Palliative and Supportive Care in Cystic Fibrosis

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Overview

1. Cystic Fibrosis – Clinical Aspects
2. Oxford Experience of CF End of Life Care
3. End of Life Care in CF – Six Key Challenges
4. A Way Forward?
“Woe to the child who tastes salty from a kiss on the brow, for he is cursed and soon must die”
“Cystic Fibrosis of the Pancreas”
Andersen DH. Am J Dis Child 1938;56:344

Dorothy Andersen
The CF Gene: First Identified in 1989
Improving Survival in Cystic Fibrosis

Median Predicted Survival Age, 1987–2011 in 5-Year Periods

US CF Foundation Registry Report 2012
Improving Survival in Cystic Fibrosis

Life expectancy of babies born with CF in the 21st century now exceeds 50 years

Survival in CF is Improving Even Among Patients with FEV$_1$ <30% Predicted

George et al BMJ 2011
CF for the Geriatrician?

- Age-Related Macular Degeneration
- Cataracts
- Hypertension
- Glaucoma
- Duodenal Ulcer
- ACE-I Related Angioedema
- Chronic Kidney Disease
- Benign Prostatic Hypertrophy
- Colonic Polyps
- Hydrocele
- Ciprofloxacin-Related Achilles Tendonitis
CF Multi-Disciplinary Team

Exercise Assistant
Social Worker
Administrator

Doctors
Dietitians

Specialist Nurses
Pharmacists

Physiotherapists
Clinical Psychologist
Cystic Fibrosis in 2017: Cause for Optimism?
Clinical Effect of Ivacaftor in Patients with CF and the G551D Mutation

McKone et al. Lancet Resp Med 2014
CF: A Life Unlimited?

Nathan Charles
Western Force & Wallabies Hooker
CF: A Life Unlimited?

Nick Talbot
Mountaineer & CF Patient
CF: The Reality for Many

Median age at death = 28 yrs
CF Lung Disease
CF Lung Disease
Microbiology of CF Lung Disease

**Pseudomonas aeruginosa**

**Haemophilus influenzae**

**Chronic S. aureus; n=1511 (17.2%)**

**Chronic P. aeruginosa; n=2960 (33.5%)**

**B. cepacia; n=326 (3.6%)**

**MRSA; n=310 (3.4%)**

**H. influenzae; n=1121 (12.4%)**

UK CF Registry Data 2013
CF Pulmonary Exacerbations

• Acute increase in respiratory symptoms:
  – Cough
  – ↑sputum
  – Chest pain
  – Fatigue
  – Haemoptysis
  – Malaise
  – Fever
  – ↑ upper airway symptoms
CF Pulmonary Exacerbations & Mortality

De Boer et al. Thorax 2011
Pulmonary Exacerbation Management

- Hydration
- Bronchodilators
- Controlled Oxygen
- NIV
- Glycaemic Control
- Nutrition
- Chest Physio
- Broad-Spectrum IV Antibiotics
Intravenous Antibiotics for *Pseudomonas aeruginosa* in CF

- **β-Lactam**
  - Ceftazidime 2-3g TDS
  - Meropenem 2g TDS
  - Tazocin 4.5g TDS/QDS
  - Fosfomycin 4g QDS

- **Aminoglycoside or Polymixin**
  - Tobramycin 7mg/kg OD
  - Colistin 1-2 megaunits TDS
Oxford Experience:
End of Life Care in CF
2011-2016
Oxford Experience 2011-16

- 16 deaths
  - 3 post-lung transplant
  - 1 death on Tx list
- Median age at death 30.5 yrs (range 21-73)
- 8/16 (50%) received specialist palliative care input

![Cause of Death Pie Chart]
Oxford Experience 2011-16

- 16 deaths
  - 3 post-lung transplant
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Place of Death
Palliative Care in CF: Challenges & Opportunities
1. Unpredictable Disease Progression

When to discuss End of Life & Transplantation?
1. Unpredictable Disease Progression

![Lung Function Graph]

- FVC
- FEV₁
2. Increasing Treatment Burden
2. Increasing Treatment Burden

### Daily Routine

<table>
<thead>
<tr>
<th>Time</th>
<th>Activity</th>
</tr>
</thead>
<tbody>
<tr>
<td>0700</td>
<td>Take DNase neb - wait 45mins whilst having breakfast</td>
</tr>
<tr>
<td>0730</td>
<td>Take tablets &amp; insulin</td>
</tr>
<tr>
<td>0745</td>
<td>Airway Clearance: Salbutamol MDI, HFCWO (8mins), AD + NIV +/- PEP ~ 1hr</td>
</tr>
<tr>
<td>0845</td>
<td>Take Seretide, Tiotropium inhalers</td>
</tr>
<tr>
<td>0900</td>
<td>Take Promixin or Bramitob via Ineb – then clean equipment ~ 30mins</td>
</tr>
<tr>
<td>1000</td>
<td>Work 10am-4:30pm</td>
</tr>
<tr>
<td>1730</td>
<td>Go to sort horses ~ 1hr, have dinner</td>
</tr>
<tr>
<td>1930</td>
<td>Take DNase neb</td>
</tr>
<tr>
<td>2030</td>
<td>Airway Clearance: Salbutamol MDI, HFCWO (8mins), AD + NIV +/- PEP ~ 1hr</td>
</tr>
<tr>
<td>2130</td>
<td>Take Seretide inhaler, Promixin or Bramitob via Ineb – clean equipment</td>
</tr>
<tr>
<td>2200</td>
<td>Bed</td>
</tr>
</tbody>
</table>
2. Increasing Treatment Burden: Non-Invasive Ventilation

- Nocturnal NIV traditionally used as a bridge to lung transplantation
- May help stabilise disease progression

Flight et al. JCF 2012
Acute Complications of CF

- Pulmonary Exacerbations
- Acute Abdomen
- Respiratory Failure
- Massive Haemoptysis
- Vascular Access Issues
- Anaphylaxis
- Pneumothorax
Pneumothorax in CF

• Lifetime incidence = 3.4%
• Risk factors\(^1\)
  – severe lung disease (FEV\(_1\) <30%)
  – increasing age
  – chronic *Pseudomonas aeruginosa* infection
• Management → chest drain

1: Flume et al. AJRCCM 2008
Massive Haemoptysis in CF

• Definition:
  >240ml blood in 24 hrs
  >100ml per 24 hrs for several days

• Lifetime incidence of 4.1% in CF\(^1\)

• Risk factors:
  – Increasing age
  – Increasing severity of lung disease
  – CF-related diabetes
  – *Staph. aureus* in sputum

Massive Haemoptysis in CF: Bronchial Artery Hypertrophy
3. Impact of Lung Transplantation
3. Impact of Lung Transplantation

![Graph showing impact of lung transplantation with key events marked: Half marathon, Prolonged Hospital Admission, and Active transplant list.](image-url)
3. Impact of Lung Transplantation
Extracorporeal Membrane Oxygenation (ECMO)

Hayes et al. JCF 2012
Awake ECMO with Physical Rehabilitation

Hayes et al JCF 2012
4. Geography
5. Infection Control & Isolation
6. Emotional Toll
6. Emotional Toll

“...being a dad would happen. Growing up as a young teenager and reading into CF gave me the impression that this could never happen. Fast forward to 2013 and our fertility journey to Oscar being born, began and what a journey it has been.”

Source: CF Trust 2016
End of Life Care in CF: A Way Forward

• Training opportunities for CF MDT staff

• “Transplant arena” meetings

• Move towards integrated palliative care in CF
  – symptom control alongside active therapy
End of Life Care in CF: A Way Forward
Summary

• Prognosis in CF continues to improve

BUT

• Many people with CF still die as young adults
• Complexity of the disease and treatments is increasing
• We have real opportunities to develop excellent end-of-life care for all with CF
Dr William Flight

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