Guideline for the child with possible arthritis
(joint swelling/pain, loss of function)

Definition:
Juvenile Idiopathic Arthritis (JIA) is defined as arthritis in 1 or more joints for 6 continuous weeks occurring before 16 years of age, after the exclusion of other causes

Prevalence:
1:1000

Classification (ILAR 2001):
- Systemic Onset JIA [SOJIA] (10%)
- Oligoarticular - ≤ 4 joints (60%)
  - persistent (60%)
  - extended – i.e. > 4 joints involved after 6 months (40%)
- Polyarticular > 4 joints (20%)
  - Rf negative (90%)
  - Rf positive (10%)
- Enthesitis Related Arthritis (includes Ankylosing Spondylitis) (5%)
- Psoriatic arthritis (5%)
- Unclassified (meets criteria for 2 or more of above)

Presenting Complaint:
- A patient with JIA more commonly presents with joint swelling and/or loss of function/gait disturbance
- Joint pain may be absent or mild in proportion to the amount of inflammation and has a high negative predictive value of inflammatory rheumatic diseases in children
- Isolated musculoskeletal pain in the absence of other signs or symptoms is almost never a presenting complaint of JIA
- Malignancy should be considered in any patient presenting with musculoskeletal pain out of proportion to clinical findings as pain associated with malignancy tends to precede arthritis by weeks or months
- Presenting features in the toddler:
  - Unsettled with early morning nappy change
  - Refusing to weight bear
  - Walking with a limp
  - Delayed motor milestones

Diagnosis:
JIA is a clinical diagnosis based on a full history and examination
Differential Diagnosis:

JIA is a **diagnosis of exclusion** and other conditions need to be ruled out such as:

- Malignancy
- Acute/chronic bone and joint infections
- Streptococcal infection
- Lyme disease
- Reactive Arthritis
- Inflammatory Bowel Disease
- SLE / Juvenile Dermatomyositis / Vasculitis
- Bleeding Disorders
- NAI
- Trauma
- Hypermobility
- Growing pains

Reactive Arthritis: typically follows a gastroenteritis or urethritis, is more common in males 13 years and over, is self limiting and tends to resolve by 6 weeks

History of JIA:

- Early morning stiffness / gelling with inactivity
- Preceding viral illness, sore throat, gastroenteritis, urethritis
- Exposure to TB / family history of TB
- Other swollen joints
- Difficulty chewing food
- Constitutional symptoms – fever, weight loss, rash, pallor, mucosal ulcers, fatigue, easy bruising / bleeding
- Eye symptoms
- Activity in school and at home
- Difficulty writing at school
- Family History of rheumatological diseases

Examination:

- Physical findings take precedence over laboratory results
- A full general and musculoskeletal examination is a must including
  - Skin
  - Hair
  - Eyes
  - Mucous membranes
  - Nails
- **Focal bony tenderness suggests an orthopaedic cause or malignancy**
- **Lymphadenopathy, hepatosplenemegaly suggest malignancy, SOJIA, SLE**
Pitfalls in the Diagnosis of JIA:

- Inadequate history taking
- Failure to examine the child fully including the entire musculoskeletal system
- Referred pain e.g. hip to knee, costovertebral to sternum
- Uveitis with silent arthritis
- Earache with TMJ joint arthritis
- Infections
- Malignancy mimicking JIA
- Discitis
- Inflammatory Bowel Disease

Investigations:

- **Laboratory investigations are not diagnostic of JIA but are necessary to exclude differentials**
- FBC, film, ESR, CRP should be requested in all patients
- Hb : if anaemia suggests **SOJIA, malignancy, SLE**
- WBC : if low / norm in child with fever think **malignancy**, in adolescent think **SLE** – absolute lymphopenia in **SLE**
- Plt : Low / norm Plt with ↑ ESR / CRP – think **SLE or malignancy**
- ESR : reflects activity in SLE / JIA but may be normal in JIA
- The ANA and Rheumatoid Factor tests have a low PPV and are **not** recommended as routine in the initial investigations of arthritis
- ANA : up to 30% healthy child are +1:40 and 5% 1:160
- RF is also present in a small percentage (2-7%) of healthy children
- Both ANA and RF may be positive in other disease conditions such as viral and bacterial infections, chronic illnesses
- RF is only useful as a predictor for disease progression in Polyarticular JIA
- ANA should only be requested to help confirm SLE
- A positive ANA test of ≤ 1:160 in a child with musculoskeletal pain in the absence of other signs / symptoms of inflammatory or autoimmune disease is at low risk of developing such a disease
- Plain X-Rays are useful in excluding orthopaedic problems and malignancy and are not helpful in the diagnosis of arthritis

Management:

- Any delay in treatment of arthritis puts the child at risk of ongoing joint, muscle, bone damage, gross/fine motor delay
- Uveitis is often asymptomatic and can occur in up to 30% of JIA
- Undiagnosed uveitis can cause blindness (visual loss in 50%, blindness 15-40%)

Risk factors for Uveitis in JIA:

- Age < 7 years  
  - Oligoarticular subtype
- Female gender  
  - ANA positivity

- All patients suspected of having JIA should be referred to the paediatric rheumatology team within 6 weeks of onset of symptoms and must be seen by the team within 4 weeks of referral (BSPAR Standards of Care April 2010)
- All patients with suspected JIA must be referred to the paediatric physiotherapists
- All patients with suspected JIA should be started on regular NSAIDS (Ibuprofen or Naproxen)
References:


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Consider:

- Malignancy
- Acute Joint & Bone Infection
- Streptococcal Infection
- Reactive Arthritis
- Lyme Disease
- Bleeding Disorders
- IBD
- NAI
- SLE
- JDM
- Vasculitis

If ≥ 6 wks also consider JIA

Check:
- FBC
- Film
- ESR
- CRP

Consider Malignancy in patient with:
- Pain out of proportion to clinical findings
- Constitutional Symptoms
- Focal Tenderness on examination
- Rash
- Lymphadenopathy
- Hepatosplenomegaly

If < 6 wks:

- Blood tests specific for differential
- Plain x-ray of affected joints
- Treat as Appropriate
  - If Reactive Arthritis treat with NSAIDS – see BNF for dose

≥ 6 wks and suspected rheumatologic condition:

- Start Regular NSAIDS:
  - Ibuprofen 30-40mg/kg/day
  - Naproxen 20mg/kg/day
- Refer to physiotherapy Linda or Sarah bleep 5157
- Discuss case with Dr. Baksh (bleep 8418, ext. 2546) re: further investigation / management