

## **Growing up in CF world – Transitioning with Success, you can have a say**

Jude Morton – Senior Respiratory Consultant, Royal Adelaide

- 50% of CF population are adults so transition important
- Transition is young people taking responsibility for their own care, a gradual process of empowerment (not just a transfer from paediatric setting to adult clinic)
- In transition process, look at whole person and build relationship – they discuss goals and aims in life (short term and long term), how CF affects life now and how it may in future, time management strategies, career/hobbies, family/friends, drugs/smoking/alcohol (drug use is a contraindication to successful transplant), sexual health/genetics/fertility, survival/transplantation, finances
- Suggest process should start at 11-12 years of age (Note: this is beyond taking responsibility for Creon dosing and should encompass the whole treatment).

## **Sexual Health for Adolescents with CF**

Katherine Frayman – Respiratory Medicine, Royal Children's Melbourne

- Patients should be offered assessment of fertility status (semen analysis)
  - Often confusion between infertility and impotence
  - Still need to use condoms to prevent STI's
  - Should be topic of ongoing discussion, a combination of parents and CF team (often too embarrassed to discuss properly)
  - Victoria is launching a new web site called "spill" late August which is a digital resource for adolescents about sexual and reproductive health (note: sneak preview looked unreal). Will be rolled out throughout Australia after launch
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## **Transplants: the Work Up! What is required?**

Allan Glanville - Professor and Medical Director, Lung Transplantation, St Vincent's, Sydney

- Third Consensus Document (published in the Journal of Heart Lung Transplant Jan 2015) forms the basis for ANZ lung transplantation criteria - essentially, FEV1 less than 30% and falling, all viable treatment options exhausted, acknowledgement that have limited life expectancy
- Referral and placement on waiting list are two distinct processes; referral not an automatic endorsement to be placed on list
- Key message: Referral should occur early to enable enough time to assess and work up patient
- Key message: non adherence to medical therapies is a contraindication (so important to demonstrate adherence to CF management regime)
- Having Burkholderia cepacia is a major issue to survival after transplant
- After referral, patient assessed, and placed on list. Then commences the "work up" of tests and education over about a 6 month period. Once completed, then go on active list.
- Once transplanted, important to continue exercise to clear mucous, but don't need to continue with CF management regime (while the CF patients' cells populate the new lungs, it is not at the epithelial level so new lungs not affected by CF)
- So far the professor's longest surviving CF transplant patient is 25 years post transplant
- Lots of immunosuppression drugs to take after transplant but QOL is much better notwithstanding

## Energy expenditure, hydration and nutrition in the active person with CF

Andrea Kench, Dietician and Anna Middleton, Physiotherapist, Westmead

- Dehydration is defined as a greater than 2 % body weight loss
- Exercise is one of the most common causes of low blood sugar (blood glucose) i.e. exercise-related hypoglycemia.
- Colour of urine is a layman indication of dehydration in the absence of clinical measures
- CF kids observed to drink less during exercise (not sure if there is a different physiological response in CF). Can't rely on thirst as an indicator.
- Therefore electrolyte replacement is important. No evidence based practice guidelines.
- Encourage sports drinks which have greater than 50mmol/L (but note they also contain carbohydrate). Hydralyte Sport is a new product and recommended.
- CF kids have a higher resting energy expenditure. Therefore 110-200% of Recommended Daily Intake required.
- In CF, Energy Expenditure is heightened post exercise. Need carbohydrate and fat as fuel source for exercise, which metabolises into ATP (talked about substrate utilization which was unintelligible to me). But importantly need extra calorific intake after exercise to replace stores and high protein for muscle repair.