

# Focus on Juvenile Idiopathic Arthritis

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# Aims

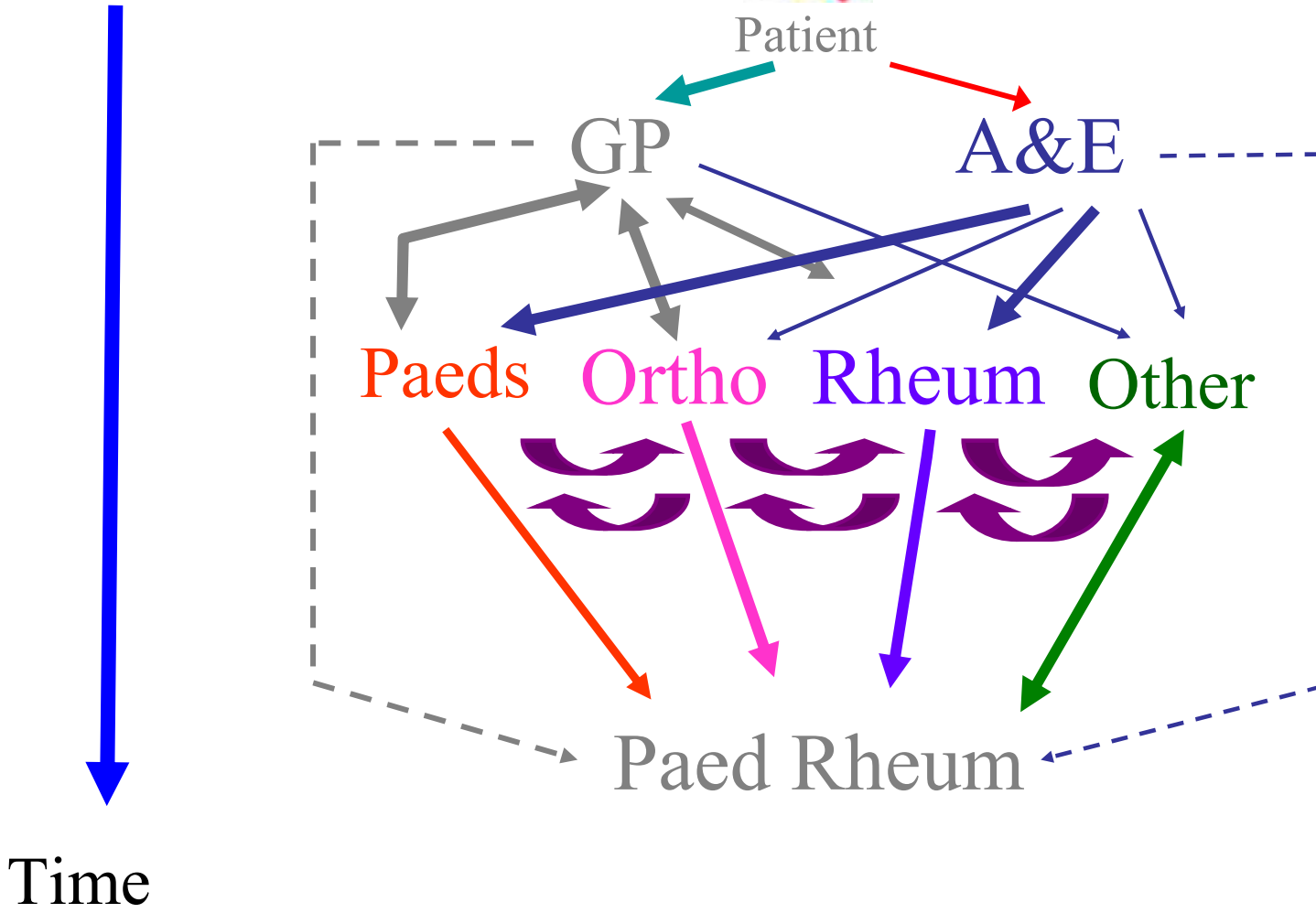
- Raise awareness about JIA
- Early diagnosis and referral does make a difference to outcome
- Your role in facilitating access to care

# Incident regional cohort of JIA (n=152)

- Prolonged interval of untreated disease
  - 74% > 10 weeks
  - median interval 20 weeks (0-416)
- Unnecessary and invasive investigations
  - 18% (arthroscopy / synovial biopsy)
- Greatest delay in those referred via orthopaedics
  - *Similar reports from elsewhere (Australia, N America)*



Patient



# Objectives

- List 3 common causes of swollen knee
- List 3 presenting features of JIA
- How to diagnose JIA
- List 3 complications of JIA
- How to contact paed rheum locally

# Definition of Juvenile Idiopathic Arthritis (JIA)

- Persistent joint swelling >6/52
- Age at onset <16years
- Diagnosis of exclusion

***Prevalence 1:1000, Incidence 1:10,000***

*Comparison with epilepsy, diabetes*

***Commonest cause of chronic arthritis***

*Northern Region approx 500 children affected*

# Terminology

- Juvenile Idiopathic Arthritis
- Juvenile Chronic Arthritis
- Juvenile Rheumatoid Arthritis
- Still's Disease

**A**vascular necrosis (eg Perthes/orthopaedic)

**R**eactive (viral, streptococcal, post GI)

**T**rauma (including Non accidental injury)

**H**aematological (eg Hb.opathies, leukaemia)

**R**ickets (ie metabolic)

**I**diopathic (reflex sympathetic dystrophy, fibromyalgia)

**T**umour (benign, malignant)

**I**nfection (viral, bacterial)

**S**ystemic (SLE / Vasculitis, Dermatomyositis, Sarcoidosis, IBD)

**JIA is a  
diagnosis of exclusion**

# Single swollen joint

- Trauma
  - ? Child protection / NAI
- Infection
  - Septic (viral, TB, Lyme)
  - Reactive
- Inflammatory
  - Juvenile Idiopathic arthritis
  - Inflammatory bowel disease
- Haemarthrosis
  - Bleeding disorders - very young (males)
  - Post trauma (adolescents)
- Tumour



If not septic or trauma, then JIA  
most likely diagnosis

# Juvenile idiopathic arthritis

- Systemic
- Polyarticular (RF neg)
- Polyarticular (RF pos)
- **Oligo-articular**
- Extended oligo
- Enthesitis related / spondyloarthropathy
- Psoriatic

*J Rheumatol 1995;22:1566-9*

# Oligo-articular JIA

- Commonest sub-type (60%)
- Knee most common affected joint
- Preschool children (girls>boys)
- Often a well child (limp)
- Generally good prognosis with early referral and treatment
  - BUT ....

# Chronic anterior uveitis



1/3 of children with JIA  
Potentially blinding  
Asymptomatic in early stages



Regular eye screening for  
many years is needed

# Making a diagnosis....

- Clinical skills most important
- Investigations
- Pattern recognition (cases)

# Presentations of musculoskeletal disease

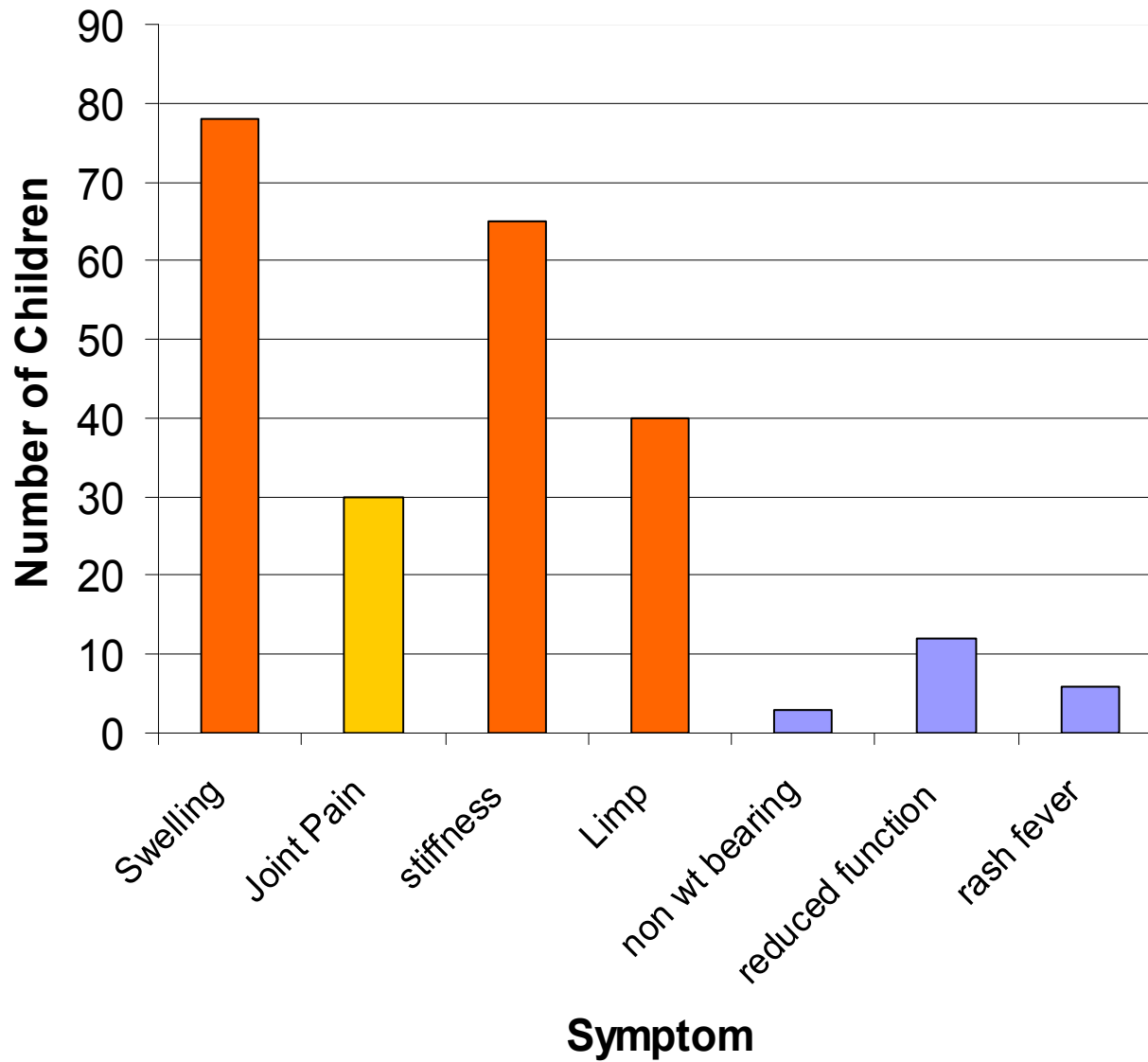
## Musculoskeletal

- Limp
- Sore joint
- Swollen joint
- Limited movement
- Painful muscles
- Morning stiffness
- Muscle weakness

## Non-musculoskeletal

- Fever without focus
- Abdominal pain
- School refusal
- Change behaviour / activities
  - *play, mood, motor skills,*
- Deteriorating school work
- Rash, 'unwell child'

## Presenting Features of Children with JIA



# Musculoskeletal history

- History of trauma (daily occurrence in a young child !!)
  - Are you happy with the story?
- Pain history
  - Mechanical versus inflammatory
- Functional history
  - What can the child do / not do / no longer do ?
- Site – think radiation & referred pain
- Swelling, deformity, 'change'
- Developmental / milestones (change?)
- Systemic features
- Family history, drug history, past illnesses

# Musculoskeletal assessment

- Examination
  - Co-operation may be difficult!
  - Changes may be subtle (comparing sides may be falsely reassuring)
  - Knowledge of 'normal' is important
- The history is not always localising
  - Often given by parent / carer – vague and non-specific
  - Need to screen all joints (and often more ! – other systems)
  - The cause of a limp is very broad

Worth looking elsewhere....  
screening all joints and for other clues



# pGALS

- A musculoskeletal screening examination
  - school age children (but younger children often can do)
- Derived from adult GALS screen
  - Simple additions, manoeuvres used in clinical practice
- Quick, sensitive & specific, acceptable

[www.arc.org.uk](http://www.arc.org.uk)



# ? Inflammatory joint disease

- Often a history of mild trauma
  - ? Incongruent to physical findings
- “Pain” may be absent / mild
  - “irritable”, poor sleeping especially young child
- Reduced function
  - School
  - Play / sports
  - Regression of motor milestones
- Gelling rather than morning stiffness
- Signs of arthritis can be subtle and asymptomatic
  - Joint Swelling, Limited joint movement
  - Look for “asymptomatic” involvement elsewhere (pGALS)
- Extra-articular features (psoriasis, GI symptoms, eye symptoms)
- Family history (autoimmune disease)
- ***Don't be reassured by a well child and normal FBC, ESR***

# Investigations – keep simple !

- **FBC (film) , ESR or CRP**
  - **Urinalysis**
  - **Radiographs / USS**
    - hips (frog view)
  - With non-specific aches and pains, consider
    - Muscles enzymes, thyroid function,
    - Bone chemistry (?rickets)
  - Autoantibodies
    - ANA rarely diagnostic
    - RF rarely helpful
  - Uric acid
    - “never” helpful
- **Consider paed rheum referral BEFORE**
  - Further imaging (MRI / Bone scan / USS ) and especially if elective / GA required
  - Arthroscopy and synovial biopsy
  - **Don't delay referral to paed rheum awaiting investigations**

# Why is early referral important ?

- Diagnosis confirmed
- Access multi-disciplinary team
- Early use of joint injections and consider immunosuppression (methotrexate)
- Access eye screening
- Optimise outcome

- JIA *not* a benign disease
- Joint damage occurs early
- “Window of Opportunity”

# JIA – Prognosis

- Early diagnosis and treatment vital
- 1/3 continue to have problems in adult life
- 1/3 “remission” but have residual problems
- 1/3 “remission” without residual problems

# JIA – drug treatments

- < 5 joints affected – mild disease
- Joint injection(s) – with corticosteroid
- NSAIDs

# Early Intervention in JIA – Does It Matter?

## Joint Injections

- Safe
- Rapidly ease symptoms
- Induce remission in many cases
- Prevent deformity



# Early Intervention in JIA – Does It Matter?

## Chronic Anterior Uveitis

- ◆ Symptomless, treatable, blinding if not treated.
- ◆ Often presents shortly before / just after onset of arthritis.



# JIA – drug treatments

- > 5 joints affected = severe disease
  - Pulsed IV steroids / multiple joint injections
  - Methotrexate (often high dose parenteral route)
  - Anti-TNF / biologics / immunomodulatory treatments  
(Autologous Haematopoietic stem cell rescue)

# Methotrexate

- Evidence base with clinical trials
  - Reduces joint damage, allows steroid sparing
  - Efficacy better with earlier use
  - Safety in short / medium term
    - *No evidence of increased malignancy / infertility / liver or lung damage*
  - No long term data >25yrs

# Multi-Disciplinary team

- ◆ Nurses / Physio / Occupational Therapists
- ◆ Education and support
- ◆ Liase with primary care / school / social services
- ◆ Shared care / Transitional care to adult services

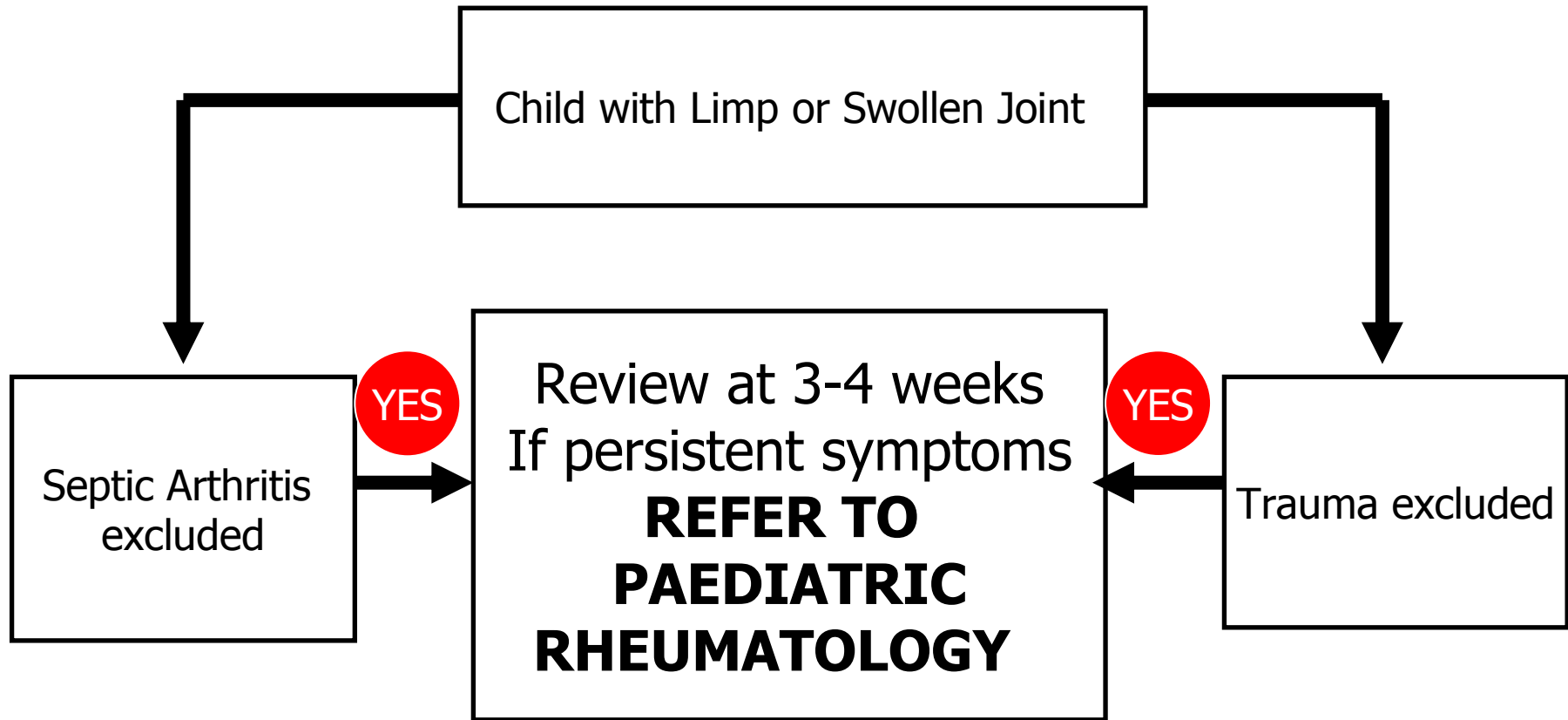
# Who to refer to paed rheum?

- **Inflammatory symptoms / signs**
  - Don't be put off by normal investigations
- **Persistent aches and pains / limp**
  - With / without systemic complaints
  - With / without abnormal examination
  - Persistence / severity (effect on function)
  - Delayed / regression milestones
  - School absenteeism + / wheelchair
- **Before contemplating invasive tests !**

# Referral to paed rheum

- No out of hours on call BUT
- Ring us / email us
  - Mark Friswell – RVI
  - Helen Foster – FRH / RVI
  - Mario Abinun – NGH
  - Secretary – Terry Bell 25318 RVI
- Fax us
  - 0191 2825485
- Clinic appt usually < 2/52

# Limping Child Protocol



# Questions

- List 3 common causes of swollen knee
- List 3 presenting features of JIA
- How to diagnose JIA
- List 3 complications of JIA
- How to contact paed rheum locally

# Key learning points

- Trauma is common but may not be the explanation
- If not trauma or septic, JIA most likely
- Apparent absence of pain doesn't exclude JIA
- Need to examine all joints
- Consider paed rheum before invasive investigations
- Think beyond hips in the limping child
- Early diagnosis of JIA does make a difference

# Summary

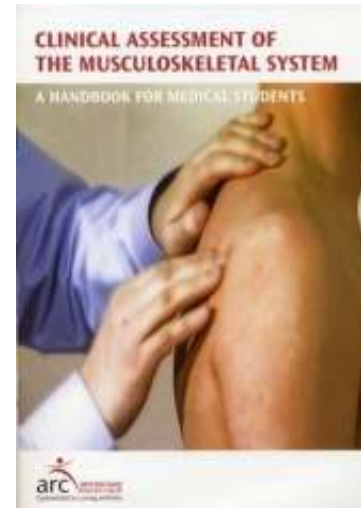
- JIA is a heterogeneous group of diseases
  - Diagnosis made by good clinical skills, judicious use *and* interpretation of relatively simple investigations
  - Sepsis mandatory to exclude but shouldn't delay referral to paed rheum (and before invasive tests)
- Management multidisciplinary
  - Trend to early and aggressive therapy
  - Outcome is variable *but...Not a benign disease*
- *Early* referral does make a difference.....

# Further resources

Patient information booklets

Growing pains

- When Tim Has Arthritis
- Teenagers with arthritis
- Hypermobility
- Fibromyalgia
- Anterior knee pain



“Hands On” – [for health professionals](#)  
Hypermobility, growing pains, pGALS

DVDs – GALS, REMS, pGALS  
Interactive cases



All free !!!!!

# Useful references

- pGALS DVD, available free on [www.arc.org.uk](http://www.arc.org.uk), web-based resources (pGALS, hands on “pGALS” as pdf - pGALS <http://www.arc.org.uk/arthritis/emedial.asp> and hand on “growing pains” <http://www.arc.org.uk/arthritis/medpubs/6541/6541.asp>) with parent info sheet on growing pains
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# Handout / further information

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