

Patients with juvenile idiopathic arthritis become adults: the role of transitional care

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ABSTRACT

Most juvenile idiopathic arthritis (JIA) patients need to attend adult rheumatology centres to continue the clinical management of their disease and to receive adequate long-term treatment. Transition from the paediatric to the adult health care team is a critical moment in the clinical history of these patients, but unfortunately, about 50% of the transfer processes to adult rheumatology are not successful, putting these patients at high risk of unfavourable outcomes. There are several obstacles to the success of transitional care for JIA patients, such as the absence of specific criteria for the assessment of disease activity, the lack of specific treatment recommendations for JIA adult patients, the poor adolescent-specific training for adult rheumatologists, and the shortage of resources. The improvement in the transition process in medical care has become a priority in many health care systems, but not many studies evaluating transition models, and common methodologies for measuring transition outcomes are available. The aim of this review is to identify and describe the models of transitional care in JIA, providing insights and recommendations to develop effective transitional care models in this disease.

Introduction

Juvenile idiopathic arthritis (JIA) is a leading cause of acquired functional disability in children and adolescents (1). Over the past few years, functional outcomes have markedly improved, but the long-term physical, psychological and socioeconomic burden of JIA is still substantial. Indeed, a sizeable proportion of young adults with this disease have limited functional abilities. Studies consistently show that JIA is not only associated with chronic disa-

bility and restricted participation in social activities, but also with significant morbidity due to articular and extra-articular manifestations, and even with premature mortality (2).

The course of JIA often continues into adulthood; approximately half of the patients have persistently active disease or experience disease flares after adolescence, and many of them still require anti-rheumatic therapy (3). A number of these patients need to attend adult rheumatology centres in order to be monitored and to receive adequate long-term treatment (4).

Juvenile idiopathic arthritis

The term juvenile idiopathic arthritis encompasses a group of heterogeneous forms of chronic arthritis of unknown aetiology that have their onset before the age of 16 years. With an incidence of 1:10,000 and a prevalence of 0.1–0.4%, it is the most common childhood rheumatic disease (1, 5).

The current International League of Associations for Rheumatology (ILAR) classification categorises JIA into 7 subtypes, based on the clinical and laboratory characteristics observed in the first 6 months after disease onset (6) (Table I).

Oligoarthritis, which is defined by the involvement of 4 or less joints in the first 6 months, is the most common subtype of JIA ($\geq 50\%$ of cases). It generally affects young children (more often females), and usually involves large joints, particularly the knee and the ankle, and occasionally the wrist and the elbow, but rarely the hip. Extension of arthritis to 5 or more joints (so-called extended oligoarthritis) after the first 6 months of disease occurs more often in patients with involvement of the upper limb joints and high ESR at onset, and has a more severe prognosis in terms of

Table I. ILAR classification of JIA.

ILAR juvenile idiopathic arthritis classification
Systemic (4-17%)
Oligoarthritis (50%)
persistent
extended
Polyarthritis (rheumatoid factor negative) (15-25%)
Polyarthritis (rheumatoid factor positive) (2-7%)
Psoriatic arthritis (7-10%)
Enthesitis-related arthritis (10-20%)
Undifferentiated arthritis (15%)

risk of joint damage. Up to 30% of patients with JIA will develop uveitis, and the most important predictive factor for its occurrence is ANA positivity (7).

Polyarticular JIA is defined by the involvement of 5 or more joints during the first 6 months after presentation, with a frequently symmetrical distribution. Rheumatoid factor (RF)-positive polyarthritis is the paediatric equivalent of adult rheumatoid factor-positive rheumatoid arthritis (RA), and the presence of anti-cyclic citrullinated peptide (CCP) antibodies is often associated with a more severe prognosis (5).

Systemic JIA (sJIA) is uncommon (4-17% of cases). Patients often present with symmetrical polyarticular involvement. However, joint disease can be absent at the onset of disease and develop later. Arthritis is accompanied or preceded by daily intermittent fever, and one or more of the following: characteristic skin rash (salmon pink, evanescent, macular), generalised lymphadenopathy, enlargement of the liver or spleen, and serositis (pericarditis, or more rarely pleural effusion, rarely peritonitis) (5). Children with sJIA are at risk of developing macrophage activation syndrome (MAS), a potentially life-threatening complication caused by a massive hyperproduction of pro-inflammatory cytokines (8, 9).

The diagnosis of juvenile psoriatic arthritis (JPsA) requires the coexistence of arthritis and psoriasis. Alternatively, when psoriatic rash is missing, the presence of arthritis and two of the following are required: family history of psoriasis in a first-degree relative, dactylitis (sausage-like swelling of individual digits that extends beyond the joint margins), and nail pitting or onycholy-

sis. The rash may appear years after the presentation of arthritis, which may involve both large and small joints (5).

ERA (enthesitis-related arthritis) mainly affects male patients older than 6 years and is defined by the coexistence of enthesitis and arthritis, or arthritis and more than two of the following: sacroiliac joint tenderness and/or inflammatory lumbar pain, HLA-B27 positivity, acute anterior uveitis or a first-degree relative with a spondyloarthropathy (6, 10). It is often characterised by asymmetrical lower limb involvement, and may progress to involve the joints of the axial skeleton (sacroiliac or lumbar spine). Patients carrying HLA-B27 frequently develop sacroiliitis.

About 20% of children with chronic arthritis do not meet the criteria for any category or have overlapping features between subtypes (*e.g.* RF-positive polyarthritis with psoriasis). These forms are classified as undifferentiated arthritis and have a variable course (5). Currently, there is limited understanding of the underlying pathogenetic mechanisms of the various JIA phenotypes. The clinical heterogeneity of JIA suggests that at least some subtypes represent distinct disease entities with their own genetic and immunopathogenetic background. It is thought that JIA is due to disrupted immune regulation, and that the course of the disease is influenced by either genetic or environmental factors. The major immunopathogenic distinction in JIA is between oligo- and polyarticular JIA, which are characterised by an imbalance between regulatory T cells and effector T cells, and systemic JIA, which is believed to be caused by dysregulation of innate immunity (11).

The first-line pharmacological treatment of JIA is based on anti-inflammatory drugs, *i.e.* NSAIDs, systemic or intra-articular glucocorticoids, and on disease-modifying anti-rheumatic drugs (DMARDs), particularly methotrexate (MTX) (12, 13).

Patients who are refractory to DMARD therapy are candidates to receive treatment with biologic agents. Several biologics have been approved for the treatment of patients with polyarticular course JIA or systemic JIA or both, including etanercept (14), followed by

adalimumab (15), abatacept (16), tocilizumab (17, 18), and canakinumab (19). In addition, the persistence of pain is a considerable problem for some children with JIA despite treatment with biologic DMARDs and good disease control (20).

Despite early treatment with DMARDs and biological drugs, in more than one-third of patients the disease continues and still requires immunosuppressive treatment after adolescence (21, 22). The analysis of 437 patients revealed that only 6% of treatment free remissions were maintained for at least 5 years (23).

Currently, there are no guidelines on how to select, taper or discontinue biologic treatments in adult patients with JIA, and adult rheumatologists usually manage JIA as they manage RA. However, although RA and JIA share some clinical features and can both become disabling if left untreated, childhood arthritis is much more heterogeneous than adult RA. The management of the different forms is in part different and is based on the specific biologic and clinical features of each JIA form.

A number of therapies are used for the management of adults with JIA. However, there are no evidence-based guidelines for the treatment of JIA in adult patients. The factors that drive the choice of therapy are patient preference, clinician experience, compliance expectations, clinical manifestations and, last but not least, the JIA subtype (4, 5, 24, 25).

The management of adults with JIA usually mimics the management of RA, with MTX and, more recently, biologic drugs as mainstay therapies, but the therapeutic approach often does not take into account that many forms of JIA differ considerably from RA. Vidquist retrospectively evaluated the relationship between the use of DMARDs and biologics and disease activity in 154 patients with JIA referred to an adult rheumatology clinic: after the transition, 29% of patients were on biologic therapy, and 44% of them had been receiving such therapy for more than 5 years, with a mean duration of treatment of 4.2 years. 29% of disease recurrences were observed after discon-

tinuation of medication, with a median interval of 1.4 years (4).

The British Society for Rheumatology Biologics Registry (BSRBR) studied 225 JIA patients retrospectively, in order to describe the pattern of biologic use among patients beginning therapy in adulthood and its relationship with the distribution of ILAR subtypes. Although all patients satisfied the diagnostic criteria for JIA, the ILAR subtype could not be defined in 32% of them, due to missing information. 56% of patients were treated with a biologic agent in combination with MTX (alone or with another DMARD). In 35% of cases the DMARDs had been withdrawn when biologic therapy was introduced. Biologic therapy did not differ significantly across ILAR subtypes. Fifty percent of patients were treated with more than one anti-TNF agent during the study period. The authors concluded that adults with JIA are often treated with biologic therapies, but the choice of the first biologic agent is frequently inconsistent, due to the lack of *ad hoc* guidelines (26).

A preliminary Position Statement on the prescription of biologic therapies to adults with JIA has been issued by the British Society for Paediatrics and Adolescent Rheumatology, together with the British Society for Rheumatology (BSPAR/BSR). The key statement is that patients with JIA requiring biologic treatment at the age of 17 years probably will have to continue this treatment when they are adults. Moreover, the disease could become active during adulthood and patients may need treatment with a biologic agent for the first time. It is important that adults with JIA who require a biologic agent, either for the first time or because of a disease relapse (*e.g.* after a period of remission or pregnancy) have access to such treatment, and that it should not be discontinued once a patient reaches 18 years of age (27).

Clinical and social issues

JIA is associated with increased mortality, significant morbidity due to articular and extra-articular manifestations, chronic disability, and restricted participation in normal social activities.

The disease itself or its treatment are associated with a number of potential complications, which include osteopenia, osteoporosis, and growth retardation. The risk of malignancy may be increased, although whether it is a consequence of JIA itself and/or of therapy with DMARDs or biologics is up for debate. Patients with JIA may be also exposed to premature atherosclerosis, with increased risk of early cardiovascular disease. Other causes of morbidity are represented by temporomandibular joint disease, which can lead to micrognathia and malocclusion; nonreversible joint damage requiring orthopaedic surgery for refractory single-joint disease, and ocular damage caused by refractory uveitis. In some instances, ocular inflammation may persist into adulthood and need continued ophthalmologic monitoring and specific therapies; notably, the uveitis process may be exacerbated after a change in systemic treatment (5).

The optimal management of JIA is based on the creation of a multidisciplinary team, coordinated by a rheumatologist and including several specialists. In particular, the gynecologist has a critical role both in young and adult JIA patients. He/she provides counselling about sexuality, contraception and reproductive issues. Little information is available about the sexuality of adolescents and young adults with JIA. De Avila Lima Souza found that the sexuality of 32 patients with JIA did not differ from controls (28). Nevertheless, this factor should not be neglected owing to its major impact on patient quality of life (QoL). The behaviour of female patients has been found to be similar to that of healthy controls in terms of sexual activity, contraception, wish for children, age at first child, and duration of breast feeding. Fertility is not impaired, but in women with JIA a significantly increased rate of miscarriages and poorer pregnancy outcomes have been reported. Moreover, in adult females with JIA, a higher rate of metrorrhagia, pelvic inflammatory disease, and ovarian cysts has been noticed (29). Finally, in adult women with JIA, some anti-rheumatic therapies could affect pregnancy. Caesar-

ean section may be necessary as a result of hip destruction, short stature or other deformities (5). In JIA patients, pregnancy generally induces a reduction in disease activity, but flares often occur after childbirth, particularly if the woman decides to breast feed the infant, or had postpartum flares after previous pregnancies or active disease before pregnancy (30). For all these reasons, the collaboration of the obstetrician/gynaecologist with the rheumatologist is crucial.

No congenital malformations or adverse pregnancy outcomes have been related to paternal treatment with anti-rheumatic drugs, but some drugs can induce oligospermia, reduce sperm motility, and semen abnormalities (31). Proper counselling should be offered to patients about potential reproductive issues, and pregnancy should be ideally planned during a period of clinical remission or low disease activity. Treatments should be chosen not only for their efficacy, but also for their compatibility with pregnancy planning.

Because the disease may cause severe musculoskeletal complication, the multidisciplinary team should include an orthopaedic surgeon, whose intervention is sometimes necessary for the correction of joint deformities and/or joint ankyloses: in a Scandinavian report, 22 out of 154 patients underwent joint surgery (4). Other specialists that should be included in the optimal multidisciplinary team are an ophthalmologist, a cardiologist, a physiotherapist, and possibly a psychologist.

The QoL of JIA patients can be severely impaired. Wipff reported that QoL was significantly worse in 161 patients with JIA than in controls matched for age and sex (32). Approximately 10–20% of 20- to 35-year-old JIA patients suffer physical, functional and social restrictions (21). Many studies have shown that although adults with JIA have school outcomes comparable to those of the general population, they have more occupational difficulties (33–35).

An important limitation for the quantitative clinical assessment of the level of disease activity at follow-up visit is the lack of validated tools for use in adults

with JIA. The British Society of Rheumatology recommends that adult JIA patients should not be inappropriately re-categorised as having RA, ankylosing spondylitis or another condition, since the tools designed for assessing other inflammatory diseases have not been validated for JIA (26, 27).

Altogether, these findings indicate that JIA patients referred to adult rheumatology centers have special needs and require a tailored multidisciplinary approach as well as a long-term specialist follow-up and medical treatment. Organisational and therapeutic improvements in this area might optimise the management and the control of the disease, and contribute to enhance patient QoL making it similar to that of the general population (21).

Transitional care in JIA

A critical moment in the clinical history of patients with childhood-onset rheumatic disorders continuing into adulthood is the transition from the paediatric to the adult health care team.

Transition has been defined as “the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centred to adult-oriented health care systems” (36). Transition from paediatric to adult care is currently recognised as one of the key factors for the correct management of childhood onset-chronic illnesses (37). There is evidence that morbidity and mortality increase following an inadequate transition from paediatric to adult services (37). Young patients are particularly vulnerable and can experience worsening of their disease. Nakhla found that hospitalisation rates are higher among diabetic adolescents during the first 2 years after transfer, compared with the 2 years before. The physiological endocrine changes during puberty, including possible development of insulin resistance, are associated with behavioural attitudes: adolescents need more autonomy and are prone to risk-taking behaviours. These threatens may be mitigated when physician continuity is ensured: after the transition, patients who were successfully transferred to a new team had a 77% lower risk of hospitalisation

than patients simply entrusted to a new physician (38).

Many authors observed that patients directly transferred from a paediatric to an adult clinic were disappointed. When diabetic patients met the adult physician before the transition, a more rigorous clinic attendance was seen post-transfer, and in these patients HbA1c levels were more often found to be within the normal range (39).

A study on children who underwent renal transplant found that, despite stable renal parameters during the year before transition, 35% of grafts were lost within one year after transfer to adult care. Many patients reported that their understanding of the prescribed medications and their potential side effects was poor and that there were problems in family dynamics, which resulted in decline in therapy adherence. In some patients, low cyclosporine levels were noted, and this could have greatly contributed to graft loss. The authors emphasised the importance of improving the dialogue between the patient and the medical team (40).

Increased disease activity and increased number of disease flares have been reported in adolescents with rheumatic diseases at the time of transition. A recent retrospective study has found that out of 58% of the patients who had active disease at the time of transfer, 30% were hospitalised because of a disease flare in the year before the transfer, and 30% had increased disease activity in the year after transfer (41).

Other factors, such as repeated and/or prolonged hospitalisations, physical limitations, changes in physical appearance and overprotective families, contribute to make the situation more challenging (42). In addition, adolescents often exhibit behaviours that can jeopardise the control of a previously well-managed condition (43).

All authors agree that transfer of patients from the paediatric to the adult system can affect their short and long-term outcomes, and underline that the lack of a well-coordinated transition process can lead to suboptimal clinical management.

Ideally, the process should be continuous, coordinated and adapted to the

psychological and social features of the patient, the complex and varied clinical context, and the readiness of the patient and the family to the change. This objective can be achieved through the institution of ‘joint clinics’ including both paediatricians and adult physicians.

The transition needs of chronically ill adolescents and their families have been assessed in several studies (44). Adolescents need autonomy in their care, details about the adult health care system and meaningful social support to better manage their disease (45, 46).

An effective transition process increases the patient’s ability to manage personal health responsibilities (47) and, even more importantly, improves long-term outcomes, as seen in several conditions, such as cystic fibrosis (48), renal failure (49) and transplanted children (50).

Co-management of the patient, and coordinated communication between paediatric and adult services, along with shared patients’ registries, are crucial for the long-term monitoring of the safety and efficacy of therapies, especially biologic agents, in JIA extended from childhood to adulthood (51).

A poor transition process, with lack of cooperation and communication between the paediatric and adult rheumatologic team, and without shared databases, often leads to loss of information on the disease onset and its subtyping, as well as on previous therapies and treatment response: these factors prevent a reliable evaluation of long-term outcomes and their correlation with potential predictors (4).

Unfortunately, in rheumatic diseases about 50% of the transfer processes to adult rheumatology are not successful. These patients are, therefore, at high risk of unfavourable outcomes (22, 52, 53).

There are several obstacles to the success of transitional care for JIA patients, such as the absence of specific criteria for the assessment of disease activity, the lack of specific treatment recommendations for JIA adult patients, the poor adolescent-specific training for adult rheumatologists, and the shortage of resources (22, 54).

A further element that hampers the implementation of a successful transi-

tion process is the difficulty for adult settings to set up the aforementioned multidisciplinary team that is needed to assist optimally children with JIA who have become adults. The achievement of this goal requires additional collaboration and coordination efforts (1, 55). The improvement in the transition process in medical care has become a priority in many health care systems. The American Academy of Pediatrics, the American Academy of Family Physicians and the American College of Physicians have underscored the need for effective transition of care (56) and this activity has been designated a public health goal for the US Department of Health and Human Services' Healthy People 2020 (57). In the US, transition has been included in the Standards and Guidelines of the National Committee for Quality Assurance (58), and it is included among the Leading Health Indicators (57).

Although no standard model for transitional care has been defined, several position statements and guidelines have been developed, in order to assist health care providers in delivering the best care to transitioning patients and to reduce the risks associated with incorrect transfer to adult services (59-62).

A clinical report from the American Academy of Pediatrics (AAP), the American College of Physicians (ACP), and the American Academy of Family Physicians (AAFP) includes guidance for health care professionals regarding young patients with special health care needs. Its recommendations are as follows: the transition process should start between 12 and 14 years, the adolescent should be guided towards the adult health care system, and the transfer from paediatric care to the adult health care system should occur between 18 and 21 years. It also suggests that the young patient should be increasingly involved in decision-making, and underlines the value of communication and coordination between the paediatric and adult system, as well as the proper sharing of medical information (56).

In 2016, an international panel of experts of EULAR developed recommen-

dations for transitional care in juvenile-onset rheumatic and musculoskeletal diseases. The panel identified significant limitations in the transition services, such as unmet training needs for healthcare professionals, lack of preparation of patients and families, and absence of robust quality indicators both for outcomes and strategy cost-effectiveness. The main recommendations that were formulated are the following: access to high-quality and coordinated transitional care, provided by means of partnership between paediatric and adult healthcare professionals; multidisciplinary care starting in early adolescence; a transition coordinator; transition policies and protocols; transfer of the documentation; open electronic-based platform to access resources; appropriate training for paediatric and adult health caregivers and an increase in evidence-based knowledge to improve outcomes. Understanding the patient's individual characteristics and the role of the families, written communication, agreed procedures, training and precise roles within the team have been identified as the main factors that ensure success (3).

Several healthcare institutions have developed and implemented transition programs (Table II).

Stringer reported the results of a 2-year transition program, ongoing at the Paediatric Rheumatology Transition clinic at the IWK Center, Canada (37). At the first transition clinic visit and at follow-up visits, the adolescent is examined at the same time by the adult and the paediatric rheumatologist who has followed the patient until then; they review the case together, while other members of the paediatric team discuss the management and transition plans, and address the potential transition issues with the adolescent. The patient is seen alone, but the parents can join at the end of the visit. The process begins about 2 years before the end of high school, and the actual transition occurs between 17 and 20 years in 88% of patients. Overall, most of the participants are satisfied with the whole program, and believe that the age of transition (17-20 year) is appropriate. However, some deficiencies in care have been re-

ported, especially in counselling about common adolescent problems, such as substance use and sexual health (37).

In the UK, the program Ready Steady Go, implemented by a NHS teaching hospital, is designed to empower patients to take control of their disease (64). The program begins at about 11 years of age, with the assessment of the knowledge of the disease and its treatments, as well the skills of self-management, the understanding of the need for a healthy lifestyle, and potential psychosocial issues. The activities of the centre and the team are tailored to the patient's skills and needs, based on the answers to specific questionnaires (64). The National Alliance to Advance Adolescent Health (Washington, DC) has realised the Got Transition Program, based on the recommendations of the American Academy of Pediatrics/American Academy of Family Physicians/American College of Physicians Clinical Report on Transition (66). The program identifies the "Six Core Elements of Health Care Transition (HCT)", to be carried out within predefined timelines:

1. Patient and family awareness of transition policy (12 years);
2. Start of the health care transition planning (14 years);
3. Discussion with patient and parents about adult model of care (16 years);
4. Transition to adult model of care (18 years);
5. Complete transfer to adult medical system, with transfer package (18-22 years);
6. Integration of the young adults into adult care (23-26 years).

All or only a few of the elements of the program can be implemented according to the judgment of the provider. More time and effort are necessary for patients with more complicated diseases, few resources or lack of family assistance, whereas patients with a higher maturity level, more family support, or lower disease severity (such as oligoarticular JIA in remission) require less support (66).

The Shared Management Model of Transition is a planned systematic program for gradually shifting the responsibilities from the health care provider

Table II. Transition models.

Name	Place	Methodology/tools	Timing
NA (37)	Health Centre Paediatric Rheumatology Transition Clinic, Halifax, Canada	<ol style="list-style-type: none"> First transition clinic visit: the adult and the paediatric rheumatologist jointly see the patient and review his/her case in consultation. The assessment of the patient is completed by both physicians, and other members of the paediatric team discuss the transition issues with him/her. The patient is seen alone, and parents join only after the visit. Follow up visits: the assessment is performed by either the adult or paediatric rheumatologist, as well the discussion with the patient about transition plans and management; other members of the team address possible transition issues. 	The first transition clinic appointment in the 2 years prior to completion of high school. Patients transferred when they have finished high school.
DON'T RETARD (63)	Department of Paediatric Rheumatology, University Hospitals, Leuven, Belgium.	<p>A five steps program:</p> <ol style="list-style-type: none"> two out-patient appointments (6 months apart) with transition coordinator (discussing leisure activities, school, friends and medication compliance); information day for adolescents and their parents (with the adult rheumatology team); individualised transfer plan; actual transfer. 	A brief transition program for young people with JIA. Patients included at a median age of 16 years.
Ready Steady Go (64)	National Health Service teaching hospital, Southampton Children's Hospital, Southampton, UK.	<ol style="list-style-type: none"> At around 11 years of age: introductory video and the 'Transition: moving into adult care' information leaflet. About every two years young patients complete a series of questionnaires: 'Ready' (around age 11–13 years), 'Steady' (around age 13–14 years), 'Go' (around age 16–18 years) to ensure that they have all the skills and knowledge in place to 'Go' to adult services. At their first clinic appointment in adult services, young patients complete 'Hello' questionnaire. http://www.uhs.nhs.uk/readysteadygo 	Starting at around 11 years of age, if developmentally appropriate; ending at their first clinic appointment in adult services. Timing mutually agreed by the young patient, family and medical professionals.
RAP transitional care program (44)	10 paediatric rheumatology centers (UK)	<ol style="list-style-type: none"> Local programme coordinator, a clinical lead (consultant rheumatologist) identified within each centre, individualised transition plan templates, focused on home, health, and school: Rheumatology Adolescent planner (RAP) Filofax, RAP resource book for parents, RAP resource book for local program coordinators, adolescent rheumatology resource directories for health professionals. 	Ideally starting at 11–12 years of age, overall duration individualised.
NA (65)	The Ohio State University Wexner Medical Center and Nationwide Children's Hospital, Columbus, OH, USA	<ol style="list-style-type: none"> Process coordinated by a social worker that meets young patients at the beginning of the transition process and during the following visits to assess transition awareness and progression, and provide information about the process and the available resources. Transfer to adult rheumatologic care when considered appropriate by the treating paediatric rheumatologist; the social worker coordinates the appointment with the adult rheumatologist and follows up with the participants. 	Patients ≥16 years of age (median age 18 years). Transition completed when the patient has seen the adult provider twice.
Got Transition (66)	Paediatric and adult academic health centres, District of Columbia, USA	<p>The Six Core Elements of Health Care Transition (HCT):</p> <ol style="list-style-type: none"> Development of transition and young adult privacy and consent policies; Creation of transitioning and young adult patient registries to monitor progress and outcomes; Transition preparation including identification of gaps in transition readiness; Transition planning including identification of adult providers and the development of a Health Care Transition Action Plan; Transition and Transfer of Care including communication between paediatric and adult providers; Transition completion 	Transition process should begin early in adolescence (ages 12–14 years), including the patient/family awareness evaluation and transition planning.
Shared Management Model (67)	Transition to Adult Healthcare Services Workgroup, Toronto, ON, (Canada)	<p>Transition program outlined to facilitate the implementation of a shared management approach.</p> <ol style="list-style-type: none"> Professional Education on Shared Management Family facilitators Transition Tools and Resources: <ul style="list-style-type: none"> ✓ Timetable for Growing Up (Developing the Skills for Growing Up, not disease specific) ✓ transition-planning checklists ✓ professional checklist ✓ medical summary 	Starting with the first tool (Timetable for Growing Up) at young age, <i>i.e.</i> 7 years.

to the parents and finally to the patient, as the young patient matures in age, developed by the Transition to Adult Healthcare Services Workgroup of the Provincial Council for Maternal and Child Health (Canada) (67).

The SHARE (Single Hub and Access point for paediatric Rheumatology in Europe) initiative is not only aimed to develop consensus guidelines for minimum recommended standards of care of paediatric rheumatic diseases in European countries, but also to establish platforms for the sharing of information and for linking the existing networks (68).

Studies on evaluation of transition effectiveness and indicators of success

Despite agreement on the importance of transition, there is a lack of rigorous research on the impact of transition programs on outcomes: not many studies have precisely evaluated transition models, and common methodologies for measuring transition outcomes are missing (40, 69-72).

The Adult Healthcare Services Work Group of the Canadian Provincial Council for Maternal and Child Health has published a list of the specific tools developed to facilitate the transition process, but has also pointed out that very few of them are supported by the literature (73).

Among the potential indicators of a 'successful' transition, clinical parameters (disease activity and status), adherence to treatment and attendance of adult healthcare as well as patient and family experience, work achievements and quality of life measures should be included. Some authors indicate that the "patient not lost to follow-up" is the most important indicator of a positive outcome (74).

Studies of transitional care programs for young adults with rheumatic diseases have highlighted the need for improved consistency of practice between paediatric and adult rheumatologic teams, have identified rheumatology training and staff induction programs as the key success factor to guarantee information continuity, and have underlined that the process could be improved through the

implementation of a structured, coordinated program (54, 75).

The comparison of the clinical outcomes of JIA patients included in a brief transition program (Devices for Optimization of Transfer and Transition of Adolescents with Rheumatic Disorders - DON'T RETARD) with those of patients receiving the usual care showed that the program has improved the patient physical, psychosocial and health status and quality of life (63).

According to Cruikshank, the healthcare facility and healthcare professional factors associated with a successful transition program are enthusiasm, motivation, interest and collaboration between paediatric and adult services (76).

Several surveys have highlighted the most common challenges, which include lack of written transition plans, low familiarity with transition guidelines, disappointing medical history information received by adult rheumatology centres from paediatric providers, and inadequate preparation of young adults to transfer to adult care (77, 78). Insufficient training and scarce resources (reimbursement, time and personnel), leading to lack of information continuity, have been identified as the major obstacles to the implementation of the transition process (79).

Conclusion

The long-term management of adults with JIA is a challenging process, and adult rheumatologists should be prepared to the high number and variety of clinical problems, such as co-morbidities, sexual health and reproduction, and work and employment issues.

Transition is a complex process and patients with special health care needs are at risk of worsening of their health and disease status and disruption in care. Several studies in paediatric rheumatic patients (as well as in those with other chronic diseases), have reported that about 50% of patients do not experience effective transfer to an adult rheumatologist and are at risk of poor outcomes. After the transition, young patients are often lost to follow up and/or their compliance to treatment

is inadequate. A successful transition is, therefore, crucial to ensure optimal care and better outcomes. Appropriate resources must be allocated to achieve this goal.

The timing of transition, particularly when patients have persistent disease activity, is a serious challenge for patients, families and healthcare professionals. The transition should begin long time before the actual transfer: in all reported experiences, the program starts at about 14 years, with co-management conducted by paediatric and adult rheumatologic teams in coordination, then drives the adolescent to the adult health care system, and finally entrusts the patient's management to the adult rheumatologists at about 21 years.

The lack of collaboration and shared information between the paediatric and the adult rheumatology system can jeopardise the entire process, as it may lead to loss of information related to disease onset and medical history, including previous treatments and patient's response.

A successful transition needs collaborative efforts and improved communication between paediatric and adult healthcare providers. Only close collaboration and co-management of the patient by paediatric and adult rheumatology teams can ensure a smooth transition process, which must be timely planned, coordinated and multidisciplinary. Shared or, at least, connected medical record databases enable the proper evaluation of long-term outcomes, and this is particularly important when new treatments become available.

Both adult and paediatric rheumatology communities are committed to improve the existing transitional care services, to amend the shortcomings of existing programs and to overcome the scarcity of resources, but there is a great need for rheumatology-specific guidelines across pediatric, adolescent and adult rheumatology centres.

An effective transition program for JIA patients could enable a real advancement in terms of quality of life, work experience, satisfaction with care, and long-term outcomes.

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