

Oligoarticular onset juvenile idiopathic arthritis as the most common cause of disability of children and young adults



Zbigniew Żuber

Department of Pediatrics, Faculty of Medicine and Health Sciences, Andrzej Frycz Modrzewski Krakow University, Poland
Ward for Older Children with Neurology and Rheumatology Subdivision, St. Louis Regional Specialised Children's Hospital in Krakow, Poland

Juvenile idiopathic arthritis (JIA) is not a homogeneous disease entity, but a group of diseases with several distinct clinical forms and different etiopathogenesis [1].

At the age of 0–6 years, the oligoarticular form of juvenile idiopathic arthritis (o-JIA) accounts for up to approximately 70% of all JIA cases, but, as the age at which the disease develops progresses, the profile of the JIA patients changes. The clinical picture of JIA becomes more diverse – at the age 12–16 years o-JIA accounts for less than 40% of all new JIA diagnoses [2].

Oligoarthritides JIA is a seemingly benign form of the disease, with asymmetrical involvement of 1 to 4 joints, mainly of the lower limbs. Approximately 80% of patients have a single joint or both knee joints' involvement; less often hip or ankle joints are affected, and the involvement of other peripheral joints is also possible [1–4]. The delay in establishing JIA diagnosis results from the fact that in children problems with lower limb joints are often considered to be of post-traumatic etiology, because „every child may have sustained a trauma”. Long-term treatment of „post-traumatic” joints, however, does not bring significant improvement; multiple joint punctures – with or without glucocorticosteroids (GCs) – yield only temporary improvement. Joints are often immobilized in a plaster dressing. Sometimes improperly targeted rehabilitation procedures are performed or even surgical procedures are performed as successive elements of treatment.

In some cases such therapy brings partial improvement, but it can also result in permanent damage, contractures, muscle atrophy or deformities. Diagnostic problems with JIA are common. While this is an obvious truth for rheumatologists, physicians of other specialties are often misled by the fact that in JIA basic biochemi-

cal test results may stay within the normal range [5, 6]. Intuitively this phenomenon seems to contradict the fact that we are dealing with an inflammatory process. How can a child's disease be considered „inflammation”, when the erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP) levels are normal and the synovial fluid assessment is within established standards? [4–6].

The problem lies in the slow intensification of o-JIA symptoms. The swelling of the joint masks the existing inflammation. General symptoms – such as fatigue and apathy – dominate, often being mistakenly considered to be related to infections [4, 5]. The problem can also be erroneously identified as the asymmetry of the body structure, gait disturbances such as tripping, falling or limping, which leads doctors to consider trauma as the cause of such abnormalities.

Oligoarthritides JIA is a systemic disease, affecting the normal development of the child, the growth, mass and proportions of the body. Growth disturbances are a huge emotional problem for the patients, worsening the social functioning and lowering the quality of life. The child's development should be assessed systematically and precisely by performing two simple, easy to perform, measurements: body weight and height. The results of these measurements should be plotted on a centile grid. Unexplained disturbances of growth and deviations from the population age norm on centile grids should lead to a thorough assessment of the musculoskeletal system. The inflammatory process, gait disturbances and developmental disturbances that continue may remain unnoticeable, even for the patient's close family.

Recent advances of the knowledge considering JIA and progress of imaging capabilities allow physicians

Address for correspondence:

Zbigniew Żuber, Department of Pediatrics, Faculty of Medicine and Health Sciences, Andrzej Frycz Modrzewski Kraków University, 1 Herlinga-Grudzińskiego St., 30-705 Kraków, Poland, e-mail: zbyszekzuber@interia.pl

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to quickly diagnose inflammatory changes of the joints. The general availability of ultrasound (US) and the increasing availability of magnetic resonance imaging (MRI) allow us to obtain information about the ongoing inflammatory process in the joint [7–9]. Ultrasound examination and assessment of hip joints seem particularly important. However, in the assessment with ultrasound the skills and experience presented by the person performing the examination are vital for its quality. These joints, due to their construction and location, are difficult to fully assess in a physical examination. Involvement of the hip joints is considered to be a „malignant localization” of JIA, being a predictive factor of poor prognosis in JIA. Temporomandibular joints are considered as another malicious location of changes. In their case MRI is the diagnostic imaging method of choice.

Although in physical examination of children it is the basic rule to examine both symmetrical joints, the assessment of only one joint, without a comparative assessment of the other one, is a common error. The most common non-joint general symptom in JIA is uveitis (occurring in 75% of patients with JIA, and in 20–30% of o-JIA patients). This is a serious complication, due to the initially asymptomatic course and a risk of permanent damage to eye structures (affecting up to 60% of patients with eye complications) [10] with visual impairment and even blindness.

It has also been proved that the presence of anti-nuclear antibodies (ANA), age under 6 years, and female gender are bad prognostic factors of the ocular involvement in the course of JIA.

As has already been mentioned above, an important problem in o-JIA is growth disorders.

In the active phase of the disease, the inhibition of growth occurs particularly frequently, while in the period of decline of the disease activity and the use of lower doses of GCs, we observe the effect of an accelerating (overtaxing) growth rate – a so-called „catch-up”. Despite the availability of new therapeutic options, still about 10–20% of patients with severe forms of the disease show permanent growth disorders leading to short stature. The knowledge of this can be particularly useful for pediatricians performing periodic assessment of the children’s health. The occurrence of growth disorders should lead the pediatrician to perform a detailed interview with the child’s carers, regarding changes in the osteoarticular system in particular. There is no doubt that the early diagnosis of the disease and the prompt implementation of an appropriate personalized therapy in order to achieve clinical remission as soon as possible is particularly important. It has been proven that the largest structural changes in active inflamed joints occur in the first years of the disease.

A diagnostic delay, as described above, is a crucial obstacle for the introduction of proper treatment. The therapy is based on the recommendations of the American College of Rheumatology (ACR, 2013 and 2019) [11, 12]. According to them, the treatment starts with the initial use of NSAIDs during the diagnosis period (6 weeks). In addition, administration of GCs is recommended on delivery. After the diagnosis is proven, it is advisable to include disease-modifying antirheumatic drugs (DMARDs) in the therapy – mainly methotrexate (MTX) as the gold standard. In subsequent stages of the treatment, if previous medications are ineffective, subsequent DMARDs can be used. In the absence of remission, despite intensification of the treatment, biological drugs are recommended [9, 11–13].

In Poland, the initial stage of the treatment is problematic as a result of the fact that the summaries of product characteristics of drugs used in o-JIA (such as methotrexate), which were compiled for the purposes of the registration of those medications, do not literally list JIA treatment as one of indications for the use of those drugs (only the polyarthritis form being included in the indications).

Another problem in JIA involves the so-called malignant locations of the changes and uveitis. In cases in which one of those features occurs, it is necessary to modify the rules of conduct. The treatment must be more aggressive, with the goal of achieving remission – the improvement of the indicator ACR Pediatrics playing a marginal role in such cases [13, 14]. It should be remembered that the disease may result in disability, even as serious as the loss of sight in the case of eye complications.

An important issue in the treatment of o-JIA is the management of the proper conventional DMARD treatment, mainly with MTX. Often too low drug doses are used or there are unjustified time gaps in the therapy. This results in DMARD therapy being considered ineffective, which may lead to the premature introduction of biological treatment, without full exploitation of the potential of the therapy with DMARDs.

The publicly funded programs of biological drug use in active rheumatoid arthritis (RA) and JIA (program B.33) do not cover the whole population of o-JIA patients [13]. Especially when one joint is involved, the patient’s qualification for standard biological treatment in accordance with The National Health Fund (NHF) program is difficult, and the presence of negative prognostic factors is vital for obtaining the approval for treatment from the NHF.

Biological treatment depends on the o-JIA form and whether the diagnosis is expected to evolve in the future towards RA, psoriatic arthritis (PsA), ankylosing spondylitis (AS) or spondyloarthritis (SpA).

Coexistence of autoimmune diseases such as idiopathic inflammatory bowel diseases, diabetes type 1, autoimmune thyroiditis and other rheumatic connective tissue diseases (RCTD) is an issue that needs to be discussed separately.

The patient in the remission period requires systematic monitoring with the assessment of the course of the disease. The recurrence of the disease process is possible at any age. Currently, a lot of attention is devoted to the transition of a patient with o-JIA from pediatric care to the care system provided for adults. The work on this subject is still ongoing.

Conclusions

The problem of o-JIA as a specific and separate „childhood” form of rheumatic disease and its comparison with the „adult” disease entities still carries a lot of unanswered questions.

Thus this article can be summarized by the following key points:

1. Changes in a single joint are most often wrongly associated with an alleged injury, leading to a delay in the diagnosis of o-JIA and to the inappropriate treatment of children.
2. Oligoarthritis JIA yields high risk of uveitis and risk of disability due to blindness.
3. The hip and temporomandibular joints are considered malicious locations of the o-JIA changes.
4. Oligoarthritis JIA leads to developmental disorders, including short stature.
5. The knowledge of this form of JIA is sparse not only among pediatricians and orthopedic doctors, but also in the rheumatologist community.

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