Clinical guideline for the diagnosis and management of juvenile idiopathic arthritis

August 2009

Approved by NHMRC
on 12 June 2009

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Clinical guideline for the diagnosis and management of juvenile idiopathic arthritis

The National Health and Medical Research Council (NHMRC) is Australia's leading funding body for health and medical research. The NHMRC also provides the government, health professionals and the community with expert and independent advice on a range of issues that directly affect the health and wellbeing of all Australians.

The NHMRC provided support to this project through the Guidelines Assessment Register (GAR) process. The GAR consultant on this project was Karen Grimmer-Somers.

The guidelines were approved by the Chief Executive Officer of the NHMRC on 12 June 2009 under section 14A of the *National Health and Medical Research Council Act* 1992. Approval for the guidelines by NHMRC is granted for a period not exceeding 5 years, at which date the approval expires. The NHMRC expects that all guidelines will be reviewed no less than once every 5 years.

This publication was supported by funding from the Australian Government. The publication reflects the views of the authors and not necessarily reflects the views of the Australian Government.

Published by:
The Royal Australian College of General Practitioners
College House
1 Palmerston Crescent
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Fax 03 8699 0400
www.racgp.org.au

ISBN 978-0-86906-298-2

Published August 2009

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INTRODUCTION

In Australia, at least 5000 children are affected by juvenile idiopathic arthritis (JIA) at any one time.[1,2] Disease prevalence in Australia is between one and four cases per 1000 children.[3] Juvenile idiopathic arthritis can have significant associated morbidity and mortality. Long term follow up studies have revealed JIA carries the potential for longer term inflammatory activity and complications, leaving a lasting impact on the patient's function, growth and quality of life. Accurate and early diagnosis along with appropriate management and referral are essential for maximising patient outcomes and quality of life.

General practice plays an important role within the Australian health care system in prevention, early detection and chronic disease management (CDM). To manage chronic illness effectively requires well coordinated, patient centred care that is continuous, comprehensive, and consistent. General practitioners are well placed to provide this care and undertake this role in consultation with other medical specialists as required.[4–6] The role GPs play in CDM through multidisciplinary care coordination and long term care planning is recognised within the national Medicare rebate framework. Children with arthritis are eligible for broader funding arrangements under CDM items for GP Management Plans and associated reviews.[6]

As part of the Australian Federal Government's Better Arthritis and Osteoporosis Care (BAOC) 2006–2007 budget initiative,[7] guidelines for the management of osteoarthritis, rheumatoid arthritis, and juvenile idiopathic arthritis have been developed to inform evidence based primary care of chronic disease in general practice.

It is important that children presenting with JIA are diagnosed early; have initial management commenced by their GP; and are referred promptly to a paediatric rheumatologist. Because of the relatively low prevalence of JIA in the general population, GPs often develop little experience with the diagnosis or management of JIA. The *Clinical guideline for the diagnosis and management of juvenile idiopathic arthritis* has been developed to fill that gap.

The guideline presents recommendations to assist GPs managing patients with JIA. It focuses on short term care, long term care planning and management, and coordination of multidisciplinary care needs. The guideline includes algorithms and resources to assist with the implementation of the recommendations.

The guideline has been endorsed by the National Health and Medical Research Council (NHMRC).

This project was supported by The Royal Australian College of General Practitioners (RACGP) and the Australian Department of Health and Aging (DoHA). The following experts were involved in the development of the guideline as part of the RACGP Juvenile Idiopathic Arthritis Working Group:

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NOTE: All website references were current at the time of publication.

Expiry date for the recommendations

This guideline presents a comprehensive review of pharmacological and non-pharmacological management of JIA within the Australian health care context, based on the best available evidence up to January 2007. Evidence published after this date has not been reviewed for the guideline.

The guideline was approved by the CEO of the NHMRC on 12 June 2009, under section 14A of the *National Health and Medical Research Council Act* 1992. Approval for the guidelines by the NHMRC is granted for a period not exceeding 5 years. It is expected that the guideline will be reviewed, and revised if necessary, no less than once every 5 years. Review should be more frequent in areas where clinical practice or research is known to be changing rapidly. Readers should check with the RACGP for any reviews or updates of the guideline.

Commonly used abbreviations

ACR 30/50/70	American College of Rheumatology Paediatric 30/50/70 criteria
AE	adverse event
ANA	antinuclear antigen
BMC	bone mineral content
BMD	bone mineral density
CDM	chronic disease management
CHAQ	Childhood Health Assessment Questionnaire
CI	confidence interval
COX-2	cyclo-oxygenase-2 selective inhibitors
CRP	C-reactive protein
DMARDs	disease modifying antirheumatic drugs
EPC	enhanced primary care
ES	effect size: 0.2 small effect, 0.5 moderate effect, 0.8 large effect
ESR	erythrocyte sedimentation rate
FBC	full blood count
GIT	gastrointestinal
GP	general practitioner
HRQOL	Health related quality of life (usually measured on a self reported 10 cm VAS)
HLA	human leukocyte antigen
JIA	juvenile idiopathic arthritis
LFTs	liver function tests
NS	not statistically significant
NSAIDs	non-steroidal anti-inflammatory drugs
NHMRC	National Health and Medical Research Council
RACGP	[The] Royal Australian College of General Practitioners
RCT	randomised controlled trial
RhF	rheumatoid factor
ROM	range of movement/motion
SR	systematic review (also used in this report to describe meta-analysis)
TENS	transcutaneous electrical nerve stimulation
VAS	visual analogue scale
WMD	weighted mean difference

BACKGROUND

Juvenile idiopathic arthritis is a chronic, autoimmune, inflammatory joint disease. It is the most common rheumatic disease in children and adolescents. It is defined as 'persistent arthritis of unknown aetiology that begins before the age of 16 years and persists for at least 6 weeks'. It is diagnosed after excluding other causes.[8]

The cause of JIA is unknown. It is suspected that environmental factors such as viral infections may trigger the condition in genetically susceptible children.[9] However, it is unusual for more than one child in a family to have arthritis.

The aim of treatment is the induction of remission and control of the disease to minimise pain and function loss, and maximise quality of life. There is currently no cure for JIA. Treatment has altered as a result of recent research into the best practice approach to managing children. This guideline reflects the current evidence based approach to managing children with JIA.

Classification

There are seven subtypes of JIA:[8]

1. **Oligoarticular** – affects four or less joints. This is the most common subtype
2. **Polyarticular (RhF negative)** – when five or more joints are affected. Rheumatoid factor (RhF) antibody is not found on blood testing
3. **Polyarticular (RhF positive)** – when five or more joints are affected and RhF is found on blood testing. This subtype may behave similarly to rheumatoid arthritis in adults
4. **Systemic** – a chronic arthritis associated with systemic features, including high spiking fever, transient episodic erythematous rash, lymphadenopathy and hepatosplenomegaly (systemic features often precede the arthritis)
5. **Enthesitis related** – (previously known as juvenile spondyloarthropathy). This is a chronic arthritis associated with enthesitis (inflammation at insertion of tendons, ligaments or fascia to bone), or with lower axial skeletal involvement. HLA B27 is present or there is a family history of a first degree relative with a HLA B27 related disease. A significant proportion of patients will develop sacroiliitis as adults, but back and sacroiliac joint involvement is uncommon during childhood
6. **Psoriatic** – chronic arthritis, usually with asymmetrical involvement of small and large joints, and either the development of psoriasis or other evidence of a psoriatic diathesis (two of the following: family history in a first degree relative, nail pits or onycholysis, or dactylitis).
7. **Undifferentiated arthritis**.

Prognosis

For many years it was believed that most children eventually outgrow JIA. Now it is known that half of children with JIA will still have active arthritis 10 years after diagnosis unless treated appropriately. In moderate to severe cases, JIA can produce serious joint and tissue damage and cause problems with bone development and growth.[10] In some cases, JIA symptoms are mild and do not cause progressive joint disease and deformities.

In the past, JIA has often been seen as a benign condition, which it is not. Children presenting with JIA may be diagnosed inappropriately as having non-specific joint paints, 'growing pains' or recurrent musculoskeletal 'sprains'. As a result appropriate referral and treatment is delayed.

The outcomes for children with JIA are improved if managed by a multidisciplinary team with the input of a paediatric rheumatologist.[11,12]

Aim of the guideline

This guideline seeks to provide recommendations for the early diagnosis and multidisciplinary management of JIA in the primary care setting. The recommendations focus on the primary care practitioner's role in:

- early identification of JIA
- early referral to a paediatric rheumatologist
- prevention of complications associated with JIA
- alleviation/minimisation of pain
- optimal management of acute exacerbations of JIA
- prevention and minimisation of joint damage
- maximisation of function

- optimisation of normal growth and development
- improved quality of life.

Scope and target population

This guideline is intended for use in the primary health care setting by health care professionals working within a multidisciplinary team, including GPs and the following allied health professionals: physiotherapists; occupational therapists; sports medicine personnel; podiatrists; dieticians; psychologists; pharmacists; nurses and community health workers.

It is intended for patients under the age of 16 years presenting with arthritic symptoms, as well as those diagnosed as having JIA. It does not cover patients over the age of 16 years, treatment of extra-articular disease, or surgical interventions.

This guideline has been developed for use in primary care settings in metropolitan, regional, rural and remote areas of Australia.

Focus of the guideline

The guideline focus is on JIA. It does not cover the management of other forms of arthritis, or complex or unusual conditions. The following process model (*Figure 1*) identifies the stages in CDM and the focus of the guideline.

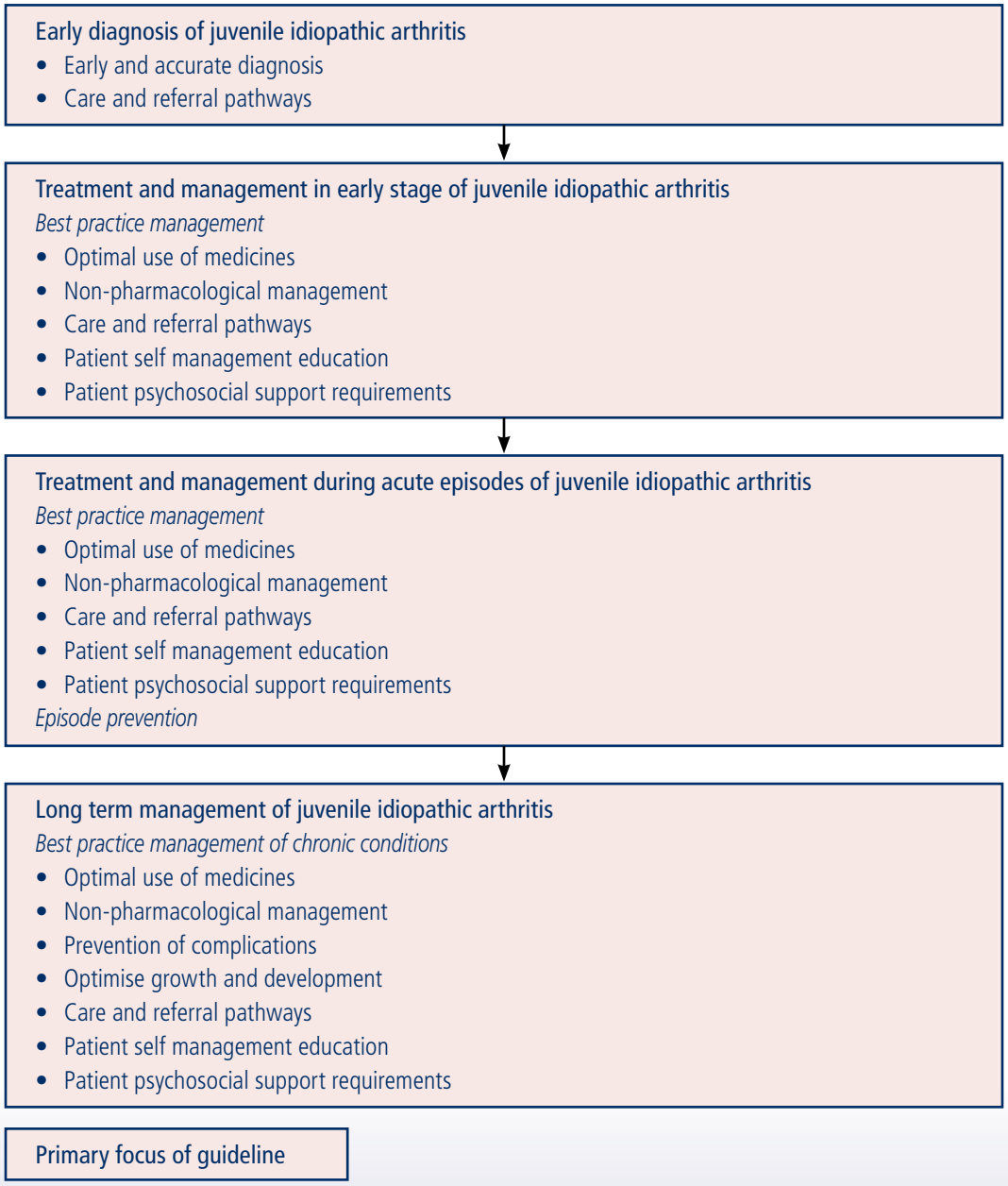


Figure 1. Stages in CDM and the focus of the JIA guideline

Methods

The process used to develop the guideline is outlined in full detail in the Process Report (*Appendix A*). This guideline is based on an evidence based literature review conducted to NHMRC requirements. The RACGP JIA Working Group, who has overseen the development of the guideline and supporting documents, comprised rheumatologists, GPs, consumer representatives, arthritis organisation representatives and an NHMRC advisor. The evidence for the guideline is based on:

1. A review of the literature identified through a systematic search of evidence published from January 2000 to January 2007
2. A national guideline for JIA[3] which was assessed using the AGREE instrument[13] and identified as being the most appropriate, recently published guideline to use as a primary reference
3. The Working Group's expert opinion
4. Australian paediatric rheumatology expert opinion.

Literature review

The method used to conduct the evidence based literature review is outlined in full in the Process Report (*Appendix A*). A search of MEDLINE, EMBASE, CINAHL and the Cochrane Library for English language publications from January 2000 to January 2007 was performed. An additional manual search was used to identify evidence for interventions not represented in the initial search or not covered by the primary reference guidelines. Articles were also identified through review of reference lists of retrieved papers and research known to RACGP Working Group members. Papers were initially selected for inclusion based on reading the title and/or the abstract. Included literature was limited to Level 1 and Level 2 evidence graded according to the *NHMRC additional levels of evidence and grades for recommendations for developers of guidelines* (2005).[14] For areas where randomised controlled trials (RCTs) or systematic reviews (SRs) were not available, lesser levels of evidence and expert opinion were sourced. Papers that met the inclusion criteria were critically appraised using checklists developed by SIGN and given an overall quality grade of high, moderate or low. Findings from the literature were reported descriptively and in a tabulated format. The full methods and findings are presented in *Juvenile idiopathic arthritis: a literature review of recent evidence* (www.racgp.org.au/guidelines/juvenileidiopathicarthritis/literaturereview).

Recommendations

The method used to develop and grade recommendations is outlined in full in the Process Report (*Appendix A*). Recommendations were based on the literature review and primary reference guideline. The RACGP Working Group developed evidence statements from which each recommendation was developed, available in *Recommendations for the diagnosis and management of juvenile idiopathic arthritis* (www.racgp.org.au/guidelines/juvenileidiopathicarthritis/recommendations). Each recommendation statement is supported by a grading that reflects the strength of the recommendation and implementability in terms of trust or confidence practitioners can use in a clinical situation. The recommendation gradings used throughout the guideline are based on *NHMRC additional levels of evidence and grades for recommendations for developers of guidelines*[14] presented in *Table 1*.

Table 1. Recommendation grades[14]

A	Excellent evidence – body of evidence can be trusted to guide practice
B	Good evidence – body of evidence can be trusted to guide practice in most situations
C	Some evidence – body of evidence provides some support for recommendation(s) but care should be taken in its application
D	Weak evidence – body of evidence is weak and recommendation must be applied with caution

The overall grade of each recommendation is based on a summation of an appraisal of individual components of the body of evidence on which the recommendation is based, including volume and consistency of the evidence. *Table 2* shows the body of evidence assessment matrix, listing all the components that were considered when assessing the body of evidence, together with the grades used.[14] The volume of evidence was defined to reflect the levels of evidence considered for this project. In reaching an overall grade, recommendations did not receive a grading of A or B unless the volume and consistency of evidence components were both graded either A or B. Overall grades were reached through consensus consideration of the grading for each component listed below.

Table 2. Body of evidence assessment matrix[14]

Component	A	B	C	D
	Excellent	**Good**	**Satisfactory**	**Poor**
Volume of evidence	Several Level I or Level II studies with low risk of bias	One or two Level II studies with low risk of bias or a SR of multiple Level III studies with low risk of bias	Level III studies with low risk of bias or Level II studies with moderate risk of bias	Level IV studies or Level I–III studies with high risk of bias
Consistency	All studies consistent	Most studies consistent and inconsistencies may be explained	Some inconsistency reflecting genuine uncertainty around clinical question	Evidence is inconsistent
Clinical impact	Very large	Substantial	Moderate	Slight or restricted
Generalisability	Population/s studied in body of evidence are the same as the target population for the guideline	Population/s studied in the body of evidence are similar to the target population for the guideline	Population/s studied in the body of evidence different to the target population for the guideline but it is clinically sensible to apply this evidence to the target population (eg. results in adults that are clinically sensible to apply to children)	Population/s studied in the body of evidence different to the target population for the guideline and hard to judge whether it is sensible to generalise to the target population
Applicability	Directly applicable to Australian health care context	Applicable to Australian health care context with few caveats	Probably applicable to Australian health care context with some caveats	Not applicable to Australian health care context

The guideline

The guideline has been designed to provide clear information to assist clinical decision making and support optimal patient care. It is based on the best evidence available up to January 2007. Where appropriate, the evidence has been interpreted with regard to the Australian context in which the guideline will be implemented. It is intended that the guideline be considered according to the limitations outlined in section 7 and used in conjunction with clinical judgment and patient preference. The guideline consists of:

Algorithms

The two algorithms summarise the main recommendations of the guideline and provide an accessible desktop reference. The algorithms provide detailed flow charts for the diagnosis and the management of JIA.

Recommendations

The 21 recommendations contained in this guideline are limited to patients under the age of 16 years presenting with arthritic symptoms, as well as those diagnosed as having JIA. They do not cover the management of other forms of arthritis, complex or unusual conditions, or give detailed guidance on pharmacological therapy in JIA. The recommendations have been developed on the basis of the best evidence available up to January 2007.

Each recommendation has been graded according to the *NHMRC additional levels of evidence and grades*[14] (from A to D).The grade reflects the degree of 'trust' that the clinician can place in the clinical application of the recommendation. Each recommendation is supported by a summary of the evidence.

The RACGP Working Group supports all 21 recommendations and intends that they be used in conjunction with clinical judgement and patient preferences. The full grading and evidence base for each recommendation is available in *Recommendations for the diagnosis and management of juvenile idiopathic arthritis* (www.racgp.org.au/guidelines/juvenileidiopathicarthritis/recommendations).

Good practice points

Where appropriate, recommendations are followed by good practice points. The good practice points are essential tips on how to effectively implement the recommendations. These points are followed by a summary of the evidence for each recommendation.

Resources

Useful references and supporting information are provided throughout the guideline. *Appendix B* contains additional resources, as well as contact details for organisations providing services and support to both patients with JIA and their carers.

The RACGP Working Group recommends consulting the National Prescribing Service (www.nps.org.au), the Rheumatology Therapeutic Guidelines (www.tg.com.au), the Paediatric Handbook (www.rch.org.au/paed_handbook) and the Australian Medicines Handbook (amh.hcn.net.au) for detailed prescribing information including:

- ***indications***
- ***drug dosage***
- ***method and route of administration***
- ***contraindications***
- ***supervision and monitoring***
- ***product characteristics.***

Limitations of the guideline

Medication information

The literature search was not designed to retrieve safety trials for pharmacological interventions. The guideline does not seek to provide full safety and usage information on pharmacological interventions. The pharmacological interventions outlined in the guideline should not be applied without consideration to the patient's clinical profile and personal preferences. The Working Group recommends consulting the sources listed above.

Search date

The guideline is based on the best evidence available up to January 2007. Evidence published after this date has not been reviewed or considered for the guideline.

Interventions included

The initial search strategy was limited to include only papers graded as Level 1 or Level 2 evidence and expanded to include lower levels of evidence for interventions where no high level evidence was found. Interventions that have not been included in the recommendations may not have had readily identifiable literature related to their use. The guideline is not intended to confirm or refute the effectiveness, nor provide guidance on the use of interventions that have not been included, as the evidence has not been reviewed.

Lack of evidence

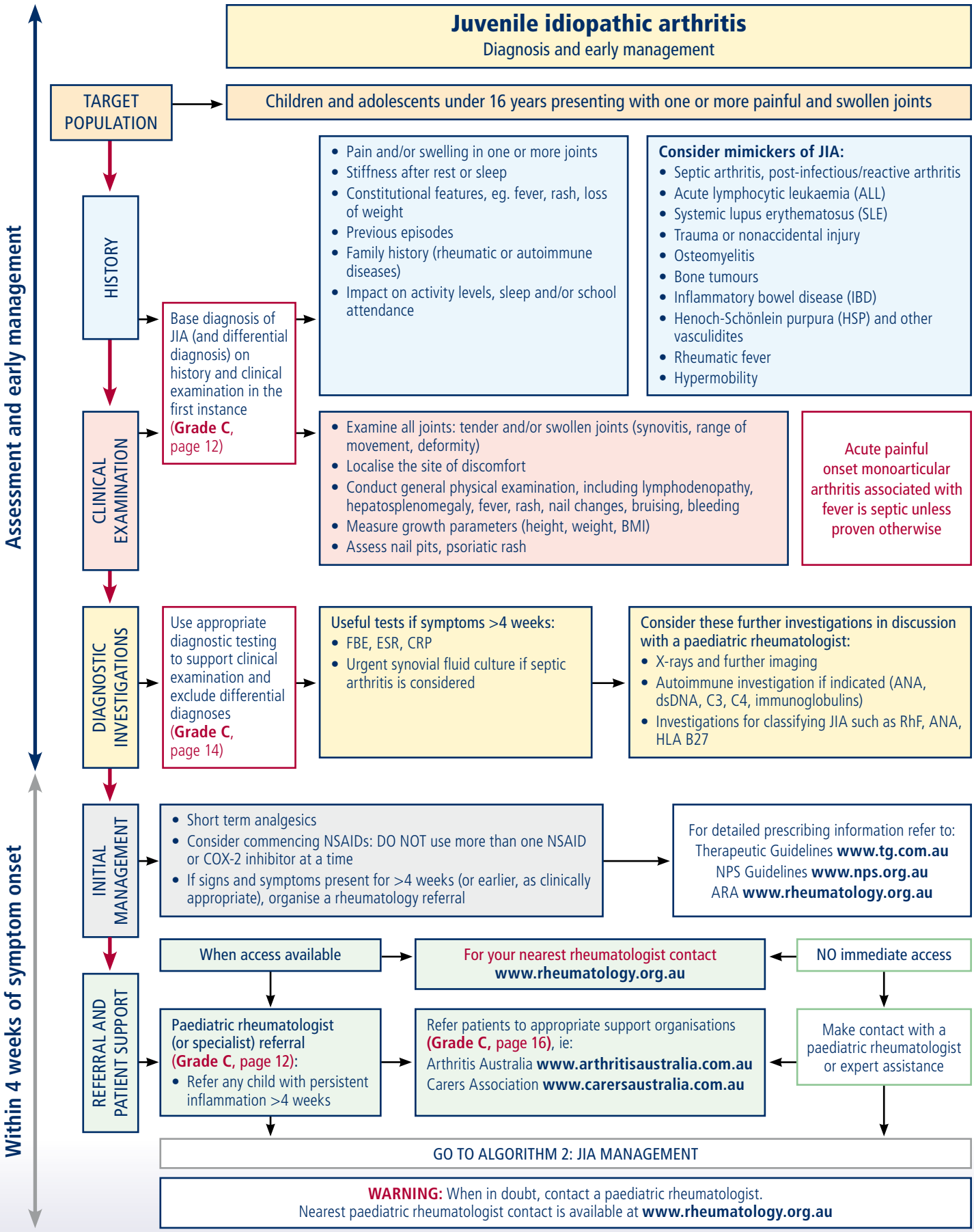
For some interventions included in the recommendations there was limited evidence from which to draw conclusions on the intervention's effectiveness. The Working Group acknowledges that lack of evidence is not evidence of lack of effect, and has attempted to reflect this in the strength of the grading given to recommendations on interventions that are not supported. In addition, some interventions were not supported in the recommendations due to lack of evidence of effect. The Working Group acknowledges that this refers to lack of evidence of effect over placebo, that is, patients may receive some beneficial outcomes from the intervention, however these do not exceed beneficial effects that can be expected from a placebo therapy.

Cost effectiveness

As part of the brief, the evidence for this guideline does not include the cost effectiveness of the recommended practice versus current/established practice. It also does not cover the economic feasibility of the recommendations.

ALGORITHMS

Diagnosis and early management of JIA



Juvenile idiopathic arthritis

Management

The goals of treatment for JIA are to control inflammation, relieve pain, prevent or control joint damage, maximise functional abilities and manage complications

0–6 weeks

DIAGNOSIS (see Algorithm 1) → Confirm diagnosis, start initial treatment and organise paediatric rheumatology referral

MANAGEMENT

- Assess individual's needs and access to a range of health professionals including but not limited to paediatric rheumatologist, physiotherapist, occupational therapist, mental health worker, ophthalmologist, orthopaedic surgeon, dietician, podiatrist, chiropodist, pharmacist, dentist, school counsellor, hand therapist, indigenous health worker and social worker (**Grade D**, page 15)
- All patients should have a care plan (**Grade D**, page 15)

PHARMACOLOGICAL INTERVENTIONS

Basic therapy (in consultation with paediatric rheumatologist)
Each JIA subtype may require a different approach to therapy. The main three subtypes of JIA are: oligoarthritis, polyarthritis, and systemic onset.

- Simple analgesics (eg. paracetamol) (**Grade C**, page 17)
- Traditional NSAIDs (**Grade B**, page 18)
- Consider weak opioids (eg. codeine) (**Grade D**, page 17)

For detailed prescribing information refer to:
Therapeutic Guidelines
www.tg.com.au
NPS Guidelines
www.nps.org.au
ARA
www.rheumatology.org.au

Advanced therapy (Prescription by paediatric rheumatologist)

- Intra-articular, oral or IV corticosteroids
- DMARDs (methotrexate, leflunomide, sulfasalazine, hydroxychloroquine)
- Biological modifying agents (bDMARDs) (page 19)

NON-PHARMACOLOGICAL INTERVENTIONS

- Adequate diet including daily calcium and vitamin D intake (**Grade B**, page 20)
- Calcium supplementation (together with vitamin D when on corticosteroids) (**Grade B**, page 20)
- Land based exercise programs (**Grade C**, page 20)
- Aquatic exercise (**Grade C**, page 21)
- Thermotherapy (heat/ice packs, warm baths) (**Grade D**, page 23)
- Consider splints (**Grade D**, page 22)
- Consider foot orthotics (**Grade D**, page 22)

6–12 weeks

MONITORING AND SUPPORT

Psychosocial support and education (**Grade C**, page 16)

- Psychosocial support for patients, family members, and carers
- Ongoing education on disease management and medications
- Refer patients to support organisations such as Arthritis Australia **www.arthritisaustralia.com.au** and Carers Association **www.carersaustralia.com.au** Tel 1800 242 636

Ongoing monitoring of JIA (**Grade C**, page 24)

- Optimise multidisciplinary communication
- Assess arthritis activity at least three times per year and adjust therapy to maintain swollen and tender joint count as low as possible
- Monitor toxicity and medication side effects
- Paediatric rheumatology review at least twice per year
- Adjust medication in consultation with paediatric rheumatologist

Manage comorbidities (**Grade C**, page 24)

- Manage comorbidities
- Regular ophthalmological review for JIA related diseases treated with long term corticosteroids
- Preventive medicine: immunisations, weight, growth, nutrition, dental
- Monitor sleep, fatigue, mood, school progress, peer and age appropriate activity

Ongoing

MANAGEMENT OF ACUTE FLARE EPISODES

- Determine if persistent flare
- Manage short term flares symptomatically with analgesics +/– NSAIDs, heat, and relative rest
- Beware of infection if on immunosuppressive medication and evaluate for co-infection
- If persisting flare or other concerns, contact a paediatric rheumatologist or rheumatology team for advice

WARNING: Treatment may cause serious adverse effects. Physicians and patients/carers must monitor for signs and symptoms of potential toxicity through regular clinical and laboratory review.

SUMMARY OF RECOMMENDATIONS

Note: There is one recommendation advising GPs not to use topical NSAIDs (highlighted in RED).

Recommendation 1 – Early diagnosis (Grade C)

General practitioners should aim to diagnose JIA as early as possible in order to optimise outcomes for patients.

Recommendation 2 – Referral (Grade C)

Referral to a rheumatologist is advised for patients with confirmed or suspected JIA whose symptoms persist beyond 4 weeks. Early referral enables aggressive intervention with disease modifying drugs, which reduces long term joint damage and disability.

Recommendation 3 – Clinical examination (Grade C)

General practitioners should base diagnosis of JIA (and differential diagnosis) on history and clinical examination in the first instance, with strong suspicion of JIA indicated by:

- pain and swelling of single or multiple joints
- persistent or worsening loss of function
- fever of at least 10 days with unknown cause, often associated with transient erythematous rash
- decreased range of motion (ROM)
- joint warmth
- effusion.

Recommendation 4 – Diagnostic investigations (Grade C)

In early assessment of patients presenting with painful and swollen joint(s), GPs should support clinical examination with appropriate tests to assist in increasing diagnostic certainty, excluding differential diagnoses, and predicting patients likely to progress to erosive disease. Base investigations usually include:

- ESR or C-reactive protein (CRP)
- Full blood count (FBC).

Consider rheumatoid factor (RhF), antinuclear antigen (ANA), human leukocyte antigen (HLA) B27, and plain radiographs. Depending on the clinical picture, additional investigations may be required.

Recommendation 5 – Multidisciplinary care (Grade D)

General practitioners should encourage and support a management approach based on individual patient need and involving a multidisciplinary team of health professionals.

Recommendation 6 – Care plans (Grade D)

Involvement of the multidisciplinary team should be managed through the development of individual care plans.

Recommendation 7 – Patient information (Grade C)

General practitioners should provide ongoing, tailored information to support their patients' understanding of their disease, treatment options, possible outcomes and their role in self management, and encourage patients and their carers to seek appropriate information and education opportunities according to their individual needs.

Recommendation 8 – Patient support (Grade C)

General practitioners should provide ongoing psychosocial support and encourage patients and their carers to seek support from appropriate sources according to their individual needs.

Recommendation 9 – Simple analgesics (Grade C)

General practitioners should consider using paracetamol in regular divided doses for treating moderate pain in children and adolescents with JIA.

Recommendation 10 – Weak opioids (Grade D)

General practitioners could consider prescribing codeine in regular divided doses, in addition to paracetamol, for treating moderate articular pain in children and adolescents with JIA.

Recommendation 11 – Traditional NSAIDs (Grade B)
General practitioners should prescribe NSAIDs as the initial drug of choice for reducing inflammation and associated pain in the treatment of JIA.

Recommendation 12 – Topical NSAIDs (Grade D)
General practitioners should NOT prescribe topical NSAIDs to treat patients with JIA.

Recommendation 13 – Complementary/alternative medicines (Grade D)
General practitioners could inform patients and their families that, although there has been no research in children with JIA, there is limited or no evidence of effectiveness above placebo of complementary/alternative medicines in adult populations with arthritis.

Recommendation 14 – Nutritional therapy – calcium (Grade B)
General practitioners should monitor calcium intake in children with JIA, and provide advice on increasing daily calcium intake. General practitioners could consider treating some patients with JIA with oral calcium and vitamin D supplementation.

Recommendation 15 – Land based exercise (Grade C)
General practitioners should encourage patients with JIA to engage in regular physical activity compatible with their general abilities and restrictions of their disease.

Recommendation 16 – Aquatic exercise (Grade C)
General practitioners could inform patients about aquatic exercise for children and adolescents, and its limited effects.

Recommendation 17 – Splints (Grade D)
General practitioners could inform patients about use of splints and make individualised recommendations in conjunction with appropriately trained multidisciplinary health professionals.

Recommendation 18 – Foot orthoses (Grade D)
General practitioners could inform patients with JIA in the lower limb about the role of comfortable, supportive shoes. General practitioners could inform patients about the use of foot orthotics based on an individualised assessment, safety, and personal preference, in conjunction with appropriately trained multidisciplinary health professionals.

Recommendation 19 – Thermotherapy (Grade D)
General practitioners could consider recommending the use of heat and cold packs, warm baths and/or ice massage for the symptomatic relief of JIA in children and adolescents.

Recommendation 20 – Complementary/alternative physical therapies (Grade D)
General practitioners could inform patients and their families who seek advice that there is no research on complementary/alternative physical therapies in children with JIA.

Recommendation 21 – Disease monitoring (Grade C)
General practitioners should be involved in monitoring disease progression and managing comorbidities in conjunction with the treating paediatric rheumatologist.

RECOMMENDATIONS FOR PRIMARY CARE OF JIA

Diagnosis of JIA

Early diagnosis and referral

Recommendation 1 – Early diagnosis (Grade C)
General practitioners should aim to diagnose JIA as early as possible in order to optimise outcomes for patients.

Recommendation 2 – Referral (Grade C)
Referral to a rheumatologist is advised for patients with confirmed or suspected JIA whose symptoms persist beyond 4 weeks. Early referral enables aggressive intervention with disease modifying drugs, which reduces long term joint damage and disability.

Good practice points

- For advice about accessing a paediatric rheumatologist, either privately or through public clinics, contact the Australian Rheumatology Association at www.rheumatology.org.au.
- When making a referral for a newly diagnosed or suspected case of JIA, make initial telephone contact with a rheumatologist and mark the referral **URGENT** (recent onset JIA).

In JIA, persistent synovitis leads to joint deformity and destruction, and may occur less than 2 years following onset of disease. Disruption of proper joint function predisposes children and young adults to premature osteoarthritis and a potential of lifetime disability.[15] An early, accurate diagnosis and disease modifying therapy is essential in order to commence appropriate management aimed at promoting normal growth and development, and to minimise disability and deformity.[2]

Early referral to a paediatric rheumatologist is recommended in the literature. With a large range of differential diagnoses for possible JIA, assessment by a paediatric rheumatologist at an early stage should be sought to confirm diagnosis.[15]

It is the consensus of the RACGP Working Group that patients with symptoms persisting beyond 4 weeks and indicative of JIA be referred to a rheumatologist to enable early initiation of therapy.

History and clinical examination

Recommendation 3 – Clinical examination (Grade C)
General practitioners should base a diagnosis of JIA (and differential diagnoses) on history and clinical examination in the first instance, with strong suspicion of JIA indicated by:

- pain and swelling of single or multiple joints
- persistent or worsening loss of function
- fever of at least 10 days with unknown cause, often associated with transient erythematous rash
- decreased range of motion (ROM)
- joint warmth
- effusion.

The international literature[15,16] and the Australian national JIA guideline[3] recommends that diagnosis of JIA should be based primarily on comprehensive history taking, complete clinical examination, and the ordering appropriate diagnostic tests.

Patients commonly present with pain and stiffness in one or more joints. Juvenile idiopathic arthritis should be particularly suspected in patients who present with persistent joint pain and swelling. In most patients, symptoms emerge over weeks to months. Musculoskeletal symptoms and signs are common in children and adolescents and may be the presenting feature of a broad spectrum of conditions. Clinical features and laboratory findings may be relatively non-specific in rheumatological conditions, and it is important to look for disease patterns when evaluating the presenting complaint and conducting a systems review.

Primary care physicians should particularly note that the absence of key symptoms, signs, or positive test results, does not necessarily rule out a diagnosis of JIA. Thus, ongoing monitoring and early specialist referral should be considered for patients with persistent symptoms.

It is the consensus of the RACGP Working Group and Australian paediatric rheumatology experts that diagnosis of JIA should be based primarily on careful history taking and clinical examination. Patients commonly present with pain and stiffness in one or more joints. Juvenile idiopathic arthritis should be particularly suspected in patients who present with persistent joint pain and swelling. In most patients, symptoms emerge over weeks to months.

The Working Group has developed the following history and clinical examination checklist (see also *Appendix B*):

History

- Pain or swelling in one or more joints is present
- Check the nature of onset – is it acute or insidious? Previous episodes?
- **Acute onset monoarticular arthritis associated with fever is septic until proven otherwise**
- Check the timing of symptoms during the day – as a general guide:
- Early morning stiffness/stiffness after rest or sleep = inflammatory
- Post-activity pain = mechanical
- Check duration of illness – if >6 weeks it is less likely to be reactive/post-viral arthritis
- Are there any concurrent infections (respiratory, enteric or skin)? Post-viral infections are probably the most common cause of transient arthritis
- Check constitutional features (eg. fever, rash, loss of weight)
- Has the child been taking any medications (eg. cefaclor)?
- What does the child, or parent consider to be the most symptomatic site – is it in the joint, muscle, adjacent bone or a more diffuse area?
- Check for extra-articular symptoms – ensure a thorough systems review and keep the following diagnoses in mind:
  - septic arthritis
  - post-infectious/reactive arthritis
  - systemic lupus erythematosus (SLE)
  - acute lymphoblastic leukaemia (ALL)
  - trauma/non-accidental injury
  - osteomyelitis
  - bone tumour
  - inflammatory bowel disease (IBD)
  - Henoch-Schönlein purpura and other vasculitides
  - rheumatic fever
- Assess whether normal activity levels or interests have been interrupted
- Assess the functional milieu of the patient (eg. school progress and attendance, sleep pattern, family and peer relationships and stress experiences)
- Check the family history for other types of inflammatory arthritis, particularly the spondyloarthropathies, autoimmune disorders and pain syndromes (eg. fibromyalgia or other models for pain behaviour).

Examination

Observe the patient as they move about the room looking for limitations or alterations in function and be opportunistic when examining them.

- Examine all joints, not only the site of the presenting complaint. There may be inflammation without symptoms in JIA
- Aim to localise the site of maximal discomfort (eg. is it the joint capsule, adjacent bone or muscle belly, tendon or ligament attachments?)
- Examine for signs of systemic diseases with an articular component, extra-articular features of JIA, or both. In particular examine the skin, eyes, abdomen, nails and lymph nodes.

A **musculoskeletal assessment** should include:

- Joints – signs of inflammation such as swelling or tenderness, the range of movement and deformity. Joints affected by JIA are typically swollen, may be tender to touch and warm but are usually not erythematous

- Entheses – bone attachment sites of ligaments/tendons (eg. Achilles tendon)
- Tendon sheaths of fingers and toes (eg. dactylitis in psoriasis)
- Gait – antalgic (pain) or limp, Trendelenburg sign
- Muscle tenderness, muscle wasting or weakness (eg. inability to toe or crouch walk)
- Patellar tracking pattern – does the patella move vertically on walking?
- Shoe sole and heel wearing pattern
- Leg length measurement
- Spinal flexion, including Schober test (the measurement of the lumbosacral range should increase in distance by at least 6 cm on maximal flexion with knees straight; the starting range is a point 5 cm below and 10 cm above the lumbosacral junction (use the Dimples of Venus as a guide)
- Assessment of growth parameters.

Differential diagnosis

Juvenile idiopathic arthritis can resemble any disorder causing acute or chronic polyarthritis in children. Elimination of other diseases is therefore a necessary step in the diagnosis of JIA.

Diagnostic investigations

Recommendation 4 – Diagnostic investigations (Grade C)

In early assessment of patients presenting with painful and swollen joint(s), GPs should support clinical examination with appropriate tests to assist in increasing diagnostic certainty, excluding differential diagnoses, and predicting patients likely to progress to erosive disease. Base investigations usually include:

- ESR or CRP
- FBC.

Consider RhF, ANA, HLA B27, and plain radiographs. Depending on the clinical picture, additional investigations may be required.

Good practice points

- Before JIA is diagnosed all other known conditions and causes of childhood arthritis must be excluded.
- Absence of any key symptoms, signs, or test results does not necessarily rule out a diagnosis of JIA.

There is no single test that accurately diagnoses JIA. A number of tests are useful in increasing diagnostic certainty, excluding other forms of arthritis, predicting patients likely to progress to erosive disease, and monitoring disease progression. Various tests also have a role in monitoring disease progression.

Given the level of distress and anxiety procedures may have for children, coordinating procedures and optimising procedural pain management through appropriate preparation of child and parent, use of topical local anaesthetic agents, and non-pharmacological techniques such as distraction and relaxation/breathing should be implemented.

Erythrocyte sedimentation rate and CRP indicate an inflammatory process but have low specificity for JIA. One of these tests is usually performed.[8] These markers are usually elevated in JIA but may be normal. The RhF test is not conclusive. RhF is positive in only a small percentage of JIA patients (unlike rheumatoid arthritis). However, when present in combination with other factors, the level of RhF may indicate the likelihood for aggressive disease progression and a poorer prognosis.[17,18]

A FBC is usually undertaken to provide general information relating to inflammation and anaemia. (Working Group)

Plain X-rays have been key investigations in identifying erosions, and predicting disease; however, erosions are not often apparent in disease of less than 3 months duration.[7] Serial X-rays over years may show disease progression and therefore indicate the need for a change in treatment strategy. Other imaging is unlikely to be helpful at diagnosis and should be ordered only in discussion with a paediatric rheumatologist.

It is the consensus of the RACGP Working Group and Australian paediatric rheumatology experts that GPs should support clinical examination with appropriate testing to support diagnostic certainty, exclude differential diagnoses and to predict patients likely to develop erosive disease.

Management of JIA

General principles

General practitioners should contribute to a management approach based on the following therapeutic goals:

- preservation of function and quality of life
- minimisation of pain and inflammation
- joint protection, and
- control of systemic complications.

Multidisciplinary care and care planning

Recommendation 5 – multidisciplinary care (Grade D)

General practitioners should encourage and support a management approach based on individual patient need and involving a multidisciplinary team of health professionals.

Recommendation 6 – care plans (Grade D)

Involvement of the multidisciplinary team should be managed through the development of individual care plans.

Good practice points

- GPs may utilise Enhanced Primary Care (EPC) items to facilitate access to appropriate services (see *Resources*). Eligible services include, but are not limited to, those provided by physiotherapists, osteopaths, occupational therapists, podiatrists, mental health workers, indigenous health workers, chiropractors, chiropodists, audiologists and speech pathologists.
- GPs should refer carers to their state/territory Carers' Association for information and support as part of the ongoing management plan (see *Resources*).
- Include pain management and assessment of sleep quality in the treatment plan.

There is strong support for a multidisciplinary approach in the management of JIA to ensure all of the child's needs are met, including normal growth, social development and physical functioning.[2,15,19,20] A wide range of interventions implemented by multidisciplinary health care providers were reviewed for these recommendations. In the vast majority of trials the intervention of interest was implemented by a health care provider with specific training and qualifications. Seeking health advice and management from an appropriately trained health care provider is considered to be a component of effective and safe therapy. A wide range of multidisciplinary health care providers may be considered in the co-management of patients with JIA, including but not limited to:

- physiotherapists
- occupational therapists
- mental health specialists
- ophthalmologists
- podiatrists or orthotists
- orthopaedic surgeons
- social workers
- pain management teams
- indigenous health workers
- community nursing teams.

The child's family and school are also essential elements in ongoing management of the child. Family should be involved in all aspects of care, and the psychosocial needs of carers should also be considered, as this influences the ongoing management of the child patient.[19] Liaison with the school (eg. principal, physical education teacher, school liaison worker) is important, particularly if special arrangements are relevant (eg. extra time for school work).[2,15,20]

It is the consensus of the RACGP Working Group that the GP, rheumatologist and multidisciplinary team should aim to engage the patient in an individualised care plan, agreeing on treatment goals that include an objective measure of disease.

Patient education and psychosocial support

Recommendation 7 – patient information (Grade C)

General practitioners should provide ongoing, tailored information to support patient understanding of their disease, treatment options, possible outcomes and their role in self management, and encourage patients and their carers to seek appropriate information and education opportunities according to their individual needs.

Recommendation 8 – patient support (Grade C)

General practitioners should provide ongoing psychosocial support and encourage patients and their carers to seek support from appropriate sources according to their individual needs.

Good practice points

- Joint protection, energy conservation, and problem solving skills training should be taught early on in the disease course.
- GPs may access medication information for patients from the Australian Rheumatology Association (ARA) or may refer patients and their carers (see *Resources*).
- Referral to Arthritis Australia is recommended for general disease and treatment information, as well as support services (see *Resources*).

The evidence of the impact of patient education and psychosocial support remains limited, but most literature agrees that while it is important to appropriately manage the medical aspects of arthritis in children, it is equally important to provide psychosocial interventions such as:[15]

- patient/family education
- psychosocial interventions/support services
- community resources
- school based resources
- information and referral regarding insurance coverage and benefit coordination.

Education and behavioural interventions are important with specific interventions. For example, there is strong evidence that a behavioural intervention involving group education sessions can have a positive impact on increasing dietary calcium intake.[21]

Munro and Murray[2] include education and support among the important aspects of effective JIA treatment. Hashkes and Laxer[18] agree that the treatment of JIA combines anti-inflammatory and immunomodulatory medications with physical and occupational therapy, an occasional need for surgery, nutritional support, and psychosocial and educational partnership with patients and parents.

It is the consensus of the RACGP Working Group that the above interventions represent important aspects of the general management of JIA and should be encouraged among all members of the multidisciplinary team. When in doubt, contact a paediatric rheumatologist. The nearest paediatric rheumatologist contact is available from www.rheumatology.org.au.

Pharmacological interventions

The RACGP Working Group recommends consulting the National Prescribing Service (www.nps.org.au), the Rheumatology Therapeutic Guidelines (www.tg.com.au), the Paediatric Handbook (www.rch.org.au/paed_handbook) and the Australian Medicines Handbook (amh.hcn.net.au/) for detailed prescribing information.

Simple analgesics (eg. paracetamol)

Recommendation 9 – simple analgesics (Grade C)
General practitioners should consider using paracetamol in regular divided doses for treating moderate pain in children and adolescents with JIA.

Good practice points

- Use the correct dose for the child's weight. In overweight children, the ideal body weight, rather than actual weight, should be used to calculate dose.
- Chronic dosing should not exceed 48 hours without medical advice.
- Maximum daily dose is 90 mg/kg to a maximum of 4 g/day for treating persistent pain in children and adolescents with JIA.

Cautions

- Paracetamol has few side effects, but dosing is limited by possible hepatotoxicity.
- Do not rely on dosing charts supplied by the manufacturer as these tend to underdose the child.
- Do not use slow release preparations in infants and children.

There is an accepted role for simple analgesics in managing pain in JIA, although the evidence supporting effectiveness specifically in JIA is limited. Paracetamol remains the analgesic of choice for treating persistent pain in children and adolescents.[22]

The recommended dose in children is 15 mg/kg orally, every 4 hours. Maximum daily dose is 90 mg/kg up to a maximum of 4 g (60 mg/kg/day maximum for infants aged less than 6 months). A dose of 10 mg/kg is no more effective than placebo for minor pain in children. General practitioners are reminded to use the correct dose for the child's weight.[23,24]

It is the consensus of the RACGP Working Group and Australian paediatric rheumatology experts that paracetamol be used for treating persistent pain in children and adolescents with JIA. Paediatric rheumatology experts recommend paracetamol should be used in the short term and it is rarely needed in the long term in patients with JIA.

Weak opioids (eg. codeine)

Recommendation 10 – Weak opioids (Grade D)
General practitioners could consider prescribing codeine in regular divided doses in addition to paracetamol for treating moderate articular pain in children and adolescents with JIA.

Good practice points

- Use the correct dose for the child's weight. The oral dose for codeine in children is 0.5 to 1.0 mg/kg every 4–6 hours up to a maximum of 3 mg/kg/day.
- Chronic dosing should not exceed 48 hours without medical advice.
- In overweight children, the ideal body weight, rather than actual weight, should be used to calculate dosage.

Cautions

- Adverse effects of codeine may occur in the absence of analgesia in poor metabolisers.
- Rapid metabolisers may experience excessive sedation.
- Be alert to paracetamol and codeine doses in combination preparations (eg. Painstop mixtures and Panadeine tablets).

There are no SRs or RCTs of codeine for treating persistent pain in children and adolescents with JIA published between January 2000 and January 2007.

Codeine is the weak opioid of choice for treating persistent pain in children and adolescents. The oral dose for codeine in children is 0.5 to 1.0 mg/kg every 4–6 hours up to a maximum of 3 mg/kg/day.[22]

The cytochrome p450 enzyme responsible (CYP2D6) shows genetic polymorphism and age dependent activity. The implications are that codeine is likely to be ineffective in poor metabolisers (9% English, 1% Swedish, German and mainland Chinese, 30% Ethiopian and Hong Kong Chinese). Adverse effects of codeine may occur in the absence of analgesia in poor metabolisers.[23]

It is the consensus of the RACGP Working Group and Australian paediatric rheumatology experts that codeine be used for treating moderate articular pain in children and adolescents with JIA in regular divided doses, in addition to paracetamol.

Traditional non-steroidal anti-inflammatory drugs (NSAIDs)

Recommendation 11 – traditional NSAIDs (Grade B)
General practitioners should prescribe NSAIDs as the initial drug of choice for reducing inflammation and associated pain in the treatment of JIA.

Good practice points

- Prescribe only one NSAID or COX-2 inhibitor at a time.
- Long term use of NSAIDs or COX-2 inhibitors should be at the lowest effective dose.
- NSAIDs may be prescribed with methotrexate.
- Use NSAIDs in the liquid form for children unable to swallow tablets.
- Base the selection of NSAID on dosing requirements, availability, and patient preferences (eg. taste).
- Consider stopping NSAIDs and COX-2 inhibitors 7–10 days before any major surgical procedure, particularly orthopaedic surgery. Discuss with the surgeon and make a decision on a case-by-case basis.

Cautions

- NSAIDs are generally well tolerated by children, but toxicity can occur. Caution should be applied in view of the known side effects, although these tend to be less common or severe than in adults: increased sleep disturbance and non-specific abdominal pain. A pseudoporphyria-like skin reaction occurs in about 5% of children taking naproxen. This latter complication is more common in fairer skinned children living in sun exposed latitudes. The antiplatelet effect of the NSAIDs predisposes to excessive bruising in particularly active children.[25]
- Most children with asthma can take NSAIDs safely. However, those with diagnosed or suspected aspirin induced asthma – symptoms of asthma usually accompanied by facial flushing and rhinitis within 3 hours of exposure to an NSAID – should avoid all NSAIDs.[25]
- Aspirin is not recommended in children because of the link with Reye syndrome.[23]

NSAIDs are the first line drug for the treatment of inflammation in children with JIA. They have well established analgesic and anti-inflammatory effects; however, they do not influence progression of the disease and do not prevent joint damage. Unlike adults, children tolerate NSAIDs very well, with few side effects.[23,26]

Table 3. Recommended paediatric doses for oral NSAIDs for juvenile idiopathic arthritis[22,26]

Drug *(Not in order of preference)*	Dose
Celecoxib	2–4 mg/kg twice daily
Diclofenac	1 mg/kg twice daily
Ibuprofen	10 mg/kg 3–4 times daily
Indomethacin	0.5–1.0 mg/kg 2–3 times daily
Meloxicam	0.15–0.30 mg/kg once daily
Naproxen	5–7.5 mg/kg twice daily
Piroxicam	0.2–0.4 mg/kg once daily

NSAIDs are valuable medications when used appropriately in carefully selected patients. Side effects are well recognised and include gastrointestinal disturbance, rash, mood changes, and sleep disturbance. No significant differences have been reported in the safety profiles of various NSAIDs.[22,26]

In both a good quality RCT[27] and a low quality SR of 14 low quality studies,[18] participants receiving all forms and doses of NSAIDs achieved significant improvements in outcome measures, and no individual NSAID was shown to have a clear advantage over others.

COX-2 inhibitors have not been studied extensively in children and potential adverse effects are not clear.

It is the consensus of the RACGP Working Group and Australian paediatric rheumatology experts that NSAIDs are the initial drug of choice for reducing inflammation and associated pain in the treatment of JIA.

Topical NSAIDs

Recommendation 12 – Topical NSAIDs (Grade D)
General practitioners should NOT prescribe topical NSAIDs to treat patients with JIA.

There are no studies of topical NSAIDs and JIA (2000–2007). Evidence from two literature reviews states there is no evidence available on the use of topical NSAIDs for treating persistent pain in children and adolescents with JIA.[2,20]

It is the consensus of the RACGP Working Group and Australian paediatric rheumatology experts that using topical NSAIDs in children and adolescents with JIA is NOT recommended.

Disease modifying anti-rheumatic drugs (DMARDs)

In caring for children with arthritis, commencing and altering DMARD medication is seen as the role of the specialist paediatric rheumatologist. Therefore, evidence on these drugs is not presented in this guideline.

General practitioners have an important role to play in monitoring adherence to DMARDs and their side effects. For example, common side effects of methotrexate treatment include nausea, anticipatory nausea, mouth ulcers, and abdominal discomfort; less commonly, altered liver function (increased transaminases), infection, or haematologic toxicity.[22,26]

Other commonly used DMARDs include hydroxychloroquine, sulfasalazine, leflunomide and biological DMARDs (bDMARDs).

Complementary/alternative medicines

Recommendation 13 – complementary/alternative medicines (Grade D)
General practitioners could inform patients and their families, that although there has been no research in children with JIA, there is limited or no evidence of effectiveness above placebo of complementary/alternative medicines in adult populations with arthritis.

Good practice points

- GPs should ask their patients about use of complementary medicines when prescribing treatment for JIA.
- GPs may consult a pharmacist if concerns arise regarding medication interactions related to complementary medicines.

There are no RCTs or SRs (2000–2007) on the use of complementary/alternative medicines including herbal and mineral supplements in children with JIA, nor is there commentary in relevant literature reviews.

Families/patients often seek complementary medicines for treatment of arthritis, particularly if they have had insufficient results from conventional medication. Alternative therapies used for the treatment of arthritis include herbs, vitamins and/or mineral supplements, aromatherapy, naturopathic and homeopathic products. These products are widely available without prescription in Australia.[28]

A number of SRs on the use of a wide range of complementary medicines in the treatment of arthritis in adult populations have not demonstrated any clinical benefits above placebo.[29–31] Until further research is conducted, these findings are likely to apply to paediatric populations with arthritis.

Although generally considered to have low risk of serious side effects, herbal and nutritional supplements may have harmful effects, particularly through interaction with other medications.[28]

It is the consensus of the RACGP Working Group and Australian paediatric rheumatology experts that GPs ask about complementary/alternative medicines when conducting medication reviews for children and adolescents with JIA. General practitioners may inform patients and their families that, although there has been no research in children, there is limited or no evidence of effectiveness above placebo of complementary medicines in adult populations with arthritis.

Non-pharmacological interventions

Nutritional therapies – calcium supplementation

Recommendation 14 – nutritional therapy – calcium (Grade B)

General practitioners should monitor calcium intake in children with JIA, and provide advice on increasing daily calcium intake. General practitioners could consider treating some patients with JIA with oral calcium and vitamin D supplementation.

Good practice points

- Encourage appropriate caloric and calcium intake.
- Children with JIA on corticosteroid therapy are at increased risk of osteoporosis and osteopenia.[22,25] Additional consideration should be given to calcium and vitamin D supplements when on corticosteroid course.

Children with JIA have been reported to have low BMD early in the disease and independent of steroid use. Additionally, 15–26% of JIA patients have pathologic fractures before adulthood.[32]

There is evidence from one good quality RCT that calcium supplementation can improve total BMD in patients with JIA. Over 2 years, patients taking calcium supplements had net improvement in total body bone mass density of 1% above those taking placebo. Some patients experienced nausea as a side effect.[33]

Evidence from a moderate quality RCT investigated achievement of daily calcium intake without dietary supplement in children with JIA. Increase in calcium intake translated into statistically superior increases at 6 and 12 months in some measures of BMD compared to those who received standard care. In the RCT, families with younger children with JIA were more prepared to participate in the interventions. In this RCT, a behavioral intervention had a positive impact on increasing dietary calcium intake. The improvement in calcium intake was achieved without compromising dietary intake in other areas.[21,32]

In addition to risks associated with disease, treatment with corticosteroids results in bone loss in a range of ways and increases the risk of osteoporosis. Steroids decrease absorption of calcium and increase urinary calcium loss, leading to bone resorption. Preventive therapy with calcium and vitamin D supplementation has been suggested for all patients taking corticosteroids.[34]

There are no SRs or RCTs conducted in children on the use of vitamin D in the protection against osteoporosis (2000–2007). In a Cochrane review of five studies that investigated the role of vitamin D supplements in adults receiving corticosteroid therapy, the meta-analysis showed that treatment with calcium and vitamin D is more effective at preventing bone loss than placebo or calcium alone.[34]

It is the consensus of the RACGP Working Group and Australian paediatric rheumatology experts that GPs monitor calcium intake in children with JIA and provide advice to families on increasing daily calcium intake. In some patients, particularly those on corticosteroids, consider calcium and vitamin D supplementation.

Land based exercises

Recommendation 15 – land based exercises (Grade C)

General practitioners should encourage patients with JIA to engage in regular physical activity compatible with their general abilities and restrictions of their disease.

Good practice points

- GPs may utilise EPC items to facilitate access to appropriate services (see *Resources*). Eligible services include, but are not limited to, those provided by physiotherapists, occupational therapists, and exercise physiologists.
- GPs could refer patients to Arthritis Australia for information and services relating to exercise (see *Resources*).
- Promote physical activity to decrease the risk of osteoporosis and osteopenia.
- Specific and individualised exercise therapy may be initiated with input from a paediatric trained physiotherapist or occupational therapist (or other appropriately trained therapists).
- The choice of exercise may depend on the child's specific needs and preferences.

One moderate quality RCT (80 children) evaluated the effects of exercise training on physical function in children with JIA. Participation in a 12 week exercise program of both low and high intensity resulted in improved physical function as measured by the self reported CHAQ, but not other self reported measures. There was no differences achieved between low or high intensity exercise, but those in low intensity programs were more likely to adhere to the regimen.[35]

A literature review summarised the findings from four low quality, small studies on land based exercise. It suggested that participation in moderate physical activity for at least 6 weeks (1–3 exercise sessions/ week) can improve both muscle function and aerobic fitness in children with JIA. Additional points from the reviewed literature suggest that:[36]

- children with JIA can participate in exercise without disease exacerbation
- participation in land based exercise at least twice a week for at least 6 weeks may help reduce disease symptoms and improve endurance
- land based exercise may lead to greater improvements in muscle strength, performance on timed tasks, and functional status than aquatic exercise
- weight bearing exercise is needed to develop optimal bone width and density during childhood
- individualised and supervised resistance exercise appears to be safe for children with JIA
- the choice of exercise may depend on the child's specific needs and preferences
- with proper screening, children with mild disease should be able to participate in most sports. However, highly competitive contact sports pose a potential risk for damage to the joint surface and growth plate, and should be avoided during periods of active joint disease.

At least 1 yearly review by a specialised paediatric physiotherapist is recommended in the Australian Paediatric Rheumatology standards of care.[3]

It is the consensus of the RACGP Working Group and Australian paediatric rheumatology experts that regular physical activity compatible with the child's general abilities and the restrictions of their disease is recommended. Regular physical activity promotes normal childhood development and may combat the adverse effects of disease on muscle strength, endurance and aerobic capacity.

Aquatic exercise

Recommendation 16 – aquatic exercise (Grade C)
General practitioners could inform patients about aquatic exercise for children and adolescents, and its limited effects.

Good practice points

- Not all children will have access to hydrotherapy.
- Aquatic exercises can be performed in a standard swimming pool.

There is evidence from one low quality RCT (54 patients with JIA) that aquatic exercise is safe, although it has no significant impact on functional outcome measures. No statistically significant differences were found on any outcome measures between the control group (standard care) and the intervention group (aquatic training program consisted of aerobic exercises, and flexibility and intensity training in a heated pool in a group setting by a physical therapist conducted for 1 hour per week) after 20 weeks of therapy. Participants had water confidence before commencing the program.[37]

One literature review suggested that participation in an exercise program at least twice per week for at least 6 weeks may help to reduce disease symptoms and improve general exercise endurance.[36]

At least 1 yearly review by a specialised paediatric physiotherapist is recommended in the Australian Paediatric Rheumatology standards of care.[3]

It is the consensus of the RACGP Working Group and Australian paediatric rheumatology experts that hydrotherapy on an individualised basis is recommended for some patients with JIA, and this is considered a safe form of exercise.

Orthotics management (splints ready made and custom made)

Recommendation 17 – splints (Grade D)

General practitioners could inform patients about use of splints and make individualised recommendations in conjunction with appropriately trained multidisciplinary health professionals.

Good practice points

- GPs should refer patients with JIA experiencing limitations in function for skilled occupational therapy advice.
- Splints should be fitted by an experienced therapist including, but not limited to, a physiotherapist, occupational therapist, orthotist or hand therapist.
- Consideration of the cost in the context of growing children is important, particularly in the face of limited high grade evidence.

A range of splints may be used in children with JIA, including resting and functional splints. These aim to maintain good joint position, support inflamed joints, stretch contracted joints, relieve pain and maintain function.[19] Splints may also be considered as an adjunct to pharmacological management to prevent contractures and increase the range of movement.[38]

There is conflicting evidence[39] from low quality studies to suggest that ready made splints and custom made splints may result in an improvement of dexterity. To date, no RCTs have been undertaken for splinting in JIA, and although splinting may have some effect, it appears to be highly dependent on the age of the child, the type of orthosis used and the location of the affected joint.[39]

It is the consensus of the RACGP Working Group and Australian paediatric rheumatology experts that splints be recommended on an individualised basis, in conjunction with specialised, paediatric trained multidisciplinary health professionals.

Orthotics management (foot orthotics)

Recommendation 18 – foot orthoses (Grade D)

General practitioners could inform patients with JIA in the lower limb about the role of comfortable, supportive shoes. General practitioners could inform patients about the use of foot orthotics based on an individualised assessment, safety and personal preference, in conjunction with appropriately trained multidisciplinary health professionals.

Good practice points

- Most children who have JIA of the lower limb only need comfortable, supportive shoes.
- Consideration of the cost in the context of growing children is important, particularly in the face of limited high grade evidence.
- Refer patients to allied health clinicians with specialised paediatric experience, preferably in managing children with JIA.

One low quality, non-blinded RCT (47 children with JIA) investigated the efficacy of custom made foot orthotics in improving pain and functional status. All children in the study received supportive comfortable shoes and those randomised to intervention groups also wore either custom made or ready made shoe inserts for 3 months. Participants in the custom orthotics group showed small but significantly greater improvements in overall pain (p=0.009), speed of ambulation (p=0.013), activity limitations (p=0.002), foot pain (p=0.019), and level of disability (p=0.024) on non-validated tools when compared with the two other groups (ready made shoe inserts and supportive athletic shoes alone). Children in all groups showed improvements in outcomes measures.[40]

It is the consensus of the RACGP Working Group and Australian paediatric rheumatology experts that use of foot orthotics be considered on an individualised basis, in conjunction with specialised, paediatric trained multidisciplinary health professionals.

Thermotherapy

Recommendation 19 – thermotherapy (Grade D)
General practitioners could consider recommending the use of heat or cold packs, warm baths and/or ice massage for symptomatic relief in children and adolescents with JIA.

Good practice points

- A warm bath or shower in the morning may reduce stiffness and pain.
- Use a larger piece of ice for massage (eg. freeze water in a paper or styrofoam cup, then cut around the top of the cup to expose the ice surface). Massage the ice gently in a circular motion. Limit ice massage to about 5 minutes at a time to avoid ice burn.
- When using thermotherapy, be alert to the patient's comfort and ability to tolerate.

There are no SRs or RCTs of thermotherapy for treating children and adolescents with JIA published between 2000 and 2007.

In a literature review of physical therapy and rehabilitation for JIA, heat treatments including heat packs, deep heat ultrasound and warm baths are suggested for decreasing joint rigidity and increasing joint flexibility, decreasing pain and decreasing muscle spasms. The authors suggested that effectiveness of heat therapy depends on the temperature, duration, rate of temperature change and the area being treated, but did not provide evidence or specify specific guidelines for each type of heat therapy. Massage, which is often used in conjunction with heat therapy, may be used to relieve pain, decrease anxiety, promote relaxation and prevent adhesions in subcutaneous tissues, although there is no evidence that it produces these outcomes in children with JIA. Potential adverse events include burns.[41]

In the same literature review, cold treatment in the form of cold packs is suggested to relieve pain and vasoconstriction in inflamed joints, although there is no evidence that it produces these outcomes in children with JIA. Potential adverse events include cold urticaria, cryoglobulinaemia, Raynaud phenomenon, and protest from the child.[41]

It is the consensus of the RACGP Working Group and Australian paediatric rheumatology experts that heat (warm/hot packs, warm baths) and/or cold (ice massage/cold packs) be recommended for symptomatic relief in children and adolescents with JIA.

Complementary/alternative physical therapies

Recommendation 20 – complementary/alternative physical therapies (Grade D)
General practitioners could inform patients and their families who seek advice that there is no research on complementary or alternative physical therapies in children with JIA.

Good practice points

- Complementary or alternative physical therapies should not replace pharmaceutical therapies and exercise in the management of JIA.
- GPs could refer patients who request information or advice on complementary or alternative physical therapies, including any risks, to Arthritis Australia (see *Resources*).

There are no SRs or RCTs of complementary and alternative physical therapies for treating children and adolescents with JIA published between 2000 and 2007, nor is there any commentary in any relevant literature reviews.

In adult populations with arthritis, research on complementary and alternative physical therapies including acupuncture, laser therapy, TENS and ultrasound has shown widely varying results depending upon the type of therapy, population of patients (eg. type/location of arthritis, duration of disease) and various therapy regimens (eg. length of therapy sessions, frequency of therapy). It is unclear whether these therapies provide any benefit to children with JIA.[42–45]

It is the consensus of the RACGP Working Group and Australian paediatric rheumatology experts that GPs should inform patients and their families who seek advice that there is no research on complementary or alternative physical therapies in children with JIA.

Disease monitoring and comorbidities

Recommendation 21 – Disease monitoring (Grade C)
General practitioners should be involved in monitoring disease progression and managing comorbidities in conjunction with the treating paediatric rheumatologist.

Good practice points

- Paediatric rheumatology review should take place at least twice a year.
- Regular screening for uveitis is recommended (*Table 4*).
- Arthritis activity should be assessed at least three times per year and treatment should be adjusted to keep the swollen and tender joint count as low as possible.
- Patients need to be monitored for potential toxicity and side effects of medications.
- Frequency and type of monitoring will depend on the DMARD prescribed, but most require FBC (to monitor for marrow suppression) and LFTs (to look for raised transaminases as a sign of hepatotoxicity) every 1–3 months (once on a stable dose). Adverse effects are less common than in adults with rheumatoid arthritis on DMARDs.

Juvenile idiopathic arthritis patients should be seen sufficiently frequently to monitor and appropriately adjust medications and therapy, and assess overall wellbeing. Depending on severity of disease and medications prescribed, visits may vary from every 2 weeks to every 3 months.[15]

General literature reviews suggest specific monitoring regimens. For example, tests to monitor complete blood cell counts, liver enzymes, and renal function are recommended for those using methotrexate, although the optimal frequency of testing is unclear.[18] Patients taking NSAIDs for more than 3–4 weeks should have monitoring laboratory tests (FBC, UE&C and LFTs) even in the absence of clinically apparent adverse effects.[15]

Uveitis occurs in approximately 20% of patients with oligoarthritis; 5–10% of those with polyarthritis and rarely in systemic arthritis. Patients who are ANA positive (particularly females) have a higher likelihood of developing uveitis. Due to the high rate of uveitis in JIA, patients should be screened regularly to prevent complications from undetected disease. Frequency of screening is determined by the risk of developing uveitis for the particular patient.[15,38]

It is the consensus of the RACGP Working Group and Australian paediatric rheumatology experts that disease monitoring and careful consideration of comorbidities by GPs is important in the management of JIA.

Table 4. Suggested timeframe for ophthalmology referral[38,46]

	Disease onset at <7 years of age	Disease onset at 7+ years of age
Pupils not round or precipitate on cornea	Immediate referral to rule out iritis	
Oligoarthritis +ve ANA	3–4 monthly for 4 years then every 6 months for 3 years then yearly	6 monthly for 4 years then yearly
Oligoarthritis –ve ANA	6 monthly for 7 years then yearly	
Polyarthritis +ve ANA	3–4 monthly for 4 years then every 6 months for 3 years then yearly	
Polyarthritis –ve ANA	6 monthly for 7 years then yearly	
Systemic onset	Yearly	

FURTHER INFORMATION

Full details of the evidence on which the guideline is based are presented in the companion documents *Recommendations for the management of juvenile idiopathic arthritis* (www.racgp.org.au/guidelines/juvenileidiopathicarthritis/recommendations) and *Juvenile idiopathic arthritis: a literature review of recent evidence* (www.racgp.org.au/guidelines/juvenileidiopathicarthritis/literaturereview).

The Process Report (*Appendix A*) outlines the full method used in the development of these recommendations.

Appendix B contains additional resources, as well as contact details for organisations providing services and support to people with JIA, and their carers.

The RACGP Working Group strongly recommends consulting the National Prescribing Service (www.nps.org.au), the Rheumatology Therapeutic Guidelines (www.tg.com.au), the Paediatric Handbook (www.rch.org.au/paed_handbook) and the Australian Medicines Handbook (amh.hcn.net.au) for detailed prescribing information including:

- ***indications***
- ***drug dosage***
- ***method and route of administration***
- ***contraindications***
- ***supervision and monitoring***
- ***product characteristics.***

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APPENDIX A. PROCESS REPORT

This report outlines the process used for the development of the evidence based *Clinical guideline for the diagnosis and management of juvenile idiopathic arthritis*.

The project consisted of the following major phases:

- formation of a multidisciplinary expert Working Group and Australia Paediatric Expert Group (see *Appendix C*)
- development of a scoping document outlining the scope and objectives of the project, including the process to be used in the guideline development
- identification and appraisal of relevant existing clinical guidelines, leading to the selection of an existing guideline for use as a primary reference
- systematic literature searches to identify more recent evidence
- synthesis of new evidence and evidence from the primary reference guideline into graded clinical recommendations and algorithms
- peer review and appraisal through a public consultation process
- response to feedback and completion of final guideline.

Figure 1 provides an overview of the primary phases in guideline development.

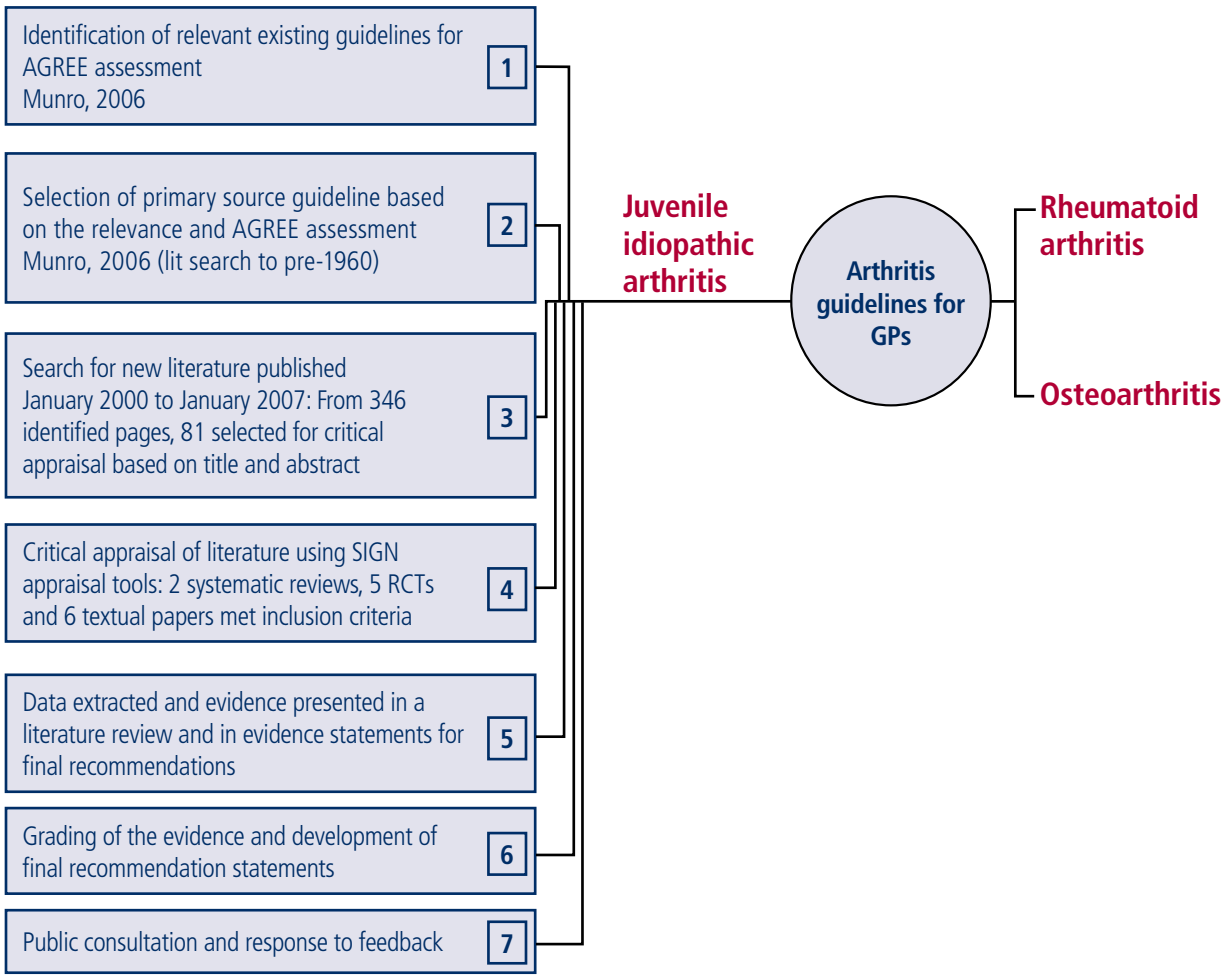

Figure 1. Process of guideline development

Identification of the guideline focus

A process model developed by the RACGP Steering Committee was used to identify the primary focus of the guideline (see the guideline background). The Working Group reached consensus opinion on the primary focus of the Guideline through discussion of areas considered most important for the primary audience (Australian GPs), with consideration to the feasibility of completing the guideline within the prescribed timeframe and budget. Clinical questions relevant to the area of guideline focus were developed to focus the search for relevant literature.

Identification, appraisal and selection of existing clinical guidelines

As part of the overall project to develop guidelines for arthritis management, the RACGP Steering Committee determined that the most feasible methodology would be to use an appropriate existing guideline as a primary reference and conduct a literature search to identify newly available evidence.

Existing guidelines were identified through database searches for the years 2005 to 2006 and those known to the Working Group. Guidelines considered to be the most relevant were selected for appraisal using the AGREE instrument.[1] Developers of the AGREE tool propose its use to assess '…the confidence that the potential biases of guideline development have been addressed adequately and that the recommendations are both internally and externally valid, and are feasible for practice'.[1] The AGREE tool includes 21 questions organised into six quality domains: scope and purpose; stakeholder involvement; rigour of development; clarity and presentation; applicability; and editorial independence. Each question is scored on a 4 point Likert scale (strongly agree, agree, disagree and strongly disagree) and the scores from multiple reviewers are used to calculate an overall quality percentage for each domain.

The Working Group identified only one relevant existing guideline, highlighting the paucity of evidence based recommendations in this field. This guideline was assessed by two reviewers using the AGREE tool:

- Munro J. Juvenile idiopathic arthritis management guidelines (Provisional). Australian Paediatric Rheumatology Group, 2006.

The AGREE assessment results for the guideline are outlined in *Table 1*. The guideline was selected for use as the primary source of information as it presented a comprehensive review of pharmacological and non-pharmacological management of JIA within the Australian health care context.

The Chair, Dr Munro acknowledged her potential conflict of interest as Project Director for development of *Juvenile idiopathic arthritis management guidelines (Provisional)*[2] and was not involved in the assessment of existing guidelines using the AGREE instrument, nor in the decision to use *Juvenile idiopathic arthritis management guidelines (Provisional)*[2] as the primary reference guideline.

Table 1. AGREE scores for identified guidelines

Guideline	AGREE domain scores					
	Domain 1. Scope and purpose	Domain 2. Stakeholder involvement	Domain 3. Rigour of development	Domain 4. Clarity and presentation	Domain 5. Applicability	Domain 6. Editorial independence
Munro J, 2006	72%	28%	24%	67%	11%	25%

Identification, appraisal and synthesis of new evidence

A search was conducted for new evidence to support that presented in *Juvenile idiopathic arthritis management guidelines (Provisional)*.[2] The process used for the literature search is reported in more detail in *Juvenile idiopathic arthritis: a literature review of recent evidence* (www.racgp.org.au/guidelines/juvenileidiopathicarthritis/literaturereview).

Search strategy

The MEDLINE, EMBASE and CINAHL databases and the Cochrane Library including CENTRAL Cochrane Controlled Trial Register were initially searched for evidence published between January 2000 and January 2007. Articles identified via personal contact with authors were also considered for inclusion. The following initial search strategies applied to the MEDLINE database and were adapted to apply to the other databases.

Search for evidence on diagnosis

- 'juvenile spondyloarthropathy'[Title/Abstract]) OR ('seronegative enthesopathy'[Title/Abstract]) OR ('juvenile chronic arthritis'[Title/Abstract])) AND ('Diagnosis'[MeSH])
- ('Spondylitis, Ankylosing/diagnosis'[MeSH]) OR ('Arthritis, Psoriatic/diagnosis'[MeSH]) OR ('Arthritis, Juvenile Rheumatoid/diagnosis'[MeSH]) OR ('spondylarthropathies/diagnosis'[MeSH])
- Limited to: Clinical Trial, Meta-Analysis, Practice Guideline, Review, Controlled Clinical Trial, Systematic Reviews.

Search for evidence on management

- ('juvenile spondyloarthropathy'[Title/Abstract]) OR ('seronegative enthesopathy'[Title/Abstract]) OR ('juvenile chronic arthritis'[Title/Abstract]) AND ('Diet Therapy'[MeSH]) OR ('Nursing'[MeSH]) OR ('Rehabilitation'[MeSH]) OR ('Surgery'[MeSH]) OR ('Therapeutics'[MeSH]) OR ('diet therapy' [Subheading]) OR ('nursing'[Subheading]) OR ('rehabilitation'[Subheading]) OR ('surgery'[Subheading]) OR ('therapy'[Subheading) OR ('Spondylitis, Ankylosing/dt,nu,rh,su,th'[MeSH]) OR ('Arthritis, Psoriatic/dt,nu,rh,su,th'[MeSH]) OR ('Arthritis, Juvenile Rheumatoid/dt,nu,rh,su,th'[MeSH]) OR ('spondylarthropathies/dt,nu,rh,su,th'[MeSH])
- Limited to: Clinical Trial, Meta-Analysis, Practice Guideline, Randomized Controlled Trial, Review, Controlled Clinical Trial, Systematic Reviews.

Inclusion/exclusion criteria

Types of studies

For evidence related to the diagnosis of JIA, initially only studies considered to be of NHMRC Level 1 or Level 2 evidence (*Table 2*) that evaluated diagnostic strategies for JIA were considered for inclusion. Due to the paucity of evidence available, the search was expanded to include lower levels of evidence such as diagnostic case control studies and literature reviews. Studies were reported in SRs already selected for inclusion were not subjected to individual critical appraisal to prevent replication of data.

For evidence related to the management of JIA, initially only papers considered to be of NHMRC Level 1 or Level 2 evidence (*Table 2*) that evaluated the effectiveness and/or safety of interventions for JIA in children aged 16 years or under were considered for inclusion. Due to the paucity of evidence available, the search was expanded to include lower levels of evidence such as comparative studies, case control studies, time series, case series and literature reviews. Randomised controlled trials that were reported in SRs already selected for inclusion in this literature review were not subjected to individual critical appraisal to prevent replication of data.

Types of participants

Studies that included children aged 16 years or under presenting with arthritic symptoms, as well as those diagnosed as having JIA were considered for inclusion.

Types of interventions

Interventions included any therapies used to manage JIA. Both pharmacological and non-pharmacological interventions were eligible for inclusion.

Table 2. NHMRC levels of evidence for intervention studies[3]

Level of evidence	Intervention studies	Diagnosis
I	Evidence obtained from a systematic review of all relevant randomised controlled trials	A systematic review of level II studies
II	Evidence obtained from at least one properly designed randomised controlled trial	A study of test accuracy with independent blinded comparison with a valid reference standard, among consecutive patients with a defined clinical presentation
III–I	Evidence obtained from well designed pseudo-randomised controlled trials (alternate allocation or some other method)	A study of test accuracy with independent blinded comparison with a valid reference standard, among non-consecutive patients with a defined clinical presentation
III–2	Evidence obtained from comparative studies with concurrent controls and allocation not randomised (cohort studies), case control studies, or interrupted time series with a control group	A comparison with reference standard that does not meet the criteria for Level II or Level III–1 evidence
III–3	Evidence obtained from comparative studies with historical control, two or more single arm studies, or interrupted time series without a parallel control group	Diagnostic case control evidence
IV	Evidence obtained from case series, either post-test or pre-test and post-test	Study of diagnostic yield (no reference standard)

Critical appraisal

Two reviewers critically appraised all studies that met the inclusion criteria. There was a high level of consensus between reviewers, with 100% agreement on Jadad scoring and minor discrepancies in SIGN scoring resolved by a third reviewer.

The following critical appraisal tools were used:

- SIGN appraisal tool for SRs (www.sign.ac.uk/guidelines/fulltext/50/checklist1.html – cited September 2006)
- SIGN appraisal tool for RCTs (www.sign.ac.uk/guidelines/fulltext/50/checklist2.html – cited September 2006)
- A checklist developed by the RACGP project team and approved by the NHMRC advisor for lower quality evidence.

Systematic reviews and RCTs were graded as being of good, moderate or low quality based on the results of appraisal using the SIGN tools. Literature reviews were those considered by the two independent reviewers to be of sufficient quality, given the lack of evidence in this field, with consideration given to the rigour of literature searching, selection of references, the author's background (where known) and peer review.

Data extraction

Two reviewers used the NHMRC RCT data extraction tool (www.nhmrc.gov.au); the Joanna Briggs Institute data extraction tool for SRs (available on request from JBI or NHMRC); or the checklist for lower quality evidence to extract data from the included studies in a systematic manner. Data from included studies was presented in a descriptive literature review, as well as a tabulated format. (Available in *Juvenile idiopathic arthritis: a literature review of recent evidence* (www.racgp.org.au/guidelines/juvenileidiopathicarthritis/literaturereview).

Special populations

The search strategy was designed to retrieve all available evidence meeting the inclusion criteria, including research specific to special populations identified by the NHMRC – Indigenous Australians (Aboriginal and Torres Strait Islanders), rural and remote communities, Muslim Australians, and Vietnamese Australians – (requirement of the contract). The literature searches identified minimal to no evidence directly related to these populations, thus a broader search was conducted to identify any research that addressed management of arthritis in the special population groups.

The following search was conducted in MEDLINE, CINAHL, EMBASE and Cochrane Library to identify relevant information:

1. Aboriginal.mp. OR Aborigine.mp. OR koori.mp. OR indigenous.mp. OR torres strait.mp. OR Vietnam/ OR Vietnamese.mp. OR rural health centers/ OR Hospitals, Rural/ OR Rural Health/ OR Rural Health Services/ OR Rural Areas/ OR Rural Health Nursing/ OR muslim.mp. OR Islam/
2. Arthritis/ OR Arthritis, Juvenile Rheumatoid/ OR Arthritis.mp
3. 2 and 3.

Ten papers were identified for retrieval – five papers related to Australian Aborigines, three related to rural health and two papers focused on Muslim populations. All 10 papers were excluded as they did not directly relate to JIA, or were historical health information.

Development and grading of the recommendations

Through group meetings, email circulation and feedback, the Working Group used the new evidence, together with evidence from the primary reference guideline and expert opinion to develop recommendations relevant to general practice within Australia. Throughout the process expert opinion was sought from the Australian Paediatric Experts Group (see *Appendix C*).

Evidence statements were developed that represented a summary of the most relevant evidence from the literature, or where there had been no newly published research, from *Juvenile idiopathic arthritis management guidelines (Provisional)*.[2] A body of evidence assessment matrix developed by the NHMRC[3] (*Table 3*) was used to assess the volume and consistency of evidence supporting each recommendation; as well as the clinical impact, generalisability and applicability of the recommendation.

Each recommendation was given a final grading (*Table 4*) representing its overall strength. The gradings reflect implementability in terms of confidence practitioners can use in a clinical situation. The overall grade of each recommendation was reached through consensus and is based on a summation of the grading of individual components of the body of evidence assessment. In reaching an overall grade, recommendations did not receive a grading of A or B unless the volume and consistency of evidence components were both graded either A or B.

Table 3. NHMRC body of evidence assessment matrix[3]

Component	A	B	C	D
	Excellent	Good	Satisfactory	Poor
Volume of evidence	At least one good quality SR that has at least one good quality RCT	At least one good quality RCT or a moderate quality SR that has at least two moderate to good quality RCTs	Moderate or low quality RCTs	General reviews published in a refereed journal, or expert committee reports or opinions (consensus) and/or clinical experience of respected authorities
Consistency	All studies consistent	Most studies consistent, and inconsistencies may be explained	Some inconsistency reflecting genuine uncertainty around clinical question	Evidence is inconsistent
Clinical impact	Very large	Substantial	Moderate	Slight or restricted
Generalis-ability	Population/s studied in the body of evidence are the same as the target population for the guideline	Population/s studied in the body of evidence are similar to the target population for the guideline	Population/s studied in the body of evidence different to the target population for the guideline but it is clinically sensible to apply this evidence to the target population	Population/s studied in the body of evidence different to the target population for the guideline and hard to judge whether it is sensible to generalise to the target population
Applicability	Directly applicable to Australian health care context	Applicable to Australian health care context with few caveats	Probably applicable to Australian health care context with some caveats	Not applicable to Australian health care context

Table 4. NHMRC grade of recommendations[3]

Grade	Description
A	Body of evidence can be trusted to guide practice
B	Body of evidence can be trusted to guide practice in most situations
C	Body of evidence provides some support for recommendation(s) but care should be taken in its application
D	Body of evidence is weak and recommendation must be applied with caution
Note: A recommendation cannot be graded A or B unless the volume and consistency of evidence components are both graded either A or B	

Consultation phase

Draft versions of the *Clinical guideline for the diagnosis and management of juvenile idiopathic arthritis, Recommendations for the diagnosis management juvenile idiopathic arthritis* and *Juvenile idiopathic arthritis: a literature review of recent evidence* were presented for public feedback via the RACGP website. An interactive survey was designed to collect comments from all potential stakeholders. The public consultation period was advertised in major national newspapers and over 200 known stakeholders (eg. members of RACGP musculoskeletal groups, consumer groups) were sent personal invitations to review the material. Feedback collected from the survey and independent submissions were collated and addressed by the Working Group.

The Working Group would like to thank respondents who provided feedback during the consultation phase of the project.

Dissemination

Final versions following consultation of the *Clinical guideline for the diagnosis and management of juvenile idiopathic arthritis, Recommendations for the diagnosis and management juvenile idiopathic arthritis* and *Juvenile idiopathic arthritis: a literature review of recent evidence*, together with supporting resources, will be made available to Australian GPs and the public on the RACGP website.

The RACGP has submitted to the Australian Department of Health and Aging (DoHA) a detailed dissemination plan based on the NHMRC standards. The dissemination process is based upon four lines of deliberate action:

- specified target groups
- the most appropriate media
- resources allocated for the design, production and distribution of materials
- the design, production and distribution process managed as a project, with appropriate evaluation and feedback.

Process report references

1. AGREE Collaboration. Appraisal of guidelines for research & evaluation (AGREE) instrument. 2001. Available at www.agreecollaboration.org. [Accessed November 2006].
2. Munro J. Juvenile idiopathic arthritis management guidelines (provisional). Australian Paediatric Rheumatology Group, 2006.
3. Coleman K, Norris S, Weston A, et al. NHMRC additional levels of evidence and grades for recommendations for developers of guidelines. Canberra: NHMRC, 2005.

APPENDIX B. RESOURCES

Useful publications

The National Health and Medical Research Council. Making decisions about tests and treatments: Principles for better communication between healthcare consumers and healthcare professionals. Canberra: NHMRC, 2005.

National Prescribing Service Limited. Indicators of quality prescribing in Australian general practice. Sydney: National Prescribing Service Limited (NPS), 2006.

The RACGP Working Group recommends consulting the National Prescribing Service (www.nps.org.au), the Rheumatology Therapeutic Guidelines (www.tg.com.au), the Paediatric Handbook (www.rch.org.au/paed_handbook) and the Australian Medicines Handbook (amh.hcn.net.au/) for detailed prescribing information.

Useful electronic sources

Note: website addresses were accurate at the time of publication.

Organisation	Website
Australian Medicines Handbook	www.amh.hcn.net.au
Australian Rheumatology Association (ARA)	www.rheumatology.org.au
ARA patient medication information sheets	www.rheumatology.org.au/community/PatientMedicineInformation.asp#medicine
Enhanced Primary Care (EPC)	www.health.gov.au/epc
National Health and Medical Research Council (NHMRC)	www.nhmrc.gov.au
National Prescribing Service (NPS)	www.nps.org.au
Paediatric Handbook	www.rch.org.au/paed_handbook
The Royal Australian College of General Practitioners (RACGP)	www.racgp.org.au
Therapeutic Guidelines	www.tg.com.au

Patient services

Note: Website addresses and contact details were accurate at the time of publication.

Organisation	Website	Telephone
Arthritis Australia	www.arthritisaustralia.com.au	1800 111 101 toll free
Australian Association of Occupational Therapists	www.ausot.com.au	+61 3 9415 2900 National office
Australian Hand Therapy Association	www.ahta.com.au	+61 8 9578 3348 National office
Australian Physiotherapy Association	www.physiotherapy.asn.au	+61 3 9092 0888 National office
Australian Rheumatology Association	www.rheumatology.org.au	+61 2 9256 5458 National office
Carers Australia	www.carersaustralia.com.au	1800 242 636 toll free Call put through to local branch

JIA history and clinical examination checklist

History

- Pain and/or swelling in one or more joints
- Check the nature of onset – is it acute or insidious? Previous episodes?
- Acute onset monoarticular arthritis associated with fever is septic until proven otherwise
- Check the timing of symptoms during the day – as a general guide:
  - early morning stiffness/stiffness after rest or sleep = inflammatory
  - post-activity pain = mechanical
- Check duration of illness – if >6 weeks it is less likely to be reactive/post-viral arthritis
- Are there any intercurrent infections (respiratory, enteric or skin)? Post-viral infections are probably the commonest cause of transient arthritis
- Constitutional features (eg. fever, rash, loss of weight)
- Has the child been taking any medications (eg. cefaclor)?
- What does the child, or parent, consider to be the most symptomatic site – is it in the joint, muscle, adjacent bone or a more diffuse area?
- Assess whether the normal activity levels or interests have been interrupted
- Check for extra-articular symptoms – ensure a thorough systems review and keep the following diagnoses in mind:
  - Septic arthritis
  - Post-infectious/reactive arthritis
  - Systemic lupus erythematosus
  - Acute lymphoblastic leukaemia
  - Trauma/non-accidental injury
  - Osteomyelitis
  - Bone tumour
  - Inflammatory bowel disease
  - Henoch-Schönlein purpura and other vasculitides
  - Rheumatic fever
  - Hypermobility
- Assess the functional milieu of the patient (eg. school progress and attendance, sleep pattern, family and peer relationships and stress experiences)
- Check family history for other types of inflammatory arthritis, particularly the spondyloarthropathies, autoimmune disorders and pain syndromes (eg. fibromyalgia or other models for pain behaviour)

General examination

Observe the patient as they move about and be opportunistic when examining them:

- Look for limitations or alterations in function in general movement
- Examine all joints, not only the site of the presenting complaint. There may be inflammation without symptoms in JIA
- Aim to localise the site of maximal discomfort (eg. is it the joint capsule, adjacent bone or muscle belly, tendon or ligament attachments?)
- Examine for signs of systemic diseases with an articular component, extra-articular features of JIA, or both. In particular examine the skin, eyes, abdomen, nails and lymph nodes

Musculoskeletal assessment

- Joints – signs of inflammation such as swelling or tenderness, the range of movement and deformity. Joints affected by JIA are typically swollen, may be tender to touch and warm but are usually not erythematous
- Entheses – bone attachment sites of ligaments/tendons (eg. Achilles tendon)
- Tendon sheaths of fingers and toes (eg. dactylitis in psoriasis)
- Patellar tracking pattern – does the patella move vertically on walking?
- Shoe sole and heel wearing pattern
- Leg length measurement
- Spinal flexion, including Schober test (the measurement of the lumbosacral range should increase in distance by at least 6 cm on maximal flexion with knees straight; the starting range is a point 5 cm below and 10 cm above the lumbosacral junction (use the Dimples of Venus as a guide for this)
- Assessment of growth parameters
- Muscle tenderness, muscle wasting or weakness (eg. inability to toe or crouch walk)
- Gait – antalgic (pain) or limp, Trendelenburg sign

Differential diagnosis

Juvenile idiopathic arthritis can resemble any disorder causing acute or chronic polyarthritis in children. Elimination of other diseases is therefore a necessary step in JIA diagnosis. Keep the following diagnoses in mind:

- Septic arthritis
- Post-infectious/reactive arthritis
- Systemic lupus erythematosus
- Acute lymphoblastic leukaemia
- Trauma/non-accidental injury
- Osteomyelitis
- Bone tumour
- Inflammatory bowel disease
- Henoch-Schönlein purpura and other vasculitides
- Rheumatic fever
- Hypermobility

APPENDIX C. MEMBERSHIP AND TERMS OF REFERENCE OF THE RACGP JIA WORKING GROUP

Aim of the RACGP Juvenile Idiopathic Arthritis Working Group

The aim of the Working Group was to undertake activities required to fulfill the aims of the project, as outlined in the funding agreement, including:

- carrying out a review of literature as per NHMRC requirements, and
- developing clinical practice guidelines based on the evidence obtained within the literature review.

Establishment of Working Group

In accordance with the project contract, membership of the Working Group endeavoured to include:

- three or more experts in each field – medical (including one GP) and allied health
- one expert NAMSCAG member
- one consumer representative
- one departmental representative
- a consultant appointed by the NHMRC.

In addition, the following groups were represented in accordance with the project contract:

- a nominee of the Australian Rheumatology Association or the Australian and New Zealand Bone and Mineral Society, and
- a nominee of the Endocrine Society of Australia and of the Faculty of Rehabilitation Medicine.

Australian Paediatric Rheumatology Expert Group

To ensure that this guideline received input from paediatric rheumatologists throughout Australia, an Expert Group was established to provide feedback on the evidence for recommendations and to contribute to the development of consensus recommendations. The Expert Group was consulted throughout the development of the guideline.

Membership of the RACGP Juvenile Idiopathic Arthritis Working Group

Member	Representation	Qualifications
Dr Jane Munro (Chair) Paediatric rheumatologist Royal Children's Hospital Melbourne, Victoria	Paediatric Rheumatology Association	MBBS, MPH, FRACP
Dr Shane Brun Associate Professor Musculoskeletal and Sports Medicine (Qld)	RACGP School of Medicine and Dentistry, James Cook University, Queensland	FFSEM(UK), FACRRM, FASMF, FARGP, MSpMed, DCH, GradDipRural.Med, GradDipEd, BAppSc, FRACGP
Dr Morton Rawlin Project Director	RACGP – Director of Educational Services	BMed, MMedSci, DipPracDerm, DipFP, DipMedHyp, DipBusAdmin, FACRRM, FRACGP
Pam Webster Consumer representative (NSW)	Consumer and NAMSCAG Carers Australia	MCH, BA, DipT, AUA
Prof Karen Grimmer-Somers	NHMRC Advisor	PhD, MMedSc, BPhty, LMusA, CertHlthEc
Amy Jasper Project Manager	RACGP – Manager, Education Evaluation	MBA, GDipHumServRes, BAppSci(AdvNsg)
Dr Jiri Rada	RACGP Project Officer	PhD, FRSH, MSc, BPHE, BA
Emily Haesler	RACGP Project Officer	BNurs, PGradDipAdvNsg

Membership of the Australian Paediatric Rheumatology Expert Group

Member	Location	Qualifications
Dr Roger Allen Paediatric rheumatologist	Department of General Medicine Royal Children's Hospital Melbourne, Vic	MBBS, FRACP
Dr JD Akikusa Paediatric rheumatologist, general paediatrician	Department of General Medicine Royal Children's Hospital Melbourne, Vic	MBBS, FRACP
Dr Christina Boros Senior Lecturer	University of Adelaide Discipline of Paediatrics Head, Rheumatology CYWHS Adelaide, SA	MBBS, PhD, FRACP
Dr Sue Piper Clinical Associate Professor	Head, Paediatric Rheumatology Unit, Monash Medical Centre Melbourne, Vic	MBBS, FRACP
Dr Navid Adib Paediatric rheumatologist	Brisbane, Qld	MBBS, PhD, FRACP
Dr Kevin Joseph Murray Consultant paediatric and adolescent rheumatologist	West Perth, WA	MBBS, FRACP
Dr Jeff Chaitow	Sydney, NSW Westmead Children's Hospital	MMBCh, FRACP

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