BARRIERS TO WORK IN CYSTIC FIBROSIS

Dr Katherine Targett
CYSTIC FIBROSIS

- Commonest fatal inherited disease in Caucasian populations of European origin

- Autosomal recessive inheritance

- In the UK: Gene frequency is 1 in 25
  Incidence 1 in 2,500 births

First described by Anderson in 1938. But……

Rochholz 1857, Almanac of children’s songs from Switzerland

‘The child will soon die whose brow tastes salty when kissed’
Molecular genetics

Gene localised to long arm of chromosome 7 in 1985

Gene locus isolated in 1989

Gene product:

Cystic Fibrosis Transmembrane Conductance Regulator (CFTR)
CFTR

CFTR Apical (luminal) membranes of

Airways
Pancreas
Liver
Intestine
Sweat glands
CF IN ADULTS

**CLINICAL**

- 80% recurrent pulmonary infections and pancreatic insufficiency
- 15% recurrent pulmonary infections
- 5% gastrointestinal problems only

Diabetes, liver disease, osteoporosis, fertility

**PSYCHOSOCIAL**

- Schooling and educational attainment
- Parental attitudes and expectations
- Emotional and psychological development
- Mental health issues

FOM 2013
EPIDEMIOLOGY

**Survival**

<table>
<thead>
<tr>
<th>Year</th>
<th>Age</th>
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</thead>
<tbody>
<tr>
<td>1930’s</td>
<td>1-2 years</td>
</tr>
<tr>
<td>1969 (USA)</td>
<td>14 years</td>
</tr>
<tr>
<td>1978 (USA)</td>
<td>21 years</td>
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<tr>
<td>1990 (USA)</td>
<td>28 years</td>
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<tr>
<td>1994 (UK)</td>
<td>31 years</td>
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Estimated survival of CF patient born in 1990s >40 years

Adults > children

Improved survival: antibiotics, nutrition
EMPLOYMENT
AND CYSTIC FIBROSIS

Aim
To evaluate which physical and psychosocial factors are associated with employment status of adults with CF
EMPLOYMENT AND CYSTIC FIBROSIS

- Multicentre study (Aberdeen, Birmingham and Newcastle)
- Adults with CF age 16 years and over
- Questionnaires:
  - Health-related quality of life, CFQ-UK. 12 QoL domains: physical, role, vitality, emotional, social, body image, eating, treatment burden, health perceptions, weight, respiratory and digestive symptoms
  - Working with CF
  - Presenteeism – Stanford Presenteeism Scale (SPS)
- Clinical markers of disease severity:
  - FEV$_1$%; BMI
  - Colonising organism in sputum;
  - CF-related diabetes;
  - Courses of IV antibiotics in preceding year
  - Transplant status

FOM 2013
Median age 26yrs old  
(range 16-70)  
Female 46%  
Education “school only” 60%  

Mean FEV$_1$ % predicted 60%  
(range 12-136%)  
Mean BMI 22  
Colonising organism Ps. Aeruginosa in 74%  
Median number of courses of IV antibiotics in preceding year = 2  
CF-related diabetes 34%  
Transplant (lung or liver) 5%  

254 subjects
EMPLOYMENT STATUS

- 30% unemployed of whom 24% seeking work
- Median hours worked = 37hrs/wk
- Mean SPS score = 25

“**My CF does not affect my ability to do the work I am asked to do, but a few employers that I have told about my CF have then said that they have found someone more suitable and are not willing to give me a chance or any support. I have not had any paid work since I left school**”
## WORK DISABILITY

<table>
<thead>
<tr>
<th>Work Disability Impact</th>
<th>Advice and Support</th>
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<tbody>
<tr>
<td>40% stopped a job because of their CF</td>
<td>42% had received advice regarding work – most from their respiratory consultant and only 6% from occupational health</td>
</tr>
<tr>
<td>24% changed duties because of CF</td>
<td>92% were allowed time off for appointments;</td>
</tr>
<tr>
<td>10% taken a cut in salary because of CF</td>
<td>30% had their hours adjusted</td>
</tr>
<tr>
<td>47% CF influenced choice of career</td>
<td>54% were allowed to work flexibly</td>
</tr>
<tr>
<td>23% discrimination because of CF</td>
<td>17% were allowed to work from home</td>
</tr>
</tbody>
</table>

FOM 2013
DIFFERENCES BETWEEN CENTRES

- Birmingham recorded consistently lower CFQ-UK scores
- Most likely to be in work in Birmingham; least likely in Newcastle
- Statistically significant between centre difference in presenteeism scores: highest in Aberdeen, lowest in Birmingham.
- For those not currently in paid employment Newcastle had the lowest percent “ever worked and currently seeking employment”, Aberdeen the most
**PREDICTORS OF EMPLOYMENT STATUS (univariate analysis)**

<table>
<thead>
<tr>
<th>Centre</th>
<th>Gender (p=0.011)</th>
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<tbody>
<tr>
<td></td>
<td>FEV$_1$% (p&lt;0.001)</td>
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<tr>
<td></td>
<td>BMI (p=0.022)</td>
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<tr>
<td>Number of courses of IV antibiotics (p&lt;0.001)</td>
<td></td>
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<tr>
<td>Ps. Aeruginosa in sputum (p=0.017)</td>
<td></td>
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<tr>
<td>Educational status (p=0.001)</td>
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<tr>
<td>All 12 QoL domains</td>
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“I have always felt scared to tell employers about my CF as it has held me back from promotions in the past. Employers have felt I might be unreliable as I may have to have time off for treatments and illness….I think employers can be very ignorant of the condition……I feel I put more effort in my job to have to prove myself which does make me very tired at the end of the day”

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QUALITY OF LIFE

CFQ-UK QoL domains

ratio employed: unemployed

FOM 2013
PREDICTORS OF EMPLOYMENT STATUS

(multivariate analysis)

Best fit model (ROC 0.856)

- Education
- Role perception
- Health perception
- Centre

- Does not include any recognised markers of disease severity
THE FUTURE

- Strategies to improve coping strategies and resilience
- Psychological interventions
- Targeting the paediatric population
- Role for occupational health advice in out-patient clinics

“I find working to be therapeutic. I also find that however tiring it is to work, the structure of the work day helps me better plan and carry out my treatments at regular intervals. I would hate to think what I would have missed out on if I had never worked, my life would have been all about CF……I find work normalises me and I don’t think about CF all day”

FOM 2013
WITH THANKS TO

Professor Graham Devereux, Aberdeen Royal Infirmary

Dr Ed Nash, Birmingham Heartlands Hospital

Dr Steve Bourke, Royal Victoria Infirmary, Newcastle upon Tyne

Society of Occupational Medicine, Scottish Group, Sandy Elder Award 2010

All the patients from Aberdeen, Birmingham and Newcastle who took part in this study