Plan

- Cystic Fibrosis (CF)
- Challenges of adolescence and young adulthood
- Interaction with chronic disease
- Providing developmentally appropriate, psychologically minded services
Cystic Fibrosis

• CF is one of the UK's most common life-threatening inherited diseases
  – 9,000 people in the UK.

• Lungs
  – Frequent lung infections cause lung damage, gradually impairing the ability of the lungs to function properly.

• Digestive system
  – CF affects the pancreas' ability to release enzymes necessary to digest food, this can cause malnutrition, which can lead to poor growth, physical weakness and delayed puberty.
Treatment Regimen

- Heavy treatment burden \(^{(\text{Sawicki et al, 2009})}\)
- Preventative
  - Acute exacerbations: IV antibiotics
    - 2+ weeks at hospital or home
  - Advanced disease
    - Oxygen, non-invasive ventilation
    - Lung transplant
The Adult CF Service

- Patients aged 16-81 (median = 26)
- 200+ patients
- Wide geographical area (SE London, Kent, Sussex & Surrey)
- Inpatient & outpatient care
  - Clinics, home visits, annual reviews

- Multidisciplinary team
  - Consultants
  - CNSs
  - Physios
  - Dietitians
  - Pharmacists
  - Social Workers
  - Psychologists
  - Ward Staff
What was important to you when you were 17 years old?

What about when you were 21 or 25?
• Physical, emotional, social and cognitive development
• Peer influence on risk perception (Knoll et al, 2015)
• ... and on risk taking behaviour (Chein et al, 2011)
• The social brain
<table>
<thead>
<tr>
<th>Development</th>
<th>Chronic Illness</th>
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<tbody>
<tr>
<td>Delayed puberty</td>
<td>Poor adherence</td>
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<tr>
<td>Identity affected by the “sick role”</td>
<td>Poorly developed abstract thinking/planning</td>
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<td>Impact on body image/perceived sexual attractiveness</td>
<td>Perception of self as “bullet proof”</td>
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<tr>
<td>Reduced independence and reliance on parents</td>
<td>Smoking, alcohol/drug use, sexual risk taking</td>
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<tr>
<td>Impact on education and employment</td>
<td>Increased risk taking behaviour</td>
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<tr>
<td>Social isolation</td>
<td>Rejection of medical professionals as part of separation from parents</td>
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Suris, Michaud & Viner (2004)
Life Cycle

Independence

Dependence

Adolescence
“puberty to relative self sufficiency”
Life Cycle

With thanks to Eileen Reynolds, CF Social Worker
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• Multidisciplinary support
• Home visits to promote self management and reduce impact of health care.
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• The CF MDT and patients
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