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# Juvenile Idiopathic Arthritis

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## 1. WHAT IS JIA

### 1.1 What is it?

Juvenile idiopathic arthritis (JIA) is a chronic disease characterised by persistent joint inflammation; the typical signs of joint inflammation are pain, swelling and limitation of movement. "Idiopathic" means that we do not know the cause of the disease and "juvenile", in this case, means that the onset of the symptoms usually occurs before 16 years of age.

### 1.2 What does chronic disease mean?

A disease is said to be chronic when the condition persists – this means that the appropriate treatment does not necessarily bring about a cure of the condition but results in an improvement of symptoms and laboratory tests.
This also means that when the diagnosis is made, it is impossible to predict for how long that the child may be ill.

### 1.3 How frequent is it?

JIA is a relatively rare disease that affects about 1-2 individuals in every 1,000 children.

### 1.4 What are the causes of the disease?

Our immune system protects us from infections caused by various microbes such as viruses or bacteria. It is able to distinguish what is potentially foreign and harmful and should be destroyed from what

belongs to us.
Chronic arthritis is believed to be an abnormal response of our immune system, which to a degree, loses capacity to distinguish "foreign" from "self" cells and results in attack to its own body components leading to inflammation (e.g. of the joint lining). For this reason, diseases such as JIA are also called "autoimmune", meaning that the immune system reacts against the patient’s own body.
However, like most human chronic inflammatory diseases, the precise mechanisms that cause JIA are unknown.

## 1.5 Is it a hereditary disease?

JIA is not a hereditary disease since it cannot be transmitted directly from parents to their children. Nevertheless there are some genetic factors, with more as yet undiscovered, that predispose individuals to the disease. The scientific community agrees that JIA is the result of a combination of genetic predisposing factors and exposure to environmental influences (probably infections). However, even when there may be a genetic predisposition, it is very rare to have two children affected in the same family.

## 1.6 How is it diagnosed?

The diagnosis of JIA is based on the presence and persistence of arthritis and by careful exclusion of any other disease through the medical history, physical examination and laboratory tests.
JIA is when the disease starts before the age of 16, symptoms last for more than 6 weeks and all other diseases that may be responsible for arthritis have been ruled out.
The reason for this 6 week period is to allow the exclusion of other forms of transient arthritis such as those which may follow various infections. The term JIA includes all forms of persistent arthritis of unknown origin with onset in childhood.
JIA includes different forms of arthritis that have been identified (see below).
The diagnosis of JIA is therefore based on the presence and persistence of arthritis and on the careful exclusion of any other disease through evaluation of medical history, physical examination and laboratory tests.

## 1.7 What happens to the joints?

The synovial membrane is the thin inner lining of the joint capsule; in arthritis this becomes much thicker, filled with inflammatory cells and tissue resulting in an increased amount of synovial fluid inside the joint. This causes swelling, pain and limitation of movement. A characteristic feature of joint inflammation is joint stiffness which occurs after prolonged rest periods; it is therefore particularly pronounced in the morning (morning stiffness).

The child often tries to reduce pain by keeping the joint in a semi-flexed position; this position is called "antalgic" to emphasise the fact that it is aimed to reduce pain. If maintained for prolonged periods (usually more than 1 month), this abnormal position leads to the shortening (called contracture) of muscles and tendons and to the development of flexion (bent) deformity.

If not properly treated, joint inflammation may cause joint damage through two main mechanisms: the synovial membrane gets very thick, becoming boggy (with the formation of what is known as the synovial pannus) and through the release of various substances that provoke damage and loss of joint cartilage and bone. On x-rays this appears as holes in the bone that are called bone erosions. The prolonged maintenance of the antalgic position causes muscle atrophy (wasting and loss of muscle), stretching or retraction of muscles and soft tissues, leading to flexion deformity.