Transitional Care of Adolescents with Kidney Disease

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“Thank you for seeing Rob, who we would like to transfer to your care as he is now an adult.”
- 18 year old male
- Nephropathic cystinosis
- LRD transplant 5 years ago. Creat 83 µmol/L
  - cyclosporine, prednisone & azathioprine
- Raised TSH, just commenced thyroxine

“...he detests phosphocysteamine and refuses to take it.”

<table>
<thead>
<tr>
<th>Cystinosis</th>
<th>Extra-renal Manifestations</th>
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</thead>
<tbody>
<tr>
<td>Pathogenesis</td>
<td>Extra-renal Manifestations</td>
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</table>
| Defect in cystine transport through lysosomal membrane | Corneal and conjunctival deposits
  - photophobia, watering and blepharospasm |
| Accumulation of cystine crystals in lysosomes | Retinal deposits |
| Infantile Cystinosis (nephropathic form) | Insulin dependent diabetes |
| Renal Manifestations | Hypothyroidism |
| Onset 3 – 6 months | Male infertility |
| Impaired proximal tubular reabsorption-Fanconi’s Syndrome | Cardiac arrhythmias |
| End stage kidney failure before age 10 | CNS deposits |
| | Hepatosplenomegaly |

“Thank you for seeing Rob, who we would like to transfer to your care as he is now an adult.”

“Rob is a somewhat difficult boy in that he has taken very little interest in his own care until now, but he is slowly maturing and works full-time.”
Waiting Time and Outcome of Kidney Transplantation in Adolescents.

Kennedy, Sean; Mackie, Fiona; Rosenberg, Andrew; McDonald, Stephen


Q. What is an Adolescent?

A. 10 to 19 years

Tasks

- Consolidate identity
- Establish relationships outside the family
- Achieve independence from parents
- Set & achieve educational & vocational goals
- Participate in community life
- Be happy – have intact mental health

The Average Public Hospital Patient

The Patient with Childhood Onset CKD

Aetiology of ESKD

Patients who commenced RRT before 18 years of age

<table>
<thead>
<tr>
<th>Primary Disease</th>
<th>Number</th>
<th>Age at Referral</th>
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<tbody>
<tr>
<td>Hypoplasia/dysplasia</td>
<td>15 (20%)</td>
<td>0.1 (birth -15.0)</td>
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<tr>
<td>Posterior urethral valves</td>
<td>16 (14%)</td>
<td>0.2 (birth-2.7)</td>
</tr>
<tr>
<td>Congenital nephrotic</td>
<td>4 (5%)</td>
<td>0.2 (4 days – 0.4)</td>
</tr>
<tr>
<td>Reflux nephropathy</td>
<td>7 (9%)</td>
<td>1.9 (0.1-10.8)</td>
</tr>
<tr>
<td>FSGS</td>
<td>3 (4%)</td>
<td>2.0 (0.7-5.5)</td>
</tr>
<tr>
<td>Nephrosochistis</td>
<td>7 (9%)</td>
<td>4.3 (1.5 – 15.9)</td>
</tr>
<tr>
<td>Glomerulonephritis</td>
<td>10 (14%)</td>
<td>7.8 (1.2-13.4)</td>
</tr>
<tr>
<td>BMT nephropathy</td>
<td>3 (4%)</td>
<td>10.4 (4.0-13.2)</td>
</tr>
<tr>
<td>Other</td>
<td>9 (12%)</td>
<td>1.1 (5 days – 15.9)</td>
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</tbody>
</table>
Paediatric Nephrologist’s View

- First met Rob when he was 10 months old
- Frequent admissions during first 2 years for fluid management and feeding
- At least monthly visits throughout childhood with repeated admissions during intercurrent illnesses
- Commenced PD aged 11, extended admissions
- LRD transplant from father aged 13
- Ongoing challenge of poor adherence

Rob’s View

Paediatric vs Adult Health Care

**Paediatric Services**
- Maximising growth & development
- Strongly family centred
- Parental relationships with patients
- Interdisciplinary care

**Adult Services**
- Battling decline & senescence
- Expect patients to function autonomously
- Empower patients with information & expectations
- Multidisciplinary care

Practical Differences

- Spectrum of disease
- Volume of patients
- Resources & workload
- Prescribing practices
- Role of GPs
- Attitude to pain

Adapted from Rosen D. 1995

Problems with Transfer of Adolescents

- Fail to adjust to their new role
- Slip through the cracks
- Specific needs may not be met

Adolescent patients & their families may evaluate the care provided in adult clinics according to the model of care provided in paediatric clinics → Dissatisfaction &/or distrust

Health Care Transition

“The purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centered to adult-oriented health care system.”

Transition is a process not an event

Transition

- A process that begins with paediatric care and continues into the adult clinic
- Start early
- Foster independence
- Teach and promote self-management
- Thorough handover of care
- Listen & support

Handy Hints

- Adolescents value privacy and expect confidentiality
- Comfortable with familiar people and places
- Parents frequently maintain a vital interest and may remain intimately involved but...
  - Adolescent responsibility is undermined when parents are asked to supervise care at home.
- Physical, emotional, social, and psychosexual development are often delayed in this patient population

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Principles of Successful Transition

1. Health care services …need to be developmentally appropriate and inclusive of the young person’s family where appropriate.
2. …need to holistically address a range of concerns such as growth and development, mental health, sexuality, nutrition, exercise and health risking behaviours such as drug and alcohol use.
3. …require flexibility …actual process of transition needs to be tailored to each individual adolescent or young person.

Goal of Transition

- “Maximize lifelong functioning and potential through the provision of high quality, developmentally appropriate health care services that continue uninterrupted as the individual moves from adolescence to adulthood.”

Consensus statement on health care transition for young adults with special health care needs. Pediatrics 2002
What is non-adherence?

- Adherence is the extent to which patients take medications as prescribed by their healthcare providers.
- Non-adherence is a continuum.
- Non-adherence is difficult to detect and quantify.
- What degree of non-adherence is clinically important?


Does non-adherence matter?

- Risk of graft failure increased seven-fold in nonadherent transplant recipients.
  (Butler JA et al Transplantation 2004)
- Non-adherence with various aspects of haemodialysis regimens increases mortality risks.
  (Denhaerynck et al American Journal of Critical Care 2007)

Non-adherence

- Transplantation
  - Missed drug doses
  - Late drug doses
  - Incorrect drug doses
- Dialysis
  - Exceeding fluid restrictions
  - Ignoring dietary restrictions
  - Missed or shortened sessions
  - Missed medications
  - Smoking

Reasons for non-adherence

- Socio-demographic
  - Disrupted families, lack of support
  - Distance to clinic
- Psychological
  - Depression
- Treatment specific
  - Cosmetic side-effects
  - Unpleasant taste
- Systemic reasons.
  - Cost of medications
  - Beliefs about illness and medication
Why are adolescents non-adherent?
- Consolidate identity
  * I am more than this illness.
- Establish relationships outside the family
  * I don't want to be different from my friends.
- Achieve independence from parents
  * I make my own rules.
- Set & achieve educational & vocational goals
  * I don't have time for this.

Risk taking

What are the Barriers to Adherence?
- Reasons for missing medications
  - 56% I just forgot
    (Mornings are the most difficult time.)
  - ~ 40% I wasn't home or interferes with activity
  - ~ 17% I hate the taste
- "What makes it difficult to take your medication on time?"

Who is at Risk?
- Transplants performed 1985 - 2009
- Functioning transplant for at least 3 months
- Spent some time aged between 10 and 30 years.
- 3,166 transplants in 2,936 recipients
- Age at transplantation: median 23 y (IQR 18-27)
- Transplanting centre
  - Adult 2,613 (83%)
  - Paediatric 553 (17%)

Causes of Kidney Graft Failure

<table>
<thead>
<tr>
<th>Cause</th>
<th>N</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>Chronic allograft nephropathy</td>
<td>443</td>
<td>61%</td>
</tr>
<tr>
<td>Acute rejection or non-compliance</td>
<td>105</td>
<td>15%</td>
</tr>
<tr>
<td>Glomerulonephritis recurrence</td>
<td>61</td>
<td>8%</td>
</tr>
<tr>
<td>Death with functioning graft</td>
<td>67</td>
<td>9%</td>
</tr>
<tr>
<td>Other</td>
<td>45</td>
<td>6%</td>
</tr>
<tr>
<td>Total</td>
<td>721</td>
<td></td>
</tr>
</tbody>
</table>

Hazard ratio

Graft loss from acute rejection/non-compliance
How should it be managed?

Managing Non-adherence

- Self-management
  - Effective in asthma, diabetes & hypertension
  - More effective than knowledge based education alone in asthma
- “An ultimate goal of transition to adult health care services is to facilitate the development of successful self-management in young people with chronic conditions.”

What works?

Supporting Self Management

- Assess
  - Assessment of beliefs, behaviour & knowledge
- Advise
  - Provide information
- Agree
  - Set collaborative goals
- Assist
  - Provide support for self management
- Arrange
  - Develop a specific plan for follow-up

“What makes it difficult to take your medication on time?”

Goals of Adolescent Health Care

- Manage acute and chronic illness
- Reduce risks
- Promote normal social & emotional development
- Promote positive self-concept and sense of competence
- Promote independent living
- Support long term planning and life goals

Psychosocial Screening

- H Home
- E Education & employment
- A Activities & peers
- D Drugs
- S Sexual health
- S Suicide & depression

M S M Yeo, L M Bond and S M Sawyer MJA 2005
originally by Goldenring & Cohen 1988
Survival of pre-emptive grafts in adolescents is similar to other age groups. Kennedy et al., Transplantation 2006.

Kaplan-Meier graft survival: Pre-emptive Grafts
Australia & New Zealand, 1980 to 2003, n=240

Acknowledgements
- Angus Ritchie
- Phil Clayton
- Stephen McDonald