

ملاحظات اختصاصی در انتقال بیماران

JIA , JSLE, JDM

به سرویس بالغین

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Clinical and social issues

JIA is associated with :

- Increased , significant morbidity due to articular and extra-articular manifestations
- chronic disability
- restricted participation in normal social activities
- osteopenia, osteoporosis
- growth retardation
- The risk of malignancy
- exposed to premature atherosclerosis
- nonreversible joint damage
- ocular damage caused by refractoryuveitis

JIA Transition

Current research suggests that approximately **one third** of youth with JIA will continue to have active disease well into adulthood

About **50%** of the transfer processes to adult rheumatology are not successful, putting these patients at high risk of unfavourable outcomes several obstacles :

- 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
- the shortage of resources

JIA

- (ACR) core outcome variables for juvenile arthritis :
 - (1) physician global assessment of disease activity (10-cm visual analog scale (VAS))
 - (2) parent/patient assessment of overall well-being (10-cm VAS)
 - (3) functional ability (childhood health assessment questionnaire (CHAQ))
 - (4) number of joints with active arthritis (defined as joint effusion or limitation of motion accompanied by heat, pain, or tenderness)
 - (5) number of joints with limited ROM
 - (6) erythrocyte sedimentation rate (ESR)

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- Improvement : 30% improvement from baseline in three of six variables, with no more than one remaining variable worsening by >30%
 - Flare : worsening of two variables by 40% without improvement in more than one variable by 30%

JADAS

- (1) physician global assessment of disease activity
- (2) parent/patient global assessment of well-being
- (3) number of active joints
- (4) ESR

- three distinct health outcomes for JIA patients
 - “Inactive disease” when a patient on medical therapy has:
 - (1) no joints with active arthritis
 - (2) no fever, rash, serositis, splenomegaly, or generalized lymphadenopathy
 - (3) no active uveitis
 - (4) normal ESR and/or CRP
 - (5) physician global assessment indicates no disease activity
- If patients meet the inactive disease criteria for 6 continuous months while taking medication, they are classified as being in “clinical remission on medication”
- if they meet the criteria for 12 continuous months off medication, they are considered to be in “clinical remission off medication”

JIA Disease Damage Index

- articular (JADI-A) :
 - evaluates damage in 36 joints (maximum score 72)
- extra-articular (JADI-E) :
 - assesses 13 items grouped into five organ systems scored
 - as absent (0) or present (1)
 - with the exception of the ocular damage which can be given a score up to 3 – for a maximum total score of 17

JIA Transition

- 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”
- Timeline :
11- and 14-yearold patients with JIA showing maximum improvement in disease knowledge after 12 months of participation in a transitional care program

AAP & ACP & AAFP

- 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

international panel of experts of – EULAR - 2016

The main recommendations :

1. YP with RMD should have access to high-quality, co-ordinated transitional care, delivered through partnership with healthcare professionals, YP and their families, to address needs on an individual basis
2. The transition process should start as early as possible; in early adolescence or directly after the diagnosis in adolescent-onset disease
3. There must be 'direct' communication between the key participants (and as a minimum, to include the YP, parent/carer, and a member each of the paediatric and adult rheumatologist teams) during the process of transition. Before and after the actual transfer, there should be 'direct' contacts between paediatric and adult rheumatologist teams

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4. Individual transition processes and progress should be carefully documented in the medical records and planned with YP and their families
 5. Every rheumatology service and clinical network—paediatric and adult—must have a written, agreed and regularly updated transition policy
 6. There should be clear written description of the MDT involved in transitional care, locally and in the clinical network. The MDT should include a designated transition co-ordinator

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- 7. Transition services must be YP focused, be developmentally appropriate and address the complexity of YP development
 - 8. There must be a transfer document
 - 9. Healthcare teams involved in transition and adolescent-young adult care must have appropriate training in generic adolescent care and childhood-onset RMD
 - 10. There must be secure funding for dedicated resources to provide uninterrupted clinical care and transition services for YP entering adult care
 - 11. There must be a freely accessible electronic-based platform to host the recommendations, standards and resources for transitional care
 - 12. Increased evidence-based knowledge and practice is needed to improve outcomes for YP with childhood-onset RMD



Six Core Elements of Health Care Transition (HCT)

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)

Table II. Transition models.

Name	Place	Methodology/tools	Timing
NA (37)	Health Centre Paediatric Rheumatology Transition Clinic, Halifax, Canada	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.	A five steps program: 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.	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)	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	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	The Six Core Elements of Health Care Transition (HCT): 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)	Transition program outlined to facilitate the implementation of a shared management approach. Professional Education on Shared Management Family facilitators Transition Tools and Resources: ✓ 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.

Disease activity measures in JSLE

- Systemic Lupus Erythematosus Disease Activity Index (SLEDAI)
 - British Isles Lupus Assessment Group index (BILAG)
 - Systemic Lupus Activity Measure (SLAM)
 - European Consensus for Lupus Activity Measurement (ECLAM)
- each of which consists of several attributes grouped into 8–10 organ systems
 - ✓ the SLEDAI and SLAM are most user-friendly
 - ✓ ECLAM appears to be most sensitive to change in JSLE
 - ✓ The BILAG is complicated and takes longer to score

Inactive JSLE

- Not having features of active SLE on physical examination (some features due to damage are permissible) and may have two or fewer mild non-limiting symptoms
- Patients must have normal CBC, C₃, and renal function tests (unless due to damage)
- however, they are allowed a positive ANA, elevated ESR and/or antiphospholipid antibody levels (within two times the upper limit of normal), and an abnormal C₄ (if presumed to be due to genetic variability)

Disease Damage in JSLE

- The Systemic Lupus International Collaborating Clinics/ACR Damage Index (SDI), which was initially developed and validated in adult SLE patients, has also been found to be valid and reliable to capture damage in JSLE
- The SDI covers damage in 12 organ systems, and a modified version for JSLE (Peds-SDI) has been proposed by the PRINTO group to include two additional organ systems:
 - growth failure
 - delayed puberty

The Peds-SDI consists of 43 items, with a maximum total score of 49

Lupus In Children

- More aggressive (especially renal and neuropsychiatric involvement)
- Needs more aggressive therapy (more medication-related morbidity)
- Less social maturity because of disease and overprotective family)
- risk-taking behaviours and rebellion because of chronic illness often manifest as non-adherence

Table 1 Common differences between paediatric and adult clinic settings

	Paediatric healthcare	Adult healthcare
Care orientation	▶ Family-centred (triadic).	▶ Patient-centred (dyadic).
Team approach	▶ Multidisciplinary.	▶ Reliant on referrals to other services.
Social work involvement	▶ Social worker often onsite.	▶ Social worker less available.
Appointment length	▶ Follow-up typically 30 min.	▶ Follow-up often 20 min or less.
Insurance	▶ Parental private insurance (until age 26 in the US). ▶ Medicaid (US governmental insurance assistance) available as a safety net until age 19.	▶ Not eligible for parental insurance past a certain age (27 in the US). ▶ Medicaid available only after burden of proof met demonstrating disability.
Educational/vocational needs	▶ Addressed and actively supported during clinic visits.	▶ Less commonly addressed or actively supported in clinic.
Late/no-show policy	▶ Patients typically accommodated despite tardiness or missed appointments.	▶ Patients often not seen if late; patients may be released from clinic for repeated missed appointments.
Trainee supervision	▶ Patients staffed in real time.	▶ Less direct supervision.
Medical care of SLE	▶ Vaccines often provided during specialty clinic visit. ▶ Patients' primary care needs often addressed during specialty clinic visit. ▶ Aggressive steroid and cyclophosphamide dosing. ▶ Annual screening echo and PFTs common. ▶ Emphasis on minimising radiation during imaging studies.	▶ Unlikely to provide vaccines during specialty clinic visit. ▶ Patients usually required to see PCP for comorbidities and health maintenance. ▶ Less intravenous steroid pulsing; often lower doses of cyclophosphamide. ▶ Rarely screened with echo or PFTs unless indicated by history or suggestive symptoms. ▶ More likely to use CT.
Medication non-adherence	▶ Teams often 'work around' non-adherence by using intravenous medications.	▶ Physicians likely to hold patient accountable for non-adherence.
Follow-up interval	▶ Typically every 2-3 months.	▶ Typically every 3-6 months.

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- **how to frame transfer**
 - **developing self-management skills**
 - **transition readiness assessments(Transition Readiness Assessment Questionnaire(TRAQ) or the Got Transition Readiness Assessment)**
 - **establishing a transfer date (at least 12 months in the future)**

Strategies for improving transition and transfer outcomes

Problem/Obstacle	Paediatrics clinic strategies	Adult clinic strategies
AYA does not schedule with adult provider or misses the first adult rheumatology appointment.	► Frame transfer as 'graduating to adult care'; express confidence in adult provider	► Communicate with paediatric provider when transferred AYA misses the first appointment.
AYA drops out of care after the first adult rheumatology clinic visit.	► Prepare AYA for what to expect from adult clinic and provider, highlighting: – Process differences. – Differences in clinic culture. – Medical practice differences. ► Communicate with receiving adult provider before the first visit, using the TRANSFER mnemonic.	► Use the WELCOME mnemonic: – Use the first visit to develop rapport with AYA and parent(s). – Align with paediatrics provider the WELCOME mnemonic. – Minimise medical changes during the first visit. ► Communicate back to paediatric clinic after AYA's first visit.
AYA lost to follow-up due to poorly timed transfer	► Avoid transfer during medical flare, pregnancy, other major life events. ► Consider adult comanagement rather than full transfer of care.	

AYA lost to follow-up due to scheduling policies.	▶ Prepare AYA/family regarding scheduling expectations and rules of adult clinic.	▶ Delineate expectations to AYA/family. ▶ Encourage AYA to save office phone number in cell phone. ▶ Advise AYA not to cancel without rescheduling
AYA struggle in adult clinic due to lack of self-management skills.	▶ Ask parent to leave room for all AYAs >16 years (or >12 years). ▶ Encourage AYA to ask and answer questions independently. ▶ Connect AYA with community resources prior to transfer.	▶ Establish expectation that AYA answers questions first. ▶ Perform social history with parent(s) out of the room; use HEADSS or other adolescent-focused approach to the history. ▶ Assess potential barriers to care (health literacy, transportation, adherence, contraception and others). ▶ Use available social work and community resources.

Adolescent medicine aspects of health not explicitly addressed in the adult clinic

- ▶ Ask parent to leave room for all AYAs >16 years (or >12 years).
- ▶ Encourage AYA to ask and answer questions independently.
- ▶ Connect AYA with community resources prior to transfer.

- ▶ Establish expectation that AYA answers questions first.
- ▶ Perform social history with parent(s) out of the room; use HEADSS or other adolescent-focused approach to the history.
- ▶ Assess potential barriers to care (health literacy, transportation, adherence, contraception and others).
- ▶ Use available social work and community resources

Poor medication adherence.

- ▶ Consolidate medications (eg, number, dosing frequency).
- ▶ Provide pillbox or consider prescribing blister packs.
- ▶ Encourage free medication reminder apps like Mango Health.
- ▶ Help patient overcome financial barriers by using a drug company patient assistance programme, applying for other medication support (www.needymeds.org) and identifying the least expensive pharmacy (www.goodrx.com, www.lowestmed.com).

Disease activity measures in JDM

- Childhood Myositis Assessment Scale (CMAS) :

- 14 maneuvers (score 0–52) to measure muscle strength, physical function, and endurance and has excellent measurement properties

Global disease activity in JDM is measured with the Disease Activity Score (DAS) and the Myositis Disease Activity Assessment Tool (MDAAT)

- The DAS assesses muscle, cutaneous, and vasculopathic features of JDM , while the MDAAT includes extramuscular manifestations and assesses disease activity in seven organ systems
- The cutaneous assessment tool (CAT) assesses the full range of cutaneous manifestations of JDM and provides both skin disease activity and skin damage scores

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- inactive disease in JDM (on or off therapy), requiring three or more of the following:
 - (1) creatine kinase 150 U/I
 - (2) CMAS 48
 - (3) MMT 78
 - (4) physician global assessment 0.2 (0–10-cm scale)

The myositis damage index (MDI)

- measure manifestations in 11 organ systems:
- muscle, skeletal, skin, gastrointestinal, pulmonary, cardiovascular, peripheral vascular, endocrine, ocular, infection, and malignancy
- Each organ system includes 2–8 items scored as present or absent to give an MDI extent-of-damage score (scored 0–35 in children and 0–37 in adolescents)
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- The long-term outcome for childhood-onset JDM has not been documented in the literature
- One small review of 16 patients over a 30-year period in one center :
 - 6 patients were reported to be in long-term remission of their diseases with average follow-up of 3 to 4.5 years
 - 6 patients had disease flares requiring additional treatment
- Another small review studied 18 adults an average of 18.5 years after diagnosis of JDM. The majority of the patients did well, with only three having significant residual disability. These subjects enjoyed a good level of educational and employment attainment

Content of a transition policy for a pediatric rheumatology department

Definition of transition
Overview of transitional care in rheumatology at host institution
NamEd team members who will coordinate the transition program
Age at main transition events
 First discussion
 Start of transition program
 Transfer to adolescent service
 Transfer to adult service
 Exceptions to stated timings, e.g., when young people are under multiple specialties
Target adult services
 Rheumatology
 Ophthalmology
 Orthopedic
 Renal

- Transition program elements
 - For young person
 - For parent/guardian
 - In pediatric rheumatology clinics
 - In adolescent rheumatology clinics
- Will include
 - Individualized transition plans
 - Multidisciplinary team documentation process
 - Informational resources
- Transfer process elements
 - Contact with adult service
 - Involvement of primary care
 - Medical summary and administrative transfer
- Aftermath
 - Policy regarding contact with patients following transfer to adult services
- Evaluation procedures
 - Audit
 - Regular review of policy
 - Participation of young people and their parents in evaluation and future development of the service

Goals of a pediatric rheumatology transition program

1. Education

Knowledge about disease, medications, and roles of health care providers

Skills in communication with health care providers

2. Assist with separation from parents with respect to medical issues

3. Encourage adherence to medical recommendations

4. Provide assistance/guidance with issues of education, jobs/ career, finances/health care coverage, independent living, and relationships outside the family

5. Implement final transfer to adult health care providers (rheumatologist and others as necessary), providing adequate records to new care providers

Determining patient's readiness to transference to an adult rheumatologist

1. Patient has relatively stable disease
2. Patient has adequate understanding of the disease and its treatments
3. Patient has demonstrated ability to make and attend appointments and take care of medication needs
4. Patient has developed an independent adult relationship with the health care providers in the YARD clinic
5. Patient has a family doctor in the community and demonstrates the ability to use the family doctor appropriately

