

INFORMATION

information

Arthritis

VICTORIA

inc. OSTEOPOROSIS VICTORIA

JUVENILE ARTHRITIS

An information sheet for the family of a child/young person diagnosed with arthritis:

- What is Arthritis?
- Diagnosis
- Different Forms of JA
- Outlook
- Treatment / Management

What is arthritis?

Arthritis means "inflammation of a joint". A joint is where bones meet. The term arthritis derives from the Greek words "Arthron" meaning joint and "Itis" meaning inflammation.

The symptoms (signs) of arthritis within a joint include:

- swelling
- tenderness
- stiffness
- redness and/or
- Warmth

One of these symptoms, all of them, or any combination of them, may be apparent in a joint at any particular point in time when arthritis is present.

Juvenile Arthritis

Different terms are often used when referring to arthritis in childhood/adolescence. This can be very confusing. The simple explanation is that different countries use different names but, they are all describing the *same* condition. In Australia, the United Kingdom and Europe it is common to use the term Juvenile Chronic Arthritis (JCA), in the USA and Canada, Juvenile Rheumatoid Arthritis (JRA) is used. In some literature childhood arthritis may be called Still's Disease, after a

children's medical specialist, George Frederic Still, who first described the differences between adult and childhood arthritis in 1896. Idiopathic Arthritis of Childhood (IAC) or Juvenile Arthritis (JA) may also be used. An international committee is currently finalizing appropriate and common terminology for the various conditions of childhood arthritis. In this information sheet we will be using the acronym JA.

JA is used as an 'umbrella' term for arthritis in childhood and is the diagnosis when the child's symptoms occur between birth and sixteen years of age. Seven forms of JA have been identified; therefore children with JA may be further classified into one of these sub-groups. Each of the seven forms of JA has its own particular features and management options, however, there are also many commonalities amongst the seven forms.

Each child who has symptoms of JA may present quite differently. Just as each child's personality and interests are unique, so may their particular symptoms of JA be unique. To complicate things further, a child may not 'fit' the general pattern of one of the sub-groups, making it unclear which type of JA they have. JA is a condition where symptoms may alter over the course of the disease, or they may mimic other childhood conditions. Reaching a diagnosis can take what will sometimes seem a drawn out process.

Diagnosis

Obtaining a diagnosis is the first, and possibly most important step in management. Paediatric Rheumatologists specialize in the diagnosis and management of JA. In Victoria we are fortunate to have three paediatric Rheumatologists linked with Victoria's two major paediatric and adolescent

centres in Melbourne. Regular visits to country regions are also scheduled for these specialists. Even with specialist attention however, diagnosis is often difficult, requiring careful monitoring of the child over a period of time. This 'waiting period' can be a difficult time for parents and you may start to question whether the specialist knows what he/she is doing. This is an understandable reaction.

As there is no known cause of JA, there is no one test which determines whether a child has JA. This is a complex condition and diagnosis may involve:

- a physical examination
- monitoring of symptoms (JA is termed a *chronic* condition, meaning that symptoms are present for more than six weeks)
- blood and urine tests
- X-rays
- electrocardiogram (ECG) - for children with systemic onset arthritis
- the exclusion of other conditions whose symptoms are similar to JA.

The seven forms of JA

1. Systemic onset arthritis
2. Oligoarthritis, Pauciarticular arthritis or Monoarticular arthritis
3. Extended Pauciarticular arthritis
4. Polyarticular arthritis (Rheumatoid Factor negative)
5. Polyarticular arthritis (Rheumatoid Factor positive)
6. Entesitis related arthritis
7. Psoriatic arthritis

These are all big words, but what do they mean? Following is an explanation of the characteristics of these seven forms of JA.

Systemic onset arthritis

Systemic means "affecting many parts of the body." Children with this condition may be very ill at the onset with a fever being the first sign noticed. This may fluctuate during the day and night and persist for days or possibly weeks. An obvious rash may be present, often coinciding with the high temperature fluctuations. This rash (which comes and goes in red blotches of various sizes), is one of the most distinctive features. The child's glands in the neck, under the arms and in the groin are usually swollen. These symptoms can sometimes be confused with infections such as measles.

The spleen and liver may also be enlarged. Very occasionally inflammation of the covering of the heart (pericarditis) occurs and, even more rarely, inflammation of the covering of the lungs (pleurisy).

Often children with systemic onset arthritis require hospitalization to rule out other childhood infections or serious illnesses and to stabilize their condition. At this stage the child may have little or no pain and possibly slight or no swelling of the joints. Some children, however, will show signs of arthritis in a few or many joints right from the outset.

A child with systemic onset arthritis will be quite listless and unwell particularly when their temperature is high. As the temperature drops, often during the day, they may feel better, only to feel unwell again as their temperature rises. It is not uncommon (during the period when a child's temperature is fluctuating), for him/her to lose weight, eat very little, feel weak and look pale and anaemic. This is a very stressful and worrying time for families and the doctor should be able to tell you how long this period-of the condition is likely to last. Your specialist will be working hard to bring these symptoms under control.

Children with systemic onset arthritis can expect:

- that symptoms may go away within a short period of time
- that symptoms may persist into adulthood, but with little joint damage occurring
- that symptoms may go away in childhood and recur later in adolescence or adulthood

It is the least common form of JA, and is more common in children under the age of five, and affects boys and girls equally.

Oligoarthritis, Pauciarticular arthritis and Monoarticular arthritis

The terms oligoarthritis and pauciarticular mean that arthritis is present in four or fewer joints during the first six months after the onset of symptoms. These two terms describe the same condition. If only one joint is affected, the term monoarticular arthritis is often used. Commonly, oligoarthritis will be used as a blanket term for this diagnosis.

Oligoarthritis is generally not symmetrical, meaning that it does not usually affect the same joint on both sides of the body. Although the

swelling may first appear after a minor injury, such as a fall, there is no evidence to suggest that the injury caused the arthritis. The child's general health is not commonly affected by oligoarthritis, and the child may not complain of any pain. Often the first sign parents will notice is that their child walks with a limp, or is reluctant to bend or use the affected joint.

There is a risk of an eye condition called uveitis (also known as iritis) in approximately 30% of children with oligoarthritis. As there is no obvious signs of uveitis developing, it is essential that the child's eyes are checked regularly by an eye specialist (ophthalmologist). This remains important for many years after the disease is officially in remission. An instrument called a slit lamp is used to make the diagnosis. This eye condition, if undetected and untreated, can cause severe eye damage, so it is very important that appointments with the ophthalmologist are kept.

The AV fact sheet entitled 'The Eyes and Juvenile Arthritis' gives more detail about uveitis.

Children with Oligoarthritis can expect:

- the symptoms to go away after a few years
- there will be no further recurrences in most cases
- that some will have a longer lasting involvement

Oligoarthritis is the most common form of JA, affecting girls more commonly than boys. It often commences between the ages of two and three.

Extended Oligoarthritis

Sometimes a child with oligoarthritis can develop arthritis in more than four joints after the initial six month period. In this case, the term Extended Oligoarthritis is used. The joint involvement is usually symmetrical, affecting the same joints on both sides of the body. As with Oligoarthritis, there is a risk of the eye condition, uveitis occurring, and regular checks are required.

Children with Extended Oligoarthritis can expect:

- that symptoms will often persist over a longer period of time
- but eventually will usually then go away

Girls are more commonly affected than boys.

Polyarticular arthritis (Rheumatoid factor negative)

Polyarticular means "many joints." In this form of JA, five or more joints will be affected. Rheumatoid factor is an antibody in the blood detected by a simple blood test. Blood tests in the majority of children with JA will show a negative result for rheumatoid factor.

The onset may be sudden, with a number of joints being affected at one time or in rapid succession. The joint involvement is usually symmetrical, meaning that the same joint is affected on both sides of the body.

At the beginning, as with systemic onset arthritis, the child may be generally unwell. For other children however, the arthritis may develop with little or no affect on general health. Later, symptoms of listlessness and fatigue may occur, and the child may be reluctant to move his/her joints. Reluctance to move painful, stiff or swollen joints can result in the joint stiffening in a bad position for normal functioning. It is important that the child is made aware of this possibility and that appropriate management strategies are in place to prevent this happening wherever possible.

The eye condition uveitis is not common in this form of juvenile arthritis, but it can occur.

Children with polyarticular arthritis - rheumatoid factor negative can expect:

- that the majority of children will have little or no active JA after 16 years
- that some children will have ongoing JA into adulthood

It can develop at any age from a few months to adolescence. It is more commonly diagnosed in girls aged between six and ten years.

Polyarticular arthritis (Rheumatoid factor positive)

This form of Polyarticular Arthritis also affects five or more joints. The joint involvement is usually symmetrical, meaning that the same joint is affected on both sides of the body. The rheumatoid factor antibody is present in the blood but may not show at the onset of symptoms.

It is important to note that rheumatoid factor is also present in a percentage of the population who do not show signs of having arthritis, therefore it is not a test for making a diagnosis.

Girls are more commonly affected than boys and it occurs more often from the age of ten, but it may also affect some younger children. This can be a severe form of JA, but is quite rare, affecting approximately 1% of children with arthritis. This form of JA appears to be very similar to adult type rheumatoid arthritis, but with a very early onset. Sometimes there may be involvement of internal organs or a child may be anaemic. The eye condition uveitis is not common in this form of JA but can occur.

Children with Polyarticular Arthritis - rheumatoid factor positive can expect:

- that in some children the condition will be well controlled with treatment
- that a significant proportion of children will have severe, active disease into adulthood with resultant limitations in joint function

Enthesitis related arthritis

Entheses are areas where tendon and other connecting tissues - join to bone. Pain at these points of connection is known as enthesopathy, and inflammation at these sites is known as enthesitis. Therefore, Enthesitis Related Arthritis is the name given to a group of conditions in which there may be inflammation particularly of the hips, knees, and ankles, the spine and also of the entheses around the heel. Other names, which have been used, for this group of conditions include juvenile spondylitis, juvenile spondylarthropathies and Seronegative-enthesopathy-arthropathy syndrome.

Enthesitis related arthritis is more common in boys than girls and often begins between the ages of nine and twelve years. Sometimes other family members may have the same condition or an associated condition, for example, Ankylosing Spondylitis. There is a higher prevalence of the HLA-B27 genetic marker in children who have Enthesitis Related Arthritis but, as with the rheumatoid factor antibody, a percentage of the general population who show no signs of arthritis also have this marker.

These children are at risk of the eye condition acute uveitis but this eye condition is quite different to the one that children who have pauciarticular arthritis may develop. Acute uveitis results in a painful, red eye so it becomes quite obvious when they have this condition. For this reason, it is not necessary to be watched as carefully for eye disease.

Children with Enthesitis Related Arthritis can expect:

- that the symptoms may go away after a short period of time
- that some children's symptoms may go away and then recur later in adolescence or adulthood
- that some children may go on to have ankylosing spondylitis in adulthood.

Psoriatic arthritis

This is an inflammatory arthritis affecting several or numerous joints, and is usually associated with the skin condition psoriasis. The psoriasis may not appear for many years after the onset of arthritis, or may precede symptoms. There is commonly a family history of psoriasis, and less commonly, a family history of arthritis. Psoriatic arthritis is often characterized by the appearance of sausage shaped fingers or toes and a 'pitted' appearance to the finger and toenails.

It can occur at any age in childhood. It generally affects girls more often than boys where onset is before the age of six, and boys more often than girls where onset is around the age of puberty. As with Pauciarticular Arthritis, there is a risk of the eye condition uveitis occurring, and regular checks are required.

Children with Psoriatic Arthritis can expect:

- the symptoms may go away after a short period of time
- in some children the symptoms may go away and then recur later in adolescence or adulthood

Other related conditions

There are a number of other conditions in which persisting arthritis may be seen. These include:

- Systemic Lupus Erythematosus (often called Lupus or SLE)
- Scleroderma
- Vasculitis
- Reactive Arthritis
- Juvenile Dermatomyositis
- Septic Arthritis
- Haemophilia
- Cystic Fibrosis

This is by no means an inclusive list. For information on these and other conditions, please contact Arthritis Victoria.

Outlook

The outlook for children with JA is very good in the majority of instances. Most children will grow up without any obvious disability. The good news is that approximately 75% of children affected will not have their JA by the time they become adults. It is not possible to determine however, which child will “grow out” of their JA and which child’s JA will continue into adulthood. For this reason, the management of JA is vital.

What we do know with some certainty is that JA is a condition that is unpredictable in its nature. It does not tend to run a single course. There will be periods where the symptoms are exacerbated, (called a ‘flare’), and times when symptoms will be slight or even absent. Sometimes a flare will coincide with an infection, injury or when the child is over-tired, at other times there will be no apparent reason. Being unable to predict the ups and downs that are likely to happen makes it difficult to be sure of the exact outcome. Appropriate management, both during and in between flares, is vital in maintaining as ‘normal’ a life for the child and his/her family.

Despite good care, there will be a number of children who will develop serious joint deformities and resultant limitations. Under these circumstances, joint replacement surgery may be required at a time when the child’s skeleton has completed its growth.

The particular form of JA that a child is diagnosed with will give some general indication of the long-term outlook but it is important to note that these are just *generalizations*. Discuss the outlook further with your specialist.

Treatment/Management

There is no cure, wonder drug or diet for JA. However, appropriate treatment or ‘management’ as it is commonly called, is vital. Each child’s management program will be individualized according to their age, condition type, its severity and other symptoms. The aim of management is to:

- control or relieve the amount of pain experienced
- reduce inflammation in the joint
- maintain or improve the range of joint movement
- maintain muscle strength surrounding the joint

- prevent as much disability as possible into adulthood
- allow ‘normal’ physical and emotional growth and development
- help the child to live as normally as possible

The team approach

The management of JA is a team effort. This team may at any given time include: paediatric rheumatologist, pediatrician, general practitioner, physiotherapist, occupational therapist, ophthalmologist, podiatrist, dentist, pharmacist and most importantly, the child and his/her parents and family.

As JA can be unpredictable and there is never one management solution, a child is likely to undergo a range of treatments throughout the course of their condition.

Medication

The majority of children with JA will require medication at one time or another. Medications play a major role in the management of JA. The medications prescribed aim to:

- control inflammation
- relieve pain
- reduce fever
- reduce other symptoms of the disease
- assist in the maintenance of the child’s ‘normal’ growth

Many parents feel anxious at the prospect of their children taking medications. This is a natural and understandable response. Children too may not want to take their medications for any number of reasons, particularly when their symptoms improve. The medications used in the management of JA are known to be helpful and have side effects that are easily recognized and detected. Please talk to your child’s specialist about any concerns that you may have. For medications to be effective, it is important that it be taken in the way it is prescribed.

Be sure you talk to the specialist before giving your child any other preparation, even over the counter and ‘natural’ remedies. Natural treatment options are not necessarily safe alternatives as it is still possible to have ill effects or negative interactions with prescribed medications. For pharmaceutical medications to be prescribed in Australia, they must meet many requirements and natural therapies do not necessarily go through the same rigorous testing procedures. It is always

best to consult your specialist before giving your child other preparations.

It may take some time for your child's specialist to determine the right medication or combination of medications for your child. This can be very frustrating, especially if your child is in pain. JA varies greatly from child to child and there are a large range of medications available for the management of JA.

The four main groups of medications used to treat JA:

- Non-steroidal anti-inflammatory drugs (NSAID's)
- Corticosteroids
- Slow acting anti-rheumatic drugs (SAARD's)
- Pain relievers (analgesics)

The type of medication recommended for use will depend on numerous factors including:

- the type of JA
- the severity of the JA
- the child's tolerance of a medication

For more specific information on medications please talk to your child's specialist, pharmacist or Arthritis Victoria. It is a legislative requirement in Australia that there be a Consumer Medicine Information (CMI) brochure available for every prescribed medication. These are readily available (at no cost) from pharmacists and the drug supplier but most medications will have a CMI distributed automatically within its packaging.

Doctors, pharmacists and libraries also hold books which contain current information relevant to all medications available.

The importance of rest

As JA can cause fatigue, it is important that a child with JA has a good nights sleep and when necessary rest during the day –rest does not necessarily have to mean lying in bed. Alternating active and more passive activities throughout the day, wherever possible, can assist a child in actively participating in their normal day events. Rest can be achieved by listening to music, reading a book, drawing a picture or a myriad of other simple activities.

Prolonged bed rest, except when a child is in considerable pain or overly-tired is not generally recommended. It is also important to watch that joints are maintained in an appropriate position

when a child is at rest. Consult your physiotherapist and occupational therapist about ways to ensure this.

Children with JA will often require resting splints, working splints and/or modified footwear. These help to support and protect painful joints, hold joints in a functional position, help to prevent disability and ease joint fatigue.

The importance of exercise and play

Arthritis tends to restrict joint movement, which can result in muscle wasting and weakened bone. Special exercises, which help to keep joints moving, maintain their flexibility, build up muscle and help prevent or limit disability are an important part of a common management strategy for JA. It is important that an appropriate exercise routine is developed by a physiotherapist. Exercise cannot, as is commonly thought, damage a joint. *Inappropriate* exercise can result in increased pain but *appropriate* exercise is, in fact, beneficial in relieving some of the pain associated with JA.

Once an exercise regime has been established it is important that it is maintained at home. A few sessions with a physiotherapist in a hospital does not make up for an ongoing program at home. If you are unsure about how to work through the prescribed exercises, discuss this with the physiotherapist. Building an exercise time into your daily routine will help you to establish their importance.

For their physical and emotional well being it is important for a child with JA to be encouraged to be as active as possible and play games that they find enjoyable. The amount, type and extent of activity may need to be monitored by a parent but remember that children with JA don't want to be different to their peers and may therefore over-extend themselves. Conversely, some 'bad' experiences are not necessarily a reason to halt any particular activity and adjustments, alternatives or compromise, with the child with JA being an integral part of the decision making team, is a far better alternative. A position where the child with JA learns to monitor and manage their own condition is the ideal goal to aim for.

Swimming is an excellent form of exercise as it is also improves muscle strength and joint movement whilst providing buoyancy. This buoyancy effect takes much of the weight from painful joints, allowing greater freedom of

movement. An added and important benefit with swimming is that it often doesn't feel to the child that they are exercising; it's fun and they can do it with friends and family. Warm water pools or spas achieve the best benefits as the warmth of the water helps to relax the muscles.

Hospitalisation

A child with JA may require hospitalization at some stage during the course of their disease.

Hospitalization may occur when:

- the child is experiencing a severe flare-up and a review of medications is required
- joint injections are necessary. These are often done under anesthesia in children.
- as part of an intensive rehabilitation program

FURTHER INFORMATION

Additional resources, information and community assistance are available from Arthritis Victoria.

Resource Centre

This is a small list of items the Arthritis Victoria Resource Centre holds on juvenile arthritis. For a complete list, please contact the Resource Centre.

All listed books are available on loan from the Arthritis Victoria Resource Centre to members. If you would like to find out more about becoming a member, call (03) 8531 8000.

- Brewer, Earl J and Angel, Kathy Cochran 1995, *Parenting a child with arthritis*, Lowell House, Los Angeles, CA.
- Bruce, Elizabeth J; Schultz, Cynthia L 2001, *Nonfinite loss and grief : A psychoeducational approach*, MacLennan & Petty, Sydney, NSW.
- *Juvenile arthritis: A handbook for parents* 1997, Arthritis Foundation of South Australia, Fullarton, South Australia.
- Lehman, Thomas J A 2004, *It's not just growing pains: A guide to childhood muscle, bone and joint pain, rheumatic diseases and the latest treatments*, Oxford University Press, Melbourne, 2004.
- *Raising a child with arthritis: A parent's guide* 1998, Arthritis Foundation of America, Atlanta, GA.
- *Speaking from experience: Arthritis in childhood: A parents perspective* 1999, Tribal. [Video recording]
- Information sheets (varied) pertaining to JA

ARTHRITIS VICTORIA

If you would like further information please contact us on

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or

Toll Free on 1800 011 041 if you are outside the metropolitan area

or

Email: anne@arthritisvic.org.au

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Prepared by Education & Services staff, Arthritis Victoria, November 2002. This material is for your information and is not intended to be medical advice. You are encouraged to review the information provided with your doctor or relevant health professional.