



# CHILDHOOD ARTHRITIS

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My Granny had arthritis...children  
don't get arthritis!



Do They???

# Musculoskeletal Examination

- PGALS
  - Paediatric
  - Gait
  - Arms
  - Legs
  - Spine

REPORTS ON THE RHEUMATIC DISEASES SERIES 5

# *Hands On*

Practical advice on management of rheumatic disease



## **pGALS – A SCREENING EXAMINATION OF THE MUSCULO-SKELETAL SYSTEM IN SCHOOL-AGED CHILDREN**

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# Joint swelling





'Try and touch your shoulder with your ear'

- Cervical spine lateral flexion



**'Open wide and put three (child's own) fingers in your mouth'**

- Temporomandibular joints (and check for deviation of jaw movement)



Passive movement of hip  
(knee flexed to 90°, and  
internal rotation of hip)

- Hip flexion and internal rotation



'Bend forwards and touch  
your toes?'

- Forward flexion of thoraco-lumbar spine (and check for scoliosis)





**Practical Tip – while performing the pGALS screening examination**

- Get the child to copy you doing the manoeuvres
- Look for verbal and non-verbal clues of discomfort (e.g. facial expression, withdrawal)
- Do the full screen as the extent of joint involvement may not be obvious from the history
- Look for asymmetry (e.g. muscle bulk, joint swelling, range of joint movement)
- Consider clinical patterns (e.g. non-benign hypermobility and Marfanoid habitus or skin elasticity) and association of leg-length discrepancy and scoliosis)

**Practical Tip – normal variants: indications for referral**

- Persistent changes (beyond the expected age ranges)
- Progressive or asymmetrical changes
- Short stature or dysmorphic features
- Painful changes with functional limitation
- Regression or delayed motor milestones
- Abnormal joint examination elsewhere
- Suggestion of neurological disease or developmental delay

# Diagnosis?

- Arthritis?
  - Trauma?
  - Septic?
  - Haemarthrosis?
  - Malignancy?
  - Infection-TB,HIV
  - Rheumatic Fever?
- Avascular Necrosis?
- Reactive Arthritis?
- Benign Hypermobility Syndrome?
- Growing Pains?

### **Practical Tip – when inflammatory joint disease is suspected**

- The lack of reported pain does not exclude arthritis
- There is a need to probe for symptoms such as
  - gelling (e.g. stiffness after long car rides)
  - altered function (e.g. play, handwriting skills, regression of motor milestones)
  - deterioration in behaviour (irritability, poor sleeping)
- There is a need to examine all joints as joint involvement is often 'asymptomatic'



### **RED FLAGS**

(Raise concern about infection, malignancy or non-accidental injury)

- Fever, malaise, systemic upset (reduced appetite, weight loss, sweats)
- Bone or joint pain with fever
- Refractory or unremitting pain, persistent night-waking
- Incongruence between history and presentation (such as the pattern of the physical findings and a previous history of neglect)

# Differential

## – Trauma?

- History, Don't be fooled
- NAI

## – Septic?

- Sick, fever
- Single joint ( not always!!)
- Severe pain-joint frozen

## – Haemarthrosis?

- Family history
- Boys
- bruising

## • Infectious

- TB ( Poncets or Joint infection)
- HIV
- Rubella, Parvovirus

## • Malignancy?

- FTT
- Fever, constant symptoms
- Pallor/Petechiae
- Bone pain vs joint

## • Rheumatic Fever?

- Jones Criteria
- Flitting Polyarthritits

# Differential ctd

## Growing Pains?

- Benign Nocturnal Musculoskeletal pains
- Never in the day
- Always recovered by the morning

## Benign Hypermobility

## Reactive Arthritis

- HLA B27 associated- shigella, salmonella, campylobacter, chlamydia
- Post streptococcal
- Post viral (irritable hip)

## • Mechanical

- Osgood Schlatter
- SUFE
- Overuse

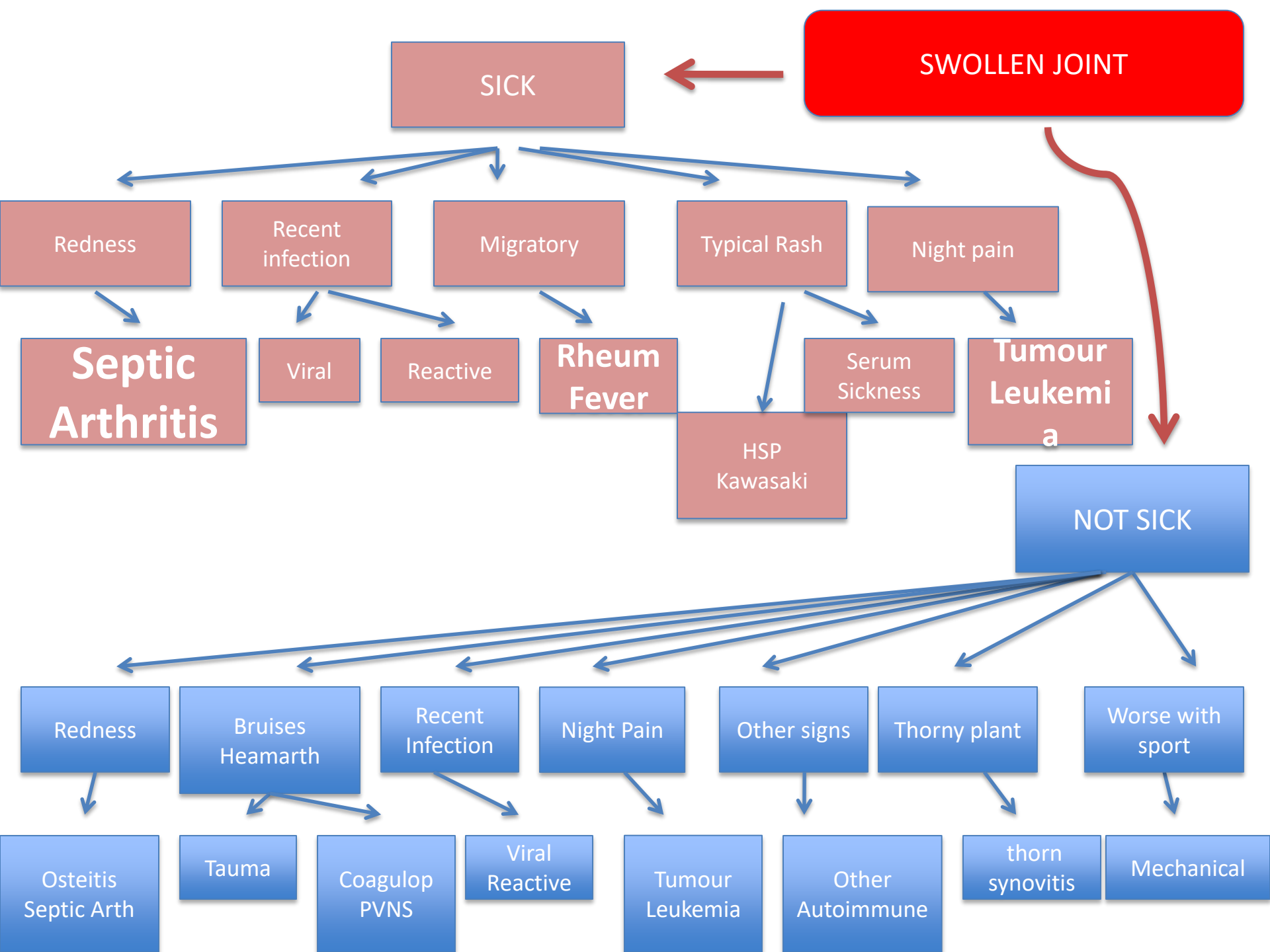
## • Avascular Necrosis

- Mostly hips
- Kohler( Navicular)
- Freibergs (2nd metatarsal)

# Differential CTD

- SLE
- Juvenile dermatomyositis
- Scleroderma
- Mixed Connective Tissue disease





**SWOLLEN JOINT**

**SICK**

Redness

Recent infection

Migratory

Typical Rash

Night pain

**Septic Arthritis**

Viral

Reactive

**Rheum Fever**

HSP  
Kawasaki

Serum Sickness

**Tumour Leukemi a**

**NOT SICK**

Redness

Bruises Hearnarth

Recent Infection

Night Pain

Other signs

Thorny plant

Worse with sport

Osteitis Septic Arth

Tauma

Coagulop PVNS

Viral Reactive

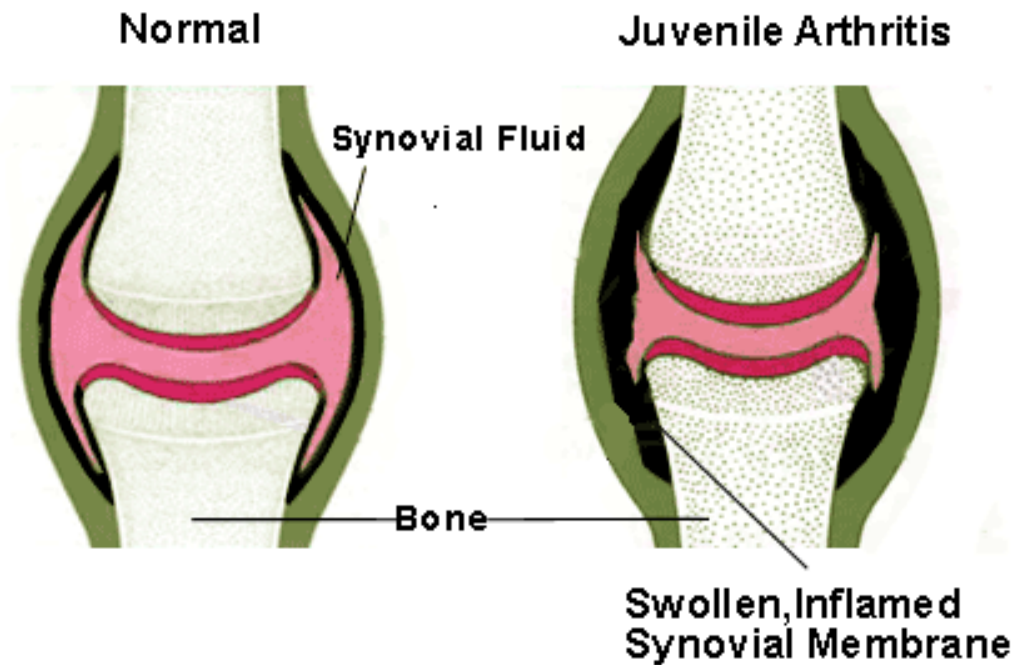
Tumour Leukemia

Other Autoimmune

thorn synovitis

Mechanical

# Juvenile Idiopathic Arthritis

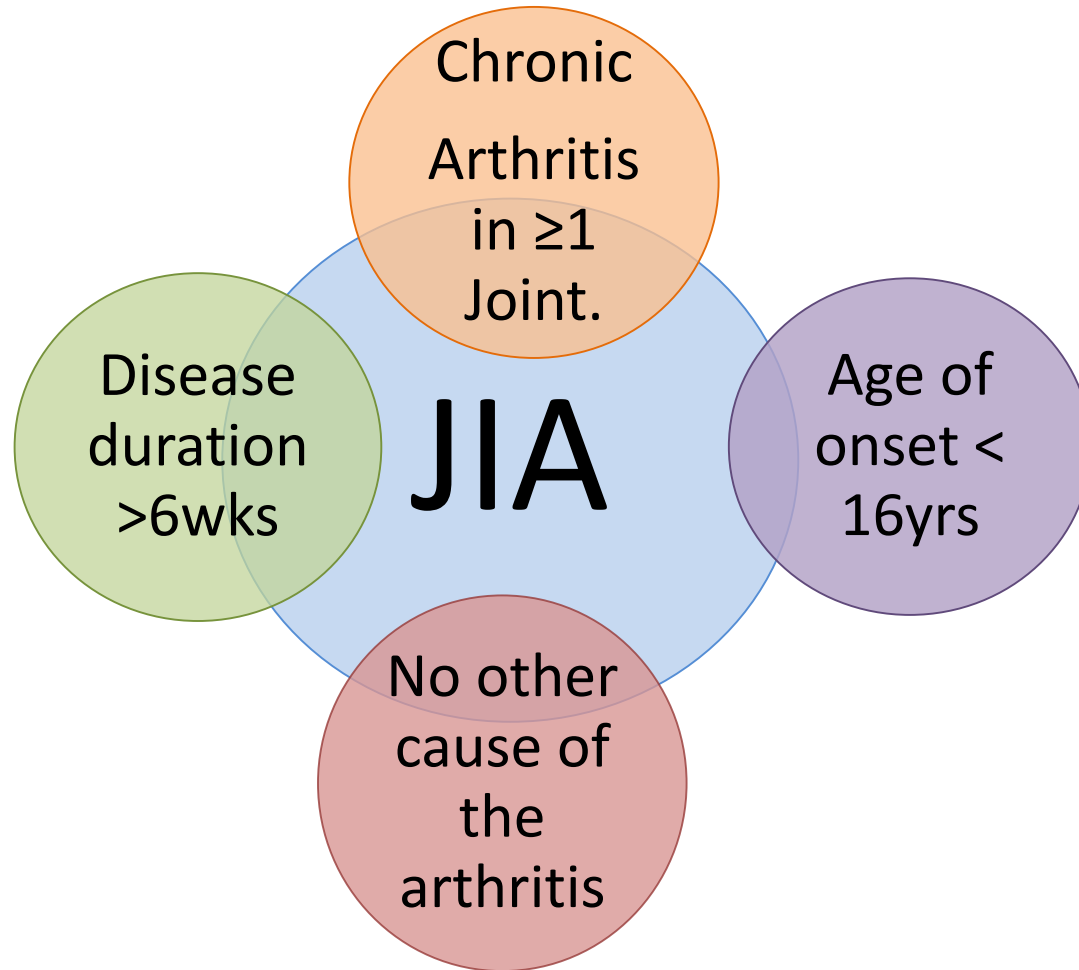




# JIA: Introduction

- is the most common paediatric rheumatologic disease and an important cause of long term morbidity
- World wide the incidence ranges from 0.8-22.6/100,000 children/yr and the prevalence ranges from 1-4/000 children.
- Described in all races and geographic areas
- Females predominate 2:1 , but with no variation in systemic type

# JIA: Definition



# Etiology & Pathogenesis

## Immunogenetic susceptibility

HLA I, HLA II alleles

Non HLA candidate loci

TNF- $\alpha$ , MIF, IL-6, IL-1 $\alpha$  encoding gene polymorphisms



## External trigger

Bacterial and viral infections

(Parvo virus B19, Rubella, EBV)

Bacterial or mycobacterial Heat Shock Proteins

Abnormal reproductive hormone levels

Joint trauma



**ABNORMAL IMMUNE  
RESPONSE**

## ABNORMAL IMMUNE RESPONSE

### Humoral

#### B-Cell activation

- Complement consumption
- Immune complex formation
- Auto antibodies (ANA etc.)
- Elevated serum Ig

### Cell mediated

TNF- $\alpha$ , IL-6, IL-1 are pro-inflammatory cytokines

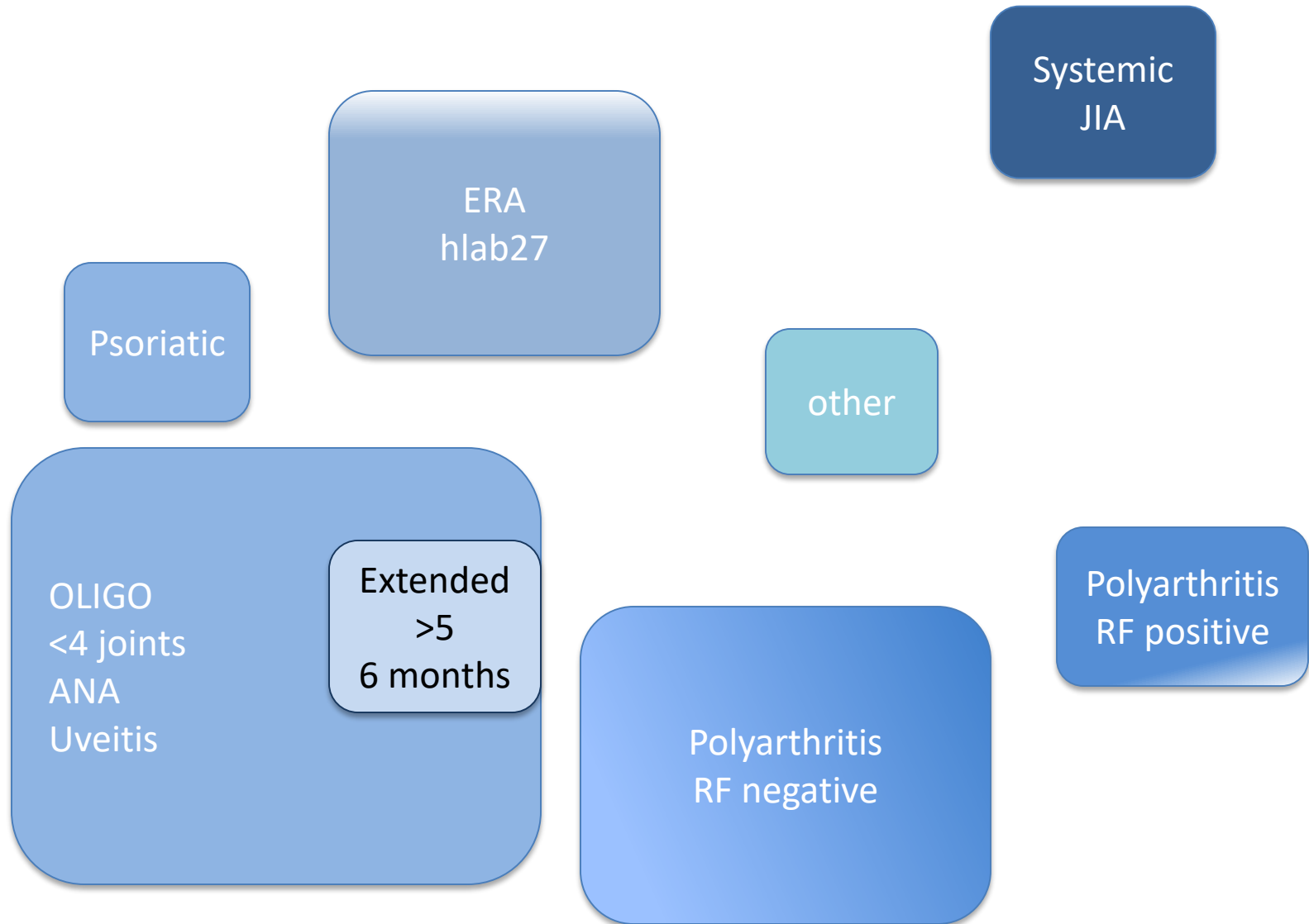
Favour TH<sub>1</sub> response over TH<sub>2</sub>

- Enhanced adhesion molecules expression in synovium results in T cell recruitment into synovium. >> Dense infiltrates, inflammatory synovitis; villous hyperplasia, hypertrophy, hyperemia & edema; vascular endothelial hyperplasia and pannus formation.
- Activation of metalloproteinase enzymes which lead to damage of synovium and adjacent tissues.

# Clinical Presentation

- May differ according to subtype.
- Morning stiffness or post activity gelling: Duration of stiffness proportional to degree of inflammation.
- Joint pain and swelling.
- May manifest as increase irritability, guarding of involved joint or refusal to walk
- Low grade fever, fatigue, malaise, wt loss may occur.
- **Painless anterior uveitis in upto 20% of JIA patients.**

# JIA SUBTYPES

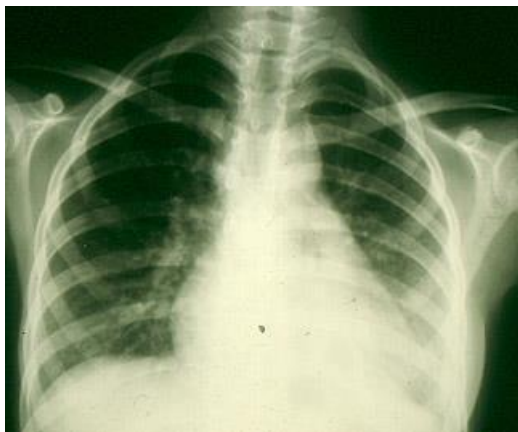
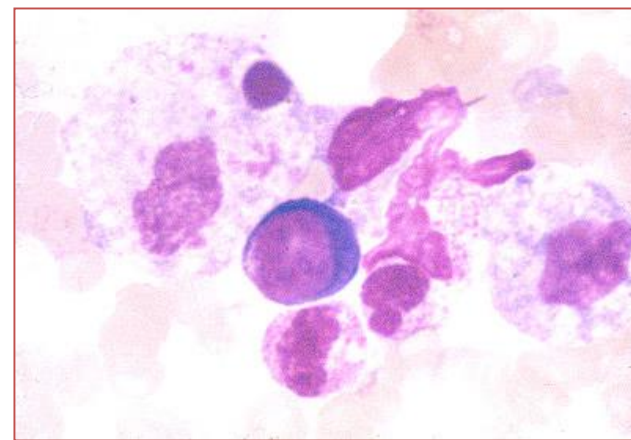




# Systemic JIA

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# Laboratory investigations

- **RF**.....Classification of Polyarticular disease
- **ACCP**.... Not recommended. Most ACCP positive children are RF positive.
- **ANA**.... For uveitis risk stratification; and rule out differentials.
  - ANA seen in up to 80% of children with JIA
  - ANA seen in up to 10% of children without arthritis.
- **ESR/CRP**: Acute phase reactants for assessing disease activity. ESR most commonly used.
- **Other tests**: as per specific differential.



# Imaging in JIA

- Radiograph is still the Gold Standard
- Ultrasonography
  - Increasing use in the clinic
  - An extension of the physical examination
- Magnetic Resonance Imaging
  - Most sensitive imaging modality
  - Can show “sub-clinical” joint inflammation
  - Utility limited by cost and access

# Management

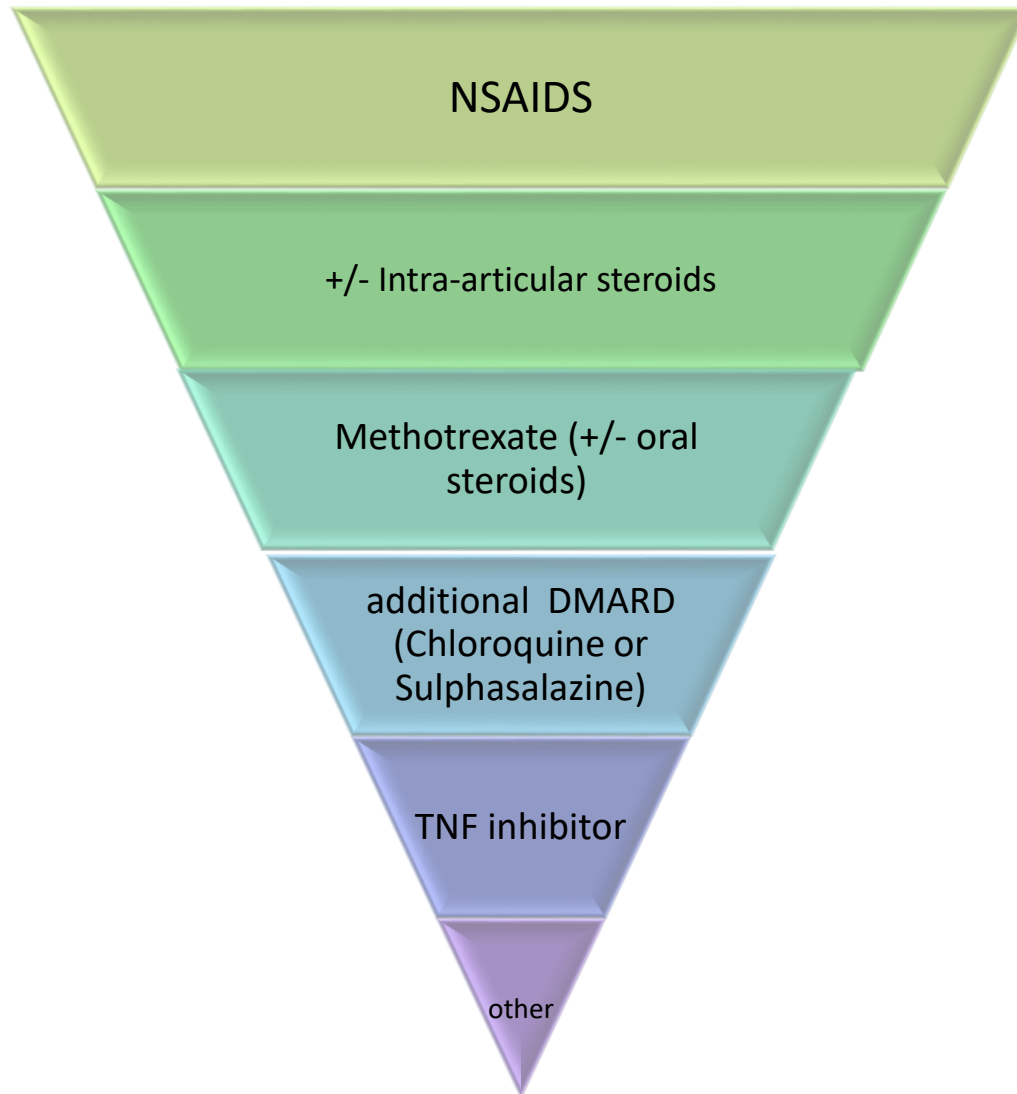
- Non pharmacological management includes
  - physiotherapy
  - occupational therapy
  - social support
  - pain management



# Counseling

- paramount importance
  - shock, disbelief and fear
- prognosis and implications of the diagnosis
  - goals of therapy
  - side effect of medication.
- well considered, scientifically valid sources of information on JIA
  - avoid bewildering/misleading information and alternative therapies
- counseling and advice regarding sporting activities and schooling

# Pharmaceutical Treatment of JIA



# Intra-articular steroids

- Can inject multiple joints at once
- May need anaesthetic



# Disease Modifying Drugs (DMARDs)

- Methotrexate
  - The first revolution in JIA
  - Massive improvement in outcome, Low toxicity
  - Start 0,3-0.5mg/kg once a week
  - take with Folic Acid
  - Nausea and GIT side effects a problem
  - Monitor FBC and LFT 3 - 6 monthly
  - Can wean after 6 months of inactive disease

## OTHER AGENTS

- Sulphasalazine, Chloroquine, Leflunamide,

# Biologics

- Anti-TNF agents

- The second revolution
- Very effective, very expensive

- Etanercept:

- TNF receptor: FC fusion protein 0.4 mg/kg subcutaneously two times per week

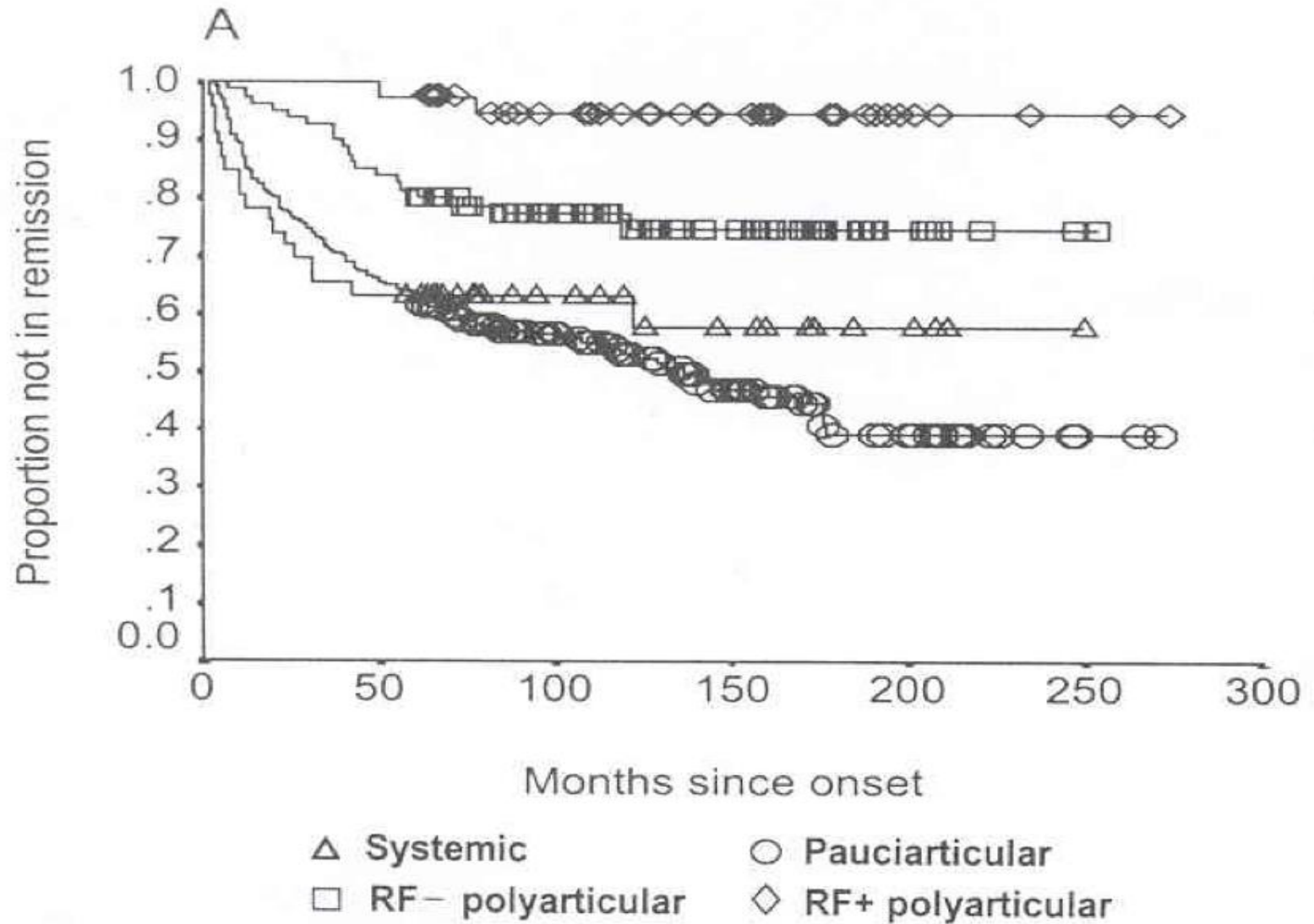
- Infliximab:

- chimeric IgG1 monoclonal antibody to TNF- $\alpha$  3 6 mg/kg IV baseline, 2, 6 weeks, 8-weekly

- Adalimumab:

- humanized monoclonal antibody to TNF- $\alpha$  24 mg/m<sup>2</sup> subcutaneously every other week (0.4-0.8 mg/kg/14d)

# Outcomes



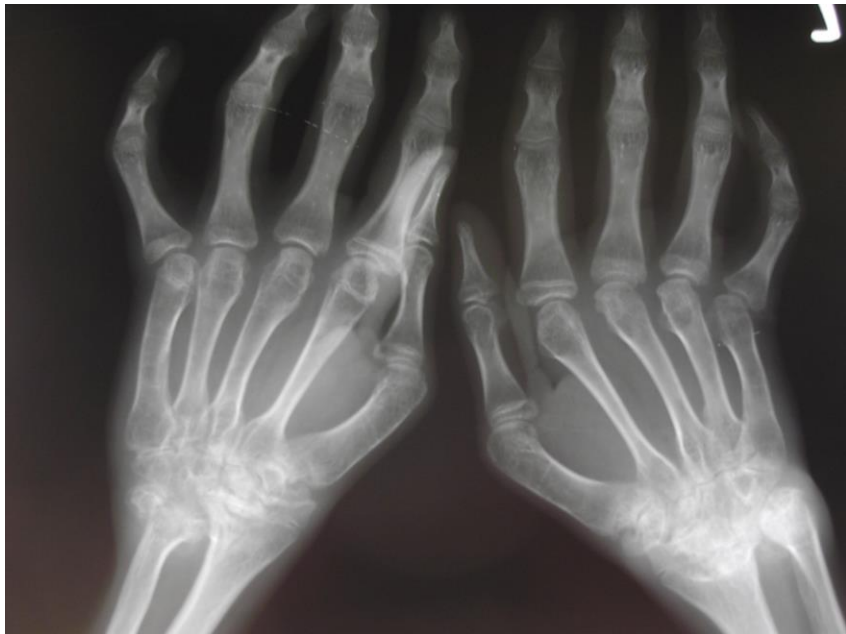
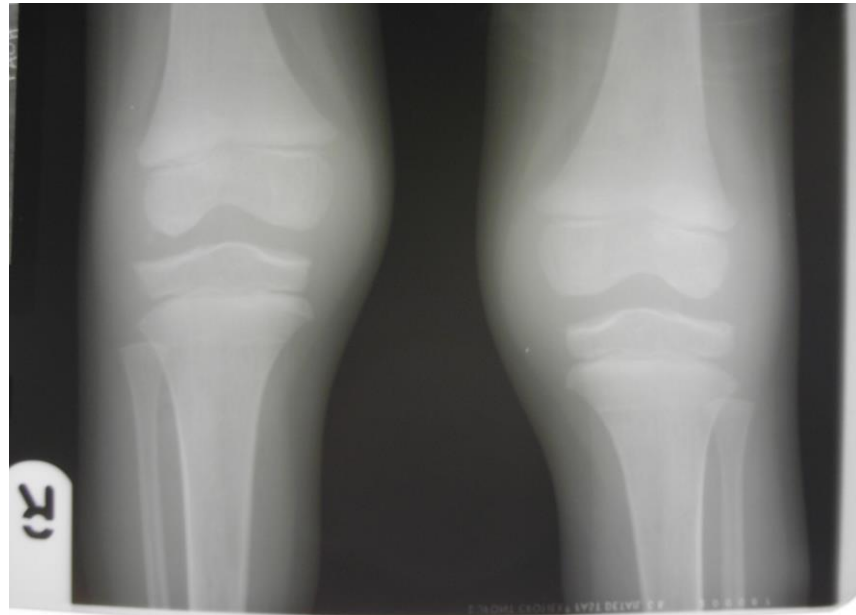
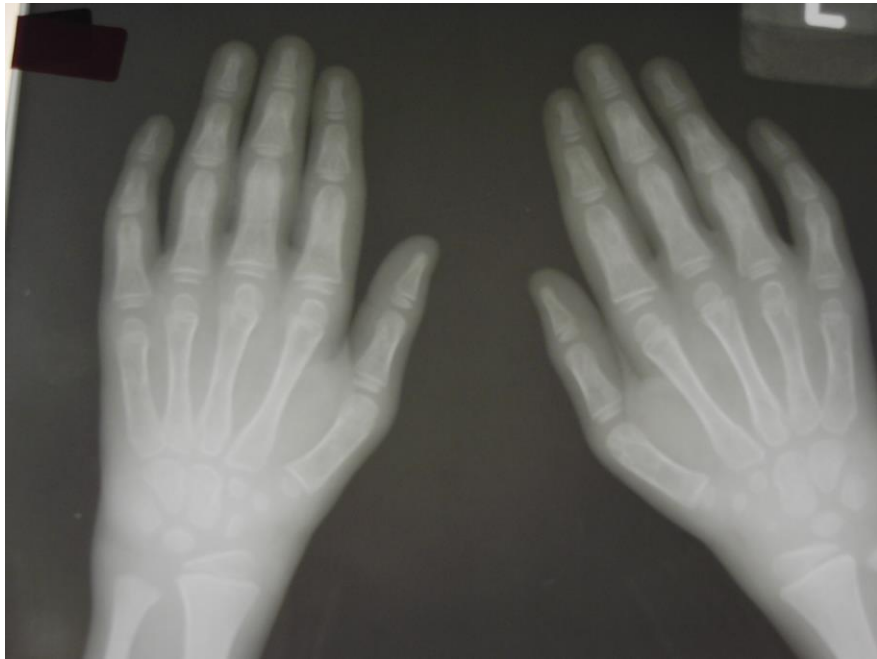


# Childhood arthritis; the disease hasn't changed



# Outcomes





# Message

- JIA is a chronic but manageable disease
- If not managed effectively, the outcome can be devastating
- Children with joint symptoms should be seen and referred early to maximise chances of good recovery.
- IT'S NOT JUST GROWING PAINS!!