FROM CHILDHOOD TO ADULTHOOD: RITE OF PASSAGE OR TRANSITION?

Lucile HOUYEL
Hôpital Marie-Lannelongue-M3C
Le Plessis Robinson, France

FCPC- RFMCC,
Schoelcher, La Martinique,
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Transition : what’s in a name?

« A purposeful, planned process that addresses the medical, psychological, educational and vocational needs of adolescents and young adults with chronic physical and medical conditions as they move from child-centered to adult-oriented healthcare systems »

Rosen J Adolesc Health 2003
Williams J Pediatr 2015

Transfer : physical movement from one system to the other
Survival of adult CHD: evolution with time

Direct link with advances in cardiac surgery and postoperative care

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Estimation of the number of patients with clinically significant CHD in the next decades

Number (n)

- Live birth with CHD (0.8%)
- Children with CHD
- Young adults (18 yr) with CHD
- Adults with CHD

Données Kompetenznetz-AHF Allemagne
Long-term potential problems

• Arrhythmias, conduction disorders
• Hemodynamic complications
  – Interventional catheterisation
  – Surgery
• Cardiac insufficiency
  – Transplantation
• Endocarditis
• Contraception, pregnancy
• Coronary disease
Figure 2. Lifetime cumulative incidence of atrial arrhythmias in ACHD. A, Overall 20-year risk of 7% of developing atrial arrhythmia (AA) from the age of 20 years and 38% from the age of 55 years.

Transition: why?

- CHD are repaired, not cured
- AHA: less than 30% of adult CHD have an appropriate follow-up
- Drop-out of care (or gaps in care) between 18 and 30 years of age: 21% to 76% among series
- Reid 2004 (Toronto):
  - only 47% of adult CHD experienced an appropriate transition (high quality specialized care without gaps in follow-up)
Transition : why ?

• Wacker 2005 (Munich): >76% of adult CHD had no cardiology visit within 5 years or more
• Yeung 2008 (Denver, Colorado): drop-out of care in 63% of patients (median 10 yrs)
• Mackie 2009 (Québec): drop-out of care
  – 61% of CHD > 18 yrs
  – 47% of moderately severe CHD
  – 21% of complex CHD
Figure 2. The proportion of patients in contact with the healthcare system.
Figure 3. Outpatient cardiology follow-up stratified by severity of CHD. “Univentricular hearts” includes hypoplastic left heart syndrome. “Conotruncal anomalies” refers to tetralogy of Fallot, truncus arteriosus, and transposition of the great arteries. “L...
Transition: aims

- To ensure continuity (and ongoing quality) of care, taking into account the problems specific to adulthood.
- To promote:
  - Independence of the patient relative to the clinicians... and to her/his parents!
  - Greater independence relative to treatments, medical and surgical decisions (control over healthcare decisions)
  - Skills in communication, decision-making, self-care and self-advocacy
- To optimize:
  - Quality of life
  - Life expectancy
  - Future productivity ....

Sable Circulation 2011
Transition: when?

• When should the process begin?
  – From 12 yrs (AHA)…… actually well before
  – From the diagnosis (antenatal!)
• Continuous process, without lapse in care
• Patient-tailored, addressed to an individual with specific needs
• According to age and developmental status
• Educational and information program

FLEXIBILITY

Williams J Pediatr 2015
Transfer: when?

- Usually: 16-18 yrs
- For some: 18-30 yrs
- When the patient is ready (physical, emotional and social stability)
- > 50% of ACHD: mood or anxiety disorders
- Deficit in executive functions
- Better results if transfer > 18 yrs

References:
- Viner Eur J Cancer 2003
- Kovacs Int J Cardiol 2009
- Spijkerboer J Pediatr Surg 2008
- Calderon J Pediatr 2012
- Bellinger Circulation 2011
- Cordina Neuroimage Clin 2014
- Reid Pediatrics 2004

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Transition : how to do it?

• Continuous process involving:
  – The child
  – Her/his parents
  – Pediatric cardiologist

• Anticipate the transfer
  – Progressive involvement of the child according to developmental milestones
  – Diagnosis, treatments, future
  – Personnalized health record
Knowledge of the disease

• Children 7-18 ans : only 22% are able to identify their CHD (median age 13 yrs)

• Adult CHD
  – 35%: nothing !
  – 79% have heard about prevention of endocarditis, but only 50% understand why.....

• The degree of knowledge is related to parental educational level
  – Constant educational reinforcement (parents and child), from the diagnosis of the defect
Information, education, about issues of life management

• Education, employment, career planning
  – Full-time job: 71% vs 84% (Simko 2006)
  – 73% if appropriate counseling vs 46% (Crossland 2005)

• Assurability

• Sports

• Risky behaviors

• Psychological issues, anxiety disorders

• Life expectancy, incurable disease
Parents-child relationship

• Parents
  – Tend to be overprotective, lack of trust
  – Reaction of depression, anxiety, stress, anger, culpability…. Not always correlated with the medical severity of the defect!

• Children
  – Adhesion… or rejection! Risky behaviors

• Parents’ acceptance of the disease and transition has a positive effect of the adolescent psychological well-being

• Importance of a good parents-child relationship
• Preserve the family..

Cohen Psychol Health Med 2008
JM, 27 yrs

• Heart transplantation at 16 months of age (dilated myocardiopathy)
• Follow-up in the pediatric HT center
• 18 yrs : transfer in adult HT center
• Lost to follow-up after the first appointment
• March 2009 (21 yrs): hemoptysis : CCML
• Severe systemic hypertension
• Creatinin 111, urea 9.2, ciclo 128 ng/ml
• Echo : no rejection, coroscan normal
JM, 27 year old

- Follow-up = 0
- Prescriptions: general practitioner
- Full-time job
- Ciclosporin... once a week!
- Disappears until 18 months later (hypertensive crisis)
  - Echo: no rejection
  - No treatment at all
JM, 27 year old

- Several hospitalizations for systemic hypertension (hemoptysis...)
- Finally accepts to take antihypertensive drugs.... But not the ciclosporin!
- January 2015 : terminal renal failure, dialysis
- Echo : acute rejection, coroscan : graft disease
- Observance : perfect!
- June 2015 : on the waiting list for heart + kidney transplantation
Transition: the Belgian model

- Louvain, Belgique: among 794 patients born between 1984 and 1988:
  - Drop-out of care = 7.3%
  - Non-specialized care = 10%
- Risk factors: male, never operated on... and complex CHD!
- Key to success:
  - Same building, common health record
  - Shared database for pediatric and adult patients
  - Complete team (specialized nurses, psychologists)
  - Reminders for missed appointments
  - Health system

Goossens J Am Coll Cardiol 2011
Transition: the keys to success

- Importance of a structured transition process
- Must begin early in childhood
- Insist on the necessity of a life-long follow-up
- Promote a perfect knowledge of the disease, with regular evaluations (parents and child)
- Dialogue with the parents, and private dialogue with the child
- « build the future »
- Positive messages (prevention, sports...)
Transition: the keys to success

• Transition: passage from
  – A pediatric management and follow-up (family-centered)
  – To an adult management and follow-up (patient-centered)

• Perfect coordination between pediatric and adult CHD cardiologist

• Increase the number of specialized adult CHD cardiologists
The ideal transition

• Continual process beginning at the first diagnosis

• Time of transfer:
  – No rules (flexibility)
  – Test: TRAQ (transition readiness assessment questionnaire) \( (Zhang \ BMC \ Pediatr \ 2014) \)

• Unique specialized heart center, from the fetus to the adult

• Paramedical team

• Close collaboration with extracardiac adult specialists
The ideal transition

- Creation of a subspecialty?
- Network of ACHD cardiologists
- Pediatric cardiology? Or congenital cardiology?

• *Williams RG.* « Transitioning youth with congenital heart disease from pediatric to adult health care ». *J Pediatr* 2015;166;15-19.