



# Juvenile Chronic Arthritis (JCA)

Englisch

During a meeting in Oslo in 1977 pediatric rheumatologists from Europe agreed on the term »juvenile chronic arthritis«. »Chronic« implies the risk of bone or cartilage destruction due to the long-standing inflammation, the term »juvenile« shall express the special aspects of childhood forms of arthritis. Thus, the formerly used term »juvenile rheumatoid arthritis« was replaced in Europe, while it is still used in the United States and some other parts of the world.

Presently there are efforts to establish an internationally accepted nomenclature and classification. An international committee of pediatric rheumatologists has suggested the term »juvenile idiopathic arthritis« and has newly defined the different subgroups. The new nomenclature and classification, however, must still prove practicable. We therefore stick to the name which has been valid in Europe up to now. The diagnosis »juvenile chronic arthritis« or »juvenile idiopathic arthritis« comprises different forms of childhood arthritis.

Though we still do not know the cause of the disease we can define several subgroups depending on age at onset, gender, number and pattern of arthritis, involvement of eyes or inner organs. Laboratory values can contribute to the diagnosis. Subgroup division occurs during the first six months of the disease.

## Subgroups

### Systemic juvenile chronic arthritis (SJCA)

This is the most severe form of juvenile arthritis. Systemic means that inner organs can be involved. The disease usually manifests in early childhood with high fever. A rheumatic rash as well as enlargement of liver, spleen and lymph nodes are common. Involvement of the heart muscle (myocardium) means a threatening complication.

### Rheumatoid-factor negative polyarthritis

This form of arthritis often presents with a lingering onset and can begin at any age. At least five joints must be involved, usually ten or more small and big joints are affected.

### Rheumatoid-factor positive arthritis

This disease resembles adult rheumatoid arthritis. Big and small joints can be affected, often symmetrically. Onset usually occurs during the second decade around puberty, girls predominate. The course is often more severe than in rheumatoid-factor negative polyarthritis with the risk of early joint destruction. As the name implies the rheumatoid factor is detected in the blood.

### Oligoarthritis type I and II

Oligoarthritis means that usually not more than 4 joints are involved at onset. We can define two forms of oligoarthritis. The early onset form manifests before school age and occurs predominantly in girls. They have a high risk for chronic eye disease (iridocyclitis). The other form concerns mainly boys who become ill after the age of eight to ten years. They often complain of heel pain. The disease can also affect the pelvic joints (sacroiliitis) or the spine.

### Psoriatic arthritis

This is diagnosed if a child suffers from psoriasis as well as arthritis. The course is usually oligoarticular. Involvement of a single digit (finger or toe) is a typical presentation. Psoriasis in close relatives is common.

### Rheumatic eye disease (iridocyclitis)

Iridocyclitis is a special problem of childhood arthritis. It is an inflammation of the anterior eye chamber and occurs in 10 to 20% of children, especially those with oligoarthritis. Onset and course are insidious since no signs of inflammation become visible and the child suffers no pain. Regular controls by the ophthalmologist are necessary who detects the inflammation by a slit lamp examination and can start effective treatment. There is a high risk for permanent damage with visual loss.



## Diagnosis

Arthritis is usually the leading symptom. It is characterized by joint swelling often combined with hyperthermia as well as functional impairment and pain. Joint pain is easily overlooked in children since they do not complain of overt arthritic pain. If parents are alert they may recognize an abnormal position of the affected joints. These are pain relieving positions which the children readily acquire to reduce joint pain. Depending on the joints involved the children may limp or have difficulties with eating, writing, dressing or other activities of daily life. They seem to be clumsy, refuse to walk, want to be carried or have a disturbed sleep. These subtle signs of arthritis are often overlooked and misunderstood. The children may be called lazy or naughty.

The diagnosis of a rheumatic disease and its classification can be difficult even for a doctor. There are few findings to prove the diagnosis. A lot of other diseases can go along with joint problems and it may be hard to differentiate them from a rheumatic disease. All these other diseases must be considered and excluded as best as possible, before establishing the diagnosis juvenile chronic arthritis.

Laboratory findings can give important hints. Altogether the value of lab results, however, are overestimated. Most often they only complete or confirm the clinical findings. They may be helpful to exclude other diseases. In the beginning x-rays also are mainly required for exclusion of non-rheumatic disorders. X-ray changes of arthritis can be expected only after months or even years from onset. For a definite diagnosis all findings must be put together like in a puzzle to receive a clear picture of the disease process.

## Therapy

The form of arthritis, the activity of the disease and a consequent treatment determine the course of juvenile chronic arthritis. In the long run many children can become free of symptoms. It is very important to avoid functional impairment and malpositions of the joints. An effective therapy comprises several tasks and all therapists must co-operate to achieve a good outcome. Medical treatment alone is insufficient. Intensive physiotherapy, occupational therapy, physical modalities and an individual social care are equally important.

### Medical therapy

Treatment of arthritis begins with so-called **non-steroidal antiinflammatory drugs (NSAIDs)**. These drugs reduce pain, inflammation and also fever. For children with a mild form of arthritis, especially when oligoarticular, this medication alone is sufficient for symptom relief. More severe forms require long-acting drugs which are also called **disease modifying anti-rheumatic drugs (DMARDs)**. The way of acting of these DMARDs is not yet understood in detail.

It is however clear that they act on the immune system and can regulate immunologic disturbances which have led to the rheumatic process. Long-acting drugs usually require several weeks or even months to show efficacy. It is therefore important to take the medication regularly and consequently for a longer period of time. Introduction of a DMARD therapy requires close clinical and laboratory controls to notice possible side effects early enough. All side effects are usually reversible when the drug is reduced or discontinued. If the disease has come into remission under medical treatment the drugs should be tapered very slowly. Early interruption of therapy may result in a bout of the disease.

**Corticosteroids** should be used with care in children due to their severe long-term side effects. Systemic application especially with high doses must be restricted to severely ill children. Local treatment for eyes and joints predominates. Eye drops or ointments containing corticosteroids are urgent for therapy of iridocyclitis. Ocular inflammation may rapidly lead to eye defects and therefore requires early and prompt reduction of the inflammatory process.

Injection of corticosteroids into a joint is very effective. This form of local therapy is mainly indicated for oligoarticular disease or for children with polyarthritis if arthritis of single joints predominates. The intra-articular injection of corticosteroids can resolve joint inflammation within days. The good result is often kept

for a long time. In case of a further bout of arthritis the injection can be repeated.

New forms of therapy like the so-called Biologicals (for example anti-TNF $\alpha$ - therapy), new immunosuppressants (like Leflunomide) or even autologous stem cell transplantation are still in a phase of trial for children with arthritis. First promising results must be calculated against still unknown possible long-term side effects.

### Physiotherapy

Each child with arthritis requires regular physiotherapeutic assessment. Impairment of joint function and pain-relieving positions must be treated as early as possible. Physiotherapy concentrates on restoration of full joint mobility and prevention of malpositions. Each child must be treated according to his or her age. Even very small children can receive effective physiotherapy.

Since treatment usually must be kept over a longer period of time it is important to establish an atmosphere of trust and friendship between child, parent and therapist. The child's initial fear of pain will cause him or her to refuse therapy and requires the therapist's full empathy. Early recognition of the pain threshold is the basis for the child's trust. Physiotherapy must never be painful. If possible parents should be integrated into the treatment. They can be trained to do some of the exercises at home.

### Local cold and hot packs

A painful swollen joint usually means inflammation. This can be treated with local cold. Ice packs are practical to use and should be applied three or even four times a day for 10 to 15 minutes. For some children ice packs feel unpleasant. They might prefer compresses with alcohol or other cooling substances. If inflammation has subsided but has left functional impairment local warmth is indicated. Warm fomentations relax muscles and increase the blood flow.

### Occupational therapy

Functional therapy trains joint function with playful activities or handicraft. Special exercises when working with clay or other materials can activate muscles which counteract the malpositions. Individual splints are adapted to stabilize affected wrist joints. These joints allow a proper hand function while the wrist is kept in a physiologic good position. Joint protection means to avoid over- or misloading of affected joints. In occupational therapy the children learn proper use of muscle forces to protect their joints during daily activities.

### Additional Aids

Devices to avoid loading of affected leg joints are indicated according to the child's age. Small children can ride on wooden horses or tricycles. For older children we have developed a scooter with a saddle, but an ordinary bicycle is also appropriate. The children can use these vehicles for transportation and also train their physical endurance. Older children and adolescents can use crutches for shorter distances at home or at school. However, if shoulder, elbow or wrist joints are also affected the use of crutches is limited.

Doctor, physiotherapist and occupational therapist decide together whenever functional wrist splints or positioning splints become necessary. Each splint must be adapted individually. Therapists must closely control the fabrication. The splints must fit perfectly and keep the joint in proper position. A lot of experience and good co-operation between doctor, therapist and orthopedic technician are of utmost importance. Splints which are not adapted to the actual joint situation may be harmful.

## Social Care

Every day life with a child who suffers from chronic arthritis is often difficult to master. It requires a lot of inventive faculty to integrate all regular therapeutic demands into the course of a day. Joint protection, joint relieve and therapy must become part of life. Social care deals with these aspects and tries to find solutions considering the individual family situation. Part of the task concentrates on practical advices. Parents for example can order a double set of school-books to avoid carrying heavy loads on the way to and from school. It may also be possible to transfer the child's classroom to the first floor to prevent climbing stairs. Children with affected wrist or finger joints often have problems with writing. They should receive more time for classwork.

Adolescents require early vocational guidance. Together with patient, family and therapists a vocational profile is established which considers the patient's personal desires within the spectrum of his potentialities and limitations. Depending on disease severity financial support and compensation can be applied for. There exist legal claims for such support to compensate for disease induced disadvantages and to improve integration of the handicapped person into community life.

Social care, however, exceeds such plain advisory tasks. Apart from practical support in daily life it must also deal with psychologic aspects which usually consider all family members in their own way. Mothers are often

torn between the demands of daily life, the care for the ill child as well as for their healthy children who often feel neglected. Education of the children is complicated by the disease. Parents are often in doubt when to follow the child's wants and demands and when to set limits. Especially those parents who have intensely cared for their chronically ill child over many years will have difficulties to find the necessary distance when the child strives after independency during adolescence.

All these aspects may cause parents to neglect their own needs. Some of them become absorbed in work to forget about their worries, or they renounce spare time for themselves and their partner. Intensive talks offer the possibility to express one's worries and problems within a sheltered atmosphere.

Social care within the specialized centers is supported by the work of parent associations which exist in all parts of Germany under the roof of the German Rheuma-Liga. They offer support, information, training and advice. Interested parents can join an association close to their home, thus receiving possibilities for exchange with other parents which often means an important help to master daily life with a chronically ill child.

If you need any further information or want to get into contact with one of our self-help-groups please write – email – phone us:

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Herausgeber:  
Deutsche Rheuma-Liga Bundesverband e.V.

1. Auflage 2002 – 5.000 Exemplare  
Drucknummer: MB 2.1/BV/11/2002

Mit freundlicher Unterstützung der Selbsthilfe-Fördergemeinschaft der Ersatzkassen.

