

# Recommendations for the diagnosis and management of juvenile idiopathic arthritis

## August 2009



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## INTRODUCTION

This supporting document provides a summary and grading of the evidence underpinning the recommendations outlined in the *Clinical guideline for diagnosis and management of juvenile idiopathic arthritis* ([www.racgp.org.au/guidelines/juvenileidiopathicarthritis](http://www.racgp.org.au/guidelines/juvenileidiopathicarthritis)) and is intended to be read in conjunction with the guideline. The process used to develop these recommendations is outlined in full in the Process Report (Appendix A). Further information on the evidence presented in this report is available in *Juvenile idiopathic arthritis: a literature review of recent evidence* ([www.racgp.org.au/guidelines/juvenileidiopathicarthritis/literaturereview](http://www.racgp.org.au/guidelines/juvenileidiopathicarthritis/literaturereview)) and in the guideline.

The recommendations are intended for children aged 16 years and under diagnosed with juvenile idiopathic arthritis (JIA) and those presenting with arthritic symptoms. The Royal Australian College of General Practitioners (RACGP) Juvenile Idiopathic Osteoarthritis Working Group supports all 21 recommendations and intends that they be used in conjunction with clinical judgement and patient preferences.

This project was supported by the 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 Osteoarthritis Working Group:

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***The RACGP Juvenile Idiopathic Osteoarthritis Working Group recommends consulting the National Prescribing Service ([www.nps.org.au](http://www.nps.org.au)), the Therapeutic Guidelines – Rheumatology ([www.tg.com.au](http://www.tg.com.au)), the Paediatric Handbook ([www.rch.org.au/paed\\_handbook](http://www.rch.org.au/paed_handbook)) and the Australian Medicines Handbook ([www.amh.hcn.net.au](http://www.amh.hcn.net.au)) for detailed prescribing information including:***

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

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

## Evidence sources

The evidence for the recommendations is based on:

- an Australian provisional guideline for JIA<sup>1</sup> which was assessed using the AGREE instrument<sup>2</sup> and identified as being an appropriate, high quality guideline to use as a primary reference
- a review of the literature identified through a systematic search for evidence published from January 2000 to January 2007
- the RACGP Working Group's expert opinion, and
- opinion from an Australian paediatric expert group.

### Primary reference guideline

The RACGP Working Group identified only one existing JIA guideline, which was assessed using the AGREE assessment tool.<sup>2</sup> This guideline, *Juvenile idiopathic arthritis management guidelines (Provisional)*,<sup>1</sup> was selected 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. AGREE scores for the selected guideline are presented in *Table 1*.

**Table 1. AGREE scores for the primary guideline reference<sup>1</sup>**

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 <sup>1</sup>	72%	28%	24%	67%	11%	25%

### Literature review

The method used to conduct the literature review is outlined in full in the Process Report (*Appendix A*) and in *Juvenile idiopathic arthritis: a literature review of recent evidence* ([www.racgp.org.au/guidelines/juvenileidiopathicarthritis/literaturereview](http://www.racgp.org.au/guidelines/juvenileidiopathicarthritis/literaturereview)).

The literature review extended the search conducted in the primary reference guideline, *Juvenile idiopathic arthritis management guidelines (Provisional)*.<sup>1</sup> A search was performed on Medline, EMBASE, CINAHL and the Cochrane Library for English language publications published between January 2000 and January 2007 that contained research on JIA. Papers were initially selected for inclusion based on their title and abstract. Systematic reviews (SRs) and randomised controlled trials (RCTs) that met the inclusion criteria were critically appraised using checklists developed by SIGN<sup>3</sup> and given an overall quality grade of high, moderate or low. Lower levels of evidence (eg. consensus guidelines, literature reviews) were assessed with a checklist designed for this project that considered the rigour of literature searching, selection of references, the author's background (where known) and peer review.

### Grading of the recommendations

Each recommendation has been graded (from A to D) according to the *National Health and Medical Research Council additional levels of evidence and grades for recommendations for developers of guidelines*<sup>4</sup> (*Table 2*). The grade reflects the degree of 'trust' that the clinician can place in the clinical application of the recommendation. Each recommendation is supported by an evidence statement.

**Table 2. Recommendation grades<sup>4</sup>**

<b>A</b>	Excellent evidence – body of evidence can be trusted to guide practice
<b>B</b>	Good evidence – body of evidence can be trusted to guide practice in most situations
<b>C</b>	Some evidence – body of evidence provides some support for recommendation(s) but care should be taken in its application
<b>D</b>	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 3* shows the body of evidence assessment matrix, listing all the components that were considered when assessing the evidence, together with the grades used.<sup>4</sup> 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 3. Body of evidence assessment matrix<sup>4</sup>**

<b>Component</b>	<b>A</b>	<b>B</b>	<b>C</b>	<b>D</b>
	<b>Excellent</b>	<b>Good</b>	<b>Satisfactory</b>	<b>Poor</b>
<b>Volume of evidence</b>	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
<b>Consistency</b>	All studies consistent	Most studies consistent and inconsistencies may be explained	Some inconsistency reflecting genuine uncertainty around clinical question	Evidence is inconsistent
<b>Clinical impact</b>	Very large	Substantial	Moderate	Slight or restricted
<b>Generalisability</b>	Directly generalisable to target population	Directly generalisable to target population with few caveats	Not directly generalisable to target population but could be sensibly applied	Not directly generalisable to target population and hard to judge whether it is sensible to apply
<b>Applicability</b>	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

## Limitations of the guideline

### Medication information

The literature search was not designed to retrieve safety trials for pharmacological interventions. The recommendations do not seek to provide full safety and usage information on pharmacological interventions. The pharmacological interventions outlined in the recommendations should not be applied without consideration given to the patient's clinical profile and personal preferences. The RACGP Working Group recommends consulting the National Prescribing Service ([www.nps.org.au](http://www.nps.org.au)), the *Therapeutic Guidelines – Rheumatology* ([www.tg.com.au](http://www.tg.com.au)), the *Paediatric Handbook* ([www.rch.org.au/paed\\_handbook](http://www.rch.org.au/paed_handbook)) and the *Australian Medicines Handbook* ([www.amh.hcn.net.au](http://www.amh.hcn.net.au)) for detailed prescribing information including:

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

### 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 RACGP 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 RACGP 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.

## Commonly used abbreviations

ACR	American College of Rheumatology paediatric 30/50/70 criteria
30/50/70	
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

## SUMMARY OF RECOMMENDATIONS

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:

- erythrocyte sedimentation rate (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. They should 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 non-steroidal anti-inflammatory drugs (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 the 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 the 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 symptomatic relief in children and adolescents with JIA.

**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.

**RECOMMENDATION 21 – DISEASE MONITORING (Grade C)**

General practitioners should be involved in monitoring disease progression and comorbidities in conjunction with the treating paediatric rheumatologist.

## FULL RECOMMENDATIONS

### Diagnosis of juvenile idiopathic arthritis

#### Early diagnosis and referral

##### EVIDENCE STATEMENT

In JIA, persistent synovitis leads to joint deformity and destruction and may occur within 2 years of the onset of disease. Disruption of proper joint function predisposes children and young adults to premature osteoarthritis and potential lifetime disability.<sup>5</sup>

One literature review reported that within the first 6 months of diagnosis, the type of JIA is determined by assessing the presentation, clinical signs and symptoms and disease course.<sup>6</sup> It is important to diagnose JIA and initiate disease modifying therapy as soon as possible.<sup>7</sup> An early, accurate diagnosis is essential in order to commence appropriate management aimed at promoting normal growth and development, and to minimise disability and deformity.<sup>5</sup>

The literature recommends early referral to a paediatric rheumatologist. With a large range of differential diagnoses, assessment by a paediatric rheumatologist at an early stage should be sought to confirm diagnosis.<sup>5</sup>

***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.***

	Component	Descriptor	Grade
Two consensus guidelines <sup>1,5</sup> and consensus between paediatric rheumatology experts and the RACGP Working Group	Volume of evidence	Poor	D
The consistency of evidence is good and there is consensus	Consistency	Good	B
Potentially significant impact on disease progression and long term outcome if JIA is diagnosed early in primary care and appropriate referrals made to enable early commencement of disease modifying antirheumatic drug (DMARD) therapy	Clinical impact	Excellent	A
Applicable to Australian health care context with few caveats. The studies did not include, or did not report, data specific to Aboriginal and Torres Strait Islander populations; these populations may be disadvantaged because of lack of access to weight loss interventions	Generalisability	Good	B
Applicable to the Australian health care context with few caveats	Applicability	Good	B

RECOMMENDATIONS	GRADE
General practitioners should aim to diagnose JIA as early as possible in order to optimise outcomes for patients.	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.	C
<b>Good practice points</b> <ul style="list-style-type: none"> <li>For advice about accessing a paediatric rheumatologist, either privately or through public clinics, contact the Australian Rheumatology Association at <a href="http://www.rheumatology.org.au">www.rheumatology.org.au</a></li> <li>When making a referral for a newly diagnosed or suspected case of JIA, make initial telephone contact with a rheumatologist and mark the referral <b>URGENT</b> (recent onset JIA).</li> </ul>	

## History and clinical examination

### EVIDENCE STATEMENT

Assessing a child presenting with signs and symptoms suggesting a diagnosis of arthritis requires taking a comprehensive patient history, conducting a complete physical examination, and ordering appropriate diagnostic tests.<sup>5</sup> Current literature agrees that a complete history should be taken, including a full description of symptoms. Common considerations include:<sup>5,7,8</sup>

- pain characteristics, eg. severity, onset, timing and duration of pain
- aggravating and alleviating factors
- early morning stiffness or presence of stiffness after resting
- presence of limp
- concurrent systemic illness, presence and duration of fever and/or rash
- preceding infections
- sleep history
- vaccination history
- any treatments that have been used and response to treatment
- impact on daily living and school.

*It is the consensus of the RACGP Working Group and Australian paediatric rheumatology experts that a 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; JIA should be particularly suspected in patients who present with persistent joint pain and swelling. In most patients, symptoms emerge over weeks to months.*

	Component	Descriptor	Grade
One Australian guideline <sup>1</sup> and consensus between paediatric rheumatology experts and the RACGP Working Group	Volume of evidence	Poor	D
The consistency of evidence is satisfactory and there is consensus	Consistency	Satisfactory	C
Potentially significant impact on long term outcome and adverse events if proper history taking and clinical examination support early diagnosis and treatment of JIA	Clinical impact	Excellent	A
Directly generalisable to the general Australian population with few caveats. There is no evidence	Generalisability	Good	B

specific to Aboriginal and Torres Strait Islander populations. These populations may be disadvantaged because of lack of access to services			
Applicable to the Australian health care context with few caveats	Applicability	Good	B

RECOMMENDATION	GRADE
<p>General practitioners should base a diagnosis of JIA (and differential diagnosis) on history and clinical examination in the first instance, with strong suspicion of JIA indicated by:</p> <ul style="list-style-type: none"> <li>• pain and swelling of single or multiple joints</li> <li>• persistent or worsening loss of function</li> <li>• fever of at least 10 days with unknown cause, often associated with transient erythematous rash</li> <li>• decreased ROM</li> <li>• joint warmth</li> <li>• effusion.</li> </ul>	C

## Diagnostic investigations

### EVIDENCE STATEMENT

There is no single test that accurately diagnoses JIA; however, 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.

**Erythrocyte sedimentation rate** and **CRP** indicate an inflammatory process but have low specificity for JIA. One or other of these tests is usually performed.<sup>5,7,8</sup> These markers are usually elevated in JIA but may be normal. They may be useful in monitoring disease activity and response to treatment (RACGP Working Group).

The **rheumatoid factor** (RhF) test is not conclusive. Rheumatoid factor is positive in only a small percentage of JIA patients (unlike in rheumatoid arthritis). However, when present in combination with other factors, the level of RhF may indicate likelihood for aggressive disease progression and poorer prognosis.<sup>5,7,8</sup>

A **full blood count** is usually undertaken to provide general information relating to inflammation, anaemia and raised white cell count.<sup>5,7,8</sup>

**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.<sup>7</sup> However, radiographs are useful to exclude trauma, malignancy or infection and to corroborate clinical findings.<sup>9</sup> Serial X-rays over years may show disease progression and therefore indicate need for change in treatment strategy. Other imaging is unlikely to be helpful at diagnosis and should be ordered in discussion with the paediatric rheumatologist.

The **antinuclear antigen** (ANA) test is positive in 40–50% of patients with polyarthritis, 70–85% of patients with oligoarthritis and 10% of patients with systemic JIA. Patients with enthesitis related JIA are usually ANA negative.<sup>5</sup>

**Human leukocyte antigen** (HLA) B27 testing may be helpful in the diagnosis of enthesitis related arthritis.<sup>7,9</sup>

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

	<b>Component</b>	<b>Descriptor</b>	<b>Grade</b>
One Australian guideline <sup>1</sup> and consensus among paediatric rheumatology experts and the RACGP Working Group	Volume of evidence	Poor	D
The consistency of evidence is satisfactory and there is consensus	Consistency	Satisfactory	C
Potentially large clinical impact with reduction in adverse effects if appropriate diagnostic investigations are used to diagnose JIA	Clinical impact	Excellent	A
Generalisable to target population with few caveats. There is no evidence specific to Aboriginal and Torres Strait Islander populations. These populations may be disadvantaged because of lack of access to services	Generalisability	Good	B
Applicable to the Australian health care context with few caveats	Applicability	Good	B

<b>RECOMMENDATION</b>	<b>GRADE</b>
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: <ul style="list-style-type: none"> <li>• ESR or CRP</li> <li>• FBC.</li> </ul> Consider RhF, ANA, HLA B27, and plain radiographs. Depending on the clinical picture, additional investigations may be required.	<b>C</b>
<b>Good practice points</b>	
<ul style="list-style-type: none"> <li>• Before JIA is diagnosed, all other known conditions and causes of childhood arthritis must be excluded</li> <li>• Absence of any key symptoms, signs, or test results does not necessarily rule out JIA.</li> </ul>	

# Management of juvenile idiopathic arthritis

## GENERAL PRINCIPLES

### Multidisciplinary care and care planning

#### EVIDENCE STATEMENT

There is strong support for a multidisciplinary approach in the management of JIA to ensure all of the needs of the child are met, including normal growth, social development and physical functioning.<sup>5,7,9,10</sup> 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
- paediatric rheumatology nurses (currently in Victoria and Western Australia only)
- 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 the 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 patient.<sup>10</sup> 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).<sup>5,7,9</sup>

***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.***

	Component	Descriptor	Grade
One Australian guideline <sup>1</sup> and consensus among paediatric rheumatology experts and the RACGP Working Group	Volume of evidence	Poor	D
The consistency of evidence is satisfactory and there is consensus	Consistency	Satisfactory	C
Satisfactory clinical impact if patients receive multidisciplinary input from appropriately trained health care providers according to individually assessed need	Clinical impact	Satisfactory	C
Generalisable to the target population with few caveats. However, multidisciplinary teams trained in JIA management are not widely available in Australia (particularly in rural and remote areas) and there is no evidence specific to Aboriginal and Torres Strait Islander populations. These populations may be disadvantaged because of lack of access to the interventions	Generalisability	Satisfactory	C
Applicable to the Australian health care context with few caveats	Applicability	Good	B

RECOMMENDATIONS	GRADE
General practitioners should encourage and support a management approach based on individual patient needs and involving a multidisciplinary team of health professionals.	<b>D</b>
Involvement of the multidisciplinary team should be managed through the development of individual care plans.	<b>D</b>
<b>Good practice points</b>	
<ul style="list-style-type: none"> <li>• GPs may utilise Enhanced Primary Care (EPC) items to facilitate access to appropriate services (<a href="http://www.health.gov.au/epc">www.health.gov.au/epc</a>). Eligible services include those provided by physiotherapists, osteopaths, occupational therapists, podiatrists, mental health workers, indigenous health workers, chiropractors, chiropodists, audiologists and speech pathologists</li> <li>• GPs should refer carers to their state/territory carers' association for information and support as part of the ongoing management plan (telephone 1800 242 636)</li> <li>• Include pain management and assessment of sleep quality in the treatment plan.</li> </ul>	

## Patient education and psychosocial support

### EVIDENCE STATEMENT

The evidence of the impact of patient education and psychosocial support remains limited, but most low level 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:

- patient/family education
- psychosocial interventions/support services
- community resources
- school based resources
- information and referral regarding insurance coverage and benefit co-ordination.<sup>5</sup>

Education, behavioural interventions and psychosocial support are important with specific interventions. In a moderate quality RCT investigating the role of dietary calcium in improving bone accretion in children with JIA, separate child and parent group education sessions focusing on a different meal in each session may have contributed to treatment success. Behavioural intervention may have a positive impact on increasing dietary calcium intake.<sup>11,12</sup> Both a systematic review<sup>13</sup> and a literature review<sup>7</sup> included education and psychosocial support among the important aspects of effective JIA treatment.

***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.***

	Component	Descriptor	Grade
One moderate quality RCT, <sup>11,12</sup> one Australian guideline <sup>1</sup> and consensus between Australian paediatric rheumatology experts and the RACGP Working Group	Volume of evidence	Satisfactory	C
The consistency of evidence is good and there is consensus	Consistency	Good	B
Satisfactory clinical impact associated with patient education and psychosocial support for JIA patients	Clinical impact	Satisfactory	C

The JIA population is relevant to the Australian context. However, the quality of patient education cannot be guaranteed. There is no evidence specific to Aboriginal and Torres Strait Islander populations. These populations may be disadvantaged because of lack of access to services	Generalisability	Satisfactory	C
The evidence is probably applicable to the Australian health care context with some caveats	Applicability	Satisfactory	C

RECOMMENDATIONS	GRADE
General practitioners should provide ongoing, tailored information to support patient understanding of their disease, treatment options, possible outcomes and their role in self management. They should encourage patients and their carers to seek appropriate information and education opportunities according to their individual needs.	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.	C
<b>Good practice points</b>	
<ul style="list-style-type: none"> <li>Joint protection, energy conservation, pain management and problem solving skills training should be taught early on in the disease course</li> <li>GPs may access information for patients from the Australian Rheumatology Association (ARA) website (<a href="http://www.rheumatology.org.au">www.rheumatology.org.au</a>) or may refer patients and their carers to the site</li> <li>Referral to Arthritis Australia is recommended for general disease and treatment information, as well as support services (<a href="http://www.arthritisaustralia.com.au">www.arthritisaustralia.com.au</a>).</li> </ul>	

## PHARMACOLOGICAL INTERVENTIONS

*The RACGP Working Group recommends consulting the National Prescribing Service ([www.nps.org.au](http://www.nps.org.au)) and the Therapeutic Guidelines – Rheumatology ([www.tg.com.au](http://www.tg.com.au)) for detailed prescribing information, including information on adverse events. When in doubt, contact a paediatric rheumatologist. The nearest paediatric rheumatologist contact is available from [www.rheumatology.org.au](http://www.rheumatology.org.au).*

### Simple analgesics (eg. paracetamol)

EVIDENCE STATEMENT
<p>There is an accepted role for simple analgesics in managing pain in JIA, although the evidence supporting effectiveness specifically in JIA is limited. There are no SRs or RCTs of paracetamol for treating persistent pain in children and adolescents with JIA published between 2000 and 2007. Paracetamol remains the analgesic of choice for treating persistent pain in children and adolescents.<sup>14</sup></p> <p>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.<sup>14-16</sup> <b>Do not rely on dosing charts supplied by the manufacturer as these tend to underdose the child.</b></p> <p><i>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</i></p>

<b>should be used in the short term; it is rarely needed long term in patients with JIA.</b>			
	<b>Component</b>	<b>Descriptor</b>	<b>Grade</b>
One Australian guideline <sup>1</sup> and consensus between Australian paediatric rheumatology experts and the RACGP Working Group	Volume of evidence	Poor	D
The consistency of evidence is satisfactory and there is consensus	Consistency	Satisfactory	C
Satisfactory clinical impact if simple analgesics are used appropriately for pain relief in JIA	Clinical impact	Satisfactory	C
Generalisable to the Australian health care context with few caveats. There is no evidence specific to Aboriginal and Torres Strait Islander populations	Generalisability	Good	B
Applicable to the Australian health care context with few caveats	Applicability	Good	B

<b>RECOMMENDATION</b>	<b>GRADE</b>
General practitioners should consider using paracetamol in regular divided doses for treating moderate pain in children and adolescents with JIA.	<b>C</b>
<b>Good practice points</b>	
<ul style="list-style-type: none"> <li>• 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 dosage</li> <li>• Chronic dosing should not exceed 48 hours without medical advice</li> <li>• Maximum daily dose is 90 mg/kg to a maximum of 4 g/day for treating persistent pain.</li> </ul>	
<b>Cautions</b>	
<ul style="list-style-type: none"> <li>• Paracetamol has few side effects, but dosing is limited by possible hepatotoxicity</li> <li>• Do not rely on dosing charts supplied by the manufacturer as these tend to underdose the child</li> <li>• Do not use slow release preparations in infants and children.</li> </ul>	

## Weak opioids (eg. codeine)

<b>EVIDENCE STATEMENT</b>
There are no SRs or RCTs of codeine for treating persistent pain in children and adolescents with JIA published between 2000 and 2007.
Codeine is the weak opioid of choice for treating persistent pain in children and adolescents. The oral dose for codeine in children is 0.5–1.0 mg/kg every 4–6 hours up to a maximum of 3 mg/kg/day. General practitioners are reminded to use the correct dose for the child's weight. <sup>14</sup>
The cytochrome p450 enzyme (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% Ethiopians and Hong Kong Chinese). Adverse effects of codeine may occur in the absence of analgesia in poor metabolisers. Rapid metabolisers may experience excessive sedation. <sup>15</sup>
<b><i>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.</i></b>

	<b>Component</b>	<b>Descriptor</b>	<b>Grade</b>
There are no recent studies of codeine and JIA (2000–2007). There is consensus between Australian paediatric rheumatology experts and the RACGP Working Group	Volume of evidence	Poor	D
The consistency of evidence is satisfactory and there is consensus	Consistency	Satisfactory	C
The clinical impact is unknown	Clinical impact	Unknown	N/A
There is evidence that use of codeine is not directly generalisable to different racial populations. There is no evidence specific to Aboriginal and Torres Strait Islander populations	Generalisability	Poor	D
The evidence is probably applicable to the Australian health care context with some caveats	Applicability	Satisfactory	C

<b>RECOMMENDATION</b>	<b>GRADE</b>
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.	<b>D</b>
<p><b>Good practice points</b></p> <ul style="list-style-type: none"> <li>• Use the correct dose for the child's weight. The oral dose for codeine in children is 0.5–1.0 mg/kg every 4–6 hours up to a maximum of 3 mg/kg/day</li> <li>• Chronic dosing should not exceed 48 hours without medical advice</li> <li>• In overweight children, the ideal body weight, rather than actual weight, should be used to calculate dose.</li> </ul> <p><b>Cautions</b></p> <ul style="list-style-type: none"> <li>• Adverse effects of codeine may occur in the absence of analgesia in poor metabolisers</li> <li>• Rapid metabolisers may experience excessive sedation</li> <li>• Be alert to paracetamol and codeine doses in combination preparations (eg. Painstop mixtures and Panadeine tablets).</li> </ul>	
<p><b>Traditional non-steroidal anti-inflammatory drugs (NSAIDs)</b></p>	

#### **EVIDENCE STATEMENT**

Traditional non-steroidal anti-inflammatory drugs are the first line drug for the treatment of inflammation in children with JIA.<sup>14</sup> Traditional non-steroidal anti-inflammatory drugs are medications with both analgesic and anti-inflammatory effects with a low risk of serious adverse effects when used appropriately in carefully selected patients. They are used for the initial treatment of pain, inflammation and stiffness. Side effects include gastrointestinal disturbance, rash, mood changes, and sleep disturbance.<sup>9</sup>

There was one good quality, active controlled RCT of meloxicam and naproxen in 232 participants (aged 2–16 years) with active JIA. Meloxicam (0.125–0.25 mg/kg/day) was as effective as naproxen (5 mg/kg twice daily) in producing a positive response on the ACR paediatric 30/50/70 and on other outcome measures when used for up to 12 months. Patients taking both NSAIDs showed significant improvement from baseline, with improvement rates ranging from 58–77% for meloxicam (depending on dose) and 64–74% for naproxen, without consideration to placebo effect.<sup>17</sup>

A low quality systematic review of 14 low quality studies reported on NSAIDs for patients under 16 years of age with arthritis (type not reported). Many of the trials presented were not placebo controlled and did not use randomisation and/or blinding techniques. The

effectiveness of aspirin was investigated in seven small trials (471 participants) for between 2 and 24 weeks. The effectiveness of naproxen was investigated in eight trials (943 participants) for between 4 and 52 weeks. Other small trials investigated the effectiveness of various NSAIDs. In these trials, 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. No significant differences were reported in the safety profiles.<sup>13</sup>

COX-2 inhibitors have not been extensively studied 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.***

	<b>Component</b>	<b>Descriptor</b>	<b>Grade</b>
One good quality RCT, <sup>17</sup> one low quality SR, <sup>13</sup> an Australian guideline <sup>1</sup> and consensus between Australian paediatric rheumatology experts and the RACGP Working Group	Volume of evidence	Good	B
The consistency of evidence is excellent and there is consensus	Consistency	Excellent	A
There is likely to be substantial clinical impact	Clinical impact	Good	B
The evidence is generalisable to the Australian health care context with few caveats. There is no evidence specific to Aboriginal and Torres Strait Islander populations	Generalisability	Good	B
The evidence is applicable to the Australian health care context with few caveats	Applicability	Good	B

<b>RECOMMENDATION</b>	<b>GRADE</b>
General practitioners should prescribe NSAIDs as the initial drug of choice for reducing inflammation and associated pain in the treatment of JIA.	<b>B</b>

#### **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
- Use NSAIDs; a liquid form is available for children who are unable to swallow tablets
- NSAIDs may be prescribed with methotrexate
- 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 decisions on a case-by-case basis.

#### **Caution**

- 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. Studies have described increased sleep disturbance and non-specific abdominal pain. A pseudoporphyrin-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<sup>18</sup>
- 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<sup>18</sup>
- Aspirin is not recommended in children because of the link with Reye syndrome.<sup>15</sup>

## Topical NSAIDs

### EVIDENCE STATEMENT

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.<sup>7, 9</sup>

***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.***

	Component	Descriptor	Grade
There are no studies of topical NSAIDs and JIA (2000-2007). There is consensus between Australian paediatric rheumatology experts and the RACGP Working Group	Volume of evidence	Poor	D
The consistency of consensus is satisfactory	Consistency	Satisfactory	C
There is no evidence in this area	Clinical impact	Unknown	N/A
There is no evidence in this area	Generalisability	Poor	D
The evidence is probably applicable to the Australian health care context with some caveats	Applicability	Satisfactory	C

RECOMMENDATION	GRADE
General practitioners should not prescribe topical NSAIDs to treat patients with JIA.	D

## Complementary and alternative medicines

### EVIDENCE STATEMENT

There are no RCTs or SRs (2000–2007) on the use of complementary and 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, but are not limited to, herbs, vitamins and/or mineral supplements, aromatherapy, and naturopathic and homeopathic products. These products are widely available without prescription in Australia.<sup>19</sup>

A number of SRs on a wide range of complementary medicines in the treatment of arthritis in adult populations have not demonstrated any clinical benefits above placebo.<sup>20–22</sup> Until further research is conducted, these findings are likely to apply to paediatric populations with arthritis. Although generally considered to have a low risk of serious side effects, herbal and nutritional supplements may have harmful effects, particularly through interaction with other medications the patient may be taking.<sup>19</sup>

***It is the consensus of the RACGP Working Group and Australian paediatric rheumatology experts that GPs ask about complementary and 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.***

	<b>Component</b>	<b>Descriptor</b>	<b>Grade</b>
There is consensus between Australian paediatric rheumatology experts and the RACGP Working Group	Volume of evidence	Poor	D
The consistency of opinion is satisfactory	Consistency	Satisfactory	C
The clinical impact is unknown	Clinical impact	Unknown	N/A
Unknown due to lack of evidence in this field	Generalisability	Unknown	N/A
The evidence is probably applicable to the Australian health care context with few caveats	Applicability	Satisfactory	C

<b>RECOMMENDATION</b>	<b>GRADE</b>
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 of complementary and alternative medicines in adult populations with arthritis.	<b>D</b>
<b>Good practice points</b>	
<ul style="list-style-type: none"> <li>• GPs should ask about their patients' use of complementary medicines when prescribing treatment for JIA</li> <li>• GPs may consult a pharmacist if concerns arise regarding medication interactions related to complementary medicines.</li> </ul>	

## NON-PHARMACOLOGICAL INTERVENTIONS

### Nutritional therapy – calcium supplementation

#### EVIDENCE STATEMENT

Children with JIA have been reported to have low BMD early in the disease independent of steroid use. Approximately 15–26% of JIA patients have pathologic fractures before adulthood.<sup>11</sup> 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.<sup>23</sup>

#### Dietary calcium

Evidence from one moderate quality RCT suggests that required daily calcium intake can be achieved without dietary supplement and may result in improvements in some measures of BMD in patients with JIA. Sixty-five families were included in the study; all families were from English speaking backgrounds and had a child aged 4–10 years with a diagnosis of JIA who was not taking oral calcium supplements or systemic corticosteroids. Children whose families participated in both the standard care educational program (control group) and the specific behavioural intervention (experimental group) maintained calcium levels above the recommended intake of 1500 mg/day after the 8 week intervention programs; this effect was sustained at 6 and 12 months. Increase in calcium intake and some measures of BMD translated into statistically superior increases at 6 and 12 months. This RCT suggested that the recommended daily calcium intake can be achieved without dietary supplement. Families with younger children with JIA were more prepared to participate in the intervention.<sup>11,12</sup>

#### Calcium supplementation

There is evidence from one good quality RCT (198 participants aged 6–18 years) that calcium supplementation can improve total BMD in patients with JIA. The population in this RCT had a mean age of 11 years, were primarily Caucasian females, and were not taking oral corticosteroids. Taking daily calcium supplements (1000 mg in conjunction with 400 IU vitamin D) had a small positive effect above placebo on total body BMD. Over 2 years,

patients taking calcium supplements had a net improvement in total body BMD of 1% above those taking placebo. Some patients experienced nausea as a side effect.<sup>24</sup>

### Calcium in combination with vitamin D

There were no SRs or RCTs conducted in children between 2000–2007 on the use of vitamin D in protecting against osteoporosis. In a Cochrane Review of five RCTs 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.<sup>23</sup>

***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.***

	Component	Descriptor	Grade
One good quality RCT, <sup>24</sup> one moderate quality RCT <sup>11,12</sup> and consensus between Australian paediatric rheumatology experts and the RACGP Working Group	Volume of evidence	Good	B
The consistency of evidence is good and there is consensus among the expert group	Consistency	Good	B
There is a satisfactory clinical impact	Clinical impact	Satisfactory	C
Directly generalisable to the Australian health care context with few caveats. There is no evidence specific to Aboriginal and Torres Strait Islander populations	Generalisability	Good	B
Applicable to the Australian health care context with few caveats	Applicability	Good	B

RECOMMENDATION	GRADE
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.	<b>B</b>
<b>Good practice points</b>	
<ul style="list-style-type: none"> <li>• Encourage appropriate caloric and calcium intake</li> <li>• Children with JIA on corticosteroid therapy are at an increased risk of osteoporosis and osteopenia. Additional consideration should be given to calcium and vitamin D supplements when on corticosteroid course.</li> </ul>	

### Land based exercise

#### EVIDENCE STATEMENT

There is evidence of reduced aerobic fitness in children with JIA. A literature review reported on a meta-analysis of five studies (type unspecified) that indicated relative peak oxygen was 21.8% lower in children with JIA than in healthy controls. The review reported support from other studies for this finding and found children with JIA also had shorter exercise times and lower peak heart rates during exercise than healthy controls. These differences appear to be related to disease duration (signs of reduced aerobic fitness were more common in patients with disease duration >2 years) but not disease activity or severity. The literature review

suggested that children with JIA experience muscular deficits including reduced muscle strength and thickness that may be related to disease duration, disease activity and disease severity. Despite these findings, other studies have found that there is no significant difference in participation in physical activity between children with JIA and healthy controls.<sup>25</sup>

One moderate quality RCT (80 children) evaluated the effects of exercise training on physical function in children with JIA. Participation in a 12 week high intensity exercise program resulted in improved physical function as measured by the self reported CHAQ, but not on objective measurement scales. There was no extra improvement in self reported measures for those who participated in high intensity aerobic training compared to those who participated in low intensity training. The exercise regimen may not have been intensive enough to show significant improvement in the time frame. Adherence was significantly greater in the low intensity exercise group (78 vs. 56%).<sup>26</sup>

A literature review that summarised findings from four low quality small studies on land based exercise suggested participation in moderate physical activity for at least 6 weeks (1–3 exercise sessions per week) can improve both muscle function and aerobic fitness in children with JIA.<sup>25</sup> The literature review included additional points:

- 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 to reduce 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.<sup>25</sup>

At least a 1 yearly review by a specialised paediatric physiotherapist is recommended in the Australian Paediatric Rheumatology standards of care.<sup>1</sup>

***It is the consensus of the RACGP Working Group and Australian paediatric rheumatology experts that regular physical activity compatible with children'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.***

	Component	Descriptor	Grade
One moderate quality RCT, <sup>26</sup> an Australian guideline <sup>1</sup> and consensus between Australian paediatric rheumatology experts and the RACGP Working Group	Volume of evidence	Satisfactory	C
The consistency of evidence is satisfactory and there is consensus	Consistency	Satisfactory	C
There is likely to be satisfactory clinical impact	Clinical impact	Satisfactory	C
Directly generalisable to the Australian health care context with few caveats. There is no evidence specific to Aboriginal and Torres Strait Islander populations	Generalisability	Good	B

Applicable to the Australian health care context with few caveats	Applicability	Good	B
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RECOMMENDATION	GRADE
General practitioners should encourage patients with JIA to engage in regular physical activity compatible with their general abilities and the restrictions of their disease.	C
<b>Good practice points</b>	
<ul style="list-style-type: none"> <li>• GPs may utilise EPC items to facilitate access to appropriate services (<a href="http://www.health.gov.au/epc">www.health.gov.au/epc</a>). Eligible services include, but are not limited to, those provided by physiotherapists, occupational therapists, and exercise physiologists</li> <li>• GPs could refer patients to Arthritis Australia for information and services relating to exercise in juvenile arthritis (<a href="http://www.arthritisaustralia.com.au">www.arthritisaustralia.com.au</a>)</li> <li>• Promote physical activity to decrease the risk of osteoporosis and osteopenia</li> <li>• Specific and individualised exercise therapy may be initiated with input from a paediatric trained physiotherapist or occupational therapist (or other appropriately trained therapist)</li> <li>• The choice of exercise may depend on the child's specific needs and preferences.</li> </ul>	

## Aquatic exercise

EVIDENCE STATEMENT			
<p>There is evidence from one non-blinded 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 consisting of aerobic exercises and flexibility and intensity training in a heated pool; program took place in a group setting, was conducted by a physical therapist and consisted of 1 hour a week) after 20 weeks of therapy. Participants had confidence in water before commencing the program.<sup>27</sup></p> <p>A literature review summarised findings from three low quality small studies on aquatic exercise that suggested children with JIA may achieve an increase in physical fitness through aquatic exercise.<sup>25</sup> The literature review included additional points:</p> <ul style="list-style-type: none"> <li>• children with JIA can participate in exercise without disease exacerbation</li> <li>• land based exercise may lead to greater improvements than aquatic exercise</li> <li>• weight bearing exercise is needed to develop optimal bone width and density during childhood</li> <li>• individualised and supervised resistance exercise appears to be safe for children with JIA</li> <li>• the choice of exercise may depend on the child's specific needs and preferences</li> <li>• children with mild disease should be able to participate in most sports with proper screening. 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.<sup>25</sup></li> </ul> <p>At least a 1 yearly review by a specialised paediatric physiotherapist is recommended in the Australian Paediatric Rheumatology standards of care.<sup>1</sup></p> <p><b><i>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 that this is considered a safe form of exercise.</i></b></p>			
Component			
Descriptor			
Grade			
One moderate quality RCT, <sup>27</sup> an Australian guideline <sup>1</sup> and consensus between Australian paediatric rheumatology experts and the RACGP Working Group	Volume of evidence	Satisfactory	C

The consistency of evidence is satisfactory and there is consensus	Consistency	Satisfactory	C
The clinical impact is unknown	Clinical impact	Unknown	N/A
Directly generalisable to the Australian health care context with few caveats. There is no evidence specific to Aboriginal and Torres Strait Islander populations	Generalisability	Good	B
Applicable to the Australian health care context with few caveats	Applicability	Good	B

RECOMMENDATION	GRADE
General practitioners could inform patients about aquatic exercise for children and adolescents and its limited effect.	C
<b>Good practice points</b>	
<ul style="list-style-type: none"> <li>Not all children will have access to hydrotherapy</li> <li>Aquatic exercises can be performed in a standard swimming pool.</li> </ul>	

### Orthotics management (ready and custom made splints)

EVIDENCE STATEMENT
One SR evaluated three before/after intervention case series as no studies of higher evidence level met the review criteria. The findings were of low level of evidence, differences were small, and findings conflicted between studies. Both ready made splints and custom made splints may have a role in the management of JIA for some patients. Although splinting seems to 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; the findings cannot be generalised to other children with JIA. <sup>28</sup>

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

	Component	Descriptor	Grade
One good quality SR of three low quality studies <sup>28</sup> and consensus between Australian paediatric rheumatology experts and the Working Group	Volume of evidence	Satisfactory	C
The findings from three low quality studies were inconsistent	Consistency	Poor	D
The potential impact is only slight or limited	Clinical impact	Poor	D
Generalisable between different patients and types of orthosis. There is no evidence specific to Aboriginal and Torres Strait Islander populations	Generalisability	Poor	D
Probably applicable to Australian health care context with some caveats	Applicability	Satisfactory	C

RECOMMENDATION	GRADE
General practitioners could inform patients about the use of splints and make individualised recommendations in conjunction with appropriately trained multidisciplinary health professionals.	D
<b>Good practice points</b>	
<ul style="list-style-type: none"> <li>GPs should refer patients with JIA experiencing limitations in function for skilled occupational therapy advice</li> <li>Splints should be fitted by an experienced therapist, including, but not limited to, physiotherapists, occupational therapists, orthotists and hand therapists</li> </ul>	

- Consideration of the cost in the context of growing children is important, particularly in the face of limited high grade evidence.

## Orthotics management (foot orthoses)

### EVIDENCE STATEMENT

One low quality, non-blinded RCT (47 children with JIA) investigated the efficacy of custom foot orthotics in improving pain and functional status. All children in the study received supporting 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 compared with the two other groups (ready made shoe inserts and supporting athletic shoes alone). Children in all groups showed improvements in outcome measures.<sup>29</sup>

***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.***

	Component	Descriptor	Grade
One low quality RCT <sup>29</sup> and consensus between Australian paediatric rheumatology experts and the RACGP Working Group	Volume of evidence	Poor	D
The consistency of evidence is satisfactory and there is consensus	Consistency	Satisfactory	C
The potential impact is low and no adverse effects reported	Clinical impact	Poor	D
Findings may not be generalisable between different patients and types of orthosis. There is no evidence specific to Aboriginal and Torres Strait Islander populations	Generalisability	Poor	D
Probably applicable to the Australian health care context with some caveats	Applicability	Satisfactory	C

RECOMMENDATION	GRADE
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.	D
<b>Good practice points</b>	
<ul style="list-style-type: none"> <li>• Most children who have JIA of the lower limb only need comfortable, supporting shoes</li> <li>• Consideration of the cost in the context of growing children is important, particularly in the face of limited high grade evidence</li> <li>• Refer patients to allied health clinicians with specialised paediatric experience, preferably in managing children with JIA.</li> </ul>	

## Thermotherapy

### EVIDENCE STATEMENT

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, heat treatments including heat packs, deep heat ultrasound and warm baths are suggested for decreasing joint rigidity, pain and muscle spasms and increasing joint flexibility. The authors suggested that effectiveness of heat therapy depends on the temperature, duration, rate of temperature change and the area being treated, but did 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.<sup>30</sup>

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.<sup>30</sup>

***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.***

	Component	Descriptor	Grade
There are no studies of thermotherapy and JIA (2000–2007). There is consensus among Australian paediatric rheumatology experts and the Working Group	Volume of evidence	Poor	D
The consistency of expert opinion is satisfactory	Consistency	Satisfactory	C
There are no studies	Clinical impact	Unknown	N/A
Not directly generalisable to the target population but could be sensibly applied. There is no evidence or data specific to racial subgroups such as Aboriginal and Torres Strait Islander populations	Generalisability	Satisfactory	C
Probably applicable to the Australian health care context with some caveats	Applicability	Satisfactory	C

RECOMMENDATION	GRADE
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.	D
<b>Good practice points</b>	
<ul style="list-style-type: none"><li>• A warm bath or shower in the morning may reduce stiffness and pain</li><li>• 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</li><li>• When using thermotherapy, be alert to the patient's comfort and ability to tolerate the treatment.</li></ul>	

## Complementary and alternative physical therapies

### EVIDENCE STATEMENT

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 relevant literature reviews.

In adult populations with arthritis, research on complementary and alternative physical therapies including acupuncture, laser therapy, transcutaneous electrical stimulation (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 therapy regimens (eg. length of sessions, frequency of therapy). It is unclear whether these therapies provide any benefit to children with JIA.<sup>31-34</sup>

***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.***

	Component	Descriptor	Grade
There is consensus among Australian paediatric rheumatology experts and the RACGP Working Group	Volume of evidence	Poor	D
The consistency of opinion is satisfactory	Consistency	Satisfactory	C
The clinical impact is unknown	Clinical impact	Unknown	N/A
Generalisability is unknown due to lack of evidence	Generalisability	Unknown	N/A
Probably applicable to the Australian health care context with few caveats	Applicability	Satisfactory	C

RECOMMENDATION	GRADE
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.	D
<b>Good practice points</b> <ul style="list-style-type: none"><li>• Complementary or alternative physical therapies should not replace pharmaceutical therapies and exercise in the management of JIA</li><li>• GPs could refer patients who request information or advice on complementary or alternative physical therapies, including information on any risks, to Arthritis Australia (<a href="http://www.arthritisaustralia.com.au">www.arthritisaustralia.com.au</a>).</li></ul>	

## DISEASE MONITORING AND COMORBIDITIES

### EVIDENCE STATEMENT

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.<sup>5</sup>

Literature reviews suggested 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.<sup>13</sup> Patients taking NSAIDs for more than 3–4 weeks should have monitoring laboratory tests (FBC; urea, electrolytes and creatinine; LFTs) even in the absence of clinically apparent adverse effects.<sup>5</sup>

Uveitis is chronic and usually asymptomatic. It 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.<sup>5,8</sup>

***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.***

	Component	Descriptor	Grade
There is consensus between Australian paediatric rheumatologists and the RACGP Working Group	Volume of evidence	Poor	D
The consistency of opinion is satisfactory	Consistency	Satisfactory	C
The impact is likely to be good	Clinical impact	Good	B
Directly generalisable to the target population with few caveats	Generalisability	Good	B
Applicable to the Australian health care context with few caveats	Applicability	Good	B

RECOMMENDATION	GRADE
General practitioners should be involved in monitoring disease progression and comorbidities in conjunction with the treating paediatric rheumatologist.	C
<b>Good practice points</b>	
<ul style="list-style-type: none"><li>• Paediatric rheumatology review should take place at least twice per year</li><li>• Regular screening for uveitis is recommended (see the JIA guideline for further details)</li><li>• 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</li><li>• Patients need to be monitored for potential toxicity and side effects of medications</li><li>• Frequency and type of monitoring will depend on the DMARD prescribed, but most require a FBC (to monitor for marrow suppression) and LFTs (to look for raised transaminases as a sign of hepatotoxicity) approximately every 1–3 months (once on a stable dose). Adverse effects are less common than in adults with rheumatoid arthritis on DMARDs.<sup>1,5,6</sup></li></ul>	

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## APPENDIX A. PROCESS REPORT

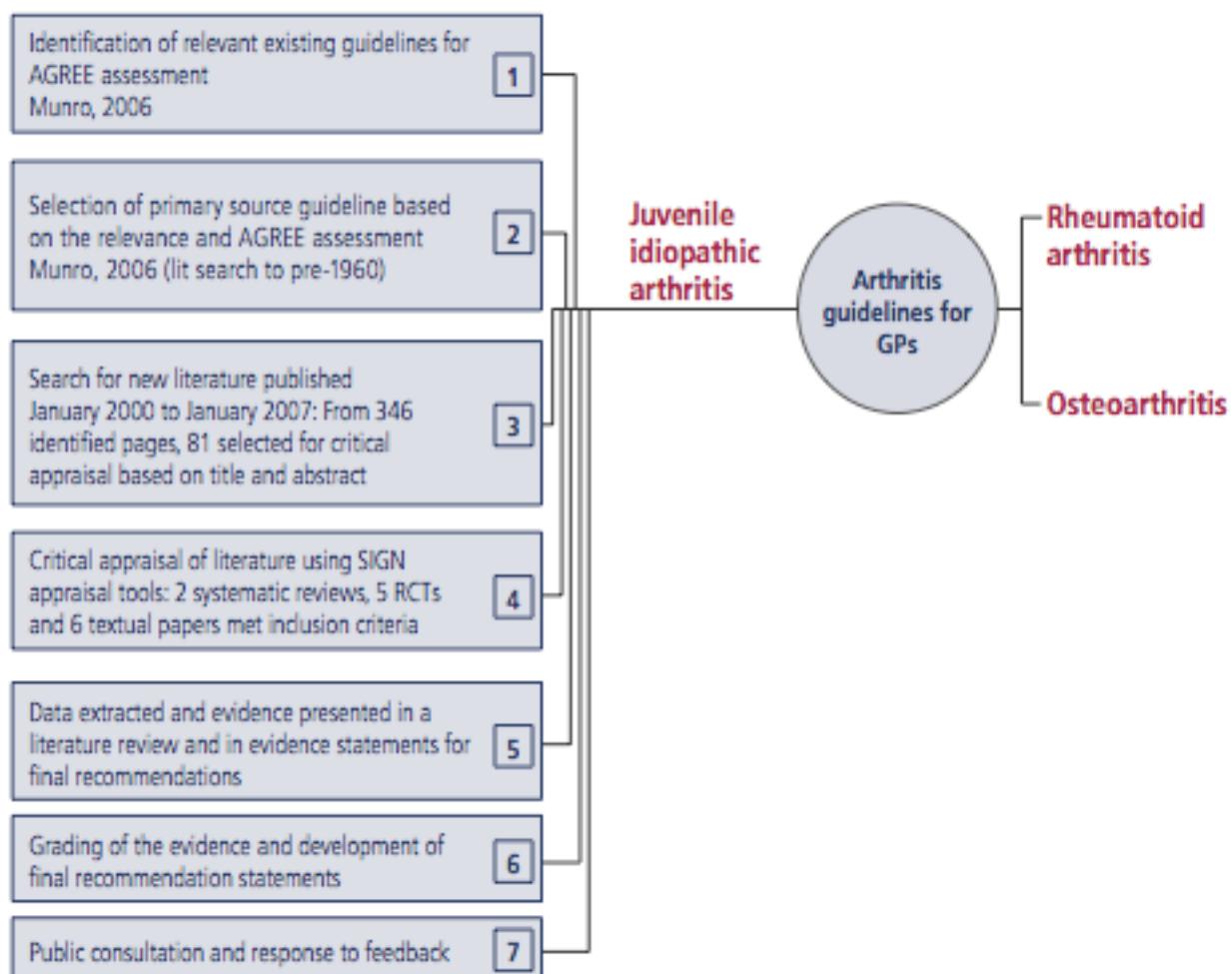
This report outlines the process used for the development of the evidence based *Recommendations for the diagnosis and management of juvenile idiopathic arthritis*.

The project consisted of the following major phases:

- formation of a multidisciplinary expert working group and an Australian paediatric expert group (see *Appendix B*)
- development of a scoping document outlining the scope and objectives of the project, including the process to be used in 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 *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 2005–2006 and through those known to the RACGP Working Group. Guidelines considered to be the most relevant were selected for appraisal using the AGREE instrument.<sup>1</sup> 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'.<sup>1</sup> 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 four 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 RACGP Working Group identified only one relevant existing guideline, highlighting the paucity of evidence based recommendations in this field. This guideline was:

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

This guideline was assessed by two reviewers using the AGREE tool. 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 of the RACGP Working Group, Dr Munro, acknowledged her potential conflict of interest as project director for development of *Juvenile idiopathic arthritis management guidelines (Provisional)*<sup>2</sup> 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)*<sup>2</sup> 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)*.<sup>2</sup> 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](http://www.racgp.org.au/guidelines/juvenileidiopathicarthritis/literaturereview)).

## Search strategy

The Cochrane Library (including the CENTRAL Cochrane Controlled Trial Register) and the MEDLINE, EMBASE, and CINAHL databases 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 reported in systematic reviews 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 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 were considered for inclusion, as were studies that included children diagnosed as having JIA.

## 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<sup>3</sup>**

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-1	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 systematic reviews ([www.sign.ac.uk/guidelines/fulltext/50/checklist1.html](http://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](http://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](http://www.nhmrc.gov.au) – cited September 2006); the Joanna Briggs Institute data extraction tool for systematic reviews (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](http://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 people and Torres Strait Islanders); rural and remote communities; Muslim Australians; and Vietnamese Australians – (which was a 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 the 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 related to Australian Aborigines, three related to rural health and two 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 RACGP 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 (*Appendix B*).

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, a summary of *Juvenile idiopathic arthritis management guidelines (Provisional)*.<sup>2</sup> A body of evidence assessment matrix developed by the NHMRC<sup>3</sup> (*Table 3*) was used to assess the volume and consistency of evidence supporting each recommendation, as well as the recommendation's clinical impact, generalisability and applicability.

Each recommendation was given a final grading (*Table 4*) representing its overall strength. The gradings reflect implementability in terms of the confidence with which practitioners can use the

recommendations 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<sup>3</sup>**

Component	A	B	C	D
	Excellent	Good	Satisfactory	Poor
<b>Volume of evidence</b>	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
<b>Consistency</b>	All studies consistent	Most studies consistent, and inconsistencies may be explained	Some inconsistency reflecting genuine uncertainty around clinical question	Evidence is inconsistent
<b>Clinical impact</b>	Very large	Substantial	Moderate	Slight or restricted
<b>Generalisability</b>	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
<b>Applicability</b>	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<sup>3</sup>**

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

## Consultation phase

Draft versions of the *Clinical guideline for the diagnosis and management of juvenile idiopathic arthritis*, *Recommendations for the diagnosis and management of 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 RACGP Working Group.

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

## Dissemination

Final versions of the *Clinical guideline for the diagnosis and management of juvenile idiopathic arthritis*, *Recommendations for the diagnosis and management of 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.

## Process report references

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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. MEMBERSHIP AND TERMS OF REFERENCE OF THE RACGP JUVENILE IDIOPATHIC ARTHRITIS WORKING GROUP

### Aim of the RACGP Working Group

The aim of the RACGP Working Group was to undertake activities required to fulfil 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 the RACGP Working Group

In accordance with the project contract, membership of the RACGP 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
<b>Dr Jane Munro</b> (Chair) Paediatric rheumatologist Royal Children's Hospital Melbourne, Victoria	Paediatric Rheumatology Association	MBBS, MPH, FRACP
<b>Dr Shane Brun</b> Associate Professor Musculoskeletal and Sports Medicine Queensland	RACGP School of Medicine and Dentistry, James Cook University, Queensland	FFSEM(UK), FACRRM, FASMF, FARGP, MSpMed, DCH, GradDipRuralMed, GradDipEd, BAppSc, FRACGP
<b>Dr Morton Rawlin</b> Project Director	RACGP – Director of Educational Services	BMed, MMedSci, DipPracDerm, DipFP, DipMedHyp, DipBusAdmin, FACRRM, FRACGP
<b>Pam Webster</b> Consumer representative	Consumer and NAMSCAG Carers Australia	MCH, BA, DipT, AUA

New South Wales		
<b>Prof Karen Grimmer-Somers</b>	NHMRC Advisor	PhD, MMedSc, BPhty, LMusA, Cert HlthEc
<b>Amy Jasper</b> Project Manager	RACGP – Manager, Education Evaluation	MBA, GDipHumServRes, BAppSci(AdvNsg)
<b>Dr Jiri Rada</b>	RACGP Project Officer	PhD, FRSH, MSc, BPHE, BA
<b>Emily Haesler</b>	RACGP Project Officer	BN, PGradDipAdvNsg

## Membership of the Australian Paediatric Rheumatology Expert Group

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