

# Delay in diagnosing childhood arthritis – does it matter?

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# Background

- Musculoskeletal (MSK) problems in childhood are common
- Wide differential diagnosis from benign to life-threatening
- Identification of the underlying condition relies on the skills and knowledge of the assessing doctor
- ***Children with MSK diseases have a delay in diagnosis\****

\*JIA Foster 2008, Schiff 2009, Tzaribachev 2009; Muscular dystrophy Ciafaloni 2009;  
Childhood cancer Dang Tan 2007, Lethaby 2013; SUFE Weigall 2010

# Juvenile Idiopathic Arthritis (JIA)

- Inflammatory arthritis of childhood, prevalence of 1:1000
- Presents with swelling and stiffness
- Can lead to restricted joint movement, joint damage, impact on function
- Trend towards early, aggressive treatment to improve outcomes



# Delay in JIA

- Median time from symptom onset to first paediatric rheumatology appt unchanged 2001 – 2010\*
- Many patients have circuitous routes to diagnosis
  - Multiple specialists
  - Multiple ‘diagnoses’ given
- Reasons for delay likely multifactorial

\*McErlane F, data from the Childhood Arthritis Prospective Study, presented at the BSR annual meeting, April 2014

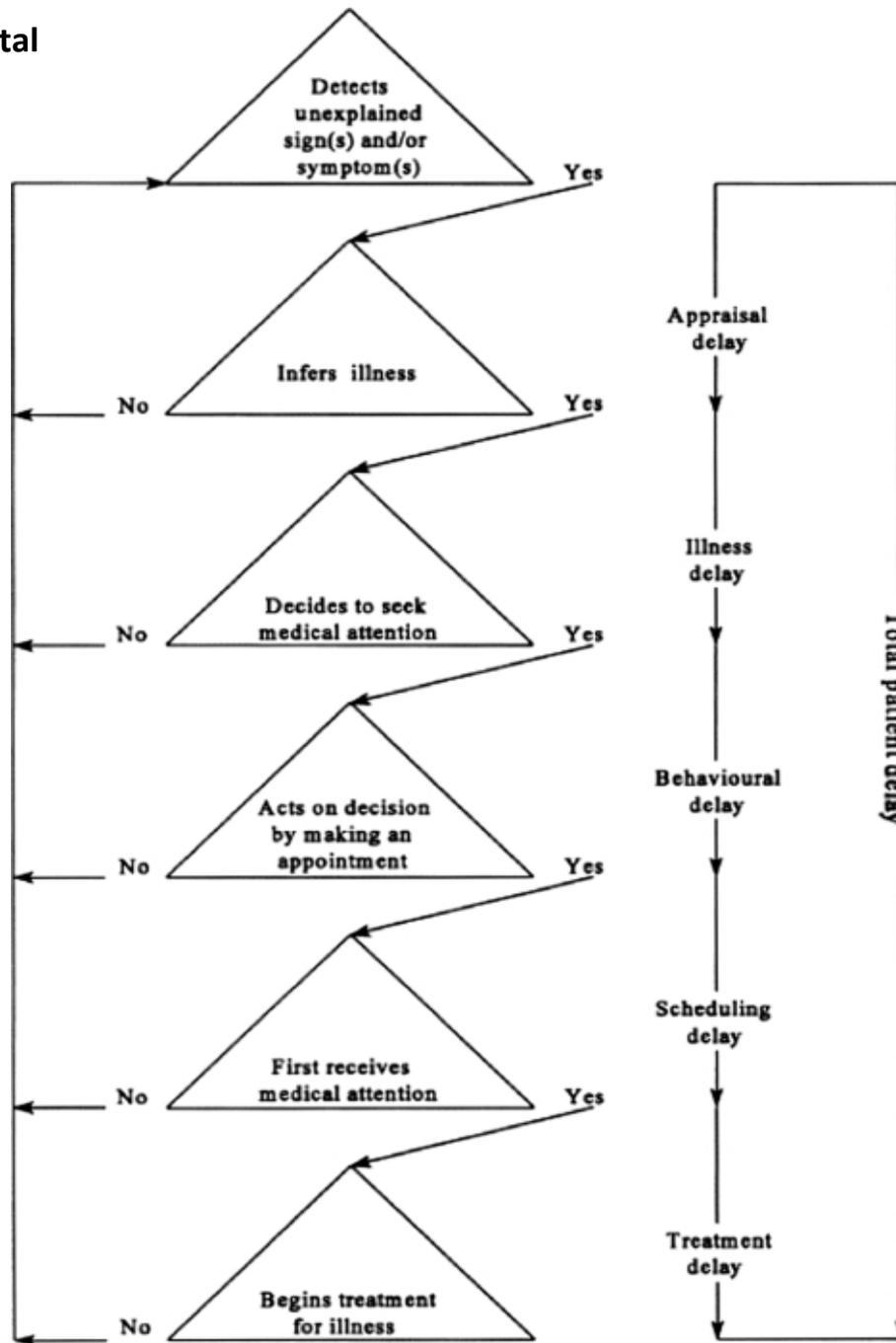
# Why do we need to reduce delay?

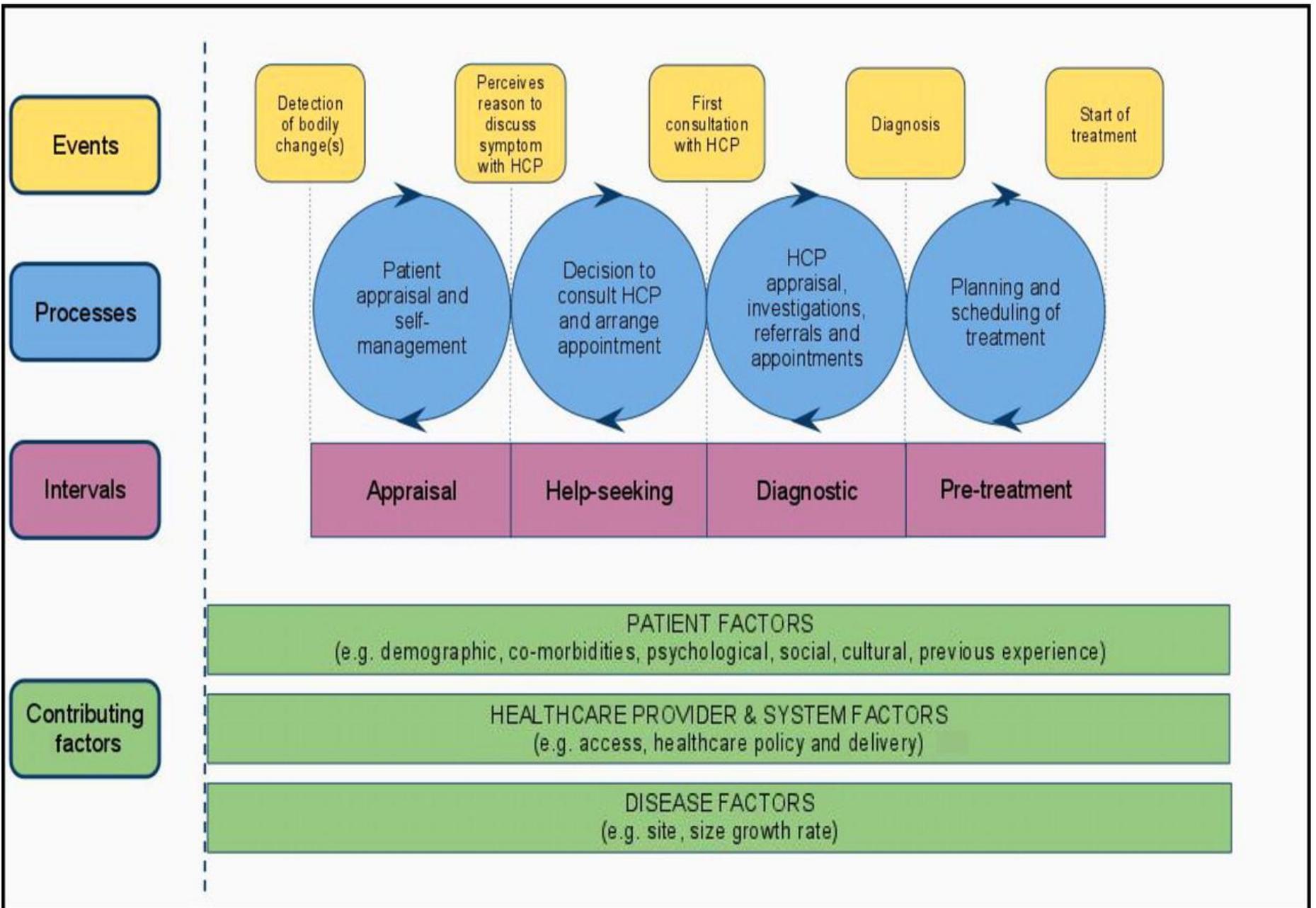
- ‘Window of opportunity’ to reduce inflammation
  - Shown in rheumatoid arthritis\*
  - JIA: TREAT study – greater chance of achieving clinically inactive disease when treated earlier in disease course<sup>+</sup>
  - Systemic-onset JIA<sup>#</sup>
- Reduce joint restriction, damage and improve quality of life
- Patient experience

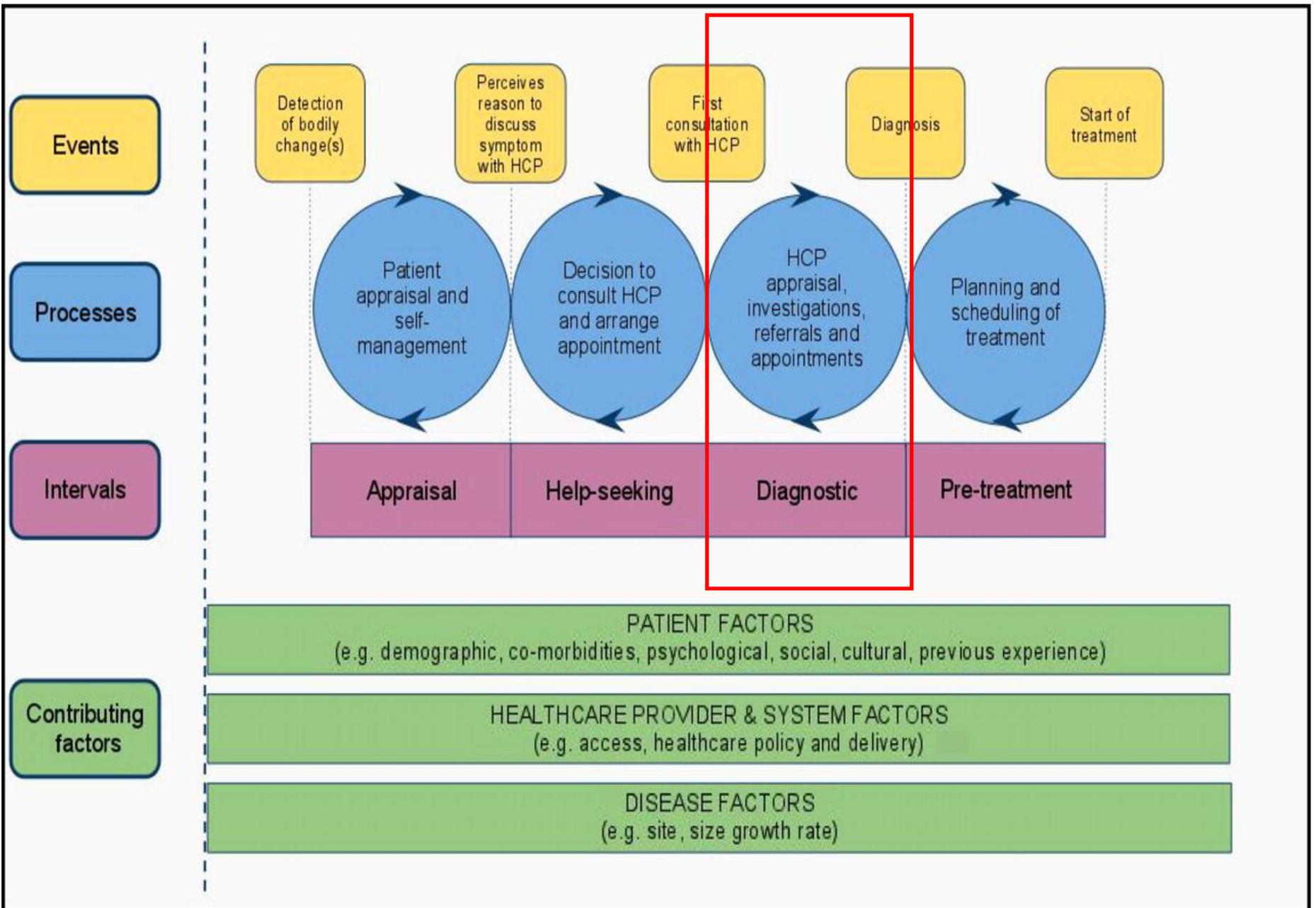
\*van Nies 2014, <sup>+</sup>Wallace et al 2012, <sup>#</sup>Nigrovic 2011

# The General Model of Total Patient Delay

Andersen *et al.* (1995).







Diagnostic  
interval

Doctors low in  
confidence in  
MSK  
assessment

Challenges of  
paediatric  
assessment

Uncertain  
referral  
pathways

Lack of  
awareness  
'children  
don't get  
arthritis'

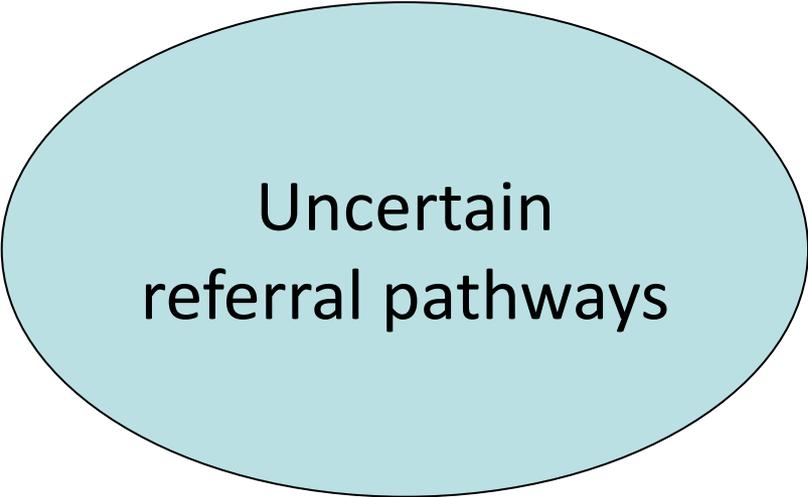
Reliance on  
history alone

Clinical signs  
not being  
identified or  
understood



Challenges of  
paediatric  
assessment

- Pre-verbal child
- Sleeping
- Crying
- Fear



Uncertain  
referral pathways

- Paediatrics
- Physiotherapy
- Orthopaedics
- Rheumatology
- Neurology
- Psychology

Hsu 2012, Carli 2012, Reeder 2004

Patient narratives

Rapley T, qualitative work



Clinical signs not  
being identified  
or understood

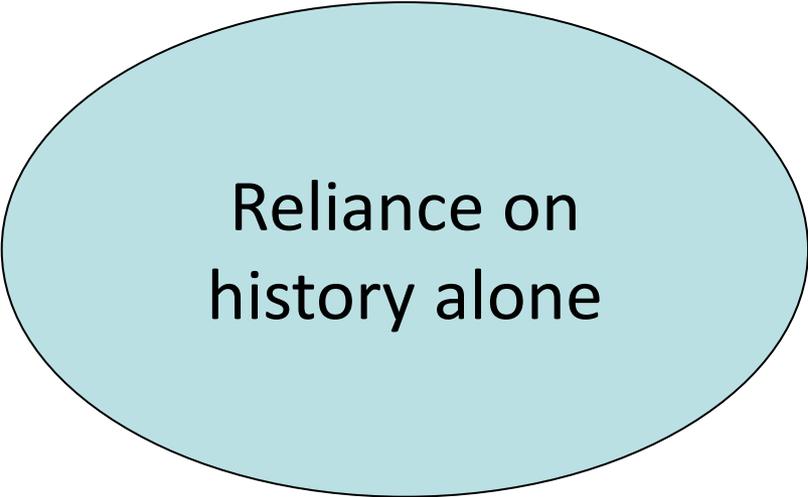
- Is this normal?
- Chubby ankles,  
hypermobility
- Different to adults

Hsu 2012, Carli 2012, Reeder 2004

Goff 2012

Patient narratives

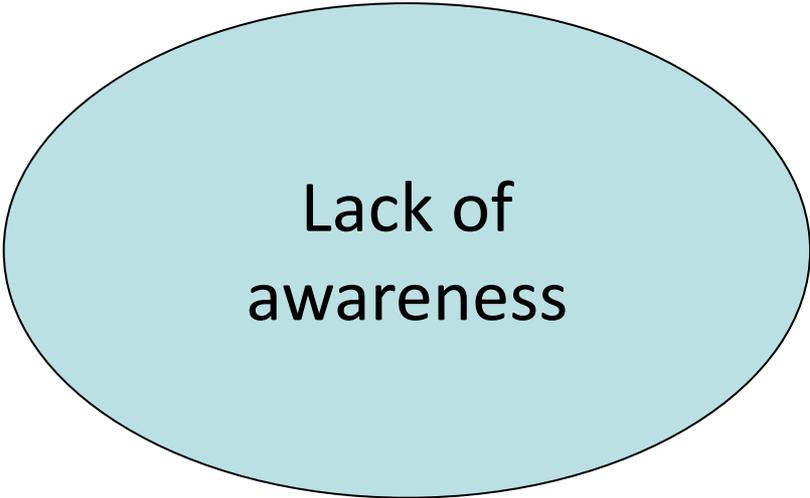
Observation from clinical practice



Reliance on  
history alone

- History not always localising
- Children may not articulate pain
- Reliant on carer knowing history

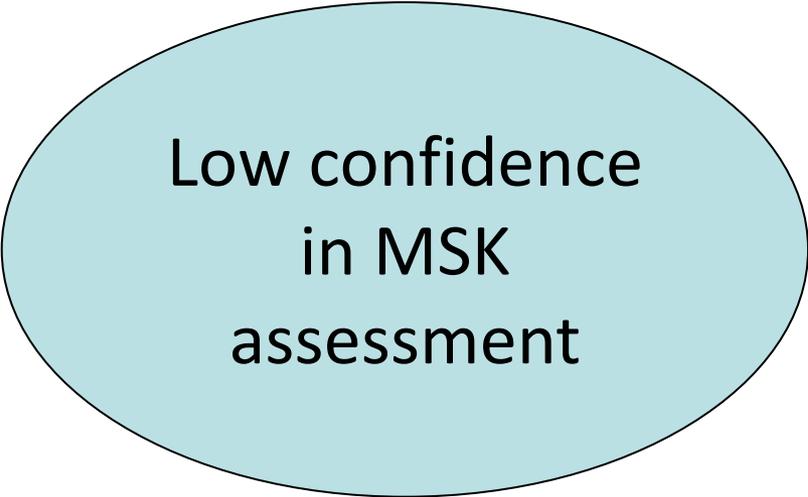
Foster 2007, Goff 2012



Lack of  
awareness

- Arthritis doesn't happen in children
- It must be growing pains

Family experiences and feedback  
Patient narratives



Low confidence  
in MSK  
assessment

- What examination should be done?
- What am I looking for?
- What does it mean?
- How long will it take?

Jandial 2007

Hergoenroeder 2001

Diagnostic interval

Doctors low in confidence in MSK assessment

Challenges of paediatric assessment

Pre-verbal child  
Co-operation  
Sleeping, grumpy, can't sit still

Uncertain referral pathways

Not happy to examine joints  
How do I know if this is normal?  
Seems to be happy - prob ok

Orthopaedics  
Rheumatology  
Paediatrics  
Physiotherapy  
Psychology

Lack of awareness  
'children don't get arthritis'

Probably growing pains

Reliance on history alone

Carer may not know history  
Children may not admit to pain/stiffness

Clinical signs not being identified or understood

Chubby ankles  
Difficult examination

# Knowledge to be shared

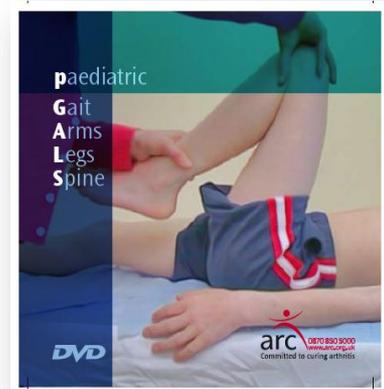
- Children get arthritis (and other serious conditions)
- Diagnosis relies on competent clinical skills (history taking and examination), which then need interpreted within the context of the presentation
- The right referral, at the right time, to the right service, makes a difference

# The challenges of sharing knowledge

- Contextually dependent
- Multiple barriers and facilitators
- Complex pathway – at what point to intervene?

# What has been tried?

- Educational interventions developed
  - pGALS, pREMS focussed on clinical skills
  - Written and electronic resources focussed on knowledge
- Focussed at different levels
  - Undergraduate, postgraduate, different specialties
  - Assessments, e-learning and formal teaching
- Linear model
- Success in some areas – but not in others



# The example of pGALS



- Validated examination of the MSK system in school age children
- Series of composite manoeuvres assessing range of joint movement
- Sensitive and specific in the hands of specialists and generalists
- Quick (2 mins) and acceptable to families
- ?Easy to implement

# pGALS in primary care

Uncertainty re interpretation

Time pressure

Infrequent examination



Patient/family expectations

Will it change management?#

- Complexity around changing practice in primary care\*
- How strong is the evidence and the context?#

\*Gabbay & le May 2004 Evidence based guidelines or collectively constructed mindlines

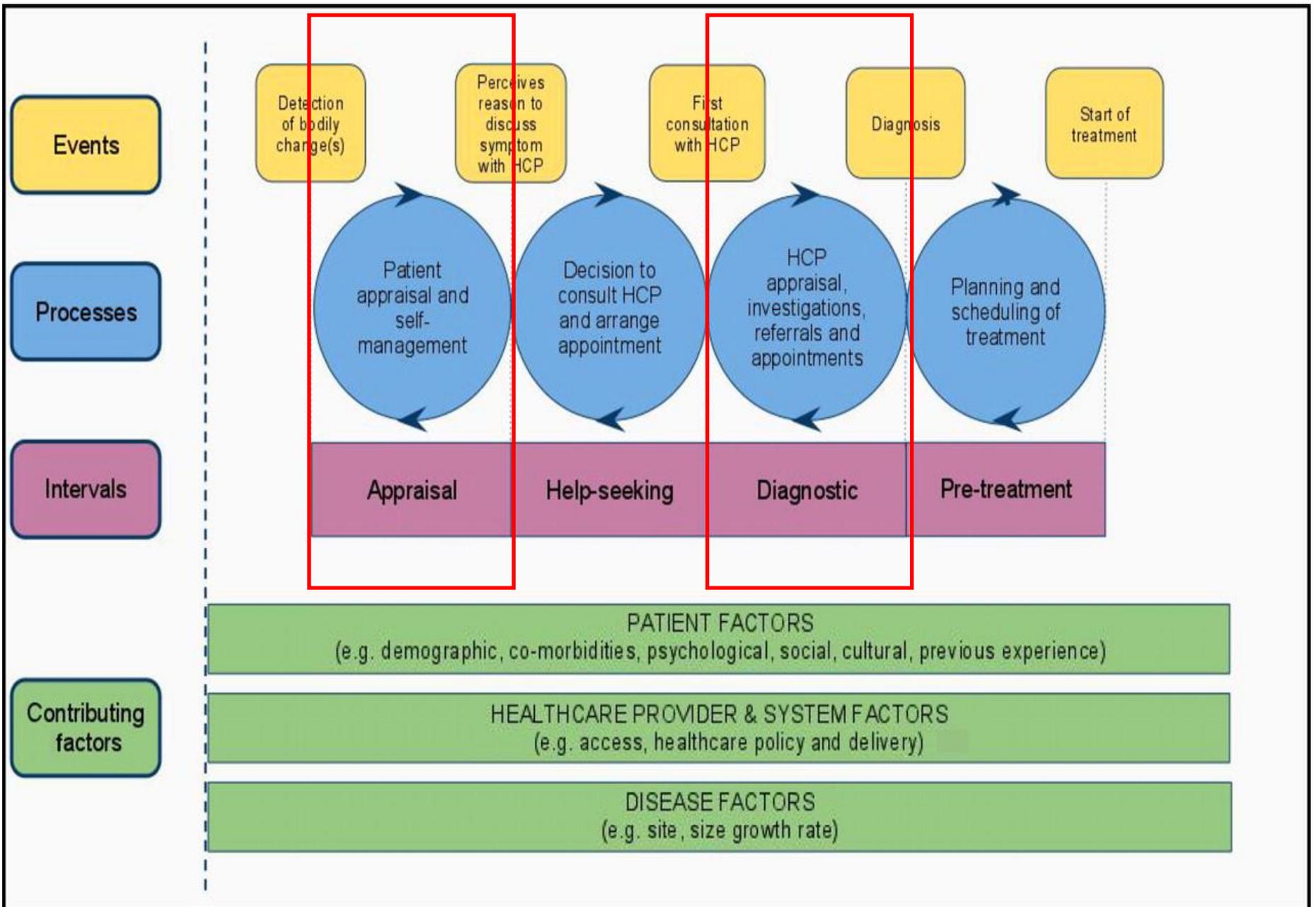
#Kitson et al 2008 PARiHS framework

# How can we achieve change?

- Need to look at the problem, knowledge and context differently
- Understand the process of delay and points of intervention
- Engage with stakeholders, including patients and families

# Contextual factors

- ‘the environment or setting in which the proposed change is to be implemented’
- Primary care – a challenging setting
  - Time-pressured
  - Competing demands
  - Politically complex
- Influence of parent, carer, significant others



# Areas of interest

- Understand barriers to paediatric assessment
- Education around referral pathways and guidance
- Clinical decision-making in paediatric practice
- Improve awareness of JIA and MSK conditions
- Targetting patients, families, schools, public



# What could be gained?

- Transferable messages
  - Other MSK diseases
  - Other chronic diseases in childhood
- Understanding knowledge exchange in the paediatric setting
- Potential to reduce delay

# Acknowledgements

- Academic paediatric rheumatology team led by Professor Helen Foster
- Clinical paediatric rheumatology team, Great North Children's Hospital
- Prof Rushmer
- Arthritis Research UK
- Patients and families

**Delay in diagnosing childhood arthritis – does it matter?****Sharmila Jandial, Consultant Paediatric Rheumatologist  
Great North Children's Hospital, Newcastle Upon Tyne**

Thursday 3rd July, 2014

*These are summary notes to accompany the presentation made by the above named speaker, as one of the knowledge exchange seminar series organised by the Fuse knowledge exchange group and is to be read in conjunction with the slide set, available on the Fuse website.*

Sharmila Jandial commenced her presentation by explaining that one of her interests was in how, and with what timeliness, research findings are adopted in practice and how clinicians' behaviours change. Slide 2 (headed Background) was used to illustrate the point that delays matter because musculoskeletal (MSK) problems are common, and untreated can lead to unavoidable joint damage and significant associated morbidity. The history and assessment are critical in guiding the treatment path taken from the initial consultation. Many MSK diseases are prone to delays in diagnosis, and this is also a global issue.

Slide 3 (headed Juvenile Idiopathic Arthritis) describes the main symptoms of this condition. Identifying this condition depends very much on the child themselves noticing and pointing out problems, if they are capable. The condition is more common than the public think and there is a significant potential for improvement, as there are now lots of treatment options, and chances of improvement are increased with early and aggressive treatment. Slide 4 (headed Delay in JIA) shows that delays in appointments have hardly changed in a decade (and are between 4-5 months and a year) and this is despite more treatments being available. The route to the consultant is complex and the reasons for delay are multi-factorial.

Slide 5 poses the question, "Why do we need to reduce delay?" The main reasons are about taking advantage of a treatment window to reduce inflammation, increase the quality of life and reducing joint restriction. In addition the experience for patients (and their doctors) is much better because patients are affected emotionally, as well as physically by the delay. Slide 6 depicts a general model of total patient delay (Andersen et al 1995), followed by a more expanded version breaking down the process in greater detail after Walter et al (2012, see Slide 7). On Slide 8, the element within the chain from first awareness of a problem to treatment initiation which the presentation then concentrated on, is highlighted in a red box. The element is HCP (*health care practitioner?*) appraisal, investigations, referrals and appointments. Dr Jandial pointed out that any reference to the patient means the patient and the carer for children. Slides 6-8, taken together, show the different stages of delay and complexity within each stage affected by how people make decisions at each stage. The whole process of decision making can be circuitous and messy.



Slide 9 shows six factors in diagnosis that have evidence to indicate their role in influencing the diagnostic interval. The following six slides examined each of these factors in more detail. Slide 10 lists the challenges of paediatric assessment. In respect of pre-verbal children understanding if anything is wrong, and if so, what, is a particular barrier. Clinician's reluctance to wake a sleeping child or inadvertent fears about making children cry also come into play, meaning that a full examination may not be undertaken. Slide 11 refers to uncertain referral pathways. One possibility is referral to going to the "wrong" speciality due to either complexity and/or sub-specialisation. Clinical signs may not be understood (Slide 12) it's important to know the normal changes in human development to be able to make the distinction from abnormal. Relying on history alone (Slide 13) can be insufficient, as the answers offered by either the child or the parent may not truly reflect the position and children find it difficult to speak about pain as a concept. Slide 14 covers the common but inaccurate misconception that children can't suffer from arthritis and putting problems erroneously down to growing pains. Slide 15 explores the low confidence some doctors have in their ability to undertake a MSK assessment. Primary care doctors in particular can be uncertain and in training sessions the context of their practice (physical arrangements and their working practices) need to be accommodated. Slide 16 draws all these factors together in one graphic and the key messages to be shared are listed on Slide 17. These are that children get arthritis (and other serious conditions), diagnosis relies on competent clinical skills and interpretation and that the right referral at the right time to the right service makes a difference.

Slide 18 moves on to the challenges of sharing knowledge in relation to enabling clinicians to be more effective in their diagnosis of juvenile arthritis. There are multiple barriers and facilitators and it's also important to know when it's best to intervene, given the complex diagnostic pathway. There has been a particular emphasis on education in clinical skills at many levels and this has largely been linear (ie; passing on the knowledge in a one way fashion) and results have been patchy (Slide 19). The example of a physical examination known by the acronym pGALS was then discussed. It consists of a two minute series of physical manoeuvres which is easier to implement in the secondary than the primary care setting (due to different pressures and constraints), consequently it's used in some practices but not all. This raises questions about how change can be brought about and what contextual factors are at work in the primary care setting. (See slides 20-23).

Slide 24 returns to the diagram depicted in Slide 8 with the addition of a box highlighting the first stage of patient appraisal and self-management which is the starting point. Slides 25 and 26 open up the possibility of widening awareness particularly in schools and amongst the public at large with the gains this could yield for reducing delay. Slide 27 lists acknowledgements Dr Jandial made on the day, to individuals, supporting clinical teams and the charity Arthritis UK.

## **Discussion**

Discussion took place on the following topics:

- The potential for links with other childhood conditions, for example, in the case of children with mental health problems there were similar issues connected with GP's



lack of confidence in diagnosis and it was suggested that there could be mutual learning across the two fields.

- Work about making services more accessible under the banner of the “You’re Welcome” project was raised. A GP young peoples’ group has been formed under the “You’re Welcome” heading. Reference was made to a sociological literature about doctors being afraid of making children cry in the clinical setting.
- There was a discussion about variations in how doctors talk to parents and children and debate about how this is learned and practiced. The relevance of the significant other (generally the parent) in determining the route the patient takes was noted and also the importance of schools and individual teachers in identifying problems.
- It was reinforced that diagnosis relied heavily on physical examination, this was critical in the absence of a diagnostic test/s.
- There was a discussion about the position of GPs in the whole diagnostic scenario. At the time of the seminar, the news about publication of cancer referral rates from primary care was to the fore and a point was made that there might be a tendency developing to scapegoat GPs for any issues to do with delayed referral. GPs face the task of sifting out the rare and it could be argued already recognise the issues about ascertaining the right referral to make. This led on to a discussion about some GPs being early adopters for pGALS and others not. Some GPs became engaged once they realised that this was a childhood condition. It was noted that pGALS had not been validated in primary care to date. Another suggestion made was that there could be a triage process in primary care whereby there is a GP or a therapist who has a special interest in the field and to whom other colleagues can direct their patients in the early stages.
- There was a discussion about the issue of standardisation and people being resistant to being told that there is a ‘standard’ approach. One solution to this could be a portfolio of approaches to suit different peoples’ ways of working. In addition what would success look like, in the absence of a clear measure? The scope for identifying success criteria and the links this might have with applications for future research funding was touched on, in conclusion.