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Juvenile Idiopathic Arthritis

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2. DIFFERENT TYPES OF JIA

2.1 Are there different types of the disease?

There are several forms of JIA. They are mainly distinguished by the number of joints affected (e.g oligoarticular – less than 5 joints - or polyarticular JIA – 5 or more joints) and by the presence of additional symptoms such as fever, rash and others (see following paragraphs). Diagnosis of the different forms is made by observing the symptoms during the first 6 months of the disease. For this reason, they are also often referred to as onset-forms.

2.1.1 Systemic JIA

Systemic means that various organs of the body may be involved, in addition to the arthritis.
Systemic JIA is characterised by the presence of fever, rash and intense inflammation of various organs of the body that may appear before arthritis or during the course of arthritis. There is recurrent high fever and a rash that appears mainly during fever spikes. Other symptoms may include muscle pain, enlargement of liver, spleen or lymph nodes and inflammation of membranes around the heart (pericarditis) and lungs (pleuritis). Arthritis, usually involving 5 or more joints, may be present at disease onset or can appear several weeks later. The disease may affect boys and girls at any age, but it is especially common in toddlers and preschool children.
About half of patients have limited periods of fever and arthritis; these patients tend to have the best long-term prognosis. In the other half, fever often tends to subside, while arthritis becomes more important

and sometimes difficult to treat. In a minority of these patients, fever and arthritis persist together. Systemic JIA accounts for less than 10% of all JIA cases; it is typical of childhood and is seldom observed in adults.

2.1.2 Polyarticular JIA

Polyarticular JIA is characterised by the involvement of 5 or more joints during the first 6 months of the disease in the absence of fever. There are blood tests that evaluate Rheumatoid Factor (RF) that can distinguish between two types: RF negative and RF positive JIA.
RF positive polyarticular JIA: this form is very rare in children (less than 5% of all JIA patients). It is the equivalent of adult RF positive rheumatoid arthritis (the most common type of chronic arthritis in adults). It often causes symmetrical arthritis affecting initially mainly the small joints of the hands and feet and then extending to the other joints. It is much more common in females than in males and has onset usually after 10 years of age. It is often a severe form of arthritis.
RF negative polyarticular JIA: this form accounts for 15-20% of all JIA cases. It can affect children at any age. Any joint can be affected and usually both large and small joints are affected.
For both forms, the treatment must be planned early, as soon as the diagnosis is confirmed. It is believed that early and appropriate treatment leads to better results. Nevertheless, response to treatment is difficult to predict in its early stages. The response to treatment varies greatly from one child to another.

2.1.3 Oligoarticular JIA (persistent or extended)

Oligoarticular JIA is the most frequent JIA subtype, accounting for almost 50% of all cases. It is characterised by the presence, in the first 6 months of the disease, of fewer than 5 joints involved in the absence of systemic symptoms. It affects large joints (such as knees and ankles) asymmetrically. Sometimes only one joint is affected (monoarticular form). In some patients, the number of joints affected increases after the first 6 months of disease to 5 or more; this is called extended oligoarthritis. If the joints involved are less than 5 throughout the course of the disease then this is termed persistent oligoarthritis. Oligoarthritis usually has its onset before the age of 6 and is primarily observed in females. With timely and appropriate treatment, joint

prognosis is often good in patients in which the disease remains limited to a few joints; it is more variable in those patients who develop an extension of articular involvement into polyarthritis.
A significant proportion of patients may develop eye complications, such as the inflammation of the eye (anterior uveitis). Since the anterior part of the uvea is formed by the iris and the ciliary body, the complication is named either chronic iridocyclitis or chronic anterior uveitis. In JIA, this is a chronic condition which develops insidiously without causing any overt symptoms (like pain or redness). If unrecognized and left untreated, anterior uveitis progresses and may cause very serious damage to the eye. Early recognition of this complication is therefore of utmost importance. Because the eye does not become red and the child does not complain of blurred vision, anterior uveitis may not be noticed by parents or clinicians. Risk factors for developing uveitis are early onset of JIA and positive ANA (Anti-Nuclear Antibody).
It is therefore imperative for children at high risk to have regular eye checks by an ophthalmologist using a special appliance known as a slit-lamp. The frequency of examinations is usually every 3 months and should be maintained long-term.

2.1.4 Psoriatic arthritis

Psoriatic arthritis is characterised by the presence of arthritis associated with psoriasis. Psoriasis is a skin inflammatory disease with patches of scaling skin often located over elbows and knees. Sometimes only the nails are affected by psoriasis or there is a family history of psoriasis. The skin disease may precede or follow the onset of arthritis. Typical signs suggestive of this JIA subtype include swelling of the whole finger or toe (so called "sausage" finger or dactylitis) and nail changes (pitting). Presence of psoriasis in a first degree relative (a parent or sibling) can also occur. Chronic anterior uveitis may develop and therefore regular eye checks are recommended.
Disease outcome varies, as response to treatment may be different for skin and joint disease. If a child has arthritis in fewer than 5 joints the treatment is the same as for the oligoarticular type. If the child has more than 5 affected joints, the treatment is the same as for the polyarticular forms. The outcome may be related to the treatment response for both arthritis and psoriasis.

2.1.5 Arthritis associated with enthesitis

The most common manifestations are arthritis affecting mainly the large joints of the lower limbs and enthesitis. Enthesitis means inflammation of the "enthesis", the point of insertion of tendons over bones (the heel is an example of enthesis). Localised inflammation in this area is usually associated with intense pain. Most commonly enthesitis is located on the soles and the heels, where the Achilles tendons are inserted. Sometimes these patients develop acute anterior uveitis. Unlike uveitis with other JIA forms, it usually presents with red and watery eyes (lachrymation) and increased sensitivity to light. Most patients are positive for a laboratory test called HLA B27: this tests for a family predisposition to the disease. This form affects predominantly males and the arthritis usually begins after 6 years of age. The course of this form is variable. In some patients, the disease becomes quiescent after time, while in others it also spreads to the lower spine and to the joints attached to the pelvis, the sacroiliac joints, limiting the movements of back. Low back pain present in the mornings and associated with stiffness is highly suggestive of spinal joint inflammation. Indeed, this form resembles some spine diseases occurring in adults called ankylosing spondylitis.

2.2 What causes chronic iridocyclitis? Is there a relationship with arthritis?

Eye inflammation (iridocyclitis) is caused by an abnormal immune response against the eye (autoimmune). However, the precise mechanisms are unknown. This complication is mainly observed in patients with early onset JIA and a positive test for ANA.

The factors linking eye to articular disease are unknown. However, it is important to remember that arthritis and iridocyclitis may follow an independent course; periodic slit-lamp examinations must be continued even if the arthritis goes into remission as the eye inflammation can relapse without symptoms and even when the arthritis is better. The course of iridocyclitis is characterised by periodic flare-ups that are also independent from those of arthritis.

Iridocyclitis usually follows the onset of arthritis or may be detected at the same time as arthritis. More rarely it precedes arthritis. These are

usually the most unfortunate cases; since the disease is asymptomatic, late diagnosis may result in visual impairment.

2.3 Is the disease in children different from the disease in adults?

Mostly yes. The polyarticular RF positive form, which is responsible for about 70% of adult rheumatoid arthritis cases, accounts for less than 5% of cases of JIA. The oligoarticular form with early onset represents about 50% of JIA cases and is not seen in adults. Systemic arthritis is characteristic of children and is seldom observed in adults.