Cystic Fibrosis & Transition from Paediatric to Adult care

Jan Tate
CF CNS
Starship Children's Health

2009

Cystic Fibrosis

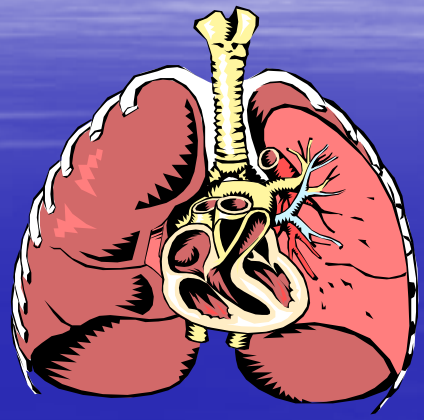
- most common life-threatening genetic disease – predominately European
- affects 1:2,500 births in NZ
- autosomal recessive disorder - both parents asymptomatic carriers
- 1:25 people in NZ are carriers
- approx 380 people in NZ with CF
- caused by a defect on Chromosome 7 - discovered 1989
- CF gene mutations in NZ
  - Delta F508 - 75% of CF population
  - Over 1000 different mutations described

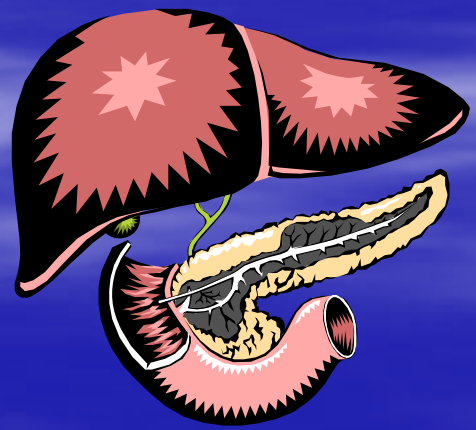
Abnormality of the CF Gene

- Cystic Fibrosis Transmembrane Regulator (CFTR)
  - functions as chloride channel
  - normally located in epithelial cells
  - causes abnormalities in transport of chloride ions across cell membranes
  - ↑ in levels of sodium & chloride
- characterised by abnormal thick secretions that block ducts in the exocrine glands
- main cause of morbidity & mortality is bacterial lung infection

Gene Causes Abnormalities

- Upper airway
  - Sinusitis, nasal polyps
  - Recurrent pneumonias
- Failure to thrive
- Diabetes
- Liver Disease
  - Gall stones
  - Portal hypertension
- Distal obstructive syndrome, constipation
- Vitamin deficiencies
- Bone disease
- Fertility concerns





- Normal neurology, development

Respiratory System

Lungs “normal” at birth

Bacterial infections → **- *Staphylococcus aureus***
- *Pseudomonas aeruginosa*
- *Burkholderia cepacia*

↓

Inflammation

↓

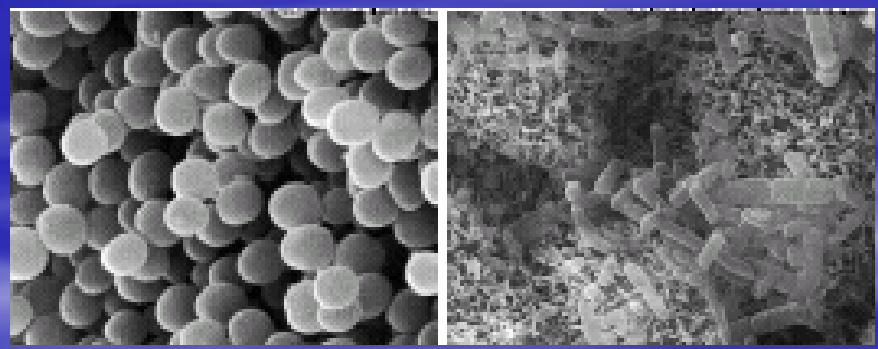
Mucocillary plugging

↓

Bronchiectasis

↓

Respiratory failure → **Premature Death**



CF Management

Aimed at delaying progression of the disease – main goal is preventing & controlling lung and other complications

Treatments are rigorous, complex & time consuming

- chest physiotherapy BD
- sport & exercise daily
- antibiotics: oral, nebulised, IV
- pancreatic enzymes with all food
- nutritional supplements & vitamins

Frequent outpatient visits

Frequent hospitalisations

Segregation issues

Regular Reviews

Respiratory

- clinical signs
- auscultation
- respiratory testing: lung function, oximetry
- regular check for respiratory infection
- Radiology: CXR, CT,

Nutrition

- weight & height percentiles
- review of caloric intake
- review of stool

Other

- annual blood testing
- annual OGTT
- abdo U/S every 3 years
- DEXA scans biannually

Preventing Cross Infection in Outpatients

Outlook of CF

- great variation in disease
- generally improved from 50 years ago
- median life expectancy now 36.5 yrs of age

(CF Foundation, 2008)

- changed from chronic childhood illness to adolescent & adult management with 90% of children with CF becoming adults and transitioning to adult services

Adolescents with CF

- not invincible
- delayed puberty
- friendships with peers
- dependence vs independence
- compliance / adherence issues
- career choices

Quality of life

Depends on

- **early treatments**
- **more aggressive treatments**
- **contact with respiratory organisms**
- **frequent follow-up & review**
- **parents & adolescent knowledge of CF**
- **adherence / compliance**
- **family attitudes**
- **daily routines**
- **environment**

Transition

"Transition from a paediatric to an adult setting is the purposeful and planned movement of adolescents with chronic physical and medical conditions from child to adult centered healthcare systems"

Blum, et. al 2002

Transition vs Transfer

- **transition refers to skills, processes and attitudes to facilitate movement from paediatric to adult care**
- **transfer refers to the physical movement from paediatric to adult services**

Principles for Successful Transition

- start early
- knowledge of adolescent
- communication between teams & adolescent & family
- identified person
- flexibility
- written information

Problems for Transition

Family & Adolescent

- **bonded to the paediatric team**
- **fear of the unknown**
- **resistance to change**
- **distrustful of skills of the new team – they have to "prove themselves"**
- **adolescents not quite ready to grow up**
- **parents concerned of lack of control**

Problems for Transition

Paediatric Team

- bonded to patients & their families
- family centered care vs individual focus
- patients are not perceived as adults
- concern re skills & knowledge of adult team

Adult Team

- not used to dealing with adolescent's family centered care
- emotional burden of the end-of-life patients
- budget – CF patients are high users

Preparation for transition is essential

- start about 13 yrs of age
- clear expectations of transfer
- transition checklist
- expectations from the adolescent
- meeting the adult team
- opportunities for adolescent to ask questions and give feedback

Timing for Transition

- requires a flexible policy
  - on leaving school?
  - on turning 16? or 18 ?
  - when the young person is ready ?
- factors influencing age of transition
  - emotional & intellectual maturity
  - disease severity & stability
  - lifestyle situations

Challenges & Success to Transition

- **Challenges**
  - sudden transfer without preparation
  - transfer during time of crisis or instability
  - transition when palliative
  - transition to a disinterested adult team

- **Success**
  - planned & orderly transfer
  - preparation of adolescent & family early in adolescence
  - presented as a positive milestone
  - target age with flexibility
  - strong communication between teams, adolescent & family
  - health histories from all paediatric disciplines

Transition at Starship - RBP

- between 16 – 18 yrs of age
- nurse specialists – key people
- planned preparation & a co-ordinated approach
- transition meetings
- joint transition clinic at Starship
- tour of adult facilities
- final summary letters
- first appointment with adult team to be within 3 months
- family meetings at adult services after transitioning to discuss transfer
- feedback from adult team to paediatric team
- not transferred if palliative

Key Points for Transition & Transfer of adolescents with Cystic Fibrosis

- empowerment - patient & family
- interdisciplinary communication
- key people identified in process
- flexibility for each individual
- numerous transitions along the way
- goal = seamless transfer to adult care before the age of 18 years

Literature List

- RACP Joint Adolescent Health Committee – Transition guidelines, 2007
- Doring, G., Hoiby, N. (2005) Standards of Care for patients with Cystic Fibrosis: A European consensus. *Journal of Cystic Fibrosis*, Vol4 (1), pp7-26
- Guidelines for Transition Pathway, Royal Children's Hospital, Melbourne, Australia
- ATS 2000 Protocols for Transitional Care from Paediatric to Adult
- Clinical Guidelines for the care of Children with Cystic Fibrosis, Paediatric CF Team, Starship Children's health, 2007
- NZ Standards of Care, in print, 2008
- Cowlard, J. (2003) Cystic Fibrosis: transition from paediatric to adult care. *Nursing Standard*, Vol18, pp39-41.
- Blum, R.W et al. (2002) A consensus Statement on health care transitions for young people with special health needs. *Pediatrics*, 110 (6): 1304-1306
- Rosen, D.S., Sawyer, S.M. et al (2003) Transition to Adult Health Care for adolescents and young adults with chronic conditions. *Journal of Adolescent Health*, 33:309-311
- Romola, S., Hawkins, K., et al (2009) Adherence to treatment in adolescents with CF: the role of illness perceptions & treatment beliefs. *Journal of Pediatric Psychology*, Feb, 09