Juvenile arthritis in Australia

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Juvenile arthritis in Australia

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Abbreviations

ABS	Australian Bureau of Statistics
AIHW	Australian Institute of Health and Welfare
CURF	confidentialised unit record file
ILAR	International League Against Rheumatism
K10	Kessler 10-item Psychological Distress Scale
NHS	National Health Survey
SDAC	Survey of Disability, Ageing and Carers

Summary

Juvenile idiopathic arthritis is the general term used to describe inflammatory arthritis in children. An estimated 4,600 Australian children under the age of 16 had arthritis in 2004–05. Based on these self-reports, almost 22,000 adults had been diagnosed with arthritis in childhood.

More common in girls, than in boys, juvenile arthritis, is estimated to be the main disabling condition for about 1,600 children. A large majority (83%) of children with this form of disability had severe activity limitation. Much disability was also noted in adults with disease onset in childhood.

Juvenile arthritis is not a 'mini' version of adult arthritis. Both its presentation and course are considerably different from those of the adult form of the disease. Because of its early onset, juvenile arthritis seriously affects the growth and development of the child. There is also a greater and long-term need for health care services throughout life in some cases. The burden of juvenile arthritis on the family is also high through reduced social interaction, role restrictions, and the cost of ongoing management and treatment. Some of the significant impacts of juvenile arthritis along its life course, based mostly on data from the 2004–05 National Health Survey and 2003 Survey of Disability, Ageing and Carers, both conducted by the Australian Bureau of Statistics (ABS) are given below.

Key findings

Impact in childhood

- Almost all children with arthritis report chronic/recurrent pain.
- A large majority of children with arthritis are restricted in physical activity or unable to do physical work (69%). Almost half have limited use of arms or fingers and difficulty gripping or holding things.
- Children with arthritis are more likely to have days away from school (33%) and days of reduced activity (16%) compared with children without arthritis (18% and 11% respectively).
- These children had significant problems at school, with almost all having difficulty participating in sports, 86% not fitting in socially and 57% unable to sit properly.
- Long periods of active arthritis may impair muscle development, resulting in generalised growth retardation, uneven limb lengths, joint erosion and lower aerobic capacity.
- The disease process and medication regime can also alter physical appearance, resulting in small jaw bones and shortening of fingers, forearms, toes or feet.

Impact later in life

- Adults with arthritis that was juvenile-onset are more likely to consider their health as fair or poor (33%) compared with those with adult-onset (28%).

- Around 32% of adults have severe activity limitation. Some (11%) are limited or restricted in physical activities, 7% report chronic pain, and 5% have difficulty gripping or holding things.
- These adults are also more likely to have days away from study (17%) and days of reduced activity (27%) than those with adult-onset arthritis (7% and 20% respectively).
- Adults with a history of juvenile-onset arthritis have higher rates of unemployment compared with their healthy peers, with around 40% not being in the workforce and 14% of those in the workforce working part-time. Another 17% have difficulty getting full or part-time work.
- Around 27% of adults with juvenile-onset arthritis have a high or very high level of psychological distress. It is reported more commonly by those in the 16–34 years age group.
- Adults with juvenile-onset arthritis are more likely to live by themselves and be less sexually active, and are less likely to be in stable relationships. Fertility is reduced in females with juvenile arthritis. They also have increased rates of miscarriage and may suffer from other reproductive problems.

Impact on the family

- Parents of children with arthritis are more likely to report health problems, family conflict and stressful life events. Mothers are affected the most, experiencing higher levels of stress and depression.

1 Introduction

Arthritis is not only a disease of the elderly; it affects younger people as well. An estimated 4,600 Australian children in 2004–05 had arthritis. While remission is common, the disease can become chronic and result in complications over time. Juvenile arthritis also has adverse effects on children's growth and musculoskeletal development.

Juvenile arthritis—juvenile rheumatoid arthritis or juvenile chronic arthritis as it is sometimes referred to—was declared a focus area under the Better Arthritis and Osteoporosis Care (BAOC) budget initiative of the Australian Government in 2006. The disease deserves focused attention because:

- It affects children in the prime of their growth and development. This may lead to activity limitations and physical deformities more commonly and much earlier in life, requiring a greater and long-term need for health care services.
- The limitations imposed by chronic pain, in particular the inability to run and play with other children, create an acute sense of helplessness. This may limit participation in school activities, affect many day-to-day activities and cause much anxiety.
- The disability associated with juvenile arthritis may also have a significant impact on a child's own health perception and body image.
- The management of juvenile arthritis invariably involves parents and other family members, which may impact on family dynamics.

Over the last several years, there has been a greater awareness of the need to manage the disease early and minimise its impact. With advanced treatment options now available it is important to recognise and treat this disease as soon as possible.

What is juvenile arthritis?

Juvenile arthritis is an umbrella term used to describe arthritis in children. The diagnosis is considered if the disease begins before the 16th birthday and lasts at least 6 weeks.

The disease is extremely diverse in its features. Many of the children are affected in only one or a few joints, commonly referred to as monoarthritis or oligoarthritis, and often as an acute event. Some may have arthritis in multiple joints, or polyarthritis, which tends to become chronic. Many children are also affected in areas other than joints—eyes, skin and other body tissues. In some cases, the symptoms may alter over the course of the disease.

Juvenile vs adult arthritis

Juvenile arthritis is not a mini-version of adult arthritis. The conceptualisation of arthritis as an outcome of ageing or exposure over the long-term to a variety of risk factors does not strictly apply to arthritis in children.

To better understand juvenile arthritis it is important to view it in the context of arthritis over the lifetime. Arthritis (Greek *arthr* = joint, *itis* = inflammation) is marked by inflammation of the joints, causing pain, stiffness and damage to the joint cartilage and surrounding structures. In some cases it is associated with problems in other tissues and organs of the

body. The damage to a joint and its surrounding structures can lead to joint weakness, instability and visible deformities. Depending on the joint involved, arthritis can interfere with many tasks of daily living, such as dressing, walking and climbing stairs, etc.

In adults, arthritis is generally considered to be a disease of older persons. This is largely because the development of its major form, osteoarthritis, is due to the destruction of cartilage and bony changes over long time. In fact, the symptoms and clinical feature of most forms of adult arthritis develop over a much shorter duration, manifesting across all age ranges. A characteristic feature of arthritis is its extreme diversity. More than 150 forms of the disease are known. Both acute and chronic phases exist, with a tendency to become life long. Females have the disease more commonly than males.

Like the adult forms of arthritis, the main symptoms of juvenile arthritis are pain, swelling and stiffness. Also, both systemic and localised forms of arthritis are encountered in children. However:

- The age-limited definition of juvenile arthritis ensures that some forms of later onset arthritis (for example, osteoarthritis) are not covered by this category.
- While children develop many of the same forms of arthritis that affect adults, some forms are unique to children.
- Many children with arthritis will outgrow their illness, whereas adults will usually have lifelong symptoms.
- Many cases of juvenile arthritis lead to early onset of physical disability.
- Congenital, genetic and immunological processes play a greater role in juvenile arthritis than in adult arthritis.

History and classifications

Juvenile arthritis has been recognised since the mid-1800s. In 1864, four cases of juvenile chronic arthritis in children aged 12 years were described by Cornil. In 1897 George Frederick Still provided clinical description in 19 cases that formed the basis for diagnosis of juvenile arthritis. That condition is known as Still's disease.

The European League Against Rheumatism (EULAR) and the American College of Rheumatology (ACR) developed diagnostic criteria and classifications for the disease (Brewer et al. 1977; Wood 1978). These classification systems however have created some problem as neither of them is able to identify uniform subgroups of the disease.

The International League Against Rheumatism (ILAR) devised a unifying set of criteria, using the umbrella term 'juvenile idiopathic arthritis', to refer to arthritis in children (Petty et al. 1997, 2004). First proposed in 1995, and later revised in 1997 and 2001, the ILAR classification system is now used in many countries. However, much of the scientific and medical literature still uses the EULAR and ACR systems.

The EULAR, ACR and ILAR classifications and their diagnostic criteria are outlined in Table 1.1 for comparison. Succinct descriptions of various forms of juvenile arthritis, as detailed under the ILAR classification system, are given in Appendix A.

Table 1.1: Comparison of various classifications of juvenile arthritis and their diagnostic criteria

	Classification		
	European League Against Rheumatism	**American College of Rheumatology**	**International League Against Rheumatism**
Entity	Juvenile chronic arthritis (JCA)	Juvenile rheumatoid arthritis (JRA)	Juvenile idiopathic arthritis (JIA)
Age at onset	Under 16 years	Under 16 years	Under 16 years
Subgroups	Pauciarticular (few joints: 1 to 4)	Pauciarticular JRA (1 to 4 joints)	Oligoarthritis (persistent), (1 to 4 joints)
			Oligoarthritis (extended), eventually affecting 5 or more joints
	Polyarticular (5 or more joints, RF negative)	Polyarticular JRA (5 or more joints)	Polyarthritis (RF positive)
			Polyarthritis (RF negative)
	Systemic; arthritis with characteristic fever	Systemic; arthritis with characteristic fever	Systemic; arthritis with characteristic fever
Other subgroups	Psoriatic arthritis		Psoriatic arthritis
	Ankylosing spondylitis		Enthesitis-related arthritis
			Undifferentiated arthritis
Duration[(a)]	3 months	6 weeks	6 months
Other features	Includes spondyloarthropathies	Excludes spondyloarthropathies	Includes spondyloarthropathies

(a) Length of illness before diagnosis.

Source: Petty et al. 2004.

Monitoring juvenile arthritis

Epidemiological and other information on juvenile arthritis in Australia is insufficient due to its diverse nature and problems related to accurate diagnosis. Nonetheless, juvenile arthritis is a source of much ill health, pain and deformity contributing to functional limitations. It can also have a significant impact later in life. It is important, therefore, that accurate, reliable and comprehensive information about juvenile arthritis becomes available.

Effective monitoring can facilitate the management of juvenile arthritis by determining its extent and impact as well as any trends in its occurrence. This in turn can help with service planning, inform national policies and strategies, and identify children with special needs.

Recently, juvenile idiopathic arthritis has been added as a focus condition under the National Health Priority Area (NHPA) and the Better Arthritis and Osteoporosis Care Program.

This report

This baseline report attempts to put together information on juvenile arthritis in Australia. The magnitude of the problem, its impact and information about prevention, treatment and management are described.

Most of the information included in this report is based on self-reports (or reports by parents or other members of the family) in the 2004–05 National Health Survey (NHS) and 2003 Survey of Disability, Ageing and Carers (SDAC), both conducted by the Australian Bureau of Statistics (ABS). Additional information, where relevant, has been derived from administrative data sources (see Appendix B). In view of data limitations, the report is generally expository in nature. The term juvenile arthritis is used consistently in the document from here onwards as an abbreviated form of juvenile idiopathic arthritis.

The report is organised into five chapters.

1. This introductory chapter provides background information on juvenile arthritis in Australia.
2. Chapter 2 provides an overview of the disease including its clinical presentation, risk factors, prognosis, associated complications and comorbidities.
3. The magnitude of the problem, using prevalence, morbidity, activity limitation, and health service use and costs data, is detailed in Chapter 3.
4. Chapter 4 outlines the effects of the disease on the child as well as on the family. The effects of arthritis on a young person's physical and mental health, growth and development, education, social interaction and quality of life are also described.
5. Treatment and management options of juvenile arthritis are described in Chapter 5. Information on the use of medications is also presented.

2 Nature of the problem

The clinical problem

As described in Chapter 1, juvenile arthritis is a diverse disease with much variation in its presentation, prognosis and complications. It is therefore difficult to provide a single description of juvenile arthritis. However, there are some common clinical features to provide an overview.

Like the adult forms of arthritis, the main symptoms of juvenile arthritis are pain, swelling and stiffness. Joint stiffness is usually common after rest or decreased activity level (also referred to as morning stiffness), and weakness in muscles and in other soft tissues around the joints involved.

Most of the children with arthritis are affected in a few joints. Arthritis affecting four or fewer joints is commonly referred to as oligoarthritis. In some cases, only one joint (monoarticular) is affected. Mostly asymmetrical in nature, arthritis in children mainly affects joints of long bones such as the knees, elbows, wrists and ankles. Some children are also affected, to varying degrees, in areas other than joints such as the eyes, skin or other body tissues.

The signs and symptoms of juvenile arthritis vary from child to child, and even from day to day in the same child. Joint stiffness and pain may be mild one day but the next day it may be so severe that the child has great difficulty moving or is unable to move at all. Periods when the arthritis is particularly active are called 'flares'. Children usually have fever and a general feeling of being unwell. Some have skin rashes, anaemia and vision problems.

The major symptoms of different types of juvenile arthritis, as detailed by ILAR, are given in Table 2.1.

Table 2.1: Symptoms and effects of different ILAR types of juvenile arthritis

ILAR type	Subcategory/ alternative name	Symptoms/presentation	Other affected areas
Oligoarthritis	Monoartcicular or pauciarticular	Can cause long bones to grow at different rates, causing a limp and damage to the affected joints.	Inflammatory eye condition (iridocyclitis)
	Extended oligoarthritis	After 6 months 5 joints may be affected.	Eye condition (uveitis)
Systemic onset arthritis	Sometimes called Still's disease	Illness, fever, swollen lymph glands, skin rash.	Heart, spleen, lymph nodes
Polyarticular arthritis	Rheumatoid factor –ve	Illness, fatigue and unwillingness to move the affected joints.	
	Rheumatoid factor +ve	Low-grade fever, anaemia, in some children inflammation of internal organs. Damage to other joints, mainly the jaw causing jaw pain and discomfort with chewing, affecting dental care and eating habits. In the spine, neck stiffness and difficulty turning the head side to side may occur.	Chronic uveitis
Enthesitis-related arthritis		The symptoms may resolve and come back in later years. Sometimes the child may develop ankylosing spondylitis.	
Psoriatic arthritis		Sausage-shaped toes or fingers and 'pitting' or dints in the nails.	Skin rashes

Source: Miller 2006.

Diagnosis

An accurate diagnosis is required for proper treatment of juvenile arthritis. Early diagnosis and treatment can control inflammation, relieve pain, prevent joint damage and maintain the affected child's ability to function. If left undiagnosed and untreated this can result in poor prognosis of the condition, in many cases causing irreversible damage to the joints, bones and organs.

Medical history and physical examination

Diagnosis of juvenile arthritis is mostly made on the basis of medical history and physical examination. There is no single defining test although several tests are used to help with the diagnosis (see below). A positive diagnosis is made when there has been persistent arthritis in one or more joints for at least 6 weeks after other possible illnesses have been ruled out. Many other potential conditions may resemble the joint pain associated with arthritis. These conditions may be:

- non-inflammatory causes such as trauma, slipped epiphysis, osteochondrosis
- blood disorders such as leukaemia or haemophilia with recurrent intra-bleeds inside the joints
- connective tissue disorders such as systemic lupus erythematosus (SLE), scleroderma
- malignancy including bone tumours, neuroblastoma

- other pain such as growing pains, fibromyalgia, inflamed spinal discs, and inherited or congenital syndromes.

Laboratory tests

X-rays and blood tests also help identify juvenile arthritis. Sometimes, a variety of tests such as erythrocyte sedimentation rate (ESR), rheumatoid factor (RF), antinuclear factor (ANA) and bone scan, may be necessary for a firm diagnosis of juvenile arthritis. The usefulness of various tests is shown in Box 2.1.

Box 2.1: Diagnostic tests for juvenile arthritis

Erythrocyte sedimentation rate (ESR)

This blood test is indicative of active inflammation in the body. ESR is always elevated in children with systemic juvenile arthritis. The test is useful to monitor the success of treatment.

Rheumatoid factor (RF)

A determination of RF in the blood is one of the significant tests for the diagnosis of juvenile arthritis as well as to determine its type.

Antinuclear factor (ANA)

Identifying ANA in the blood is important as it can be observed in children with pauciarticular juvenile arthritis.

Complete blood count (CBC)

CBC is done to detect anaemia that is common in children with juvenile arthritis. This test can also indicate various abnormalities in white blood cells or platelets that are associated with the immune system and blood clotting.

X-ray, medical resonance imaging (MRI), computer topography (CT) scan, dual-energy x-ray absorptionmetry (DXA), and bone scan

All these imaging tests may help to detect damage in the affected joints. A bone scan can also detect the inflammation of joints and bone as well as other abnormalities.

Source: Miller 2006.

Causal mechanisms and risk factors

The causal mechanisms and risk factors for juvenile arthritis are largely unknown. The diverse nature of the disease—in symptoms, age of onset, sex ratio and so forth—suggests several distinct causal mechanisms. The varying progression of the disease and associated complications further point to the role of other factors.

Given this variation in disease development and progression, a clear picture of 'what goes wrong' has not emerged. However, three major possibilities—genetic susceptibility, autoimmunity and environmental triggers—come into view.

Genetic susceptibility

Several studies have revealed familial aggregation of juvenile arthritis. The disease and its concomitants are noted more commonly in siblings and relatives of children with arthritis. Family members of children with arthritis are also reported to have other musculoskeletal disorders such as ankylosing spondylitis, Reiter's disease, spondylitis with inflammatory bowel disease and psoriatic arthritis, all disorders with high genetic content, more commonly (Yamamoto 2003).

Multiple genes contribute at various points in the arthritis pathology. Juvenile arthritis is associated with a range of human leukocyte antigens (HLA) (Forre & Smerdel 2002). These HLA associations differ in various forms of juvenile arthritis. HLA are important for presenting parts of foreign proteins to the immune system. Many non-HLA factors may also contribute to the disease process.

Environmental triggers

The role of environmental triggers in precipitating the disease, either directly or through autoimmune mechanisms, also remains poorly understood. Infection has been implicated in the onset and exacerbation of some forms of arthritis in children with known genetic predisposition, as described below. However, microbiological studies have failed to reveal the mechanisms for infections in juvenile arthritis (Schneider & Passo 2002).

Viral agents

Common viral agents including rubella, especially after immunisation with the measles-mumps-rubella vaccine, are implicated in the pathogenesis of arthritis (Benjamin et al. 1992). In children, infection by rubella virus is associated with chronic arthritis as well as other musculoskeletal problems. Viral infections such as influenza A (Life et al. 1993), parvovirus B19 (Nocton et al. 1993), herpes simplex virus (Williams & Malone 1992), cytomegalovirus (Ansell 1983), coxsackieviruses and adenoviruses (Rahal et al. 1976) have also been occasionally implicated. A recent study has found a causal relationship between HIV infection and arthritis in children, with 78% of HIV-infected children developing arthritis (Chinniah et al. 2005).

Bacterial agents

Bacteria associated with reactive arthritis, notably yersinia, salmonella, shigella and campylobacter, have been suggested to play a causative role in pauciarticular juvenile chronic arthritis (Sieper et al. 1992). Several studies have proposed an association between serological evidence of borrelia infection and juvenile chronic arthritis (Saulsbury & Katzmann 1990; Banerjee et al. 1992). Although *Chlamydophila pneumoniae*, when carried to the joint fluid, can evoke a local inflammatory response (Gerard et al. 2000), Altun and colleagues (2004) did not find this infection as a significant triggering factor in juvenile arthritis.

Prognosis, complications and comorbidity

The prognosis of juvenile arthritis is favourable overall, although, depending on the type, there are some variations (Minden et al. 2000). Some forms of juvenile arthritis are extremely

variable in their rate of progression. In many cases, the inflammation may become inactive after several years; however, about 85% of children may have the condition for months to 3 or 4 years. Arthritis may be severe in about 15% of cases, either from the onset or later. The prognoses of some subgroups of juvenile arthritis are shown in Table 2.2.

Table 2.2: Prognoses of various subgroups of juvenile arthritis

Subgroup	General prognosis	Prognosis for severe cases
Oligoarthritis, pauciarticular and monoarticular arthritis	In 80% of cases, the disease resolves within 15 years.	Severe joint involvement: 15% Eye involvement a major problem at 10 years after presentation: Reduced visual acuity: 50% Cataracts and glaucoma: 25%
Extended oligoarthritis	Symptoms often persist over a longer period of time, but eventually go away.	
Systemic onset	50% remit without recurrence. Prognosis is worst in those with disease onset before the age of 5. About 4% die from infection and amyloidosis.	50% have polyarticular arthritis and 33% have severe destruction.
Polyarticular arthritis (RF negative)	Majority go into remission with little erosive disease.	10–15% have severe limitation at 15 years after presentation.
Polyarticular arthritis (RF positive)	Only 33% are independent after 15 years.	63% have severe joint destruction, in many cases leading to total loss of independence.
Enthesitis- related arthritis	Symptoms may go away within a short period. In some children the symptoms may go away and then recur later in adolescence or adulthood.	Some children may go on to develop ankylosing spondylitis.
Psoriatic arthritis	The symptoms may go away after a short period of time. Some children's symptoms may go away and then recur later in adolescence or adulthood.	

Source: Pachman & Poznanski 1998.

Children with juvenile arthritis generally suffer from a variety of growth disorders (details in Chapter 4). These range from general growth retardation to local acceleration of growth in the affected limb (MacRae et al. 2006). Many are likely to develop scoliosis.

Juvenile arthritis may lead to or be associated with various complications. The disease increases the risk of fractures, especially during adolescence (Burnham et al. 2006). A study in the United Kingdom has reported that 6.7% of children with juvenile arthritis had a fracture compared with 3.3% of other children of the same age. The most common fracture sites were the arm and leg bones. Low bone mass, which may increase the possibility of minimal trauma fracture, is another known complication of juvenile arthritis. This may be because of chronic inflammation, delayed puberty, malnutrition, weakness, inactivity, and treatment with corticosteroids. Heart complications such as pericarditis, aortic valve regurgitation, and cardiac tamponade (which can in particular be a life-threatening complication if untreated) are also noted. The complications may vary with the type of juvenile arthritis (see Table 2.3).

Table 2.3: Selected subgroups of juvenile arthritis and their complications

Subgroup	Possible complications
Systemic-onset	pericarditis
	haemolytic anaemia
	disseminated intravascular coagulation
	endarteritis in the fingers and toes resulting in very poor circulation
Pauciarticular	knee flexion contractures
	reduced length
Polyarticular	skeletal abnormalities—accelerated bone age
	narrowed joint spaces
	swan-neck and/or boutonniere deformities
	joint subluxation
	cervical spine involvement

Source: Pachman & Poznanski 1998.

Children with juvenile arthritis are more likely to have diseases such as autoimmune thyroiditis, subclinical hypothyroidism and coeliac disease (Stagi et al. 2005). Persistent arthritis may be seen in several other childhood diseases or congenital malformations, including SLE, scleroderma, vasculitis, reactive arthritis, juvenile dermatomyositis, septic arthritis, haemophilia and cystic fibrosis. The damage to growing joints in childhood may persist into adult life, causing difficulty in movement and joint use.

The physical impairments, activity limitations and long-term outcomes of juvenile arthritis are described in detail in Chapter 4.

3 Magnitude of the problem

As described in the preceding chapter, juvenile arthritis is a source of much ill health, pain and deformity, contributing to activity limitation, time off school, and low participation in physical and social activities in children. Over the course of the disease, there may also be many complications.

No single measure can provide an overview of the various dimensions of this problem, so a set of measures is needed to map these dimensions. While measures of incidence and prevalence provide information about disease occurrence and persistence, measures of disease severity, use of health services and quality of life can broaden the view of the problem's impact. Disease costs provide an added perspective.

An additional dimension to mapping the extent of the problem is that childhood disorders such as juvenile arthritis not only cause significant burden in childhood but also entail much illness, disability and poor health outcomes in adulthood. The use of health care services increases over time with substantial costs. Premature mortality is also not uncommon.

The mosaic of information about juvenile arthritis in Australia that follows has been derived from a variety of data sources. An attempt has been made to integrate this somewhat disparate information to create an overall picture.

Incidence and prevalence

Incidence

There are no established national data about the incidence of juvenile arthritis in Australia. In the absence of a disease registry, it is difficult to determine the incidence of a disease that is low in occurrence and difficult to fully characterise. The diverse nature of the disease and the use of overlapping definitions further make the estimation of incidence difficult.

The Burden of Disease and Injury Study has indirectly estimated the incidence of juvenile arthritis in Australia (Begg et al. 2007). Using DISMOD (an epidemiological disease model linking populations exposed to risk of disease with incident cases, prevalent cases, case fatality and the duration of time lived with a disease or injury, including its sequelae), the study estimated 368 new cases (92 males; 276 females) among those aged 0 to 14 years in 2003. This number equates broadly to 24 new cases per 100,000 person years.

The global incidence of juvenile arthritis ranges between 7 and 23 per 100,000 person years in the United States of America and Northern Europe (Table 3.1). The DISMOD estimate for Australian children is at the upper end of this range.

Table 3.1: World-wide incidence of juvenile arthritis

Geographical region/country	Study type	Incidence range (per 100,000 person years)	Reference/source
USA	Population-based	7–21	Peterson et al. 1996
Northern Europe	Population-based	7–21	Kaipiainen & Savolainen 1996; Bernston et al. 2003
Norway and France	Retrospective	1–23	Prieur et al. 1987; Moe & Rygg 1998
Scandinavia	Population-based	7–23	Burgos-Vargas & Clark 1989
Germany	Prospective	7	von Koskull et al. 2001
Berlin, Germany	Retrospective	4	Kiessling et al. 1998
Australia	DISMOD	24	Begg et al. 2007

Prevalence

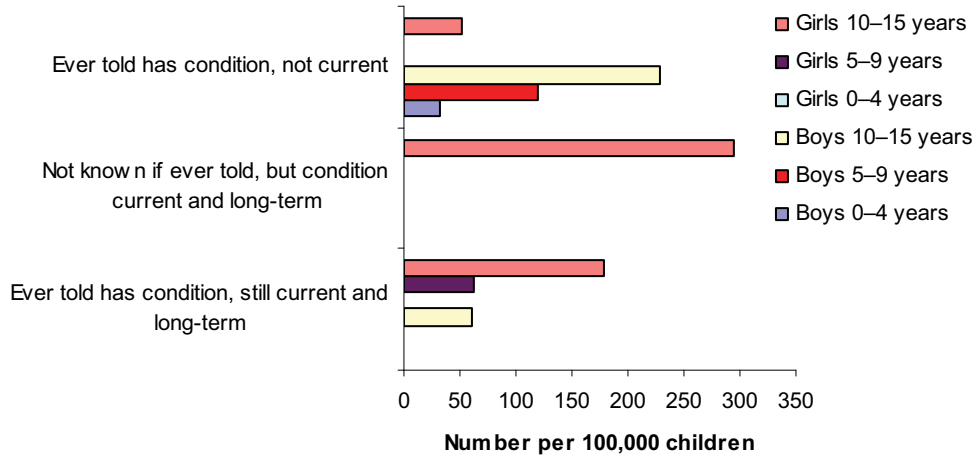
Because of its age-limited definition, estimation of the prevalence of juvenile arthritis is often based on the number of children with arthritis in the population. Most of the studies reporting prevalence of juvenile arthritis have taken this approach. However, this notion runs counter to the traditional meaning of disease prevalence as the product of incidence and disease duration. There are many adults in the population with juvenile arthritis who need to be taken into consideration in determining the extent of the problem.

In view of this, two different estimates of the prevalence of juvenile arthritis in Australia are provided. In addition to reporting prevalence in children, some insight to the extent of juvenile arthritis in the population as a whole is given.

Disease prevalence in children

An estimated 7,900 Australian children (aged 0–15 years), based on the 2004–05 NHS, reportedly ever had arthritis, a point prevalence rate of 173 per 100,000 children. There were more girls than boys (199 compared with 148 per 100,000; a sex ratio of 1.3:1.0; f:m). The point prevalence exceeded 400 per 100,000 among those aged 10–15 years.

Of these, an estimated 4,600 children had arthritis at the time of survey, a point prevalence of 99 per 100,000, or 1 in 1,000, children. Interestingly, the sex ratio among those with current arthritis was highly deviant (9:1; f:m). Most of the reported cases were aged 10–15 years, with a smaller proportion in the age range 5–9 years (Figure 3.1). It must be noted here that not all children reporting arthritis at the time of survey were told by their doctor that they had the condition. Only one-half of those reporting current arthritis had their disease diagnosed by a doctor. Besides, low sample size may have caused deviations in the sex ratios.



Note: Based on information reported by a parent or carer.

Source: AIHW analysis of the 2004–05 ABS National Health Survey CURF.

Figure 3.1: Prevalence of arthritis in Australian children, 2004–05

These estimates of the prevalence of juvenile arthritis are based on interviews, not on diagnostic tests or medical examinations, with a parent or carer reporting whether the child has had arthritis. The results broadly represent their understanding or recall of the disease. It is well recognised that respondents are more likely to report conditions that have had some impact on a child's activities. In the NHS, the information collected about the disease is supplemented by the question if it was ever diagnosed by a doctor.

The considerable difference in sex ratios between the two sets of estimates in Figure 3.1 (ever had arthritis and current arthritis) would suggest that a large proportion of past cases may not be genuine cases of arthritis. A large proportion of cases reported among boys may have resulted from injury. The issue of recall bias, particularly for past medical problems, also needs to be taken into consideration.

International comparisons

The reported prevalence of juvenile arthritis varies considerably worldwide. There are an estimated 300,000 children living with arthritis in the United States of America (Arthritis Foundation 2007). On the other hand, no cases of juvenile arthritis were found in a survey of 20,000 Chinese children living in British Columbia, Canada (Manners & Bower 2002).

Prevalence rates of between 121 and 220 per 100,000 children have been reported in various population-based studies from around the world. An overall figure of 132 per 100,000 children was arrived at in a meta-analysis including practitioner-based and clinic-based studies (Oen & Cheang 1996). The range of estimated prevalence of juvenile arthritis is much smaller in some European countries (Norway and France), between 10 and 80 per 100,000 children (Prieur et al. 1987; Moe & Rygg 1998).

Table 3.2: Worldwide prevalence of juvenile arthritis

Disease/ condition	Country/ place of study	Study type	Prevalence per 100,000 children	Year of study	Source
Juvenile arthritis overall	Germany	Prospective	15		von Koskull et al. 2001
Treated chronic arthritis	Berlin, Germany	Retrospective	20		Kiessling et al. 1998
Juvenile arthritis overall	North America	Population-based	94	1980	Peterson et al. 1996
		Population-based	86	1990	Peterson et al. 1996
		Population-based	121		Oen & Cheang 1996
		Practitioner- and clinic-based	132		Oen & Cheang 1996

Overall prevalence

The overall prevalence of juvenile arthritis in Australia can be estimated using the age of disease onset information. In the 2004–05 NHS, the survey respondents reporting arthritis were asked about the age at which their disease was diagnosed. This historical information was sought not only from parents or carers of children with arthritis but also obtained from adult respondents with arthritis. Similar information is available from the 2003 SDAC.

The overall prevalence of juvenile arthritis in Australia was estimated to be about 103 per 100,000 persons. This corresponds to an estimated total of about 22,000 persons with juvenile arthritis in Australia. The prevalence in females was more than twice the rate in males.

Activity limitation

Activity limitation is a useful indicator of the impact of arthritis on a child's health and functional status. Arthritic pain restricts a child's ability to participate in school activities as well as in other activities of daily living. In younger children, it adds to the caretaking demands of the parents.

In assessing the role of arthritis in limiting activities, it must be recognised that in most of the children the disease may not have run its full course to the extent of causing severe/profound disability. Arthritis is therefore not a major cause of childhood disability. Of 318,000 Australian children with disability (ages 0–14 years) in 2003, only 1,600 are estimated to have arthritis and related disorders as their main disabling condition (Table 3.3). This is in line with the general observation that arthritis-related disability takes much longer to develop.

Table 3.3: Juvenile arthritis as a disabling condition, 2003

	Children with disability (arthritis as the main disabling condition)		Children with disability (conditions other than arthritis)	
Disability status	**Number '000**	**Per cent**	**Number '000**	**Per cent**
Profound	0.3	17.0	77.7	35.5
Severe	1.3	83.0	86.3	39.4
Moderate	0.0	0.0	12.8	5.8
Mild	0.0	0.0	42.2	19.3

Source: AIHW analysis of ABS 2003 Survey of Disability, Ageing and Carers CURF.

On the other hand, in some forms of juvenile arthritis (in particular RF +ve polyarticular arthritis), the disease may cause severe activity limitations soon after onset. The distribution of arthritis-associated disability in children as being either severe or profound in nature (Table 3.3) in comparison with disability caused by other conditions supports that line of argument.

Adverse outcomes for child development and wellbeing associated with arthritis in children may also have long-term detrimental effects on social and economic status in adulthood.

Health care service use

Health care service use is another indirect measure of the extent of the problem. Most of the children with juvenile arthritis recover from the disease early and may not need to use health services long term. However, in a small proportion of cases it is difficult to treat and manage the disease effectively without some medical intervention.

The management of juvenile arthritis is usually about pain relief and preserving joint function. Medicines are central to this disease management strategy. For painful, eroded joints, surgical operation is indicated. Children on a variety of drugs need to be monitored closely for any side effects. There is also an increased risk of complications over time with certain forms of juvenile arthritis. Regular follow-up and contact with a variety of specialists is therefore important. Managing this whole gamut of problems in fact requires a multi-therapist approach.

While the first point of contact is a general practitioner, the disease needs to be primarily managed by a paediatric rheumatologist with support from physical and occupational therapists, social workers and nurse specialists. Their efforts can be further coordinated with other specialists such as ophthalmologists or orthopaedic surgeons. In some cases, hospitalisation may be required. To ensure that the child settles well into the school environment and other community settings, the support of other health professionals such as psychologists, school counsellors and various other community resources may also be called for.

Quantification of the role of all these professionals from the available data sources is rather difficult. Nonetheless, an attempt is made below to put the need for, and use, of health care services in perspective.

Medical consultations

The 2004–05 NHS reveals that the main point of contact for children with arthritis, in almost 46% of cases, was a general practitioner. Visits to day clinics are also common (Table 3.4). Around 18% consulted a specialist or a dentist. Several other health professionals were also consulted. These consultations however, may not be arthritis specific.

Table 3.4: Medical consultations for children with arthritis, 2004–05

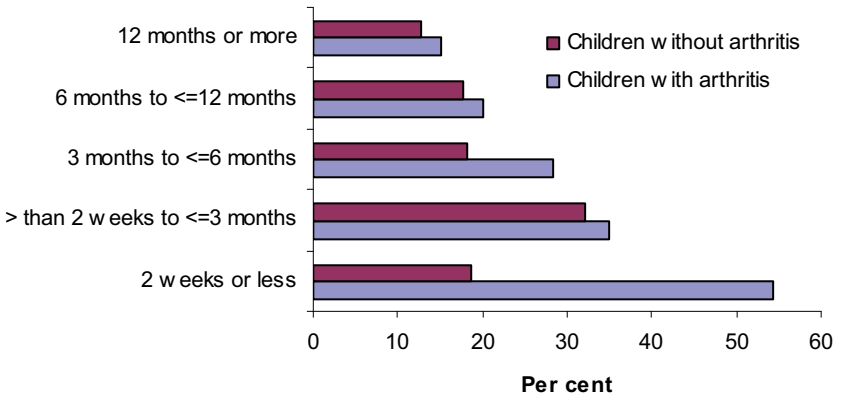
	Children with arthritis		Children without arthritis	
Type of consultation	**Number '000**	**Per cent**	**Number '000**	**Per cent**
Discharged from hospital inpatient episode	0.0	0.0	35.9	1.2
Visited casualty/emergency	0.0	0.0	30.4	1.0
Visited outpatients	0.0	0.0	85.8	2.8
Visited day clinic	0.9	19.6	42.5	1.4
Consulted general practitioner	2.1	45.7	462.6	15.0
Consulted specialist	0.4	8.7	187.3	6.1
Consulted dentist	0.4	8.7	295.9	9.6
Consulted other health professional	1.1	23.9	513.6	16.6

Notes

1. The children (or their parents and carers) were asked about any actions taken in relation to their disease in the 2 weeks before the survey. Actions taken during 6 months and 12 months before the survey were also recorded.
2. Percentages do not add up to 100 as more than one action may have been taken.
3. Other health professionals include accredited counsellors, chemists, nurses, physiotherapists etc.

Source: AIHW analysis of the ABS 2004–05 National Health Survey CURF.

Children with arthritis needed to consult a doctor (either a general practitioner or a specialist) more often than those without arthritis. The frequency of visiting a doctor varied, with about 30% visiting at least once and another 13% twice. Most of these visits were within 2 weeks or less of the survey. In around 28% of children a doctor's consultation took place between 3 months to less than 6 months (Figure 3.2).



Source: AIHW analysis of the ABS 2004–05 National Health Survey CURF.

Figure 3.2: Time since last consultation with a doctor by children with arthritis, 2004–05

Hospital use

Hospitalisation for juvenile arthritis is not common. Table 3.4 shows that in the 2 weeks before their NHS interview, there was no inpatient episodes reported for children with arthritis. There were no reports of visits to a casualty/emergency department either. On the other hand, a large proportion of children with arthritis visited day clinics.

A large proportion of hospitalisations for arthritis are for procedures to relieve pain and improve function. Surgery has a limited but important role in the management of arthritis in younger children (Glueck & Gellman 2005). However, in the older child, surgical approaches to joint contractures, dislocations or joint replacement become more significant.

The number of children with arthritis visiting a hospital could not be ascertained reliably from the NHS data because of the low prevalence of the disease. However, this information can be supplemented from the AIHW National Hospital Morbidity Database.

In 2004–05, juvenile arthritis was the principal diagnosis in 586 hospitalisations, with an average length of stay of 1.2 days. The hospital separation rate was higher in 10–16 year olds, accounting for almost half of all hospitalisations. Girls were hospitalised more often than boys. A total of 1,690 procedures were performed on these children. Over half of the procedures were related to the musculoskeletal system (56%). Non-invasive procedures, cognitive and other interventions accounted for about 40% of the procedures.

Other health professionals

Allied health professionals, in particular physiotherapists, are integral to the management of juvenile arthritis. The objectives of using these services are to minimise pain and maintain as well as restore function.

Based on the 2004–05 NHS, about 23% of children with juvenile arthritis had consulted an allied or other health professional within the previous 2 weeks of the survey. The professionals most frequently consulted were nurses, chemists, and optometrists. Other professionals mostly consulted were accredited counsellors, social workers/welfare officers, physiotherapists/hydrotherapists, all accounting for about 9% of consultations (Table 3.5).

Table 3.5: Allied and other health professional consultations by children with arthritis, 2004–05

Health professional	Children with arthritis		Children without arthritis	
	Number '000	Per cent	Number '000	Per cent
Accredited counsellor	0.4	9.2	62.9	2.0
Chemist	1.0	22.5	152.9	4.9
Acupuncturist	0.0	0.0	0.0	0.0
Chiropodist/podiatrist	0.0	0.0	7.6	0.2
Chiropractor	0.0	0.0	58.9	1.9
Dietitian/nutritionist	0.0	0.0	3.3	0.1
Naturopath	0.0	0.0	16.1	0.5
Nurse	1.2	26.0	62.3	2.0
Optician/optometrist	0.9	19.5	23.8	0.8
Physiotherapist/hydrotherapist	0.4	9.2	33.9	1.1
Psychologist	0.0	0.0	42.9	1.4
Social worker/welfare officer	0.4	9.2	37.5	1.2
Speech therapist/pathologist	0.0	0.0	110.7	3.6
Other	0.4	9.2	45.7	1.5
Did not consult	3.5	76.1	2576.1	83.3

Notes

1. The children (or their parents and carers) were asked about any actions taken in relation to their disease in the 2 weeks before their interview. Actions taken during 6 months and 12 months before the survey were also recorded.
2. Percentages do not add up to 100 as more than one action may have been taken.
3. Other health professionals include Aboriginal health worker, alcohol/drug worker, audiologist, herbalist, hypnotherapist, occupational therapist, osteopath and traditional healer.

Source: AIHW analysis of the ABS 2004–05 National Health Survey CURF.

Successful management of juvenile arthritis includes considering a variety of associated issues. For example, ensuring normal growth and psychosocial development of the child is important. Similarly, there is a need to accommodate everyday tasks and school activities. The skeletal immaturity further adds to the child's needs. All these issues make the use of health care services much more intensive.

Mortality

Juvenile arthritis is an uncommon primary cause of death. When it is listed as a cause of death, it is mostly listed as an associated cause of death for adults who had had juvenile arthritis. This signifies that the arthritis was considered to have been a contributory cause, although not the main one.

Over the 10-year period 1996–2005, juvenile arthritis was listed as an underlying cause for only six deaths. In 10 other deaths, it was listed as an associated cause of death.

Juvenile arthritis contributes to mortality in a variety of ways, and estimating that contribution requires sophisticated modelling. The Australian Burden of Disease study, for example, uses an increased risk of mortality of 1.6 in its modelling (Begg et al. 2007). A significant proportion of children with pauciarticular juvenile arthritis (about 3%) die of kidney failure due to amyloidosis, or following overwhelming infection.

4 Impact on growth, functioning and quality of life

Living with juvenile arthritis is challenging both for children and their parents. The pervasive nature of its symptoms, especially pain, has a significant impact on a child's health and functional status. The impact can be early on, happening in the initial phases, or it can be long-term, persisting into adulthood or emerging later in life. Not all children are affected in the same way. The overall outcome depends on the age at onset, form and course of the disease, type and number of joints involved, treatment received and drug side effects.

Due to the generally self-limiting nature of the disease, a large proportion of children with juvenile arthritis (around 60%) reach adulthood without much functional impairment (Packham & Hall 2002a). In several cases, however, the disease has considerable harmful effects. Growth disturbances, persistent pain, poor aerobic function and impaired exercise tolerance are common. There are also difficulties with activities of daily living. Physical disability affecting work, social and sexual life may continue into adulthood. Long-term outcomes include joint deformity and destruction, postural abnormalities, muscle atrophy and weakness, and psychological issues. The earlier the onset of arthritis, the sooner these problems may arise.

This chapter provides a statistical overview of children's experience of arthritis; its impact on their functioning, including activities of daily life; psychosocial health; and their growth and development. The chapter also describes outcomes later in life such as morbidity, disability and psychosocial health. Its impact on family members is also reviewed.

The experience of arthritis

Arthritis can have a significant impact on the daily life of a child. Pain, stiffness, limited joint motion and endurance arrive early. While remission is common, the prognosis can be poor.

In early phases of the disease, the illness limits participation in certain activities only. However, with time the impact may turn into a severe or profound disability. The experience of arthritis described below is mostly based on information about the severe end of the spectrum, given the lack of suitable empirical data about milder forms/earlier stages of the disease.

Pain

Pain is a key expression of arthritis. It generally tends to be mild to moderate but causes much discomfort and limits many types of activities. Children usually describe the pain of arthritis as hurting, stinging, warm and uncomfortable. The experience varies greatly from joint to joint, not only daily but even hourly (Box 4.1). The source of pain in juvenile arthritis is mostly inflammatory in nature.

Sherry et al. (1990) point out that almost 86% of children with arthritis report pain at routine clinic visits. A study by Schanberg and colleagues (2003) noticed that over a 2-month period children reported pain on average on 73% of days, and 76% of children reported having pain on all of the days. A large majority of children described the intensity of their pain as mild to

moderate, but almost one-third (31%) reported it as severe. The pain reportedly lasted from 30 minutes to 24 hours during any one day.

Box 4.1: Pain in children with juvenile arthritis

Source

- *Inflammation of synovial membrane, tendons or ligaments*
- *Weakening of muscles and other soft tissues around the joint*
- *Fatigue*
- *Combination of the above*

Variation

- *From joint to joint*
- *Degree of swelling*
- *Extent of heat or redness*
- *Damage within the joint*

Timing

- *Morning (stiffness)*

Characterising the experience of pain in children is difficult. Older children, especially those diagnosed recently, report more pain than their younger counterparts. Besides, children with long-standing disease report less pain. The perception of pain therefore often reflects a child's adjustment to the disease rather than the extent of pain itself.

Limited information is available about the experience of pain in Australian children. The ABS Survey of Disability, Ageing and Carers (SDAC) collects information about pain through the screening question: 'Whether has chronic pain or discomfort, limiting activities'. The rider 'limiting activities' restricts the scope of the information generated.

Based on the 2003 SDAC, about 11,300 children aged 0–14 years had chronic or recurrent pain and were limited in activities. Arthritis was the largest contributor for this pain in 1,555 children. At the more severe end of the spectrum, all children with arthritis as their main disabling condition that limited activities reported chronic/recurrent pain.

Physical limitations

Children with arthritis often have physical limitations. In addition to joint erosions, long periods of active arthritis may impair muscle development. Inability to move freely and difficulty in gripping are some of the common outcomes. Gait deviations, including decreased velocity, cadence, and step and stride length, may also result. Due to lack of physical activity, children with arthritis may become quite unfit.

The SDAC collects information about some of these physical limitations. The information provided below is limited to children with arthritis who have profound or severe activity restrictions to qualify for the designation of disability. There are likely to be lesser physical limitations in children with more recent onset of disease. The most common limitation reported in the 2003 SDAC was restricted physical activity or inability to do physical work (Table 4.1). One out of every two children with arthritis reported limited use of their arms

and fingers and difficulty gripping or holding things. Many children with disability due to arthritis had lower body limitations such as limited use of their feet and legs. These impacts compare unfavourably with disability caused by other medical conditions.

Table 4.1: Physical limitations in children with disability, 2003

	Main disabling condition			
	Arthritis		Other	
Limitation	Number '000	Per cent	Number '000	Per cent
Limited or restricted in physical activities or in doing physical work	1.1	68.7	31.3	0.8
Has difficulty gripping or holding things	0.8	50.0	21.9	0.6
Has limited use of arms or fingers	0.8	50.0	10.5	0.3
Has limited use of feet and legs	0.6	37.5	14.2	0.4
Has a disfigurement or deformity	0.0	0.0	17.7	0.5

Notes

1. The proportions are based on the estimated number of children with disability due to arthritis and related disorders as the main disabling condition (N= 1,555) and those with other main disabling conditions (N= 3,849,094).
2. Children aged 0–14 years.
3. Percentages do not add up to 100 as more than one limitation may have been reported.

Source: AIHW analysis of ABS 2003 Survey of Disability, Ageing and Carers CURF.

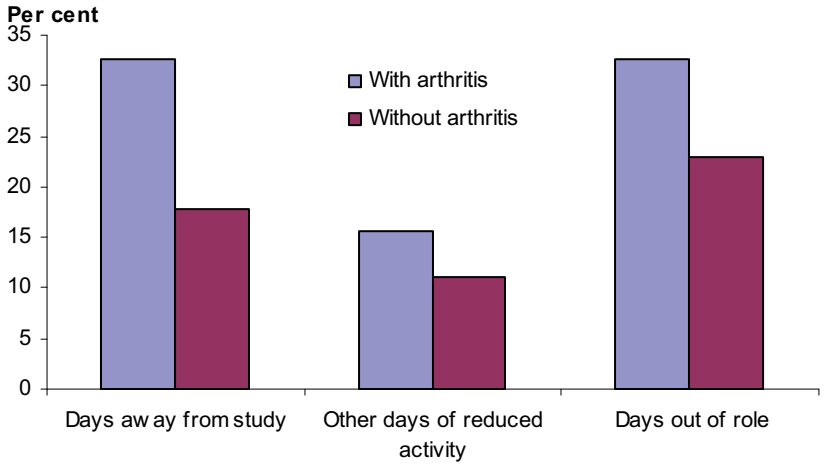
Uveitis, inflammation inside the eye that causes reduced vision, is another source of physical limitation. Lung function abnormalities due to respiratory muscle weakness are also a cause of activity limitation in some children with arthritis (Knook et al. 1999).

Several of these limitations worsen in adulthood, leading to physical impairments. These poor outcomes stress the need to manage the problems early in life. In addition to maintaining joint function and preservation, there is a need for muscle strengthening and aerobic exercise. Weight bearing and continuing to walk are important for satisfactory bone growth and density, joint stability and muscle development. In affected children physical activity such as standing and walking should also be encouraged at the expected age.

Reduced activity

Arthritis often puts children out of their daily routine (out of role). Children with arthritis are more likely to have days of reduced activity, including being away from school. The varying degrees of pain and stiffness can affect a child's mobility, strength and endurance. For example, children may find it difficult to sit on the floor or hold pencils, crayons or paint brushes, or may have a problem carrying books or opening lunch boxes.

Based on 2004–05 NHS, Australian children with arthritis are almost twice as likely to have days away from study than other children (Figure 4.1). Out of school, they are also more likely to have days of reduced activity.



Notes

1. Days away from study refers to days on which the respondent was away from school for at least half the day due to own illness or injury.
2. Other days of reduced activity refers to other days on which activity was reduced for more than half the day due to own illness or injury.
3. Days out of role refer to the combination of days away from school or study or other days on which activity was reduced for more than half the day.
4. Children aged 0–15 years.

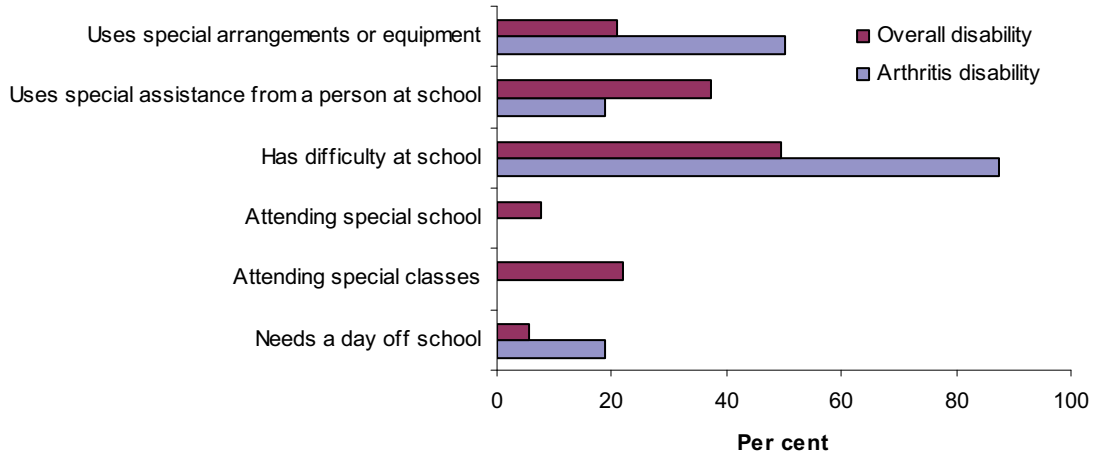
Source: AIHW analysis of the ABS 2004–05 National Health Survey CURF.

Figure 4.1: Reduced activity among children, 2004–05

Activity restrictions at school

Older children with arthritis experience several activity restrictions at school. Many activity restrictions are likely to be first identified at school or be obvious or disabling in the school situation. Children with disability associated with arthritis are likely to experience these restrictions more often (94%) than those with disability in general (82%), although a vast majority report moderate or mild schooling restrictions.

A large proportion of children with arthritis disability (85%) report limitations in participating fully in school activities compared with 50% of those with disability in general. About one-half use special arrangements or equipment. One out of four needs a day off or uses special assistance from a person at school (Figure 4.2).



Note: Children aged 5–14 years.

Source: AIHW analysis of ABS 2003 Survey of Disability, Ageing and Carers CURF.

Figure 4.2: Schooling restrictions experienced by children with arthritis-associated disability, 2003

Sports participation, fitting in socially and problems in sitting are the three most commonly reported difficulties in children with arthritis disability (Table 4.2). This contrasts with difficulties generally reported by children with disability at school which include, in particular, communication and learning difficulties.

Table 4.2: Difficulties experienced at school by children with disability, 2003

	Arthritis disability		Overall disability	
Difficulty	**Number '000**	**Per cent**	**Number '000**	**Per cent**
Difficulty in sitting	0.8	57.1	20.6	13.1
Hearing or sight problems	0.0	0.0	12.8	8.1
Communication difficulties	0.0	0.0	57.7	36.6
Learning difficulties	0.0	0.0	110.0	69.8
Intellectual difficulties	0.0	0.0	25.5	16.2
Fitting in socially	1.2	85.7	76.6	48.6
Sports participation	1.4	100.0	32.2	20.4
Other, including access difficulties	0.0	0.0	15.8	10.0

Notes

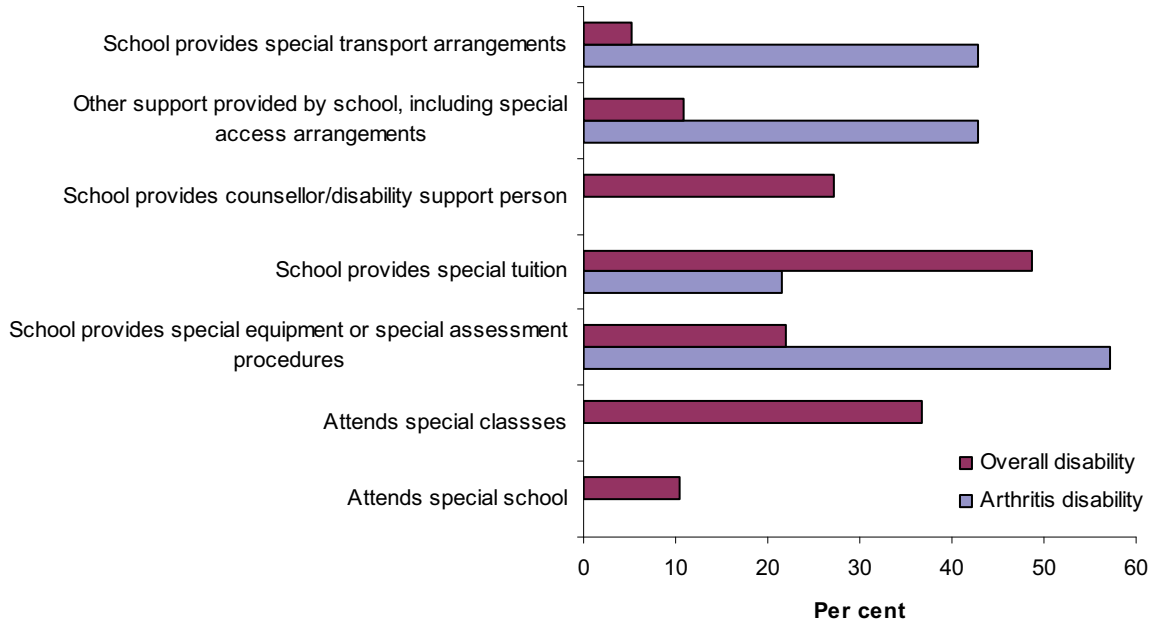
1. Children aged 5–14 years.
2. More than one difficulty may be reported.

Source: AIHW analysis of ABS 2003 Survey of Disability, Ageing and Carers CURF.

However, many of the difficulties experienced by children can be reduced by effective support from school. According to the 2003 SDAC, a large proportion of children receive

some sort of special support from school (74% with disability in general and 59% with disability associated with arthritis).

Those with disability associated with arthritis receive support in areas of access and mobility such as special equipment or special assessment procedures, special access arrangements and special transport arrangements (Figure 4.3). In comparison, children with disability in general receive support for learning and communication. This is to be expected, as a greater proportion of children with general disability have learning and communication difficulty.



Note: Children aged 5–14 years.

Source: AIHW analysis of ABS 2003 Survey of Disability, Ageing and Carers CURF.

Figure 4.3: Special support received by children with disability at school, 2003

Psychosocial impact

Chronic pain, physical limitations, inability to undertake certain activities, health perception and body image affect the psychology of children. Indeed, pain, stiffness and fatigue are significant predictors of impaired psychological wellbeing, causing increased functional disability, anxiety, and reducing participation in school and social activities (Schanberg et al. 2003).

Contributing factors for psychosocial problems include age of disease onset, duration of illness and disability, repeated hospital admissions and physical deformity (David et al. 1994). Isolation, inadequacy and insecurity further add to the mix (Packham 2004). Restrictions imposed by the disease or embarrassment about having the disease fuel the anger of the child. Most want to be like their peers and may resent other children, including their own siblings, who do not have the disease.

Social isolation

Children with juvenile arthritis can become socially isolated or experience social difficulties because they are less likely to be able to participate in a variety of physical activities including play and sports. Alteration in physical appearance (such as swollen or deformed joints) may also attract negative reactions from other children, preventing the affected child from participation in peer group activities.

School performance

The greatest social impact of arthritis is likely to be on a child's school performance. Some children may find it difficult to participate in regular school activities. Tiredness can be a problem for some children due to sleep deprivation from the pain. Others may have side effects from their medication (for example, nausea and vomiting from anti-inflammatory drugs) or from medical procedures (Lovell et al. 1990). The nature of the condition may cause irregular absences from the school.

According to the 2003 SDAC, all children aged 5–14 years with disability due to arthritis attended school. This contrasts with children with disability due to other conditions among which almost one out of five children was not attending school at the time of the survey (Table 4.3).

Table 4.3: School attendance by children with disability, 2003

	Arthritis disability		Any disability		No disability		All children	
Measure	Number '000	Per cent	Number '000	Per cent	Number '000	Per cent	Number '000	Per cent
At school								
Attending school	1.6	100.0	256.1	80.6	2274.5	96.4	2530.6	96.5
Not attending school	0.0	0.0	53.7	16.9	84.7	3.6	92.7	3.5
Type of school								
Primary	1.0	62.8	193.2	75.4	1711.6	75.3	1904.7	75.3
Secondary	0.6	37.2	62.9	24.6	562.9	24.7	625.9	24.7
Reason for not attending school								
Condition prevents school attendance	0.0	0.0	2.1	28.3	0.0	0.0	2.1	28.3
Too young	0.0	0.0	5.3	71.7	0.0	0.0	5.3	71.7

Note: Children aged 5–14 years.

Source: AIHW analysis of ABS 2003 Survey of Disability, Ageing and Carers CURF.

Body image

The disease process and medication regime of juvenile arthritis can alter physical appearance. Some children may have small jaw bones, or micrognathia, which can profoundly change the look of the face. In some cases shortening of fingers, hands, forearms, toes or feet may occur. Some children may feel embarrassed by the weight gain and skin changes due to drug therapy such as oral corticosteroids. Other cosmetic changes may be

surgical scars, awkward movements such as limping, or the necessity to wear splints or braces. Children with arthritis often develop gait impairments, including decreased gait velocity, arrhythmic movement, and unevenness in step and stride length. They may also develop an increased pelvic tilt and decreased hip extension.

These changes may affect the body image of the child. No Australian data are currently available to provide information about these issues in relation to juvenile arthritis.

Growth and development

Children with arthritis suffer from a variety of growth disorders, ranging from general growth retardation to local acceleration of growth in the affected limb (Packham & Hall 2002a; MacRae et al. 2006). Puberty and appearance of secondary sex characteristics may be delayed (Fraser et al. 1988).

Retardation of linear growth is associated with extended periods of active disease, exacerbated by long-term use of systemic steroids. Consequently, short stature is common among those with a history of juvenile arthritis.

Leg-length discrepancy is common. Unilateral knee arthritis may result in overgrowth of the distal femur, as increased blood supply to the inflamed joint causes accelerated growth of the ossification centres. Micrognathia or undergrowth of the mandible may also result from temporomandibular arthritis.

The disease process, medication and lack of physical activity often contribute to inadequate bone formation, low bone turnover and depressed bone formation (McDonagh 2001). Children with arthritis are more likely to develop osteopenia or bone loss. Those with severe arthritis are at an increased risk of bone fracture.

Structural scolioses are also associated with juvenile arthritis (Ross et al. 1987). These may arise from postural curves associated with asymmetrical involvement of a lower limb joint, which in turn retards bone growth.

The long-term impact of juvenile arthritis varies by disease subtype; those with the polyarticular and systemic subtypes are the most affected.

Long-term outcomes

Juvenile arthritis often continues into adulthood. Between 10% and 20% enter adulthood with active disease, some with moderate to severe functional disabilities. The disease also recurs in adulthood.

Poor outcomes of juvenile arthritis fester up in adult years. With the passage of time (and active arthritis), a variety of physical impairments and health problems emerge. Some of these problems are listed in Box 4.2.

Box 4.2: Health problems in adults with a history of juvenile arthritis

General growth

- *Leg-length discrepancy*
- *Structural growth*

Facial appearance

- *Micrognathia (small mandible, and overbite and poor mouth opening)*
- *Restricted temporomandibular joint damage (TMJ – mouth opening less than 3 cm)*

Eye problems

- *Uveitis*
- *Glaucoma*
- *Cataract*
- *Dry eyes*

Note: Study sample: 246 adults, 19 years or over with juvenile arthritis.

Source: Packham & Hall 2002a.

Early onset of disability and increased need for surgery is common in persons with juvenile arthritis. Many experience considerable functional limitations after 10 to 20 years of active disease (Cassidy & Petty 1995; Zak & Pedersen 2000). Almost 20% of adults—with average disease duration of 20 years—suffer from depression (David et al. 1994). Anxiety is also common—Packham and colleagues (2002b) suggest anxiety to be significantly more prevalent than depression in adults with juvenile arthritis.

Australian data

No follow-up studies of juvenile arthritis have been undertaken in Australia. However, health and functioning profiles of adults with a history of juvenile arthritis can be generated from various population health surveys. We use information from two of these surveys, namely the National Health Survey (NHS) and Survey of Disability, Ageing and Carers (SDAC) to generate one such profile (see Box 4.3).

The impact of juvenile arthritis can be seen in a variety of physical and psychological health domains. Based on data from the NHS and the SDAC, three different domains of health may be viewed:

- health status
- psychological distress
- functional limitations.

Box 4.3: Data sources for monitoring long-term impact of juvenile-onset arthritis in Australian adults

National Health Survey

Data from the 2004–05 NHS has been used in this report to draw together information about the long-term consequences of juvenile arthritis. The survey collects information about the age of onset of the disease. In conjunction with information on the current age of the respondent, a profile of the long-term consequences of the disease in the Australian population can be generated. It also offers the opportunity to compare this profile with that of respondents with adult-onset arthritis and those without arthritis.

Based on the 2004–05 NHS, an estimated 32,000 Australian adults aged 16 years and over developed arthritis in childhood (age of onset: 1 to 15 years). Almost 70% of these were females. This ratio was much higher than those noted among respondents with adult-onset arthritis (1.5) and those without arthritis (1.1).

Survey of Disability, Ageing and Carers

Similar information about the long-term impact of juvenile arthritis on functioning and disability may be generated from the ABS Survey of Disability, Ageing and Carers (SDAC).

In addition to providing a self-assessed overview of physical health and psychological wellbeing, information is provided on the impact of juvenile arthritis on functioning and disability, employment and social life in adulthood. Where possible, comparisons are made with persons with onset of arthritis in adulthood and those reporting no arthritis from the same data sources.

Self-assessed health status is a powerful predictor of overall health outcomes. The other domains provide insight into the impact that juvenile arthritis has on day-to-day life—on the ability to function in society and to do the things they want to do.

Self-assessed health status

Adults with juvenile-onset arthritis are less likely to report their health as 'excellent/very good' than those with adult-onset arthritis. Although slightly more adults with juvenile arthritis rate their health to be 'good' and 'fair', the proportion of those rating their health as 'poor' is lower (Table 4.4).

Table 4.4: Self-assessed health status of adults with long-term arthritis, 2004–05

	Juvenile-onset arthritis		Adult-onset arthritis		No arthritis	
Health status	Number '000	Per cent	Number '000	Per cent	Number '000	Per cent
Excellent/very good	9.1	28.3	1,018.1	34.1	1,830.8	40.7
Good	12.4	38.4	927.2	31.2	1,348.8	29.9
Fair	8.5	26.5	675.3	15.9	871.9	19.4
Poor	2.2	6.8	362.8	12.1	51.0	10.0

Note: Adults aged 16 years or over.

Source: AIHW analysis of the ABS 2004–05 National Health Survey CURF.

More persons with adult-onset arthritis consider their health to be 'poor' compared with those with juvenile-onset arthritis. This may be due to the presence of several highly debilitating forms of arthritis in adulthood, such as rheumatoid arthritis, where functional limitations arrive soon after disease onset and worsen quickly, resulting in loss of independence.

In both groups, the proportion of respondents reporting 'poor' health increases with age, which may be taken as a proxy for years of active disease. However, this response may be confounded by a variety of other factors.

Psychological health

Based on information collected during the 2004–05 NHS, adults with juvenile-onset arthritis are more likely than those with adult-onset arthritis to report 'very high' and 'high' levels of psychological distress (Table 4.5). This determination of psychological wellbeing is based on the Kessler Psychological Distress Scale 10 (K10), which measures non-specific psychological distress. A 'very high' level of psychological distress, as measured by K10, may indicate a need for professional help.

Table 4.5: Psychological distress in adults with arthritis, 2004–05

	Juvenile-onset arthritis (per cent)				Adult-onset arthritis (per cent)			
Level of distress	16–34 years	35–64 years	65 or over	All ages	16–34 years	35–64 years	65 or over	All ages
Low (10–15)	47.4	48.7	43.3	47.0	58.3	66.7	75.3	64.6
Moderate (16–21)	23.1	30.6	29.2	26.4	28.7	21.8	17.1	23.8
High (22–29)	20.7	12.6	27.5	19.7	10.0	7.8	5.6	8.4
Very high (30–50)	8.8	8.1	0.0	6.9	3.0	3.7	2.0	3.2

Note: Adults aged 16 years or over.

Source: AIHW analysis of the ABS 2004–05 National Health Survey CURF.

A high degree of psychological distress is noted more commonly in the younger age groups (ages 16–34 and 35–64 years). Those aged 65 years and over appear more likely to take the disease in their stride.

Functional limitations

As described earlier, between 10% and 20% of children with arthritis enter adulthood with moderate to severe functional limitations. Many others develop these limitations later in life.

Over time, these limitations progress towards defined disability. An estimated 25,000 Australian adults had disability associated with juvenile-onset arthritis in 2003 (Table 4.6). Those in the age group 45–64 years constituted more than one-half of those affected.

The age and sex profile of juvenile arthritis-associated disability is considerably different from that of adult-onset arthritis-associated disability. The early arrival of disability among those with juvenile-onset arthritis reflects generally the relatively longer duration of illness. Females outnumber males (5:1) in this segment of the population (Table 4.6).

Table 4.6: A demographic profile of adults with arthritis-associated disability, 2003

	Adults with juvenile arthritis		Adult-onset arthritis	
Demographic characteristic	**Number '000**	**Per cent**	**Number '000**	**Per cent**
Gender				
Males	4.5	18.4	163.8	30.7
Females	20.3	81.6	369.8	69.3
Total	*24.8*	*100.0*	*533.6*	*100.0*
Age group (in years)				
15–29	1.7	7.0	8.5	1.6
30–44	3.9	15.8	27.4	5.1
45–64	13.1	52.7	211.2	39.6
65 and over	6.1	24.5	286.5	53.7
Total	*24.8*	*100.0*	*533.6*	*100.0*

Note: Adults aged 15 years or over; with arthritis as the main disabling condition.

Source: AIHW analysis of ABS 2003 Survey of Disability, Ageing and Carers CURF.

Table 4.7 details physical impairments reported by adults with arthritis. The impairments are more pronounced in adults with juvenile-onset arthritis compared to those with adult-onset arthritis. Except for limitations or restrictions in physical activities, the proportion of persons with impairments is larger in the juvenile-onset arthritis group.

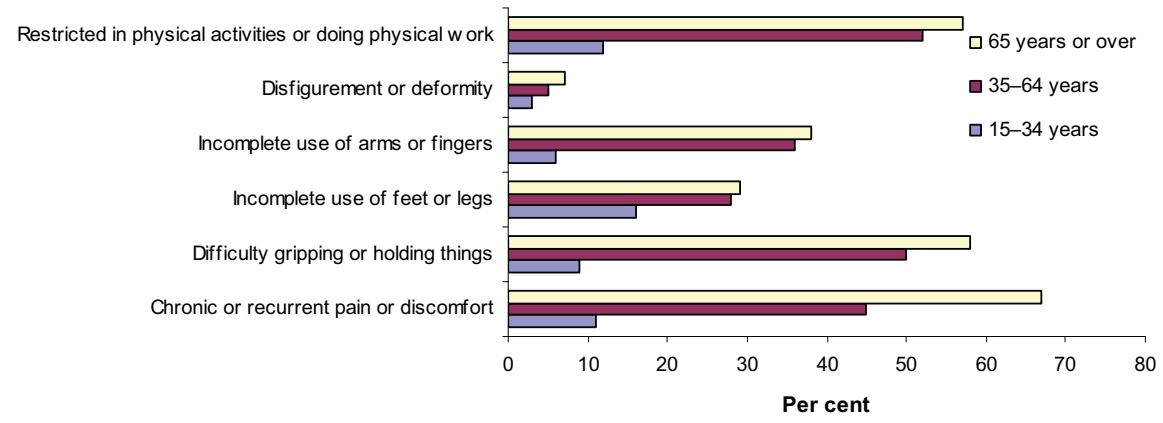
Table 4.7: Physical impairments in adults with arthritis, 2003

	Juvenile-onset arthritis		Adult-onset arthritis	
Physical impairment	**Number '000**	**Per cent**	**Number '000**	**Per cent**
Chronic or recurrent pain or discomfort	15.2	61.3	295.7	55.4
Difficulty gripping or holding things	13.5	54.4	263.2	49.3
Incomplete use of feet or legs	7.1	28.5	129.5	24.3
Incomplete use of arms or fingers	5.5	22.0	88.6	16.8
Disfigurement or deformity	0.8	3.3	15.2	2.9
Limited or restricted in physical activities	13.2	53.3	298.1	55.7

Note: Adults aged 15 years or over, with arthritis as the main disabling condition.

Source: AIHW analysis of ABS 2003 Survey of Disability, Ageing and Carers CURF.

Age remains the strongest predictor of these impairments (Figure 4.4). More than one-half of persons in the age group 35–64 years report chronic or recurrent pain, difficulty holding or gripping things and restriction in physical activities.



Note: Adults aged 15 years or over.

Source: AIHW analysis of ABS 2003 Survey of Disability, Ageing and Carers CURF.

Figure 4.4: Physical impairments in adults with juvenile arthritis, 2003

Unemployment

Juvenile arthritis has a detrimental effect on employment (David et al. 1994; Foster et al. 1999). Despite good academic attainment, adults with a history of juvenile-onset arthritis are more likely to remain unemployed. A large majority of this is attributed directly to disease-related problems (Packham & Hall 2002b).

The 2004–05 NHS confirms the existence of this pattern of disadvantage in Australia (Table 4.8). Much lower employment rates are reported by persons with juvenile-onset arthritis in the working age groups than those with adult-onset arthritis. The gap widens with age.

Table 4.8: Employment status of adults with arthritis, 2004–05

Employment status	Adults with juvenile-onset arthritis (per cent)			Adult-onset arthritis (per cent)		
	16–34 years	35–64 years	16–64 years	16–34 years	35–64 years	16–64 years
Employed, working full-time	35.9	42.2	31.4	51.9	56.3	48.9
Employed working part-time	11.8	17.7	13.9	24.6	20.8	20.8
Unemployed, looking for full-time work	15.6	0.5	8.7	3.5	1.6	2.3
Unemployed, looking for part-time work	13.6	0.0	7.5	1.4	0.5	0.8
Not in the labour force	23.1	39.6	38.5	18.6	20.8	27.2
Total	**100.0**	**100.0**	**100.0**	**100.0**	**100.0**	**100.0**

Note: Adults aged between 16 and 64 years.

Source: AIHW analysis of the ABS 2004–05 National Health Survey CURF.

Social life

Because of functional restrictions, pain and poor body image, social and sexual relationships may be harder to develop and maintain by persons with a history of juvenile-onset arthritis. Young adult males with juvenile-onset arthritis are more likely to live by themselves without dating, report less frequent sexual activity, and are also less likely to be in stable relationships (Ostensen et al. 2000). About 30% attribute their problems to physical disability and body image (Packham & Hall 2002c).

In adult females with juvenile-onset arthritis, fertility is significantly reduced, with an increased rate of miscarriage. A higher proportion of females have had metrorrhagia (uterine bleeding occurring at completely irregular intervals), pelvic inflammatory disease and/or surgery for ovarian cysts.

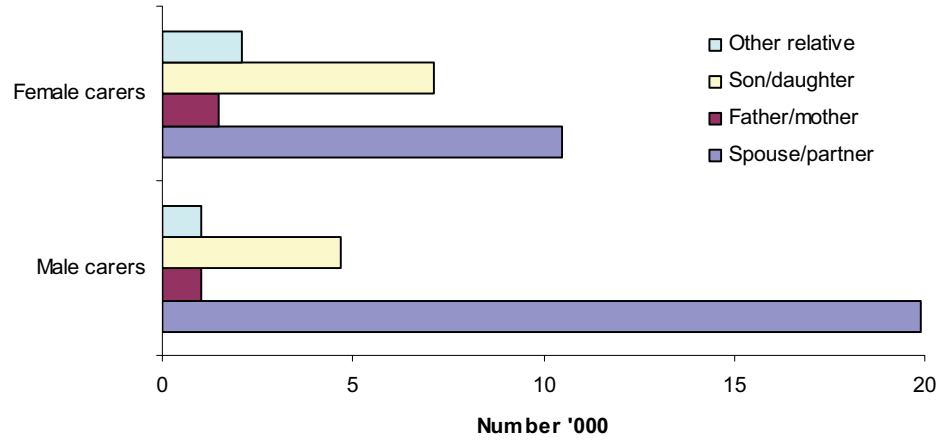
No data are available to report this aspect of juvenile arthritis in Australia.

Juvenile arthritis and family

Families, especially parents, are an important source of support for children with arthritis. The family becomes involved in providing help and care in a variety of ways. There is an ongoing need to manage the child's pain and suffering, help with mobility and activity limitations, and provide regular support and care. Daily treatment, monitoring and dealing with a wide variety of health, social care and education issues require much time and attention.

Some insight into caring for children with juvenile arthritis in Australia can be obtained from the 2003 SDAC. The focus in these cases is usually on primary carers with particular attention to ensuing disability.

The 2003 SDAC indicates that more than 45,000 co-resident primary carers were providing assistance to people with arthritis-associated disability. Although a large proportion of these carers were looking after an adult, a significant proportion was parent carers—looking after a son or daughter with arthritis. In most of these cases, the disability is most likely to result from juvenile arthritis (Figure 4.5).



Note: Arthritis as the main disability.

Source: AIHW analysis of ABS 2003 Survey of Disability, Ageing and Carers CURF.

Figure 4.5: Carers of people with arthritis-associated disability, 2003

Impact on parents

Arthritis in children, especially the younger ones, may require parents to take on more protective, more controlling or more intrusive parenting roles. Parents can be emotionally or physically affected by the strain of dealing with their child's pain and physical limitations.

Over two-thirds of parents of children with arthritis report personal health problems, family conflict and lack of social support (Vandvik et al. 1989). Greater functional impairment among children has also been associated with increased family stress (McCormick et al. 1986), problematic adjustment and parental strain (Jessop et al. 1988; Silver et al. 1995), and poorer concurrent functioning among parents (Timko et al. 1992).

Because of low sample size, information cannot be provided for carers of children with juvenile arthritis in Australia. However, information available on carers of children with disability (Table 4.9) can provide some insight into the effects of caring for children with chronic illnesses. Most of these carers are more likely to be parents.

Based on the 2003 SDAC, a large proportion of carers feel dissatisfied with the nature of caring. A profound sense of helplessness, infused with anger, is noted (Table 4.9). A large proportion of carers report change to their income or financial situation, with their income decreasing (43%). Almost 23% report having difficulty meeting everyday expenses. The caring role, however, may not affect their relationship with co-residents and friends.

Table 4.9: Effects on carers of children with disability, 2003

Type of effect	Number '000	Per cent
Physical or emotional effects		
Feeling dissatisfied	45.5	87.8
Fatigue and weariness	26.1	50.3
Change in physical and emotional wellbeing	21.0	40.5
Feeling of worry and depression	18.3	35.3
Feeling of anger and resentment	9.6	18.5
Main effects on relationship with co-residents and friends		
Relationship unaffected	25.4	49.0
Brought closer together	19.5	37.6
Lost touch with existing friends	14.1	27.2
Circle of friends changed	8.9	17.2
Relationship strained	6.9	13.3
Financial effects		
Income has decreased	22.2	42.9
Income not affected	15.8	30.5
Has difficulty meeting everyday expenses	12.0	23.2

Note: Carers of children aged 0–15 years with disability.

Source: AIHW analysis of ABS 2003 Survey of Disability, Ageing and Carers CURF.

Mothers are affected the most and are more likely to report higher levels of stress and depression as they appear to have greater responsibility for the day-to-day care and medical needs of their ill children (Lustig et al. 1996). Mothers of children with mild disease activity experience more distress compared with those with none or moderate/severe disease activity (Daltroy et al. 1992). They were mainly worried about the child's peer relationships, morale and practical daily challenges, feeling that the effect on family function was more disruptive (Ennett et al. 1991).

The type of medications used by their children may also significantly affect a mother's mental health. Higher levels of psychological problems are noted by mothers whose children were taking non-steroidal anti-inflammatory drugs (NSAIDs) in combination with steroids. This may be because the type of medication is an indicator of disease severity (steroids are prescribed only to those with severe disease). Other reasons may be the daily hassles of medication-giving or other activities that may affect the daily functioning of the family (for example, getting the child to school on time), and potential adverse side effects (for example, mood swings or fluid retention). The mental strain of mothers depends upon the child's functional status (Timko et al. 1992).

Affected fathers also experience many emotions and perceive their child's condition as a catalyst for meaningful involvement (McNeill 2004). The child's visible symptoms, such as short stature, apparent physical weakness or steroid-induced weight gain, affects fathers the most and they are mainly worried about the child coping with school or college, employment and boyfriends or girlfriends.

Impact on siblings

The chronic pain of juvenile arthritis also puts strain on siblings (Britton 2002). They could be concerned about their own health, worrying if they would ever develop the condition themselves. The siblings are also troubled by the relationship with the ill sibling as well as with their parents. Many resent their family not having enough energy and time for play and other shared activities, as parents quite often have to resort to separate activities to accommodate both the well and unwell children. Many siblings feel left out and try to compete for parental attention. As the ill child is emotionally volatile, there is often less communication between siblings. Despite all this hostility among the siblings, a recent study has found that some siblings develop a more compassionate and protective role towards their ill sibling (Britton & Moore 2002).

5 Treatment and management

The central goal of any treatment and management strategy for juvenile arthritis is to restore normal childhood activities, ensure proper growth and development, and reduce joint erosion. There is also the need to ensure that the transition from the paediatric to the adult phase of juvenile arthritis is well managed.

This chapter reviews various strategies used to treat and manage juvenile arthritis, taking into consideration the guidelines developed by the Royal Australian College of General Practitioners (RACGP 2008). The chapter also touches upon the rehabilitation of adults with a history of juvenile arthritis. Due to the lack of relevant Australian data, this chapter is more expository than statistical in nature.

Management strategies

There are no well-recognised management strategies that consistently achieve the best results for juvenile arthritis. Any management strategy needs to be integrated with the dynamics of skeletal growth and development. The approach also needs to take into account the child's varied medical, rehabilitation and psychological needs.

Four major objectives of the management of juvenile arthritis are:

- early diagnosis
- minimise pain
- keep the joints mobile
- prevent deformity.

These objectives are best achieved by a multidisciplinary team, ideally, a paediatric multidisciplinary team. The team should include a paediatric rheumatologist, a nurse, a physical and occupational therapist, a paediatric ophthalmologist, a paediatric orthopaedic surgeon, a paediatric gastroenterologist, a paediatric haematologist and a social worker.

The management team should not only educate the affected child but also the child's parents in understanding the disease process and its potential complications. Family members are therefore an integral part of this team. Parents need to work very closely with the medical personnel involved in the treatment and management of the child's condition.

To ensure a smooth transition from the paediatric to the adult stage of juvenile arthritis, a team approach is again highly recommended in view of the long-term impact of juvenile arthritis on later life. In addition to problems posed by the illness, associated disability, employment difficulties and psychosocial issues need to be aggressively addressed in adults. Rehabilitation is also central to any management and treatment strategy in adults with a history of juvenile arthritis.

General treatment

The first line of management for juvenile arthritis is drug treatment, which plays a major role. The aim is to control inflammation and relieve pain. Most children with juvenile arthritis will require medication at one time or other during the course of their illness. Some of the medications used to treat arthritis in children are described in Box 5.1.

Box 5.1: Medications commonly used for treating juvenile arthritis

Paracetamol

Paracetamol is usually the first line of treatment for pain as well as discomfort. It does not, however, reduce inflammation. Paracetamol relieves pain probably by acting on the brain, but not in other areas. Pain-relieving properties are comparable to aspirin and other non-steroidal anti-inflammatory agents such as Brufen and Voltaren.

Non-steroidal anti-inflammatory drugs (NSAIDs)

NSAIDs can block the activity of enzymes known as Cyclooxygenase (COX enzymes) that play a role in reducing pain and inflammation. Aspirin, ibuprofen and naproxen are some of the common NSAIDs.

Disease-modifying antirheumatic drugs (DMARDs)

This is a group of drugs that have the capacity to alter the course of the disease. Common drugs to treat juvenile arthritis include methotrexate, sulfasalazine, cyclospori, and azathioprine. These drugs do not act in the same way but produce similar effects. DMARDs reduce inflammation and pain and can retard the erosion of bone and facilitate healing. However, they only rarely induce true remission.

Corticosteroids

These synthetic versions of natural hormones reduce both inflammation and the activity of the immune system. Corticosteroids can provide rapid, dramatic improvement in some children with juvenile arthritis who do not respond to other medications, such as NSAIDs or methotrexate. Unfortunately, symptoms often return when therapy is stopped which can induce a strong rebound effect.

Hydroxychloroquine

Hydroxychloroquine fights some kinds of infections but it is also useful in arthritis, helping relieve inflammation, swelling, stiffness and joint pain. It may be given alone or with one or more other medicines.

Biologics

Biological agents are a type of medications used to block the action of a naturally occurring protein in the body that helps cause inflammation. Etanercept (Enbrel) and infliximab are examples, but only Enbrel is approved for use in Australia.

Source: Miller 2006.

The drug treatment of juvenile arthritis often needs to be individualised, as the response to a medicine may vary even with the same subtype. Adjustments have to be made to the type and dose of the medication with changes in the child's condition or if any side effects develop. Depending on the severity of the condition, some children may need to take a combination of medications.

A brief overview of the various drug treatment strategies for juvenile arthritis follows.

Non-steroidal anti-inflammatory drugs

NSAIDs are often used in conjunction with paracetamol as the first line of treatment in juvenile arthritis, mainly to relieve pain, stiffness and inflammation. NSAIDs are mostly useful in children with mild disease. A combination of NSAIDs and injection into the joint space known as intra-articular corticosteroid injection (IAS) has also been found to be effective in children with functional limitations (Ruperto et al. 2004).

Disease-modifying antirheumatic drugs

DMARDs are usually used as the second line of treatment for juvenile arthritis. DMARDs are proven to be most effective in severe cases of juvenile arthritis. The safety and effectiveness of methotrexate has been established by many studies on children with systemic arthritis, oligoarthritis and polyarthritis, but not enthesitis-related arthritis (Woo et al. 2000; Ruperto et al. 2004). Single weekly doses of methotrexate with food, in particular the use of parenteral solution of methotrexate (25 mg/ml) taken orally, has been reported to be most effective (Wallace 2006).

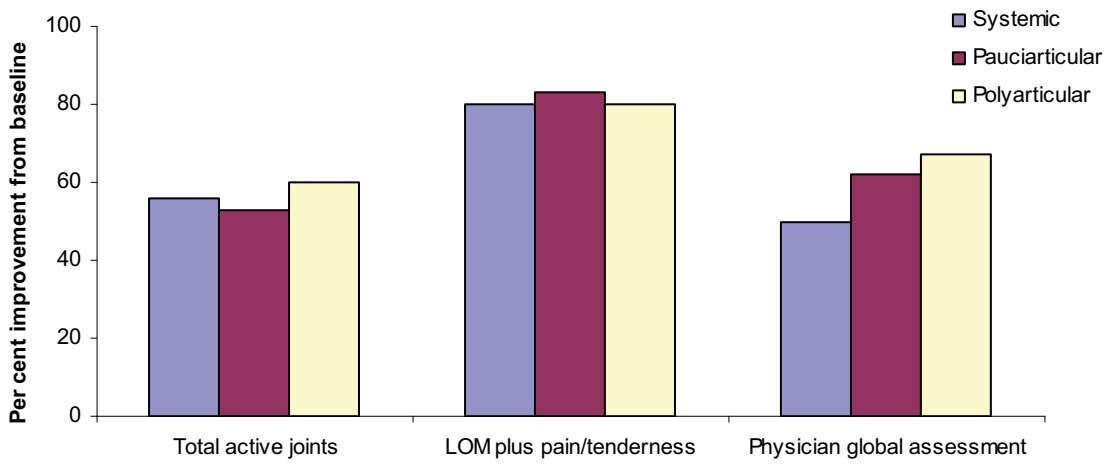
Corticosteroids

Corticosteroids that are taken by mouth or injected into the joint spaces are most often used to control the initial stages of pauciarticular (oligoarthritis) or polyarticular disease with severe morning stiffness or night pain. Computed tomography (CT)-guided injection of corticosteroid into an affected temporomandibular joint (TMJ) has been found to be very effective (Arabshahi et al. 2005).

Oral corticosteroids may also be used as 'bridge' therapy when starting a stronger second-line medication, such as methotrexate, to control symptoms while the new medication takes effect. After a period, the corticosteroid is slowly withdrawn to see whether the other medication is effective. Injections of corticosteroids may be used to treat specific joints when conservative therapy has otherwise controlled symptoms well. Corticosteroid eye drops are used in children who develop inflammatory eye disease. However, long-term use of oral corticosteroids causes major side effects, including a weakened immune system and weakened muscles, among many other problems.

Biologics

Certain biologics, etanercept in particular, have been demonstrated to be effective in treating several types of juvenile arthritis. It is considered to be most beneficial in children who do not respond adequately to conventional management, have severe and aggressive disease, and adverse side effects of long-term medication including steroids. A multicentre randomised, double-blind, placebo-controlled trial found etanercept to be effective and safe in children with the polyarticular subtype found to be resistant to methotrexate (Lovell et al. 2000). The safety and long-term effectiveness of etanercept was confirmed by Lovell and his colleagues again in a different multicentre, open-label, extended-treatment trial in 2001 and also in other open-label pilot trials (Ilowite 2002; Henrickson & Reiff 2004). Significant improvement was noticed in people with three juvenile arthritis subtypes after open-label etanercept treatment (Figure 5.1).



Notes

1. Percentage improvement from baseline at day 90.

2. LOM: loss of motion.

Source: Ilowite 2002.

Figure 5.1: Effectiveness of etanercept in children with juvenile arthritis

The use of etanercept, however, does not always lead to sustained improvement over a longer period, especially in children with the systemic subtype (Quartier et al. 2003). This is mainly due to severe side effects such as increased risk of infections, injection site reactions and upper respiratory tract infections. Further clinical trials are required to demonstrate the efficacy and safety of infliximab in the treatment of juvenile arthritis.

The introduction of these various medications has markedly improved the treatment of arthritis in children. The effectiveness of some of the medications is shown in Table 5.2. There is, however, still a lack of evidence to guide the treatment of some subtypes of juvenile arthritis.

Table 5.2: Effectiveness of common medication used to treat selected types of juvenile arthritis

	Efficacy	
Medication	**Persistent oligoarthritis**	**Polyarthritis**
Non-steroidal anti-inflammatory drugs (NSAIDs)	Mild–moderate	Mild[(a)]
Intra-articular corticosteroids[(b)]	Significant[(c)]	Moderate[(d)]
Methotrexate	Unknown	Significant
Sulfasalazine	Unclear	Moderate
Etanercept	Unknown	Significant
Infliximab	Unknown	Significant
Adalimimab	Unknown	Significant

(a) Effective in up to 25% of children.

(b) For intra-articular steroids efficacy was measured as benefit for >6 months.

(c) 'Significant' denotes effective in >50% of children.

(d) 'Moderate' denotes effective in 25% to 50% of children.

Note: There is a lack of evidence on the utility of medications in other subtypes of juvenile arthritis.

Source: Hashkes & Laxer 2005.

Local treatment

Local treatment aims to prevent stiffness and deformity. Physiotherapy is found to be particularly useful. Mild heat techniques may be helpful in easing joint symptoms, especially stiffness and protective muscle spasm in acute stages of the disease. Prone lying for some time each day may prevent flexion contracture of the hips. Night splints for various joints are also useful.

A moderate impact aerobic exercise program improves the pain and increases the cardio-respiratory performance of children. Hydrotherapy programs are also safe and beneficial.

Children experiencing increased pain during routine physical exercise should, however, self-limit their activities. Children and their families should be educated in basic joint protection techniques. For example, static flexion positions should be avoided; affected joints should be maintained in an extended position during rest. Activities such as cartwheels, handstands, chin-ups and rope climbing should be avoided so that the total body weight is not placed on the non-weight-bearing affected joints.

Surgical interventions

Surgery is considered as a last resort in the management of juvenile arthritis, but it can be necessary if joints become eroded and pain cannot be adequately controlled. It is particularly useful in children with pauciarticular or polyarticular-onset subtypes, where there is bone and soft-tissue deformity, leg length inequality or joint destruction, and medical treatment has failed. It has, however, been recommended that, if possible, reconstructive surgery should be delayed until completion of skeletal growth (Glueck & Gellman 2005).

Some of the commonly used surgical procedures are:

- soft tissue releases of contractures, involving cutting the muscles attached to an abnormally bent joint. As the muscles and other shortened tissues are released, the affected joint can return to a more normal position
- total joint replacement: arthroplasties of hip and knee may be required.

Other surgical procedures recommended for selected cases include:

- osteotomy, involving removing a wedge of bone to allow more normal alignment of the joint. An osteotomy may be recommended for children who have severe joint contractures
- epiphysiodesis: the portion in a long leg bone where growth occurs is removed in order to stop growth
- cataract extraction with intraocular lens in children with juvenile rheumatoid-related uveitis (Lam et al. 2002).

Other therapies

Some other therapies have also been found to be useful in managing pain and maximising the functional abilities of children with arthritis.

Diet

No specific diet helps in the treatment of juvenile arthritis. However, as children with juvenile arthritis are at risk of osteopenia, the inclusion of at least three servings of calcium-rich foods each day may help to strengthen their bones. Some children may gain too much weight due to limited activity and the side effects of medication, which in turn puts more stress on weight-bearing joints such as knees, hips and ankles. A well-balanced diet with appropriate exercise can help children keep a normal body weight.

Cognitive-behavioural pain management

Cognitive-behavioural pain management sessions can sometimes be effective. A 6- and 12- month follow-up study of the use of cognitive-behavioural self-regulatory techniques such as progressive muscle relaxation, guided imagery and meditative breathing, taught to children with pauciarticular or systemic onset subtypes consistently decreases the pain and improves adaptive functioning (Walco et al. 1992).

Complementary and alternative medicines

The use of complementary and alternative medicines (CAM) to manage the pain in the paediatric population has been found to be useful. Techniques such as acupuncture, biofeedback, herbal medicine, homeopathy and hypnosis helps reduce chronic pain such as headaches (mainly tension headache and migraines), back pain, ear pain, functional abdominal pain, procedural pain and post-operative pain in children (Tsao & Zeltzer 2005). However, evidence-based recommendations for CAM to manage juvenile arthritis are not currently available.

Rehabilitation

As juvenile arthritis impairs joint function and can result in severe physical disability, a comprehensive rehabilitation program must start early to restore loss of function and prevent permanent disability. The program, however, will differ depending on the child's age and developmental status. Small children need adequate mobility for psychosocial development. The next step is integration into school life and peer group. Adolescents will require help for adequate vocational training and self-care. In this case vocational rehabilitation services can be useful by beginning career counselling with adolescents during their senior year in high school.

Appendix A

The International League Against Rheumatism classification of juvenile arthritis

ILAR has classified juvenile arthritis into five different subgroups as well as an unclassifiable category. The subgroups are distinguished by the number and site of joints affected during the first 6 months, and the presence of other symptoms (Petty et al. 2004). All subgroups are characterised by chronic articular inflammation. The features and symptoms of each subgroup are described below.

1 Oligoarthritis, pauciarticular and monoarticular arthritis

These forms of juvenile arthritis are the most common, persistent and mild. The disease affects four or fewer joints, in an asymmetrical fashion, usually the knees, elbows, wrists and ankles. The arthritis starts when the child is 2 or 3 years old, and girls are more often affected than boys. Although general health and growth are not affected by this type of arthritis, in some children the disease may cause the long bones of arms or legs to grow at different rates. For example, uneven leg lengths and limping can occur in children with arthritis in one leg.

Extended oligoarthritis

Extended oligoarthritis has the features of persistent oligoarthritis, but with a more severe outcome. More than four joints are affected in the 6 months after the onset of symptoms.

2 Systemic onset arthritis

This least common but most serious form of juvenile arthritis affects many areas of the body at the same time, mainly internal organs as well as joints. It occurs mostly under the age of 5 and affects girls and boys equally. The common symptoms and effects of systemic arthritis are:

- spiking fever (103° F or higher) followed by a rash that may last for weeks or even months
- joint inflammation, which usually begins at the same time as the fever and rash but may also begin weeks or months later
- swollen lymph glands and enlargement of the liver and spleen
- a feeling of listlessness and unwellness during the fever, mostly in the late afternoon or evening
- fatigue, aching limbs, anaemia and weight loss
- inflammation of internal organs causing stomach pain, and often affecting the heart or lungs.

3 Polyarthritis

Also known as polyarticular arthritis, polyarthritis affects five or more joints during the first 6 months of disease. This type of arthritis can occur at any age, and is more common in girls than in boys. The arthritis usually starts in several joints at the same time. In some cases, however, it will begin in only one or two joints, and then spread to other joints. The symptoms of polyarthritis include mild fever, loss of appetite and anaemia. There are two subtypes: rheumatoid factor positive and rheumatoid factor negative. The rheumatoid factor (RF or RhF) is an autoantibody (antibody direct against an organism's own tissues) generally associated with rheumatoid arthritis.

4 Enthesitis-related arthritis

Enthesitis means inflammation of the place where the tendons attach to the bone. Other names for this type of arthritis are juvenile spondylitis, juvenile spondyloarthropathies and seronegative enthesopathy-arthropathy syndrome. This type of arthritis affects the large joints of the legs and arms. It is common in children between the ages of 9 and 12 years, mainly boys. Symptoms may include:

- pain or tenderness in the sacroiliac region (the lower back and across the top of the buttocks)
- spinal pain
- stiffening of the joints and ligaments leading to fusion of the vertebrae and loss of mobility.

5 Psoriatic arthritis

Psoriatic arthritis is inflammatory arthritis of the joints associated with the skin condition psoriasis. It often affects only one or a few joints and may involve the hips, back or the fingers and toes. This type of arthritis can occur at any age, but is more common in girls under 6 years of age and boys around the onset of puberty. In about half of children with psoriatic arthritis, the disease starts before any sign of the skin condition. Symptoms of psoriasis include:

- scaling red rash commonly seen behind the ears, on the eyelids, elbows, knees, and at the scalp line or navel
- occasional pitting or ridging of the fingernails.

6 Undifferentiated arthritis

This category of arthritis does not represent a separate subset but includes children who do not fit into any, or fulfil criteria for more than, one subtype.

Appendix B

Data sources

Most of the information included in this report is drawn from population surveys conducted by the ABS: the 2004–05 NHS and the 2003 SDAC. Where applicable, other information has been drawn from administrative databases, including the AIHW National Mortality Database and the AIHW National Hospital Morbidity Database.

A brief description of these data sources follows.

Population surveys

These surveys are designed to gather information about the characteristics and features of the general population. The two commonly used population surveys for health and disability related information are the NHS and the SDAC.

National Health Survey

The NHS, conducted every three years by the ABS, is designed to obtain national information on the health status of Australians, their use of health services and facilities, and health-related aspects of their lifestyle. The most recent NHS was conducted in 2004–05, with previous surveys being conducted in 1977, 1983, 1989–90, 1995 and 2001. The survey is community-based and does not include information from people living in nursing homes or otherwise institutionalised.

Data available from the NHS include self-reports of various forms of arthritis, back pain, osteoporosis and other diseases of the musculoskeletal system and connective tissues. Some information on age at diagnosis, medications used and other actions taken for arthritis is also available.

Survey of Disability, Ageing and Carers

Also conducted by the ABS, the SDAC collects national information on people with disabilities, older people (aged 60 years or over) and their carers. The survey is conducted every 5 years (with surveys in 1988, 1993, 1998 and 2003), and covers people in both private and non-private dwellings, including those in cared accommodation establishments, but excluding those in correctional institutions. The survey collects data on disability due to impairments, activity limitations and/or participation restrictions, and also collects information about the role of various diseases and health conditions in causing disability.

Administrative data sources

In recent years, administrative databases have been increasingly used for statistical analysis by health officials and academics, both at the national and international level. The data in these databases are collected systematically and regularly and have broad population coverage.

AIHW National Hospital Morbidity Database

This database contains demographic, diagnostic, procedural and duration of stay information on episodes of care for patients admitted to hospital. The collection is maintained by the AIHW using data supplied by state and territory health authorities. The database is episode-based and does not have individual-specific information. In this report, disease data relate to the principal diagnosis reported for hospitalisations unless otherwise specified.

AIHW National Mortality Database

The mortality data contains information on the cause of death supplied by the medical practitioner certifying the death or by a coroner. Registration of deaths is the responsibility of the state and territory registrars of Births, Deaths and Marriages. Registrars provide the information to the ABS for the coding of cause of death and the encoded data are then made available to AIHW. In this report, unless otherwise specified, death data relate only to the underlying cause of death.

Appendix C

Definition of medical terms

Adenovirus A group of viruses that cause symptoms similar to those of the common cold. They infect the tissue linings of the respiratory tract, the eyes, the intestines, and the urinary tract.

Ankylosing spondylitis (AS) A seronegative polyarthritis that can progressively stiffen the spine and sacroiliac joints. Strongly associated with the tissue-type antigen *HLA* B27. AS mainly affects males.

Aortic valve regurgitation Condition occurring when the heart's aortic valve does not close tightly with each heart beat after blood is pumped out of the heart to the rest of the body. As a result, some of the blood just pumped out leaks back into the heart. Also known as *aortic insufficiency* or *aortic incompetence.*

Autoimmunity A disorder in which the body's immune system reacts against some of a person's own body tissues or products by producing antibodies that damage them.

Boutonniere deformities A deformed position of the finger in which the joint nearest the knuckle is permanently bent toward the palm while the furthest joint is bent back away.

Campylobacter A type of digestive tract bacterium that causes gastroenteritis, mainly among children and young adults. May be involved in the development of some cases of *reactive arthritis.*

Cardiac tamponade A condition in which excessive fluid accumulates between the heart and its protective covering, the pericardium.

Cervical spine The neck bones of the vertebral column, made up of seven vertebrae, beginning at the base of the skull. The cervical vertebrae contain and protect the spinal cord, support the skull, and enable diverse head movements.

Chlamydophila pneumoniae A respiratory pathogen that causes mild pneumonia or bronchitis in adolescents and young adults.

Coeliac disease An autoimmune disorder of the small intestine, marked by sensitivity to gluten-containing foods and causing poor digestion, diarrhoea and other potentially serious symptoms. Can be well treated by a strict and life long gluten-free diet.

Coxsackievirus A group of viruses that can multiply in the human digestive tract and can spread from person to person, usually on unwashed hands and surfaces contaminated by faeces. Type A coxsackieviruses usually cause less severe illnesses but Type B can seriously damage the heart, brain and other parts of the body.

Cystic fibrosis A serious and chronic hereditary disease that affects the mucus glands of the lungs, liver, skin, pancreas and intestines, making the mucus thick and sticky. Despite great improvements in treatment, life expectancy is markedly reduced, with respiratory complications most prominent.

Cytokines Small proteins that are released by cells and play a key role in immunity and inflammation.

Cytomegalovirus A member of the herpes family that occurs often in humans and mostly causes mild symptoms. But it can lead to congenital problems in the babies of females infected during pregnancy.

Dermatomyositis An inflammatory muscle disease that causes weakness and also involves skin problems such as rash on the eyelids.

Disseminated intravascular coagulation A serious condition in which the blood starts to clot throughout the body. It can occur in response to various large disturbances such as major injury or severe infection. Also called consumptive coagulopathy.

Endarteritis Chronic inflammation of the inner layer of blood vessels.

Epiphysis The end piece of the bone, where the new bone is formed as a person grows.

Fibromyalgia A condition marked by muscle pain, stiffness and fatigue, but with no demonstrable changes in the tissues.

Haemolytic anaemia Anaemia due to the abnormal breakdown of red blood cells.

Herpes simplex virus Causes several distinct disorders based on the site of infection. Oral herpes, also known as cold sores, infects mainly the lips. The other form of herpes mainly affects the genitals.

Human leukocyte antigens (HLA) A set of antigens that act as sentries to the human tissues. Central to distinguishing self from non-self, HLA show strong associations with a variety of diseases in particular those with immunological components.

Hypothyroidism Reduced activity of the thyroid gland; low levels of thyroid hormone may lead to cretinism and goitre. Occurs in endemic form in areas of iodine deficiency. The child with the condition may become severely dwarfed if not treated early.

Inflammation Local response to injury or infection, marked by local redness, heat, swelling and pain. Can also occur when there is no clear external cause and the body reacts against itself, as in auto-immune diseases.

Influenza A common and very contagious viral disease that causes sneezing, coughing, other symptoms such as fever and headache, and can sometimes have serious complications.

Joint subluxation A partial dislocation of a joint, which can result from acute injury and conditions such as severe rheumatoid arthritis.

Leukaemia A cancer in which far too many white blood cells (leukocytes) are produced, those cells are immature or abnormal, and the production of other vital blood components such as red cells and platelets is suppressed.

Measles A highly contagious infection that causes flu-like symptoms, fever, a typical rash and sometimes serious secondary problems such as brain damage. Usually occurring in children, measles is preventable by vaccination.

Mumps A contagious viral disease marked by acute and painful swelling of the saliva-producing glands, often similarly affecting the testicles and sometimes other parts of the body. Preventable by vaccination.

Neuroblastoma A cancer of nerve tissue outside the brain, that occurs mostly in young children. It can spread to several other parts of the body including the bones, where the resulting symptoms may be confused with arthritis.

Osteochondrosis The interruption of the blood supply of a bone, in particular to the epiphysis, which leads to localised death of bone tissue.

Parvovirus B19 A respiratory virus that has reportedly been a cause of arthritis with mild and transient joint symptoms. In some cases of juvenile arthritis, B19 virus was isolated from the synovial fluid.

Pericarditis An inflammation of the pericardium (the fibrous sac that encloses the heart).

Reactive arthritis An autoimmune disease that develops in response to digestive tract infections. Associated with *HLA* B27 but less often than with Reiter's disease. Reported almost equally in both sexes. Mild end of the severity spectrum of spondyloarthropathies.

Reiter's disease An autoimmune *spondyloarthropathy* with syndromic presentation of joint inflammation, conjunctivitis and genital lesions. Strongly associated with *HLA* B27, the disease is more common in males.

Rheumatoid factor (RF or RhF) An abnormal autoantibody in the blood that can act against one of a person's other antibody types (IgG). Rheumatoid factor is commonly present in people with rheumatoid arthritis; hence it is termed seropositive arthritis. It is strongly associated with *HLA* DR4.

Rubella A viral disease of children and young adults, with mild symptoms. Often causes serious birth defects if mother contracts it during the first 3 months of pregnancy. Preventable by vaccine.

Salmonella A foodborne bacterium that leads to sudden onset of headache, abdominal pain, fever, diarrhoea, nausea and sometimes vomiting. A common cause of 'food poisoning' outbreaks.

Scleroderma A usually chronic skin disease marked by thickened skin due to excessive amounts of collagen, the main protein in white connective tissue.

Scoliosis An abnormal sideways curvature of the spine.

Seronegative Absence of rheumatoid factor in the serum. A biocharacteristic used for distinguishing spondyloarthropathies from rheumatoid arthritis.

Seropositive Presence of rheumatoid factor in the serum. A biomarker for rheumatoid arthritis. Some forms of juvenile arthritis are also seropositive.

Shigella A bacterium causing acute diarrhoea with fever, nausea and sometimes other symptoms, usually transmitted directly from person to person via the faecal-oral route.

Slipped epiphysis When the femur slips out of its hip socket to a weakening of its *epiphyseal* growth plate. In advanced cases, the slippage shortens the leg and pushes the foot point outwards.

Spondyloarthropathies A group of seronegative arthritic conditions that cause inflammation of the spine and sacroiliac joints. In addition to the classical *ankylosing spondylitis*, the category includes various diseases such as *Reiter's disease* and is strongly associated with the tissue type *HLA* B27.

Subclinical hypothyroidism A condition in which the thyroid gland is only just able to produce enough thyroid hormone. The person has no symptoms of thyroid deficiency but the condition is detected by raised levels of the pituitary hormone that is needed to stimulate the thyroid glands.

Systemic lupus erythematosus (SLE) An autoimmune connective tissue disease with a typical facial rash and symptoms often similar to rheumatoid arthritis. More common in females, SLE can affect many body organs.

Temporomandibular joint (TMJ) The joint of the upper jaw (maxilla) and lower jaw (mandible), located directly in front of each ear.

Thyroiditis Inflammation of the thyroid gland.

Vasculitis Inflammation of the wall of smaller blood vessels, including capillaries and smaller veins and arteries.

Yersinia A bacterium that causes acute diarrhoea, fever and other abdominal symptoms, and may be involved in *reactive arthritis*.

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