# JIA

PRESENTED BY : NKOMO LIBERTY ZIKHAI

# PLAN OF PRESENTATION

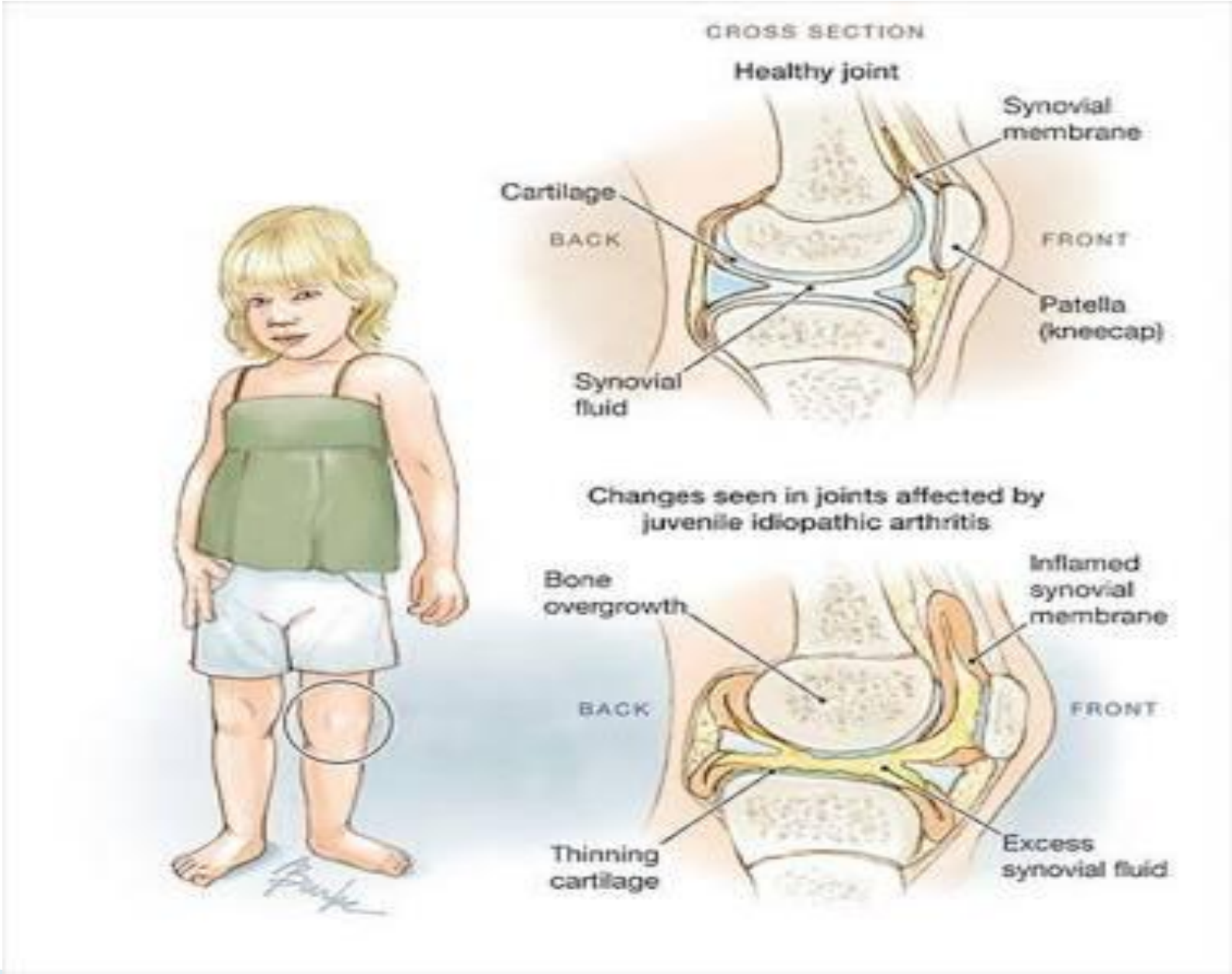
- DEFINITION
- EPIDERMIOLOGY
- SIGNS AND SYMPTOMS
- CAUSES
- CLASSIFICATION
- DIAGNOSIS
- TREATMENT
- PROGNOSIS

# DEFINITION

Juvenile idiopathic arthritis (JIA) , also known as juvenile rheumatoid arthritis (JRA), is the most common form of arthritis in children and adolescents.

Juvenile in this context refers to an onset before age 16 , idiopathic refers to a condition with no defined cause, and arthritis is the inflammation of the synovium of a joint.)

JIA is an autoimmune, non-infective, inflammatory joint disease of more than 6 weeks duration in children less than 16 years of age. The disease commonly occurs in children from the ages of 7 to 12, but it may occur in adolescents as old as 15 years of age, as well as in infants. It is a subset of arthritis seen in childhood, which may be transient and self-limited or chronic. It differs significantly from arthritis commonly seen in adults (osteoarthritis, rheumatoid arthritis), and other types of arthritis that can present in childhood which are chronic conditions (e.g. psoriatic arthritis and ankylosing spondylitis). Aetiopathology is similar to rheumatoid arthritis but with less marked cartilage erosion, and joint instability and absent rheumatoid factor.


CROSS SECTION
Healthy joint
Synovial membrane
Cartilage
BACK
FRONT
Patella (kneecap)
Synovial fluid
Changes seen in joints affected by juvenile idiopathic arthritis
Bone overgrowth
Inflamed synovial membrane
BACK
FRONT
Thinning cartilage
Excess synovial fluid

# EPIDEMIOLOGY

- JIA affects approximately 1 in 1,000 children in any given year, with about 1 in 10,000 having a more severe form.
- JIA occurs in both sexes, but, like other rheumatological diseases, is more common in females. Symptoms onset is frequently dependent on the subtype of JIA and is from the pre-school years to the early teenage years.
- Juvenile idiopathic arthritis affects somewhere between 8 and 150 of every 100,000 children, depending on the analysis. Of these children, 50 percent have pauciarticular JIA, 40 percent have polyarticular JIA and 10 percent have systemic JIA. It has been shown, that in a preselected group (children under 16 years with orthodontic treatment need) prevalence rises to 1 out of 100 (0.88% out of 1024 children).

# SIGNS AND SYMPTOMS

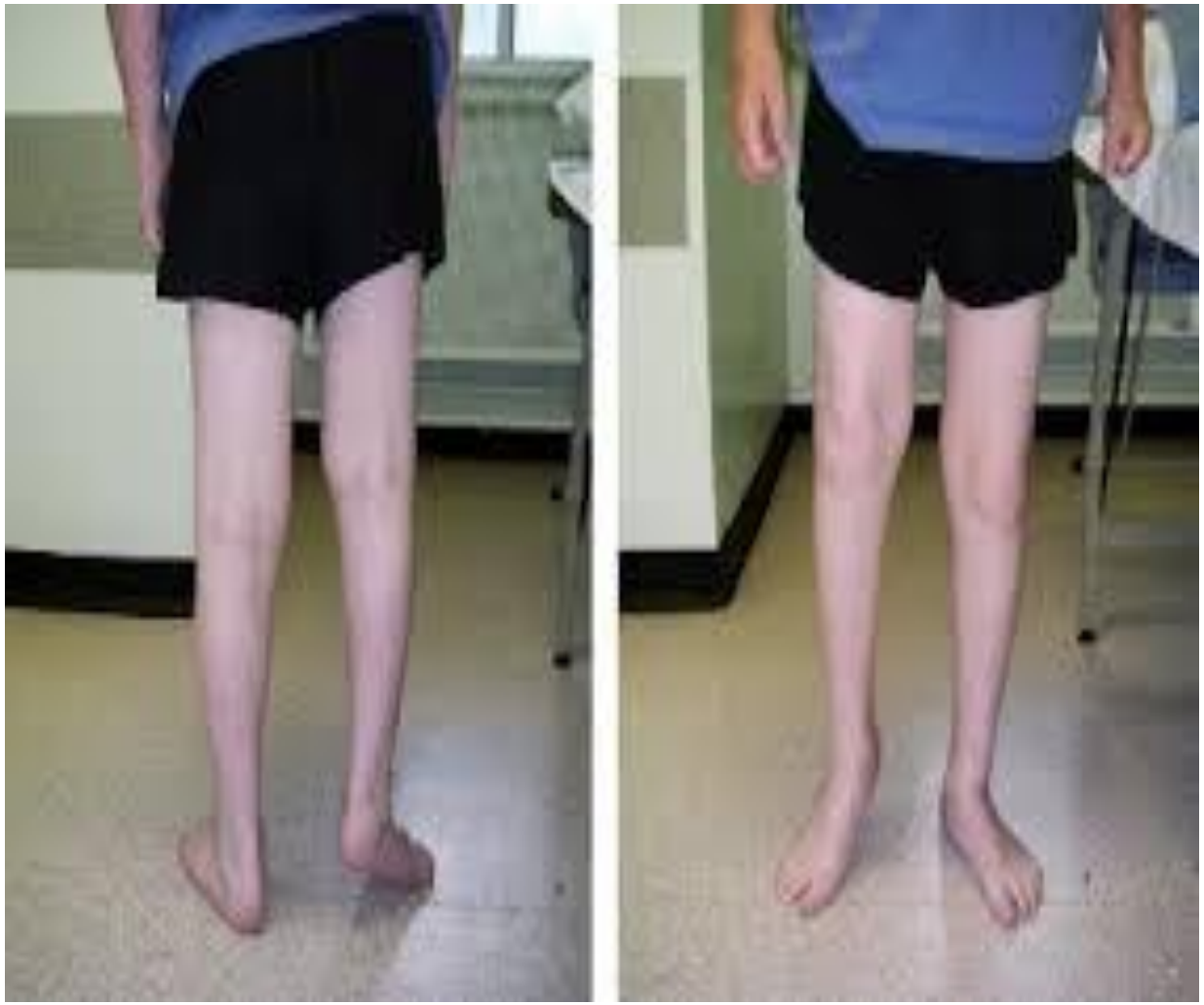
Symptoms of JIA are often non-specific initially, and include lethargy, reduced physical activity, and poor appetite. The first manifestation, particularly in young children, may be limping. Children may also become quite ill, presenting with flu-like symptoms that persist. The cardinal clinical feature is persistent swelling of the affected joint(s), which commonly include the knee, ankle, wrist and small joints of the hands and feet. Swelling may be difficult to detect clinically, especially for joints such as those of the spine, sacroiliac joints, shoulder, hip and jaw, where imaging techniques such as ultrasound or MRI are very useful.

Pain is an important symptom. Morning stiffness that improves later in the day is a common feature (this implies inflammatory type joint pain versus mechanical type joint pain). Late effects of arthritis include joint contracture (stiff, bent joint due to fibrosis) and joint damage. Children with JIA vary in the degree to which they are affected by particular symptoms. Children may also have swollen joints (inflammatory swelling, or in chronic arthritis due to synovial proliferation and thickening, and periarticular soft tissue swelling).

# ETRA ARTICULAIRE

- Eye disease: JIA is associated with inflammation in the front of the eye (specifically iridocyclitis, a form of chronic anterior uveitis), which affects about one child in five who has JIA, most commonly girls. This complication is usually asymptomatic and can be detected by an experienced optometrist or ophthalmologist using a slit lamp. Later slit lamp features include synechiae. Most children with JIA are enrolled in a regular slit lamp screening program, as poorly controlled chronic anterior uveitis may result in permanent eye damage, including blindness.
- Growth disturbance: Children with JIA may have reduced overall rate of growth, especially if the disease involves many joints or other body systems. Paradoxically, individually affected large joints (such as the knee) may grow faster, due to inflammation – induced, increased blood supply to the bone growth plates situated near the joints. This can result in leg length discrepancy, and also deformities such as genu valgum. Asymmetrical growth can also affect other bones e.g. discrepancy in digit length. There can be marked difference in bone age (skeletal maturation).

Fig. 1 Effects of uveitis on the eye.



# COMPLICATIONS

- JIA is a chronic disorder which if neglected can lead to serious complications. Proper follow up with health professionals can significantly reduce the chance of developing complications.
- A form of eye inflammation called uveitis is common with some types of JIA. The inflamed eyes, if left untreated, can result in glaucoma, scars, cataracts and even blindness. Often the eye inflammation occurs without symptoms, or while the JIA is otherwise in remission, and thus it is important for all children to get regular eye check-ups from an eye physician. The presence of ANA is a predictor of eye involvement (common in oligoarthritis type JIA).

- Growth retardation is common in children with JIA. Moreover, the medications (corticosteroids) used to treat JIA have potent side effects that can limit growth. Other musculoskeletal issues may include joint contractures, muscle weakness or muscle loss, and osteoporosis.
- Children who delay treatment or do not participate in physical therapy can often develop joint deformities of the hand and fingers. Over time hand function is lost and almost impossible to recover.

# CAUSES

- The cause of JIA remains a mystery. However, the disorder is autoimmune — meaning that the body's own immune system starts to attack and destroy cells and tissues (particularly in the joints) for no apparent reason. It is believed that the immune system gets provoked by changes in the environment, in combination with mutations in many associated genes and/or other causes of differential expression of genes. Experimental studies have shown that certain viruses that have mutated may be able to trigger JIA. JIA appears to be more common in girls and the disease is most common in Caucasians.

- Associated factors that may worsen or have been linked to rheumatoid arthritis include the following:

.genetic predisposition; it appears that when one family member has been diagnosed with rheumatoid arthritis or another autoimmune disorder, the chances are higher that other family members or siblings may also develop arthritis

.females are more likely to develop rheumatoid arthritis than males at all ages

.there is a strong belief that psychological stress may worsen the symptoms of rheumatoid arthritis. However, when the emotional stress is under control the arthritis symptoms do not always disappear suggesting that the association is not straightforward

.even though no distinct immune factor has been isolated as a cause of arthritis, there are some experts who believe that the triggering factor may be something like a virus which then disappears from the body after permanent damage is done

.because rheumatoid arthritis is more common in women, there is a belief that perhaps sex hormones may be playing a role in causing or modulating arthritis. Unfortunately, neither sex hormone deficiency nor replacement has been shown to improve or worsen arthritis.

- The cause of JIA, as the word idiopathic suggests, is unknown and an area of active research. Current understanding of JIA suggests that it arises in a genetically susceptible individual due to environmental factors.

# CLASSIFICATION

- The 3 major types of JIA are oligoarticular JIA, polyarticular JIA and systemic JIA.

Oligoarticular / Pauciarticular JIA (60%)

Systemic JIA/Still's disease (15%)

Polyarticular JIA/Juvenile RA (10%)

Seronegative spondylarthritis Juvenile AS (5%)

# OLIGOARTICULAR

- Oligoarticular (or pauciarticular) JIA affects 4 or fewer joints in the first 6 months of illness. The prefixes oligo- and pauci- mean "few". Oligoarticular is used with JIA terminology, and pauciarticular is used with JRA terminology. Patients with oligoarticular JIA are more often ANA positive, when compared to other types of JIA. Accounts for about 50% of JIA cases. Usually involves the large joints such as the knees, ankles, and elbows but smaller joints (such as the fingers and toes) may also be affected. The hip is not affected unlike polyarticular JIA. It is usually not symmetrical, meaning the affected joints are on one side of the body rather than on both sides simultaneously. Length discrepancy & muscles atrophy often happens which leads to asymmetric growth and risk of flexion contracture. Early childhood onset are at risk for developing a chronic iridocyclitis or an anterior uveitis, which is inflammation of the eye. This condition often goes unnoticed; therefore these children should be closely monitored by an ophthalmologist. If ANA+, patient need routine eye exam every 3 months. If ANA- and older than 7 years old, can have eye exam every 6 months. Children with late childhood onset are at risk for sacroilitis and spondyloarthropathy.

# POLYARTICULAR

- Polyarticular JIA affects 5 or more joints in the first 6 months of disease. This subtype can include the neck and jaw as well as the small joints usually affected. This type of JIA is more common in girls than in boys. Usually the smaller joints are affected in polyarticular JIA, such as the fingers and hands, although weight-bearing joints such as the knees, hips, and ankles may also be affected. The joints affected are usually symmetrical, meaning that it affects both joints on both sides of the body (such as both wrists.) Children with polyarticular JIA are also at risk for developing chronic iridocyclitis or uveitis (inflammation of the eye) and should also be monitored by an ophthalmologist. Rheumatoid factor may be positive i.e. seropositive in children with polyarticular JIA occurring between 9-16 years of age and is associated with HLA DR4 and HLA DW4. This group has poorer prognosis with about 50% progressing to severe disabiling arthritis, persisting into adulthood. It is generally seronegative in JIA occurring below 10 years of age with a milder disease process and responds better to treatment. Seropositivity is rare in children with systemic JIA. Due to the greater number of joints affected by polyarticular JIA as well as the tendency to worsen over time, polyarticular JIA needs to be treated aggressively.

# SYSTEMIC

- Systemic JIA is characterized by arthritis, fever, which typically is higher than the low-grade fever associated with polyarticular and a salmon pink rash. It accounts for 10-20% of JIA and affects males and females equally, unlike the other two subtypes of JIA, and affects adolescents . It generally involves both large & small joints. Systemic JIA can be challenging to diagnose because the fever and rash come and go. Fever can occur at the same time every day or twice a day (often in late afternoon or evening) with a spontaneous rapid return to baseline (vs. Septic Arthritis of continuous fever). The rash often occurs with fever. It is a discrete, salmon-pink macules of different sizes. It migrates to different locations on skin, rarely persisting in one location more than one hour. The rash is commonly seen on trunk and proximal extremities or over pressure areas.
- Systemic JIA may have internal organ involvement: Hepatosplenomegaly, Lymphadenopathy, Serositis, Hepatitis, Tenosynovitis, etc.
- It is also known as "systemic onset juvenile rheumatoid arthritis".

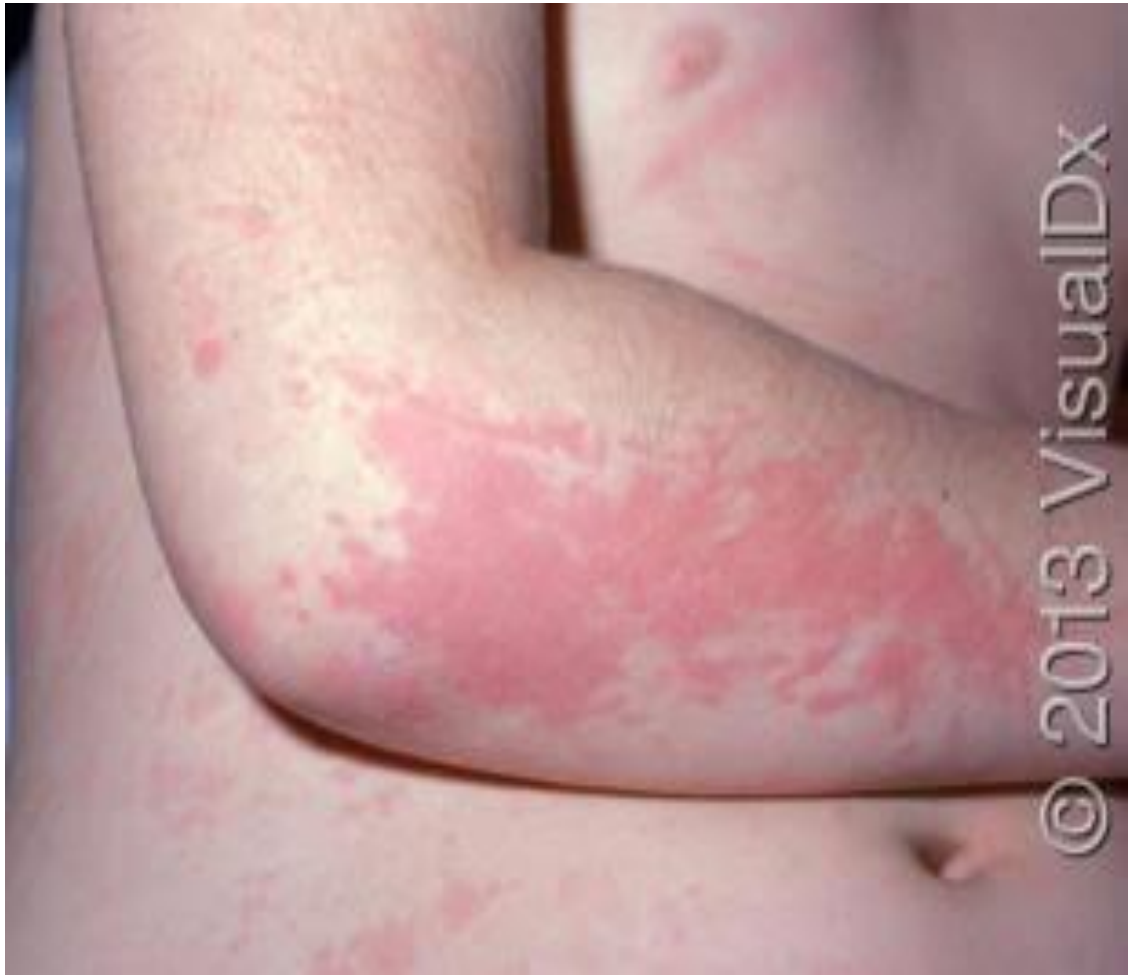
- A polymorphism in macrophage migration inhibitory factor has been associated with this condition.
- It is sometimes called "adolescent-onset Still's disease", to distinguish it from adult-onset Still's disease. However, there is some evidence that the two conditions are closely related.
- Rheumatoid factor and ANA are generally negative in systemic JIA.

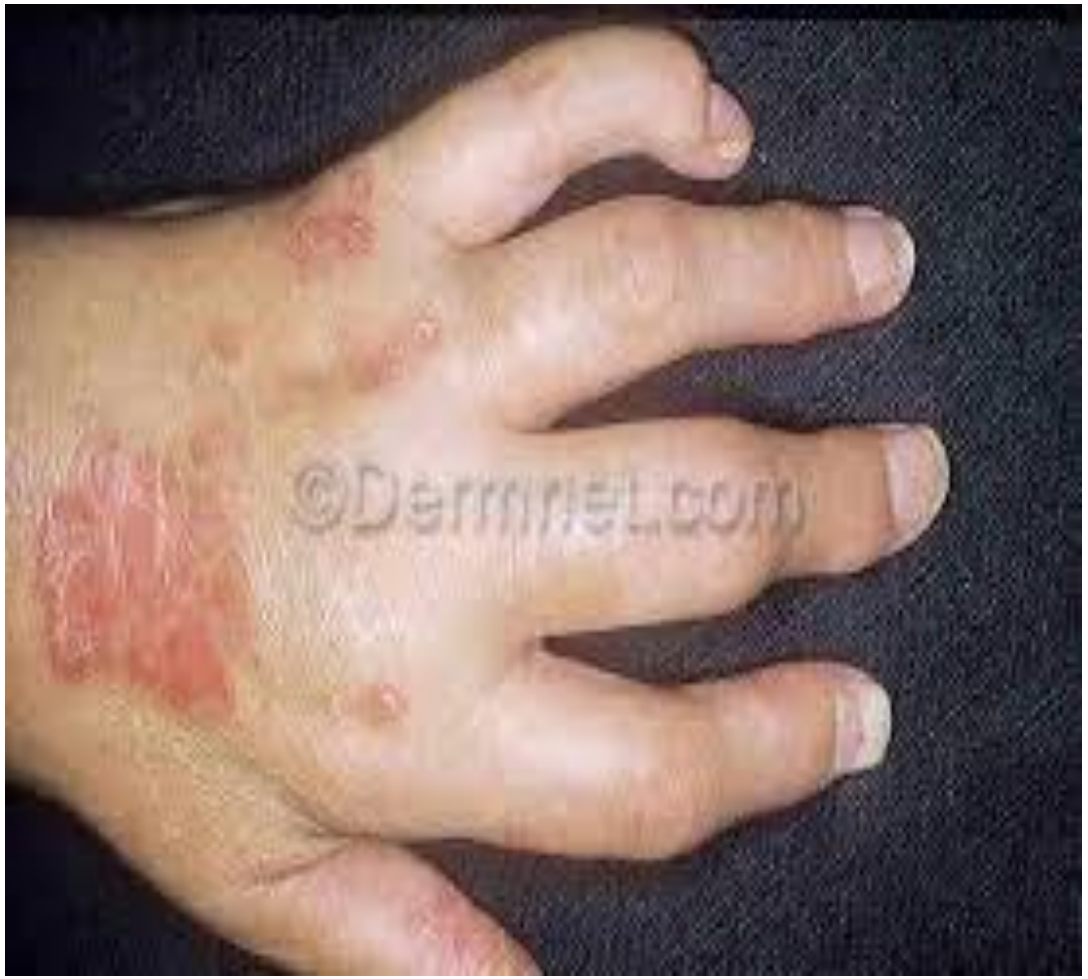
# OTHER TYPES

- Some doctors include two other, less common forms: enthesitis –related arthritis and psoriatic JIA. Enthesitis is an inflammation of the insertion points of the tendons. This form occurs most often in boys older than 8, characteristically causes back pain, and is linked to ankylosing spondylitis and inflammatory bowel disease. Psoriatic JIA occurs most often in girls, in conjunction with psoriasis, although joint problems may precede the skin manifestations by several years.

# DIAGNOSIS

- Diagnosis of JIA is difficult because joint pain in children can be from many other causes. There is no single test that can confirm the diagnosis and most physicians use a combination of blood tests, x rays and the clinical presentation to make an initial diagnosis of JIA. The blood tests measure antibodies and the rheumatoid factor. Unfortunately, the rheumatoid factor is not present in all children with JIA. Moreover, in some cases the blood work is somewhat normal. X rays are obtained to ensure that the joint pain is not from a fracture, cancer, infection or a congenital abnormality.
- In most cases, fluid from the joint is aspirated and analysed. This test often helps in making a diagnosis of JIA by ruling out other causes of joint pain.


© 2013 VisualDx


©DermNet.com

# DIFFERENTIAL DIAGNOSIS

- One possible differential diagnosis for JIA is Farber disease. Farber disease is a rare, fatal, genetic lysosomal disorder caused by a deficiency of the enzyme acid ceramidase . It has symptoms similar to JIA including swelling, stiffness and pain at the joints. These joint abnormalities are progressive and will develop during early infancy in a patient with Farber disease. Patients with Farber disease typically have subcutaneous nodules and a hoarse or weak voice due to growth of nodules on the larynx. A Farber disease diagnosis can be confirmed via gene sequencing.
- RAA
- Long fever duration
- Malign hemopathy

# TREATMENT

- JIA is best treated by a multidisciplinary team. The major emphasis of treatment for JIA is to help the child regain normal level of physical and social activities. This is accomplished with the use of physical therapy, pain management strategies and social support. Another emphasis of treatment is to control inflammation as well as extra-articular symptoms quickly. Doing so should help to reduce joint damage, and other symptoms, which will, help reduce levels of permanent damage leading to disability.
- There have been very beneficial advances in drug treatment over the last 20 years. Most children are treated with non-steroidal anti-inflammatory drugs and intra-articular corticosteroid injections. Methotrexate, a disease modifying anti-rheumatic drug (DMARD) is a powerful drug which helps suppress joint inflammation in the majority of JIA patients with polyarthritis (though less useful in systemic arthritis). Newer drugs have been developed recently, such as TNF alpha blockers, such as etanercept . There is no controlled evidence to support the use of alternative remedies such as specific dietary exclusions, homeopathic treatment or acupuncture. However, an increased consumption of omega-3 fatty acids proved to be beneficial in two small studies.

- Celecoxib has been found effective in one study.
- Other aspects of managing JIA include physical and occupational therapy. Therapists can recommend the best exercise and also make protective equipment. Moreover, the child may require the use of special supports, ambulatory devices or splints to help them ambulate and function normally.
- Surgery is only used to treat the most severe cases of JIA. In all cases, surgery is used to remove scars and improve joint function.
- Home remedies that may help JIA includes getting regular exercises to increase muscle strength and joint flexibility. Swimming is perhaps the best activity for all children with JIA. Stiffness and swelling can also be reduced with application of cold packs but a warm bath or shower can also improve joint mobility.

# THERAPY

- The best approach to treating a child with JIA involves a team of medical professionals including a rheumatologist, occupational therapist (OT), physical therapist (PT), nurse and social worker.[citation needed]
- The role of the OT/PT is to help children participate as fully and independently as possible in their daily activities or "occupations“ , by preventing psychological and physical dependency. The aim is to maximize quality of life, and minimize disruption to the child’s and family’s life. OTs work with children, their families and schools, to come up with an individualized plan which is based on the child’s condition, limitations, strengths and goals. This is accomplished by ongoing assessments of a child’s abilities and social functioning. The plan may include the use of a variety of assistive devices, such as splints, that help a person perform tasks. The plan may also involve changes to the home, encouraging use of uninvolved joints, as well as providing the child and their family with support and education about the disease and strategies for managing it. OT interventions will be changed depending on the progression and remission of JIA. Early OT involvement is essential. Interventions taught by an OT can help a child adapt and adjust to the challenges of JIA throughout the rest of their life.


Physical therapy
can help kids
juvenile arthritis.

# PROGNOSIS

- With proper therapy, some children do improve with time and lead normal lives. However, severe cases of JIA which are not treated promptly can lead to poor growth and worsening of joint function. In the last two decades, significant improvements have been made in treatment of JIA and most children can lead a decent quality of life. The prognosis of JIA depends on prompt recognition and treatment. Many children with JIA have gone on to play professional sports and have a variety of successful careers.

# REFERENCES


- Juvenile Arthritis: Arthritis Is A Children's Disease Too". Retrieved 2010-04-19.
- "Farber Disease". Retrieved 2014-01-21.
- "What is the outlook (prognosis) for children with arthritis?". Retrieved 2010-04-19.