveitis occurs in up to 20% of patients. Highest risk in young, ANA-positive girls. Urgent ophthalmology review required.

POLYARTICULAR JIA: CLINICAL BLUEPRINT

CORE DEFINITION

≥5 joints in the first **6 months.**
Accounts for **20-30% of cases.**
Prominent **morning stiffness.**



RF-NEGATIVE

- **Bimodal onset** (2-4 yrs & 6-12 yrs)
- F:M ratio 3:1
- **Symmetric** small and large joints
- **Variable prognosis**
- **ANA+** in 40-50%

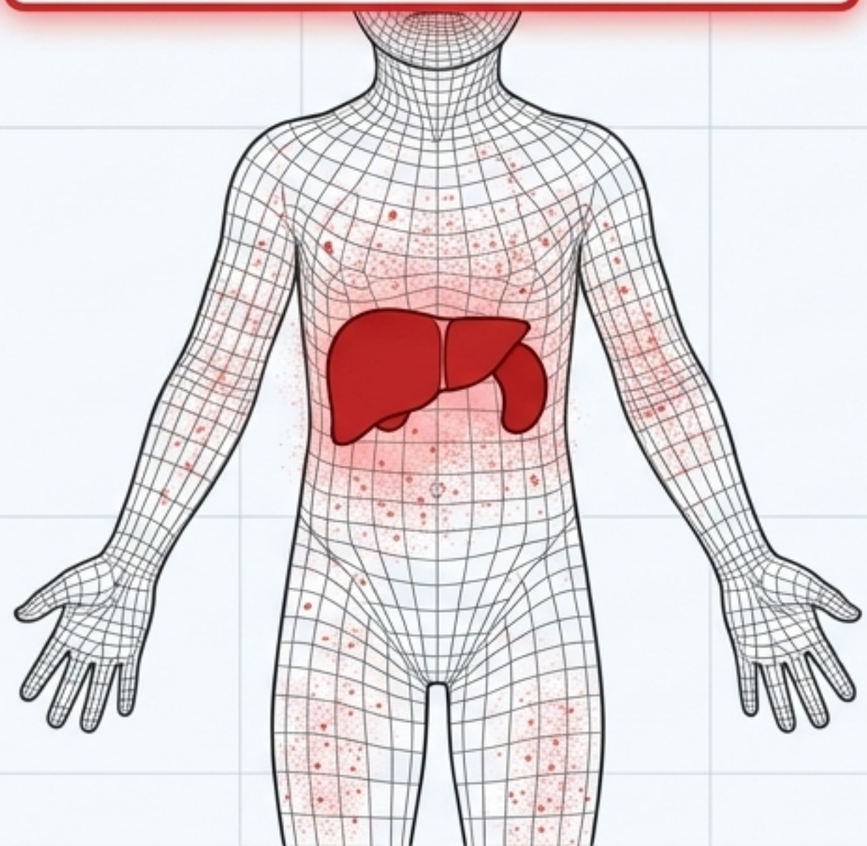
RF-POSITIVE

- **Late childhood/ adolescence** (≥9 yrs)
- F:M ratio 9:1
- **Prominent erosive disease in small joints** (hands/feet)
- Resembles **adult RA**
- **Highest risk of severe disability**

SYSTEMIC JIA: CLINICAL BLUEPRINT

CORE DEFINITION:

10-20% of cases. **Biphasic** age (1-5 yrs and re). Requires **arthritis** plus specific **systemic features**.



MAJOR CRITERIA 1

- **Quotidian Fever:** Daily spikes ≥39°C for ≥2 weeks, returning to baseline rapidly.

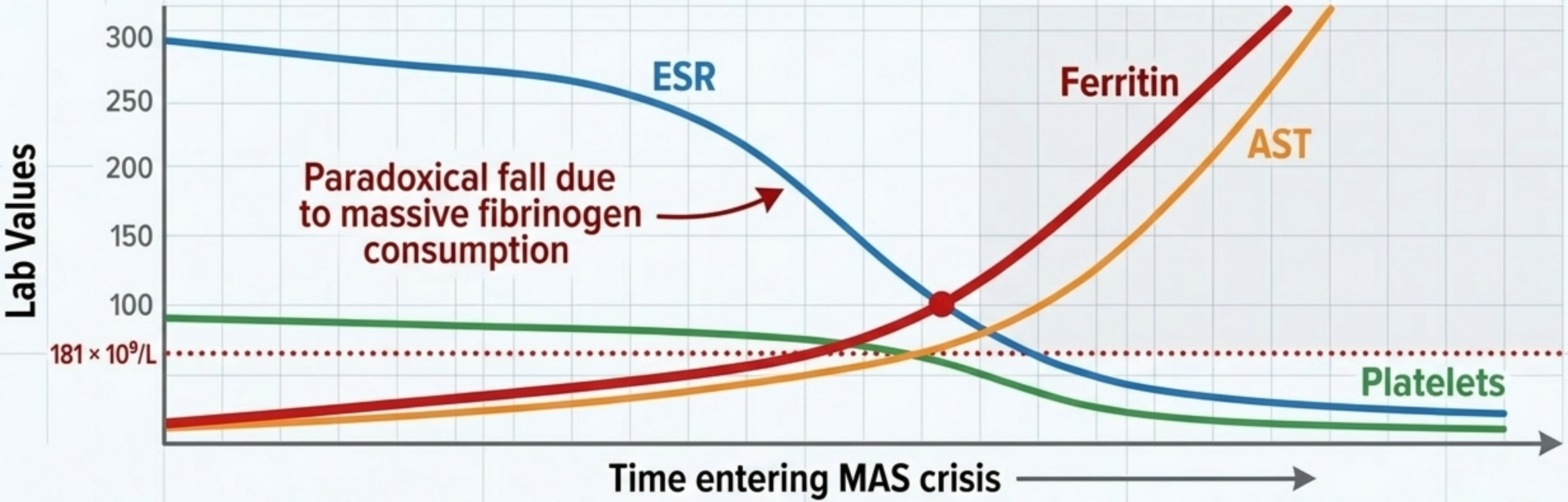
MAJOR CRITERIA 2

- **Evanescent Rash:** Salmon-pink, macular, non-pruritic on trunk/proximal limbs appearing with fever.

OTHER FEATURES

- **Serositis** (pericarditis/pleuritis), hepatosplenomegaly, generalized lymphadenopathy.

THE MAS TIPPING POINT



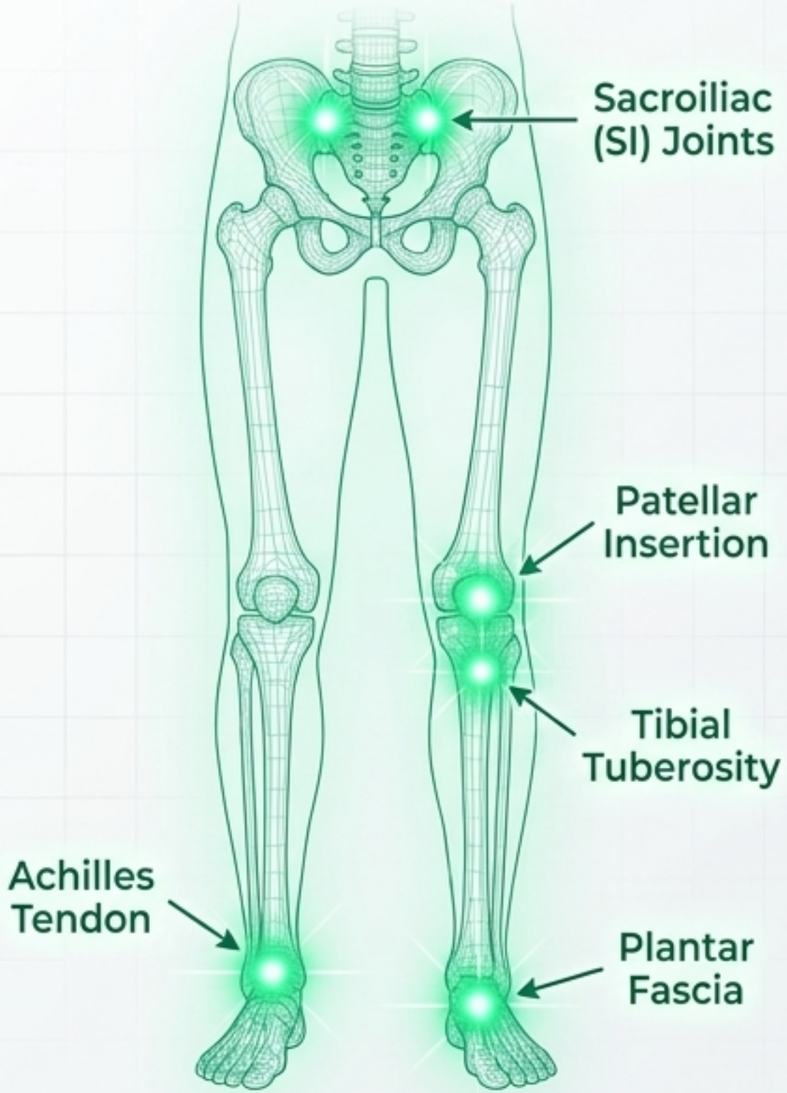
DIAGNOSTIC CRITERIA

- Triggered by flare/infection. Requires **hyperferritinaemia** (>684 ng/mL), **hypofibrinogenaemia** (<2.5 g/L), **falling ESR/platelets, elevated AST/LDH.**

IMMEDIATE ACTION

- Emergency **high-dose IV methylprednisolone** (30 mg/kg), **Ciclosporin** (PBS Auth), **Anakinra** (IL-1Ri, PBS Auth), ICU support.

ENTHESITIS-RELATED ARTHRITIS (ERA): CLINICAL BLUEPRINT



CORE PROFILE

- Part of the juvenile spondyloarthropathy spectrum. Predominantly male (8:1).

DIAGNOSTIC FEATURES

- Arthritis **AND** enthesitis, **OR** one of the two plus: SI joint tenderness, inflammatory spinal pain, HLA-B27+, male onset ≥6 years, or acute anterior uveitis.

KEY DISTINCTIONS

UVEITIS PATTERN

Acute, symptomatic, and unilateral (unlike the silent chronic uveitis in Oligo).

JOINT PATTERN

Asymmetric large lower limbs, high risk of axial progression (sacroiliitis).

Baseline Diagnostic Investigations

Essential Bloods

FBC & ESR/CRP (MBS 65070)

Identifies anemia of chronic disease, thrombocytosis, and baseline inflammation.

Immunology Markers

ANA (MBS 66580) - vital for uveitis risk.

RF (MBS 66586) - prognostic for Polyarticular.

HLA-B27 (MBS 66591) - supports ERA diagnosis.

MAS Screen (For Systemic JIA)

Ferritin, Fibrinogen, LDH, LFTs.

Imaging & Referrals

Slit-lamp biomicroscopy by Ophthalmologist (**Mandatory**).

Ultrasound for synovitis.

MRI (MBS 63001 series) for early sacroiliitis/osteitis.

Mandatory Uveitis Screening Schedule

Patient Variable	Screening Frequency
Oligo/Poly/Psoriatic JIA + ANA Positive + Age <7 years at onset	**Every 3 months**
Oligo/Poly/Psoriatic JIA + ANA Positive + Age ≥7 years at onset. **AND Enthesitis-Related Arthritis** (for acute uveitis)	**Every 6 months**
Systemic JIA (without ANA or other specific risk factors).	**Every 12 months**

Clinical Note: Screening must be done via slit-lamp by an ophthalmologist. Must continue for **4-7 years post-onset,** with many experts recommending lifelong screening. First-line treatment is topical corticosteroids.

Risk-Aligned Intervention

Lower Risk

Profile

Persistent Oligoarticular JIA. Few joints, no poor prognostic factors.

Setting

Rheumatology OPD (3-6 monthly review)

Approach

NSAIDs, Intra-articular corticosteroids (Triamcinolone hexacetonide)

Moderate Risk

Profile

Extended Oligoarticular, RF-Polyarticular, ERA

Setting

Rheumatology OPD (1-3 monthly review)

Approach

Early conventional DMARDs (Methotrexate, Sulfasalazine)

High Risk

Profile

RF+ Polyarticular, Systemic JIA (MAS risk), Refractory disease. Poor prognostic factors present (cervical spine/hip involvement, erosions)

Setting

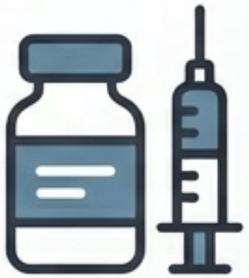
Close multidisciplinary follow-up, potential inpatient care

Approach

Aggressive biologic DMARDs (TNFi, IL-1Ri, IL-6Ri)

The Anchor Therapy: Methotrexate

Methotrexate (The Anchor DMARD)



Dose: 0.5-1 mg/kg (max 25 mg) once weekly. Subcutaneous (SC) preferred over oral for GI tolerance.

Folic Acid Support: 1 mg daily OR 5 mg once weekly (48h post-MTX) to reduce toxicity.

Monitoring: FBC, LFTs every 1-3 months.

Contraindications: Avoid in renal/hepatic impairment and pregnancy (highly teratogenic).

NSAIDs

Naproxen/Ibuprofen for symptoms. Not disease-modifying.

Corticosteroids

Intra-articular (Triamcinolone) first-line for Oligo. Systemic (low-dose oral/IV pulse) for bridging only, to prevent growth impairment/osteopenia.

The Biologic Arsenal Matrix

Pre-Treatment Screening (All Biologics)
Test for latent TB (TST/IGRA, CXR), Hep B/C, HIV, varicella. Contraindication: No live vaccines while on biologics.

Agent (Class)	Primary JIA Indication	Delivery & Dose	PBS Status
Etanercept (TNFi)	Poly, Extended Oligo, ERA	0.8 mg/kg SC weekly	PBS Authority Req.
Adalimumab (TNFi)	Poly, ERA, Refractory Uveitis	20-40 mg SC biweekly	PBS Authority Req.
Tocilizumab (IL-6Ri)	Systemic JIA, Polyarticular	IV monthly or SC weekly	PBS Authority Req.
Anakinra (IL-1Ri)	Systemic JIA (esp. MAS risk)	1-2 mg/kg SC daily	PBS Authority Req.
Abatacept (T-cell mod)	Refractory Polyarticular	IV monthly or SC weekly	PBS Authority Req.

Therapeutic Sequence Logic Tree

Pathway 1: Polyarticular JIA

Methotrexate → Inadequate response → **TNFi (Etanercept/Adalimumab)** OR **Abatacept/Tocilizumab**

Pathway 2: Systemic JIA (Active Arthritis)

Anakinra or **Tocilizumab** (Targeting IL-1/IL-6) → Add **Methotrexate** if chronic arthritis persists

Pathway 3: ERA

NSAIDs + Physio → **Methotrexate/Sulfasalazine** (peripheral joints) → **TNFi** (critical for axial disease)

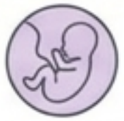
Pathway 4: Chronic Anterior Uveitis

Topical steroids → Methotrexate → **Adalimumab** (PBS-listed for CAU)

Long-Term Management & Special Populations

Paediatric Growth

Chronic inflammation and systemic steroids stunt growth. Goal is rapid steroid-sparing remission. Biologic dosing transitions to adult fixed-dose at target weight (e.g., 40-50kg).

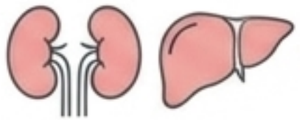


Pregnancy & Contraception

Methotrexate is heavily teratogenic; cease ≥3 months pre-conception. TNFi (Adalimumab/ Etanercept) can be continued through 1st trimester. Pre-pregnancy adolescent counseling vital.

Immunocompromise & Surgery

Live vaccines (MMR, Varicella) strictly contraindicated on biologics. Hold Methotrexate 1 week pre/post major surgery. Hold biologics 1 dose cycle pre-surgery.



Renal/Hepatic Safeguards

Avoid NSAIDs in renal impairment. Reduce/avoid Methotrexate if eGFR <30 mL/min or in hepatic disease.

Aboriginal and Torres Strait Islander Health Considerations

The Challenge

- Higher incidence, delayed diagnosis, and greater disease severity.
- **Geographic barriers:** Specialist services centered in major cities, creating medication access issues (e.g., cold-chain integrity for biologic SC injections).
- **Comorbidities:** High burden of co-occurring conditions like rheumatic heart disease.

Actionable Clinical Strategy

- **Access:** Maximize Telehealth (MBS items 91801, 91802) for outreach.
- **Partnership:** Collaborate directly with local Aboriginal Medical Services (AMS) for medication adherence and biologic monitoring.
- **Support:** Utilize Indigenous health incentive items (MBS 715) and advocate for patient-assisted travel schemes.
- **Cultural Safety:** Engage Aboriginal Health Workers, ensure clear non-jargon communication, and prioritize family-centered decision-making.