



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

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KDIGO



# 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

Aristotle. “Youth are heated by Nature and drunken men by wine”. More than 20 centuries ago.



# Our Images of Teens

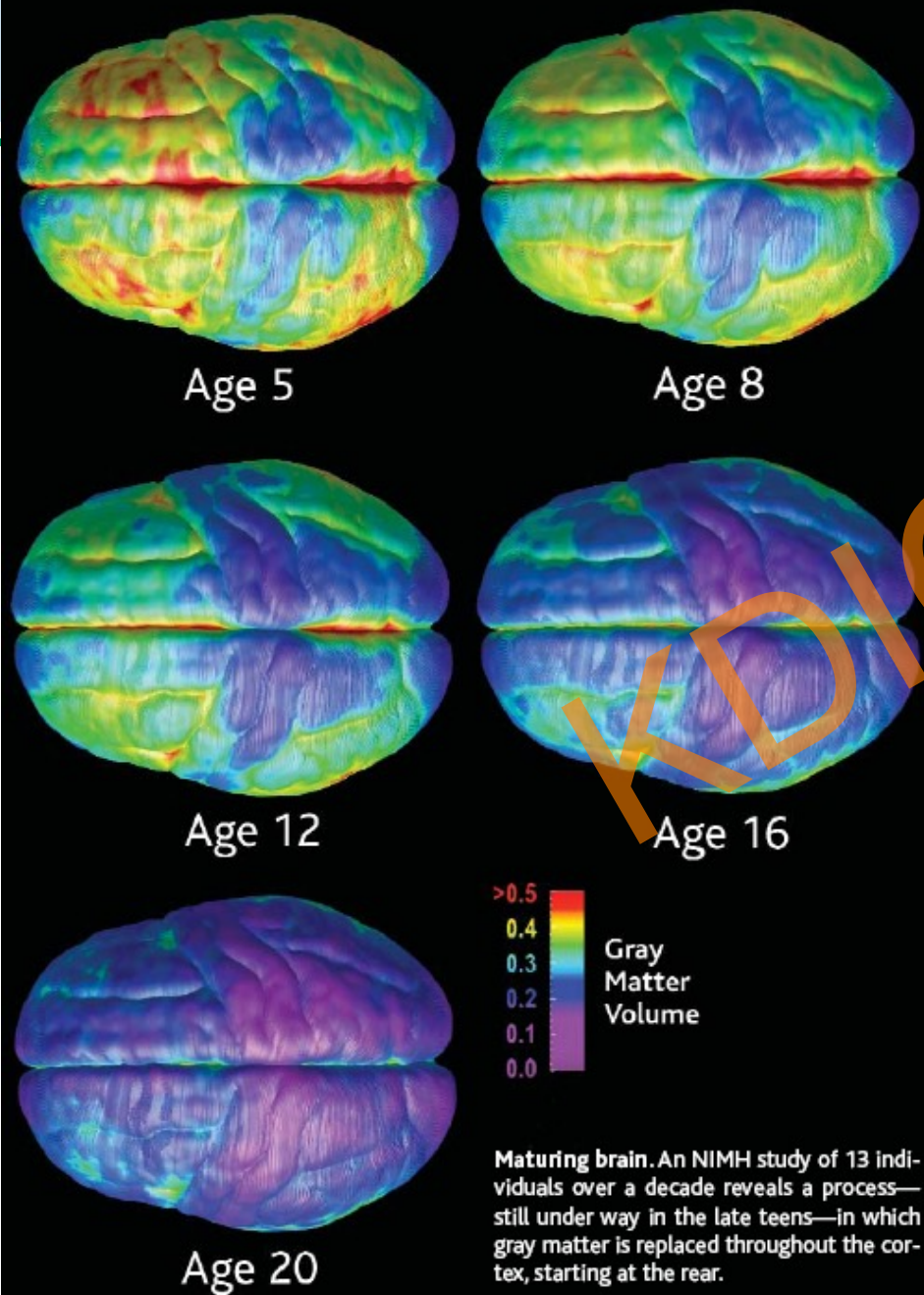


# Developmental Tasks of Adolescence

- 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**



## Normal Brain Development



**Maturing brain.** An NIMH study of 13 individuals over a decade reveals a process—still under way in the late teens—in which gray matter is replaced throughout the cortex, starting at the rear.

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. Watson<sup>1</sup>, Paul N. Harden<sup>2</sup>, Maria E. Ferris<sup>3</sup>, Peter G. Kerr<sup>4</sup>, John D. Mahan<sup>3</sup> and Maher Fouad Ramzy<sup>5</sup>, Consensus Panel Members

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

**Watson AR., Harden PN., Ferris ME., et al. Transition from pediatric to adult renal services:  
a consensus statement by the International Society of Nephrology (ISN) and the International Pediatric  
Nephrology Association. *Kidney International* 2011;80:704-707. doi: 10.1038/ki.2011.209**



# 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

Foster BJ., Platt RW., Dahhou M., et al. The impact of age at transfer from pediatric to adult-oriented care on renal allograft survival. *Pediatric Transplantation* 2011;15:750-759. doi: 10.1111/j.1399-3046.2011.01567.x



# 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

Tong A., Lowe A., Sainsbury, P., et al. Experiences of parents who have children with chronic kidney disease: A systematic review of Qualitative studies. *Pediatrics* 2008;121(2):349-360. <http://www.doi.org/> 10.1542/peds.2006-3470



# 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)

Is non-compliance among adolescent renal transplant recipients inevitable? S Feinstein et al *Pediatrics* 115:969 (2005)



# 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

Chaturvedi S., Jones CL., Walker RG., et al. The transition of kidney transplant recipients: a work in progress. *Pediatric Nephrology* 2009;24:1055-1060. doi: 10.1007/s00467-009-1124-y





# 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

A systematic review of transition readiness and transfer satisfaction measures for adolescents with chronic illness.  
J Stinson et al *International Journal of Adolescent Medicine & Health*. 26(2):159-74, 2014.



# 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  
Bloom SR. et al Journal of Adolescent Health. 51(3):213-9, 2012

