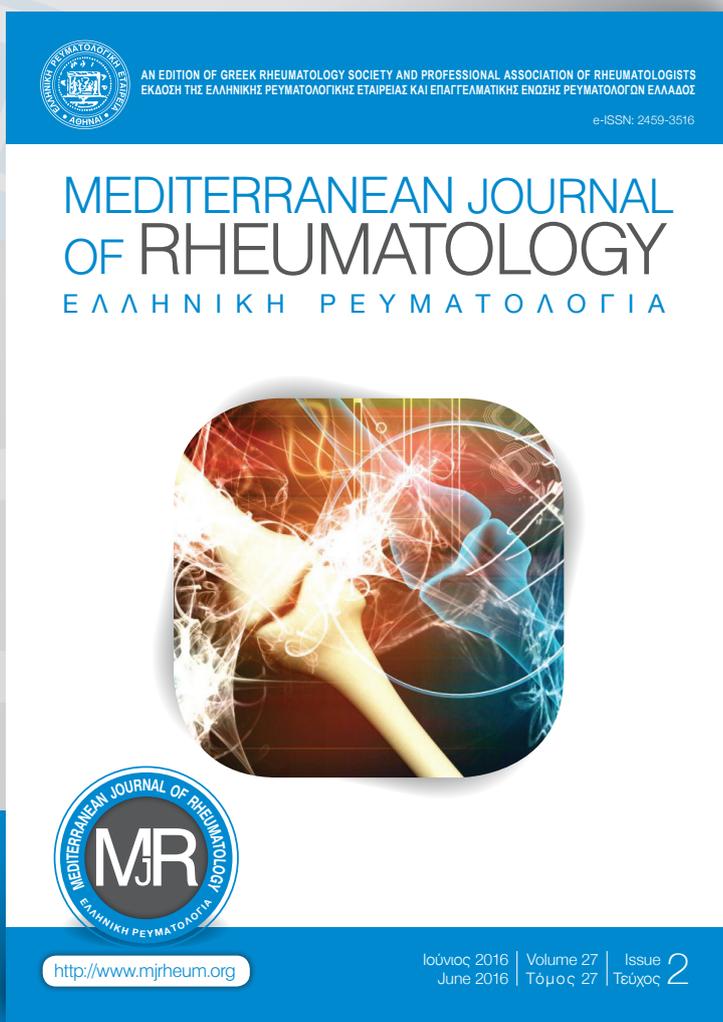

Transition of the patient with Juvenile Idiopathic Arthritis

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Mediterr J Rheumatol 2016; 27(2):63-8



E-ISSN: 2459-3516



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ABSTRACT

Juvenile Idiopathic Arthritis (JIA) is the most common pediatric rheumatic disease; when disease activity persists into adulthood, it may lead to functional disability. The transition of adolescents and young adults with JIA to an adult Rheumatology clinic is not a matter of transfer: it is a coordinated and targeted transition and integration from child/family-focused health systems to those for adults. Transition is an active, dynamic and flexible process, adapted appropriately to the patient's age, mental and physical development in respect to his/her culture. It aims to empower the patient to undertake the responsibilities of their own personal health and lifestyle, and above all, to guarantee the optimum lifelong functionality. A prerequisite for successful transition is the patient's training in development of their skills regarding communication, decision making, and self-health management. The transition process involves parents, patients and health caregivers (pediatric/adult) and, in addition, educational /social services. Patient management is holistic, physical and psychosocial, with an extension to areas of education and vocational rehabilitation and requires the full cooperation of parents, patients and health professionals at all stages. The steps for final transition take place only when the patient has completed his/her developmental process and has a proven ability for self-healthcare management, unsupported by the family.

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Mediterr J Rheumatol 2016; 27(2):63-8

<https://doi.org/10.31138/mjr.27.2.63>

Keywords: Juvenile Idiopathic Arthritis, transition-integration, young people with arthritis.

Juvenile Idiopathic Arthritis: demographic characteristics-outcome

Juvenile Idiopathic Arthritis (JIA) is the most common rheumatic disease of childhood. Its incidence is estimated to be 2-20 new cases/100,000 children annually while prevalence ranges from 16 to 150 per 100,000 children.^{1,2} It is a chronic arthritis occurring before the age of sixteen, with great heterogeneity in clinical presentation, course, and outcome. Recent data suggest that JIA is not such a "benign" disease as previously thought, as a significant proportion of patients suffer from persistent disease activity in adulthood, leading to functional disability.^{3,4}

In particular, long-term follow-up studies of patients with JIA from different countries, published after 1994, revealed that the percentage of adolescents with disease in remission varies from 35% to 60%, thus indicating that about half of those patients may have a various degree of disease activity entering adulthood (18 years of age in Greece).^{5,6} A recent study of 102 patients with JIA from Northern Greece demonstrated that after a follow-up period of approximately 17 years, 49 (48%) had inactive disease. Furthermore, 30% of these patients had moderate to severe disability on the last follow-up, while 18.7% reported impairment in their psychosocial status.^{7,8}

Bringing this all together, there is an emerging need to follow-up on these patients in adult Rheumatology clinics in order to continue providing comprehensive medical care through a specialized setting.

TRANSITION OF THE ADOLESCENT WITH JIA: DEFINITION AND PURPOSE

The transition of adolescents/young adults with JIA to an adult Rheumatology clinic is not only a matter of a simple transferal: it is a coordinated action aiming to integrate the patient from child/family focused health systems to those for adults. It aims to empower the patient to undertake the responsibilities of personal health and lifestyle and mainly maximize lifelong functionality and potential by providing high quality health care services, which continue without interruption after adulthood.⁹⁻¹¹

PROBLEMS THAT AN ADULT RHEUMATOLOGIST MAY FACE WITH JIA PATIENTS

The adult Rheumatologist will face a JIA patient with consequences that affect both physical health and psychosocial status.

Physical problems primarily include growth disorders, either localized - such as overgrowth of the affected limb with associated length discrepancy - or generalized, such as delayed physical growth. Other problems include: disorders of sex maturity such as delayed ad-

olescence, functional remnants leading to physical disability, complications of uveitis (such as cataract, glaucoma, vision loss), osteoporosis and/or osteonecrosis secondary to the disease or prolonged use of steroids as well as the effect of the long-term use of immunomodulatory drugs such as frequent infections.²⁻⁷

Regarding psychosocial problems, it is worth noting patients' insufficient autonomy due to functional disease residues, which is reflected in professional activities, every day and social life. Such issues may require modifications and adjustments during disease flares. On the other hand, the psychological burden, expressed as increased anxiety or depression, is not rare-

Table 1. The potential impact of JIA which adult rheumatologists take into consideration.

Physical health

Impaired growth (generalized or localized)

- Disorder of gender maturation
- Functional residues-physical disability
- Uveitis
- Osteoporosis-Osteonecrosis
- Immunosuppression

Psychosocial health

- Insufficient autonomy
- Anxiety/depression
- Occupational targets/potentials in relation to JIA
- Sociability

ly encountered in adolescents with JIA (**Table 1**).^{8,12,13} In addition to the problems related to the disease, an adult Rheumatologist has to deal with an adolescent or young adult who is invited to participate in a different model of patient-physician collaboration - changing from family-oriented to patient-oriented - and is expected to face the new environment with awe and concern. Furthermore, the highly critical and revolutionary spirit of this age group can lead to disputing of physicians' instructions or even non-compliance to treatment and regular follow-up. Thus, the physician is challenged to successfully manage these individuals, showing patience and goodwill, consenting and allowing enough time for dialogue.¹⁴

PREPARING FOR THE TRANSITION

Although the most appropriate period to start the formal process of transition is adolescence, the concept of transition can be introduced earlier in discussions with the family so as to allow sufficient time for parents to get used to the idea. The process of transition is a family process. The family plays a crucial role and

should be supportive and encouraging when considering the future expectations for their child's education, employment, and independent living in relation to the disease. The "vision" for a future where the child will assume alone the management of his/her own medical care, from early adulthood, will be the motivation for proper preparation.¹⁵

During the follow-up period and with age, the patient should be increasingly involved in direct discussions concerning the diagnosis, treatment, long-term effects of the disease and medications, as well as restrictions on sports. In adolescence, discussions will focus mainly on the risks of non-compliance with monitoring and treatment, risky behaviors, such as tattooing, smoking, alcohol, and drug addiction, versus the benefits of healthy living. Furthermore, relevant information must be given on specific topics, such as infections, vaccinations, and the impact of JIA on sexual behavior and reproduction.¹⁵⁻¹⁷

The patient's preparation to manage individual health is an ongoing process which should also start from early childhood and continue into adulthood, in order to complete the development of skills which are necessary for self-care. This allows independent physical and psychosocial living, which is reflected in everyday life, travel, management of leisure time, sociability/friendship, and education or employment.¹⁵⁻¹⁸

Furthermore, the patient should be able to comply with the physician's instructions concerning follow-up and treatment and to manage situations such as pain, fatigue, and emergencies or other co-morbid conditions (**Table 2**).¹⁹

TRANSITION TIME SCHEDULE

Transition timing includes specific steps:

Step 1: Talk with the parents of a child who has a confirmed diagnosis of JIA about the course and disease duration. Where there is interest in online information, trusted sources are recommended, such as PRINTO (Pediatric Rheumatology International Trials Organization, <http://www.printo.it/pediatric-rheumatology/>), where they can find information about the disease in their own language.

Step 2: Gradual training for JIA (answering questions regarding chronicity, complications, outcome of the disease and training in the quality of life through the CHAQ [Childhood Health Assessment Questionnaire] and JAMAR [Juvenile Arthritis Multidimensional Assessment Report]) questionnaires. Frequent visits, allocated time by the attending physician, and the development of a trust relationship between the doctor and the family are a prerequisite.²⁰

Step 3: Begin discussion and prompt parents to schedule training and occupation of their child in relation to JIA. Moreover, cooperation with other specialists (psychologists, occupational counselors) is encouraged in order to ensure the most appropriate option for each patient regarding education on future employment.

Step 4: Ongoing patient preparation/education to develop self-care skills and independent living. To our center's experience, participating in group events away from home, such as summer camps with the supervision of a physician and a specialist physiotherapist, is particularly useful.

Table 2. 'Handbook' of knowledge and skills for a successful JIA adolescent-young adult's transition.

I. The Knowledge for transition

- Awareness of the disease/treatment and their impact on the patient
- Risks of non-compliance
- Specific issues, e.g., infections, vaccinations
- Insurance coverage
- Healthy living/lifestyle (tattooing, smoking, alcohol, drug addiction)
- Impact of addiction (alcohol, substances) on disease-treatment
- JIA impact on sexual health and reproduction
- Academic training/inclinations

II. Skills for transition

- Confidence in the management of individual health in dependently of parents
- Ability to seek health information
- Autonomous access to follow ups, collaboration with new medical team
- Compliance with follow ups/treatments
- Management skills of pain/stress/emergencies
- Physical and psychosocial independent living (everyday, transportation, leisure time management, sociability/friendship, education-work)

The cooperation of the patient/family, doctors and the health system is necessary. Continuous and coordinated communication between patients, families and children/adult Rheumatologists is needed so as to facilitate the transition and transfer.²⁰

The transition plan should be flexible in terms of timing and it should be tailored to the developmental and psychosocial status of each candidate. The timing of the transition should be accompanied by emotional and developmental maturity. The adolescent should be involved in transition planning and should be encouraged to share his own concerns about quality of life issues, such as physical limitations, social relationships, etc. with doctors who will conduct his follow-up in the future. Transition should take place only after the new patient has been able to complete his own developmental growth and has proven his ability to manage health care independently from the family and pediatrician. An important factor is his/her access to insurance coverage.²³

When designing the transition program, academic performance of adolescents, any learning disabilities, behavioral problems, attention concentration, hyperactivity and other psychological issues should be taken into account as they may interfere with the ability to assume responsibility for self-care.

Certainly the transition to an adult Rheumatologist should be avoided during periods of flares or psychosocial dysfunction.²¹⁻²²

It is very helpful for young adults with JIA already integrated in an adult Rheumatology clinic to be involved in welcoming new patients who are still in the transition process.

OCCUPATIONAL ORIENTATION IN THE ADOLESCENT WITH JIA

The adolescent is guided early on to choose an occupation compatible with his/her physical functionality and spiritual potential. For example, the profession of a jeweler is not compatible with the polyarticular course of arthritis which affects the hands, or the profession of a salesman in a patient with affected lower limbs.²⁴⁻²⁵

Parents and patients should be informed regarding disease limitations which may affect the ability of work as patients grow up. Such training is intended to direct the adolescent in choosing a full-time career, which they will be able to maintain throughout their life.²⁴

Studies have shown that patients with JIA have similar academic performance to healthy peers. On the other hand, higher unemployment, discrimination in the workplace, difficulty in maintaining the employment during relapses and even job change due to physical dysfunction have been noted.²⁴

MEDICAL REPORT OF A PATIENT WITH JIA

The medical report of a patient with should include a complete and comprehensive medical history, which necessarily accompanies the transition. The age and disease onset type is reported. Then, after the first 6 months, the JIA phenotype is recorded (affected joints and periarticular structures), the laboratory profile - ANA, RF, HLA - as well as ocular-involving uveitis. The medication and the patient's response is also described in accordance to the specific tool for disease improvement, ACR pedi. The disease course is then described, including instances of flares and their management. Finally, during the last visit, the patient's condition is assessed, implementing the international validated tools for the quantitative assessment of the disease activity (JADAS [Juvenile Arthritis Disease Activity Score]), outcomes (ACR pedi), damage (JADI [Juvenile Arthritis Damage Index]) and the patient's functional ability (CHAQ, JAMAR).

Moreover, other co-morbidities are reported with their impact on the patient's health as well as the patient's and family's degree of compliance and cooperation. It is also helpful to describe the patient's psychosocial profile and personality characteristics (concerns/phobias, confidence or insecurity, preferences) as well as his/her academic achievement and professional orientation.^{26,27}

PEOPLE INVOLVED IN THE TRANSITION

According to international literature, the involvement of parents and the child, as well as the adult and pediatric Rheumatologist simultaneously in the transition process is initially mandatory. The presence of the family doctor, pediatrician and/or physician who will have undertaken the patient's support of general health issues (vaccinations, blood pressure control, cholesterol, tobacco use, alcohol and other substances, nutritional counseling, contraception, sexuality) is also very helpful. The adult and pediatric Rheumatologist will conduct a discussion about the patient based on the medical report which has been forwarded to the adult Rheumatologist. The involvement of other health professionals (specialized nurses, physiotherapists) who will undertake the coordination of the transition will facilitate the process. The invitation of educational and vocational representatives could be very effective in the transition process as it facilitates the patient's decision on professional orientation. Finally, social services (social workers) and support groups (associations relative to rheumatic diseases) should accompany this process in order to smoothen the integration obstacles of the new patient into the adult group.^{26,27}

EVALUATION OF THE TRANSITION PROCESS

The provision of an evaluation questionnaire, which includes questions relevant to the evaluation of patient and staff preparation in order to achieve the transition, as well as the satisfaction of this process (overall satisfaction score), which will be handed out to all involved health professionals and patients, is very important. It will lead to a revision of the transition process and prevent adverse cooperation between the patient and staff in the new medical environment. Thus through this evaluation process the optimal transition for each JIA patient can be achieved and adapted to the existing conditions, which will lead to a better disease outcome.²⁹⁻³³

In conclusion, the transition of an adolescent with JIA to an adult Rheumatologist is an active, dynamic, and flexible process, adapted appropriately to age, mental and physical development, and the cultural level of the patient. It is characterized by a holistic approach to the patient's physical and psychosocial status, with an extension to areas of education and vocational rehabilitation. It should be timely, focused on the future, and patient-centered. Prerequisites include skills training in communication, decision making, and management of individual health. Under these circumstances, the transition can be successful and contribute to optimal disease outcome.^{34,35}

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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