ADOLESCENT ISSUES: KIDNEY TRANSPLANTATION RECOGNITION OF SYSTEMIC DISEASE NEEDS TRANSITION TO ADULT CLINICS

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Scope

• How should the recognition of systemic disease impact the therapeutic approach?
• Is the bone disease of cystinosis in infancy replaced by another?
• **How should cystinosis patients be prepared for the transition to internal medicine clinics?**
• How should the paediatrician and adult teams be prepared for transition to internal medicine clinics?
• How to manage halitosis?
• How should social adaptation of cystinosis patients be supported?
• Is there a special adaptation of therapeutic education and psychological back-up to adolescents?
• Can cystinosis patients perform all jobs? Is there an effect of neural disease on job training and performance?
Scope

- Is there a preference for a particular form of RRT in cystinosis patients?
- Should cysteamine dose be adapted in patients on RRT? Optimal dose recommendations?
- Do cystinosis patients need a special preparation to renal transplantation?
- Do cystinosis patients need different immunosuppressive therapy after renal transplantation?
- How should the onset of the need for a kidney transplant impact substrate reduction therapy?
Outline

• Adolescence
  – Transition to “adult” healthcare
• Cystinosis & adolescence
  – Transplantation
  – Peer pressures: being different
What is Adolescence?

- A time of complex problems?
- A period of “storm and stress”
- A natural tendency to “risk taking”? 
- A time of developing “affect regulation”? 
- A period of increased resiliency? 
- A time of change

Our Images of Teens

- TIME
- Maclean’s
- Rave Fever
- MACLEAN’S
- TEEN TROUBLE

Research is revolutionizing our view of the adolescent mind—and explaining its mystifying ways.
Developmental Tasks of Adolescence

- Development of self-esteem and a healthy **identity**
- Emancipation from parents to **autonomous** behaviors
- Formation of a **sexual** identity
- Meaningful social and peer **relationships**
- Seeking **vocational** goals
- Establishing moral and ethical **values**
New understanding of adolescent brain development: relevance to transitional healthcare for young people with long term conditions.

Colver A. Longwell S. Archives of Disease in Childhood. 98(11):902-7, 2013
Youth with chronic illness

- More likely to report “fair” or “poor” health status
- Higher risk for emotional distress & suicidal thoughts or attempts
- More likely to be abused
- Higher risk for getting in fights, smoking, and other substance use
Systemic Disease

- What to monitor/screen for and when to start?
  - Hypothyroidism
  - Gonadal function
  - Bone mineral density
  - Nephrocalcinosis
  - Myopathy/swallowing dysfunction
  - Psycho-educational assessments
  - Diabetes
  - Eye disease
  - Lung function
Transition from pediatric to adult renal services: a consensus statement by the International Society of Nephrology (ISN) and the International Pediatric Nephrology Association (IPNA)

Alan R. Watson¹, Paul N. Harden², Maria E. Ferris³, Peter G. Kerr⁴, John D. Mahan³ and Maher Fouad Ramzy⁵, Consensus Panel Members

¹Co-Chair, IPNA, UK; ²Co-Chair, ISN, UK; ³IPNA, USA; ⁴ISN, Australia and ⁵ISN, Egypt

UNOS Database 2011

- Retrospective cohort study (1987 – 2007)
- Failure rates (FR) <21 yr (223) vs. >21 yr (217)
- Age-standardized FR
  12.9/100/year for <21 yr vs. 8.7/100 year for >21 yr
  with 58% failure rates for early transferees
- Transfer < 21 yr should be undertaken cautiously until reliable methods of assessing readiness are developed

Experiences of Parents Who Have Children With Chronic Kidney Disease:

A Systematic Review of Qualitative Studies

- Intrapersonal
  living with constant uncertainty & stress
- Interpersonal
  medicalization of the parental role
- External Issues
  management of the medical regime

Adolescent Adherence

- Less in adolescent years
- Many reasons
  - Fears of medication harm
  - Independence/invulnerability
- Techniques to improve
  - Texting
  - Education

Adherence to medical recommendations and transition to adult services in pediatric renal transplant recipients. E Shemesh et al Curr Opin Org Transp 15:288 (2010)
Transition Recommendations

(1) seeing the young person alone as well as with family
(2) respecting privacy and confidentiality
(3) actively involving the adolescent in medical decision making
(4) promoting responsible for self management, including adherence with medication
(5) addressing educational and vocational needs
(6) raising and addressing reproductive and sexual health issues, substance abuse and other risk taking behaviours

Transition & Cystinosis

• No good tools to gauge readiness/satisfaction
  – 8 Readiness and 6 Satisfaction measures

• Issues
  – All the usual ones, plus
  – Rare disease
  – CNS disease: executive function impairment
  – Halitosis
  – etc

Models for Health Care as an Adult

• Variable depending on
  – Health care system
  – Medical needs
• Continued care by same team
• Shared care
• Full transition
RRT/Transplantation

• Issues
  – Polyuria
    • Pre or post transplant nephrectomy
  – Cysteamine dosing
    • When to restart post transplant
  – Immunosuppression
    • The same as other patients?
Conclusions

• Adolescence is a challenging time
• More so with chronic disease
• With cystinosis
  – Worsening kidney function/RRT
    • Additional manifestations of cystinosis
  – Transition to “adult” health care
    • Best practices still not clear in general

Health care transition for youth with special health care needs