What is Juvenile Idiopathic Arthritis (JIA)?

JIA is really a *group* of illnesses which have been put under one name because they share similar symptoms.

'Juvenile' means a young person (under the age of 16)
'Idiopathic' means that the cause is unknown
'Arthritis' means painful inflammation and stiffness of the joints.

Arthritis doesn't just involve joints it can also involve other tissues and organs of the body. JIA is an umbrella term incorporating the forms of arthritis affecting young people, under the age of 16. The typical age of onset is at or about 6 years of age. Disease prevalence in Australia is between one and four cases for 1000 children. This means that at any one time approximately 5000 children will be affected by JIA. (http://www.racgp.org.au/your-practice/guidelines/musculoskeletal/)

Are there different types of JIA?

Yes, there are seven main types;

1. Oligoarthritis or Pauciarticular [Oligo- = a few]
2. Polyarticular (RhF* negative) [Poly- = many]
3. Polyarticular (RhF* positive) – this subtype may behave similarly to RA in adults
4. Systemic onset
5. Enthesitis- related arthritis
6. Psoriatic Arthritis
7. Undifferentiated Arthritis

*RhF = Rheumatoid factor in the blood

Each of these main types of JIA has its own set of symptoms. Your child's diagnosis will depend on which pattern of symptoms they experience and the number of joints affected during the initial six months of the disease.

What are the main symptoms of JIA?

The common symptoms for all types of JIA include:

- Pain
- Swelling
- Stiffness
- Tenderness
- Redness and warmth around the joint area.

Approximately 50 percent of young people affected by JIA also experience uveitis (inflammation of the 'middle' part of the eye).

What should I look for as a parent?

- If your child stops walking after learning to toddle, or never actually takes to walking
- Stiffness of the joints in the morning is quite common
- Some children don't complain of pain but they can appear moody and difficult.

1. Oligoarthritis or Pauciarticular

- Four joints or less are affected (Oligo = few)
- Typically it does not affect the same joints on either side of the body
- General health is not affected
- Most commonly develops in children under five
- This type has the highest chance of your child developing chronic anterior uveitis (inflammation of the eye that doesn't cause a red or painful eye but can still cause reduced vision if left untreated).
- Most commonly affects the knees, ankles and/or wrist joints and sometimes the elbows and small joints of the hands and feet.

This is the most common form of JIA. Most children will only experience a mild form of the disease.

Polyarticular (RF negative and RF positive)

- Usually comes on suddenly
- Usually affects many joints (Poly = many)
- Typically affects the same joints on either side of the body
- Usually affects the joints of the hands and feet
- General feeling of malaise, tiredness and listlessness
- May cause anaemia (shortage of red blood cells)

RF is an antibody produced by the immune system and can be detected by a blood test. However, this alone does not indicate a definite diagnosis as it is common to have RF negative arthritis.

2. Polyarticular RF negative

- About one quarter of all children affected will go into remission
- Can start at any age and can be a very mild illness, or it can be more severe
- Usually affects the joints of the hands and feet. It may affect the hips, knees, neck, elbows, shoulders or jaw.

3. Polyarticular RF positive

- Usually affects girls aged 10 years or older
- General feeling of malaise, tiredness and listlessness
- May cause anaemia (shortage of red blood cells)
- Similar presentation to adult RA
- Early treatment is important as many children can have quite a severe form of disease

4. Enthesitis- related arthritis

- Pain and inflammation in the soft tissues (tendons, ligaments etc) around the joints – where tendons attach to the bone - rather than in the joints themselves
- This form of JIA commonly affects the spine, heels, hips, knees and ankles (lower limbs)
- Increased risk of developing uveitis
- The majority with this form of JIA will have the genetic marker HLA-B27 which can be detected by a blood test. A significant proportion of patients will develop sacroiliitis as adults (inflammation at the joint between the sacrum and pelvis)
- In contrast to the uveitis seen with other types of JIA, this type is associated with a red, painful eye (acute anterior uveitis).

5. Psoriatic Arthritis

- Along with joint pain, the skin is involved in the form of a scaly rash that can cover large areas of the body
- Usually develops at around 8-9 years old and is more common in girls
- The rash may start at the same time as the joint pain, but not always
- Usually affects the fingers and toes, but it may affect other joints too
- There is a risk of developing uveitis – but it is the painless type that doesn't look red.

6. Systemic Onset

Initially the child will have:

- A high but fluctuating temperature, often in the evenings
- A pinkish coloured rash over the body
- Swollen glands in the axilla (armpits), neck and groin
- Fatigue

- May also develop inflammation of the liver and spleen (Systemic = affecting the body as a whole, rather than particular parts).

This is the rarest type of JIA. It can be confused with other childhood illnesses such as glandular fever, measles or other infections.

For many years it was thought that most children will outgrow their arthritis. However, it is now known that unless treated appropriately, half of children with JIA will still have active arthritis 10 years after diagnosis. (http://www.racgp.org.au/your-practice/guidelines/musculoskeletal/)

7. Undifferentiated

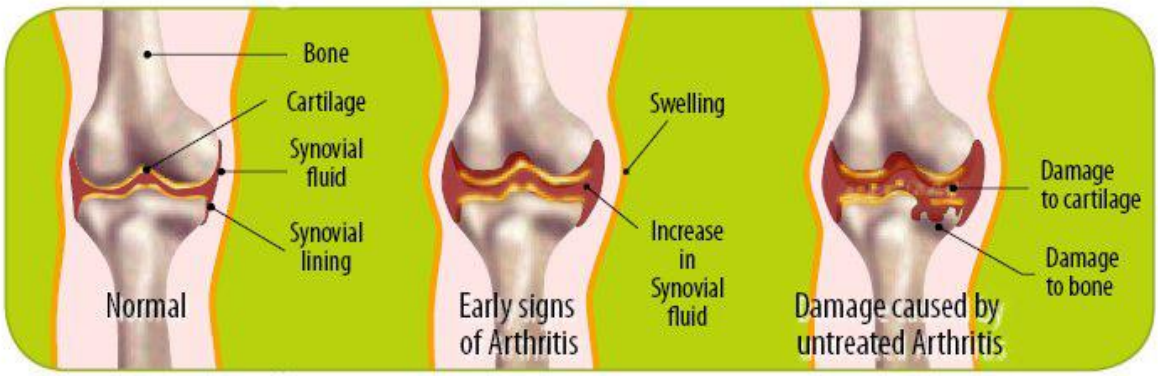
- Occasionally, children and young people can't be fitted neatly into the above groups and their condition is referred to as 'undifferentiated arthritis'.

HOW DID MY CHILD GET THIS DISEASE?

By definition (idiopathic), the causes of arthritis in children are- unknown. The recent research suggests that there may a genetic factor in some forms of arthritis, but these conditions are not classed as being hereditary. If you have one child with arthritis, it is highly unlikely your other children will also develop arthritis.

It has also been proposed that an infection could cause JIA. There has been no evidence in the research to support this. It is most likely due to a combination of genetic factors and various triggering factors from the environment. JIA cannot be passed from one child to another – it is not infectious.

Image from Royal Children's Hospital Melbourne
http://www.rch.org.au/rheumatology/information_about_rheumatological_conditions/Information_on_JIA_for_young_people/



JIA is an *autoimmune* illness. This means that, for unknown reasons, the immune system attacks normal healthy cells in the body rather than fighting foreign bodies such as bacteria or viruses. The immune system

attacks *connective tissue* cells that compose the lining of the joints (synovium). These cells are also found outside the joints, which is why JIA is not confined to the joints and can affect other parts of the body.

DIAGNOSIS

Diagnosis is usually made based on your child's medical history and physical examination. There is no definitive test for JIA. Rather, it involves several steps – that include:

- Taking a detailed medical history of the child and (where possible) their biologically related family members
- Physical examinations
- Pathology tests – including blood and urine tests
- Other tests such as X-rays of the joints.
  Some tests may be repeated several times – in order to monitor changes in the joints at different time intervals. This allows the doctor/specialist to determine rate of disease progression. It is important that your child be referred to a specialist Rheumatologist - preferably a specialist in paediatric arthritis.

  <u>Eye Tests</u>
  It is essential that all children with JIA have regular eye checks to look for inflammation in the eye. This must be conducted by an Ophthalmologist (specialist eye doctor). The type of uveitis associated with the majority of JIA is chronic anterior uveitis, which is painless and not associated with any redness of the eye. This means it is difficult to diagnose without a special test called a slit lamp examination.

If left untreated, chronic anterior uveitis can cause blindness

  It is understandable that it may take some time to reach a definite diagnosis of JIA, and the waiting period may be frustrating. However it is important to be prepared to repeat the tests etc. so a diagnosis can be made. The Paediatric Rheumatologist can then ensure your child receives the best possible treatment to control the symptoms and minimise potential joint damage. If you feel it is required, you can always seek a second opinion. You simply need to ask your GP for another referral.

IS THERE A CURE FOR JIA?

At the present time, there is no cure for JIA. However, with appropriate early management, most children have a very positive outcome and will grow up without any lasting effects. The key to

the best care for your child is early diagnosis, treatment with medicines shown to be effective, an appropriate exercise programme and keeping up usual school and leisure activities.

EVERY RESPONSE IS DIFFERENT

Each child diagnosed with JIA may respond differently to treatment. It is difficult to determine how the condition will specifically affect your child. The good news is that the treatments available for JIA are continually improving. This means that the treatment usually causes the symptoms to disappear for long time periods – called remission. It is best to ask your child's Rheumatologist for a likely prognosis.

THE TEAM APPROACH

To achieve the best possible outcome, the management of JIA will involve a team effort. This team may include;

- Rheumatologist (preferably Paediatric)
- Child with JIA
- General Practitioner
- Physiotherapist
- Occupational Therapist
- Ophthalmologist (specialist eye doctor)
- Podiatrist
- Dentist
- Pharmacist
- Counsellors/Psychologist
- Parents
- Teachers
- Other family members
- Support Groups

The particular members of your child's team will depend on several factors. Firstly, the type of JIA with which your child is diagnosed, how active the disease process is, what joints are involved and how the illness is impacting on your child's well-being.

A BALANCE BETWEEN EXERCISE AND REST IS IMPORTANT

It is a well-accepted health message that exercise is important for all children, including those with JIA. Exercising helps to move stiff joints, strengthen weak muscles and maintain a healthy heart. However, caution is required as incorrectly prescribed exercises can lead to increased pain and cause further joint damage.

Your child's therapists and doctors will advise you on suitable exercises. Swimming or exercising/playing in water is excellent as the buoyancy of the water takes the majority of the weight off painful joints. It also allows greater ease of movement and improves joint mobility and muscle strength. It is even better if the water is warm as this helps with joint stiffness and also relieves pain. There are a number of hydrotherapy pools located in the metropolitan area. You can contact *Arthritis and Osteoporosis WA* for further details. Although water-based exercise has many benefits, it does not assist with increasing bone strength. As one side-effect of some medications prescribed for JIA is osteoporosis (thinning of the bones), it is imperative that your child includes some land-based weight bearing exercise to help increase bone strength. Your child's physiotherapist should guide you on suitable exercises. Your child's teacher should also be involved to discuss how they can participate in PE classes.

JIA can cause tiredness so it is important that your child gets a good night's sleep and a rest period during the day if required. Rest doesn't necessarily mean *complete* recumbence like lying in bed. Rest can be listening to music, reading a book, drawing a picture or other 'quiet time' activities.

Overall, your child should be encouraged to live her or his life as 'normally' as possible. This strategy enables them to increase their self-esteem and feeling in control over their illness.

SUMMARY OF IMPORTANT POINTS

1. Early diagnosis is crucial, so although the number of tests and medical appointments in the initial period can be frustrating, try to persevere. This will ensure your child receives the best possible care and management from the outset.

2. A team approach is important – you and your child are a crucial part of the management team. Open lines of communication assist to coordinate all aspects of your child's care. It is also important to include your child's teachers, family members and friends.

3. JIA can be a *progressive* illness. Be prepared for flare-ups with a management plan that can be put in place when needed. Your child's healthcare team should assist with the implementation of a good management plan.

4. Understand that JIA is still being researched and there will surely be new treatments available in the future. Having a good relationship with your child's healthcare team will ensure your child receives the best available up- to- date treatment of their condition.

5. Encourage your child to keep a diary. This can boost their feeling of control over the disease. Likewise, write down any questions and/or keep a log of the efficacy and side-effects of any new medication you may wish to speak to your child's healthcare team about.

6. Just like other children, your child should maintain a healthy weight and keep active. There may be special considerations like a warm bath or shower in the morning to assist with overcoming joint stiffness, alternative exercises from the child's physiotherapist if they are unable to participate in PE sessions at school, additional rest breaks or 'quiet time' to assist with managing fatigue. However, these are relatively small adjustments in the overall scheme of things in encouraging your child to live a 'normal' life as possible.

Treatment of childhood arthritis has made great strides in recent years. There are myriad of information and resources available.

Below are some useful contacts:

Arthritis & Osteoporosis WA – Tel (08) 9388 2199;
www.arthritiswa.org.au

www.arthritisresearchuk.org – has published a number of booklets containing useful and evidence-based information regarding all aspects of JIA – from different medications to 'A Guide for Teenagers'. A selection of these booklets is available from *Arthritis & Osteoporosis WA*.

www.nps.org.au – National Prescribing Service has information on different drugs. Talking with your doctor or pharmacist is always recommended.

Medicines Information Line
Phone toll free: 1300 888 763